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Artículos originales (todos) * Original articles (all)**
Neuroendocrine tumors.

Abril - Mayo 2013 / April - May 2013

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[1]

TÍTULO / TITLE: - Association between BRAF V600E mutation and mortality in patients with papillary thyroid cancer.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - JAMA. 2013 Apr 10;309(14):1493-501. doi: 10.1001/jama.2013.3190.

●●Enlace al texto completo (gratis o de pago) [1001/jama.2013.3190](#)

AUTORES / AUTHORS: - Xing M; Alzahrani AS; Carson KA; Viola D; Elisei R; Bendlova B; Yip L; Mian C; Vianello F; Tuttle RM; Robenshtok E; Fagin JA; Puxeddu E; Fugazzola L; Czarniecka A; Jarzab B; O'Neill CJ; Sywak MS; Lam AK; Riesco-Eizaguirre G; Santisteban P; Nakayama H; Tufano RP; Pai SI; Zeiger MA; Westra WH; Clark DP; Clifton-Bligh R; Sidransky D; Ladenson PW; Sykorova V

INSTITUCIÓN / INSTITUTION: - Laboratory for Cellular and Molecular Thyroid Research, Johns Hopkins University School of Medicine, Baltimore, MD 21287, USA. mxing1@jhmi.edu

RESUMEN / SUMMARY: - IMPORTANCE: BRAF V600E is a prominent oncogene in papillary thyroid cancer (PTC), but its role in PTC-related patient mortality has not been established. OBJECTIVE: To investigate the relationship between BRAF V600E mutation and PTC-related mortality. DESIGN, SETTING, AND PARTICIPANTS: Retrospective study of 1849 patients (1411 women and 438 men) with a median age of 46 years (interquartile range, 34-58 years) and an

overall median follow-up time of 33 months (interquartile range, 13-67 months) after initial treatment at 13 centers in 7 countries between 1978 and 2011. MAIN OUTCOMES AND MEASURES: Patient deaths specifically caused by PTC. RESULTS: Overall, mortality was 5.3% (45/845; 95% CI, 3.9%-7.1%) vs 1.1% (11/1004; 95% CI, 0.5%-2.0%) (P < .001) in BRAF V600E-positive vs mutation-negative patients. Deaths per 1000 person-years in the analysis of all PTC were 12.87 (95% CI, 9.61-17.24) vs 2.52 (95% CI, 1.40-4.55) in BRAF V600E-positive vs mutation-negative patients; the hazard ratio (HR) was 2.66 (95% CI, 1.30-5.43) after adjustment for age at diagnosis, sex, and medical center. Deaths per 1000 person-years in the analysis of the conventional variant of PTC were 11.80 (95% CI, 8.39-16.60) vs 2.25 (95% CI, 1.01-5.00) in BRAF V600E-positive vs mutation-negative patients; the adjusted HR was 3.53 (95% CI, 1.25-9.98). When lymph node metastasis, extrathyroidal invasion, and distant metastasis were also included in the model, the association of BRAF V600E with mortality for all PTC was no longer significant (HR, 1.21; 95% CI, 0.53-2.76). A higher BRAF V600E-associated patient mortality was also observed in several clinicopathological subcategories, but statistical significance was lost with adjustment for patient age, sex, and medical center. For example, in patients with lymph node metastasis, the deaths per 1000 person-years were 26.26 (95% CI, 19.18-35.94) vs 5.93 (95% CI, 2.96-11.86) in BRAF V600E-positive vs mutation-negative patients (unadjusted HR, 4.43 [95% CI, 2.06-9.51]; adjusted HR, 1.46 [95% CI, 0.62-3.47]). In patients with distant tumor metastasis, deaths per 1000 person-years were 87.72 (95% CI, 62.68-122.77) vs 32.28 (95% CI, 16.14-64.55) in BRAF V600E-positive vs mutation-negative patients (unadjusted HR, 2.63 [95% CI, 1.21-5.72]; adjusted HR, 0.84 [95% CI, 0.27-2.62]). CONCLUSIONS AND RELEVANCE: In this retrospective multicenter study, the presence of the BRAF V600E mutation was significantly associated with increased cancer-related mortality among patients with PTC. Because overall mortality in PTC is low and the association was not independent of tumor features, how to use BRAF V600E to manage mortality risk in patients with PTC is unclear. These findings support further investigation of the prognostic and therapeutic implications of BRAF V600E status in PTC.

[2]

TÍTULO / TITLE: - Hijacking the neuronal NMDAR signaling circuit to promote tumor growth and invasion.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Cell. 2013 Mar 28;153(1):86-100. doi: 10.1016/j.cell.2013.02.051.

●●Enlace al texto completo (gratis o de pago) [1016/j.cell.2013.02.051](https://doi.org/10.1016/j.cell.2013.02.051)

AUTORES / AUTHORS: - Li L; Hanahan D

INSTITUCIÓN / INSTITUTION: - Swiss Institute for Experimental Cancer Research, School of Life Science, Swiss Federal Institute of Technology Lausanne (EPFL), Lausanne 1015, Switzerland.

RESUMEN / SUMMARY: - Glutamate and its receptor N-methyl-D-aspartate receptor (NMDAR) have been associated with cancer, although their functions are not fully understood. Herein, we implicate glutamate-driven NMDAR signaling in a mouse model of pancreatic neuroendocrine tumorigenesis (PNET) and in selected human cancers. NMDAR was upregulated at the periphery of PNET tumors, particularly invasive fronts. Moreover, elevated coexpression of NMDAR and glutamate exporters correlated with poor prognosis in cancer patients. Treatment of a tumor-derived cell line with NMDAR antagonists impaired cancer cell proliferation and invasion. Flow conditions mimicking interstitial fluid pressure induced autologous glutamate secretion, activating NMDAR and its downstream MEK-MAPK and CaMK effectors, thereby promoting invasiveness. Congruently, pharmacological inhibition of NMDAR in mice with PNET reduced tumor growth and invasiveness. Therefore, beyond its traditional role in neurons, NMDAR may be activated in human tumors by fluid flow consequent to higher interstitial pressure, inducing an autocrine glutamate signaling circuit with resultant stimulation of malignancy.

[3]

TÍTULO / TITLE: - Polycomb subunits Ezh1 and Ezh2 regulate the Merkel cell differentiation program in skin stem cells.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - EMBO J. 2013 May 14. doi: 10.1038/emboj.2013.110.

●●Enlace al texto completo (gratis o de pago) [1038/emboj.2013.110](#)

AUTORES / AUTHORS: - Bardot ES; Valdes VJ; Zhang J; Perdigoto CN; Nicolis S; Hearn SA; Silva JM; Ezhkova E

INSTITUCIÓN / INSTITUTION: - Black Family Stem Cell Institute, Icahn School of Medicine at Mount Sinai, New York, NY, USA.

RESUMEN / SUMMARY: - While the Polycomb complex is known to regulate cell identity in ES cells, its role in controlling tissue-specific stem cells is not well understood. Here we show that removal of Ezh1 and Ezh2, key Polycomb subunits, from mouse skin results in a marked change in fate determination in epidermal progenitor cells, leading to an increase in the number of lineage-committed Merkel cells, a specialized subtype of skin cells involved in mechanotransduction. By dissecting the genetic mechanism, we showed that the Polycomb complex restricts differentiation of epidermal progenitor cells by repressing the transcription factor Sox2. Ablation of Sox2 results in a dramatic loss of Merkel cells, indicating that Sox2 is a critical regulator of Merkel cell specification. We show that Sox2 directly activates Atoh1, the obligate regulator

of Merkel cell differentiation. Concordantly, ablation of Sox2 attenuated the Ezh1/2-null phenotype, confirming the importance of Polycomb-mediated repression of Sox2 in maintaining the epidermal progenitor cell state. Together, these findings define a novel regulatory network by which the Polycomb complex maintains the progenitor cell state and governs differentiation in vivo.

[4]

TÍTULO / TITLE: - Causes of Death and Prognostic Factors in Multiple Endocrine Neoplasia Type 1: A Prospective Study: Comparison of 106 MEN1/Zollinger-Ellison Syndrome Patients With 1613 Literature MEN1 Patients With or Without Pancreatic Endocrine Tumors.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Medicine (Baltimore). 2013 May;92(3):135-81. doi: 10.1097/MD.0b013e3182954af1.

●●Enlace al texto completo (gratis o de pago)

[1097/MD.0b013e3182954af1](#)

AUTORES / AUTHORS: - Ito T; Igarashi H; Uehara H; Berna MJ; Jensen RT

INSTITUCIÓN / INSTITUTION: - From the Department of Medicine and Bioregulatory Science (TI, HI), Graduate School of Medical Sciences, Kyushu University, Fukuoka, Japan; Digestive Diseases Branch (TI, HI, HU, MJB, RTJ), National Institute of Diabetes and Digestive and Kidney Diseases, National Institutes of Health, Bethesda, Maryland; and Hopital Kirchberg (MJB), Luxembourg, Luxembourg.

RESUMEN / SUMMARY: - Multiple endocrine neoplasia type 1 (MEN1) is classically characterized by the development of functional or nonfunctional hyperplasia or tumors in endocrine tissues (parathyroid, pancreas, pituitary, adrenal). Because effective treatments have been developed for the hormone excess state, which was a major cause of death in these patients in the past, coupled with the recognition that nonendocrine tumors increasingly develop late in the disease course, the natural history of the disease has changed. An understanding of the current causes of death is important to tailor treatment for these patients and to help identify prognostic factors; however, it is generally lacking. To add to our understanding, we conducted a detailed analysis of the causes of death and prognostic factors from a prospective long-term National Institutes of Health (NIH) study of 106 MEN1 patients with pancreatic endocrine tumors with Zollinger-Ellison syndrome (MEN1/ZES patients) and compared our results to those from the pooled literature data of 227 patients with MEN1 with pancreatic endocrine tumors (MEN1/PET patients) reported in case reports or small series, and to 1386 patients reported in large MEN1 literature series. In the NIH series over a mean follow-up of 24.5 years, 24 (23%) patients died (14 MEN1-related and 10 non-MEN1-related deaths). Comparing the causes of death with the results from the 227 patients in the pooled literature series, we

found that no patients died of acute complications due to acid hypersecretion, and 8%-14% died of other hormone excess causes, which is similar to the results in 10 large MEN1 literature series published since 1995. In the 2 series (the NIH and pooled literature series), two-thirds of patients died from an MEN1-related cause and one-third from a non-MEN1-related cause, which agrees with the mean values reported in 10 large MEN1 series in the literature, although in the literature the causes of death varied widely. In the NIH and pooled literature series, the main causes of MEN1-related deaths were due to the malignant nature of the PETs, followed by the malignant nature of thymic carcinoid tumors. These results differ from the results of a number of the literature series, especially those reported before the 1990s. The causes of non-MEN1-related death for the 2 series, in decreasing frequency, were cardiovascular disease, other nonendocrine tumors > lung diseases, cerebrovascular diseases. The most frequent non-MEN1-related tumor deaths were colorectal, renal > lung > breast, oropharyngeal. Although both overall and disease-related survival are better than in the past (30-yr survival of NIH series: 82% overall, 88% disease-related), the mean age at death was 55 years, which is younger than expected for the general population. Detailed analysis of causes of death correlated with clinical, laboratory, and tumor characteristics of patients in the 2 series allowed identification of a number of prognostic factors. Poor prognostic factors included higher fasting gastrin levels, presence of other functional hormonal syndromes, need for >3 parathyroidectomies, presence of liver metastases or distant metastases, aggressive PET growth, large PETs, or the development of new lesions. The results of this study have helped define the causes of death of MEN1 patients at present, and have enabled us to identify a number of prognostic factors that should be helpful in tailoring treatment for these patients for both short- and long-term management, as well as in directing research efforts to better define the natural history of the disease and the most important factors determining long-term survival at present.

[5]

TÍTULO / TITLE: - Tumor marker and measurement fluctuations may not reflect treatment efficacy in patients with medullary thyroid carcinoma on long-term RET inhibitor therapy.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Ann Oncol. 2013 May 14.

●●Enlace al texto completo (gratis o de pago) [1093/annonc/mdt177](#)

AUTORES / AUTHORS: - Kurzrock R; Atkins J; Wheler J; Fu S; Naing A; Busaidy N; Hong D; Sherman S

INSTITUCIÓN / INSTITUTION: - Hematology-Oncology Division, University of California San Diego Moores Cancer Center, San Diego.

RESUMEN / SUMMARY: - BACKGROUND: RET kinase inhibitors have significant activity in patients with medullary thyroid carcinoma (MTC). PATIENTS AND METHODS: We retrospectively reviewed the electronic medical record for patterns of calcitonin, carcinoembryonic antigen (CEA) and tumor measurement responses in consecutive patients with MTC who received treatment with a RET inhibitor for at least 6 months. RESULTS: Twenty-six patients who received RET kinase inhibitors for at least 6 months were included. All patients experienced an initial decline in calcitonin; 20 (77%) demonstrated later fluctuations in calcitonin, which spiked above baseline levels in 9 individuals (35%). Twenty of the 22 patients (91%) with elevated CEA experienced a decline with treatment, with 11 individuals (50%) later demonstrating transient fluctuations in CEA, including spikes above baseline in 7 patients (32%). Ten of the 26 patients (38%) also demonstrated short-lived fluctuations in RECIST measurements, including changes of over 20% from nadir values. Vacillations in calcitonin, CEA and measurements often occurred repeatedly in individual patients and did not regularly correlate with each other. CONCLUSIONS: Repeated transient fluctuations in tumor markers and measurements are a characteristic of patients with MTC receiving treatment with RET inhibitors, and such short-term vacillations may not reflect responsiveness over the long term. CLINICAL TRIALS INCLUDED: NCT00215605; NCT00244972; NCT00121680; NCT00495872.

[6]

TÍTULO / TITLE: - Treatment of Orbital Metastases From a Primary Midgut Neuroendocrine Tumor With Peptide-Receptor Radiolabeled Therapy Using ¹⁷⁷Lutetium-DOTATATE.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - J Clin Oncol. 2013 Apr 29.

●●Enlace al texto completo (gratis o de pago) [1200/JCO.2012.45.8612](https://doi.org/10.1200/JCO.2012.45.8612)

AUTORES / AUTHORS: - Dobson R; Vinjamuri S; Hsuan J; Banks M; Terlizzo M; Wiesmann H; Daousi C; Poston GP; Cuthbertson DJ

INSTITUCIÓN / INSTITUTION: - University Hospital Aintree, Liverpool, United Kingdom.

[7]

TÍTULO / TITLE: - Body composition variation and impact of low skeletal muscle mass in patients with advanced medullary thyroid carcinoma treated with vandetanib: results from a placebo-controlled study.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - J Clin Endocrinol Metab. 2013 Jun;98(6):2401-8. doi: 10.1210/jc.2013-1115. Epub 2013 Mar 29.

●●Enlace al texto completo (gratis o de pago) [1210/jc.2013-1115](https://doi.org/10.1210/jc.2013-1115)

AUTORES / AUTHORS: - Massicotte MH; Borget I; Broutin S; Baracos VE; Leboulleux S; Baudin E; Paci A; Deroussent A; Schlumberger M; Antoun S

INSTITUCIÓN / INSTITUTION: - 3001 12 Avenue Nord, Sherbrooke, Quebec, Canada, J1H 5N4. marie-helene.massicotte@usherbrooke.ca.

RESUMEN / SUMMARY: - Objectives: Vandetanib was approved by the U.S. Food and Drug Association for the treatment of advanced medullary thyroid cancer (MTC). Because body weight (BW) loss is observed in MTC and because low skeletal muscle mass (SM) is associated with drug toxicity, this study assessed effects of vandetanib on SM and adipose tissue (AT) and explored the association between SM, toxicity, and serum concentration of vandetanib. Methods: Thirty-three patients with MTC received vandetanib (n = 23) or placebo (n = 10) in the ZETA study. Visceral AT (VAT), sc AT (SAT), and SM were assessed with computed tomography imaging by measuring tissue cross-sectional areas (square centimeters per square meter). Dose-limiting toxicities (DLTs) were prospectively recorded. Results: Early at 3 months, compared with placebo group who lost BW, muscle, and SAT, patients treated with vandetanib gained 1.5 kg BW (P = 0.02), 1.3 cm²/m² (approximately 0.7 kg) of SM (P = 0.009), and 4.5 cm²/m² (approximately 0.5 kg) of SAT (P = 0.004) and gained more VAT, 5.1 cm²/m² (approximately 0.7 kg) (P = 0.02). Patients with DLT had lower SM index (37.2 vs 44.3 cm²/m², P = 0.003) and a higher vandetanib serum concentration (1091 vs 739 ng/mL, P = 0.03). Patients with SM index <43.1 cm²/m² had a higher probability of DLT (73% vs 14%, P = 0.004) and a higher vandetanib serum concentration (1037 vs 745 ng/mL, P = 0.04). Patients with the highest compared with the intermediate and lower levels of vandetanib serum concentration experienced more DLT, respectively, 78% vs 40% vs 20% (P = 0.04). Conclusions: Muscle and adipose tissues are restored after only 3 months of vandetanib treatment. Patients with low muscle mass had high vandetanib serum concentration and high incidence of toxicities.

[8]

TÍTULO / TITLE: - Testing new susceptibility genes in the cohort of apparently sporadic pheochromocytoma/paraganglioma patients with clinical characteristics of hereditary syndromes.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Clin Endocrinol (Oxf). 2013 Apr 1. doi: 10.1111/cen.12218.

●●Enlace al texto completo (gratis o de pago) [1111/cen.12218](https://doi.org/10.1111/cen.12218)

AUTORES / AUTHORS: - Peczkowska M; Kowalska A; Sygut J; Waligorski D; Malinoc A; Janaszek-Sitkowska H; Prejbisz A; Januszewicz A; Neumann HP

INSTITUCIÓN / INSTITUTION: - Department of Hypertension, Institute of Cardiology, Warsaw, Poland.

RESUMEN / SUMMARY: - BACKGROUND: Pheochromocytoma (PCC) and paraganglioma (PGL) can occur sporadically or as a part of familial cancer syndromes. Red flags of hereditary syndromes are young age and multifocal tumours. We hypothesized that such patients are candidates for further molecular diagnosis in case of normal results in 'classical' genes. MATERIAL AND METHODS: We selected patients with PCC/PGL under the age of 40 and/or with multiple tumours. First, we tested the genes RET, VHL, NF1, SDHB, SDHC and SDHD. Patients without mutations in these genes were tested for mutations in MAX, TMEM127 and SDHAF2. RESULTS: In 153 patients included, mutations were detected in the classical genes in 72 patients (47%) [RET-22 (14%), VHL-13 (9%), NF1-3 (2%), SDHB-13 (9%), SDHC-3 (2%), SDHD-16 (11%), SDHB large deletions- 2 (1%)]. One patient with MAXc.223C>T (p.R75X) mutation was detected. It was a male with bilateral, metachronous pheochromocytomas diagnosed in 36 and 40 years of age. Remarkably, he showed in the period before the MAX gene was detected, a RET p. Y791F variant. During 10-year follow-up, we did not find any thyroid abnormalities. LOH examination of tumour tissue showed somatic loss of the wild-type allele of MAX. CONCLUSION: Analysis of the MAX gene should be performed in selected patients, especially those with bilateral adrenal pheochromocytoma in whom mutations of the classical genes are absent. Our study provides with further support that Y791F RET is a polymorphism.

[9]

TÍTULO / TITLE: - NeuroD1 regulates survival and migration of neuroendocrine lung carcinomas via signaling molecules TrkB and NCAM.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Proc Natl Acad Sci U S A. 2013 Apr 16;110(16):6524-9. doi: 10.1073/pnas.1303932110. Epub 2013 Apr 3.

●●Enlace al texto completo (gratis o de pago) [1073/pnas.1303932110](https://doi.org/10.1073/pnas.1303932110)

AUTORES / AUTHORS: - Osborne JK; Larsen JE; Shields MD; Gonzales JX; Shames DS; Sato M; Kulkarni A; Wistuba II; Girard L; Minna JD; Cobb MH

INSTITUCIÓN / INSTITUTION: - Department of Pharmacology, University of Texas Southwestern Medical Center, Dallas, TX 75390-9041, USA.

RESUMEN / SUMMARY: - Small-cell lung cancer and other aggressive neuroendocrine cancers are often associated with early dissemination and frequent metastases. We demonstrate that neurogenic differentiation 1 (NeuroD1) is a regulatory hub securing cross talk among survival and migratory-inducing signaling pathways in neuroendocrine lung carcinomas. We find that NeuroD1 promotes tumor cell survival and metastasis in aggressive neuroendocrine lung tumors through regulation of the receptor tyrosine kinase tropomyosin-related kinase B (TrkB). Like TrkB, the prometastatic signaling molecule neural cell adhesion molecule (NCAM) is a downstream target of

NeuroD1, whose impaired expression mirrors loss of NeuroD1. TrkB and NCAM may be therapeutic targets for aggressive neuroendocrine cancers that express NeuroD1.

[10]

TÍTULO / TITLE: - Risk of thyroid cancer in first-degree relatives of patients with non-medullary thyroid cancer by histology type and age at diagnosis: a joint study from five Nordic countries.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - J Med Genet. 2013 Jun;50(6):373-82. doi: 10.1136/jmedgenet-2012-101412. Epub 2013 Apr 13.

●●Enlace al texto completo (gratis o de pago) [1136/jmedgenet-2012-101412](#)

AUTORES / AUTHORS: - Fallah M; Pukkala E; Tryggvadottir L; Olsen JH; Tretli S; Sundquist K; Hemminki K

INSTITUCIÓN / INSTITUTION: - Division of Molecular Genetic Epidemiology, German Cancer Research Center, Im Neuenheimer Feld 580, Heidelberg 69120, Germany; m.fallah@dkfz.de.

RESUMEN / SUMMARY: - BACKGROUND: We aimed to estimate lifetime cumulative risk of thyroid cancer (CRTC) in first-degree relatives of patients with non-medullary thyroid cancers (NMTC), including papillary (PTC)/follicular/oxyphilic/anaplastic thyroid carcinoma, by histology and age at diagnosis in patients and their relatives. DESIGN: A population-based cohort of 63 495 first-degree relatives of 11 206 NMTC patients diagnosed in 1955-2009 in Nordic countries was followed for cancer incidence. Standardised incidence ratios (SIRs) were calculated using histology-specific, age-specific, sex-specific, period-specific and country-specific incidence rates as reference. RESULTS: The 0-84-year CRTC in female relatives of a patient with PTC was 2%, representing a threefold increase over the general population risk (SIR=2.9, 95% CI 2.4 to 3.4; Men: CRTC=1%, SIR=2.5, 95% CI 1.9 to 3.3). When there were ≥ 2 PTC patients diagnosed at age < 60 years in a family, CRTC for female relatives was 10% (male 24%). Twins had a 23-fold increased risk of concordant PTC. Family history of follicular/oxyphilic/anaplastic carcinoma increased CRTC in relatives to about 1-2%. Although no familial case of concordant oxyphilic/anaplastic carcinoma was found, familial risks of discordant histology types of NMTC were interchangeably high for most of the types, for example, higher risk of PTC when a first-degree relative had follicular (SIR=3.0, 95%CI 1.7 to 4.9) or anaplastic (SIR=3.6, 95% CI 1.2 to 8.4) carcinoma. The earlier a patient was diagnosed with PTC in a family, the higher was the SIR in his/her younger relatives. There was a tendency towards concordant age at diagnosis of thyroid cancer among relatives of PTC patients.

CONCLUSIONS: This study provides clinically relevant risk estimates for family members of NMTC patients.

[11]

TÍTULO / TITLE: - Are Circulating Tumor Cells a New, Valid Prognostic Marker in Neuroendocrine Tumors?

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - J Clin Oncol. 2013 May 28.

●●Enlace al texto completo (gratis o de pago) [1200/JCO.2013.49.2132](https://doi.org/10.1200/JCO.2013.49.2132)

AUTORES / AUTHORS: - Antonuzzo L; Meoni G; Di Costanzo F

INSTITUCIÓN / INSTITUTION: - Azienda Ospedaliera Universitaria Careggi, Florence, Italy.

[12]

TÍTULO / TITLE: - The genomic landscape of small intestine neuroendocrine tumors.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - J Clin Invest. 2013 Jun 3;123(6):2502-8. doi: 10.1172/JCI67963. Epub 2013 May 15.

●●Enlace al texto completo (gratis o de pago) [1172/JCI67963](https://doi.org/10.1172/JCI67963)

AUTORES / AUTHORS: - Banck MS; Kanwar R; Kulkarni AA; Boora GK; Metge F; Kipp BR; Zhang L; Thorland EC; Minn KT; Tentu R; Eckloff BW; Wieben ED; Wu Y; Cunningham JM; Nagorney DM; Gilbert JA; Ames MM; Beutler AS

RESUMEN / SUMMARY: - Small intestine neuroendocrine tumors (SI-NETs) are the most common malignancy of the small bowel. Several clinical trials target PI3K/Akt/mTOR signaling; however, it is unknown whether these or other genes are genetically altered in these tumors. To address the underlying genetics, we analyzed 48 SI-NETs by massively parallel exome sequencing. We detected an average of 0.1 somatic single nucleotide variants (SNVs) per 106 nucleotides (range, 0-0.59), mostly transitions (C>T and A>G), which suggests that SI-NETs are stable cancers. 197 protein-altering somatic SNVs affected a preponderance of cancer genes, including FGFR2, MEN1, HOOK3, EZH2, MLF1, CARD11, VHL, NONO, and SMAD1. Integrative analysis of SNVs and somatic copy number variations identified recurrently altered mechanisms of carcinogenesis: chromatin remodeling, DNA damage, apoptosis, RAS signaling, and axon guidance. Candidate therapeutically relevant alterations were found in 35 patients, including SRC, SMAD family genes, AURKA, EGFR, HSP90, and PDGFR. Mutually exclusive amplification of AKT1 or AKT2 was the most common event in the 16 patients with alterations of PI3K/Akt/mTOR signaling. We conclude that sequencing-based analysis may provide provisional grouping of SI-NETs by therapeutic targets or deregulated pathways.

[13]

TÍTULO / TITLE: - Adjuvant Radiation Therapy Increases Disease-Free Survival in Stage IB Merkel Cell Carcinoma.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Ann Plast Surg. 2013 May 6.

●●Enlace al texto completo (gratis o de pago)

[1097/SAP.0b013e31827f4c58](#)

AUTORES / AUTHORS: - Sexton KW; Poteet SP; Hill JB; Schmidt A; Patel A; Del Corral GA; Axt J; Kelley MC; Thayer WP; Shack RB

INSTITUCIÓN / INSTITUTION: - From *Division of General Surgery, daggerDepartment of Plastic Surgery, double daggerSchool of Medicine, Vanderbilt University Medical Center, Nashville, TN; section signDivision of Plastic Surgery, Albany Medical College, Albany, NY; and parallelDivision of Surgical Oncology, Vanderbilt University Medical Center, Nashville, TN.

RESUMEN / SUMMARY: - Merkel cell carcinoma (MCC) is a rare and aggressive cutaneous malignancy. Adjuvant radiation increases survival in advanced stages, but efficacy in stage I disease is unknown. A retrospective review included all patients treated for stage I MCC during a 15-year period at Vanderbilt University Medical Center. Among 42 patients, 26 (62%) had a negative sentinel lymph node biopsy (stage IA) and 16 (38%) had clinically negative lymph nodes (stage IB) at the time of resection. Analysis using Cox regression revealed that higher stage and absence of adjuvant radiation are associated with increased disease recurrence (hazard ratio, 6.29; P = 0.003 and hazard ratio, 4.69; P = 0.013, respectively). Controlling for stage, radiation therapy significantly increased disease-free survival among patients with stage IB disease (P = 0.0026) in a log-rank test comparing Kaplan-Meier curves. These findings support adjuvant radiation therapy in stage IB MCC patients with clinically negative lymph nodes who do not undergo sentinel lymph node biopsy.

[14]

TÍTULO / TITLE: - Patient with adult-onset type II citrullinemia beginning 2 years after operation for duodenal malignant somatostatinoma: Indication for liver transplantation.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Hepatol Res. 2013 May;43(5):563-8. doi: 10.1111/j.1872-034X.2012.01098.x.

●●Enlace al texto completo (gratis o de pago) [1111/j.1872-034X.2012.01098.x](#)

AUTORES / AUTHORS: - Tazawa K; Yazaki M; Fukushima K; Ogata S; Makuuchi M; Morita K; Hiraishi H; Iwasaki Y; Kita J; Kubota K; Ikeda S

INSTITUCIÓN / INSTITUTION: - Department of Medicine (Neurology and Rheumatology), Shinshu University School of Medicine, Matsumoto.

RESUMEN / SUMMARY: - We report a 51-year-old female patient with adult-onset type II citrullinemia (CTLN2) who had a history of pancreatoduodenectomy for duodenal somatostatinoma with metastases to regional lymph nodes at age 49 years, paying special attention to indications for liver transplantation. At age 50 years, she developed hepatic encephalopathy with elevation of plasma ammonia and citrulline levels. A diagnosis of CTLN2 was made by DNA analysis of the SLC25A13 gene and treatment with conservative therapies was begun, including a low-carbohydrate diet and supplementation with arginine and sodium pyruvate. However, despite these treatments, frequent attacks of encephalopathy occurred with markedly elevated plasma ammonia levels. While we were apprehensive regarding the risk of recurrence of somatostatinoma due to immunosuppressive therapy after liver transplantation, the patient was in a critical condition with CTLN2 and it was decided to perform living-donor liver transplantation using a graft obtained from her son. Her postoperative clinical course was uneventful and she has had an active life without recurrence of somatostatinoma for 2 years. This is the first case of CTLN2 with somatostatinoma. As the condition of CTLN2 patients with rapidly progressive courses is often intractable by conservative therapies alone, liver transplantation should be considered even after surgery for malignant tumors in cases with neither metastasis nor recurrence.

[15]

TÍTULO / TITLE: - Long-term Survival of a Patient with Neuroendocrine Carcinoma of the Ampulla of Vater with Lymph Node Micrometastasis.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Am Surg. 2013 May;79(5):199-201.

AUTORES / AUTHORS: - Watanabe Y; Yamada D; Ogawa Y; Fujino M; Kobayashi K; Ryu S; Tanaka M

INSTITUCIÓN / INSTITUTION: - Department of Surgery, Chihaya Hospital, Fukuoka, Japan.

[16]

TÍTULO / TITLE: - Octreotide LAR and bolus octreotide are insufficient for preventing intraoperative complications in carcinoid patients.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - J Surg Oncol. 2013 Jun;107(8):842-6. doi: 10.1002/jso.23323. Epub 2013 Apr 16.

●●Enlace al texto completo (gratis o de pago) 1002/jso.23323

AUTORES / AUTHORS: - Massimino K; Harrskog O; Pommier S; Pommier R

INSTITUCIÓN / INSTITUTION: - Division of General Surgery, Department of Surgery, Oregon Health and Science University, Portland, Oregon.

RESUMEN / SUMMARY: - BACKGROUND AND OBJECTIVES: Surgery in carcinoid patients can provoke a carcinoid crisis, which can have serious sequelae, including death. Octreotide prophylaxis is recommended to prevent carcinoid crisis, however there are few reports of outcomes and no large series examining its efficacy. We hypothesized that a 500 microg prophylactic octreotide dose is sufficient to prevent carcinoid crisis. METHODS: Records of carcinoid patients undergoing abdominal operations during years 2007-2011 were retrospectively reviewed. Octreotide use and intraoperative and postoperative outcomes were analyzed. RESULTS: Ninety-seven intraabdominal operations performed by a single surgeon were reviewed. Ninety percent of patients received preoperative prophylactic octreotide. Fifty-six percent received at least one additional intraoperative dose. Twenty-three patients (24%) experienced an intraoperative complication. Intraoperative complications correlated with presence of hepatic metastases but not presence of carcinoid syndrome. Postoperative complications occurred in 60% of patients with intraoperative complications versus 31% of those with none (P = 0.01). CONCLUSIONS: Significant intraoperative complications occur frequently in patients with hepatic metastases regardless of presence of carcinoid syndrome and despite octreotide LAR or single dose prophylactic octreotide. Occurrence of such events correlates strongly with postoperative complications. Randomized controlled trials are needed to determine whether the administration of prophylactic octreotide is beneficial. J. Surg. Oncol. 2013;107:842-846. © 2013 Wiley Periodicals, Inc.

[17]

TÍTULO / TITLE: - Resection of At-Risk Mesenteric Lymph Nodes Is Associated with Improved Survival in Patients with Small Bowel Neuroendocrine Tumors.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - World J Surg. 2013 May 9.

●●Enlace al texto completo (gratis o de pago) [1007/s00268-013-1918-](http://1007/s00268-013-1918-8)

[8](#)

AUTORES / AUTHORS: - Landry CS; Lin HY; Phan A; Charnsangavej C; Abdalla EK; Aloia T; Nicolas Vauthey J; Katz MH; Yao JC; Fleming JB

INSTITUCIÓN / INSTITUTION: - Department of Surgical Oncology, Baylor University Medical Center, 3410 Worth Street, Suite 235, Dallas, TX, 75246, USA, Christine.Landry@Baylorhealth.edu.

RESUMEN / SUMMARY: - BACKGROUND: Neuroendocrine tumors of the small intestine commonly metastasize to regional lymph nodes (LNs). Single-

institution reports suggest that removal of LNs improves outcome, but comprehensive data are lacking. We hypothesized that the extent of lymphadenectomy reported in a large administrative database would be associated with overall survival for jejunal and ileal neuroendocrine tumors. METHODS: A search of the Surveillance Epidemiology and End Results database was performed for patients with jejunal and ileal neuroendocrine tumors from 1977 to 2004. Descriptive patient characteristics were collected to include age at diagnosis, sex, race, grade, primary tumor size, LN status, number of LNs resected, presence of distant metastasis, and the type of operation. Statistical analyses were limited to patients with only one primary tumor to exclude patients with other malignancies. Univariate and multivariate analyses were performed to analyze the number of LNs resected and the LN ratio (number of positive LNs/total number of LNs removed) to determine the effect on cancer-specific survival. RESULTS: Altogether, 1,364 patients were included in this analysis. Removal of any LNs was associated with improved cancer-specific survival when compared to patients with no LN removal reported ($p = 0.0027$) on univariate analysis. Among those who had any LNs removed, a median of eight LNs were identified in resection specimens with a median LN ratio of 0.29 (range 0-1). On multivariate analysis (adjusting for age and tumor size), patients with >7 LNs removed experienced better cancer-specific survival than those with ≤ 7 LNs removed (median survival not reached vs. 140 months): hazard ratio and 95 % confidence interval were 0.573 (0.402, 0.817) ($p = 0.002$). CONCLUSIONS: This review of a large number of surgical patients demonstrates that regional mesenteric lymphadenectomy in conjunction with resection of the primary tumor is associated with improved survival of patients with small bowel neuroendocrine tumors.

[18]

TÍTULO / TITLE: - A comprehensive next generation sequencing based genetic testing strategy to improve diagnosis of inherited pheochromocytoma and paraganglioma.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - J Clin Endocrinol Metab. 2013 May 10.

●●Enlace al texto completo (gratis o de pago) [1210/jc.2013-1319](#)

AUTORES / AUTHORS: - Rattenberry E; Vialard L; Yeung A; Bair H; McKay K; Jafri M; Canham N; Cole TR; Denes J; Hodgson SV; Irving R; Izatt L; Korbonits M; Kumar AV; Lalloo F; Morrison PJ; Woodward ER; Macdonald F; Wallis Y; Maher ER

INSTITUCIÓN / INSTITUTION: - 1Centre for Rare Diseases and Personalised Medicine, School of Clinical and Experimental Medicine, College of Medical and Dental Sciences, University of Birmingham, Edgbaston, Birmingham, B15 2TH.

RESUMEN / SUMMARY: - Context:Pheochromocytomas and paragangliomas are notable for the high frequency of inherited cases, many of which present as apparently sporadic tumors.Objective:The objective of this study was to establish a comprehensive next generation sequencing (NGS)-based strategy for the diagnosis of pheochromocytoma and paraganglioma patients by testing simultaneously for mutations in MAX, RET, SDHA, SDHB, SDHC, SDHD, SDHAF2, TMEM127 and VHL.Design:After designing and establishing the methodology for the assay, it was validated on DNA samples with known genotype and then patients were studied prospectively.Setting:The study was performed in a diagnostic genetics laboratory.Patients:DNA samples from 205 individuals affected with PPGL/HNPGL (adrenal or extra-adrenal pheochromocytoma/head and neck paraganglioma) were analyzed. A proof of principle study was performed using 85 samples known to contain a variant in one or more of the genes to be tested, followed by prospective analysis of an additional 120 samples.Main Outcome Measure(s):We assessed the ability to use an NGS-based method to perform comprehensive analysis of genes implicated in inherited PPGL/HNPGL.Result:The proof of principle study showed that the NGS assay and analysis gave a sensitivity of 98.7%. A pathogenic mutation was identified in 16.6% of the prospective analysis cohort of 120 patients.Conclusions:A comprehensive NGS-based strategy for the analysis of genes associated with predisposition to PPGL and HNPGL was established, validated and introduced into diagnostic service. The new assay allows simultaneous analysis of nine genes and allows more rapid and cost-effective mutation detection than the previously used conventional Sanger sequencing based methodology.

[19]

TÍTULO / TITLE: - Cytoplasmic staining of OCT4 is a highly sensitive marker of adrenal medullary-derived tissue.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Am J Surg Pathol. 2013 May;37(5):727-33. doi: 10.1097/PAS.0b013e3182793dc2.

●●Enlace al texto completo (gratis o de pago)

[1097/PAS.0b013e3182793dc2](#)

AUTORES / AUTHORS: - Alexander RE; Cheng L; Grignon DJ; Idrees M

INSTITUCIÓN / INSTITUTION: - Department of Pathology and Laboratory Medicine, Indiana University School of Medicine, Indianapolis, IN 46202, USA.

RESUMEN / SUMMARY: - OCT4 immunostaining has become an essential resource in diagnosing germ cell neoplasia. OCT4 is a transcription factor with a characteristic nuclear staining pattern specific to germ cell neoplasms. Our institution has observed that paraganglionic tissue consistently displayed intense cytoplasmic staining by utilizing monoclonal OCT4 antibody, and we

intended to determine whether OCT4 could provide additional diagnostic utility in adrenal tumors. We used monoclonal and polyclonal OCT4 antibodies for comparison of staining patterns and intensities. Thirty-eight pheochromocytomas (8 metastatic), 22 adrenal cortical carcinomas (2 metastatic), 15 metastatic tumors to the adrenal glands, and 10 normal adrenal glands containing cortical and medullary tissue were immunostained with OCT4. A 4-tier system (0 to 3), for recording intensity and extent of cytoplasmic staining, was used. All 30 primary pheochromocytomas displayed strong and diffuse (3+3) cytoplasmic immunoexpression. Six of 8 metastatic pheochromocytomas showed strong immunoexpression (3+3), whereas the remaining 2 showed moderate intensity (2+3). All 22 adrenal cortical carcinomas, including metastatic cases, were completely negative. Only 2 metastatic tumors to the adrenal gland showed weak, cytoplasmic positivity: a small cell carcinoma and a Merkel cell carcinoma. Controls stained in an appropriate nuclear manner. Immunoelectron microscopy demonstrated the antibody interacting with neurosecretory granules. To our knowledge, the cytoplasmic expression of OCT4 in adrenal medulla and pheochromocytoma has not been specifically studied. The goal of this study is to analyze the immunoreactivity of adrenal cortical carcinoma and pheochromocytoma to OCT4 and determine the sensitivity and specificity of this particular staining pattern and to compare monoclonal and polyclonal antibodies.

[20]

TÍTULO / TITLE: - Menin epigenetically represses Hedgehog signaling in MEN1 tumor syndrome.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Cancer Res. 2013 Apr 15;73(8):2650-8. doi: 10.1158/0008-5472.CAN-12-3158. Epub 2013 Apr 11.

●●Enlace al texto completo (gratis o de pago) [1158/0008-5472.CAN-12-3158](#)

AUTORES / AUTHORS: - Gurung B; Feng Z; Iwamoto DV; Thiel A; Jin G; Fan CM; Ng JM; Curran T; Hua X

INSTITUCIÓN / INSTITUTION: - Department of Cancer Biology, Abramson Family Cancer Research Institute, Abramson Cancer Center, University of Pennsylvania Perelman School of Medicine, Philadelphia, PA 19104, USA.

RESUMEN / SUMMARY: - Multiple endocrine neoplasia type 1 (MEN1) is an inherited tumor syndrome that includes susceptibility to pancreatic islet tumors. This syndrome results from mutations in the MEN1 gene, encoding menin. Although menin acts as an oncogenic cofactor for mixed lineage leukemia (MLL) fusion protein-mediated histone H3 lysine 4 methylation, the precise basis for how menin suppresses gene expression and proliferation of pancreatic beta cells remains poorly understood. Here, we show that menin

ablation enhances Hedgehog signaling, a proliferative and oncogenic pathway, in murine pancreatic islets. Menin directly interacts with protein arginine methyltransferase 5 (PRMT5), a negative regulator of gene transcription. Menin recruits PRMT5 to the promoter of the Gas1 gene, a crucial factor for binding of Sonic Hedgehog (Shh) ligand to its receptor PTCH1 and subsequent activation of the Hedgehog signaling pathway, increases repressive histone arginine symmetric dimethylation (H4R3m2s), and suppresses Gas1 expression. Notably, MEN1 disease-related menin mutants have reduced binding to PRMT5, and fail to impart the repressive H4R3m2s mark at the Gas1 promoter, resulting in its elevated expression. Pharmacologic inhibition of Hedgehog signaling significantly reduces proliferation of insulinoma cells, and expression of Hedgehog signaling targets including Ptch1, in MEN1 tumors of mice. These findings uncover a novel link between menin and Hedgehog signaling whereby menin/PRMT5 epigenetically suppresses Hedgehog signaling, revealing it as a target for treating MEN1 tumors.

[21]

TÍTULO / TITLE: - The targeted therapy revolution in neuroendocrine tumors: in search of biomarkers for patient selection and response evaluation.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Cancer Metastasis Rev. 2013 Apr 16.

●●Enlace al texto completo (gratis o de pago) [1007/s10555-013-9421-](#)

[0](#)

AUTORES / AUTHORS: - De Dosso S; Grande E; Barriuso J; Castellano D; Tabernero J; Capdevila J

INSTITUCIÓN / INSTITUTION: - Oncology Institute of Southern Switzerland, Bellinzona, Switzerland.

RESUMEN / SUMMARY: - The molecular events of tumorigenesis in neuroendocrine tumors are poorly understood. Understanding of the molecular alterations will lead to the identification of molecular markers, providing new targets for therapeutics. The purpose of this review was to critically analyze the genetic abnormalities in neuroendocrine tumors, with the aim of identifying biomarkers that indicate a response to agents developed against these targets and to serve as an understanding for the combinations of different active compounds. Human epidermal growth factor receptor 1/2 (EGFR and HER2), vascular endothelial growth factor receptors, hepatocyte growth factor receptor (c-Met), platelet-derived growth factor receptor, insulin-like growth factor, phosphatidylinositol 3-kinase-Akt-mammalian target of rapamycin pathway, and heat shock proteins are all interesting candidate biomarkers with involvement in carcinogenesis and tumor evolution of several neoplasms, including neuroendocrine tumors. Some of them have already been evaluated both as targets and also as biomarkers in clinical trials conducted in advanced

neuroendocrine tumor settings, and others should encourage further investigations into innovative therapeutic opportunities.

[22]

TÍTULO / TITLE: - Uncommon breast malignancies: presentation pattern, prognostic issue and treatment outcome in an Italian single institution experience.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Tumori. 2013 Jan-Feb;99(1):39-44. doi: 10.1700/1248.13786.

●●Enlace al texto completo (gratis o de pago) [1700/1248.13786](#)

AUTORES / AUTHORS: - Bareggi CM; Consonni D; Galassi B; Gambini D; Locatelli E; Visintin R; Runza L; Giroda M; Sfondrini MS; Onida F; Tomirotti M

INSTITUCIÓN / INSTITUTION: - Medical Oncology Unit, Fondazione IRCCS Ca' Granda Ospedale Maggiore Policlinico, Università degli Studi di Milano, Milan, Italy. claudia.bareggi@policlinico.mi.it

RESUMEN / SUMMARY: - AIMS AND BACKGROUND: Often neglected by large clinical trials, patients with uncommon breast malignancies have been rarely analyzed in large series. PATIENTS AND METHODS: Of 2,052 patients diagnosed with breast cancer and followed in our Institution from January 1985 to December 2009, we retrospectively collected data on those with uncommon histotypes, with the aim of investigating their presentation characteristics and treatment outcome. RESULTS: Rare histotypes were identified in 146 patients (7.1% of our total breast cancer population), being classified as follows: tubular carcinoma in 75 (51.4%), mucinous carcinoma in 36 (24.7%), medullary carcinoma in 25 (17.1%) and papillary carcinoma in 10 patients (6.8%). Whereas age at diagnosis was not significantly different among the diverse diagnostic groups, patients with medullary and papillary subtypes had a higher rate of lymph node involvement, similar to that of invasive ductal carcinoma. Early stage diagnosis was frequent, except for medullary carcinoma. Overall, in comparison with our invasive ductal carcinoma patients, those with rare histotypes showed a significantly lower risk of recurrence, with a hazard ratio of 0.28 (95% CI, 0.12-0.62; P = 0.002). CONCLUSIONS: According to our analysis, patients with uncommon breast malignancies are often diagnosed at an early stage, resulting in a good prognosis with standard treatment.

[23]

TÍTULO / TITLE: - Melatonin-mediated insulin synthesis during endoplasmic reticulum stress involves HuD expression in rat insulinoma INS-1E cells.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - J Pineal Res. 2013 May 3. doi: 10.1111/jpi.12064.

●●Enlace al texto completo (gratis o de pago) 1111/jpi.12064

AUTORES / AUTHORS: - Yoo YM

INSTITUCIÓN / INSTITUTION: - Department of Biomedical Engineering, College of Health Science, Yonsei University, Wonju, Korea.

RESUMEN / SUMMARY: - In this study, we investigated how melatonin mediates insulin synthesis through endoplasmic reticulum (ER) via HuD expression in rat insulinoma INS-1E cells. Under ER stress condition (thapsigargin with/without melatonin, tunicamycin with/without melatonin), phosphorylation of AMP-activated protein kinase (p-AMPK) was significantly increased when compared with only with/without melatonin (control/melatonin). Insulin receptor substrate (IRS) two protein was significantly reduced under conditions of ER stress when compared with control/melatonin, but no expression of IRS1 protein was observed. In thapsigargin treatment, melatonin (10, 50 μ m) increased IRS2 protein expression in a dose-dependent manner. p-Akt (Ser473) expression significantly decreased under ER stress condition prior to control/melatonin. Melatonin (10, 50 μ m) significantly reduced nuclear and cellular p85 α expressions in a dose-dependent manner when compared with only thapsigargin or tunicamycin. These results indicate the activation of the aforementioned expressions under regulation of the pathway, AMPK --> IRS2 --> Akt/PKB --> PI3K (p85 α). However, mammalian target of rapamycin and raptor protein, mTORC1, was found to be independent of the ER stress response. In thapsigargin treatment, melatonin increased nuclear mammalian RNA-binding protein (HuD) expression and reduced cellular HuD expression and subsequently resulted in a decrease in cellular insulin level and rise in insulin secretion in a dose-dependent manner. In tunicamycin treatment, HuD and insulin proteins showed similar expression tendencies. These results indicate that ER stress/melatonin, especially thapsigargin/melatonin, increased nuclear HuD expression and subsequently resulted in a decrease in intracellular biosynthesis; it is hypothesized that extracellular secretion of insulin may be regulated by melatonin.

[24]

TÍTULO / TITLE: - Early B-cell differentiation in Merkel cell carcinomas: clues to cellular ancestry.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Cancer Res. 2013 Apr 10.

●●Enlace al texto completo (gratis o de pago) 1158/0008-5472.CAN-13-0616

AUTORES / AUTHORS: - Zur Hausen A; Rennspiess D; Winnepenninckx V; Speel EJ; Kurz AK

INSTITUCIÓN / INSTITUTION: - Department of Pathology, Maastricht University Medical Center.

RESUMEN / SUMMARY: - Merkel cell carcinoma (MCC) is a highly malignant neuroendocrine non melanoma skin cancer, which is associated with the Merkel cell polyoma virus (MCPyV). Recently, expression of the terminal deoxynucleotidyl transferase (TdT) and the paired box gene 5 (PAX 5) has been consistently reported in the majority of MCCs. We tested 21 MCCs for the expression of MCPyV, TdT, PAX5, IgG, IgM, IgA, kappa and lambda by immunohistochemistry and assessed IgH and Igk rearrangement in all 21 MCCs. All of the MCCs revealed specific expression of PAX5 and 72.8% of the MCCs expressed TdT. In addition, most of the MCC revealed specific expression of one or more Ig subclasses and kappa or lambda. One MCC did reveal monoclonal IgH and Igk rearrangement next to 2 other MCCs showing Igk rearrangement. Since co-expression of TdT and PAX5 under physiological circumstances is restricted to pro/pre- and pre-B cells we propose - based on our results - that the cell of origin of MCC is a pro/pre- or pre-B cell rather than the postmitotic Merkel cells. MCPyV infection and transformation of pro-/pre- B cells is likely to induce the expression of simple cytokeratins as has been shown for SV40 in other non epithelial cells. This model of cellular ancestry of MCC might impact therapy and possibly helps to understand why approximately 20% of MCC are MCPyV negative.

[25]

TÍTULO / TITLE: - Multiple microvascular alterations in pancreatic islets and neuroendocrine tumors of a men1 mouse model.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Am J Pathol. 2013 Jun;182(6):2355-67. doi: 10.1016/j.ajpath.2013.02.023. Epub 2013 Apr 12.

●●Enlace al texto completo (gratis o de pago)

1016/j.ajpath.2013.02.023

AUTORES / AUTHORS: - Chu X; Gao X; Jansson L; Quach M; Skogseid B; Barbu A

INSTITUCIÓN / INSTITUTION: - Department of Medical Sciences, Uppsala University, Uppsala, Sweden.

RESUMEN / SUMMARY: - Vascular therapeutic targeting requires thorough evaluation of the mechanisms activated in the specific context of each particular tumor type. We highlight structural, molecular, and functional microvascular aberrations contributing to development and maintenance of pancreatic neuroendocrine tumors (NETs), with special reference to multiple endocrine neoplasia 1 (MEN1) syndrome, using a Men1 mouse model. Tissue samples were analyzed by immunofluorescence to detect vessel density and pericyte distribution within the endocrine pancreas; expression of angiogenic factors was assessed by immunohistochemistry and quantitative real-time PCR in isolated islets and adenomas cultured under normoxic or hypoxic conditions. The

increased vascular density of pancreatic NETs developed in Men1 mice was paralleled by an early and extensive redistribution of pericytes within endocrine tissue. These morphological alterations are supported by, and in some cases preceded by, fine-tuned variations in expression of several angiogenic regulators and are further potentiated by hypoxia. By combining two novel ex vivo and in vivo single-islet and tumor perfusion techniques, we demonstrated that both vascular reactivity and blood perfusion of tumor arterioles are significantly altered in response to glucose and l-nitro-arginine methyl ester. Our findings unravel multiple potential molecular and physiological targets differentially activated in the endocrine pancreas of Men1 mice and highlight the need for in-depth functional studies to fully understand the contribution of each component to development of pancreatic NETs in MEN1 syndrome.

[26]

TÍTULO / TITLE: - Mechanisms of p53 Restriction in Merkel Cell Carcinoma Cells Are Independent of the Merkel Cell Polyoma Virus T Antigens.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - J Invest Dermatol. 2013 Apr 5. doi: 10.1038/jid.2013.169.

●●Enlace al texto completo (gratis o de pago) [1038/jid.2013.169](#)

AUTORES / AUTHORS: - Houben R; Dreher C; Angermeyer S; Borst A; Utikal J; Haferkamp S; Peitsch WK; Schrama D; Hesbacher S

INSTITUCIÓN / INSTITUTION: - Department of Dermatology, Venereology and Allergology, University Hospital Würzburg, Würzburg, Germany.

RESUMEN / SUMMARY: - Merkel cell carcinoma (MCC) is a rare and very aggressive skin cancer with viral etiology. The tumor-associated Merkel cell polyoma virus (MCPV) belongs to a group of viruses encoding T antigens (TAs) that can induce tumorigenesis by interfering with cellular tumor-suppressor proteins like p53. To explore possible modes of p53 inactivation in MCC p53 sequencing, expression analysis and reporter gene assays for functional analyses were performed in a set of MCC lines. In one MCPV-negative and one MCPV-positive cell line, p53 inactivating mutations were found. In the majority of MCC lines, however, wild-type p53 is expressed and displays some transcriptional activity, which is yet not sufficient to effectively restrict cellular survival or growth in these cell cultures. Interestingly, the MCPV TAs are not responsible for this critical lack in p53 activity, as TA knockdown in MCPV-positive MCC cells does not induce p53 activity. In contrast, inhibition of the ubiquitin ligase HDM-2 (human double minute 2) by Nutlin-3^a leads to p53 activation and p53-dependent apoptosis or cell cycle arrest in five out of seven p53 wild-type MCC lines, highlighting p53 as a potential target for future therapies of this aggressive tumor. *Journal of Investigative Dermatology* (2013) 0,000-000. advance online publication, 16 May 2013; doi:10.1038/jid.2013.169.

[27]

TÍTULO / TITLE: - Cocoon Formation in Patients With Midgut Neuroendocrine Tumors: A Rare and Unrecognized Final Pathway.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Pancreas. 2013 Apr 12.

●●Enlace al texto completo (gratis o de pago)

[1097/MPA.0b013e318287ce77](#)

AUTORES / AUTHORS: - Wang YZ; King H; Diebold A

INSTITUCIÓN / INSTITUTION: - From the Department of Surgery, Louisiana State University Health Sciences Center, New Orleans, LA.

RESUMEN / SUMMARY: - **OBJECTIVES:** Neuroendocrine tumors (NETs) are relatively rare with an indolent nature. As a result, treatment is often delayed and passive. The most commonly recognized disease progression leading to death is from the sequelae of bowel obstruction, ischemia, or liver failure secondary to liver metastasis. We recently recognized a rare cocoon-like formation in patients with metastatic gastroenteropancreatic NETs and hypothesize that this may be a distinct, final pathway for these patients. **METHODS:** Ten patients with stage IV gastroenteropancreatic NETs, seen at our center between October 2008 and November 2011, who developed a cocoon were identified. Patient's charts, operative reports, pathology, and tumor markers were reviewed. **RESULTS:** No discernable predictors were identified as precursors to this condition. One patient survived 13 months after cocoon diagnosis, and the remaining 9 patients were all deceased within 5 months. Surgical treatment was attempted in 6 patients and was only partially successful in 1 patient who had the earliest stage of cocoon formation (type 1). **CONCLUSIONS:** Cocoon-like formations in patients with stage IV gastroenteropancreatic NETs is rare and may be a terminal disease progression that has not been previously recognized. The best treatment option remains unknown. Surgical treatment is not advisable, with the exception of type 1 abdominal cocoons.

[28]

TÍTULO / TITLE: - Merkel cell carcinoma concurrent with Bowen's disease: two cases, one with an unusual immunophenotype.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - J Cutan Pathol. 2013 Apr 24. doi: 10.1111/cup.12176.

●●Enlace al texto completo (gratis o de pago) [1111/cup.12176](#)

AUTORES / AUTHORS: - Ishida M; Okabe H

INSTITUCIÓN / INSTITUTION: - Division of Diagnostic Pathology, Department of Clinical Laboratory Medicine, Shiga University of Medical Science, Shiga, Japan.

RESUMEN / SUMMARY: - The concurrence of Merkel cell carcinoma (MCC) and squamous cell carcinoma (SCC) is well known, and MCC concurrent with Bowen's disease has also been documented. Herein, we describe two cases of MCC concurrent with Bowen's disease, and one case exhibited an unusual immunophenotype. An 86-year-old male (Patient 1) and an 87-year-old female (Patient 2) presented with nodules of the chest and cheek, respectively. Histopathologic study revealed Bowen's disease and a proliferation of small round cells in the dermis and/or subcutis. Immunohistochemically, the round cells expressed endocrine markers. 'Dot' immunopositivity for cytokeratin (CK) (AE1/AE3) was observed in both patients. However, dot-like CK20 positivity was present only in the second tumor, and thyroid transcription factor-1 (TTF-1) was only positive in the first. Both cases were negative for Merkel cell polyomavirus (MCPyV). MCC concurrent with SCC usually does not involve detectable MCPyV infection, which suggests that combined MCC may develop through different tumorigenic pathways, such as chronic ultraviolet exposure, as compared to pure MCC. Additionally, concurrent tumors may exhibit an unusual immunophenotype, such as TTF-1+ /CK20(-) .

[29]

TÍTULO / TITLE: - Rectal Neuroendocrine and L-cell Tumors: Diagnostic Dilemma and Therapeutic Strategy.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Am J Surg Pathol. 2013 May 2.

●●Enlace al texto completo (gratis o de pago)

[1097/PAS.0b013e3182819f0f](#)

AUTORES / AUTHORS: - Lee SH; Kim BC; Chang HJ; Sohn DK; Han KS; Hong CW; Lee EJ; Lee JB; Lee DS; Lee IT; Youk EG

INSTITUCIÓN / INSTITUTION: - Departments of *Pathology double daggerSurgery, Daehang Hospital, Seoul daggerCenter for Colorectal Cancer, National Cancer Center, Goyang, Republic of Korea.

RESUMEN / SUMMARY: - Rectal neuroendocrine tumors (NETs) are currently divided into L-cell and non-L-cell types. In the World Health Organization 2010 classification, L-cell tumors are defined as borderline, whereas non-L-cell tumors are considered to represent malignancies. To establish differential diagnostic criteria and therapeutic strategy, we investigated the pathologic features of rectal NETs associated with lymph node metastasis and the clinicopathologic significance of the L-cell phenotype. We analyzed 284 patients with rectal NETs. Factors, including T stage, mitosis, histologic pattern, lymphatic invasion, tumor border, and lymph node metastasis, were retrospectively evaluated. We also evaluated tumor immunoreactivity for L-cell markers, including glucagon-like peptide 1, pancreatic peptide, and peptide YY, in 240 cases. L-cell immunoreactivity was detected in 189 of 240 NETs (79%).

Of the factors evaluated, only age and the frequency of lymphatic invasion were significantly different between patients with L-cell and non-L-cell tumors. Of the 284 patients, 18 (6.3%) had lymph node metastases. Lymphatic invasion and T stage were independent risk factors for lymph node metastasis. Subgroup analysis based on tumor size showed lymph node metastasis in 0%, 4%, 24%, and 100% of patients with NETs with a size of <5, 5 to 9, 10 to 14, and ≥ 15 mm, respectively. Depth of tumor invasion, lymphatic invasion, and mitosis were correlated with tumor size ($P < 0.0001$). In conclusion, L-cell phenotype alone does not guarantee favorable biological characteristics. The clinical management of rectal NETs should depend on tumor size. Careful pathologic examination of lymphatic invasion is necessary.

[30]

TÍTULO / TITLE: - Multiple gastrinomas of the duodenum in a patient with sporadic Zollinger-Ellison syndrome.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Endocrine. 2013 Apr 24.

●●Enlace al texto completo (gratis o de pago) [1007/s12020-013-9965-](#)

[3](#)

AUTORES / AUTHORS: - Campana D; Fusaroli P; Cacciari G

INSTITUCIÓN / INSTITUTION: - Department of Medical and Surgical Sciences, S. Orsola-Malpighi Hospital, University of Bologna, Via Massarenti, 9, 40138, Bologna, Italy, davide.campana@unibo.it.

[31]

TÍTULO / TITLE: - ^{68}Ga DOTANOC PET/CT for Accurate Delineation of Disease Extent in a Case of Sinonasal Small Cell Neuroendocrine Carcinoma.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Clin Nucl Med. 2013 Apr 18.

●●Enlace al texto completo (gratis o de pago)

[1097/RLU.0b013e318279b976](#)

AUTORES / AUTHORS: - Singhal A; Singla S; Sharma P; Dhull VS; Khangembam BC; Kumar R

INSTITUCIÓN / INSTITUTION: - From the Department of Nuclear Medicine, All India Institute of Medical Sciences, New Delhi, India.

RESUMEN / SUMMARY: - Neuroendocrine tumors constitute a heterogeneous group of neoplasms arising from the cells of the neural crest. We present a rare case of primary sinonasal neuroendocrine carcinoma in a 61-year-old male patient where somatostatin receptor PET/CT with ^{68}Ga -DOTANOC correctly delineated the extent of primary tumor as compared to contrast-enhanced CT, thereby changing patient management.

[32]

TÍTULO / TITLE: - Human ASH-1 Promotes Neuroendocrine Differentiation in Androgen Deprivation Conditions and Interferes With Androgen Responsiveness in Prostate Cancer Cells.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Prostate. 2013 May 8. doi: 10.1002/pros.22679.

●●Enlace al texto completo (gratis o de pago) [1002/pros.22679](#)

AUTORES / AUTHORS: - Rapa I; Volante M; Migliore C; Farsetti A; Berruti A; Vittorio Scagliotti G; Giordano S; Papotti M

INSTITUCIÓN / INSTITUTION: - Department of Oncology at San Luigi Hospital, University of Turin, Orbassano, Turin, Italy.

RESUMEN / SUMMARY: - **BACKGROUND:** Neuroendocrine differentiation in prostate cancer is a dynamic process associated to the onset of hormone-refractory disease in vivo. The molecular mechanisms underlying this process are poorly recognized. Our study aimed at testing in vitro the role of hASH-1, a transcription factor implicated in neuroendocrine differentiation, in the onset of neuroendocrine phenotype in prostate cancer cells. **METHODS:** Androgen sensitive LNCAP, androgen insensitive PC-3, and three immortalized prostate cancer cell lines were cultured in standard and androgen deprivation conditions. Expression of hASH-1 was modulated by either specific lentiviral transduction or shRNA interference. Inhibitors of WNT-11, a WNT family member associated to the development of neuroendocrine differentiation in prostate cancer, were also used. Cell viability was measured using the MTS method. Neuroendocrine phenotype was assessed by morphology, immunohistochemistry and real time PCR for several neuroendocrine markers. **RESULTS:** hASH-1 was up-modulated by androgen deprivation in LNCaP cells and in androgen-sensitive immortalized prostate cancer cells, and associated with the onset of a neuroendocrine phenotype. Silencing of hASH-1 prevented neuroendocrine differentiation, as did also the selective interference with the WNT-11 pathway. Moreover, hASH-1 over-expression in LNCaP cells was sufficient to promote neuroendocrine differentiation and increased cell viability at basal and androgen-deprived growth conditions. **CONCLUSION:** In summary, the present data support previous evidence that the acquisition of a neuroendocrine phenotype is linked to androgen responsiveness profiles and suggest a pivotal role of hASH-1 transcription factor, whose activity might be explored as a potential therapeutic target in prostate cancer, with special reference to hormone refractory disease. Prostate 9999:XX-XX. © 2013 Wiley Periodicals, Inc.

[33]

TÍTULO / TITLE: - Vandetanib: A novel targeted therapy for the treatment of metastatic or locally advanced medullary thyroid cancer.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Am J Health Syst Pharm. 2013 May 15;70(10):849-55. doi: 10.2146/ajhp120253.

●●Enlace al texto completo (gratis o de pago) [2146/ajhp120253](#)

AUTORES / AUTHORS: - Ton GN; Banaszynski ME; Kolesar JM

INSTITUCIÓN / INSTITUTION: - GiangThy N. Ton is Pharm.D. student, and Megan E. Banaszynski is Pharm.D. student, School of Pharmacy, University of Wisconsin (UW)-Madison. Jill M. Kolesar, Pharm.D., BCPS, FCCP, is Professor, Division of Pharmacy Practice, School of Pharmacy, UW-Madison, and Director, Analytical Instrumentation Laboratory for Pharmacokinetics, Pharmacodynamics and Pharmacogenetics, UW Carbone Comprehensive Cancer Center.

RESUMEN / SUMMARY: - **PURPOSE:** The pharmacology, pharmacokinetics, efficacy, safety and tolerability, drug and food interactions, cost, and place in therapy of vandetanib are reviewed. **SUMMARY:** Vandetanib is a small-molecule inhibitor of vascular endothelial growth factor receptor, epidermal growth factor receptor, and receptor tyrosine kinase signaling pathways, which are involved in the pathogenesis of medullary thyroid cancer (MTC). Vandetanib is currently approved as an alternative to local therapies for both unresectable and disseminated disease. Vandetanib was evaluated in a randomized, placebo-controlled, double-blind Phase III study comparing vandetanib with placebo in adult patients with unresectable locally advanced or metastatic hereditary or sporadic MTC. Vandetanib demonstrated a statistically significant longer progression-free survival (predicted median of 30.5 months) compared with placebo (median of 19.3 months) (hazard ratio, 0.46; 95% confidence interval, 0.31-0.69; p = 0.0001). The most commonly observed adverse effects of vandetanib include nausea, diarrhea, headache, rash, prolongation of the Q-T interval, and hypertension. Because it can prolong the Q-T interval, vandetanib is contraindicated for use in patients with serious cardiac complications, including congenital long QT syndrome, bradyarrhythmias, uncompensated heart failure, and a history of torsades de pointes. **CONCLUSION:** Vandetanib has been shown to be more effective than placebo in the treatment of advanced MTC; however, it has not been compared with radiation, resection, or embolization. Vandetanib also has significant and fairly common cardiac toxicities. The cost, benefits, and risks of vandetanib for patients with MTC should be weighed, as alternative treatments remain an option for most patients.

[34]

TÍTULO / TITLE: - Sunitinib-induced hyperammonaemia in a patient with pancreatic neuroendocrine tumour.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - J Clin Pharm Ther. 2013 Apr 16. doi: 10.1111/jcpt.12054.

●●Enlace al texto completo (gratis o de pago) [1111/jcpt.12054](#)

AUTORES / AUTHORS: - Shea YF; Chiu WY; Mok MY; Hung IF; Yau CC

INSTITUCIÓN / INSTITUTION: - Department of Medicine, Queen Mary Hospital, the University of Hong Kong, Hong Kong, China.

RESUMEN / SUMMARY: - WHAT IS KNOWN AND OBJECTIVE: Sunitinib can improve progression-free survival and overall survival in patients with advanced pancreatic neuroendocrine tumor (PNET). From clinical trial, most commonly reported adverse events of sunitinib were neutropenia (12%), diarrhea (10%), asthenia (7%), erythrocytopenia (7%), hypertension (7%) and thrombocytopenia (6%). CASE SUMMARY: We report a patient with PNET with liver metastases who developed hyperammonemia with a low dosage of sunitinib probably contributed by the presence of liver metastases. WHAT IS NEW AND CONCLUSIONS: We would like to draw attention to the potential risk of sunitinib induced hyperammonemic encephalopathy even with a low dosage of sunitinib. The absence of sunitinib-induced hyperammonemia during its initial course does not rule out this possibility if there is increased in liver metastases. We suggest checking the ammonia level if patient on sunitinib presented with altered sensorium even if the liver function is normal.

[35]

TÍTULO / TITLE: - A Single Institution's 26-Year Experience With Nonfunctional Pancreatic Neuroendocrine Tumors: A Validation of Current Staging Systems and a New Prognostic Nomogram.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Ann Surg. 2013 May 12.

●●Enlace al texto completo (gratis o de pago)

[1097/SLA.0b013e31828f3174](#)

AUTORES / AUTHORS: - Ellison TA; Wolfgang CL; Shi C; Cameron JL; Murakami P; Mun LJ; Singhi AD; Cornish TC; Olin K; Meriden Z; Choti M; Diaz LA; Pawlik TM; Schulick RD; Hruban RH; Edil BH

INSTITUCIÓN / INSTITUTION: - Departments of *Surgery daggerPathology double daggerOncology, The Sol Goldman Pancreatic Cancer Research Center, the Johns Hopkins University School of Medicine, Sidney Kimmel Comprehensive Cancer Center, Baltimore, MD section signVanderbilt University Medical Center, Department of Pathology, Microbiology and Immunology, Nashville, TN paragraph signThe Johns Hopkins Bloomberg School of Public Health, Department of Biostatistics, Baltimore, MD ||The University of Colorado, Department of Surgery, Aurora, CO.

RESUMEN / SUMMARY: - OBJECTIVE:: To validate the 2010 American Joint Committee on Cancer (AJCC) and 2006 European Neuroendocrine Tumor Society (ENETS) tumor staging systems for pancreatic neuroendocrine tumors (PanNETs) using the largest, single-institution series of surgically resected patients in the literature. BACKGROUND:: The natural history and prognosis of PanNETs have been poorly defined because of the rarity and heterogeneity of these neoplasms. Currently, there are 2 main staging systems for PanNETs, which can complicate comparisons of reports in the literature and thereby hinder progress against this disease. METHODS:: Univariate and multivariate analyses were conducted on the prognostic factors of survival using 326 sporadic, nonfunctional, surgically resected PanNET patients who were cared for at our institution between 1984 and 2011. Current and proposed models were tested for survival prognostication validity as measured by discrimination (Harrel's c-index, HCI) and calibration. RESULTS:: Five-year overall-survival rates for AJCC stages I, II, and IV are 93% (88%-99%), 74% (65%-83%), and 56% (42%-73%), respectively, whereas ENETS stages I, II, III, and IV are 97% (92%-100%), 87% (80%-95%), 73% (63%-84%), and 56% (42%-73%), respectively. Each model has an HCI of 0.68, and they are no different in their ability to predict survival. We developed a simple prognostic tool just using grade, as measured by continuous Ki-67 labeling, sex, and binary age that has an HCI of 0.74. CONCLUSIONS:: Both the AJCC and ENETS staging systems are valid and indistinguishable in their survival prognostication. A new, simpler prognostic tool can be used to predict survival and decrease interinstitutional mistakes and uncertainties regarding these neoplasms.

[36]

TÍTULO / TITLE: - Somatic Mutations in H-RAS in Sporadic Pheochromocytoma and Paraganglioma Identified by Exome Sequencing.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - J Clin Endocrinol Metab. 2013 May 2.

●●Enlace al texto completo (gratis o de pago) [1210/jc.2012-4257](#)

AUTORES / AUTHORS: - Crona J; Delgado Verdugo A; Maharjan R; Stalberg P; Granberg D; Hellman P; Bjorklund P

INSTITUCIÓN / INSTITUTION: - Departments of Surgical (J.C., A.D.V., R.M., P.S., P.H., P.B.) and Medical Sciences (D.G.), Uppsala University, 75185 Uppsala, Sweden.

RESUMEN / SUMMARY: - Context:Up to 60% of pheochromocytoma (PCC) and paraganglioma (PGL) are associated with either somatic or germline mutations in established PCC and PGL susceptibility loci. Most unexplained cases are characterized by an increased activity of the RAS/RAF/ERK signaling pathway. Mutations in RAS subtypes H, K, and N are common in human cancers; however, previous studies have been inconsistent regarding the mutational

status of RAS in PCC and PGL. Objectives: To identify novel disease causing genes in PCC and PGL tumors. Design, setting, and participants: Four benign and sporadic PCC and PGL tumors were subjected to whole exome sequencing using the Illumina HiSeq Platform. Sequences were processed by CLC genomics 4.9 bioinformatics software and the acquired list of genetic variants was filtered against the Catalogue of Somatic Mutations in Cancer database. Findings were validated in an additional 78 PCC and PGL tumor lesions. Results: Exome sequencing identified 2 cases with somatic mutations in the H-RAS. In total, 6.9% (n = 4/58) of tumors negative for mutations in major PCC and PGL loci had mutations in H-RAS: G13R, Q61K, and Q61R. There were 3 PCC and 1 PGL; all had sporadic presentation with benign tumor characteristics and substantial increases in norepinephrine and/or epinephrine. H-RAS tumors were exclusively found in male patients (P = .007). Conclusions: We identified recurrent somatic H-RAS mutations in pheochromocytoma and paraganglioma. Tumors with H-RAS mutations had activation of the RAS/RAF/ERK signaling pathway and were associated with male PCC patients having benign and sporadic disease characteristics. H-RAS could serve as a prognostic and predictive marker as well as a novel therapeutic target.

[37]

TÍTULO / TITLE: - Familial SDHA Mutation Associated With Pituitary Adenoma and Pheochromocytoma/Paraganglioma.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - J Clin Endocrinol Metab. 2013 Jun;98(6):E1103-8. doi: 10.1210/jc.2013-1400. Epub 2013 Apr 30.

●●Enlace al texto completo (gratis o de pago) [1210/jc.2013-1400](#)

AUTORES / AUTHORS: - Dwight T; Mann K; Benn DE; Robinson BG; McKelvie P; Gill AJ; Winship I; Clifton-Bligh RJ

INSTITUCIÓN / INSTITUTION: - PhD, Cancer Genetics, Kolling Institute of Medical Research, Royal North Shore Hospital, St Leonards, New South Wales 2065, Australia. trisha.dwight@sydney.edu.au.

RESUMEN / SUMMARY: - Context: Reports of the coexistence of pituitary adenomas and pheochromocytoma/paraganglioma are uncommon. Recently germline mutations in 2 of the genes encoding succinate dehydrogenase, SDHC and SDHD, were associated with pituitary tumors. Objective: Our aim was to determine whether the development of a pituitary adenoma was associated with SDHA mutation. Patients: A 46-year-old female presented with carotid body paraganglioma (proband). Subsequently the proband's son was diagnosed with a nonfunctioning pituitary macroadenoma at age 30 years. Results: An immunohistochemical analysis of the resected paraganglioma and pituitary adenoma revealed the loss of succinate dehydrogenase subunit B and

succinate dehydrogenase subunit A (SDHA) expression in both tumors, with the preservation of staining in nonneoplastic tissue. Mutation analysis showed a novel SDHA mutation (c.1873C>T, p.His625Tyr) in the germline of the proband as well as in the proband's son. In the paraganglioma of the proband, in addition to the germline mutation, a somatic mutation was observed (c.1865G>A, p.Trp622*). In the pituitary adenoma of the proband's son, loss of SDHA immunoreactivity was paradoxically accompanied by loss of the mutant allele. Conclusions: This is the first report of a pituitary adenoma arising in the setting of germline SDHA mutation. The loss of SDHA protein expression in both the paraganglioma (proband) and pituitary adenoma (proband's son) argues strongly for a causative role of SDHA mutation. This report further strengthens the link between pituitary neoplasia and germline SDH mutation. Although pituitary adenomas appear rare among patients carrying SDH subunit mutations, they may have been underrecognized due to the low penetrance of disease and lack of systematic surveillance.

[38]

TÍTULO / TITLE: - Calcitonin measurement in aspiration needle washout fluids has higher sensitivity than cytology in detecting medullary thyroid cancer: a retrospective multicentre study.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Clin Endocrinol (Oxf). 2013 Apr 30. doi: 10.1111/cen.12234.

●●Enlace al texto completo (gratis o de pago) [1111/cen.12234](#)

AUTORES / AUTHORS: - Trimboli P; Cremonini N; Ceriani L; Saggiorato E; Guidobaldi L; Romanelli F; Ventura C; Laurenti O; Messuti I; Solaroli E; Madaio R; Bongiovanni M; Orlandi F; Crescenzi A; Valabrega S; Giovanella L

INSTITUCIÓN / INSTITUTION: - Section of Endocrinology and Diabetology, Ospedale Israelitico, Rome, Italy.

RESUMEN / SUMMARY: - **OBJECTIVE:** Only few studies analysed the capability of cytology in detecting medullary thyroid cancer (MTC), and they reported a low accuracy of this diagnostic technique. Recently, calcitonin (CT) measurement in aspiration needle washout (FNA-CT) of thyroid and neck lesions has been reported as a sensitive tool for MTC. The aim of this study is to compare the sensitivity of FNA-CT and cytology in detecting MTC and to assess a cut-off value of FNA-CT for clinical practice. **PATIENTS:** Thirty-eight MTC lesions from 36 patients were retrospectively studied, diagnosed and treated in four different centres. Furthermore, 52 nonmedullary lesions from subjects undergone biopsy following increased serum CT were collected as a control group. **RESULTS:** Cytology detected MTC in 21/37 lesions with 56.8% sensitivity. The median FNA-CT value was 2000 pg/ml (range 58-10 000 pg/ml) in MTC and 2.7 pg/ml (range <2-13 pg/ml) in controls (P < 0.001). Using a cut-

off of 39.6 pg/ml, MTC lesions could be identified with 100% sensitivity and specificity. As the most important finding, 14 histologically proved MTC lesions could be detected by FNA-CT, despite they were cytologically diagnosed as benign or nonconclusive. CONCLUSIONS: This study shows, as the first in a multicentre series, that FNA-CT sensitivity is higher than that of cytology in diagnosing MTC. To avoid false-negative MTC by cytology, CT measurement in aspiration needle washout is to be performed in all patients undergoing biopsy following high serum CT.

[39]

TÍTULO / TITLE: - Christia vesperilionis plant extracts as novel antiproliferative agent against human neuroendocrine tumor cells.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Oncol Rep. 2013 Jun;29(6):2219-26. doi: 10.3892/or.2013.2367. Epub 2013 Mar 26.

●●Enlace al texto completo (gratis o de pago) [3892/or.2013.2367](#)

AUTORES / AUTHORS: - Hofer D; Schwach G; Ghaffari Tabrizi-Wizsy N; Sadjak A; Sturm S; Stuppner H; Pfragner R

INSTITUCIÓN / INSTITUTION: - Institute of Pathophysiology and Immunology, Medical University of Graz, Graz, Austria.

RESUMEN / SUMMARY: - Neuroendocrine tumors respond poorly to radiation and conventional chemotherapy, hence surgical removal of the neoplastic tissue is still the most effective way of treatment. In an attempt to find new therapeutic plant extracts of *Christia vesperilionis* (CV) their antitumor potential in human medullary thyroid carcinoma (MTC) and human small intestinal neuroendocrine tumor (SI-NET) cell lines were tested. Proliferation and viability were analyzed using cell counting and WST-1 assay. Apoptosis was determined by microscopy, luminescence assays for caspases 3/7, and expression studies of apoptosis-related genes. CV extracts showed antiproliferative and proapoptotic effects in all MTC and SI-NET cell lines, whereby high growth inhibition was observed by treatment with the ethylacetate-extracts (CV-45) in tumor cell lines but not in normal human fibroblasts. Furthermore CV-45 treatment resulted in alterations of gene expression of PDCD5, MTDH and TNFRSF10b in MTC as well as in SI-NET cells. The results indicate that *Christia vesperilionis* could serve as an anticancer therapeutic for treatment of neuroendocrine tumors.

[40]

TÍTULO / TITLE: - Effects of octreotide therapy in progressive head and neck paragangliomas: Case series.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Head Neck. 2013 Apr 22. doi: 10.1002/hed.23348.

●●Enlace al texto completo (gratis o de pago) 1002/hed.23348

AUTORES / AUTHORS: - van Hulsteijn LT; van Duinen N; Verbist BM; Jansen JC; van der Klaauw AA; Smit JW; Corssmit EP

INSTITUCIÓN / INSTITUTION: - Departments of Endocrinology and Metabolic Diseases, Leiden University Medical Center, Leiden, the Netherlands.

RESUMEN / SUMMARY: - BACKGROUND: Octreotide, a somatostatin analogue, may be beneficial in the treatment of paragangliomas of the head and neck (HNPGLs). METHODS: Non-blinded, prospective intervention study. During one year, patients received a monthly intramuscular injection of 30 mg Sandostatin LAR®. Pre- and post-treatment tumor volumes were assessed by magnetic resonance imaging, urinary catecholamine secretion was measured and HNPGL-related signs and symptoms were recorded. RESULTS: In one out of four included HNPGL-patients, a stabilization of tumor growth was observed after octreotide therapy. In one patient, octreotide therapy was discontinued before the end of the study because of potential side-effects. No improvements in HNPGL-related signs and symptoms were observed. CONCLUSION: In one out of four patients, HNPGL tumor growth velocity was reduced after octreotide therapy. Research assessing the effects of somatostatin analogues targeting different sst subtypes or combined with other therapies may offer new possibilities for the treatment of HNPGLs. Head Neck, 2013.

[41]

TÍTULO / TITLE: - Heparanase promotes lymphangiogenesis and tumor invasion in pancreatic neuroendocrine tumors.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Oncogene. 2013 May 6. doi: 10.1038/onc.2013.142.

●●Enlace al texto completo (gratis o de pago) 1038/onc.2013.142

AUTORES / AUTHORS: - Hunter KE; Palermo C; Kester JC; Simpson K; Li JP; Tang LH; Klimstra DS; Vlodavsky I; Joyce JA

INSTITUCIÓN / INSTITUTION: - 1] Cancer Biology and Genetics Program, Memorial Sloan-Kettering Cancer Center, New York, NY, USA [2] Louis V. Gerstner Jr. Graduate School of Biomedical Sciences, Memorial Sloan-Kettering Cancer Center, New York, NY, USA.

RESUMEN / SUMMARY: - Heparan sulfate proteoglycans are an important and abundant component of the extracellular matrix, which undergo substantial remodeling throughout tumorigenesis via the enzymatic activity of heparanase. Heparanase has been shown to be upregulated in many human cancers; however, its specific functions in human pancreatic neuroendocrine tumors (PanNETs) and spontaneous mouse models of cancer have not been evaluated. Here, we investigated the role of heparanase in PanNETs using patient samples and the RIP1-Tag2 (RT2) PanNET-transgenic mouse model. High heparanase expression significantly correlated with more advanced tumor

stage, higher tumor grade and the presence of distant metastasis in PanNET patients. We genetically manipulated heparanase levels in the RT2 model using heparanase-transgenic mice, which constitutively overexpress heparanase, and heparanase-knockout mice. Heparanase was found to have a critical role in promoting tumor invasion, through both macrophage and cancer cell sources in the tumor microenvironment. In addition, elevated heparanase levels significantly increased peritumoral lymphangiogenesis in vivo and promoted the trans-differentiation of macrophages into lymphatic endothelial cell-like structures in culture. Conversely, we found that heparanase deletion led to increased angiogenesis and pericyte coverage. Together, these data identify important roles for heparanase in regulating several critical aspects of tumorigenesis, demonstrating that heparanase represents a potential therapeutic target for PanNET patients. Oncogene advance online publication, 6 May 2013; doi:10.1038/onc.2013.142.

[42]

TÍTULO / TITLE: - Solid-pseudopapillary pancreatic tumor mimicking a neuroendocrine neoplasm on 18F-FDOPA PET/CT.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - J Clin Endocrinol Metab. 2013 May 13.

●●Enlace al texto completo (gratis o de pago) [1210/jc.2013-1942](#)

AUTORES / AUTHORS: - Imperiale A; Addeo P; Averous G; Jacques Namer I; Bachellier P

INSTITUCIÓN / INSTITUTION: - 1Dept. of Biophysics and Nuclear Medicine, Hautepierre University Hospital, Strasbourg, France.

RESUMEN / SUMMARY: - Abstract Not Available.

[43]

TÍTULO / TITLE: - Circulating serotonin and bone density, structure and turnover in carcinoid syndrome.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - J Clin Endocrinol Metab. 2013 Apr 30.

●●Enlace al texto completo (gratis o de pago) [1210/jc.2012-4174](#)

AUTORES / AUTHORS: - Walsh JS; Newell-Price JD; Debono M; Adaway J; Keevil B; Eastell R

INSTITUCIÓN / INSTITUTION: - 1Academic Unit of Bone Metabolism at the University of Sheffield and Sheffield Teaching Hospitals NHS Foundation Trust, Sheffield, UK.

RESUMEN / SUMMARY: - Context:Gut-derived serotonin has been proposed as a regulator of bone formation, and inhibition of gut serotonin synthesis increases bone formation in rodents. Carcinoid neuroendocrine tumors can produce very

high levels of circulating serotonin and so offer a model of serotonin excess in humans. Objectives: To determine if patients with carcinoid syndrome have lower bone formation markers, lower bone density or poor bone structure compared with healthy controls. Design: We conducted a cross-sectional study of 25 patients with carcinoid syndrome and 25 healthy controls, individually matched to carcinoid patients by gender, age, height and BMI. Outcome measures: We measured circulating serotonin in blood and plasma, and 5HIAA in plasma and urine. We measured lumbar spine and hip BMD by DXA, the distal radius and tibia with high-resolution pQCT, and bone turnover with serum osteocalcin, PINP and CTX. Results: All measures of serotonin and 5HIAA were higher in carcinoid patients than in controls. No measures of bone density or bone structure differed significantly between cases and controls. Osteocalcin was higher in cases than controls (26.0 vs 21.1 ng/ml, p 0.02). PINP and CTX did not differ between cases and controls. In patients with carcinoid syndrome, plasma 5HIAA was positively correlated with osteocalcin. In controls, whole blood serotonin was positively correlated with osteocalcin, PINP and CTX. (R values 0.40 to 0.47, all p <0.05.) Conclusions: High circulating serotonin in carcinoid syndrome is not associated with clinically significant lower bone density, poorer bone structure or lower bone formation markers.

[44]

TÍTULO / TITLE: - Rutin, a bioflavonoid antioxidant protects rat pheochromocytoma (PC-12) cells against 6-hydroxydopamine (6-OHDA)-induced neurotoxicity.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Int J Mol Med. 2013 Jul;32(1):235-40. doi: 10.3892/ijmm.2013.1375. Epub 2013 May 10.

●●Enlace al texto completo (gratis o de pago) [3892/ijmm.2013.1375](#)

AUTORES / AUTHORS: - Magalingam KB; Radhakrishnan A; Haleagrahara N

INSTITUCIÓN / INSTITUTION: - Department of Pathology, Faculty of Medicine, International Medical University, Kuala Lumpur 57000, Malaysia.

RESUMEN / SUMMARY: - Free radicals are widely known to be the major cause of human diseases such as neurodegenerative diseases, cancer, allergy and autoimmune diseases. Human cells are equipped with a powerful natural antioxidant enzyme network. However, antioxidants, particularly those originating from natural sources such as fruits and vegetables, are still considered essential. Rutin, a quercetin glycoside, has been proven to possess antioxidant potential. However, the neuroprotective effect of rutin in pheochromocytoma (PC-12) cells has not been studied extensively. Therefore, the present study was designed to establish the neuroprotective role of rutin as well as to elucidate the antioxidant mechanism of rutin in 6-hydroxydopamine (6-OHDA)-induced toxicity in PC-12 neuronal cells. PC-12 cells were

pretreated with different concentrations of rutin for 4, 8 and 12 h and subsequently incubated with 6-OHDA for 24 h to induce oxidative stress. A significant cytoprotective activity was observed in rutin pretreated cells in a dose-dependent manner. Furthermore, there was marked activation of antioxidant enzymes including superoxide dismutase (SOD), catalase, glutathione peroxidase (GPx), and total glutathione (GSH) in rutin pretreated cells compared to cells incubated with 6-OHDA alone. Rutin significantly reduced lipid peroxidation in 6-OHDA-induced PC-12 cells. On the basis of these observations, it was concluded that the bioflavonoid rutin inhibited 6-OHDA-induced neurotoxicity in PC-12 cells by improving antioxidant enzyme levels and inhibiting lipid peroxidation.

[45]

TÍTULO / TITLE: - Histologic changes in type A chronic atrophic gastritis indicating increased risk of neuroendocrine tumor development: the predictive role of dysplastic and severely hyperplastic enterochromaffin-like cell lesions.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Hum Pathol. 2013 May 2. pii: S0046-8177(13)00088-9. doi: 10.1016/j.humpath.2013.02.005.

●●Enlace al texto completo (gratis o de pago)

1016/j.humpath.2013.02.005

AUTORES / AUTHORS: - Vanoli A; La Rosa S; Luinetti O; Klersy C; Manca R; Alvisi C; Rossi S; Trespi E; Zangrandi A; Sessa F; Capella C; Solcia E

INSTITUCIÓN / INSTITUTION: - Department of Pathology, University of Pavia and Fondazione Istituto di Ricovero e Cura a Carattere Scientifico (IRCCS) Policlinico San Matteo, 27100 Pavia, Italy.

RESUMEN / SUMMARY: - The role of putative preneoplastic enterochromaffin-like cell lesions, either hyperplastic or dysplastic, in the genesis of type 1 enterochromaffin-like cell neuroendocrine tumors associated with type A chronic atrophic gastritis, their actual neoplastic risk, and their precise histogenetic mechanism deserve further clarification by specific histopathologic studies coupled with patient follow-up. A total of 100 patients with severe type A chronic atrophic gastritis, enterochromaffin-like cell hyperplasia, and antral G-cell hyperplasia were endoscopically and histologically followed up for a median of 90.1 months (total of 9118 person-months). Preneoplastic enterochromaffin-like cell lesions and newly developed neuroendocrine tumors were investigated histologically and histochemically, in parallel with enterochromaffin-like cell lesions found in nontumor mucosa of another 32 well-characterized and previously reported type 1 neuroendocrine tumors. Both neuroendocrine and nonneuroendocrine mucosa changes were analyzed and statistically evaluated. During follow-up, 7 of 100 patients developed neuroendocrine tumors: 5 were in a group of 20 cases with previous enterochromaffin-like cell dysplasia and 2

were among 80 cases showing only enterochromaffin-like cell hyperplasia throughout the study (hazard ratio, 20.7; $P < .001$). The severity of enterochromaffin-like cell hyperplasia at first biopsy, with special reference to linear hyperplasia with 6 chains or more per linear millimeter, also increased the risk of neuroendocrine tumor development during follow-up (hazard ratio, 13.0; $P < .001$). Enterochromaffin-like cell microinvasive dysplastic lesions arising at the epithelial renewal zone level, in connection with immature proliferating mucous-neck cells, were found to be linked to early intramucosal neuroendocrine tumor histogenesis. Both enterochromaffin-like cell dysplasia and severe hyperplasia indicate increased risk of neuroendocrine tumor development in type A chronic atrophic gastritis with hypergastrinemia/G-cell hyperplasia.

[46]

TÍTULO / TITLE: - Systemic treatment of advanced lung carcinoid tumors: show me the data!

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Chest. 2013 Apr;143(4):884-6. doi: 10.1378/chest.12-2455.

●●Enlace al texto completo (gratis o de pago) [1378/chest.12-2455](#)

AUTORES / AUTHORS: - Jett JR; Carr LL

[47]

TÍTULO / TITLE: - Comparison of WHO Classifications (2004, 2010), the Hochwald Grading System, and AJCC and ENETS Staging Systems in Predicting Prognosis in Locoregional Well-differentiated Pancreatic Neuroendocrine Tumors.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Am J Surg Pathol. 2013 Jun;37(6):853-9. doi: 10.1097/PAS.0b013e31827fcc18.

●●Enlace al texto completo (gratis o de pago)

[1097/PAS.0b013e31827fcc18](#)

AUTORES / AUTHORS: - Liu TC; Hamilton N; Hawkins W; Gao F; Cao D

INSTITUCIÓN / INSTITUTION: - Departments of *Pathology and Immunology daggerSurgery double daggerDivision of Biostatistics, Washington University School of Medicine, St Louis, MO section signDepartment of Pathology, Key Laboratory of Carcinogenesis and Translational Research (Ministry of Education), Peking University Cancer Hospital & Institute, Beijing, China.

RESUMEN / SUMMARY: - It is difficult to predict prognosis in patients with locoregional well-differentiated (WD) pancreatic neuroendocrine tumors (PanNET). We aimed to examine commonly used stratification systems [World

Health organization (WHO) 2004 and 2010 classifications, American Joint Committee on Cancer (AJCC) and European Neuroendocrine Tumor Society (ENETS) staging, and the Hochwald grading system] for their power in predicting recurrence-free survival (RFS) in these patients. Seventy-five such patients (mean age 56 y, mean follow-up 79 mo) who underwent resection with sufficient tissue material and follow-up data were studied. RFS was correlated with variable clinicopathologic features and stratified with above-mentioned systems. Concordance index (CI) was then calculated. With the WHO 2004 classification, 16, 35, and 24 PanNETs were classified as benign behavior, uncertain behavior, and WD endocrine carcinoma, respectively. By the WHO 2010 classification, 26, 41, and 8 tumors were grade 1, 2, and 3, respectively. Using the Hochwald system, 47 were low grade, and 28 were intermediate grade. The AJCC staging information was complete for 62 patients (13 had the lymph node status Nx) and included: stages IA (19/62), IB (10/62), IIA (10/62), and IIB (23/62). The ENETS staging information was stages I (16/62), IIa (8/62), IIb (14/62), IIIa (0/62), and IIIb (24/62). The average Ki-67 proliferation index (PI) was 8.1%. Factors that predicted RFS included tumor size, nodal metastasis, vascular invasion, perineural invasion, necrosis, mitosis, and Ki-67 PI (all $P < 0.01$). The CI for each system was: 0.6361 for WHO 2004, 0.6735 for WHO 2010, 0.6495 for AJCC staging, 0.6642 for ENETS staging, and 0.6851 for the Hochwald grading system. When these systems were analyzed in conjunction with various additional important pathologic features, combination of the Hochwald grading system and Ki-67 PI achieved the highest CI (0.7946). Therefore, although all these systems predict RFS well in locoregional WD PanNETs, the Hochwald grading system achieves the highest predictive ability. Further predictive power can be achieved by combining the Hochwald grading system and Ki-67 PI.

[48]

TÍTULO / TITLE: - Germline mutations in NF1 and BRCA1 in a family with neurofibromatosis type 1 and early-onset breast cancer.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Breast Cancer Res Treat. 2013 Jun;139(2):597-602. doi: 10.1007/s10549-013-2538-6. Epub 2013 Apr 27.

●●Enlace al texto completo (gratis o de pago) [1007/s10549-013-2538-](#)

[6](#)

AUTORES / AUTHORS: - Campos B; Balmana J; Gardenyes J; Valenzuela I; Abad O; Fabregas P; Volpini V; Diez O

INSTITUCIÓN / INSTITUTION: - Center for Molecular Genetic Diagnosis (CDGM)-IDIBELL, Gran Vía de l'Hospitalet, 199, 08908, L'Hospitalet de Llobregat, España, bcampos@idibell.cat.

RESUMEN / SUMMARY: - Neurofibromatosis type 1 (NF1) is a common dominant autosomal disorder caused by mutations in the NF1 gene. The main manifestations of NF1 are cafe-au-lait spots, neurofibromas, intertriginous freckling, Lisch nodules, and malignancy, including peripheral nerve sheath tumors, central nervous system gliomas, and a variety of other tumors not so clearly defined. The association between NF1 and breast cancer or other gynecologic malignancies seems uncommon and has been scarcely referred in the literature. We describe a family with two females affected by both NF1 and early-onset breast cancer, and a male with NF1. We evaluated whether the concomitance of both disorders could be attributed to a NF1 mutation and its supposed increased risk of breast cancer or to the concurrence of two NF1 and BRCA1/2 germline mutations. Mutation analyses identified a frameshift mutation in BRCA1 and a nonsense mutation in NF1. Our findings stress the importance of considering all phenotypic features in families with both NF1 and breast tumors. To offer a specific risk assessment and management of both conditions, NF1 and BRCA1/2 cancer predisposing genes should be analyzed.

[49]

TÍTULO / TITLE: - In vitro and in vivo activity of cabozantinib (XL184), an inhibitor of RET, MET, and VEGFR2, in a model of medullary thyroid cancer.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Thyroid. 2013 May 24.

●●Enlace al texto completo (gratis o de pago) [1089/thy.2013.0137](#)

AUTORES / AUTHORS: - Bentzien F; Zuzow M; Heald N; Gibson A; Shi Y; Goon L; Yu P; Engst S; Zhang W; Huang S; Zhao L; Vysotskaia V; Chu F; Bautista R; Cancilla B; Lamb P; Joly A; Yakes M

INSTITUCIÓN / INSTITUTION: - Exelixis Inc, S San Francisco, California, United States ; fbentzie@exelixis.com.

RESUMEN / SUMMARY: - Background: A limited number of approved therapeutic options are available to metastatic medullary thyroid cancer patients, and the response to conventional chemotherapy and/or radiotherapy strategies is inadequate. Sporadic and inherited gene mutations in the tyrosine kinase RET result in oncogenic activation that is associated with the pathogenesis of medullary thyroid carcinoma. Cabozantinib is a potent inhibitor of MET, RET, and vascular endothelial factor receptor 2 (VEGFR2), as well as other tyrosine kinases that have been implicated in tumor development and progression. The object of this study was to determine the in vitro biochemical and cellular inhibitory profile of cabozantinib against RET, and in vivo antitumor efficacy using a xenograft model of medullary thyroid carcinoma. Methods: Cabozantinib was evaluated in biochemical and cell based assays that determined the potency of the compound against wild type and activating mutant forms of RET. Additionally, the pharmacodynamic modulation of RET and MET and in vivo

antitumor activity of cabozantinib was examined in a medullary thyroid carcinoma tumor model following sub chronic oral administration. Results: In biochemical assays, cabozantinib inhibited multiple forms of oncogenic RET kinase activity including M918T and Y791F mutants. Additionally, it inhibited proliferation of TT tumor cells that harbor a C634W activating mutation of RET that is most often associated with MEN 2^a and familial medullary thyroid carcinoma. In these same cells grown as xenograft tumors in nude mice, oral administration of cabozantinib resulted in dose dependent tumor growth inhibition that correlated with a reduction in circulating plasma calcitonin levels. Moreover, immunohistochemical analyses of tumors revealed that cabozantinib reduced levels of phosphorylated MET and RET, and decreased tumor cellularity, proliferation and vascularization. Conclusions: Cabozantinib is a potent inhibitor of RET and prevalent mutationally activated forms of RET known to be associated with medullary thyroid carcinoma, and effectively inhibited the growth of a medullary thyroid carcinoma tumor cell model in vitro and in vivo.

[50]

TÍTULO / TITLE: - A comparison of Ki-67 and mitotic count as prognostic markers for metastatic pancreatic and midgut neuroendocrine neoplasms.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Br J Cancer. 2013 Mar 14;108(9):1838-45. doi: 10.1038/bjc.2013.156. Epub 2013 Apr 11.

●●Enlace al texto completo (gratis o de pago) [1038/bjc.2013.156](#)

AUTORES / AUTHORS: - Khan MS; Luong TV; Watkins J; Toumpanakis C; Caplin ME; Meyer T

INSTITUCIÓN / INSTITUTION: - 1] Neuroendocrine Tumour Unit, Centre for Gastroenterology, Royal Free Hospital, London NW3 2QG, UK [2] Department of Oncology, UCL Cancer Institute, Paul O’Gorman Building, Huntley Street, London WC1E 6BT, UK.

RESUMEN / SUMMARY: - Background: The aim of this study was to compare mitotic count (MC) and Ki-67 proliferation index as prognostic markers in pancreatic and midgut neuroendocrine neoplasms (NENs). Methods: Two hundred eighty-five patients with metastatic NENs were recruited. Concordance between histological grade according to either Ki-67 or MC as defined by the European Neuroendocrine Tumour Society guidelines was assessed and the prognostic significance of Ki-67 or MC were evaluated. Results: There was a discrepancy of 44 and 38% in grade assignment when using Ki-67 or MC in pancreatic and midgut NENs, respectively. In multivariate analysis, grade using Ki-67, but not MC, was a significant prognostic factor in determining overall survival (hazard ratios: midgut G2 2.34, G3 15.1, pancreas G2 2.08, G3 11.3). The prognostic value of Ki-67 was improved using a modified classification

(hazard ratios: midgut G2 3.02, for G3 22.1, pancreas G2 5.97, G3 33.8). Conclusion: There is a lack of concordance between Ki-67 and MC in assigning tumour grade. Grade according to Ki-67 was a better prognostic marker than MC for metastatic pancreatic and midgut NENs. We suggest that Ki-67 alone should be used for grading pancreatic and midgut NENs and that the current threshold for classifying G1/G2 tumours should be revised from 2 to 5%.

[51]

TÍTULO / TITLE: - Expression of anaplastic lymphoma kinase in Merkel cell carcinomas.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Hum Pathol. 2013 Apr 8. pii: S0046-8177(13)00053-1. doi: 10.1016/j.humpath.2012.11.021.

●●Enlace al texto completo (gratis o de pago)

[1016/j.humpath.2012.11.021](#)

AUTORES / AUTHORS: - Filtenborg-Barnkob BE; Bzorek M

INSTITUCIÓN / INSTITUTION: - Department of Pathology, Naestved and Slagelse Hospital, Hospital South, Denmark. Electronic address: bfil@regionsjaelland.dk.

RESUMEN / SUMMARY: - This study examines the presence of anaplastic lymphoma kinase protein and anaplastic lymphoma kinase gene rearrangements in Merkel cell carcinomas. A total of 32 cases of Merkel cell carcinomas and 12 cases of small cell lung carcinomas were analyzed. Immunohistochemistry was performed using 3 different anaplastic lymphoma kinase antibody clones (D5F3, 5^{a4}, and anaplastic lymphoma kinase 1). Tumors were divided into high (intensity score 2-3+ in $\geq 25\%$ of the tumor cells) and low expressors (all other positive expression patterns). Anaplastic lymphoma kinase reactivity in Merkel cell carcinoma was observed in 93.8% (30/32) with clone D5F3, 87.5% (28/32) with clone 5^{a4}, and 12.5% (4/32) with clone anaplastic lymphoma kinase 1. One small cell lung carcinoma (1/12; 8.3%) showed anaplastic lymphoma kinase low expression with clone D5F3. Anaplastic lymphoma kinase high expression was observed in 81.3% (26/32) of the Merkel cell carcinomas with clone D5F3, 71.9% (23/32) with clone 5^{a4}, and none with clone anaplastic lymphoma kinase 1. The specificity of anaplastic lymphoma kinase expression in Merkel cell carcinoma versus small cell lung carcinoma was 91.7% with clone D5F3 and 100% with the clones 5^{a4} and anaplastic lymphoma kinase 1. Interphase fluorescence in situ hybridization with the anaplastic lymphoma kinase dual-color, break-apart rearrangement probe was performed on 10 randomly selected Merkel cell carcinoma anaplastic lymphoma kinase high expressors. No rearrangement or other cytogenetic aberration of the anaplastic lymphoma kinase gene locus was identified. In conclusion, the anaplastic lymphoma kinase protein was detected with high

frequency in Merkel cell carcinomas and was useful in distinguishing Merkel cell carcinoma from small cell lung carcinoma. No correlation with anaplastic lymphoma kinase rearrangement was found. Our findings could have important therapeutic consequences for patients, but the role of anaplastic lymphoma kinase in the pathogenesis of Merkel cell carcinoma needs to be further elucidated.

[52]

TÍTULO / TITLE: - Clinical and Dermoscopic Characteristics of Merkel Cell Carcinoma.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Br J Dermatol. 2013 Apr 9. doi: 10.1111/bjd.12376.

●●Enlace al texto completo (gratis o de pago) [1111/bjd.12376](#)

AUTORES / AUTHORS: - Jalilian C; Chamberlain AJ; Haskett M; Rosendahl C; Goh M; Beck H; Keir J; Varghese P; Mar A; Hosking S; Hussain I; Rich M; McLean C; Kelly JW

INSTITUCIÓN / INSTITUTION: - Victorian Melanoma Service, The Alfred Hospital, Melbourne, VIC, Australia.

RESUMEN / SUMMARY: - BACKGROUND: Merkel cell carcinoma (MCC) is an aggressive cutaneous malignancy with a high mortality rate. Diagnosis is often delayed. OBJECTIVE: We set out to characterise the dermoscopic features of Merkel Cell Carcinoma. METHODS: Clinical and dermoscopic images of 12 biopsy proven MCCs were analysed in a retrospective manner with existing dermoscopic criteria being scored independently by three dermatologists. RESULTS: The four most frequent clinical features were cherry red colour, shiny surface, sharp circumscription and nodular morphology. Significant dermoscopic features included linear irregular and polymorphous vessels, poorly focused vessels, milky pink areas, white areas and architectural disorder. Pigmented structures were absent for all lesions. CONCLUSION: The dermoscopic features described herein help the clinician to distinguish MCC from other benign and malignant red nodules. Increasing recognition of the presenting features will facilitate earlier diagnosis of MCC and reduced mortality. This article is protected by copyright. All rights reserved.

[53]

TÍTULO / TITLE: - Chromophobe renal cell carcinoma with sarcomatoid differentiation: a clinicopathologic study of 14 cases.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Anal Quant Cytol Histol. 2013 Apr;35(2):77-84.

AUTORES / AUTHORS: - Lauer SR; Zhou M; Master VA; Osunkoya AO

INSTITUCIÓN / INSTITUTION: - Department of Pathology, Emory University School of Medicine, Atlanta, GA 30322, USA.

RESUMEN / SUMMARY: - **OBJECTIVE:** To investigate the clinicopathologic features of chromophobe renal cell carcinoma with sarcomatoid differentiation. **STUDY DESIGN:** A search was made through the surgical pathology and expert consult files of two major academic institutions from 2003 to 2011 for cases of chromophobe renal cell carcinoma with sarcomatoid differentiation. **RESULTS:** Fourteen patients were identified. The patients included 9 males (64%) and 5 females (36%). The mean patient age was 60.4 years (range, 40-82 years). There was a left-sided predominance: left (9 patients) and right (5 patients). The mean tumor size was 14.6 cm (range, 9.5-28.0 cm), and the mean percentage sarcomatoid differentiation was 67% (range, 30-99%). All tumors exhibited moderate to extensive areas of necrosis. The nonsarcomatoid component in all cases demonstrated classic features of chromophobe renal cell carcinoma. Nine patients (64%) had pT3 disease and 5 patients (36%) had pT4 disease. Five patients (36%) had positive surgical margins. Three patients (21%) had tissue diagnosis of metastatic disease at the time of initial surgery. Six patients (43%) had subsequent pathologic and/or radiologic evidence of multiple or isolated metastatic disease. Follow-up information was available in all 14 patients. Mean follow-up time was 16 weeks (range, 2-84 weeks). Ten of 14 patients (71%) died of disease, 9 of those within 6 months (mean survival time of 10 weeks), 3 patients (21%) were alive with disease, and only 1 patient (7%) was alive with no evidence of disease. **CONCLUSION:** This study is one of the largest series to date specifically examining the clinicopathologic features of sarcomatoid chromophobe renal cell carcinoma in radical nephrectomy specimens and confirms the observation that these tumors behave more aggressively than conventional clear cell renal cell carcinoma or papillary renal cell carcinoma.

[54]

TÍTULO / TITLE: - Clinicopathologic Features of Primary Merkel Cell Carcinoma: A Detailed Descriptive Analysis of a Large Contemporary Cohort.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Dermatol Surg. 2013 Apr 3. doi: 10.1111/dsu.12194.

●●Enlace al texto completo (gratis o de pago) [1111/dsu.12194](#)

AUTORES / AUTHORS: - Schwartz JL; Bichakjian CK; Lowe L; Griffith KA; Frohm ML; Fullen DR; Hayman JA; Lao CD; Shah KS; McLean SA; Bradford CR; Johnson TM; Wong SL

INSTITUCIÓN / INSTITUTION: - Department of Dermatology, University of Michigan Health System, Ann Arbor, Michigan.

RESUMEN / SUMMARY: - **BACKGROUND:** Little uniformity exists in the clinical and histologic variables reported with primary Merkel cell carcinoma (MCC).

OBJECTIVE: To provide a rigorous descriptive analysis of a contemporary cohort and promote the prospective collection of detailed data on MCC for future outcome studies. METHODS AND MATERIALS: A detailed descriptive analysis was performed for clinical and histologic features of 147 patients with 150 primary MCC tumors in a prospectively collected database from 2006 to 2010. RESULTS: The majority (73.5%) of patients were at American Joint Committee on Cancer clinical stage I or II at presentation, 20.4% at stage III, and 6.1% at stage IV. Detailed descriptive clinical and histologic findings are presented. CONCLUSION: Clinical and histologic profiling of primary MCC in the literature is variable and limited. Systematic prospective collection of MCC data is needed for future outcome studies and the ability to compare and share data from multiple sources for this relatively rare tumor.

[55]

TÍTULO / TITLE: - GATA3 expression in paragangliomas: a pitfall potentially leading to misdiagnosis of urothelial carcinoma.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Mod Pathol. 2013 Apr 19. doi: 10.1038/modpathol.2013.76.

●●Enlace al texto completo (gratis o de pago) [1038/modpathol.2013.76](#)

AUTORES / AUTHORS: - So JS; Epstein JI

INSTITUCIÓN / INSTITUTION: - Department of Pathology, The Johns Hopkins Medical Institutions, Baltimore, MD, USA.

RESUMEN / SUMMARY: - GATA3 is a zinc-finger transcription factor, which is expressed in various normal and neoplastic tissues. Amongst tumors, it labels urothelial carcinoma, collecting duct carcinoma of the kidney, breast carcinoma, lymphoma and, uncommonly, endometrial carcinoma. Few studies have investigated its positivity in various neoplasms that may mimic urothelial neoplasms. In this study, we evaluated GATA3 expression in urinary bladder paragangliomas, which may closely mimic urothelial carcinomas. We retrieved 12 cases of paragangliomas from the urinary bladder and 20 cases of paragangliomas from non-urollogic sites using the Hopkins Pathology Data Base system. GATA3 was positive in 10 of the 12 (83%) urinary bladder paragangliomas studied on routine slide sections. Most (6/12) of the staining was diffusely strong (3+) staining, whereas the rest (4/12) that were positive showed mixed intensities (strong 3+ to moderate 2+). The 20 paragangliomas from other sites were constructed into tissue microarrays, wherein three cores from each tumor were taken. Fifteen out of 20 (75%) paragangliomas outside of the bladder were positive for GATA3 staining. Moderate (2+) or strong (3+) staining was seen in 13/20 (65%) of extravesical paragangliomas, ranging from 5 to 100% of the cell labeling (mean 59%, median 60%). In the remaining 7/20 (35%) cases, only weak (2/7) or negative (5/7) immunoreactivity for GATA3 was

seen. An additional 15 cases of metastatic paraganglioma from various primary sites were retrieved with 12 of 15 (80%) metastatic paragangliomas staining positively for GATA3. Overall, for paragangliomas, regardless of site, 78.7% were positive for GATA3. Recognition of this finding will aid pathologists in preventing a misdiagnosis of a urothelial tumor based on GATA3 expression, which is critical given the differences in treatment, follow-up and prognosis between bladder paragangliomas and urothelial carcinoma. Modern Pathology advance online publication, 19 April 2013; doi:10.1038/modpathol.2013.76.

[56]

TÍTULO / TITLE: - Metabolic response demonstrated by 18F-FDG-PET/CT in metastatic medullary thyroid carcinoma under sorafenib therapy.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Endocrine. 2013 Apr 7.

●●Enlace al texto completo (gratis o de pago) [1007/s12020-013-9946-](#)

[6](#)

AUTORES / AUTHORS: - Martinez-Rodriguez I; Banzo I; Carril JM

INSTITUCIÓN / INSTITUTION: - Nuclear Medicine Department, Hospital Universitario Marques de Valdecilla, Universidad de Cantabria, Avda. Valdecilla, s/n, 39008, Santander, España, mimartinez@humv.es.

RESUMEN / SUMMARY: - A 51-year-old woman with a medullary thyroid carcinoma (MTC) presented with a palpable nodule in the right breast. Serum calcitonin was 1,453 pg/ml and carcinoembryonic antigen was 201 ng/ml. Cervical ultrasound and bone scintigraphy were normal. Computed tomography (CT) showed nodules in the right breast and anterior thoracic wall and a hypodense lesion in the right hepatic lobe. Histology of the breast nodule confirmed metastasis from MTC. To evaluate the metabolic activity of these lesions, 18F-fluorodeoxyglucose positron emission tomography-CT (FDG-PET/CT) scan was requested. Axial fused images revealed high FDG uptake by the breast and thoracic wall nodules and the right hepatic lobe. FDG-PET/CT also showed uptake in sacrum and right iliac bone, undetected by CT and bone scintigraphy. After seven cycles of sorafenib PET/CT became negative. Calcitonin decreased to 82.5. A PET/CT performed 6 months later remained negative. This is the first published image of the complete metabolic response of MTC to sorafenib therapy using FDG-PET/CT.

[57]

TÍTULO / TITLE: - Vandetanib: opening a new treatment practice in advanced medullary thyroid carcinoma.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Endocrine. 2013 Apr 14.

●●Enlace al texto completo (gratuito o de pago) [1007/s12020-013-9943-](http://1007/s12020-013-9943-9)

[9](#)

AUTORES / AUTHORS: - Durante C; Paciaroni A; Plasmati K; Trulli F; Filetti S

INSTITUCIÓN / INSTITUTION: - Dipartimento di Medicina Interna e Specialita Mediche, Sapienza Universita di Roma, V.le del Policlinico, 155, 00161, Rome, Italy.

RESUMEN / SUMMARY: - Medullary thyroid cancer (MTC) is frequently diagnosed in a locally advanced or metastatic stage, and 10-year survival rates in these cases are below 20 %. Cytotoxic chemotherapy has no significant impact on overall or progression-free survival. Vandetanib (Caprelsa®, AstraZeneca) is a once-daily oral tyrosine kinase inhibitor that selectively inhibits signalling mediated by growth-factor receptor tyrosine kinase RET (constitutively activated in roughly 60 % of all MTCs), vascular endothelial growth-factor receptors 2 and 3, and epidermal growth-factor receptors. It is the first systemic drug with demonstrated anti-tumor benefits in advanced MTC, and it has recently been approved for locally advanced or metastatic MTC by the United States Food and Drug Administration (April 2011) and the European Medicines Agency (February 2012). This review, starting from the phases II and III efficacy and safety data that led to these approvals, explores important issues related to dosing, patient selection, and strategies for managing the substantial risk of toxicity associated with the drug (including life-threatening cardiac events that are the subject of a black-box warning in the United States). All these issues still remain to be defined. Vandetanib is becoming a standard of care for symptomatic, progressive, metastatic MTCs, to be used selectively in those patients who are likely to benefit from it.

[58]

TÍTULO / TITLE: - First-line irinotecan combined with 5-fluorouracil and leucovorin for high-grade metastatic gastrointestinal neuroendocrine carcinoma.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Tumori. 2013 Jan-Feb;99(1):57-60. doi: 10.1700/1248.13789.

●●Enlace al texto completo (gratuito o de pago) 1700/1248.13789

AUTORES / AUTHORS: - Du Z; Wang Y; Zhou Y; Wen F; Li Q

INSTITUCIÓN / INSTITUTION: - Department of Medical Oncology, Cancer Center, State Key Laboratory of Biotherapy, West China Hospital, Sichuan University, No. 37, Guo Xue Xiang, China.

RESUMEN / SUMMARY: - AIM AND BACKGROUND: High-grade gastrointestinal neuroendocrine neoplasms, ie, poorly differentiated neuroendocrine carcinomas, with no effective therapeutic approaches, have a high ability to metastasize. METHODS: A review of the hospital information system was performed. Patients with histologically proven gastrointestinal neuroendocrine

carcinoma who were treated with irinotecan combined with 5-fluorouracil and leucovorin in a first-line setting were eligible for analysis. We extracted information on age, sex, disease stage, laboratory findings, radiological findings, pathological findings, chemotherapy, effectiveness and adverse events of therapy, and outcomes. RESULTS: Eleven patients were included in the study. Partial response was observed in 7 patients. Median progression-free survival and overall survival were 6.5 (95% CI, 5.1-7.9) and 13.0 (95% CI, 9.8-16.2) months, respectively. No treatment-related deaths occurred. CONCLUSIONS: The results demonstrated that irinotecan combined with 5-fluorouracil and leucovorin is an active regimen with acceptable toxicity for patients with metastatic high-grade gastrointestinal neuroendocrine carcinoma that merits further investigation in prospective trials.

[59]

TÍTULO / TITLE: - Malignant head and neck paragangliomas: Treatment efficacy and prognostic indicators.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Am J Otolaryngol. 2013 May 1. pii: S0196-0709(13)00073-2. doi: 10.1016/j.amjoto.2013.03.010.

●●Enlace al texto completo (gratis o de pago)

1016/j.amjoto.2013.03.010

AUTORES / AUTHORS: - Sethi RV; Sethi RK; Herr MW; Deschler DG

INSTITUCIÓN / INSTITUTION: - Harvard Medical School, Boston, MA, USA.

RESUMEN / SUMMARY: - PURPOSE: Malignant head and neck paragangliomas (MHNPs) are rare and occur in 6%-19% of all HNPs. We sought to identify predictors of survival and compare efficacy of treatment modalities to inform management of this rare disease. MATERIALS AND METHODS: We performed a retrospective cohort study of MHNP cases in the National Cancer Institute Surveillance Epidemiology and End Results database (SEER) from 1973 to 2009. We identified 86 patients with MHNP who had documented regional or distant tumor spread with a median follow-up of 74 months. We used Cox proportional hazard models to assess the significance of demographic factors and treatment on five-year overall survival. RESULTS: The most common treatment was surgery alone (36.0%), followed by surgery with adjuvant radiation (33.7%). Five-year overall survival was 88.1% for surgery alone and 66.5% for adjuvant radiation (p=0.2251). In univariate analysis, regional (vs. distant) spread (HR 0.23, p<0.0001), surgery alone (HR 0.29, p<0.0001) and primary site in the carotid body (HR 0.32, p=0.006) conferred significant survival advantage whereas age>50 (HR 4.04, p<0.0001) worsened survival. Regional (vs. distant) spread (HR 0.42, p=0.046) and age>50 (HR 2.98, p=0.005) remained significant in multivariate analysis. In patients with regional-only disease, five-year overall survival was 95.4% for surgery alone compared to

75.6% for surgery with radiation (p=0.1055). CONCLUSIONS: This is the largest and most contemporary series of MHNP patients. Age and tumor stage are significant factors in predicting survival. Surgical resection significantly improves survival outcomes. From this analysis, the value of adjuvant radiation is not clear.

[60]

TÍTULO / TITLE: - Chromophobe hepatocellular carcinoma with abrupt anaplasia: a proposal for a new subtype of hepatocellular carcinoma with unique morphological and molecular features.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Mod Pathol. 2013 May 3. doi: 10.1038/modpathol.2013.68.

●●Enlace al texto completo (gratis o de pago) [1038/modpathol.2013.68](#)

AUTORES / AUTHORS: - Wood LD; Heaphy CM; Daniel HD; Naini BV; Lassman CR; Arroyo MR; Kamel IR; Cosgrove DP; Boitnott JK; Meeker AK; Torbenson MS

INSTITUCIÓN / INSTITUTION: - Department of Pathology, Johns Hopkins University School of Medicine, Baltimore, MD, USA.

RESUMEN / SUMMARY: - Hepatocellular carcinomas exhibit heterogeneous morphologies by routine light microscopy. Although some morphologies represent insignificant variations in growth patterns, others may represent unrecognized subtypes of hepatocellular carcinoma. Identification of these subtypes could lead to separation of hepatocellular carcinomas into discrete groups with unique underlying genetic changes, prognosis, or therapeutic responses. In order to identify potential subtypes, two pathologists independently screened a cohort of 219 unselected hepatocellular carcinoma resection specimens and divided cases into potential subtypes. One of these promising candidate subtypes was further evaluated using histological and molecular techniques. This subtype was characterized by a unique and consistent set of histological features: smooth chromophobic cytoplasm, abrupt focal nuclear anaplasia (small clusters of tumor cells with marked nuclear anaplasia in a background of tumor cells with bland nuclear cytology), and scattered microscopic pseudocysts-we designate this variant as 'chromophobe hepatocellular carcinoma with abrupt anaplasia'. Thirteen cases were identified (6% of all hepatocellular carcinomas), including 6 men and 7 women with an average age of 61 years. Six cases occurred in cirrhotic livers. Serum AFP was elevated in 6 out of 10 cases. There were a variety of underlying liver diseases, but cases were enrichment for chronic hepatitis B, P=0.006. Interestingly, at the molecular level, this variant was strongly associated with the alternative lengthening of telomere (ALT) phenotype by telomere FISH. ALT is a telomerase-independent mechanism of telomere maintenance and is found in

approximately 8% of unselected hepatocellular carcinomas. In contrast, 11/12 (92%) of the cases of chromophobe hepatocellular carcinoma with abrupt anaplasia were ALT-positive. In summary, we propose that chromophobe hepatocellular carcinoma with abrupt anaplasia represents a new subtype of hepatocellular carcinoma with unique morphological and molecular features. Modern Pathology advance online publication, 3 May 2013; doi:10.1038/modpathol.2013.68.

[61]

TÍTULO / TITLE: - United States-based practice patterns and resource utilization in advanced neuroendocrine tumor treatment.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - World J Gastroenterol. 2013 Apr 21;19(15):2348-54. doi: 10.3748/wjg.v19.i15.2348.

●●Enlace al texto completo (gratis o de pago) [3748/wjg.v19.i15.2348](#)

AUTORES / AUTHORS: - Strosberg J; Casciano R; Stern L; Parikh R; Chulikavit M; Willet J; Liu Z; Wang X; Grzegorzewski KJ

INSTITUCIÓN / INSTITUTION: - H Lee Moffitt Cancer Center and Research Institute, Gastrointestinal Tumor Department, Tampa, FL 33612, USA.

RESUMEN / SUMMARY: - AIM: To assess advanced neuroendocrine tumor (NET) treatment patterns and resource utilization by tumor progression stage and tumor site in the United States. METHODS: United States Physicians meeting eligibility criteria were provided with online data extraction forms to collect patient chart data on recent NET patients. Resource utilization and treatment pattern data were collected over a baseline period (after diagnosis and before tumor progression), as well as initial and secondary progression periods, with progression defined according to measurable radiographic evidence of tumor progression. Resource categories used in the analysis include: Treatments (e.g., surgery, chemotherapy, radiotherapy, targeted therapies), hospitalizations and physician visits, diagnostic tests (biomarkers, imaging, laboratory tests). Comparisons between categories of resource utilization and tumor progression status were examined using univariate (by tumor site) and multivariate analyses (across all tumor sites). RESULTS: Fifty-five physicians were included in the study and completed online data extraction forms using the charts of 110 patients. The physician sample showed a relatively even distribution for those affiliated with academic versus community hospitals (46% vs 55%). Forty (36.3%) patients were reported to have pancreatic NET (pNET), while 70 (63.6%) patients had gastrointestinal tract (GI)/Lung as the primary NET site. Univariate analysis showed the proportion of patients hospitalized increased from 32.7% during baseline to 42.1% in the progression stages. While surgeries were performed at similar proportions overall at baseline and progression, pNET patients, were more likely than GI/Lung NET patients to have undergone

surgery during the baseline (33.3% vs 25.0%) and any progression periods (26.7% vs 23.4%). While peptide-receptor radionuclide and targeted therapy utilization was low across NET types and tumor stages, GI/Lung types exhibited greater utilization of these technologies compared to pNET. Chemotherapy utilization was also greater among GI/Lung types. Multivariate analysis results demonstrated that patients in first progression period were over 3 times more likely to receive chemotherapy when compared to baseline (odds ratio: 3.31; 95%CI: 1.46-7.48, P = 0.0041). Further, progression was associated with a greater likelihood of having a study physician visit [relative risk (RR): 1.54; 95%CI: 1.10-2.17, P = 0.0117], and an increased frequency of other physician visits (RR: 1.84; 95%CI: 1.10-3.10, P = 0.0211). CONCLUSION: Resource utilization in advanced NET in the United States is significant overall and data suggests progression has an impact on resource utilization regardless of NET tumor site.

[62]

TÍTULO / TITLE: - Aberrant anaplastic lymphoma kinase expression in high-grade pulmonary neuroendocrine carcinoma.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - J Clin Pathol. 2013 Apr 25.

●●Enlace al texto completo (gratis o de pago) [1136/jclinpath-2012-201329](#)

AUTORES / AUTHORS: - Nakamura H; Tsuta K; Yoshida A; Shibata T; Wakai S; Asamura H; Furuta K; Tsuda H

INSTITUCIÓN / INSTITUTION: - Department of Pathology and Clinical Laboratories, National Cancer Center Hospital, Tokyo, Japan.

RESUMEN / SUMMARY: - AIMS: In lung cancer with anaplastic lymphoma kinase (ALK) gene rearrangement, accurate diagnosis is essential to identify patients who can be treated with a specific kinase inhibitor. Sensitive ALK immunostaining can provide nearly complete concordance with gene rearrangement status and is expected to serve as a surrogate biomarker for tailored treatment with an ALK inhibitor. In the present report, we describe aberrant ALK expression in a small proportion of pulmonary neuroendocrine carcinomas (NECs) that do not have ALK gene alteration. METHODS: A total of 227 pulmonary NECs were assembled on tissue microarrays and were subjected to highly sensitive ALK staining methods. RESULTS: We observed focal positivity with heterogeneous intensity in 2 (2.9%) of 69 small-cell carcinomas and 1 (0.9%) of 106 large-cell NECs. In contrast, 52 carcinoid tumours were all negative for ALK expression. Neither ALK rearrangement nor amplification was observed using fluorescence in situ hybridisation, and no somatic mutation was detected in three ALK positive NECs. CONCLUSIONS: Thus, this aberrant expression is probably of a wild-type ALK and a potential

pitfall when implementing sensitive ALK immunohistochemistry in the molecular diagnosis of lung cancer.

[63]

TÍTULO / TITLE: - SDHB gene positive metastatic paraganglioma associated with lesions which demonstrate both positive and negative uptake of 18FDG PET and 131MIBG.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - QJM. 2013 May 28.

●●Enlace al texto completo (gratis o de pago) [1093/qjmed/hct118](#)

AUTORES / AUTHORS: - Casey R; Slattery D; Prendeville S; Moore M; Maher M; O'Halloran D

INSTITUCIÓN / INSTITUTION: - From the Department of Endocrinology, Department of Pathology and Department of Radiology, Cork University Hospital, Cork, Ireland.

[64]

TÍTULO / TITLE: - Prophylactic total thyroidectomy using the minimally invasive video assisted (MIVAT) approach in children with multiple endocrine neoplasia (MEN) type 2.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Head Neck. 2013 Apr 22. doi: 10.1002/hed.23358.

●●Enlace al texto completo (gratis o de pago) [1002/hed.23358](#)

AUTORES / AUTHORS: - Glynn RW; Cashman EC; Doody J; Phelan E; Russell JD; Timon C

INSTITUCIÓN / INSTITUTION: - Department of Otorhinolaryngology, Royal Victoria Eye and Ear Hospital, Adelaide Road, Dublin 2, Republic of Ireland.

RESUMEN / SUMMARY: - BACKGROUND: There have been few reports of prophylactic thyroidectomy using the MIVAT approach in children with MEN2. METHODS: Retrospective review of a prospectively maintained database of patients who underwent MIVAT for total thyroidectomy. RESULTS: Six children underwent MIVAT; RET codon mutations identified were 634, 620, 611, and 918. Mean operative time was 93 minutes (range 68-105). Five patients were discharged on the first postoperative day; one patient suffered a postoperative haematoma and was discharged 2 days postoperatively. There were no cases of laryngeal nerve palsy or post-operative hypoparathyroidism. High levels of satisfaction with post-operative cosmesis were reported. Calcitonin levels have been undetectable at follow-up, thus far (mean follow-up 42.8 months). CONCLUSIONS: Whilst our outcomes are similar to those reported using the traditional approach, it is important to note that MIVAT is essentially the same

operation, just performed through a smaller incision, with resulting benefits in terms of pain, cosmesis and, perhaps, morbidity. Head Neck, 2013.

[65]

TÍTULO / TITLE: - Merkel cell carcinoma: diagnosis, management, and outcomes.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Plast Reconstr Surg. 2013 May;131(5):771e-8e. doi: 10.1097/PRS.0b013e3182865cf3.

●●Enlace al texto completo (gratis o de pago)

[1097/PRS.0b013e3182865cf3](#)

AUTORES / AUTHORS: - Senchenkov A; Moran SL

INSTITUCIÓN / INSTITUTION: - Rochester, Minn. From the Division of Plastic and Reconstructive Surgery and the Department of Orthopedics, Mayo Clinic.

RESUMEN / SUMMARY: - SUMMARY: : Merkel cell carcinoma is a rare, aggressive cutaneous malignancy with high rates of recurrence, metastases, and mortality. Its nonspecific clinical presentation often delays the diagnosis, and its treatment is still controversial because of the infrequent nature of the tumor. The authors provide an overview of the current literature on epidemiology, cause, pathogenesis, staging, management, and outcomes of this disease. Effective diagnostic and treatment modalities such as wide local excision of the primary tumor, importance of sentinel node biopsy for staging, evidence for the use of adjuvant radiation therapy, and emphasis on a multidisciplinary treatment approach of Merkel cell carcinoma as it pertains to surgical practice are reviewed.

[66]

TÍTULO / TITLE: - Simulation-based reconstruction of absolute activities from the (99m)Tc/(111)In dual-isotope SPECT/CT: phantom experiments and imaging of neuroendocrine tumors.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Phys Med Biol. 2013 May 21;58(10):3339-57. doi: 10.1088/0031-9155/58/10/3339. Epub 2013 Apr 25.

●●Enlace al texto completo (gratis o de pago) [1088/0031-](#)

[9155/58/10/3339](#)

AUTORES / AUTHORS: - Shcherbinin S; Chamoiseau S; Celler A

INSTITUCIÓN / INSTITUTION: - Department of Radiology, University of British Columbia, 366-828 West 10th Avenue, Vancouver BC, V5Z 1M9, Canada.

RESUMEN / SUMMARY: - We investigate the quantitative accuracy of the reconstruction of absolute (99m)Tc and (111)In activities from (99m)Tc/(111)In dual-isotope SPECT studies. The separate reconstruction of two images is achieved by applying Monte Carlo simulation-based corrections for self-scatter

and cross-talk between energy windows. For method evaluation, a series of (99m)Tc/(111)In physical phantom experiments was performed using a clinical SPECT/CT camera. The containers were filled with different ratios of (99m)Tc and (111)In activities to create cross-talk with varying severity levels. In addition, we illustrate the performance of our method by reconstructing images from four simultaneous (99m)Tc/(111)In SPECT/CT studies of neuroendocrine patients. Similarly to the phantom experiments, clinical cases provide examples with different severity of cross-talk. Phantom experiments showed that Monte Carlo simulation-based corrections improved both quantitative accuracy and visual properties of (99m)Tc and (111)In images. While the errors of absolute activities for both tracers in six containers ranged from 16% to 75% if no corrections for self-scatter and cross-talk were applied, these errors decreased to below 10% when images were reconstructed with the aforementioned corrections. These activities were measured using regions of interest larger than the true sizes of the containers in order to account for the spill-out effect. Analysis of patient studies confirmed that accurate simulation-based compensations improved resolution and contrast for both (99m)Tc and (111)In images.

[67]

- CASTELLANO -

TÍTULO / TITLE: Causa rara de hipertension arterial en la juventud: paraganglioma retroperitoneal con invasion vascular.

TÍTULO / TITLE: - An uncommon cause of high blood pressure in young people: retroperitoneal paraganglioma with vascular invasion.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Nefrologia. 2013 May 17;33(3):435-437. doi: 10.3265/Nefrologia.pre2012.Oct.11744.

●●Enlace al texto completo (gratis o de pago)

[3265/Nefrologia.pre2012.Oct.11744](#)

AUTORES / AUTHORS: - Ayllon-Teran MD; Torres-Lorite M; Benitez-Cantero JM; Sanchez-Hidalgo JM; Diaz-Iglesias C; Rufian-Pena S

[68]

TÍTULO / TITLE: - Absence of Merkel Cell Polyomavirus in Monocytic Leukemias.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Acta Haematol. 2013 Apr 19;130(3):135-137.

●●Enlace al texto completo (gratis o de pago) [1159/000347174](#)

AUTORES / AUTHORS: - Hashida Y; Imajoh M; Taniguchi A; Kamioka M; Daibata M

INSTITUCIÓN / INSTITUTION: - Department of Microbiology and Infection, Kochi Medical School, Kochi University, Kochi, Japan.

[69]

TÍTULO / TITLE: - Diencephalic syndrome as sign of tumor progression in a child with neurofibromatosis type 1 and optic pathway glioma: a case report.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Childs Nerv Syst. 2013 Apr 25.

●●Enlace al texto completo (gratis o de pago) [1007/s00381-013-2109-](#)

[5](#)

AUTORES / AUTHORS: - Cavicchiolo ME; Opocher E; Daverio M; Bendini M; Viscardi E; Bisogno G; Perilongo G; Da Dalt L

INSTITUCIÓN / INSTITUTION: - Department of Woman and Child Health, University Hospital of Padua, Via Giustiniani 3, 35128, Padua, Italy.

RESUMEN / SUMMARY: - ILLUSTRATIVE CASE: We describe the case of a 3-year-old child, diagnosed with familial neurofibromatosis type 1 (NF1) and asymptomatic optic pathway tumor at the age of two, who developed diencephalic syndrome (DS) due to tumor progression 1 year after diagnosis. Magnetic resonance imaging disclosed an enlarging hypothalamic contrast-enhanced mass. Because of the tumor progression, in terms of tumor volume and DS, chemotherapy (CT) treatment was started according to the international protocol for progressive low-grade glioma, with rapid clinical improvement in terms of gain weight and DS resolution. Interestingly, tumor volume was unchanged after CT. **CONCLUSIONS:** This case report highlights the following facts: (1) optic pathway glioma (OPG) in young children with NF1 may have definitive growth potentials and thus, they are worth an accurate clinical follow-up; (2) also, OPG occurring in NF1 patients can be responsible for DS in case of hypothalamus involvement; (3) consequently, the child's growth pattern must be included among the clinical parameters, which must be specifically evaluated during the follow-up of children, with or without NF1, bearing an OPG; and, finally, (4) that DS can improve after CT, even in face of a stable tumor volume.

[70]

TÍTULO / TITLE: - The influence of glutamate receptors on proliferation and metabolic cell activity of neuroendocrine tumors.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Anticancer Res. 2013 Apr;33(4):1267-72.

AUTORES / AUTHORS: - Haas HS; Pfragner R; Tabrizi-Wizsy NG; Rohrer K; Lueftenegger I; Horwath C; Allard N; Rinner B; Sadjak A

INSTITUCIÓN / INSTITUTION: - MA, Institute of Pathophysiology and Immunology, Center of Molecular Medicine, Medical University of Graz, Heinrichstrasse 31^a, 8010 Graz, Austria. helga.haas@medunigraz.at

RESUMEN / SUMMARY: - Neuroendocrine tumors are relatively insensitive to radiation therapy, as well as chemotherapy. Thus, new approaches for alternative therapies are needed. We found that glutamate receptor antagonists are capable of suppressing tumor growth and cell activity of different peripheral malignancies. In the present article we review scientific literature in this field of science. Subtype-specific, non-competitive, metabotropic glutamate receptor-1 antagonists differently suppressed the growth and metabolic cell activity of one human medullary thyroid carcinoma cell line, as well as of four different human midgut neuroendocrine tumor cell lines. Furthermore, PCR analyses revealed that this subtype of glutamate receptor is expressed in these cell lines. These first results indicate that specific metabotropic glutamate receptor antagonists suppress the proliferation and cell activity of neuroendocrine tumor cells, which makes them possible targets in cancer therapy.

[71]

TÍTULO / TITLE: - Expression of CK19 and KIT in resectable pancreatic neuroendocrine tumors.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Tumour Biol. 2013 May 19.

●●Enlace al texto completo (gratis o de pago) [1007/s13277-013-0850-](http://1007/s13277-013-0850-8)

[8](#)

AUTORES / AUTHORS: - Han X; Zhao J; Ji Y; Xu X; Lou W

INSTITUCIÓN / INSTITUTION: - Department of Pancreatic Surgery, ZhongShan Hospital, Fudan University, 180 Fenglin Road, Shanghai, 200032, China.

RESUMEN / SUMMARY: - The objectives of this study were to validate the immunohistochemical expression patterns of CK19 and KIT in primary pancreatic neuroendocrine tumors (pNETs) and to verify the potential biomarkers that can be used to predict the clinical behaviors and postoperative outcomes. The immunohistochemical expressions of CK19 and KIT were determined in normal pancreatic islets and resectable pNETs. Associations of the immunohistochemical features with the clinicopathologic features and prognosis were evaluated. All 20 samples from the normal control group were negative for both KIT and CK19 in normal pancreatic islets. Positive rates for KIT and CK19 in pNETs were 49.5 % (45/91) and 70.0 % (70/100), respectively. The percentages of G1, G2, and G3 tumors were 54.9, 42.9, and 2.2 %, respectively. Ki-67 index was significantly higher in the KIT-positive subgroup than in the KIT-negative subgroup ($p < 0.05$); however, no statistically significant difference of the Ki-67 index was found between the CK19-positive and the CK19-negative subgroups ($p = 0.656$). The positive CK19 expression

was significantly associated with non-functioning tumors, regional lymph nodes metastases, and advanced tumor node metastasis (TNM) stage ($p < 0.05$). Meanwhile, the positive KIT expression was significantly associated with advanced TNM grade ($p < 0.05$). In univariate analysis, the overall survival in patients with positive CK19 expression was significantly lower than that in patients with CK19-negative expression ($p < 0.05$). Also, patients with negative KIT expression showed a tendency of longer survival duration compared with those with positive KIT expression ($p = 0.188$). The high-risk subgroup (2.1 +/- 2.9) might have a higher Ki-67 index than the low-risk subgroup (1.0 +/- 1.7, $p = 0.208$). There was a significant difference in functioning status among the three risk levels ($p < 0.05$). Pairwise comparison prompted that patients at high risk were more prone to have regional lymph nodes metastases, distant metastases, and/or recurrences. In conclusion, the expressions of CK19 and KIT are associated with aggressive clinical behaviors in patients with resectable pNETs. CK19 and KIT may play a role in tumor progression and metastases.

[72]

TÍTULO / TITLE: - Usefulness of significant morphologic characteristics in distinguishing between Merkel cell polyomavirus-positive and Merkel cell polyomavirus-negative Merkel cell carcinomas.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Hum Pathol. 2013 May 10. pii: S0046-8177(13)00110-X. doi: 10.1016/j.humpath.2013.01.026.

●●Enlace al texto completo (gratis o de pago)

[1016/j.humpath.2013.01.026](#)

AUTORES / AUTHORS: - Iwasaki T; Matsushita M; Kuwamoto S; Kato M; Murakami I; Higaki-Mori H; Nakajima H; Sano S; Hayashi K

INSTITUCIÓN / INSTITUTION: - Division of Molecular Pathology, Department of Microbiology and Pathology, Tottori University Faculty of Medicine, Yonago, 683-8503 Japan. Electronic address: iwasakit-path@umin.ac.jp.

RESUMEN / SUMMARY: - Merkel cell polyomavirus (MCPyV) monoclonally integrates into genomes of approximately 80% of Merkel cell carcinomas (MCCs) and undergoes mutation. We previously demonstrated statistically significant differences in tumor cell morphology and biology between MCPyV-positive and MCPyV-negative MCCs. We reassessed the usefulness of our morphologic criteria in differentiating MCPyV-negative and MCPyV-positive MCCs for practical diagnosis. Two trainees and 4 pathologists challenged estimations (5-point confidence scale) of MCPyV infection in MCCs using hematoxylin and eosin-stained slides of 43 new MCC cases and 2 morphologic criteria: (1) nuclear polymorphism is higher and cytoplasm is more abundant in MCPyV-negative MCC cells, and (2) MCC combined with squamous cell carcinoma is defined as MCPyV negative, regardless of tumor cell morphology

of MCC. Subsequently, immunohistochemistry for MCPyV large T antigen and polymerase chain reaction for MCPyV DNA yielded concordant results (MCPyV positivity was 30/43 and 32/43, respectively) for 41 (96%) of 43 cases. The mean accuracy, sensitivity, and specificity of the trainees and pathologists were 92.4% +/- 1.5% and 81.5% +/- 11.0%, 95.6% +/- 6.2% and 90.2% +/- 8.3%, and 83.3% +/- 11.8% and 74.6% +/- 14.1%, respectively. Values of the areas under the curve were 0.80 to 0.95, indicating good informative scores. Using our morphologic criteria, observers can predict the absence of MCPyV infection and diagnose MCPyV-negative MCCs with poor prognosis. Unexpectedly, the performance of trainees was superior to that of pathologists, implying that our morphologic criteria are useful even for practitioners having little experience. Our morphologic criteria will provide pathologists with convenient and reliable hallmarks for accurate MCC diagnosis.

[73]

TÍTULO / TITLE: - Comparison of two low-dose calcium infusion schedules for localisation of insulinomas by selective pancreatic arterial injection with hepatic venous sampling for insulin.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Clin Endocrinol (Oxf). 2013 May 27. doi: 10.1111/cen.12253.

●●Enlace al texto completo (gratis o de pago) [1111/cen.12253](#)

AUTORES / AUTHORS: - Braatvedt G; Jennison E; Holdaway I

INSTITUCIÓN / INSTITUTION: - Department of Endocrinology, Greenlane Clinical Centre and Auckland City Hospital, Auckland, New Zealand.

RESUMEN / SUMMARY: - **OBJECTIVE:** Localisation of small insulinomas may be difficult. Selective pancreatic arterial injection of calcium with hepatic venous insulin measurement (SACST) has been used for this purpose, but can rarely cause hypoglycaemia. Two low-dose concentrations of calcium, 0.25 and 0.1 of the usual concentration used for the test, have been compared for sensitivity of localisation and safety. **DESIGN:** SACST was performed at calcium concentrations of 0.0025 (Protocol A) and 0.00625 (Protocol B) mEq calcium per kg. The standard concentration is 0.025mEq/kg. **PATIENTS:** 21 successive patients with biochemical evidence of insulinoma were studied. **RESULTS:** Using surgical localisation as the gold standard, Protocol A had a sensitivity of 91% and Protocol B 75% for correct localisation. The false positive localisation rate was 16%. No hypoglycaemia was observed. These results compare favorably with published data using the standard calcium concentration. SACST was superior to localisation by non-invasive imaging; in 7 cases SACST was correct when conventional imaging was negative (5) or false positive (2). **CONCLUSION:** Low concentrations of calcium are effective and safe when

performing SACST for localisation of insulinoma. This article is protected by copyright. All rights reserved.

[74]

TÍTULO / TITLE: - Specific analysis of KIT and PDGFR-alpha expression and mutational status in Merkel cell carcinoma.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - J Cutan Pathol. 2013 Apr 13. doi: 10.1111/cup.12160.

●●Enlace al texto completo (gratis o de pago) [1111/cup.12160](#)

AUTORES / AUTHORS: - Swick BL; Srikantha R; Messingham KN

INSTITUCIÓN / INSTITUTION: - Department of Dermatology, University of Iowa, Iowa City, IA, USA; Department of Pathology, University of Iowa, Iowa City, IA, USA; Iowa City Veterans Affairs Medical Center, Iowa City, IA, USA.

RESUMEN / SUMMARY: - BACKGROUND: The purpose of this study was to explore the immunohistochemical and mutational status of the tyrosine kinases KIT and platelet derived growth receptor-alpha (PDGFRA) in Merkel cell carcinoma (MCC). Specifically, we examined the mutated exons in gastrointestinal stromal cell tumors that may confer a treatment response to imatinib mesylate. METHODS: We evaluated KIT and PDGFRA immunostaining in 23 examples of MCC utilizing laser capture microdissection to obtain pure samples of tumor genomic DNA from 18 of 23 examples of MCC. PCR amplification and sequencing of KIT exons 9, 11, 13 and 17, and PDGFRA exons 10, 12, 14 and 18 for mutations was performed. RESULTS: Fifteen of 23 tumors (65%) demonstrated CD117 expression and 22 of 23 tumors (95%) demonstrated PDGFRA expression. A single heterozygous KIT exon 11 base change resulting in an E583K mutation was discovered in 12 of 18 (66%) examples of MCC. In addition, a single nucleotide polymorphism was detected in eight of 18 tumors (44%) in exon 18 of PDGFRA (codon 824; GTC > GTT). CONCLUSIONS: We discovered a novel somatic KIT exon 11 E583K mutation in 66% of tumors. This mutation has been previously described in a human with piebaldism and appears to represent an inactivating mutation. Therefore, despite expression of CD117 and PDGFRA, the absence of activating mutations in these tyrosine kinases makes KIT and PDGFRA unlikely candidates of MCC oncogenesis.

[75]

TÍTULO / TITLE: - Expression of PTEN and mTOR in pancreatic neuroendocrine tumors.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Tumour Biol. 2013 May 19.

●●Enlace al texto completo (gratis o de pago) [1007/s13277-013-0849-](https://doi.org/10.1007/s13277-013-0849-1)

[1](#)

AUTORES / AUTHORS: - Han X; Ji Y; Zhao J; Xu X; Lou W

INSTITUCIÓN / INSTITUTION: - Department of Pancreatic Surgery, Zhong Shan Hospital, Fudan University, 180 Fenglin Road, Shanghai, 200032, China.

RESUMEN / SUMMARY: - The purposes of this study were to clarify the expression patterns of phosphorylated mammalian target of rapamycin (p-mTOR), mTOR, and phosphatase and tensin homolog (PTEN) in primary pancreatic neuroendocrine tumors (pNETs) and their significance in predicting clinical behaviors and postoperative outcomes. The expressions of p-mTOR, mTOR, and PTEN were assessed in 20 normal pancreatic islets and in 90 resectable pNETs using immunohistochemistry. The associations of the biomarker expressions with clinicopathologic variables and survival duration were analyzed. The percentages of G1, G2, and G3 tumors were 54.4, 43.3, and 2.2 %, respectively. A strongly positive staining was observed for both mTOR and PTEN in normal pancreatic islets, whereas negative staining was observed for p-mTOR. In primary pNETs, the mTOR and p-mTOR positive rates were 70.8 % (63/89) and 44.4 % (40/90), respectively. p-mTOR expressions strongly correlate with mTOR expressions. No significant correlation between p-mTOR and clinicopathological features was found. The high expression rate of PTEN was 56.7 % (51/90), whereas the low expression rate was 43.4 % (39/90). PTEN loss (low expression) was significantly more frequent in patients with advanced WHO grades ($p = 0.004$) and in patients with higher Ki-67 index ($p = 0.002$). In our immunohistochemical classification system, the Ki-67 index was significantly higher in the PTEN low expression/p-mTOR-positive subgroup (2.7 ± 2.5) than in the PTEN high expression/p-mTOR-negative subgroup (1.0 ± 1.7 , $p = 0.006$). Patients in the PTEN low expression/p-mTOR-positive subgroup presented a significantly lower 5-year overall survival (OS) than those in the PTEN high expression/p-mTOR-negative subgroup ($p = 0.049$; 5-year OS = 79 vs. 100 %, HR = 7.0). ENETS TNM staging and major vascular invasion were independently associated factors for predicting the overall survival rate of patients ($p = 0.019$ and 0.011 , respectively). In conclusion, positive p-mTOR expression and PTEN loss may have a synergic effect on tumorigenesis and proliferation; targeted therapy based on mTOR/PTEN signal pathway and its associated molecular mechanism may play a role in the treatment of pancreatic neuroendocrine tumors.

[76]

TÍTULO / TITLE: - Surgical Approach to the Management of Medullary Thyroid Cancer: When Is Lymph Node Dissection Needed?

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Oncology. 2013 May 14;84(6):350-355.

●●Enlace al texto completo (gratis o de pago) [1159/000351148](https://doi.org/10.1159/000351148)

AUTORES / AUTHORS: - Stamatakos M; Paraskeva P; Katsaronis P; Tasiopoulou G; Kontzoglou K

INSTITUCIÓN / INSTITUTION: - Department of Surgery, General Hospital of Karditsa, Karditsa, Greece.

RESUMEN / SUMMARY: - Objective: Medullary thyroid cancer (MTC) is a rare and particularly aggressive type of thyroid cancer of neuroendocrine origin. It occurs in hereditary and sporadic forms and its aggressiveness is related to the clinical presentation and the type of RET mutation. Methods: In this article, we present the criteria, as reviewed in contemporary literature, regarding lymph node dissection and radical neck dissection in patients with either sporadic or hereditary MTC. Results: Early diagnosis and treatment remains the key to a 100% cure rate. Conclusions: Routine central lymph node dissection is the minimum procedure recommended for all sporadic and hereditary MTCs. Routine lateral lymph node dissection on either side is necessary when lymph node metastases are found in the central neck compartment.

[77]

TÍTULO / TITLE: - Endoscopic ultrasound (EUS)-guided fiducial placement allows localization of small neuroendocrine tumors during parenchymal-sparing pancreatic surgery.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Surg Endosc. 2013 Apr 30.

●●Enlace al texto completo (gratis o de pago) [1007/s00464-013-2975-](https://doi.org/10.1007/s00464-013-2975-7)

[7](#)

AUTORES / AUTHORS: - Law JK; Singh VK; Khashab MA; Hruban RH; Canto MI; Shin EJ; Saxena P; Weiss MJ; Pawlik TM; Wolfgang CL; Lennon AM

INSTITUCIÓN / INSTITUTION: - Division of Gastroenterology, Johns Hopkins Hospital, Baltimore, MD, USA, jlw8@jhmi.edu.

RESUMEN / SUMMARY: - BACKGROUND: Parenchymal-sparing pancreatic surgery is ideal for lesions such as small pancreatic neuroendocrine tumors (PanNET). However, precise localization of these small tumors at surgery can be difficult. The placement of fiducials under endoscopic ultrasound (EUS) guidance (EUS-F) has been used to direct stereotactic radiation therapy for pancreatic adenocarcinoma. This report describes two cases in which placement of fiducials was used to guide surgical resection. This study aimed to assess the feasibility, safety, and efficacy of using EUS-F for intraoperative localization of small PanNETs. METHODS: A retrospective study analyzed two consecutive patients with small PanNETs who underwent EUS-F followed by enucleation in a tertiary-care referral hospital. The following features were examined: technical success and complication rates of EUS-F, visibility of the fiducial at the time of surgery, and fiducial migration. RESULTS: In the study,

EUS-F was performed for two female patients with a 7-mm and a 9-mm PanNET respectively in the uncinate process and neck of the pancreas. In both patients, EUS-F was feasible with two Visicoil fiducials (Core Oncology, Santa Barbara, CA, USA) placed either within or adjacent to the tumors using a 22-gauge Cook Echotip needle. At surgery, the fiducials were clearly visible on intraoperative ultrasound, and both the tumor and the fiducials were successfully enucleated in both cases. No complications were associated with EUS-F, and no evidence of pancreatitis was shown either clinically or on surgical pathology. This investigation had the limitations of a small single-center study. CONCLUSIONS: For patients undergoing enucleation, EUS-F is technically feasible and safe and aids intraoperative localization of small PanNETs.

[78]

TÍTULO / TITLE: - Putting the Pieces Together: Necrolytic Migratory Erythema and the Glucagonoma Syndrome.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - J Gen Intern Med. 2013 May 17.

●●Enlace al texto completo (gratis o de pago) [1007/s11606-013-2490-](#)

[5](#)

AUTORES / AUTHORS: - Halvorson SA; Gilbert E; Hopkins RS; Liu H; Lopez C; Chu M; Martin M; Sheppard B

INSTITUCIÓN / INSTITUTION: - Division of Hospital Medicine, Department of Medicine, Oregon Health & Science University, 3181 SW Sam Jackson Park Rd, BTE-119, Portland, OR, 97239, USA, halvorss@ohsu.edu.

RESUMEN / SUMMARY: - Glucagonomas are slow-growing, rare pancreatic neuroendocrine tumors. They may present with paraneoplastic phenomena known together as the “glucagonoma syndrome.” A hallmark sign of this syndrome is a rash known as necrolytic migratory erythema (NME). In this paper, the authors describe a patient with NME and other features of the glucagonoma syndrome. The diagnosis of this rare tumor requires an elevated serum glucagon level and imaging confirming a pancreatic tumor. Surgical and medical treatment options are reviewed. When detected early, a glucagonoma is surgically curable. It is therefore imperative that clinicians recognize the glucagonoma syndrome in order to make an accurate diagnosis and refer for treatment.

[79]

TÍTULO / TITLE: - Rare neuroendocrine tumours: Results of the surveillance of rare cancers in Europe project.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Eur J Cancer. 2013 Mar 27. pii: S0959-8049(13)00161-5. doi: 10.1016/j.ejca.2013.02.029.

●●Enlace al texto completo (gratis o de pago) 1016/j.ejca.2013.02.029

AUTORES / AUTHORS: - van der Zwan JM; Trama A; Otter R; Larranaga N; Tavilla A; Marcos-Gragera R; Dei Tos AP; Baudin E; Poston G; Links T

INSTITUCIÓN / INSTITUTION: - Department of Registry and Research, Comprehensive Cancer Centre the Netherlands, Catharijnesingel 55-h, Utrecht, The Netherlands. Electronic address: j.vanderzwan@iknl.nl.

RESUMEN / SUMMARY: - Because of the low incidence, and limited opportunities for large patient volume experiences, there are very few relevant studies of neuroendocrine tumours (NETs). A large population-based database (including cancer patients diagnosed from 1978 to 2002 and registered in 76 population-based cancer registries [CRs]), provided by the project 'surveillance of rare cancers in Europe' (RARECARE) is used to describe the basic indicators of incidence, prevalence and survival of NETs, giving a unique overview on the burden of NETs in Europe. NETs at all cancer sites, excluding lung, were analysed in this study. In total over 20,000 incident cases of NETs were analysed and a data quality check upon specific NETs was performed. The overall incidence rate for NETs was 25/1,000,000 and was highest in patients aged 65 years and older with well differentiated endocrine carcinomas (non-functioning pancreatic and gastrointestinal) (40 per 1,000,000). We estimated that slightly more than 100,000 people were diagnosed with NETs and still alive in EU27 at the beginning of 2008. Overall, NETs had a 5-year relative survival of 50%; survival was low (12%) for poorly differentiated endocrine carcinoma, and relatively high (64%) for well differentiated carcinoma (not functioning of the pancreas and digestive organs). Within NETs, endocrine carcinoma of thyroid gland had the best 5-year relative survival (82%). Because of the complexity and number of the different disciplines involved with NETs (as they arise in many organs), a multidisciplinary approach delivered in highly qualified reference centres and an international network between those centres is recommended.

[80]

- CASTELLANO -

TÍTULO / TITLE: Carcinoma neuroendocrino de célula grande prostática. A propósito de un caso.

TÍTULO / TITLE: - Large-cell neuroendocrine carcinoma of prostate. Case report.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Arch Esp Urol. 2013 May;66(4):368-371.

AUTORES / AUTHORS: - Moratalla Charcos LM; Pastor Navarro T; Cortes Vizcaino V; Osca Garcia JM; Gil Salom M

INSTITUCIÓN / INSTITUTION: - Urology Department.Hospital Universitario Dr.Peset.Valencia.Spain.

RESUMEN / SUMMARY: - OBJECTIVE: To report a case of a neuroendocrine differentiation in a prostate cancer patient, a rare subtype. METHODS: We describe the case of a patient diagnosed with adenocarcinoma of the prostate initially, who presented hematuria due to disease progression with neuroendocrine differentiation despite androgen-deprivation therapy (ADT). DISCUSSION: Prostate cancer is the most common tumor in men. Histologically they are diagnosed as adenocarcinomas, which followed by ADT for a long time, develop neuroendocrine differentiation (NED). CONCLUSIONS: The prognostic significance of NED remains controversial. We must think in neuroendocrine differentiation in ADT-treated patient with disease progression and low PSA.

[81]

TÍTULO / TITLE: - Metastases from Neuroendocrine Tumors to the Breast Are More Common than Previously Thought. A Diagnostic Pitfall?

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - World J Surg. 2013 Apr 17.

●●Enlace al texto completo (gratis o de pago) [1007/s00268-013-2037-](http://1007/s00268-013-2037-2)

[2](#)

AUTORES / AUTHORS: - Crona J; Granberg D; Norlen O; Warnberg F; Stalberg P; Hellman P; Bjorklund P

INSTITUCIÓN / INSTITUTION: - Department of Surgical Sciences, Uppsala University, 751 85, Uppsala, Sweden, joakim.crona@surgsci.uu.se.

RESUMEN / SUMMARY: - BACKGROUND: Metastases from neuroendocrine tumors (NETs) to the breast have been described as a rare phenomenon. Presentation, imaging results, and cytopathologic findings of these tumours may closely mimic those of a mammary carcinoma. METHODS: This study was a retrospective review of 661 patients with metastatic NETs, of whom 280 were females, treated at Uppsala University Hospital, Uppsala, Sweden. Patients with pathological breast lesions were identified. Histopathological slides from available NET breast lesions were analyzed for mammary carcinoma and neuroendocrine markers. RESULTS: We have identified 20 female patients with NET metastases to the breast, 11/235 with small intestinal NETs, 8/55 with lung NETs, and 1/6 with thymic NETs. There were no male patients with NET metastatic to the breast. Four patients had their breast lesion initially diagnosed as mammary carcinoma. Retrospectively, these lesions showed negative staining for mammary carcinoma markers. CONCLUSIONS: Metastases to the breast from neuroendocrine tumors may be more common than previously thought. Patients with a lesion to the breast and symptoms typical for NET may

benefit from additional histopathological investigation, because NET metastases and mammary carcinoma have different immunohistochemical profiles.

[82]

TÍTULO / TITLE: - Ultrasonography-guided core needle biopsy for the thyroid nodule: does the procedure hold any benefit for the diagnosis when fine-needle aspiration cytology analysis shows inconclusive results?

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Br J Radiol. 2013 May;86(1025):20130007. doi: 10.1259/bjr.20130007.

●●Enlace al texto completo (gratis o de pago) [1259/bjr.20130007](#)

AUTORES / AUTHORS: - Hahn SY; Shin JH; Han BK; Ko EY; Ko ES

INSTITUCIÓN / INSTITUTION: - Department of Radiology, Samsung Medical Center, Sungkyunkwan University School of Medicine, Gangnam-gu, Seoul, Republic of Korea.

RESUMEN / SUMMARY: - **OBJECTIVE:** We evaluated the diagnostic role of ultrasonography-guided core needle biopsy (CNB) according to ultrasonography features of thyroid nodules that had inconclusive ultrasonography-guided fine-needle aspiration (FNA) results. **METHODS:** A total of 88 thyroid nodules in 88 patients who underwent ultrasonography-guided CNB because of previous inconclusive FNA results were evaluated. The patients were classified into three groups based on ultrasonography findings: Group A, which was suspicious for papillary thyroid carcinoma (PTC); Group B, which was suspicious for follicular (Hurthle cell) neoplasm; and Group C, which was suspicious for lymphoma. The final diagnoses of the thyroid nodules were determined by surgical confirmation or follow-up after ultrasonography-guided CNB. **RESULTS:** Of the 88 nodules, the malignant rate was 49.1% in Group A, 12.0% in Group B and 90.0% in Group C. The rates of conclusive ultrasonography-guided CNB results after previous incomplete ultrasonography-guided FNA results were 96.2% in Group A, 64.0% in Group B and 90.0% in Group C ($p=0.001$). 12 cases with inconclusive ultrasonography-guided CNB results were finally diagnosed as 8 benign lesions, 3 PTCs and 1 lymphoma. The number of previous ultrasonography-guided FNA biopsies was not significantly different between the conclusive and the inconclusive result groups of ultrasonography-guided CNB ($p=0.205$). **CONCLUSION:** Ultrasonography-guided CNB has benefit for the diagnosis of thyroid nodules with inconclusive ultrasonography-guided FNA results. However, it is still not helpful for the differential diagnosis in 36% of nodules that are suspicious for follicular neoplasm seen on ultrasonography. **Advances in knowledge:** This study shows the diagnostic contribution of ultrasonography-guided CNB as an alternative to repeat ultrasonography-guided FNA or surgery.

[83]

TÍTULO / TITLE: - Repeatability of Gallium-68 DOTATOC Positron Emission Tomographic Imaging in Neuroendocrine Tumors.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Pancreas. 2013 Apr 12.

●●Enlace al texto completo (gratis o de pago)

[1097/MPA.0b013e318287ce21](#)

AUTORES / AUTHORS: - Menda Y; Ponto LL; Schultz MK; Zamba GK; Watkins GL; Bushnell DL; Madsen MT; Sunderland JJ; Graham MM; O'Dorisio TM; O'Dorisio MS

INSTITUCIÓN / INSTITUTION: - From the *Department of Radiology, University of Iowa Carver College of Medicine; daggerDepartment of Biostatistics, University of Iowa College of Public Health; Departments of double daggerInternal Medicine, and section signPediatrics, University of Iowa Carver College of Medicine, Iowa City, IA.

RESUMEN / SUMMARY: - **OBJECTIVE:** To evaluate the repeatability of gallium-68 1,4,7,10-tetraazacyclododecane-N,N',N'',N'''-tetraacetic (DOTA)-D-Phe-Try-octreotide (Ga-DOTATOC) positron emission tomography (PET) in neuroendocrine tumors. **METHODS:** Five patients with neuroendocrine tumors were imaged with Ga-DOTATOC PET twice within 5 days. Maximum and mean standardized uptake values (SUVmax and SUVmean) and kinetic parameters (K-Patlak and K-influx) of target lesions were measured. The repeatability of these measurements was investigated. **RESULTS:** Forty-seven target lesions were identified on whole-body PET and 21 lesions on dynamic images. There was excellent repeatability with intraclass correlation coefficient of 0.99 for SUVmax, SUVmean, and K-Patlak, and 0.85 for K-influx. The median absolute percent differences and the interquartile ranges (IQR) between 2 scans for SUVmax and SUVmean were 7.4% (IQR, 14.1%) and 9.3% (IQR, 10.6%), respectively. The median absolute percent differences for K-Patlak and K-influx were 12.5% (IQR, 12.6%) and 29.9% (IQR, 22.4%), respectively. The SUVmax of target lesions did not differ by more than 25% between the 2 scans. **CONCLUSIONS:** Ga-DOTATOC PET imaging of neuroendocrine tumors is highly reproducible. A difference of more than 25% in SUVmax represents a change that is larger than the measurement error observed on repeated studies and should reflect a significant change in the biological character of the tumor.

[84]

TÍTULO / TITLE: - Neuroendocrine tumours—challenges in the diagnosis and classification of pulmonary neuroendocrine tumours.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - J Clin Pathol. 2013 May 17.

●●Enlace al texto completo (gratuito o de pago) 1136/jclinpath-2012-201310

AUTORES / AUTHORS: - den Bakker MA; Thunnissen FB

INSTITUCIÓN / INSTITUTION: - Department of Pathology, Maasstad Hospital, Rotterdam, The Netherlands.

RESUMEN / SUMMARY: - Pulmonary neuroendocrine (NE) proliferations are a diverse group of disorders which share distinct cytological, architectural and biosynthetic features. Tumours composed of NE cells are dispersed among different tumour categories in the WHO classification of tumours and as such do not conform to a singular group with regards to treatment and prognosis. This is reflected by the highly variable behaviour of NE proliferations, ranging from asymptomatic, for instance in diffuse idiopathic pulmonary NE cell hyperplasia and tumourlets, to highly malignant cancers such as small cell lung cancer and large cell NE carcinoma. In this review NE proliferations are described as distinct entities ranging from low grade lesions to high grade cancers. The differential diagnoses are considered with each of the entries. Finally, mention is made of tumours which may show some NE features.

[85]

TÍTULO / TITLE: - Ki-67 is a reliable pathological grading marker for neuroendocrine tumors.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Virchows Arch. 2013 May;462(5):501-5. doi: 10.1007/s00428-013-1410-8. Epub 2013 Apr 16.

●●Enlace al texto completo (gratuito o de pago) 1007/s00428-013-1410-8

AUTORES / AUTHORS: - Nadler A; Cukier M; Rowsell C; Kamali S; Feinberg Y; Singh S; Law CH

INSTITUCIÓN / INSTITUTION: - Division of General Surgery, University of Toronto, Toronto, Ontario, Canada.

RESUMEN / SUMMARY: - In neuroendocrine tumors (NETs), proliferation markers, especially Ki-67, have become increasingly important. This study was designed to examine the reproducibility of Ki-67 for use in the current classification of NETs. A retrospectively assembled integrated database with prospectively collected data of patients undergoing multidisciplinary management for NETs from 2000 to 2009 was analyzed. Original pathology was reviewed to reassess Ki-67 values. Ki-67 was then categorized to grades G1 ($\leq 2\%$), G2 (3-20%), or G3 ($>20\%$) according to the European Neuroendocrine Tumor Society (ENETS) guidelines and the 2010 World Health Organization (WHO) classification. Original Ki-67 values were compared to reviewed values. All statistical analyses were carried out using SAS 9.1.3. A total of 184 patients were included of which 48% were male. The most common primary NET site

was the small bowel, in 27 %. On pathology review, there was 94 % agreement for G1, with 4 % of cases upgraded at review to G2 and 2 % of cases upgraded to G3. For G2, there was 94 % agreement, with 6 % of cases downgraded to G1 and 0 % upgraded. For G3, there was 90 % agreement, with 10 % of cases downgraded to G2 and none to G1 (kappa = 0.89). Ki-67 is a proliferative marker for NETs that is highly reproducible when used to grade tumors according to ENETS and WHO categories. The high inter-institutional reliability in the determination of tumor grade as assessed by Ki-67 makes it a reliable tool in the assessment of patients with NETs.

[86]

TÍTULO / TITLE: - An unusual case of a calcified carcinoid tumour.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Eur J Cardiothorac Surg. 2013 May 14.

●●Enlace al texto completo (gratis o de pago) [1093/ejcts/ezt262](#)

AUTORES / AUTHORS: - Meda S; Pernazza F; Di Stasio M

INSTITUCIÓN / INSTITUTION: - Division of Thoracic Surgery, Santi Antonio e Biagio e Cesare Arrigo Hospital, Alessandria, Italy.

[87]

TÍTULO / TITLE: - PAX-8 Expression in Primary and Metastatic Merkel Cell Carcinoma: An Immunohistochemical Analysis.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Am J Dermatopathol. 2013 Jun;35(4):448-51. doi: 10.1097/DAD.0b013e318271ce53.

●●Enlace al texto completo (gratis o de pago)

[1097/DAD.0b013e318271ce53](#)

AUTORES / AUTHORS: - Sangoi AR; Cassarino DS

INSTITUCIÓN / INSTITUTION: - *Department of Pathology, El Camino Hospital, Mountain View, CA daggerDepartment of Pathology, Southern California Permanente Medical Group, Los Angeles, CA.

RESUMEN / SUMMARY: - : PAX-8, a nephric cell lineage transcription factor initially characterized in renal cell carcinomas, is also well recognized as a marker of Mullerian tract and thyroid tumors. From a previous tissue microarray study of nonrenal neoplasms including a variety of skin tumors, we identified PAX-8 staining in a small set of Merkel cell carcinomas, a finding not previously described. Herein, we explore PAX-8 immunoreactivity in 34 whole-section Merkel cell carcinomas (24 primary, 10 metastatic) using polyclonal PAX-8 (prediluted) and 2 varieties of monoclonal PAX-8 (prediluted clone MRQ-50 and 1:100 dilution clone BC12). Nuclear staining intensity and extent was semiquantitatively analyzed with a comparison of staining thresholds required

for a “positive” result ($\geq 2+$ vs. $1+$). Thirty-three of 34 (97%) cases showed positive Cell Marque polyclonal PAX-8 staining, whereas 31 of 34 (91%) cases showed positive Cell Marque monoclonal PAX-8 staining. There was no significant difference in staining between primary versus metastatic tumors. The Cell Marque polyclonal PAX-8 was superior to their monoclonal PAX-8, maintaining strong sensitivity using a $\geq 2+$ versus $1+$ staining cut point for positive results (79% vs. 18%, respectively), which may be important in cases with scant tissue or background staining. The Biocare monoclonal PAX-8 was negative in all cases. PAX-8 staining in Merkel cell carcinoma expands the spectrum of tumors showing immunoreactivity and may prove to be a useful addition to a diagnostic panel. Awareness of this immunoreactivity and recognition of the antibody source and clone are important to preclude diagnostic pitfalls with tumors in the differential diagnosis.

[88]

TÍTULO / TITLE: - Utility of the Quantitative Ki-67 Proliferation Index and CD56 Together in the Cytologic Diagnosis of Small Cell Lung Carcinoma and Other Lung Neuroendocrine Tumors.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Acta Cytol. 2013;57(3):281-90. doi: 10.1159/000346394. Epub 2013 Apr 25.

●●Enlace al texto completo (gratis o de pago) [1159/000346394](#)

AUTORES / AUTHORS: - Zheng G; Ettinger DS; Maleki Z

INSTITUCIÓN / INSTITUTION: - Department of Pathology, The Johns Hopkins Hospital, Baltimore, Md., USA.

RESUMEN / SUMMARY: - Background: Distinction of small cell lung carcinoma (SCLC) from non-small cell lung carcinoma (NSCLC) is critical because of the differences in prognosis and management. Patients with SCLC usually present with distant metastasis, and clinicians demand an accurate diagnosis in order to initiate appropriate therapy. Limited cytology material, occasionally with crush artifact, is not uncommon. Therefore, robust cytomorphologic features and a small immunostaining panel would be ideal to differentiate SCLC from NSCLC and other neuroendocrine neoplasms. We evaluated CD56 and the quantitative Ki-67 immunohistochemical panel in comparison to synaptophysin and chromogranin, along with cytomorphology to diagnose SCLC. Design: Eighty-eight cases of SCLC were retrieved from the cytology archives of The Johns Hopkins Hospital. Forty neuroendocrine neoplasms were used as control cases. Results: SCLCs included 33 lung cases and 55 metastatic lesions. The specimens were obtained by fine needle aspiration, thoracocentesis, bronchoalveolar lavage and abdominal paracentesis. CD56 was expressed in 98.9% of SCLCs, which is significantly more sensitive than synaptophysin and chromogranin. The Ki-67 labeling index was high ($>70\%$) in all cases, which is a

reliable marker to differentiate SCLC from other neuroendocrine neoplasms and NSCLC. Conclusion: CD56 and quantitative Ki-67 along with cytomorphology is a robust immunohistochemical panel to differentiate SCLC from other neuroendocrine neoplasms and NSCLC.

[89]

TÍTULO / TITLE: - Laparoscopic resection of primary midgut carcinoid tumors.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Surg Endosc. 2013 Apr 10.

●●Enlace al texto completo (gratis o de pago) [1007/s00464-013-2944-](http://1007/s00464-013-2944-1)

[1](#)

AUTORES / AUTHORS: - Reissman P; Shmailov S; Grozinsky-Glasberg S; Gross DJ

INSTITUCIÓN / INSTITUTION: - Department of Surgery, Shaare-Zedek Medical Center, The Hebrew University School of Medicine, P.O. Box 3235, Jerusalem, Israel, reissman@szmc.org.il.

RESUMEN / SUMMARY: - BACKGROUND: Laparoscopic intestinal surgery is the preferable technique for the majority of intestinal surgical disorders. However, no series on laparoscopic resection of intestinal midgut carcinoid tumors (MCTs) has been reported to date. This is related to the rarity of these tumors as well as the technical difficulties resecting the large mesenteric root lymph node mass commonly found with these tumors and the occasional difficulty identifying the primary MCT, which may be small and undetected on preoperative imaging studies. This is the first series to report the results for laparoscopic resection of MCT. METHODS: All consecutive patients with MCT (excluding appendiceal carcinoid tumor) between 2002 and 2012 underwent laparoscopic resection. The patient's clinical data, preoperative endocrine workup, imaging studies, operative data, final histology, and outcome were recorded and analyzed. RESULTS: During the study period, 35 patients underwent surgery for primary intestinal carcinoid tumor. Of the 35 patients, 20 (12 women and 8 men ages 26-86 years) had surgery for primary MCT, and the remainder had a colorectal carcinoid tumor. In the MCT group, ten patients had liver metastases at the time of surgery. In three patients, multiple synchronous MCTs were detected intraoperatively. All the patients underwent a laparoscopic resection with en bloc resection of the corresponding mesenteric root mass. No conversion to open surgery was needed, and no major morbidity occurred. Two patients (10 %) each experienced minor morbidity with wound infection and prolonged ileus. The median hospital length of stay was 6 days (range 4-9 days). During a follow-up period of 3-96 months, no patients experienced local or regional recurrence. No distant metastases were detected during the follow-up period in any patients who had surgery with intent to cure. CONCLUSION: Although technically difficult, laparoscopic resection of primary MCTs is feasible

and safe, with the additional known significant advantages of laparoscopic surgery in general. Similar to the large-scale prospective studies that proved the oncologic safety of laparoscopic surgery for colorectal cancer, this small series showed that the laparoscopic technique also may be oncologically safe for these rare tumors.

[90]

TÍTULO / TITLE: - Gastric Neuroendocrine Carcinoma Staged and Followed With 18F-FDG PET/CT-A Report of 3 Cases.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Clin Nucl Med. 2013 Jun;38(6):447-50. doi: 10.1097/RLU.0b013e31828da1f5.

●●Enlace al texto completo (gratis o de pago)

[1097/RLU.0b013e31828da1f5](#)

AUTORES / AUTHORS: - Makis W; Ciarallo A; Hickeson M; Derbekyan V; Novales-Diaz JA; Lisbona R

INSTITUCIÓN / INSTITUTION: - From the *Department of Nuclear Medicine, Brandon Regional Health Centre, Brandon, Manitoba; and daggerDepartment of Nuclear Medicine, Royal Victoria Hospital, McGill University Health Centre, Montreal, Quebec, Canada.

RESUMEN / SUMMARY: - Gastric neuroendocrine carcinomas (NEC) are very rare, aggressive tumors of the stomach that are distinct from the more benign neuroendocrine tumors, sometimes referred to as "gastric carcinoids." We present 3 cases of gastric NEC representing various histological subtypes that were successfully staged and followed with F-FDG PET/CT, impacting therapeutic management in each case.

[91]

TÍTULO / TITLE: - Surgery for metastatic neuroendocrine tumors with occult primaries.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - J Surg Res. 2013 Apr 25. pii: S0022-4804(13)00351-X. doi: 10.1016/j.jss.2013.04.008.

●●Enlace al texto completo (gratis o de pago) [1016/j.jss.2013.04.008](#)

AUTORES / AUTHORS: - Bartlett EK; Roses RE; Gupta M; Shah PK; Shah KK; Zaheer S; Wachtel H; Kelz RR; Karakousis GC; Fraker DL

INSTITUCIÓN / INSTITUTION: - Department of Surgery, University of Pennsylvania, Philadelphia, Pennsylvania. Electronic address:

Edmund.Bartlett@uphs.upenn.edu.

RESUMEN / SUMMARY: - INTRODUCTION: Neuroendocrine tumors (NETs) frequently metastasize prior to diagnosis. Although metastases are often

identifiable on conventional imaging studies, primary tumors, particularly those in the midgut, are frequently difficult to localize preoperatively. **MATERIALS AND METHODS:** Patients with metastatic NETs with intact primaries were identified. Clinical and pathologic data were extracted from medical records. Primary tumors were classified as localized or occult based on preoperative imaging. The sensitivities and specificities of preoperative imaging modalities for identifying the primary tumors were calculated. Patient characteristics, tumor features, and survival in localized and occult cases were compared. **RESULTS:** Sixty-one patients with an intact primary tumor and metastatic disease were identified. In 28 of these patients (46%), the primary tumor could not be localized preoperatively. A median of three different preoperative imaging studies were utilized. Patients with occult primaries were more likely to have a delay (>6 mo) in surgical referral from time of onset of symptoms (57% versus 27%, $P = 0.02$). Among the 28 patients with occult primary tumors, 18 (64%) were found to have radiographic evidence of mesenteric lymphadenopathy corresponding, in all but one case, to a small bowel primary. In all but three patients (89%), the primary tumor could be identified intraoperatively. **CONCLUSION:** The primary tumor can be identified intraoperatively in a majority of patients with metastatic NETs, irrespective of preoperative localization status. Referral for surgical management should not, therefore, be influenced by the inability to localize the primary tumor.

[92]

TÍTULO / TITLE: - Merkel cell carcinoma: a primer for the radiologist.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - AJR Am J Roentgenol. 2013 Jun;200(6):1186-96. doi: 10.2214/AJR.12.9884.

●●Enlace al texto completo (gratis o de pago) [2214/AJR.12.9884](#)

AUTORES / AUTHORS: - Tirumani SH; Shinagare AB; Sakellis C; Saboo SS; Jagannathan JP; Krajewski KM; Ramaiya NH

INSTITUCIÓN / INSTITUTION: - 1 Department of Imaging, Dana Farber Cancer Institute, Harvard Medical School, 450 Brookline Ave, Boston, MA 02215.

RESUMEN / SUMMARY: - **OBJECTIVE.** Merkel cell carcinoma (MCC) is an aggressive cutaneous neuroendocrine tumor. The purpose of this review is to provide a comprehensive description of the staging, workup, treatment, and follow-up of MCC. **CONCLUSION.** Sentinel lymph node mapping and PET/CT are the cornerstones of staging of MCC. MCC is a radiosensitive tumor, and hence radiotherapy plays a major role in its management. Close follow-up with PET/CT helps in detecting recurrences of MCC.

[93]

TÍTULO / TITLE: - IgE-mediated basophil tumour necrosis factor alpha induces matrix metalloproteinase-9 from monocytes.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Allergy. 2013 May;68(5):614-20. doi: 10.1111/all.12143. Epub 2013 Apr 10.

●●Enlace al texto completo (gratis o de pago) 1111/all.12143

AUTORES / AUTHORS: - Falkencrone S; Poulsen LK; Bindslev-Jensen C; Woetmann A; Odum N; Poulsen BC; Blom L; Jensen BM; Gibbs BF; Yasinska IM; Sumbayev VV; Skov PS

INSTITUCIÓN / INSTITUTION: - Department of Dermatology and Allergy Centre, Odense University Hospital, 5000 Odense C, Denmark. sfal@reflab.dk

RESUMEN / SUMMARY: - BACKGROUND: IgE-mediated activation of mast cells has been reported to induce the release of tumour necrosis alpha (TNF-alpha), which may display autocrine effects on these cells by inducing the generation of the tissue remodelling protease matrix metalloproteinase-9 (MMP-9). While mast cells and basophils have been shown to express complementary and partially overlapping roles, it is not clear whether a similar IgE/TNF-alpha/MMP-9 axis exists in the human basophil. The purpose of this study was thus to investigate whether IgE-mediated activation of human basophils induces TNF-alpha and MMP-9 release. METHODS: Human peripheral blood mononuclear cells (PBMC), isolated basophils and monocytes were stimulated up to 21 h with anti-IgE. Mediator releases were assessed by ELISA, and surface expressions of mediators were detected by flow cytometry. Upregulation of cytokine production was detected by Western blot and polymerase chain reaction (PCR). RESULTS: IgE-mediated activation of basophils induced the synthesis and release of both TNF-alpha and MMP-9 from PBMC. In contrast, IgE-mediated activation of purified basophils induced the release and cellular expression of TNF-alpha but not MMP-9. Isolated monocytes did not release MMP-9 upon anti-IgE stimulation, but MMP-9 release was induced by stimulating monocytes with supernatants from activated basophils, and this release was inhibited by anti-TNF-alpha neutralizing antibodies. CONCLUSION: Our results strongly indicate that human basophils release TNF-alpha following IgE-dependent activation and that this cytokine subsequently stimulates MMP-9 release from monocytes. These findings support a direct involvement of basophils in inflammation as well as suggesting a role for the basophil in tissue remodelling.

[94]

TÍTULO / TITLE: - Ectopic Cushing's Syndrome Secondary to Pulmonary Carcinoid Tumor.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Ann Thorac Surg. 2013 May;95(5):1797-9. doi: 10.1016/j.athoracsur.2012.09.039.

●●Enlace al texto completo (gratis o de pago)

1016/j.athoracsur.2012.09.039

AUTORES / AUTHORS: - Hashemzadeh S; Asvadi Kermani A; Ali-Asgharzadeh A; Halimi M; Soleimani M; Ladan A

INSTITUCIÓN / INSTITUTION: - Tuberculosis and Lung Diseases Research Center, Hematology and Oncology Research Center, Tabriz University of Medical Sciences, Tabriz, Iran.

RESUMEN / SUMMARY: - Adrenocorticotrophic hormone (ACTH) overproduction within the pituitary gland or ectopically leads to hypercortisolism. In this study a case of Cushing's syndrome caused by an ectopic ACTH-secreting carcinoid tumor in lung is discussed, as are the available diagnostic procedures. The patient was a 28-year-old woman with clinical features starting about 6 months previously. The results of her biochemical tests suggested ectopic Cushing's syndrome. Full-body computed tomography revealed a single nodule in the inferior lobe of the right lung. After removal of the nodule, the patient's symptoms subsided clinically, and laboratory tests confirmed remission of the hypercortisolism.

[95]

TÍTULO / TITLE: - A case of malignant insulinoma: successful control of glycemic fluctuation by replacing octreotide injections with octreotide LAR injections.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Endocr J. 2013 May 11.

AUTORES / AUTHORS: - Okamoto M; Kishimoto M; Takahashi Y; Osame K; Noto H; Yamamoto-Honda R; Kajio H; Tokuhara M; Edamoto Y; Endo H; Igari T; Kubota K; Noda M

INSTITUCIÓN / INSTITUTION: - Department of Diabetes and Metabolic Medicine, Center Hospital, National Center for Global Health and Medicine, Tokyo 162-8655, Japan.

RESUMEN / SUMMARY: - A 73-year-old woman with malignant insulinoma was treated with 100 mug/day octreotide for unresected insulinoma and liver metastases. The daily administration of the drug induced hyperglycemia after dinner in addition to existing fasting hypoglycemia possibly because this drug suppressed both insulin and glucagon secretion and its blood concentration was unstable. After replacing a daily injection of octreotide with a monthly injection of octreotide long-acting repeatable (LAR), blood glucose levels stabilized within the normal range. The findings of the present study showed that octreotide LAR could be useful for the long-term treatment of unresectable insulinomas.

[96]

TÍTULO / TITLE: - Pheochromocytoma-induced takotsubo-like cardiomyopathy and global heart failure with need for extracorporeal life support.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Intensive Care Med. 2013 May 14.

●●Enlace al texto completo (gratis o de pago) [1007/s00134-013-2942-](#)

[8](#)

AUTORES / AUTHORS: - Kaese S; Schulke C; Fischer D; Lebiez P

INSTITUCIÓN / INSTITUTION: - Division of Electrophysiology, Department of Cardiovascular Medicine, University of Muenster, Albert-Schweitzer-Campus 1, A1, 48149, Muenster, Germany, Sven.Kaese@ukmuenster.de.

[97]

TÍTULO / TITLE: - Accuracy of visual assessments of proliferation indices in gastroenteropancreatic neuroendocrine tumours.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - J Clin Pathol. 2013 May 23.

●●Enlace al texto completo (gratis o de pago) [1136/jclinpath-2012-](#)

[201217](#)

AUTORES / AUTHORS: - Young HT; Carr NJ; Green B; Tilley C; Bhargava V; Pearce N

INSTITUCIÓN / INSTITUTION: - Cellular Pathology, University Hospital Southampton, Southampton, UK.

RESUMEN / SUMMARY: - AIMS: To compare the accuracy of eyeball estimates of the Ki-67 proliferation index (PI) with formal counting of 2000 cells as recommend by the Royal College of Pathologists. METHODS: Sections from gastroenteropancreatic neuroendocrine tumours were immunostained for Ki-67. PI was calculated using three methods: (1) a manual tally count of 2000 cells from the area of highest nuclear labelling using a microscope eyepiece graticule; (2) eyeball estimates made by four pathologists within the same area of highest nuclear labelling; and (3) image analysis of microscope photographs taken from this area using the ImageJ 'cell counter' tool. ImageJ analysis was considered the gold standard for comparison. RESULTS: Levels of agreement between methods were evaluated using Bland-Altman plots. Agreement between the manual tally and ImageJ assessments was very high at low PIs. Agreement between eyeball assessments and ImageJ analysis varied between pathologists. Where data for low PIs alone were analysed, there was a moderate level of agreement between pathologists' estimates and the gold standard, but when all data were included, agreement was poor. CONCLUSIONS: Manual tally counts of 2000 cells exhibited similar levels of accuracy to the gold standard, especially at low PIs. Eyeball estimates were

significantly less accurate than the gold standard. This suggests that tumour grades may be misclassified by eyeballing and that formal tally counting of positive cells produces more reliable results. Further studies are needed to identify accurate clinically appropriate ways of calculating.

[98]

TÍTULO / TITLE: - 18F-Radiolabeled GLP-1 analog exendin-4 for PET/CT imaging of insulinoma in small animals.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Nucl Med Commun. 2013 Jul;34(7):701-8. doi: 10.1097/MNM.0b013e3283614187.

●●Enlace al texto completo (gratis o de pago)

[1097/MNM.0b013e3283614187](#)

AUTORES / AUTHORS: - Wu H; Liang S; Liu S; Pan Y; Cheng D; Zhang Y

INSTITUCIÓN / INSTITUTION: - aDepartment of Nuclear Medicine, School of Medicine, Ruijin Hospital, Shanghai Jiao Tong University bDepartment of Nuclear Medicine, Zhongshan Hospital, Fudan University, Shanghai, China.

RESUMEN / SUMMARY: - BACKGROUND: Insulinoma is a neuroendocrine tumor derived from the beta cells of pancreatic islets. They are usually relatively inaccessible for surgical intervention. High expression levels of glucagon-like peptide-1 (GLP-1) receptor have been detected in insulinoma. AIM: The aim of the study was to evaluate the potential of F-radiolabeled GLP-1 analog exendin-4 for the diagnosis of insulinoma using PET/computed tomography imaging. MATERIALS AND METHODS: The GLP-1 receptor-specific molecular probe [F]FB-exendin-4 was prepared by the conjugation of exendin-4 and N-succinimidyl-4-[F] fluorobenzoate ([F]SFB). High expression of GLP-1 by the RIN-m5f insulinoma line and GLP-1 receptor specificity were evaluated by determining the saturation curve for in-vitro binding of I-radiolabeled exendin-4 and by investigation of the competitive binding between I-radiolabeled and unlabeled exendin-4. Further, the in-vivo biodistribution and micro-PET/computed tomography images of insulinoma-bearing mice were studied. RESULTS: An overall radiochemical yield of 35.6±2.3% (decay corrected, n=5) and specific radioactivity of around 30 GBq/micromol were achieved for [F]FB-exendin-4, and the radiochemical purity was over 98%. Both in-vitro and in-vivo studies confirmed the specificity of [F]FB-exendin-4 to insulinoma cells. CONCLUSION: [F]FB-exendin-4 has been found to be an effective molecular imaging probe for detecting insulinomas.

[99]

TÍTULO / TITLE: - Immunohistochemical Characterization of Neuroendocrine Differentiation of Canine Anal Sac Glandular Tumours.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - J Comp Pathol. 2013 Apr 10. pii: S0021-9975(13)00030-3. doi: 10.1016/j.jcpa.2013.01.013.

●●Enlace al texto completo (gratis o de pago) [1016/j.jcpa.2013.01.013](#)

AUTORES / AUTHORS: - Suzuki K; Morita R; Hojo Y; Nomura K; Shibutani M; Mitsumori K

INSTITUCIÓN / INSTITUTION: - Laboratory of Veterinary Toxicology, Tokyo University of Agriculture and Technology, Tokyo, Japan. Electronic address: kzsuzuki@cc.tuat.ac.jp.

RESUMEN / SUMMARY: - Histological features and expression of neuroendocrine markers were examined in 69 samples of canine anal sac glandular carcinomas (ASGCs). The tumours were classified into solid, rosette and tubular types and mixtures of these types. Tumour-associated death in dogs with solid tumours and mixed tumours with solid components was higher than in dogs with rosette and tubular type tumours. Chromogranin A immunoreactivity was observed in 28 of 69 samples (40.6%) irrespective of histological type and was localized to the marginal areas of the tumour nest and the basal areas of the tubular and rosette structures. Neuron-specific enolase immunoreactivity in neoplastic epithelial cells was observed in 32 cases (46.4%) and was less frequently observed in the tubular type (14.3%). Synaptophysin expression was present in 15.9% of cases and was least frequent in the tubular type. Twenty-one of the 69 samples expressed more than two neuroendocrine markers and were classified as carcinomas with neuroendocrine differentiation. There was no relationship between neuroendocrine differentiation and clinical outcome. These results suggest that some ASGCs have neuroendocrine differentiation regardless of histological pattern, but clinical outcome is more related to the histological pattern than to neuroendocrine differentiation.

[100]

TÍTULO / TITLE: - Presentation, diagnostic features and glucose handling in a monocentric series of insulinoma.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - J Endocrinol Invest. 2013 Apr 18.

●●Enlace al texto completo (gratis o de pago) [3275/8942](#)

AUTORES / AUTHORS: - Toiari M; Davi MV; Dalle Carbonare L; Boninsegna L; Castellani C; Falconi M; Francia G

INSTITUCIÓN / INSTITUTION: - Department of Internal Medicine D, "G.B. Rossi" University Hospital, Verona, Italy.

RESUMEN / SUMMARY: - Background: new aspects have emerged in the clinical and diagnostic scenarios of insulinoma: current guidelines have lowered the diagnostic insulin threshold to 3FU/ml in the presence of hypoglycemia (<55 mg/dL); post-prandial hypoglycemia has been reported as the only presenting

symptom; preexisting diabetes mellitus was recognized in some patients. Aim: to evaluate clinical features, diagnostic criteria and glucose metabolic profile in a monocentric series of patients affected by insulinomas including two subgroups: sporadic and MEN-1. Subjects and methods: clinical, pathological and biochemical data regarding 33 patients were analyzed. Results: following the current guidelines the 72h-fasting test was initially positive in all cases but one. In this case the test, initially negative, became positive after a 2-year follow-up. Nadir insulin level was ≥ 3 FU/mL but < 6 FU/mL in three patients and ≥ 6 FU/mL in the remaining 30 cases. At presentation 27 patients (82%) reported only fasting symptoms, 3 (9%) only post-prandial and 3 (9%) both. Seven cases (21%) had previously been affected by type 2 diabetes mellitus or impaired glucose metabolism. Conclusions: in our series the new cut-off of insulin increased the sensitivity of the 72h-fasting test from 87% to 97%. The absence of hypoglycemia during the test cannot definitively rule out the diagnosis and the test should be repeated in every highly suspicious case. Post-prandial hypoglycaemia can be the only presenting symptom. Diabetes mellitus may be associated with the occurrence of insulinoma. A possible diagnosis of insulinoma must not be ignored if previous impaired glucose handling is evident.

[101]

TÍTULO / TITLE: - Interobserver agreement of proliferation index (Ki-67) outperforms mitotic count in pulmonary carcinoids.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Virchows Arch. 2013 May;462(5):507-13. doi: 10.1007/s00428-013-1408-2. Epub 2013 Apr 5.

●●Enlace al texto completo (gratis o de pago) [1007/s00428-013-1408-](#)

[2](#)

AUTORES / AUTHORS: - Warth A; Fink L; Fisseler-Eckhoff A; Jonigk D; Keller M; Ott G; Rieker RJ; Sinn P; Soder S; Soltermann A; Willenbrock K; Weichert W

INSTITUCIÓN / INSTITUTION: - Institute of Pathology, University Hospital Heidelberg, Im Neuenheimer Feld 224, 69120, Heidelberg, Germany, arne.warth@med.uni-heidelberg.de.

RESUMEN / SUMMARY: - Evaluation of proliferative activity is a cornerstone in the classification of endocrine tumors; in pulmonary carcinoids, the mitotic count delineates typical carcinoid (TC) from atypical carcinoid (AC). Data on the reproducibility of manual mitotic counting and other methods of proliferation index evaluation in this tumor entity are sparse. Nine experienced pulmonary pathologists evaluated 20 carcinoid tumors for mitotic count (hematoxylin and eosin) and Ki-67 index. In addition, Ki-67 index was automatically evaluated with a software-based algorithm. Results were compared with respect to correlation coefficients (CC) and kappa values for clinically relevant grouping

algorithms. Evaluation of mitotic activity resulted in a low interobserver agreement with a median CC of 0.196 and a median kappa of 0.213 for the delineation of TC from AC. The median CC for hotspot (0.658) and overall (0.746) Ki-67 evaluation was considerably higher. However, kappa values for grouped comparisons of overall Ki-67 were only fair (median 0.323). The agreement of manual and automated Ki-67 evaluation was good (median CC 0.851, median kappa 0.805) and was further increased when more than one participant evaluated a given case. Ki-67 staining clearly outperforms mitotic count with respect to interobserver agreement in pulmonary carcinoids, with the latter having an unacceptable low performance status. Manual evaluation of Ki-67 is reliable, and consistency further increases with more than one evaluator per case. Although the prognostic value needs further validation, Ki-67 might perspectivevely be considered a helpful diagnostic parameter to optimize the separation of TC from AC.

[102]

TÍTULO / TITLE: - Neuroendocrine Tumour at the Carina of a Dog.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - J Comp Pathol. 2013 Apr 11. pii: S0021-9975(13)00027-3. doi: 10.1016/j.jcpa.2013.01.010.

●●Enlace al texto completo (gratis o de pago) 1016/j.jcpa.2013.01.010

AUTORES / AUTHORS: - Gould EN; Johnson LR; Traslavina RP; Mohr FC

INSTITUCIÓN / INSTITUTION: - William R. Pritchard Veterinary Medical Teaching Hospital, University of California at Davis, Davis, CA, USA.

RESUMEN / SUMMARY: - A 10-year-old, neutered female, crossbred pit bull terrier was presented for cough, haemoptysis and rapidly progressive respiratory difficulty. Thoracic radiographs suggested a soft tissue density at the carina and bronchoscopy revealed a large, broad-based mass obstructing the entire left mainstem bronchus and half of the entrance to the right mainstem bronchus. Microscopically, the mass consisted of neoplastic cells that were packeted into small nests and had strong granular cytoplasmic immunoreactivity to synaptophysin and chromogranin A. Cytoplasmic neurosecretory granules stained strongly by the Grimelius method. A diagnosis of obstructive neuroendocrine tumour was made.

[103]

- CASTELLANO -

TÍTULO / TITLE: Carcinoma neuroendocrino de oído medio.

TÍTULO / TITLE: - Middle-ear neuroendocrine carcinoma.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Acta Otorrinolaringol Esp. 2013 May 24. pii: S0001-6519(13)00027-7. doi: 10.1016/j.otorri.2012.12.002.

●●Enlace al texto completo (gratis o de pago)

1016/j.otorri.2012.12.002

AUTORES / AUTHORS: - Martínez Subías JJ; El Uali M; Del Rey FJ; Del Valle M

INSTITUCIÓN / INSTITUTION: - Servicio de Otorrinolaringología, Hospital Santa Bárbara, Soria, España. Electronic address: jmartinezs@hotm.com.

[104]

TÍTULO / TITLE: - Hybrid oncocytic/chromophobe renal cell tumours do not display genomic features of chromophobe renal cell carcinomas.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Virchows Arch. 2013 May 25.

●●Enlace al texto completo (gratis o de pago) [1007/s00428-013-1422-](http://1007/s00428-013-1422-4)

[4](#)

AUTORES / AUTHORS: - Pote N; Vieillefond A; Couturier J; Arrufat S; Metzger I; Delongchamps NB; Camparo P; Mege-Lechevallier F; Molinie V; Sibony M

INSTITUCIÓN / INSTITUTION: - Department of Pathology, Cochin Hospital, Assistance Publique Hopitaux de Paris, Paris Descartes University, Paris, France.

RESUMEN / SUMMARY: - Hybrid oncocytic/chromophobe tumours (HOCT) are renal tumours recently described displaying histological features of both renal oncocytoma (RO) and chromophobe renal cell carcinoma (ChRCC), raising the question of their precise signification in the RO/ChRCC group. This study aimed to describe clinicopathological features of so called HOCT and to characterise their genomic profile. Five hundred and eighty-three tumours belonging to the ChRCC/RO group were retrospectively reviewed. Twelve tumours that could not be classified as RO or CHRC were considered as HOCT. Hale staining and cytokeratin 7 (CK7) immunostaining were performed. Genomic profile was established by array comparative genomic hybridisation (array-CGH) on frozen samples. Mean age at diagnosis was 70 years (range 46-83). No recurrence was observed (median follow-up: 18 months; range 9-72). Tumour size ranged from 1 to 11 cm. HOCT showed an admixture of RO- and ChRCC-like areas and/or "hybrid" cells with overlapping cytonuclear and/or histochemical features. Hale staining was apical in 50 to 100 % of cells, and CK7 was expressed in 10 to 100 % of cells. Genomic profile was balanced in seven cases or showed a limited number of random imbalances in five cases, as observed in RO. In no instances were observed the characteristic chromosome losses of ChRCC. These results suggest that so called HOCT are not true hybrid tumours and rather could represent a morphological variant of RO. From a diagnostic perspective, an array-CGH analysis could be performed in ambiguous ChRCC/RO cases to formally exclude the diagnosis of ChRCC.

[105]

TÍTULO / TITLE: - Neuroendocrine neoplasms of the jejunum: a heterogeneous group with distinctive proximal and distal subsets.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Virchows Arch. 2013 May;462(5):489-99. doi: 10.1007/s00428-013-1411-7. Epub 2013 Apr 12.

●●Enlace al texto completo (gratis o de pago) [1007/s00428-013-1411-](#)

[7](#)

AUTORES / AUTHORS: - Chopin-Laly X; Walter T; Hervieu V; Poncet G; Adham M; Guibal A; Chayvialle JA; Lombard-Bohas C; Scoazec JY

INSTITUCIÓN / INSTITUTION: - Service de Chirurgie Digestive, Hospices Civils de Lyon, Hopital Edouard Herriot, 69437, Lyon Cedex 03, France.

RESUMEN / SUMMARY: - Neuroendocrine tumors (NETs) of the jejunum are rare and usually grouped with either duodenal or ileal NETs. We aimed at better evaluating their characteristics by studying 116 cases of small-bowel NETs for which a precise anatomical location was available. Thirty-four cases were duodenal. Eighty-two were located after the duodenojejunal ligament, including ten cases in the first 50 cm, four cases between 50 and 100 cm, and six cases between 100 and 250 cm. All tumors located after 50 cm from the duodenojejunal ligament were enterochromaffin neoplasms. In contrast, the ten tumors located before this point formed a heterogeneous group. They included two cases of gastrin-expressing tumors in the first 10 cm and one case of enterochromaffin tumor located at 45 cm. The seven remaining cases were large tumors, located between 10 and 50 cm, of intermediate or high histological grade (four out of seven G2 or G3), locally invasive and usually metastatic (five out of seven with liver metastases); their survival was comparable to that of duodenal NETs. Patients with tumors located in the duodenum or the first 50 cm of the jejunum had longer survivals than those with lower jejunal and ileal tumors ($p = 0.024$). In conclusion, our study underlines the heterogeneity of jejunal NETs and supports the distinction between “upper” and “lower” jejunal tumors, which, for prognostic purposes, might be grouped with, respectively, duodenal and ileal NETs. Our data suggest that the arbitrary limit between upper and lower jejunal tumors might be fixed at 50 cm from the duodenojejunal ligament.

[106]

TÍTULO / TITLE: - Expression of CD133 in Neuroendocrine Neoplasms of the Digestive Tract: A Detailed Immunohistochemical Analysis.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Tohoku J Exp Med. 2013;229(4):301-9.

AUTORES / AUTHORS: - Mia-Jan K; Munkhdelger J; Lee MR; Ji SY; Kang TY; Choi E; Cho MY

INSTITUCIÓN / INSTITUTION: - Department of pathology, Yonsei University Wonju College of Medicine.

RESUMEN / SUMMARY: - Gastroenteropancreatic neuroendocrine tumors (GEP-NETs) are potentially malignant with variable biologic behavior that originates from neuroendocrine cells of digestive tract. Recently, the existence of cancer stem cells (CSC) was demonstrated in tumors of gastrointestinal tract. CD133 is a transmembrane glycoprotein that serves as a CSC marker in various malignancies. However, the expression of CD133 in neuroendocrine neoplasms (NEN) of digestive tract has not been studied. We evaluated tissue expression of CD133 by immunohistochemistry in 90 NENs of digestive tract with their matched non-neoplastic mucosa including stomach (n = 15), small intestine (n = 7), appendix (n = 3), colon (n = 8), rectum (n = 41), pancreas (n = 2), gallbladder (n = 4) and liver (n = 10). Tumors were divided according to 2010 WHO classification. CD133 was expressed in 30.3% (17/56) of well-differentiated neuroendocrine tumors (NET), 26.1% (6/23) of poorly-differentiated neuroendocrine carcinomas (NEC) and 63.6% (7/11) of mixed adenoneuroendocrine carcinoma (MANECs). MANEC refers to existence of both adenocarcinoma and NEC together, each one comprising at least 30% of the tumor. CD133 was expressed in cytoplasm, luminal-side of cell membrane, or both and the staining pattern correlated with tumor growth pattern. CD133 expression was not significantly correlated with tumor grade, site, expression of neuroendocrine markers (chromogranin-A and synaptophysin) and patients' survival. Thus, CD133 expression may lack prognostic significance in GEP-NETs. Importantly, CD133 was not detectable in non-neoplastic neuroendocrine cells of digestive system including pancreatic islets. In conclusion, CD133 is expressed in poorly-differentiated NECs and well-differentiated NETs of the digestive tract.

[107]

TÍTULO / TITLE: - Radiotherapy in the Management of Pancreatic Neuroendocrine Tumors (PNET): Experience at Three Institutions.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Anticancer Res. 2013 May;33(5):2175-7.

AUTORES / AUTHORS: - Saif MW; Ove R; Ng J; Russo S

INSTITUCIÓN / INSTITUTION: - Tufts University School of Medicine, Division of Hematology/Oncology, Department of Medicine, Director, GI Oncology Program 800 Washington Street, Box 245, Boston, MA 02111, U.S.A. wsaif@tuftsmedicalcenter.org.

RESUMEN / SUMMARY: - Aim: Advanced pancreatic neuroendocrine tumor (PNET) presents a therapeutic challenge as many are unresectable and

relatively resistant to systemic therapy with a high malignant potential. We share our experience using concurrent capecitabine or infusional 5-fluorouracil with radiation for patients with resected and locally advanced PNET. PATIENTS AND METHODS: Six patients (two females, four males) with PNET were treated with capecitabine or infusional 5-FU and concurrent radiation. RESULTS: The median age was 52 years (range: 38 to 63 years), with ECOG Performance Status (PS) 0-1, grade 0-1 weight loss, and grade 0-1 pain. One patient underwent resection with negative margins, two with positive margins, and three had unresectable locally advanced disease. All six patients demonstrated partial radiographic response and sustained local control. The treatment was tolerable with only grade 2 hand-foot syndrome and grade 1 mucositis observed. CONCLUSION: Prospective studies to further investigate the role of chemoradiation in this setting are warranted.

[108]

TÍTULO / TITLE: - Small cell carcinoma of the colon arising in a carcinoid tumor.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Anticancer Res. 2013 Apr;33(4):1713-5.

AUTORES / AUTHORS: - Saif MW

INSTITUCIÓN / INSTITUTION: - Section of GI Cancers and Experimental Therapeutics, Tufts University School of Medicine, Boston, MA 02111, USA.
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RESUMEN / SUMMARY: - Small cell carcinomas of the gastrointestinal tract are rare and clinically aggressive tumors. A case is presented of a 70 year-old woman who presented with small bowel obstruction and was found to have a cecal mass. She underwent right hemicolectomy, and histopathology showed a small cell carcinoma arising in the background of a carcinoid tumor. Although small cell carcinomas of the colon have frequently been found in association with colonic adenomas, this appears to be the first report of a low-grade carcinoid tumor in combination with a small cell carcinoma.

[109]

TÍTULO / TITLE: - Gastric carcinoids (neuroendocrine neoplasms).

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Gastroenterol Clin North Am. 2013 Jun;42(2):381-97. doi: 10.1016/j.gtc.2013.01.009. Epub 2013 Mar 1.

●●Enlace al texto completo (gratis o de pago) 1016/j.gtc.2013.01.009

AUTORES / AUTHORS: - Kidd M; Gustafsson B; Modlin IM

INSTITUCIÓN / INSTITUTION: - Department of Surgery, Yale University School of Medicine, PO Box 208602, New Haven, CT, USA. Electronic address:
mark.kidd@yale.edu.

RESUMEN / SUMMARY: - Gastric neuroendocrine neoplasms of the stomach can be divided into the usually well-differentiated, hypergastrinemia-dependent type I and II lesions and the more aggressively behaving gastrin-independent type III lesions. Studying menin and its complex interrelationship with gastrin may provide insight into tumor biology at the clinical level and in terms of basic cell biology (eg, the role of the epigenome in neuroendocrine cell proliferation), and lead to potential consideration of other targets that are known candidates for molecular-based therapies in other adenocarcinomas.

[110]

TÍTULO / TITLE: - Hepatic metastasis of a carotid body paraganglioma 5 years after resection of the primary tumor.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Am Surg. 2013 May;79(5):194-6.

AUTORES / AUTHORS: - Moris D; Sotiropoulos G; Vernadakis S

INSTITUCIÓN / INSTITUTION: - 1st Department of Surgery Athens University School of Medicine "Laikon" General Hospital Athens, Greece.

[111]

TÍTULO / TITLE: - Overexpression of miR-10^a and miR-375 and downregulation of YAP1 in medullary thyroid carcinoma.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Exp Mol Pathol. 2013 May 16. pii: S0014-4800(13)00058-0. doi: 10.1016/j.yexmp.2013.05.001.

●●Enlace al texto completo (gratis o de pago)

1016/j.yexmp.2013.05.001

AUTORES / AUTHORS: - Hudson J; Duncavage E; Tamburrino A; Salerno P; Xi L; Raffeld M; Moley J; Chernock RD

INSTITUCIÓN / INSTITUTION: - Department of Pathology and Immunology, Washington University School of Medicine, St. Louis, MO, United States.

RESUMEN / SUMMARY: - MicroRNAs are a primordial mechanism of gene expression control that appear to be crucial to cellular development and may play an important role in tumor development. Much is known about the genetics of medullary thyroid carcinomas, as approximately 25% are hereditary and harbor germ line activating mutations in the RET gene. Somatic RET mutations are also seen in roughly 50% of sporadic medullary thyroid carcinomas. Few studies, however, have evaluated the role of microRNA expression in these tumors. DNA and RNA were extracted from formalin-fixed paraffin-embedded tissue blocks of 15 medullary thyroid carcinomas [10 with RET mutations (3 hereditary) and 5 without RET mutations] and 5 non-tumor thyroid glands. miRNA expression of 754 targets was quantitated by real-time PCR using the

ABI OpenArray miRNA assay. Three miRNAs showed significant differential expression and were validated in a larger cohort of 59 cases by real-time PCR. Expression of potential downstream targets and upstream regulators was also investigated by real-time PCR. miR-375 and miR-10^a were significantly overexpressed, while miR-455 was underexpressed in medullary thyroid carcinomas. Expression of all 3 miRNAs was validated in the larger cohort of cases (miR-375, $p=3.3 \times 10^{-26}$; miR-10^a, $p=5.6 \times 10^{-14}$; miR-455, $p=2.4 \times 10^{-4}$). No significant differences in miRNA expression were found between RET mutation positive and negative tumors nor between sporadic and hereditary tumors. Expression of the potential downstream targets of miR-375, YAP1 (a growth inhibitor) and SLC16a2 (a transporter of thyroid hormone), was downregulated in the tumors suggesting that miR-375 is a negative regulator of the expression of these genes. Thus, differential expression of miR-375, miR-10^a and miR-455 may be important for tumor development and/or reflect C-cell lineage of medullary thyroid carcinoma. Furthermore, the growth inhibitor YAP1 is identified as a potential important downstream target of miR-375.

[112]

TÍTULO / TITLE: - Gastric neuroendocrine carcinoma with non-islet cell tumor hypoglycemia associated with enhanced production of insulin-like growth factor II.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Intern Med. 2013;52(7):757-60. Epub 2013 Apr 1.

AUTORES / AUTHORS: - Ida T; Morohashi T; Ohara H; Goto T; Inamori M; Nakajima A; Maeda S; Tsukumo Y; Sakamoto A; Ishikawa Y

INSTITUCIÓN / INSTITUTION: - Department of Gastroenterology, Omori Red Cross Hospital, Japan.

RESUMEN / SUMMARY: - A 75-year-old man was admitted to the hospital with a loss of consciousness. His blood glucose level was 24 mg/dL. Abdominal computed tomography revealed multiple metastatic lesions in the liver, while upper endoscopy disclosed advanced gastric cancer. The hypoglycemia was refractory despite the administration of glucose and steroid therapy. The patient died within one month of admission. An autopsy revealed neuroendocrine-type gastric cancer, which, on examination with immunohistochemistry, was found to be negative for insulin and insulin-like growth factor I and positive for insulin-like growth factor II (IGF-II). The patient was diagnosed as having gastric cancer with non-islet cell tumor hypoglycemia (NICTH) caused by IGF-II.

[113]

TÍTULO / TITLE: - Terminal ileal carcinoid tumor without hepatic or extrahepatic metastasis causing carcinoid syndrome.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Am Surg. 2013 Apr;79(4):439-41.

AUTORES / AUTHORS: - Datta J; Merchant NB

INSTITUCIÓN / INSTITUTION: - Department of Surgery, University of Pennsylvania Health System, Philadelphia, Pennsylvania 19104, USA.

jashodeep.datta@uphs.upenn.edu

[114]

TÍTULO / TITLE: - Endoscopic mucosal resection for rectal carcinoids under micro-probe ultrasound guidance.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - World J Gastroenterol. 2013 Apr 28;19(16):2555-9. doi: 10.3748/wjg.v19.i16.2555.

●●Enlace al texto completo (gratis o de pago) [3748/wjg.v19.i16.2555](#)

AUTORES / AUTHORS: - Zhou FR; Huang LY; Wu CR

INSTITUCIÓN / INSTITUTION: - Fu-Run Zhou, Liu-Ye Huang, Cheng-Rong Wu, Department of Gastroenterology, Yu Huang Ding Hospital affiliated to Qingdao University School of Medicine, Yantai 264000, Shandong Province, China.

RESUMEN / SUMMARY: - AIM: To assess the therapeutic value of endoscopic mucosal resection (EMR) under micro-probe ultrasound guidance for rectal carcinoids less than 1 cm in diameter. METHODS: Twenty-one patients pathologically diagnosed with rectal carcinoids following colonoscopy in our hospital from January 2007 to November 2012 were included in this study. The patients consisted of 14 men and 7 women, with a mean age of 52.3 +/- 12.2 years (range: 36-72 years). The patients with submucosal tumors less than 1 cm in diameter arising from the rectal and muscularis mucosa detected by micro-probe ultrasound were treated with EMR and followed up with conventional endoscopy and micro-probe ultrasound. RESULTS: All of the 21 tumors were confirmed by micro-probe ultrasound as uniform hypoechoic masses originating from the rectal and muscularis mucosa, without invasion of muscularis propria and vessels, and less than 1 cm in diameter. EMR was successfully completed without bleeding, perforation or other complications. The resected specimens were immunohistochemically confirmed to be carcinoids. Patients were followed up for one to two years, and no tumor recurrence was reported. CONCLUSION: EMR is a safe and effective treatment for rectal carcinoids less than 1 cm in diameter.

[115]

TÍTULO / TITLE: - Hybrid Peripheral Nerve Sheath Tumors, Including a Malignant Variant in Type 1 Neurofibromatosis.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Am J Dermatopathol. 2013 May 14.

●●Enlace al texto completo (gratis o de pago)

[1097/DAD.0b013e31827e2917](https://doi.org/10.1097/DAD.0b013e31827e2917)

AUTORES / AUTHORS: - Kacerovska D; Michal M; Kuroda N; Tanaka A; Sima R; Denisjuk N; Kreuzberg B; Ricarova R; Kazakov DV

INSTITUCIÓN / INSTITUTION: - *Sikl's Department of Pathology, Charles University in Prague, Faculty of Medicine in Pilsen, Pilsen, Czech Republic
Biological Laboratory, Pilsen, Czech Republic
Department of Diagnostic Pathology, Kochi Red Cross Hospital, Kochi, Japan
Kempf und Pfaltz, Histologische Diagnostik, Zurich, Switzerland
Department of Radiology || Department of Ophthalmology, Charles University in Prague, Faculty of Medicine in Pilsen, Pilsen, Czech Republic.

RESUMEN / SUMMARY: - : The authors report a small case series of hybrid nerve sheath tumors occurring in the setting of type 1 neurofibromatosis. Four lesions were benign and consisted of plexiform neurofibromas with considerable areas of perineuriomatous differentiation in patients with type 1 neurofibromatosis. In these lesions, biphasic (Schwannian and perineuriomatous) differentiation was apparent on immunohistochemistry, with the perineuriomatous areas staining for epithelial membrane antigen, glut-1, and claudin-1 and being negative for S-100 protein. Three patients were members of a single family, with a history of various malignant neoplasms. Included in the series is 1 hybrid lesion in which neurofibromatous and perineuriomatous areas were clearly visible on hematoxylin- and eosin-stained slides. The lesion was unique in that it manifested malignant change in the S-100 protein-positive component, which was classified as malignant peripheral nerve sheath tumor. The malignant component showed areas with an epithelioid cell morphology.

[116]

TÍTULO / TITLE: - Management of pancreatic neuroendocrine tumors.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Surg Clin North Am. 2013 Jun;93(3):675-91. doi: 10.1016/j.suc.2013.02.001. Epub 2013 Apr 3.

●●Enlace al texto completo (gratis o de pago) [1016/j.suc.2013.02.001](https://doi.org/10.1016/j.suc.2013.02.001)

AUTORES / AUTHORS: - Dickson PV; Behrman SW

INSTITUCIÓN / INSTITUTION: - Division of Surgical Oncology, Department of Surgery, University of Tennessee Health Science Center, 910 Madison Avenue, Suite 208, Memphis, TN 38163, USA.

RESUMEN / SUMMARY: - Pancreatic neuroendocrine tumors account for 1% to 2% of pancreatic neoplasms and may occur sporadically or as part of a hereditary syndrome. Patients may present with symptoms related to hormone secretion by functional tumors or to locally advanced or metastatic nonfunctional tumors. Asymptomatic pancreatic neuroendocrine tumors are

increasingly detected incidentally during abdominal imaging performed for other reasons. The management of localized pancreatic neuroendocrine tumors is surgical resection. Hepatic metastases are common and their management involves a variety of liver-directed therapies, which should be tailored according to extent of disease, symptoms, presence of extrahepatic metastases, and patient performance status.

[117]

TÍTULO / TITLE: - Growth hormone-releasing hormone-producing pancreatic neuroendocrine tumor in a multiple endocrine neoplasia type 1 family with an uncommon phenotype.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Eur J Gastroenterol Hepatol. 2013 Jul;25(7):858-862.

●●Enlace al texto completo (gratis o de pago)

[1097/MEG.0b013e32835f433f](#)

AUTORES / AUTHORS: - Sala E; Ferrante E; Verrua E; Malchiodi E; Mantovani G; Filopanti M; Ferrero S; Pietrabissa A; Vanoli A; La Rosa S; Zatelli MC; Beck-Peccoz P; Verga U

INSTITUCIÓN / INSTITUTION: - aDepartment of Clinical Sciences and Community Health, Endocrinology and Diabetology Unit bDepartment of Medicine, Surgery and Dentistry, Pathology Unit, University of Milan, Fondazione IRCCS Ca Granda Ospedale Maggiore Policlinico, Milan Departments of c2nd General Surgery dPathology, University of Pavia and IRCCS Policlinico S. Matteo, Pavia eDepartment of Pathology, Ospedale di Circolo, Varese fDepartment of Biomedical Sciences and Advanced Therapies, Section of Endocrinology, University of Ferrara, Ferrara, Italy.

RESUMEN / SUMMARY: - The objective of this study was to describe a multiple endocrine neoplasia type 1 (MEN1) family characterized by primary hyperparathyroidism, in association with acromegaly because of ectopic growth hormone-releasing hormone (GHRH) secretion by a pancreatic neuroendocrine tumor in a young man and with a bronchial carcinoid in his mother. We investigate the clinical, radiological imaging, histopathologic findings, and therapy. An 18-year-old man successfully underwent subtotal parathyroidectomy for primary hyperparathyroidism. A subsequent genetic analysis showed a MEN1 gene mutation. Three years later, acromegaly because of ectopic GHRH secretion was diagnosed (pituitary MRI negative and elevated GHRH levels). A search for an ectopic tumor was unsuccessful and somatostatin analog therapy was started. Successively, scintigraphy with somatostatin analogs (68-Ga-DOTATOC-PET) showed three focal areas in the pancreatic tail. Distal pancreatectomy showed multiple pancreatic neuroendocrine tumors and hormonal status was normalized. Afterwards, the evaluation of the patient's mother, carrying the same mutation, indicated a

primary hyperparathyroidism and a 4 cm lung mass. The patient underwent subtotal pneumonectomy and the histological analysis was consistent with the diagnosis of a typical bronchial carcinoid. In conclusion, an atypical phenotype may be recorded in MEN1 families, thus emphasizing the importance of the new imaging and surgical techniques in the diagnosis and treatment of such a rare disease.

[118]

TÍTULO / TITLE: - Bronchial Carcinoid Imaged With Cardiac Gated, 128-Slice, Dual-Source, Flash CT Scanner to Direct Operative Management.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Mil Med. 2013 Apr;178(4):e508-10. doi: 10.7205/MILMED-D-12-00357.

●●Enlace al texto completo (gratis o de pago) [7205/MILMED-D-12-00357](#)

AUTORES / AUTHORS: - Marquardt J; Vachon T; Lin T; Boswell G

INSTITUCIÓN / INSTITUTION: - Marine Corps Air Station Yuma, Marine Attack Squadron 513, PO Box 99260, Yuma, AZ 85369.

RESUMEN / SUMMARY: - Carcinoid is a rare lung cancer that typically presents with a relatively indolent clinical behavior. We present the case of a 32-year-old male with progressive respiratory symptoms, which resulted in the diagnosis of typical bronchial carcinoid. This case shows a novel imaging technique for staging a bronchial carcinoid for determination of optimal management. This case also shows the multidisciplinary approach required for management of patients with carcinoid tumors.

[119]

TÍTULO / TITLE: - Pituitary metastasis of an unknown neuroendocrine breast carcinoma mimicking a pituitary adenoma.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Pathology. 2013 Jun;45(4):422-4. doi: 10.1097/PAT.0b013e328360dfd1.

●●Enlace al texto completo (gratis o de pago) [1097/PAT.0b013e328360dfd1](#)

AUTORES / AUTHORS: - Senetta R; Castellano I; Garbossa D; Sapino A; Cassoni P

INSTITUCIÓN / INSTITUTION: - *Department of Medical Sciences, University of Turin daggerDepartment of Laboratory Diagnostic AO-U San Giovanni Battista of Turin double daggerNeurochirurgia, Department of Neuroscience, University of Turin, Turin, Italy.

[120]

TÍTULO / TITLE: - Pheochromocytoma Presenting With Remote Bony Recurrence Twenty Years After Initial Surgery: Detection With 68Ga-DOTANOC PET/CT.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Clin Nucl Med. 2013 May 1.

●●Enlace al texto completo (gratis o de pago)

[1097/RLU.0b013e31828e98f0](#)

AUTORES / AUTHORS: - Parida GK; Dhull VS; Sharma P; Bal C; Kumar R

INSTITUCIÓN / INSTITUTION: - From the Department of Nuclear Medicine, All India Institute of Medical sciences, New Delhi, India.

RESUMEN / SUMMARY: - Pheochromocytomas are rare tumors which can be malignant in 10% of cases. We present the case of a 75-year-old woman who presented with headache and palpitation for 1 year. She had a past history of right adrenalectomy for pheochromocytoma 20 years back. In between, the patient was asymptomatic. Twenty-four-hour urinary vanillylmandelic acid was raised. Noncontrast CT and ultrasound of abdomen were unremarkable. The patient underwent Ga-DOTANOC PET/CT that showed metastasis to left ilium, which was confirmed on biopsy.

[121]

TÍTULO / TITLE: - Coil embolization and surgical removal of carotid body paraganglioma.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - J Craniofac Surg. 2013 May;24(3):e242-5. doi: 10.1097/SCS.0b013e31828607ef.

●●Enlace al texto completo (gratis o de pago)

[1097/SCS.0b013e31828607ef](#)

AUTORES / AUTHORS: - Cvjetko I; Erdelez L; Podvez Z; Buhin M; Vidjak V; Borovecki A; Cvjetko T; Grsic K

INSTITUCIÓN / INSTITUTION: - From the *Departments of Vascular Surgery, daggerRadiology and double daggerPathology, University Hospital Meku, Zagreb; section signPolclinic for Speech and HearingRehabilitation, Suvag, Zagreb; and parallelInstitute for Tumors, Zagreb, Croatia.

RESUMEN / SUMMARY: - Carotid body paraganglioma has considerable malignant potential and locally aggressive behavior, so it should be treated as soon as it is discovered. We report the case of 60-year-old male patient with a carotid body paraganglioma (Shamblin group II) that was causing the carotid arteries to spread. Angiography showed 1 dominant feeding artery arising from the right external carotid artery. Selective angiography was performed 2 days before surgical removal of the tumor, and the feeding artery was successfully

embolized with coils. Literature review reveals previous reports where preoperative embolization of the feeding arteries was done using ethanol, polymers, or other liquid agents. In our case, angiography (via femoral artery) was performed 2 days before surgical removal of the tumor, and the main feeding artery (a single branch arising from external carotid artery) was successfully embolized with coils rather than liquids. Performing coil embolization before operating reduced subsequent blood loss and made it easier to identify the feeding artery during surgery. Supraselective coiling, although as difficult as embolization with liquids, may reduce the incidence of postoperative stroke. At 1 year after surgery, the patient had no signs of tumor recurrence.

[122]

TÍTULO / TITLE: - Mediastinal Paraganglioma between the Great Vessels in an 81-Year-Old Woman.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Tex Heart Inst J. 2013;40(2):189-92.

AUTORES / AUTHORS: - Ghouri MA; Krishnan E; Singh A; Zaman T; Hallman CH

INSTITUCIÓN / INSTITUTION: - Texas Heart Institute at St. Luke's Episcopal Hospital, Houston, Texas 77030.

RESUMEN / SUMMARY: - Nonfunctional paragangliomas are slow-growing, typically benign tumors that arise from the extra-adrenal paraganglion of the autonomic nervous system. They are identified and characterized with the use of computed tomography and other imaging methods; for definitive diagnosis, histopathologic evaluation is crucial. Surgical resection is the treatment of choice, and results of postoperative biochemical testing can reveal recurrence. Because of this lesion's familial association, genetic testing is suggested. We report the case of an 81-year-old woman who presented with neck pain, intermittent palpitations, hypertension, and dyspnea. Contrast-enhanced computed tomography of the chest revealed a multilobular, high-density lesion between the aorta and the pulmonary artery in the superior mediastinum. The patient's 24-hour urinary vanillylmandelic acid levels were not elevated, which suggested a nonfunctional tumor. Mediastinal exploration revealed a large, vascular, irregular, consistently firm mass that adhered to the aortic arch. Upon histopathologic analysis after complete resection, the mass was determined to be a paraganglioma with a low index of mitosis. The patient had postoperative respiratory insufficiency that necessitated tracheostomy, but she recovered well after rehabilitation. In addition to reporting our patient's case, we discuss the nature, diagnosis, and treatment of paragangliomas.

[123]

TÍTULO / TITLE: - Analysis of pheochromocytomas / paragangliomas from Eastern Slovakia.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Neoplasma. 2013;60(4):452-8. doi: 10.4149/neo_2013_059.

●●Enlace al texto completo (gratis o de pago) [4149/neo_2013_059](#)

AUTORES / AUTHORS: - Felsoci M; Lazurova I; Wagnerova H; Svajdler M

RESUMEN / SUMMARY: - This multi centre observational cohort study gives a view about the occurrence, clinical and laboratory presentation, localization, histological type and genetic background of pheochromocytoma (PHEO) and paraganglioma (PGL) in Eastern Slovakia. It included 28 patients (18 women + 10 men), of which 23 were diagnosed to have PHEO (82,1%) and 7 patients (25%) suffered from PGL with retroperitoneal, inguinal/pelvic and mediastinal distribution. Arterial hypertension was the major symptom present in 86 % with slight dominance of paroxysmal form (58%). In 3 cases (10,7%), the diagnosis was gained after differentiation of adrenal incidentaloma in asymptomatic patients. Five patients (17,8%) were classified to have malignant form of the disease. 9 patients (32,1%) were confirmed to have hereditary form - five of them (17,8%) with familiar medullar thyroid cancer (FMTC) and mutations in RET gene classified as multiple endocrine neoplasia 2^a and 4 patients (14,3%) with germline mutations of SDHB gene, respectively. There was found a relatively high occurrence of other co-morbidities: thyroid disease in 20 patients (71,4%), impairment of glucose metabolism in 11 patients (39,3%) and apart from FMTC, 4 patients (14,3%) suffered also from other malignancy. Together with a bigger size of the primary tumor (6,6 cm), higher concentrations of metanephrines and prevalence of extra-adrenal tumors, malignant and hereditary forms, we suppose genetic and environmental factors of Eastern Slovakia may play a role in the etiopathogenesis of the tumors. Keywords: pheochromocytoma, paraganglioma, thyroid, diabetes mellitus, genetics, environmental.

[124]

TÍTULO / TITLE: - Predominantly tubular chromophobe renal cell carcinoma: an unusual histological presentation reminiscent of a neuroendocrine tumor.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Int J Surg Pathol. 2013 Jun;21(3):311-3. doi: 10.1177/1066896912475080.

●●Enlace al texto completo (gratis o de pago)

[1177/1066896912475080](#)

AUTORES / AUTHORS: - Singh G; Joshi P; Rao N; Seth A

INSTITUCIÓN / INSTITUTION: - 1All India Institute of Medical Sciences, New Delhi, India.

[125]

TÍTULO / TITLE: - Characterization of neuroendocrine tumors of the pancreas by real-time quantitative polymerase chain reaction. A methodological approach.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Endocr Pathol. 2013 Jun;24(2):83-91. doi: 10.1007/s12022-013-9246-y.

●●Enlace al texto completo (gratis o de pago) [1007/s12022-013-9246-](#)

[y](#)

AUTORES / AUTHORS: - Annaratone L; Volante M; Asioli S; Rangel N; Bussolati G

INSTITUCIÓN / INSTITUTION: - Department of Medical Sciences, University of Turin, Via Santena 7, 10126, Turin, Italy.

RESUMEN / SUMMARY: - The aim of this study was to assess the suitability of using real-time quantitative PCR (RT-qPCR) to characterize neuroendocrine (NE) tumors of the pancreas. For a series of tumors, we evaluated several genes of interest, and the data were matched with the “classical” immunohistochemical (IHC) features. In 21 cases, we extracted RNA from formalin-fixed paraffin-embedded (FFPE) blocks, and in nine cases, we also extracted RNA from fresh-frozen tissue. The RT-qPCR procedure was performed using two sets of customized arrays. The test using the first set, covering 96 genes of interest, was focused on assessing the feasibility of the procedure, and the results were used to select 18 genes indicative of NE differentiation, clinical behavior, and therapeutic responsiveness for use in the second set of arrays. Threshold cycle (Ct) values were used to calculate the fold-changes in gene expression using the 2-Ct method. Statistical procedures were used to analyze the results, which were matched with the IHC and follow-up data. Material from fresh-frozen samples performed better in terms of the level of amplification, but acceptable and concordant results were also obtained from FFPE samples. In addition, high concordance was observed between the mRNA and protein expression levels of somatostatin receptor type 2^a (R = 0.52, p = 0.016). Genes associated with NE differentiation, as well as the gastrin-releasing peptide receptor and O-6-methylguanine-DNA methyltransferase genes, were underexpressed, whereas angiogenesis-associated markers (CDH13 and SLIT2) were overexpressed in tissues with malignant behavior. The RT-qPCR procedure is practical and feasible in economic terms for the characterization of NE tumors of the pancreas and can complement morphological and IHC-based evaluations. Thus, the results of the RT-qPCR procedure might offer an objective basis for therapeutic choices.

[126]

TÍTULO / TITLE: - Neurofibromatosis 1 presenting with multiple duodenal gists associated with a somatostatin-producing d cell neoplasm.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Endocr Pathol. 2013 Jun;24(2):100-5. doi: 10.1007/s12022-013-9239-x.

●●Enlace al texto completo (gratis o de pago) [1007/s12022-013-9239-](#)

[X](#)

AUTORES / AUTHORS: - Serio G; Zampatti C; Ballabio A; Ricci R; Martini M; Zurleni F

INSTITUCIÓN / INSTITUTION: - Operative Unit of Anatomic Pathology, Azienda Ospedaliera "Ospedale di Circolo" di Busto Arsizio, Via A. da Brescia 1, 21052, Busto Arsizio, Italy, gserio@aobusto.it.

RESUMEN / SUMMARY: - The co-existence of a duodenal somatostatin-producing D cell neoplasm and multiple duodenal gastrointestinal stromal tumours (GISTs) in a 61-year-old woman with neurofibromatosis type 1 is reported. Histologically, the D cell neoplasm showed a glandular pattern with psammoma bodies and was metastatic to regional lymph nodes and liver at the time of surgery. Tumour cells were monomorph and showed intense and diffuse immunoreactivity for somatostatin, focal positivity for calcitonin, while were negative for other gastroenteropancreatic hormones including insulin, glucagon, pancreatic polypeptide, serotonin and gastrin. Four submucosal and subserosal GISTs, ranging from 5 to 15 mm in diameter, were composed of uniform spindle-shaped cells lacking mitoses and contained numerous skeinoid fibres. The tumours were positive for CD117, DOG1, vimentin and CD34 and did not have KIT or PDGFRA mutations. The clinical and pathological importance of this unusual association is discussed.

[127]

TÍTULO / TITLE: - Thymic Neuroendocrine Tumor Presenting with the Ectopic ACTH Syndrome.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - J Thorac Oncol. 2013 Jun;8(6):e57-8. doi: 10.1097/JTO.0b013e31828c2900.

●●Enlace al texto completo (gratis o de pago)

[1097/JTO.0b013e31828c2900](#)

AUTORES / AUTHORS: - Barbieri JS; Seshasai R; Shemesh A; Sedrak M; Hoffman B; Alley EW

INSTITUCIÓN / INSTITUTION: - *Perelman School of Medicine at the University of Pennsylvania, Philadelphia, Pennsylvania; and daggerRenal-Electrolyte and Hypertension Division, Departments of double daggerEmergency Medicine, section signMedicine, and ||Division of Hematology Oncology, University of Pennsylvania Health System, Philadelphia, Pennsylvania.

[128]

TÍTULO / TITLE: - Targeted Therapies in Neuroendocrine Tumors (NET): Clinical Trial Challenges and Lessons Learned.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Oncologist. 2013;18(5):525-32. doi: 10.1634/theoncologist.2012-0434. Epub 2013 Apr 24.

●●Enlace al texto completo (gratis o de pago)

[1634/theoncologist.2012-0434](#)

AUTORES / AUTHORS: - Yao JC; Lagunes DR; Kulke MH

INSTITUCIÓN / INSTITUTION: - The University of Texas MD Anderson Cancer Center, Houston, Texas, USA;

RESUMEN / SUMMARY: - In the past 3 years, we have witnessed the completion of four randomized phase III studies in neuroendocrine tumors and the approval of two new drugs, everolimus and sunitinib, for the treatment of patients with well-differentiated pancreatic neuroendocrine tumors. These studies demonstrate a shift from case series and single-arm studies toward prospective, randomized controlled clinical trials and evidence-based therapy in the neuroendocrine tumor field. However, the clinical development of these agents also highlights the potential challenges awaiting other new drugs in this area. Herein, we discuss the strengths and weaknesses of the most recent phase II and phase III neuroendocrine tumor studies and discuss how limitations inherent in current trial design can lead to potential pitfalls. We also discuss how trial design can be improved, with the hope of increasing the number of drugs successfully developed to treat patients with neuroendocrine tumors.

[129]

TÍTULO / TITLE: - Therapeutic effect of sunitinib malate and its influence on blood glucose concentrations in a patient with metastatic insulinoma.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Expert Rev Anticancer Ther. 2013 Apr 10.

●●Enlace al texto completo (gratis o de pago) [1586/era.13.45](#)

AUTORES / AUTHORS: - Chen J; Wang C; Han J; Luan Y; Cui Y; Shen R; Sha D; Cong L; Zhang Z; Wang W

INSTITUCIÓN / INSTITUTION: - Department of Oncology, Provincial Hospital affiliated to Shandong University, Jinan, 250021, China.

RESUMEN / SUMMARY: - Standard cytotoxic chemotherapy has limited efficacy in advanced insulinomas, and control of blood glucose concentrations in these patients may be difficult. This article describes an elderly (74-year-old) man with metastatic insulinoma and severe hypoglycemia who was treated with repeated 6-week cycles of oral sunitinib malate (25 mg/day for 4 weeks,

followed by 2 weeks off treatment). After treatment for more than 2 years, his condition improved and he continued to have a good quality of life with no evidence of tumor progression based on PET/CT findings. Although sunitinib treatment lowered the patient's blood glucose concentrations further and induced repeated symptomatic hypoglycemic episodes, he was able to tolerate the treatment well after changing the timing of sunitinib dosing and adjusting his diet.

[130]

TÍTULO / TITLE: - Selecting patients for cytotoxic therapies in gastroenteropancreatic neuroendocrine tumours.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Best Pract Res Clin Gastroenterol. 2012 Dec;26(6):843-54. doi: 10.1016/j.bpg.2012.12.001.

●●Enlace al texto completo (gratis o de pago) 1016/j.bpg.2012.12.001

AUTORES / AUTHORS: - Costa FP; Gumz B; Pasche B

INSTITUCIÓN / INSTITUTION: - Centro de Oncologia, Hospital Sirio Libanes, Rua Dona Adma Jafet 90, Sao Paulo, SP, CEP 01308-050, Brazil.
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RESUMEN / SUMMARY: - Gastroenteropancreatic neuroendocrine tumours (GEP-NET) have heterogenic clinical presentations. The majority of GEP-NET tumours have an indolent behaviour, but patients will eventually develop symptoms of tumour progression or hormone secretion that may require systemic medical interventions. Cytotoxic chemotherapy has been tested in GEP-NETs since the 80s, but treatment recommendations are controversial in many instances. Patient selection is mandatory for optimal use of chemotherapy. Important prognostic factors such as primary tumour site, tumour differentiation, tumour staging and proliferation index have been identified and validated in retrospective and prospective series. The combination of those factors and the natural history of GEP-NET provide valuable information with respect to treatment planning. In this report we provide treatment recommendations to improve systemic therapy in patients with advanced GEP-NETs based on a comprehensive review of the literature.

[131]

TÍTULO / TITLE: - Pancreatic neuroendocrine tumors: clinical features, diagnosis and medical treatment: advances.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Best Pract Res Clin Gastroenterol. 2012 Dec;26(6):737-53. doi: 10.1016/j.bpg.2012.12.003.

●●Enlace al texto completo (gratis o de pago) 1016/j.bpg.2012.12.003

AUTORES / AUTHORS: - Ito T; Igarashi H; Jensen RT

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RESUMEN / SUMMARY: - Pancreatic neuroendocrine tumors (pNETs) comprise with gastrointestinal carcinoids, the main groups of gastrointestinal neuroendocrine tumors (GI-NETs). Although these two groups of GI-NETs share many features including histological aspects; over-/ectopic expression of somatostatin receptors; the ability to ectopically secrete hormones/peptides/amines which can result in distinct functional syndromes; similar approaches used for tumor localization and some aspects of treatment, it is now generally agreed they should be considered separate. They differ in their pathogenesis, hormonal syndromes produced, many aspects of biological behaviour and most important, in their response to certain anti-tumour treatment (chemotherapy, molecular targeted therapies). In this chapter the clinical features of the different types of pNETs will be considered as well as aspects of their diagnosis and medical treatment of the hormone-excess state. Emphasis will be on controversial areas or recent advances. The other aspects of the management of these tumors (surgery, treatment of advanced disease, tumor localization) are not dealt with here, because they are covered in other chapters in this volume.

[132]

TÍTULO / TITLE: - Role of mitogen-activated protein kinase cascades in 2,3,7,8-tetrachlorodibenzo-p-dioxin-induced apoptosis in neuronal pheochromocytoma cells.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Hum Exp Toxicol. 2013 Apr 12.

●●Enlace al texto completo (gratis o de pago)

[1177/0960327113482595](#)

AUTORES / AUTHORS: - Xu G; Duan Z; Chen G; Nie X; Liu J; Zhang Y; Li Y; Wan C; Jiang J

INSTITUCIÓN / INSTITUTION: - 1Department of Nutrition and Food Hygiene, School of Public Health, Nantong University, Nantong, Jiangsu, People's Republic of China.

RESUMEN / SUMMARY: - Mitogen-activated protein kinases (MAPKs) are involved in neuronal death caused by many cytotoxins. Conventional MAPKs consist of three family members: extracellular signal-regulated kinase-1/2 (ERK1/2), c-Jun N-terminal kinase (JNK) and p38. It has been originally shown that ERK1/2 is important for cell survival, whereas JNK and p38 are deemed stress responsive and thus involved in apoptosis. However, information describing the role of MAPKs in 2,3,7,8-tetrachlorodibenzo-p-dioxin (TCDD)-

induced neurotoxicity is insufficient. The aim of this study was to identify the role of MAPK cascades in TCDD-induced neurotoxicity using differentiated pheochromocytoma (PC12) cells as a model for neuronal cells. Cell viability assay, terminal deoxynucleotidyl transferase dUTP nick-end labeling assay and flow cytometry analysis showed that TCDD attenuated cell viability with a dose- and time-dependent manner and significantly induced apoptosis in primary cortical neurons and PC12 cells. Western blot analysis indicated that TCDD markedly activated the expression of ERK1/2, JNK and p38 in TCDD-treated PC12 cells. Furthermore, PD98059 (ERK1/2 inhibitor), SP600125 (JNK inhibitor) and SB202190 (p38 inhibitor) notably blocked the effect of TCDD on cell apoptosis. Based on the findings above, it is concluded that the activation of MAPK signaling pathways may be associated with TCDD-mediated neuronal apoptosis.

[133]

TÍTULO / TITLE: - Pancreatic Surgery with Vascular Reconstruction in Patients with Locally Advanced Pancreatic Neuroendocrine Tumors.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - J Gastrointest Surg. 2013 May 14.

●●Enlace al texto completo (gratis o de pago) [1007/s11605-013-2221-](http://1007/s11605-013-2221-6)

[6](#)

AUTORES / AUTHORS: - Haugvik SP; Labori KJ; Waage A; Line PD; Mathisen O; Gladhaug IP

INSTITUCIÓN / INSTITUTION: - Department of Hepato-Pancreato-Biliary Surgery, Rikshospitalet, Oslo University Hospital, Sognsvannsveien 20, 0372, Oslo, Norway, svhaug@ous-hf.no.

RESUMEN / SUMMARY: - INTRODUCTION: Pancreatic neuroendocrine tumors (PNET) are rare neoplasms with better prognosis than most pancreatic malignancies. Surgery of locally advanced PNET remains controversial, and the role of vascular reconstruction in this patient group has yet to be defined. The aim of this study was to evaluate the feasibility and outcome of pancreatic resections with vascular reconstruction in patients with locally advanced PNET. METHODS: Retrospective analysis of patients who underwent pancreatic surgery with vascular reconstruction for locally advanced PNET at a single institution. Furthermore, a review of the relevant literature on the topic was performed. RESULTS: Seven patients who had undergone vascular reconstruction for locally advanced PNET were identified. Four patients had liver metastases at time of surgery. Postoperative complications developed in four patients with no mortality. Median follow-up time of all patients was 21 (range, 3-58) months. Three patients had disease in remission after 58, 42 and 3 months, respectively. One patient died 35 months postoperatively due to progressive disease, whereas three patients had progression of disease after

21, 9, and 4 months postoperatively. CONCLUSION: Pancreatic surgery with vascular reconstruction in patients with locally advanced PNET is feasible with acceptable outcome.

[134]

TÍTULO / TITLE: - Neuroendocrine Tumors Arising in Meckel's Diverticula: Frequency of Advanced Disease Warrants Aggressive Management.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - J Gastrointest Surg. 2013 Jun;17(6):1084-91. doi: 10.1007/s11605-013-2191-8. Epub 2013 Apr 5.

●●Enlace al texto completo (gratis o de pago) [1007/s11605-013-2191-8](#)

AUTORES / AUTHORS: - Lorenzen AW; O'Dorisio TM; Howe JR

INSTITUCIÓN / INSTITUTION: - Department of Surgery, University of Iowa Carver College of Medicine, Iowa City, IA, USA.

RESUMEN / SUMMARY: - BACKGROUND: Meckel's diverticulum is a common anomaly of the GI tract, which occasionally gives rise to cancer. The most frequent tumors affecting these diverticula are neuroendocrine tumors (NETs), and whether these should be treated in similar fashion as small bowel NETs or appendiceal NETs is unclear. METHODS: A retrospective chart review was conducted at a single academic medical center between 1998 and 2012. Demographic, radiologic, biochemical, and clinicopathologic data were collected as well as status at last follow-up. RESULTS: Seven patients were identified with NETs involving Meckel's diverticula, including one with limited information other than management of her late metastases. Of the six other patients, all had involvement of regional nodes, including three patients with tumors <2 cm in size, and four had liver metastases at presentation. CONCLUSIONS: NETs in Meckel's diverticula are rare tumors, but when they develop, are often associated with nodal metastases and liver metastases, even when the tumors are small. Therefore, optimal management of these NETs is small bowel resection with regional lymphadenectomy and debulking of liver metastases where feasible.

[135]

TÍTULO / TITLE: - Radiolabelled somatostatin analogue treatment in gastroenteropancreatic neuroendocrine tumours: factors associated with response and suggestions for therapeutic sequence.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Eur J Nucl Med Mol Imaging. 2013 Apr 26.

●●Enlace al texto completo (gratis o de pago) [1007/s00259-013-2402-](#)

[2](#)

AUTORES / AUTHORS: - Campana D; Capurso G; Partelli S; Nori F; Panzuto F; Tamburrino D; Cacciari G; Delle Fave G; Falconi M; Tomassetti P

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RESUMEN / SUMMARY: - PURPOSE: Peptide receptor radionuclide therapy (PRRT) is a relatively new treatment modality for patients with unresectable or metastatic gastroenteropancreatic neuroendocrine tumours (GEP NETs). The aim of this study was to determine the time to progression of patients treated with PRRT and to identify the prognostic factors related to treatment response. METHODS: Patients with sporadic GEP NETs prospectively treated with PRRT were retrospectively analysed. The primary end point was progression-free survival (PFS). RESULTS: A total of 69 patients (37 men and 32 women; 45 with pancreatic and 24 with gastrointestinal lesion; 22 NET G1 and 41 NET G2) were treated with ⁹⁰Y or ¹⁷⁷Lu. The objective response rate was 27.5 % (partial response, PR), while 50.7 % had stable disease and 23.2 % had progressive disease. Significant differences in PFS were observed in relationship to the stage of the disease (44 months for stage III, 23 months for stage IV), the evidence of a PR 6 months after the end of the PRRT (39 months in patients with a PR, 22 months in patients without a PR) and previous transarterial chemoembolization (TACE, yes 13 months vs no 31 months). Stage IV, NET G2 and previous TACE were found to be significant factors for tumour progression at multivariate analysis. CONCLUSION: Low tumour burden and a low proliferation index represent independent prognostic factors for long PFS, while previous chemoembolization techniques represent independent prognostic factors for early tumour progression and shorter PFS. Our data suggest that chemoembolization techniques to reduce the hepatic tumour burden should be avoided.

[136]

TÍTULO / TITLE: - Molecular and therapeutic advances in the diagnosis and management of malignant pheochromocytomas and paragangliomas.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Oncologist. 2013;18(4):391-407. doi: 10.1634/theoncologist.2012-0410. Epub 2013 Apr 10.

●●Enlace al texto completo (gratis o de pago)

[1634/theoncologist.2012-0410](http://1634.theoncologist.2012-0410)

AUTORES / AUTHORS: - Lowery AJ; Walsh S; McDermott EW; Prichard RS

INSTITUCIÓN / INSTITUTION: - Department of Surgery, St. Vincent's University Hospital, Dublin, Ireland.

RESUMEN / SUMMARY: - Pheochromocytomas (PCCs) and paragangliomas (PGLs) are rare catecholamine-secreting tumors derived from chromaffin cells

originating in the neural crest. These tumors represent a significant diagnostic and therapeutic challenge because the diagnosis of malignancy is frequently made in retrospect by the development of metastatic or recurrent disease. Complete surgical resection offers the only potential for cure; however, recurrence can occur even after apparently successful resection of the primary tumor. The prognosis for malignant disease is poor because traditional treatment modalities have been limited. The last decade has witnessed exciting discoveries in the study of PCCs and PGLs; advances in molecular genetics have uncovered hereditary and germline mutations of at least 10 genes that contribute to the development of these tumors, and increasing knowledge of genotype-phenotype interactions has facilitated more accurate determination of malignant potential. Elucidating the molecular mechanisms responsible for malignant transformation in these tumors has opened avenues of investigation into targeted therapeutics that show promising results. There have also been significant advances in functional and radiological imaging and in the surgical approach to adrenalectomy, which remains the mainstay of treatment for PCC. In this review, we discuss the currently available diagnostic and therapeutic options for patients with malignant PCCs and PGLs and detail the molecular rationale and clinical evidence for novel and emerging diagnostic and therapeutic strategies.

[137]

TÍTULO / TITLE: - Routine preoperative In-octreotide scintigraphy in patients with medullary thyroid cancer.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Langenbecks Arch Surg. 2013 May 19.

●●Enlace al texto completo (gratis o de pago) [1007/s00423-013-1086-](http://1007/s00423-013-1086-1)

[1](#)

AUTORES / AUTHORS: - Dahlberg J; Bumming P; Gjertsson P; Jansson S

INSTITUCIÓN / INSTITUTION: - Department of Surgery, Sahlgrenska University Hospital, 413 45, Gothenburg, Sweden, jakob.dahlberg@vgregion.se.

RESUMEN / SUMMARY: - BACKGROUND: Surgery is the only potential cure for patients with medullary thyroid carcinoma (MTC). Preoperative ultrasound, computed tomography and magnetic resonance imaging are not sensitive enough for detection of microscopic disease. The aim of this study was to investigate if routine preoperative ¹¹¹In-labelled (DTPA-D-Phe¹)-octreotide scintigraphy (SRS) could be used as a staging procedure in planning primary surgery in patients with MTC. METHODS: This study included patients with primary sporadic clinically overt MTC diagnosed between 1996 and 2009. All patients underwent conventional imaging of neck and thorax and SRS prior to standardised surgery. The findings on SRS were correlated to the findings on conventional imaging, histopathology and to postoperative biochemical results

and survival. RESULTS: A total of 19 patients with sporadic MTC were enrolled. Median follow-up was 77(9-184) months. SRS visualised the primary tumour in 16 (84 %) patients. Fifteen (79 %) patients had locoregional lymph node metastases, but SRS detected metastatic lesions in only 8 (53 %) patients. In three patients with distant spread, SRS failed to detect metastatic lesions in two. At latest follow-up, six (32 %) patients had died, nine (47 %) patients were alive with elevated tumour markers, and four (21 %) patients were considered in complete biochemical remission. CONCLUSIONS: This study provided further evidence that SRS, compared to conventional imaging, is fairly sensitive for detection of primary MTC but not metastatic disease. Although preoperative SRS may be of prognostic value, there is no indication for its routine use as a staging procedure in planning primary surgery.

[138]

TÍTULO / TITLE: - Multimodality treatment for poorly differentiated neuroendocrine head and neck carcinomas - a single institution experience.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Eur J Cancer Care (Engl). 2013 May 23. doi: 10.1111/ecc.12070.

●●Enlace al texto completo (gratis o de pago) [1111/ecc.12070](#)

AUTORES / AUTHORS: - Gorner M; Brasch F; Hirnle P; Gehl HB; Scholtz LU; Wegehenkel K; Sudhoff H

INSTITUCIÓN / INSTITUTION: - Department for Hematology, Oncology and Palliative Care, Academic Teaching Hospital Bielefeld, Bielefeld.

RESUMEN / SUMMARY: - Poorly differentiated head and neck neuroendocrine neoplasms are very rare. Surgical resection alone is insufficient to control the disease because of the high incidence of metastases. However, due to the lack of randomised clinical trials, treatment recommendations for this cancer vary considerably and are based on a limited number of small retrospective studies. We performed a retrospective analysis of all patients treated at our institution between 2003 and 2011. We assessed the stage of disease, type of therapy, toxicity, treatment response, time to progression and overall survival for all cases. Ten patients received combined modality treatment with chemotherapy in addition to surgery or radiation or both. According to Response Evaluation Criteria In Solid Tumours (RECIST) criteria, six of nine evaluable patients achieved complete remission and three patients had a partial remission. The mean duration of response was 358 days, with a range from 141 to 1080 days. The overall 1-year survival rate was 88%; however, only approximately 50% of patients were alive after 2 years. Multimodality treatment concepts induce high initial remission rates in poorly differentiated neuroendocrine head and neck carcinomas. However, the time to relapse is usually short, and therefore long-term prognosis of this rare head and neck tumour remains poor.

[139]

TÍTULO / TITLE: - Peptide receptor imaging in neuroendocrine tumors: comparison between diagnostic scintigraphy and post-therapy whole-body scan.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Ann Nucl Med. 2013 May 12.

●●Enlace al texto completo (gratis o de pago) [1007/s12149-013-0732-](#)

[6](#)

AUTORES / AUTHORS: - Minutoli F; Herberg A; Sindoni A; Cardile D; Cucinotta M; Baldari S

INSTITUCIÓN / INSTITUTION: - Department of Biomedical Sciences and of Morphologic and Functional Images, University of Messina, Messina, Italy, fminutoli@unime.it.

RESUMEN / SUMMARY: - **OBJECTIVE:** Several different somatostatin analogs labeled with gamma or positron-emitting radionuclides exist for diagnostic imaging of neuroendocrine tumors (NETs). Differences between standard diagnostic scintigraphy (SDS) and post-therapy whole-body scan (PTWBS) at peptide receptor radionuclide therapy in lesion detection are known; such differences have been correlated with the varying degree of receptor subtype expression and the varying receptor affinity profile of the different ligands. The aim of this study is to investigate differences between SDS and PTWBS obtained using the same radiopharmaceutical. **METHODS:** We retrospectively reviewed clinical records of 53 patients with a diagnosis of NET, who underwent both SDS and PTWBS using ¹¹¹In-Pentetreotide. We compared the number of lesions for each body region detected by SDS and PTWBS. **RESULTS:** In 14/53 patients (26.4 %) discrepancies between SDS and PTWBS were found. PTWBS detected 68 additional lesions with respect to SDS that were distributed as follows: head and neck, 6; mediastinum, 1; liver, 10; abdomen/pelvis, 1; bone, 44; other localizations, 6. The number of lesions detected by SDS was significantly different from that revealed by PTWBS (Wilcoxon matched pairs test, P = 0.0313). The regions that contributed significantly to reach this difference were head and neck (McNemar test, P = 0.0412), liver (McNemar test, P = 0.0044), bone (McNemar test, P < 0.0001) and other localizations (McNemar test, P = 0.0412). **CONCLUSION:** PTWBS shows more lesions than SDS with a significant discrepancy. We suppose that administration of higher radiopharmaceutical activity, use of larger peptide amount and the different time interval between radiopharmaceutical administration and scan execution can determine a higher sensitivity of PTWBS.

[140]

TÍTULO / TITLE: - Current and Future Treatments for Malignant Pheochromocytoma and Sympathetic Paraganglioma.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Curr Oncol Rep. 2013 May 15.

●●Enlace al texto completo (gratis o de pago) [1007/s11912-013-0320-](#)

[X](#)

AUTORES / AUTHORS: - Jimenez C; Rohren E; Habra MA; Rich T; Jimenez P; Ayala-Ramirez M; Baudin E

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RESUMEN / SUMMARY: - Pheochromocytomas (PHs) and sympathetic paragangliomas (SPGs) are rare neuroendocrine tumors. Approximately 17 % of these tumors are malignant, but because no molecular or histologic markers for malignancy exist, patients are often diagnosed with malignant PHs or SPGs after unresectable disease has formed. Patients with progressive metastatic tumors and overwhelming symptoms are currently treated with systemic chemotherapy and radiopharmaceutical agents such as metaiodobenzylguanidine. These therapies lead to partial radiographic response, disease stabilization, and symptomatic improvement in approximately 40 % of patients, and systemic chemotherapy is associated with a modest improvement in overall survival duration. However, over the past decade, substantial progress has been made in clinical, biochemical, and radiographic diagnosis of PHs and SPGs. Approximately 50 % of patients with malignant PHs and SPGs have been found to carry hereditary germline mutations in the succinate dehydrogenase subunit B gene (SDHB), and anti-angiogenic agents such as sunitinib have been found to potentially play a role in the treatment of malignant disease, especially in patients with SDHB mutations. In some patients, treatment with sunitinib has been associated with partial radiographic response, disease stabilization, decreased fluorodeoxyglucose uptake on positron emission tomography, and improved blood pressure control. These findings have led to the development of prospective clinical trials of new targeted therapies for metastatic disease. Here, we provide an updated review of the clinical and genetic predictors of malignant disease, radiographic diagnosis of malignant disease, and information from the most relevant studies of systemic therapies, as well as proposed treatment guidelines for patients with metastatic or potentially malignant PHs and SPGs.

[141]

TÍTULO / TITLE: - Orbital Metastasis Secondary to Merkel Cell Carcinoma: Case Report and Literature Review.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Orbit. 2013 May 10.

●●Enlace al texto completo (gratis o de pago)

3109/01676830.2013.788671

AUTORES / AUTHORS: - Johnson D; Warder D; Plourde ME; Brundage M; Ten Hove M

INSTITUCIÓN / INSTITUTION: - Department of Ophthalmology, Queen's University, Kingston, Ontario and.

RESUMEN / SUMMARY: - Abstract Introduction: Merkel cell carcinoma is an aggressive malignancy often associated with metastatic spread, but has never been reported to metastasize to the orbit. Case: An 80 year old male with metastatic Merkel cell carcinoma presented with ptosis and extraocular movement abnormalities, and was found to have a lesion of the orbit consistent with metastatic spread. The lesion responded favorably to radiation therapy. Comment: Although the orbit is a frequent site of metastatic disease, this is the first reported case of presumed Merkel cell carcinoma metastasizing to the orbit.

[142]

TÍTULO / TITLE: - Radiological and nuclear medicine imaging of gastroenteropancreatic neuroendocrine tumours.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Best Pract Res Clin Gastroenterol. 2012 Dec;26(6):803-18. doi: 10.1016/j.bpg.2012.12.004.

●●Enlace al texto completo (gratis o de pago) 1016/j.bpg.2012.12.004

AUTORES / AUTHORS: - Sundin A

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RESUMEN / SUMMARY: - Neuroendocrine tumours (NETs) comprise a heterogeneous group of neoplasms with very varying clinical expression. A functioning NET, for instance in the pancreas, may be very small and yet give rise to severe endocrine symptoms whereas a patient with a small bowel tumour may present with diffuse symptoms and disseminated disease with a palpable bulky liver. Imaging of NETs is therefore challenging and the imaging needs in the various patients are diverse. The basic modalities for NET imaging are computed tomography (CT) or magnetic resonance imaging (MRI) in combination with somatostatin receptor imaging (SMI) by scintigraphy with ¹¹¹In-labelled octreotide (OctreoScan) or more recently by positron emission tomography (PET) with ⁶⁸Ga-labelled somatostatin analogues. In this review these various morphological and functional imaging modalities and important methodological aspects are described. Imaging requirements for the various types of NETs are discussed and typical image findings are illustrated.

[143]

TÍTULO / TITLE: - Risk stratification in medullary thyroid cancer: Moving beyond static anatomic staging.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Oral Oncol. 2013 Apr 16. pii: S1368-8375(13)00519-8. doi: 10.1016/j.oraloncology.2013.03.443.

●●Enlace al texto completo (gratis o de pago)

1016/j.oraloncology.2013.03.443

AUTORES / AUTHORS: - Tuttle RM; Ganly I

INSTITUCIÓN / INSTITUTION: - Endocrinology Service, Memorial Sloan-Kettering Cancer Center, New York, NY, United States. Electronic address: tuttlem@mskcc.org.

RESUMEN / SUMMARY: - Objectives: Much progress has been made over the last 10 years with regard to risk estimation in non-medullary differentiated thyroid cancer with risk of recurrence systems and response to therapy re-evaluation approaches being used to augment initial risk estimates obtained using standard anatomic staging systems. Furthermore, risk stratification is being increasingly viewed as an active, evolving, dynamic process that requires re-evaluation during follow-up rather than a single static risk estimate predicted by initial staging. As with differentiated thyroid cancer, multiple clinico-pathologic factors have been demonstrated to correlate with the risk of disease specific mortality, risk of death, likelihood of disease progression, likelihood of cure with initial therapy, and likelihood of cure with additional therapy in medullary thyroid cancer. Materials and Methods: In this review, we re-examine the clinically important initial risk factors in medullary thyroid cancer and then re-evaluate how some of these risk factors can be used to alter risk estimates over time as they reflect the response to therapy and the clinical course of the disease. Results and Conclusions: We demonstrate that the same response to therapy nomenclature that we have proposed and validated in differentiated thyroid cancer (excellent response, biochemical incomplete response, structural incomplete response, indeterminant response) can be easily applied to medullary cancer and used to guide on-going clinical management.

[144]

TÍTULO / TITLE: - Outcomes of concurrent Caesarean delivery and pheochromocytoma resection in late pregnancy.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Intern Med J. 2013 May;43(5):588-91. doi: 10.1111/imj.12118.

●●Enlace al texto completo (gratis o de pago) 1111/imj.12118

AUTORES / AUTHORS: - Song Y; Liu J; Li H; Zeng Z; Bian X; Wang S

INSTITUCIÓN / INSTITUTION: - Department of Obstetrics and Gynecology, Peking Union Medical College Hospital, Peking Union Medical College, Chinese Academy of Medical Sciences, Beijing, China.

RESUMEN / SUMMARY: - Undiagnosed pheochromocytoma in pregnancy is associated with significant maternal and foetal mortality. Herein we reviewed five cases of pheochromocytoma in pregnancy occurring during late pregnancy. The mean age at presentation was 30.6 years, and the gestational age ranged from 26 to 36 weeks. All patients had elevated levels of urinary catecholamines. In three patients, the lesion was located in the adrenal gland, in one patient the urinary bladder, and there was one case of recurrent malignant pheochromocytoma with metastases. Tumour resections were performed at the time of Caesarean section in four patients, two through laparoscopy. When pheochromocytoma is diagnosed in pregnancy after the second trimester, concurrent tumour resection with Caesarean section is feasible and in our series was achieved in four cases without adverse maternal or foetal consequences.

[145]

TÍTULO / TITLE: - Integrative analysis of miRNA and mRNA expression profiles in pheochromocytoma and paraganglioma identifies genotype-specific markers and potentially regulated pathways.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Endocr Relat Cancer. 2013 May 16.

●●Enlace al texto completo (gratis o de pago) [1530/ERC-12-0183](#)

AUTORES / AUTHORS: - de Cubas AA; Leandro-Garcia LJ; Schiavi F; Mancikova V; Comino-Mendez I; Inglada-Perez L; Perez M; Ibarz N; Ximenez-Embun P; Lopez-Jimenez E; Leton R; Maliszewska A; Gomez Grana A; Bernal C; Alvarez-Escola C; Rodriguez-Antona C; Opocher G; Munoz J; Megias D; Cascon A; Robledo M

INSTITUCIÓN / INSTITUTION: - A de Cubas, Hereditary Endocrine Cancer Group (Human Cancer Genetic Programme), Spanish National Cancer Research Centre (CNIO), Madrid, 28029, España.

RESUMEN / SUMMARY: - Pheochromocytoma (PCC) and paraganglioma (PGL) are rare neuroendocrine neoplasias of neural crest origin that can be part of several inherited syndromes. Although their mRNA profiles are known to depend on genetic background, a number of questions related to tumor biology and clinical behavior remain unanswered. Since microRNAs are key players in the modulation of gene expression, their comprehensive analysis could resolve some of these issues. Through characterization of microRNA profiles in 69 frozen tumors with germline mutations in the genes SDHD, SDHB, VHL, RET, NF1, TMEM127, and MAX, we identified microRNA signatures specific to, as well as common among, the genetic groups of PCC/PGLs. MicroRNA

expression profiles were validated in an independent series of 30 composed of VHL-, SDHB-, SDHD- and RET-related formalin-fixed paraffin-embedded PCC/PGL samples using qRT-PCR. Up-regulation of miR-210 in VHL- and SDHB-related PCC/PGL, while miR-137 and miR-382 were confirmed as generally up-regulated in PCC/PGL (except in MAX-related tumors). Also, we confirmed over-expression of miR-133b as VHL-specific, miR-488 and miR-885-5p as RET-specific, and miR-183 and miR-96 as SDHB-specific microRNAs. To determine the potential roles microRNAs play in PCC/PGL pathogenesis, we performed bioinformatic integration and pathway analysis using matched mRNA profiling data that indicated a common enrichment of pathways associated with neuronal and neuroendocrine-like differentiation. We demonstrated that miR-183 and/or miR-96 impede NGF-induced differentiation in PC12 cells. Finally, global proteomic analysis in SDHB and MAX-tumors allowed us to determine that microRNA regulation occurs primarily through mRNA degradation in PCC/PGL, which partially confirmed our miRNA-mRNA integration results.

[146]

TÍTULO / TITLE: - A multi-institutional, phase II open-label study of ganitumab (AMG 479) in advanced carcinoid and pancreatic neuroendocrine tumors.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - *Endocr Relat Cancer*. 2013 May 21;20(3):383-90. doi: 10.1530/ERC-12-0390. Print 2013 Jun.

●●Enlace al texto completo (gratis o de pago) [1530/ERC-12-0390](#)

AUTORES / AUTHORS: - Strosberg JR; Chan JA; Ryan DP; Meyerhardt JA; Fuchs CS; Abrams T; Regan E; Brady R; Weber J; Campos T; Kvolts LK; Kulke MH

INSTITUCIÓN / INSTITUTION: - Department of GI Oncology, Moffitt Cancer Center, 12902 Magnolia Drive, Tampa, Florida 33612, USA Department of Medical Oncology, Dana-Farber Cancer Institute, Boston, Massachusetts 02481, USA Division of Medical Oncology, Massachusetts General Hospital, Boston, Massachusetts 02481, USA.

RESUMEN / SUMMARY: - The IGF pathway has been implicated in the regulation of neuroendocrine tumor (NET) growth, and preliminary studies suggested that ganitumab (AMG 479), a human MAB against IGF1R, may have antitumor activity in this setting. We performed a two-cohort phase II study of ganitumab in patients with metastatic progressive carcinoid or pancreatic NETs (pNETs). This open-label study enrolled patients (≥ 18 years) with metastatic low- and intermediate-grade carcinoid or pNETs. Inclusion criteria included evidence of progressive disease (by Response Evaluation Criteria in Solid Tumors (RECIST)) within 12 months of enrollment, ECOG PS 0-2, and fasting blood sugar < 160 mg/dl. Prior treatments were allowed and concurrent somatostatin analog therapy was permitted. The primary endpoint was objective response.

Secondary endpoints included overall survival (OS), progression-free survival (PFS), and safety. Sixty patients (30 carcinoid and 30 pNETs) were treated with ganitumab 18 mg/kg every 3 weeks, among whom 54 patients were evaluable for survival and 53 patients for response. There were no objective responders by RECIST. The median PFS duration was 6.3 months (95% CI, 4.2-12.6) for the entire cohort; 10.5 months for carcinoid patients, and 4.2 months for pNET patients. The OS rate at 12 months was 66% (95% CI, 52-77%) for the entire cohort. The median OS has not been reached. Grade $\frac{3}{4}$ AEs were rare and consisted of hyperglycemia (4%), neutropenia (4%), thrombocytopenia (4%), and infusion reaction (1%). Although well tolerated, treatment with single-agent ganitumab failed to result in significant tumor responses among patients with metastatic well-differentiated carcinoid or pNET.

[147]

TÍTULO / TITLE: - Serotonin and the 5-HT₇ receptor: The link between hepatocytes, IGF-1 and small intestinal neuroendocrine tumors.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Cancer Sci. 2013 Apr 11. doi: 10.1111/cas.12174.

●●Enlace al texto completo (gratis o de pago) [1111/cas.12174](#)

AUTORES / AUTHORS: - Svejda B; Kidd M; Timberlake A; Harry K; Kazberouk A; Schimmack S; Lawrence B; Pfragner R; Modlin IM

INSTITUCIÓN / INSTITUTION: - Gastrointestinal Pathobiology Research Group, Yale University School of Medicine, New Haven, Connecticut, USA.

RESUMEN / SUMMARY: - Platelet-derived serotonin (5-HT) is involved in liver regeneration. The liver is also the metastatic site for malignant enterochromaffin (EC) cell "carcinoid" (neuroendocrine) neoplasms, the principal cellular source of 5-HT. We hypothesized that 5-HT produced by metastatic EC cells played a role in the hepatic tumor-microenvironment principally via 5-HT₇ receptor-mediated activation of hepatocyte IGF-1 synthesis and secretion. Using isolated rat hepatocytes, we evaluated 5-HT₇ receptor expression (using PCR, sequencing and western blot). ELISA, cell transfection and western blots delineated 5-HT-mediated signaling pathways (pCREB, AKT and ERK). IGF-1 synthesis/secretion was evaluated using QPCR and ELISA. IGF-1 was tested on small intestinal neuroendocrine neoplasm proliferation, while IGF-1 production and 5-HT₇ expression were examined in an in vivo SCID metastasis model. Our results demonstrated evidence for a functional 5-HT₇ receptor. 5-HT activated cAMP/PKA activity, pCREB (130-205%, P < 0.05) and pERK/pAKT (1.2-1.75, P < 0.05). Signaling was reversed by the 5-HT₇ receptor antagonist SB269970. IGF-1 significantly stimulated proliferation of two small intestinal neuroendocrine neoplasm cell lines (EC₅₀ : 7-70 pg/mL) and could be reversed by the small molecule inhibitor BMS-754807. IGF-1 and 5-HT were elevated (40-300x) in peri-tumoral hepatic tissue in nude mice, while 5-HT₇ was

increased fourfold compared to sham-operated animals. We conclude that hepatocytes express a cAMP-coupled 5-HT₇ receptor, which, at elevated 5-HT concentrations that occur in liver metastases, signals via CREB/AKT and is linked to IGF-1 synthesis and secretion. Because IGF-1 regulates NEN proliferation, identification of a role for 5-HT₇ in the hepatic metastatic tumor microenvironment suggests the potential for novel therapeutic strategies for amine-producing mid-gut tumors.

[148]

TÍTULO / TITLE: - Humoral hypercalcemia of malignancy caused by parathyroid hormone-related peptide-secreting neuroendocrine tumors: Report of six cases.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Pancreatology. 2013 May-Jun;13(3):324-6. doi: 10.1016/j.pan.2013.02.007. Epub 2013 Mar 1.

●●Enlace al texto completo (gratis o de pago) 1016/j.pan.2013.02.007

AUTORES / AUTHORS: - Milanesi A; Yu R; Wolin EM

INSTITUCIÓN / INSTITUTION: - Division of Endocrinology, Cedars-Sinai Medical Center, Los Angeles, CA, USA.

RESUMEN / SUMMARY: - We report the clinical characteristics and management of six patients with metastatic gastroentero-pancreatic neuroendocrine tumor (NET) presenting with severe hypercalcemia due to elevation of parathyroid hormone-related protein (PTHrP). All patients had histological confirmation of NET, five well-differentiated and one poorly differentiated. In 5 patients, hypercalcemia developed after years after the initial diagnosis of NET. One patient presented with concomitant elevation of PTHrP and intact parathyroid hormone (PTH) in the setting of multiple endocrine neoplasia 1 (MEN1). In all the other cases, PTH levels were low or undetectable. Management of malignant hypercalcemia due to PTHrP-producing NET is challenging, and optimal therapy depends on the extent of metastatic disease and the grade of malignancy. Aggressive tumor cytoreduction in addition to the systemic treatment modalities is frequently used to control disease progression and endocrine symptoms. To our knowledge, this is the largest series to date of hypercalcemia mediated by PTHrP-secreting NET.

[149]

TÍTULO / TITLE: - Tanycytic ependymoma of the filum terminale associated with multiple endocrine neoplasia Type 1: first reported case.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Spine J. 2013 Apr 3. pii: S1529-9430(13)00266-0. doi: 10.1016/j.spinee.2013.02.066.

- Enlace al texto completo (gratis o de pago)

[1016/j.spinee.2013.02.066](https://doi.org/10.1016/j.spinee.2013.02.066)

AUTORES / AUTHORS: - Funayama T; Sakane M; Yoshizawa T; Takeuchi Y; Ochiai N

INSTITUCIÓN / INSTITUTION: - Department of Orthopaedic Surgery, Graduate School of Comprehensive Human Sciences, University of Tsukuba, 1-1-1 Tennoudai, Tsukuba, Ibaraki 305-8575, Japan. Electronic address: funatoru3@gmail.com.

RESUMEN / SUMMARY: - BACKGROUND CONTEXT: Ependymoma associated with multiple endocrine neoplasia Type 1 (MEN-1) is an extremely rare clinical entity. To the best of our knowledge, only five cases of ependymoma associated with MEN-1 have been previously described. Furthermore, there has been no case of tanycytic ependymoma of the filum terminale associated with MEN-1. PURPOSE: The present case report illustrates a 53-year-old man with tanycytic ependymoma of the filum terminale associated with MEN-1. We review the literature on ependymoma with MEN-1 and tanycytic ependymoma of the cauda equina region and also discuss the risk of recurrence. STUDY DESIGN: A case report. METHODS: The patient presented with complaints of nocturnal pain in the lower back, accompanied by numbness around the anus and intermittent claudication for approximately 1 year. Magnetic resonance imaging (MRI) identified an intradural-enhancing, large mass lesion at the level from Th12 to L2 vertebrae, with a cranial cystic lesion. RESULTS: Open-door laminoplasty of the Th12, L1, and L2 and en bloc tumor resection with thickened filum terminale were performed. Histopathologic examination of the tumor specimens showed tanycytic ependymoma (World Health Organization Classification Grade II). At the time of the 2-year and 8-month follow-up examination, MRI did not show tumor recurrence. CONCLUSIONS: This is the first reported case of this clinical entity. A careful follow-up of patients with this unusual tumor is strongly recommended.

[150]

TÍTULO / TITLE: - Case report: recurrent acute pancreatitis secondary to papillary somatostatinoma—a new association.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Pancreatology. 2013 Mar-Apr;13(2):186-8. doi: 10.1016/j.pan.2013.01.012. Epub 2013 Feb 9.

- Enlace al texto completo (gratis o de pago) [1016/j.pan.2013.01.012](https://doi.org/10.1016/j.pan.2013.01.012)

AUTORES / AUTHORS: - Ambrose T; Stephenson TJ; Booth J; Chandra N

INSTITUCIÓN / INSTITUTION: - Department of Gastroenterology, Royal Berkshire Hospital, London Road, Reading RG1 5AN, United Kingdom.

RESUMEN / SUMMARY: - Acute pancreatitis may rarely be caused by papillary mass lesions such as adenocarcinomas and neuroendocrine tumours.

Occasionally these papillary lesions may cause recurrent episodes of acute pancreatitis and patients presenting in this way require further pancreatic investigation. We believe this to be the first reported case of a duodenal papillary somatostatinoma causing recurrent acute pancreatitis. The patient was investigated with multiple imaging modalities, both at endoscopy and with more traditional radiology, and treated with resection by Whipple's pancreaticoduodenectomy. If diagnosed early in the absence of distant metastases the prognosis of papillary somatostatinoma with tumour resection is excellent.

[151]

TÍTULO / TITLE: - Pulmonary Collision Tumor Consisting of Adenocarcinoma and Typical Carcinoid-A Case Report and Review of Literature.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Am J Ther. 2013 May 17.

●●Enlace al texto completo (gratis o de pago)

[1097/MJT.0b013e318293b0b0](#)

AUTORES / AUTHORS: - Abbi KK; Hameed MK; Jiang Y; De Las Casas LE; Schwann TA

INSTITUCIÓN / INSTITUTION: - 1Division of Hematology and Oncology, Department of Medicine, 2Department of Pathology, and 3Division of Cardiothoracic Surgery, Department of Surgery, University of Toledo Medical Center, Toledo, OH.

RESUMEN / SUMMARY: - Collision tumors are rare in nature. We report a case of a 70-year-old woman who was found to have a new mass in the right lung. Right upper and middle lobectomies with a mediastinal lymph node sampling were performed. Pathological examination of the mass revealed a collision tumor composed of micropapillary adenocarcinoma and typical carcinoid. The neoplastic cells were not intimately admixed with one another. To the best of our knowledge, this case is the first report in the English medical literature of a primary pulmonary collision tumor consisting of micropapillary adenocarcinoma and typical carcinoid.

[152]

TÍTULO / TITLE: - Imprint cytological feature of large cell neuroendocrine carcinoma of the gallbladder: A case report.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - J Med Invest. 2013;60(1-2):149-53.

AUTORES / AUTHORS: - Nakagawa T; Sakashita N; Ohnishi K; Komohara Y; Takeya M

INSTITUCIÓN / INSTITUTION: - Department of Cell Pathology, Graduate School of Medical Sciences, Faculty of Life Sciences, Kumamoto University.

RESUMEN / SUMMARY: - Large cell neuroendocrine carcinoma (LCNEC) is a rare poorly differentiated carcinoma with neuroendocrine differentiation showing aggressive clinical behavior. We herein report a case of gallbladder LCNEC, which was difficult to differentiate from poorly differentiated adenocarcinoma. An imprint cytology was very useful for the final diagnosis in this case. A 56-year-old male with left exophthalmos was admitted to the hospital. Radiological examinations revealed the presence of a left gallbladder tumor with orbital metastasis. The histological diagnosis was poorly differentiated adenocarcinoma, and intensive chemoradiotherapy was administered. Unfortunately, the patient died of extensive metastases 36 months after the initial onset of symptoms. An autopsy revealed a tumor mass in the gallbladder associated with multiple liver and peritoneal metastases. Imprint cytology of the main tumor revealed cytological features of LCNEC, and additional histological examinations confirmed this diagnosis. Although performing a histological examination is important for making a final diagnosis, imprint cytology is a powerful tool for differential diagnosis of LCNEC, especially in patients with carcinoma with poor differentiation. J. Med. Invest. 60: 149-153, February, 2013.

[153]

TÍTULO / TITLE: - Neuroendocrine neoplasms of the GI tract: the role of cytotoxic chemotherapy.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Expert Rev Anticancer Ther. 2013 Apr;13(4):451-9. doi: 10.1586/era.13.22.

●●Enlace al texto completo (gratis o de pago) [1586/era.13.22](#)

AUTORES / AUTHORS: - Khasraw M; Yap SY; Ananda S

INSTITUCIÓN / INSTITUTION: - Andrew Love Cancer Centre, Geelong Hospital, Geelong, VIC, Australia. m.khasraw@deakin.edu.au

RESUMEN / SUMMARY: - Neuroendocrine neoplasms (NENs) comprise a heterogeneous group of neoplasms derived from peptide- and amine-secreting cells of the neuroendocrine system. NENs commonly arise in the GI tract but can arise in most organs of the body. NENs in different organs share many common pathologic features. Although the incidence of NENs is not high, the prevalence is not low because many patients may live relatively long periods without major symptoms from the disease. While many of these tumors lead an indolent clinical course, they constitute a therapeutic challenge when they progress, metastasize and become symptomatic. Treatment requires a multidisciplinary approach including cytotoxic chemotherapy. Almost all clinical trials investigating cytotoxic chemotherapy in NENs are small single-arm studies

and guidelines are derived from expert opinion and from extrapolating results from small cell lung cancer studies. This article briefly reviews NENs before focusing on reviewing data on the role of cytotoxic chemotherapy studies in NENs.

[154]

TÍTULO / TITLE: - Prognostic value of Pheochromocytoma of the adrenal gland scaled score (Pass score) tests to separate benign from malignant neoplasms.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Tunis Med. 2013 Mar;91(3):209-15.

AUTORES / AUTHORS: - Mlika M; Kourda N; Zorgati MM; Bahri S; Ben Ammar S; Zermani R

RESUMEN / SUMMARY: - Background: Differentiating malignant from benign pheochromocytoma has been challenging when based on histologic features. This is due to the definition of malignant pheochromocytoma which are defined by the presence of metastases. A PASS score was developed and according to many authors, a PASS score ≥ 4 identified potentially malignant tumors. aim: To assess the prognostic value of PASS score in differentiating benign pheochromocytomas from malignant ones. methods: The records of 11 patients with tumors diagnosed as "pheochromocytoma" were identified from 1970 to 2010 in the files of the pathology, intern medicine and biochemistry departments of the Charles Nicolle hospital and Pasteur Institute. Receiver operating characteristics (ROC) curve analysis was performed to evaluate the diagnostic performance of PASS. The logistic model was developed using the 11 predictive variables. Its performance was evaluated by calculating the area under the ROC curve and comparing it with that of the PASS. results: In benign tumors, The PASS score was < 4 in 3 cases and ≥ 4 in 6 cases. In malignant tumors, the PASS score was ≥ 4 in both cases. According to the ROC curve analysis, a PASS equal or superior to 4 identifies malignant pheochromocytoma with a sensitivity of 50% and a specificity of 45%. Conclusion: I think that PASS score, despite its low sensitivity, may help to reserve the more aggressive treatment and narrow follow up for potentially malignant tumors. Widespread of this called score with complete clinical data will help to validate these findings and to add other prognostic factors of value that could be a part of this scaled score such as immunohistochemical findings.

[155]

TÍTULO / TITLE: - Lessons learned and questions unanswered from use of multitargeted kinase inhibitors in medullary thyroid cancer.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Oral Oncol. 2013 Apr 9. pii: S1368-8375(13)00518-6. doi: 10.1016/j.oraloncology.2013.03.442.

●●Enlace al texto completo (gratis o de pago)

1016/j.oraloncology.2013.03.442

AUTORES / AUTHORS: - Sherman SI

INSTITUCIÓN / INSTITUTION: - Department of Endocrine Neoplasia and Hormonal Disorders, University of Texas M.D. Anderson Cancer Center, 1400 Pressler Street, Unit 1461, Houston, TX 77030, USA. Electronic address: sisherma@mdanderson.org.

RESUMEN / SUMMARY: - **OBJECTIVES:** To review studies of novel multitargeted kinase inhibitors studied in patients with medullary thyroid cancer (MTC). **MATERIALS AND METHODS:** Search of relevant references in PubMed and Google Scholar on “chemotherapy” and “medullary thyroid cancer”. **RESULTS:** Multitargeted kinase inhibitors have revolutionized the role of chemotherapy for progressive MTC, providing for the first time tolerable therapeutic options that can improve outcomes in patients with progressive disease. Drugs thought to inhibit the RET kinase have advanced the furthest for this disease, but these agents also target the VEGF receptor along with other kinases that may be relevant to both beneficial and adverse effects. Vandetanib improved progression-free survival from 19.3 to 30.5 months compared with placebo in patients with metastatic disease, whereas cabozantinib improved progression-free survival from 4.0 months to 11.2 months in a population with more aggressive disease. However, “cure” remains elusive, adverse events frequent, and exactly how such “targeted” agents actually function within MTC remains unclear. **CONCLUSIONS:** New approaches to clinical trial design and the preclinical development of targeted agents may be required to optimize the combination of maximum efficacy with minimal toxicity for patients with metastatic MTC.

[156]

TÍTULO / TITLE: - Linking inflammation and neuroendocrine differentiation: the role of macrophage migration inhibitory factor-mediated signaling in prostate cancer.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Endocr Relat Cancer. 2013 May 21;20(3):C1-4. doi: 10.1530/ERC-13-0133. Print 2013 Jun.

●●Enlace al texto completo (gratis o de pago) 1530/ERC-13-0133

AUTORES / AUTHORS: - Savoy RM; Ghosh PM

INSTITUCIÓN / INSTITUTION: - Departments of Urology Biochemistry and Molecular Medicine, University of California Davis, Sacramento, California, USA VA Northern California Health Care System, Sacramento, California, USA.

RESUMEN / SUMMARY: - A new paper by Tawadros et al. in Endocrine-Related Cancer demonstrates a link between macrophage migration inhibitory factor and neuroendocrine differentiation in prostate cancer. This paper may have implications in explaining the effect of prostatitis and chronic inflammation on the development of aggressive prostate cancer.

[157]

TÍTULO / TITLE: - Corticotropin secreting bronchial carcinoid diagnosed after 22 years.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Tunis Med. 2013 May;91(5):364-5.

AUTORES / AUTHORS: - Hadj Ali S; Chihaoui M; Kanoun F; Lamine F; Ayadi A; El Mezni F; Kchir MN; Kilani T; Slimane H

[158]

TÍTULO / TITLE: - SDH Mutations Establish a Hypermethylator Phenotype in Paraganglioma.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Cancer Cell. 2013 May 21. pii: S1535-6108(13)00183-9. doi: 10.1016/j.ccr.2013.04.018.

●●Enlace al texto completo (gratis o de pago) 1016/j.ccr.2013.04.018

AUTORES / AUTHORS: - Letouze E; Martinelli C; Lorient C; Burnichon N; Abermil N; Ottolenghi C; Janin M; Menara M; Nguyen AT; Benit P; Buffet A; Marcaillou C; Bertherat J; Amar L; Rustin P; De Reynies A; Gimenez-Roqueplo AP; Favier J

INSTITUCIÓN / INSTITUTION: - Programme Cartes d'Identite des Tumeurs, Ligue Nationale Contre Le Cancer, 75013 Paris, France.

RESUMEN / SUMMARY: - Paragangliomas are neuroendocrine tumors frequently associated with mutations in RET, NF1, VHL, and succinate dehydrogenase (SDHx) genes. Methylome analysis of a large paraganglioma cohort identified three stable clusters, associated with distinct clinical features and mutational status. SDHx-related tumors displayed a hypermethylator phenotype, associated with downregulation of key genes involved in neuroendocrine differentiation. Succinate accumulation in SDH-deficient mouse chromaffin cells led to DNA hypermethylation by inhibition of 2-OG-dependent histone and DNA demethylases and established a migratory phenotype reversed by decitabine treatment. Epigenetic silencing was particularly severe in SDHB-mutated tumors, potentially explaining their malignancy. Finally, inactivating FH mutations were identified in the only hypermethylated tumor without SDHx mutations. These findings emphasize the interplay between the Krebs cycle, epigenomic changes, and cancer.

[159]

TÍTULO / TITLE: - Biochemical markers for gastroenteropancreatic neuroendocrine tumours (GEP-NETs).

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Best Pract Res Clin Gastroenterol. 2012 Dec;26(6):791-802. doi: 10.1016/j.bpg.2012.12.006.

●●Enlace al texto completo (gratis o de pago) 1016/j.bpg.2012.12.006

AUTORES / AUTHORS: - Kanakis G; Kaltsas G

INSTITUCIÓN / INSTITUTION: - University of Athens Medical School, Athens, Greece. geokan@endo.gr

RESUMEN / SUMMARY: - Biochemical markers are applied in gastroenteropancreatic neuroendocrine tumours (GEP-NETs) for diagnostic, prognostic or predictive purposes. Chromogranin A is the most important general marker and it is recommended to be measured in every patient with a suspected NET, whereas Neuron Specific Enolase is elevated mainly in poorly differentiated NETs. Pancreatic Polypeptide is used in the diagnosis of pancreatic non-functioning NETs, whereas Chorionic Gonadotrophin has an adjunctive role. In the case of functioning tumours, specific markers should be sought and monitored during follow up. Endogenous hyperinsulinemia is suggested in the presence of non-suppressible insulin and proinsulin levels during hypoglycemia, whereas high fasting or stimulated gastrin levels along with elevated gastric acid output are diagnostic for the Zollinger-Ellison syndrome. Glucagon, vasoactive intestinal polypeptide (VIP) and somatostatin are markers for glucagonoma, VIP-oma and somatostatinoma syndromes respectively. In case of ectopic paraneoplastic syndrome, the relevant hormone serves as a diagnostic and prognostic marker.

[160]

TÍTULO / TITLE: - Early Diffuse Coronary Artery Spasm After Heart Valve Surgery in the Carcinoid Syndrome.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - J Card Surg. 2013 May 27. doi: 10.1111/jocs.12138.

●●Enlace al texto completo (gratis o de pago) 1111/jocs.12138

AUTORES / AUTHORS: - Anselmi A; Corbineau H; Boulmier D; Ruggieri VG

INSTITUCIÓN / INSTITUTION: - Division of Thoracic and Cardiovascular Surgery, Pontchaillou University Hospital, Rennes, France.

RESUMEN / SUMMARY: - BACKGROUND: Coronary spasm has been described in the context of carcinoid heart disease, but few information are available over perioperative coronary spasm. METHODS: We describe the case of a patient developing severe spasm of the entire coronary tree immediately after tricuspid

valve surgery for carcinoid heart disease, leading to severe myocardial failure.
RESULTS: The spasm was diagnosed by coronary angiography and resolved by intracoronary injection of trinitrine. We present the angiographic features of simultaneous spasm of all coronary arteries and of its evolution.
CONCLUSION: The present case illustrates the likelihood of perioperative spasm of the entire coronary tree in carcinoid heart disease, and the usefulness of coronary angiography for both diagnosis and treatment.

[161]

TÍTULO / TITLE: - Intervention in gastro-enteropancreatic neuroendocrine tumours.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Best Pract Res Clin Gastroenterol. 2012 Dec;26(6):855-65. doi: 10.1016/j.bpg.2013.01.008.

●●Enlace al texto completo (gratis o de pago) 1016/j.bpg.2013.01.008

AUTORES / AUTHORS: - Baudin E; Planchard D; Scoazec JY; Guigay J; Dromain C; Hadoux J; Debaere T; Elias D; Ducreux M

INSTITUCIÓN / INSTITUTION: - Department of Nuclear Medicine and Endocrine Oncology, Institut Gustave Roussy, 39 rue Camille Desmoulins, 94805 Villejuif Cedex, France. baudin@igr.fr

RESUMEN / SUMMARY: - Neuroendocrine tumours require dedicated interventions to control their capacity to secrete hormones but also, antitumour growth strategies. Recommendations for early interventions in NET include the management of hormone-related symptoms and poorly differentiated neuroendocrine carcinomas. In contrast, prognostic heterogeneity is a key feature of well differentiated NET that complexified the antitumour strategy whatever the stage in this subgroup of tumour. In this review, timely therapeutic interventions to control hormone-related symptoms and tumour growth in GEP NET patients are discussed. The necessity of controlling hormone-related symptoms as the first step of any strategy affects also the tumour growth control strategy. In the absence of cure at the metastatic stage, progresses are expected in the recognition of well differentiated NET subgroups that display either excellent or poor prognosis.

[162]

TÍTULO / TITLE: - Epidemiology of gastroenteropancreatic neuroendocrine tumours.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Best Pract Res Clin Gastroenterol. 2012 Dec;26(6):691-703. doi: 10.1016/j.bpg.2013.01.006.

●●Enlace al texto completo (gratis o de pago) 1016/j.bpg.2013.01.006

AUTORES / AUTHORS: - Fraenkel M; Kim MK; Faggiano A; Valk GD

INSTITUCIÓN / INSTITUTION: - Endocrinology Unit, Soroka University Medical Center and the Faculty of Health Sciences, Ben-Gurion University of the Negev, Yitzchag Rager, Beer Sheva, Israel. meravfra@gmail.com

RESUMEN / SUMMARY: - Gastroenteropancreatic neuroendocrine tumours are a heterogeneous group of tumours arising from diffuse endocrine cells, causing unique clinical syndromes. These tumours, formerly named carcinoid, can involve any part of the gastrointestinal tract and the endocrine pancreas and have a wide range of malignant potential: from benign to poorly differentiated tumours. In this review we will summarize the data available on the epidemiology of gastroenteropancreatic tumours as it is reported from around the world. This includes annual incidence rates at the various anatomic sites, and trends in incidence rates with time. In addition age and stage at presentation, gender and racial differences and finally prognosis and survival were collected when reported.

[163]

TÍTULO / TITLE: - Multiple endocrine neoplasia type 2.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Front Horm Res. 2013;41:16-29. doi: 10.1159/000345667. Epub 2013 Mar 19.

●●Enlace al texto completo (gratis o de pago) [1159/000345667](#)

AUTORES / AUTHORS: - Lodish M

INSTITUCIÓN / INSTITUTION: - Section on Endocrinology and Genetics, Eunice Kennedy Shriver National Institute of Child Health and Human Development, National Institutes of Health, Bethesda, Md., USA.

RESUMEN / SUMMARY: - Multiple endocrine neoplasia type 2 (MEN2) is an autosomal-dominant cancer syndrome characterized by variable penetrance of medullary thyroid carcinoma (MTC), pheochromocytoma (PHEO), and primary hyperparathyroidism (PHPT). MEN2 consists of two clinical subtypes, MEN2A and MEN2B. Familial medullary thyroid cancer is now viewed as a phenotypic variant of MEN2A with decreased penetrance for PHEO and PHPT rather than a distinct entity. All subtypes are caused by gain-of-function mutations of the RET proto-oncogene. Genotype-phenotype correlations exist that help predict the presence of other associated endocrine neoplasms as well as the timing of thyroid cancer development. Recognition of the clinical entity in individuals and families at risk of harboring a germline RET mutation is crucial for the management and prevention of associated malignancies. Recent guidelines released by the American Thyroid Association regarding the management of MTC will be summarized in this chapter.

[164]

TÍTULO / TITLE: - Lung carcinoid with pulmonary vein and left atrial neoplastic thrombus.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - ANZ J Surg. 2013 Apr;83(4):289-90. doi: 10.1111/ans.12081.

●●Enlace al texto completo (gratis o de pago) [1111/ans.12081](#)

AUTORES / AUTHORS: - Anile M; Mazzei G; Diso D; Patella M; Russo E; Torromeo C; Vitolo D; Venuta F

INSTITUCIÓN / INSTITUTION: - Department of Thoracic Surgery, University of Rome Sapienza, Policlinico Umberto I, Rome, Italy.

[165]

TÍTULO / TITLE: - Multiple endocrine neoplasia type 1.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Front Horm Res. 2013;41:1-15. doi: 10.1159/000345666. Epub 2013 Mar 19.

●●Enlace al texto completo (gratis o de pago) [1159/000345666](#)

AUTORES / AUTHORS: - Agarwal SK

RESUMEN / SUMMARY: - Multiple endocrine neoplasia type 1 (MEN1) is an autosomal-dominant tumor syndrome characterized by the occurrence of tumors in multiple endocrine tissues and nonendocrine tissues. The three main endocrine tissues most frequently affected by tumors are parathyroid (95%), enteropancreatic neuroendocrine (50%) and anterior pituitary (40%). Tumors are caused by a heterozygous germline-inactivating mutation in the MEN1 gene (1st hit) followed by somatic inactivating mutation or loss of the normal copy of the gene (2nd hit), leading to complete loss of function of the encoded protein menin. Most of the disease features and tumors are recapitulated in mouse models with heterozygous germline loss of the Men1 gene. Also, tissue-specific tumors are observed in mouse models with homozygous somatic loss of the Men1 gene specifically in MEN1-associated endocrine tissues. Hence, mouse models could serve as possible surrogates for studying MEN1 and related states. To gain insights into MEN1 pathophysiology, menin-interacting partners and pathways have been identified to investigate its tumor suppressor and other functions. Also, the 3D crystal structure of menin has been deciphered which could be useful to reveal the relevance of MEN1 gene mutations and menin's interactions. This chapter covers clinical, genetic and basic findings about the MEN1 syndrome, MEN1 gene and its product protein menin.

[166]

TÍTULO / TITLE: - Shortness: an unknown phenotype of multiple endocrine neoplasia type 1.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Eur J Endocrinol. 2013 May 9.

●●Enlace al texto completo (gratis o de pago) [1530/EJE-13-0126](#)

AUTORES / AUTHORS: - Lopez CL; Langer P; Waldmann J; Fendrich V; Sitter H; Nies C; Bartsch DK

INSTITUCIÓN / INSTITUTION: - C Lopez, Department of Surgery, Philipps University Marburg, Marburg, 35043, Germany.

RESUMEN / SUMMARY: - **OBJECTIVE:** Observation of shortness among female participants of a REGULAR SCREENING PROGRAM IN MEN1 PATIENTS HAS RISED THE QUESTION,IF SHORTNESS REPRESENTS A PHENOTYPICAL CHARACTERISTIC OF THE DISEASE.**METHODS:** The body height, measured in cm, of genetically confirmed MEN1 patients at the time of diagnosis was compared to both, the body height of unaffected relatives (parents, siblings, children), the midparental body height and the age-matched German population. Univariate analysis of clinical variables were tested by t test, Mann-Whitney U test and ANOVA as appropriate, multivariate analysis was performed as logistic regression analysis. P-values <0.05 were considered statistically significant. **RESULTS:** The mean body height of 22 female MEN1 patients (mean age 33.5years) was 161 +5cm, and thus significantly smaller than the body heights of unaffected female relatives (mean 165.5 +7.3cm, p=0.027), the age-matched German female population (mean 167cm, p=0.0001) and mid parent height (177.5cm, p<0.0001). The mean body heights of 24 male MEN1 patients (mean age 34.8 years) was also smaller (177+6.5cm) than the average body heights of German males in this age group (180cm, p=0.031) and tended to be smaller than that of unaffected male relatives (178.5 +5.8cm, p= 0.0915) and the midparental body height (177.5cm, p=0.124). **CONCLUSIONS:** Small body height is a yet unrecognized phenotypic characteristic of MEN1 patients, especially in women. The mechanisms behind this phenotypic characteristic warrants further investigation.

[167]

TÍTULO / TITLE: - Pancreatic glucagonoma with pancreatic calcification.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Pancreatology. 2013 May-Jun;13(3):327-9. doi: 10.1016/j.pan.2012.11.308. Epub 2012 Nov 23.

●●Enlace al texto completo (gratis o de pago) [1016/j.pan.2012.11.308](#)

AUTORES / AUTHORS: - Gupta RA; Udwadia FE; Agrawal P; Doctor N

INSTITUCIÓN / INSTITUTION: - Department of Surgical Gastroenterology, Jaslok Hospital and Research Centre, Mumbai, Maharashtra, India.

RESUMEN / SUMMARY: - BACKGROUND: Glucagonoma is an uncommon type of pancreatic neuroendocrine tumor [NET] which is characterized by diabetes mellitus, necrolytic migratory erythema, depression and deep vein thrombosis. The typical rash is often misdiagnosed and the diagnosis is delayed by 7-8 years. Pancreatic NETs and other pancreatic tumors are known to show calcifications within the tumor but calcification of the remaining normal pancreas is very uncommon. It occurs when there is ductal obstruction leading to acute or chronic pancreatitis. CASE REPORT: We present a case of glucagonoma with coexistent pancreatic calcification. CONCLUSION: Glucagonoma should be suspected in a diabetic patient with migratory rash. Pancreatic tumor should be suspected in patient with idiopathic focal pancreatitis.

[168]

TÍTULO / TITLE: - A 9 Years Boy with MEN-2B Variant of Medullary Thyroid Carcinoma.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Mymensingh Med J. 2013 Apr;22(2):406-9.

AUTORES / AUTHORS: - Sattar MA; Hadi HI; Ekramuddoula FM; Hasanuzzaman SM

INSTITUCIÓN / INSTITUTION: - Dr Md Abdus Sattar, Associate Professor, Department of Otolaryngology-Head & Neck Surgery, Bangabandhu Sheikh Mujib Medical University, Shahbagh, Dhaka, Bangladesh.

RESUMEN / SUMMARY: - To highlight a rare disease like multiple endocrine neoplasia (MEN)-2B variant of medullary thyroid carcinoma and to optimize the management option in such cases, we present a nine year old boy with thyroid swelling, cervical lymphadenopathy and thick lips. His calcitonin level was raised. Investigation's results of the boy were as following fine needle aspiration cytology (FNAC) was medullary carcinoma of thyroid, preoperative calcitonin was >2000pg/ml, post operative histopathological report was medullary carcinoma. Total thyroidectomy with aggressive initial neck surgery may reduce the recurrence and increase better prognosis and survival rate. Calcitonin is used as diagnostic and follow-up marker.

[169]

TÍTULO / TITLE: - Primary cardiac paraganglioma arising from interatrial septum.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - J Card Surg. 2013 May;28(3):274-5. doi: 10.1111/jocs.12103.

●●Enlace al texto completo (gratis o de pago) 1111/jocs.12103

AUTORES / AUTHORS: - Wang Y; Xiao Y; Wang X

INSTITUCIÓN / INSTITUTION: - Department of Cardiovascular Surgery, Xinqiao Hospital, The Third Military Medical University, Chongqing, China.

RESUMEN / SUMMARY: - doi: 10.1111/jocs.12103 (J Card Surg 2013;28:274-275).

[170]

TÍTULO / TITLE: - Primary paraganglioma in the facial nerve canal.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Auris Nasus Larynx. 2013 May 18. pii: S0385-8146(13)00114-4. doi: 10.1016/j.anl.2013.04.007.

●●Enlace al texto completo (gratis o de pago) 1016/j.anl.2013.04.007

AUTORES / AUTHORS: - Takahashi K; Yamamoto Y; Ohshima S; Morita Y; Takahashi S

INSTITUCIÓN / INSTITUTION: - Department of Otolaryngology, Niigata University, Faculty of Medicine, Japan. Electronic address: kuniyuki@med.niigata-u.ac.jp.

RESUMEN / SUMMARY: - OBJECTIVE: To describe primary paraganglioma in the facial nerve canal and discuss the characteristics of facial nerve paraganglioma in contrast with other tumors. CASE REPORT: A 23-year-old man developed gradually progressive right facial palsy as the initial symptom. One year later, he exhibited hearing loss without tinnitus in his right ear. CT demonstrated an enlarged facial nerve canal with irregular bony erosion of the circumference. MRI showed a well-enhanced heterogeneous mass with hypo-intensity spots inside it. During surgery, a blood-rich tumor was observed along the facial nerve: however, extensive bleeding interfered with tumor removal. The surgical specimen demonstrated paraganglioma. The tumor was completely removed in the second surgery in combination with arterial embolization. Facial nerve function was reconstructed with a free muscle flap more than one year following resection. CONCLUSION: Because paraganglioma is a blood-rich tumor, it is important to perform angiography and embolization. If preoperative facial nerve palsy is demonstrated, careful management of facial nerve function is needed. Paraganglioma must be considered in the differential diagnosis of a facial nerve tumor.

[171]

TÍTULO / TITLE: - Neuroendocrine tumours of the small intestine.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Best Pract Res Clin Gastroenterol. 2012 Dec;26(6):755-73. doi: 10.1016/j.bpg.2012.12.002.

●●Enlace al texto completo (gratis o de pago) 1016/j.bpg.2012.12.002

AUTORES / AUTHORS: - Strosberg J

INSTITUCIÓN / INSTITUTION: - H. Lee Moffitt Cancer Center and Research Institute, Dept. of GI Oncology, 12902 Magnolia Dr., Tampa, FL 33612, USA.
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RESUMEN / SUMMARY: - The prevalence of intestinal neuroendocrine tumours, also known as carcinoid tumours, has increased significantly over the past three decades. Tumours of the distal small intestine (midgut) are often indolent, but are characterized by a high potential to metastasize to the small-bowel mesentery and liver. Patients with distant metastases are prone to development of the carcinoid syndrome, a constellation of symptoms which includes flushing, diarrhoea, and valvular heart disease. The carcinoid syndrome is caused by secretion of serotonin and other vasoactive substances into the systemic circulation. Treatment options for metastatic intestinal NETs have expanded in recent years. Of particular importance has been the development of somatostatin-analogue therapies. Somatostatin analogues were originally introduced for palliation of the carcinoid syndrome; however recent clinical trials have demonstrated that they can exert an inhibitory effect on tumour growth. Other novel agents targeting the VEGF and mTOR pathways have recently been evaluated in phase III trials, however their role in the management of small-intestinal NETs remains controversial. This article examines the biological characteristics of small intestinal NETs, summarizes current guidelines on classification, staging and grading, and reviews developments in locoregional and systemic therapy.

[172]

TÍTULO / TITLE: - mTOR, p70S6K, AKT and ERK1/2 levels predict sensitivity to mTOR and PI3K/mTOR inhibitors in human bronchial carcinoids.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Endocr Relat Cancer. 2013 May 7.

●●Enlace al texto completo (gratis o de pago) [1530/ERC-13-0042](#)

AUTORES / AUTHORS: - Gagliano T; Bellio M; Gentilin E; Mole D; Tagliati F; Schiavon M; Cavallesco NG; Andriolo LG; Rea F; Degli Uberti EC; Zatelli MC

INSTITUCIÓN / INSTITUTION: - T Gagliano, Department of Medical Sciences, University of Ferrara, Ferrara, Italy.

RESUMEN / SUMMARY: - Bronchial carcinoids (BC) are rare neuroendocrine tumors that are still orphan of medical treatment. Human BC primary cultures may display resistance to Everolimus, an inhibitor of the mammalian target of rapamycin (mTOR), in terms of cell viability reduction. Our aim is to assess whether the novel dual PI3K/mTOR inhibitor, NVP-BEZ235, may be effective in Everolimus-resistant human BC tissues and cell lines. In addition, we search for possible markers of mTOR inhibitors efficacy, that may help in identifying the patients that may benefit from mTOR inhibitors treatment, sparing them from ineffective therapy. We found that NVP-BEZ235 is twice as potent as

Everolimus in reducing cell viability and activating apoptosis in human BC tissues that display sensitivity to mTOR inhibitors, but is not effective in Everolimus-resistant BC tissues and cell lines, that by-pass cyclin D1 down-regulation and escape G0/G1 blockade. Rebound AKT activation was not observed in response to treatment with either mTOR inhibitor in 'resistant' BC cells. In addition to total mTOR levels, putative markers of BC sensitivity to mTOR inhibitors are represented by AKT, p70S6K and ERK1/2 protein levels. Finally, we validated these markers in an independent BC group. These data indicate that the dual PI3K/mTOR inhibitor NVP-BEZ235 is more potent than Everolimus in reducing human BC cell proliferation. 'Resistant' cells display lower levels of mTOR, p70S6K, AKT and ERK1/2, indicating that these proteins may be useful as predictive markers of resistance to mTOR and PI3K/mTOR inhibitors in human BC.

[173]

TÍTULO / TITLE: - Gastric and duodenal neuroendocrine tumours.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Best Pract Res Clin Gastroenterol. 2012 Dec;26(6):719-35. doi: 10.1016/j.bpg.2013.01.002.

●●Enlace al texto completo (gratis o de pago) 1016/j.bpg.2013.01.002

AUTORES / AUTHORS: - O'Toole D; Delle Fave G; Jensen RT

INSTITUCIÓN / INSTITUTION: - Department of Gastroenterology and Clinical Medicine, St James's Hospital and Trinity College, Dublin, Ireland.

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RESUMEN / SUMMARY: - Gastric neuroendocrine neoplasms (NENs) are increasing in frequency and have a varied spectrum with regard to histology, clinicopathologic background, stage, and prognosis. They are usually discovered incidentally, are for the most part benign and are associated with hypergastrinaemia (secondary either to chronic atrophic gastritis or rarely Zollinger-Ellison syndrome; types 1 and 2, respectively) or more rarely sporadic type 3. Applications of recent staging and grading systems - namely using Ki-67 proliferative indices - (from ENETS and WHO 2010) can be particularly helpful in further categorising these tumours. The natural history of Type 1 gastric carcinoids is generally (>95%) favourable and simple surveillance is usually recommended for small (<1 cm) T1 tumours, with local (endoscopic or surgical) resection for larger lesions. Other potential therapies such as somatostatin analogues and gastrin receptor antagonists may offer newer therapeutic possibilities. Rarely, gastric NENs have a malignant course and this is usually confined to Type 2 and especially Type 3 tumours; the latter mimic the biological course of gastric adenocarcinoma and require radical oncological therapies. Most duodenal NENs, apart from gastrinomas (that are not dealt with here) are sporadic and non functional. They are also increasing in frequency

probably due to incidental discovery at endoscopy or imaging for other reasons and this may account for their overall good prognosis. Peri-ampullary and ampullary NENs may have a more aggressive outcome and should be carefully appraised and treated (often with surgical resection).

[174]

TÍTULO / TITLE: - Endocrine tumors associated with neurofibromatosis type 1, peutz-jeghers syndrome and other familial neoplasia syndromes.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Front Horm Res. 2013;41:166-81. doi: 10.1159/000345676. Epub 2013 Mar 19.

●●Enlace al texto completo (gratis o de pago) [1159/000345676](#)

AUTORES / AUTHORS: - Kalkan E; Waguespack SG

RESUMEN / SUMMARY: - Endocrine tumors are a less common but important component of the clinical spectrum of a number of hereditary tumor syndromes such as neurofibromatosis type 1, Peutz-Jeghers syndrome, Beckwith-Wiedemann syndrome, the tuberous sclerosis complex, Li-Fraumeni syndrome, PTEN hamartoma tumor syndrome, and APC-associated polyposis. It is important to recognize the often unique clinical presentations of these tumors and possible strategies for presymptomatic screening and early diagnosis.

[175]

TÍTULO / TITLE: - Novel hereditary forms of pheochromocytomas and paragangliomas.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Front Horm Res. 2013;41:79-91. doi: 10.1159/000345671. Epub 2013 Mar 19.

●●Enlace al texto completo (gratis o de pago) [1159/000345671](#)

AUTORES / AUTHORS: - Dahia PL

INSTITUCIÓN / INSTITUTION: - Division of Hematology and Medical Oncology, Department of Medicine, San Antonio, Tex., USA.

RESUMEN / SUMMARY: - Pheochromocytomas and paragangliomas are catecholamine-secreting tumors of neural crest origin that arise from the adrenal medulla or extra-adrenal sympathetic paraganglia, respectively. Over the last decade, the extensive genetic heterogeneity of these tumors came to light with the identification of multiple susceptibility genes. These mutations account for at least one-third of pheochromocytomas and paragangliomas, the highest inheritable proportion of any known human tumor. This chapter will present an overview of genetic and molecular features of the most recently identified hereditary forms of pheochromocytoma and paraganglioma: those caused by mutations in five genes of the succinate dehydrogenase (SDH)

complex, the transmembrane-encoding gene TMEM127 and the MYC-binding partner, MAX. Initial genotype-phenotype correlations, as well as emerging functional data, have aligned the new mutants either with defects in hypoxic-angiogenic signaling (SDH-related) or kinase receptor/mTOR pathways (TMEM127 and MAX). These findings, in combination with those of the more well-established syndromes, have been relevant for guiding clinical follow-up. The progress of recent years in understanding the pathogenesis of pheochromocytomas and paragangliomas is expected to continue to improve patient screening and to become, in the long term, the catalyst for development of new therapeutic options for surgically untreatable tumors.

[176]

TÍTULO / TITLE: - Multiple endocrine neoplasia type 4.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Front Horm Res. 2013;41:63-78. doi: 10.1159/000345670. Epub 2013 Mar 19.

●●Enlace al texto completo (gratis o de pago) [1159/000345670](#)

AUTORES / AUTHORS: - Lee M; Pellegata NS

INSTITUCIÓN / INSTITUTION: - Institute of Pathology, Helmholtz Zentrum Munchen, Neuherberg, Germany.

RESUMEN / SUMMARY: - A few years ago a novel multiple endocrine neoplasia syndrome, named multiple endocrine neoplasia type 4 (MEN4), was discovered thanks to studies conducted on a MEN syndrome in the rat (named MENX). The rat and the human syndromes are both caused by germline mutations in the Cdkn1b/CDKN1B gene, respectively. This gene encodes p27Kip1, a putative tumor suppressor which binds to and inhibits cyclin/cyclin-dependent kinase complexes, thereby preventing cell cycle progression. MEN4 patients carry heterozygous mutations at various residues of p27Kip1 and present with endocrine lesions mainly belonging to a MEN1-like spectrum: their most common phenotypic features are parathyroid and pituitary adenomas. Recently, germline mutations in p27kip1 were also identified in patients with a sporadic parathyroid disease presentation. In vitro functional analysis of several CDKN1B sequence changes identified in MEN4 patients detected impaired activity of the encoded p27Kip1 variant proteins (e.g. reduced expression, mislocalization or poor binding to interaction partners), thereby highlighting the characteristics of the protein which are critical for tumor suppression. Although the number of MEN4 patients is low, the discovery of this syndrome has demonstrated a novel role for CDKN1B as a tumor susceptibility gene for neuroendocrine tumors. Here, we review the clinical characteristics of the MEN4 syndrome and the molecular phenotype of the associated p27Kip1 mutations.

[177]

TÍTULO / TITLE: - Rare treatable limb girdle muscle disease.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - J Assoc Physicians India. 2012 Sep;60:62-5.

AUTORES / AUTHORS: - Chandra SR; Shenoy RK; Karthikeyan; Suresh K; Chithr P; Annapoorni CS

INSTITUCIÓN / INSTITUTION: - NIMHANS, Bangalore.

RESUMEN / SUMMARY: - We report two cases of Limb Girdle pattern of muscle weakness caused by hyperparathyroidism due to parathyroid adenoma. It can be easily missed as early symptoms are non specific but once diagnosed it is easily treatable and complete recovery occurs over a period of time.

[178]

TÍTULO / TITLE: - Endosonographic and cyst fluid characteristics of cystic pancreatic neuroendocrine tumours: A multicentre case series.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Dig Liver Dis. 2013 Apr 9. pii: S1590-8658(13)00096-0. doi: 10.1016/j.dld.2013.02.021.

●●Enlace al texto completo (gratis o de pago) 1016/j.dld.2013.02.021

AUTORES / AUTHORS: - Ho HC; Eloubeidi MA; Siddiqui UD; Brugge WR; Rossi F; Bounds BW; Aslanian HR

INSTITUCIÓN / INSTITUTION: - Section of Digestive Diseases, Yale University School of Medicine, New Haven, CT, USA. Electronic address:

henry.ho@yale.edu.

RESUMEN / SUMMARY: - BACKGROUND: Pancreatic neuroendocrine tumours are uncommon neoplasms which may rarely be cystic. Differentiation from other more common cystic neoplasms may be difficult. AIMS: To describe the morphologic, cytologic, and cyst fluid characteristics of cystic pancreatic neuroendocrine tumours. METHODS: Retrospective analysis of consecutive patients referred for endosonographic evaluation of pancreatic cysts at four centres. RESULTS: 27 patients (12 males) with cystic pancreatic neuroendocrine tumours were identified. Prior to endosonography, this tumour was suspected in only 2 patients based on presenting symptoms (7.4%). The median cyst size was 35mm (range 8-80mm). Wall thickening was identified in 13 cases. The median carcinoembryonic antigen level was 1.25 (range 0.6-500). Fine needle aspiration cytology in 17 of 24 patients confirmed neuroendocrine tumour (71%). In 8 of 9 patients who had needle targeting of the cyst wall, cytology was consistent with neuroendocrine tumour (88.9%). 18 patients underwent surgical resection. CONCLUSIONS: Cystic pancreatic neuroendocrine tumour was rarely suspected, including by cross-sectional imaging. Wall thickening was identified in approximately half of cases on

endosonography. Cyst fluid was typically non-viscous with very low carcinoembryonic antigen levels. Targeting the wall during fine needle aspiration had a high diagnostic yield and should be performed.

[179]

TÍTULO / TITLE: - Complete remission of Merkel cell carcinoma treated with electrochemotherapy and etoposide.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - G Ital Dermatol Venereol. 2013 Jun;148(3):310-1.

AUTORES / AUTHORS: - Curatolo P; Rotunno R; Miraglia E; Mancini M; Calvieri S; Giustini S

INSTITUCIÓN / INSTITUTION: - La Sapienza University of Rome, Department of Dermatology, Policlinico Umberto I, Rome, Italy - rorot@libero.it.

[180]

TÍTULO / TITLE: - Diagnosis and localization of insulinoma in Thai patients: performance of endoscopic ultrasonography compared to computed tomography and magnetic resonance imaging.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - J Med Assoc Thai. 2013 Feb;96 Suppl 2:S187-93.

AUTORES / AUTHORS: - Pongprasobchai S; Lertwattanarak R; Pausawasdi N; Prachayakul V

INSTITUCIÓN / INSTITUTION: - Division of Gastroenterology, Department of Medicine, Siriraj Hospital, Bangkok, Thailand. supot.pon@mahidol.ac.th

RESUMEN / SUMMARY: - BACKGROUND: Endoscopic ultrasonography (EUS) has now been accepted as the most sensitive method to localize insulinoma. However the data in Thai patients is lacking and the diagnostic performances of EUS comparing to computed tomography (CT) and magnetic resonance imaging (MRI) is unknown. MATERIAL AND METHOD: Retrospective analysis of 19 patients with recurrent hypoglycemia suggestive of insulinoma who underwent EUS, CT and MRI for tumor localization during 2007 to 2012. Surgical pathology or long-term follow-up was used as gold standard. RESULTS: There were 14 patients with 15 insulinoma lesions and 5 patients without insulinoma (2 nesidioblastosis and 3 without lesion). EUS, CT and MRI were performed in 19, 11 and 10 patients, respectively. EUS could detect insulinoma with sensitivity, specificity, positive predictive value (PPV) and negative predictive value (NPV) of 93%, 80%, 93% and 80%, respectively. The corresponding performances for CT were 78%, 100%, 100%, 50% and MRI were 71%, 33%, 71%, 33%, respectively. In patients with positive CT subsequent EUS did not change diagnosis. However, EUS was able to detect insulinoma in 50% of patients with negative CT. On the other hand, in patients

with positive MRI, EUS changed and corrected the diagnosis of MRI in 29% and was able to detect insulinoma in 67% of patients with negative MRI. EUS, CT and MRI correctly localized insulinoma in 87%, 67% and 57%, respectively. The most common incorrect localization was between pancreatic body and tail. CONCLUSION: EUS has the best diagnostic performance in detection and localization of insulinoma. CT is less sensitive but very specific, therefore positive CT may preclude the need of EUS. MRI, however is less sensitive and specific than CT. Either positive or negative MRI may require further EUS.

[181]

TÍTULO / TITLE: - Clinicopathologic characteristics at diagnosis and the survival of patients with medullary breast carcinoma in China: a comparison with infiltrating ductal carcinoma-not otherwise specified.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - World J Surg Oncol. 2013 Apr 22;11:91. doi: 10.1186/1477-7819-11-91.

●●Enlace al texto completo (gratis o de pago) [1186/1477-7819-11-91](#)

AUTORES / AUTHORS: - Cao AY; He M; Huang L; Shao ZM; Di GH

INSTITUCIÓN / INSTITUTION: - Breast Cancer Institute, Cancer Centre/Cancer Institute; Department of Oncology, Shanghai Medical College, Fudan University, 270 Dong'an Road, Shanghai, 200032, People's Republic of China. didy@medmail.com.cn.

RESUMEN / SUMMARY: - BACKGROUND: Few studies have addressed the biological features of medullary breast carcinoma (MBC) in the context of clinical outcomes. We sought to compare the baseline demographics, standard pathologic factors and long-term clinical outcomes between MBC and infiltrating ductal carcinoma-not otherwise specified (IDC-NOS) using a large database. METHODS: A total of 2,202 cases with pure IDC-NOS and 188 cases with typical MBC meeting the inclusion criteria were identified. The clinical and biological features, the overall survival (OS) and recurrence/metastasis-free survival (RFS) were compared for both groups. RESULTS: There were a higher proportion of patients diagnosed prior to 40 years of age in the MBC group compared to the IDC-NOS group. MBC cases demonstrated less aggressive tumor features such as lower tumor stage, smaller tumor size and a lower proportion of nodal involvement than IDC-NOS; however, immunohistochemical staining revealed that MBC displayed the triple-negative phenotype more often than IDC-NOS cases (40.4% versus 26.2%; $P < 0.001$). Although the clinical behavior of MBC was not commensurate with its pathologic features, women diagnosed with MBC had a lower frequency of recurrence/metastasis ($P = 0.032$) and death ($P = 0.042$) than those with IDC-NOS, and the 10-year OS and RFS were significantly higher for MBC (91% and 74%) compared to IDC-NOS (81% and 64%). Moreover, multivariate analysis revealed that TNM stage

was a statistically significant factor for survival. CONCLUSIONS: MBC in Chinese women demonstrated less aggressive behavior and better prognosis than IDC-NOS. This favorable outcome was maintained after 10 years.

[182]

TÍTULO / TITLE: - Primary neuroendocrine lung tumor presenting with acute ileal obstruction. Case report.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - G Chir. 2013 Mar;34(3):78-81.

AUTORES / AUTHORS: - Li Destri G; Ferraro MJ; Vecchio G; Musumeci A; Calabrini M; Giarrizzo A

RESUMEN / SUMMARY: - The authors describe a clinical case of a patient with neuroendocrine carcinoma of the lung diagnosed after the onset of an intestinal obstruction from an ileal metastasis. A review of literature reveals that the incidence of symptomatic gastro-intestinal metastases from lung cancer has been estimated to be about 2-3% and is exceedingly rare that the intestinal symptoms may be the initial presentation of cancer of the lung. The authors emphasize the difficulty of preoperative diagnosis of gastro-intestinal metastases which is made, almost always, too late because of the lack of specific symptoms. In our case, on account of the computed tomography, we leaned towards the diagnosis of lymphoma because of the double mediastinal and abdominal localization. Furthermore, this diagnosis was supported by the fact that the pulmonary lesion did not have clear radiological features of a lung cancer. The prognosis is poor because once intestinal metastases occur, other metastatic sites, which would make surgery only a palliative measure, are already present. The review of the literature shows that the average survival rate of these patients is 136 days. In our case the patient survived 277 days.

[183]

TÍTULO / TITLE: - Genotype and tumor locus determine expression profile of pseudohypoxic pheochromocytomas and paragangliomas.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Neoplasia. 2013 Apr;15(4):435-47.

AUTORES / AUTHORS: - Shankavaram U; Fliedner SM; Elkahloun AG; Barb JJ; Munson PJ; Huynh TT; Matro JC; Turkova H; Linehan WM; Timmers HJ; Tischler AS; Powers JF; de Krijger R; Baysal BE; Takacova M; Pastorekova S; Gius D; Lehnert H; Camphausen K; Pacak K

INSTITUCIÓN / INSTITUTION: - Radiation Oncology Branch, National Cancer Institute, National Institutes of Health, Bethesda, MD 20892-1109, USA.

RESUMEN / SUMMARY: - Pheochromocytomas (PHEOs) and paragangliomas (PGLs) related to mutations in the mitochondrial succinate dehydrogenase (SDH) subunits A, B, C, and D, SDH complex assembly factor 2, and the von

Hippel-Lindau (VHL) genes share a pseudohypoxic expression profile. However, genotype-specific differences in expression have been emerging. Development of effective new therapies for distinctive manifestations, e.g., a high rate of malignancy in SDHB- or predisposition to multifocal PGLs in SDHD patients, mandates improved stratification. To identify mutation/location-related characteristics among pseudohypoxic PHEOs/PGLs, we used comprehensive microarray profiling (SDHB: n = 18, SDHD-abdominal/thoracic (AT): n = 6, SDHD-head/neck (HN): n = 8, VHL: n = 13). To avoid location-specific bias, typical adrenal medulla genes were derived from matched normal medullas and cortices (n = 8) for data normalization. Unsupervised analysis identified two dominant clusters, separating SDHB and SDHD-AT PHEOs/PGLs (cluster A) from VHL PHEOs and SDHD-HN PGLs (cluster B). Supervised analysis yielded 6937 highly predictive genes (misclassification error rate of 0.175). Enrichment analysis revealed that energy metabolism and inflammation/fibrosis-related genes were most pronouncedly changed in clusters A and B, respectively. A minimum subset of 40 classifiers was validated by quantitative real-time polymerase chain reaction (quantitative real-time polymerase chain reaction vs. microarray: r = 0.87). Expression of several individual classifiers was identified as characteristic for VHL and SDHD-HN PHEOs and PGLs. In the present study, we show for the first time that SDHD-HN PGLs share more features with VHL PHEOs than with SDHD-AT PGLs. The presented data suggest novel subclassification of pseudohypoxic PHEOs/PGLs and implies cluster-specific pathogenic mechanisms and treatment strategies.

[184]

TÍTULO / TITLE: - Long-term management of a patient with well-differentiated pulmonary neuroendocrine carcinoma: a case report.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Case Rep Oncol. 2013 Apr 6;6(1):209-15. doi: 10.1159/000350745. Print 2013 Jan.

●●Enlace al texto completo (gratis o de pago) [1159/000350745](#)

AUTORES / AUTHORS: - Beck T; Mantooth R

INSTITUCIÓN / INSTITUTION: - Highlands Oncology Group, Rogers, Ark., USA.

RESUMEN / SUMMARY: - Pulmonary neuroendocrine tumors (NET) are rare, and very few published reports have described the long-term treatment of patients with this disease. Current treatment options for patients with metastatic well-differentiated pulmonary NET are limited. This case report details the long-term treatment of a 62-year-old female patient with well-differentiated pulmonary NET and multiple liver metastases. The heavily pretreated patient achieved radiographic stability in measurable disease, improvement in nonmeasurable disease, and symptomatic improvement over 3 years while receiving the combination of everolimus and octreotide long-acting repeatable (LAR).

Treatment was well tolerated without mucositis, rash, or pneumonitis. This case report suggests that the combination of everolimus and octreotide LAR may be a novel treatment option for heavily pretreated patients with metastatic well-differentiated pulmonary NET, but these findings require further analysis in clinical trials.

[185]

TÍTULO / TITLE: - MIBG molecular imaging for evaluating response to chemotherapy in patients with malignant pheochromocytoma: preliminary results.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Cancer Imaging. 2013 Apr 15;13:155-61. doi: 10.1102/1470-7330.2013.0017.

●●Enlace al texto completo (gratis o de pago) [1102/1470-7330.2013.0017](#)

AUTORES / AUTHORS: - Maurea S; Fiumara G; Pellegrino T; Zampella E; Assante R; Mainenti P; Cuocolo A

INSTITUCIÓN / INSTITUTION: - Department of Advanced Biomedical Sciences, University Federico II, Napoli, Italy.

RESUMEN / SUMMARY: - Malignant pheochromocytomas respond to chemotherapy with a reduction in tumor size and catecholamine secretion. We investigated the usefulness of molecular imaging with meta-iodobenzylguanidine (MIBG) for evaluating the effects of chemotherapy in patients with malignant pheochromocytoma. Six patients were studied before and after 6 +/- 4 months of combination chemotherapy with cyclophosphamide, vincristine, and dacarbazine. Urinary catecholamines, metanephrines, and vanillylmandelic acid (VMA) levels were measured before and after chemotherapy. [(131)I]MIBG uptake was calculated for each tumor lesion on images before and after chemotherapy. An intensity ratio (IR) of abnormal to normal tissue count density was used to evaluate the change in lesion activity with therapy. Urinary catecholamines, metanephrines, and VMA significantly decreased with chemotherapy. MIBG uptake decreased in most lesions and the reduction in overall IR correlated with the reduction in urinary VMA. However, the change in individual lesions was variable and MIBG IR did not change or increased in a number of lesions. In conclusion, MIBG imaging is useful in the evaluation of patients with malignant pheochromocytoma who are receiving chemotherapy. It can provide not only a measure of overall effectiveness of treatment but also allows a lesion-by-lesion evaluation of the heterogeneity of response to chemotherapy.

[186]

TÍTULO / TITLE: - Merkel cell carcinoma: complete clinical remission associated with disease progression.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - JAMA. Acceso gratuito al texto completo.

●●Enlace a la Editora de la Revista <http://jama.ama-assn.org/search.dtl>

●●Cita: JAMA: <> Dermatol. 2013 Apr 1;149(4):501-2. doi: 10.1001/jamadermatol.2013.2596.

●●Enlace al texto completo (gratuito o de pago)

1001/jamadermatol.2013.2596

AUTORES / AUTHORS: - Bertolotti A; Conte H; Francois L; Dutriaux C; Ezzedine K; Melard P; Vergier B; Taieb A; Jouary T

[187]

TÍTULO / TITLE: - A case of pulmonary carcinoid tumour in a pregnant woman successfully treated with bronchoscopic (electrocautery) therapy.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - British Medical J (BMJ). Acceso gratuito al texto completo.

●●Enlace a la Editora de la Revista <http://bmj.com/search.dtl>

●●Cita: British Medical J. (BMJ): <> Case Rep. 2013 Apr 22;2013. pii: bcr2013009250. doi: 10.1136/bcr-2013-009250.

●●Enlace al texto completo (gratuito o de pago) 1136/bcr-2013-009250

AUTORES / AUTHORS: - Binesh F; Samet M; Bovanlu TR

INSTITUCIÓN / INSTITUTION: - Shahid Sadoughi University of Medical Sciences, Yazd, Iran. binesh44@yahoo.com

RESUMEN / SUMMARY: - We present an uncommon case of a carcinoid tumour of the bronchus that was diagnosed during pregnancy in a 28-year-old woman. The patient was admitted at the emergency department with massive haemoptysis. Owing to the patient's critical condition, she underwent urgent flexible bronchoscopy. Bleeding was controlled by local injection of 500 mg tranexamic acid and electrocautery. After the bleeding has stopped, multiple specimens were taken. Histological examination confirmed typical carcinoid tumour. Owing to repeated haemoptysis, she was treated with bronchoscopic (electrocautery) therapy, and, after delivery, she underwent pulmonary lobectomy. Only a few similar cases were found in the literature reporting bronchopulmonary carcinoid tumour during pregnancy and we could not find any similar case which was treated by electrocautery.

[188]

TÍTULO / TITLE: - Glomus tumours in the long finger and in the thumb of a young patient with neurofibromatosis-1 (Nf-1).

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - J Plast Surg Hand Surg. 2013 Jun;47(3):238-40. doi: 10.3109/2000656X.2012.754624. Epub 2013 Apr 3.

●●Enlace al texto completo (gratis o de pago)

[3109/2000656X.2012.754624](#)

AUTORES / AUTHORS: - Dahlin LB; Muller G; Anagnostaki L; Nordborg K

INSTITUCIÓN / INSTITUTION: - Department of Hand Surgery.

RESUMEN / SUMMARY: - Abstract Glomus tumours are rare in adults, and such tumours may be associated with neurofibromatosis-1 (Nf-1). Here we describe successful treatment of two glomus tumours initially in the long finger and, one year later, in the thumb in a 17-year young girl with Nf-1.

[189]

TÍTULO / TITLE: - Optimization of image reconstruction method for SPECT studies performed using [99mTc-EDDA/HYNIC] octreotate in patients with neuroendocrine tumors.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Nucl Med Rev Cent East Eur. 2013;16(1):9-16. doi: 10.5603/NMR.2013.0003.

●●Enlace al texto completo (gratis o de pago) [5603/NMR.2013.0003](#)

AUTORES / AUTHORS: - Sowa-Staszczak A; Lenda-Tracz W; Tomaszuk M; Glowa B; Hubalewska-Dydejczyk A

INSTITUCIÓN / INSTITUTION: - Department of Endocrinology, Jagiellonian University Medical College, Krakow, Poland. wioletta.tracz@gmail.com.

RESUMEN / SUMMARY: - BACKGROUND: Somatostatin receptor scintigraphy (SRS) is a useful tool in the assessment of GEP-NET (gastroenteropancreatic neuroendocrine tumor) patients. The choice of appropriate settings of image reconstruction parameters is crucial in interpretation of these images. The aim of the study was to investigate how the GEP NET lesion signal to noise ratio (TCS/TCB) depends on different reconstruction settings for Flash 3D software (Siemens). METHODS: SRS results of 76 randomly selected patients with confirmed GEP-NET were analyzed. For SPECT studies the data were acquired using standard clinical settings 3-4 h after the injection of 740 MBq 99mTc-[EDDA/HYNIC] octreotate. To obtain final images the OSEM 3D Flash reconstruction with different settings and FBP reconstruction were used. First, the TCS/TCB ratio in voxels was analyzed for different combinations of the number of subsets and the number of iterations of the OSEM 3D Flash reconstruction. Secondly, the same ratio was analyzed for different parameters of the Gaussian filter (with FWHM = 2-4 times greater from the pixel size). Also the influence of scatter correction on the TCS/TCB ratio was investigated. RESULTS: With increasing number of subsets and iterations, the increase of TCS/TCB ratio was observed. With increasing settings of Gauss [FWHM coefficient] filter, the decrease of TCS/TCB ratio was reported. The use of scatter correction slightly decreases the values of this ratio. CONCLUSIONS: OSEM algorithm provides a meaningfully better reconstruction of the SRS SPECT study as compared to the FBP technique. A high number of subsets

improves image quality (images are smoother). Increasing number of iterations gives a better contrast and the shapes of lesions and organs are sharper. The choice of reconstruction parameters is a compromise between image qualitative appearance and its quantitative accuracy and should not be modified when comparing multiple studies of the same patient.

[190]

TÍTULO / TITLE: - The identification of gut neuroendocrine tumor disease by multiple synchronous transcript analysis in blood.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - PLoS One. 2013 May 15;8(5):e63364. doi: 10.1371/journal.pone.0063364. Print 2013.

●●Enlace al texto completo (gratis o de pago)

[1371/journal.pone.0063364](#)

AUTORES / AUTHORS: - Modlin IM; Drozdov I; Kidd M

INSTITUCIÓN / INSTITUTION: - Department of Surgery, Yale University School of Medicine, New Haven, Connecticut, United States of America.

RESUMEN / SUMMARY: - Gastroenteropancreatic (GEP) neuroendocrine neoplasms (NENs) are increasing in both incidence and prevalence. A delay in correct diagnosis is common for these lesions. This reflects the absence of specific blood biomarkers to detect NENs. Measurement of the neuroendocrine secretory peptide Chromogranin A (CgA) is used, but is a single value, is non-specific and assay data are highly variable. To facilitate tumor detection, we developed a multi-transcript molecular signature for PCR-based blood analysis. NEN transcripts were identified by computational analysis of 3 microarray datasets: NEN tissue (n = 15), NEN peripheral blood (n = 7), and adenocarcinoma (n = 363 tumors). The candidate gene signature was examined in 130 blood samples (NENs: n = 63) and validated in two independent sets (Set 1 [n = 115, NENs: n = 72]; Set 2 [n = 120, NENs: n = 58]). Comparison with CgA (ELISA) was undertaken in 176 samples (NENs: n = 81). 51 significantly elevated transcript markers were identified. Gene-based classifiers detected NENs in independent sets with high sensitivity (85-98%), specificity (93-97%), PPV (95-96%) and NPV (87-98%). The AUC for the NEN gene-based classifiers was 0.95-0.98 compared to 0.64 for CgA (Z-statistic 6.97-11.42, p<0.0001). Overall, the gene-based classifier was significantly ($\chi^2 = 12.3$, p<0.0005) more accurate than CgA. In a sub-analysis, pancreatic NENs and gastrointestinal NENs could be identified with similar efficacy (79-88% sensitivity, 94% specificity), as could metastases (85%). In patients with low CgA, 91% exhibited elevated transcript markers. A panel of 51 marker genes differentiates NENs from controls with a high PPV and NPV (>90%), identifies pancreatic and gastrointestinal NENs with similar efficacy, and confirms GEP-NENs when CgA levels are low. The panel is significantly more accurate than the CgA assay. This reflects its utility to identify multiple diverse

biological components of NENs. Application of this sensitive and specific PCR-based blood test to NENs will allow accurate detection of disease, and potentially define disease progress enabling monitoring of treatment efficacy.

[191]

TÍTULO / TITLE: - Carcinoid tumor of the ileoanal pouch in a patient with ulcerative colitis.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - R I Med J (2013). 2013 Jan 4;96(1):30-1.

AUTORES / AUTHORS: - Resnick M; Pricolo V; Chen S

INSTITUCIÓN / INSTITUTION: - Professor of Pathology & Laboratory Medicine at the Warren Alpert Medical School of Brown University.

RESUMEN / SUMMARY: - Carcinoid tumors have been reported to occur in various locations, particularly in the gastrointestinal tract. The relationship between the development of carcinoids and ulcerative colitis has been an unclear and controversial one. The association of ulcerative colitis and the development of ileal-pouch carcinoids has not, however, been well documented. We report a case of carcinoid tumor arising in an ileoanal pouch and discuss its unique diagnostic and therapeutic considerations.

[192]

TÍTULO / TITLE: - Merkel cell polyomavirus is frequently detected in Korean patients with Merkel cell carcinoma.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Ann Dermatol. 2013 May;25(2):203-7. doi: 10.5021/ad.2013.25.2.203. Epub 2013 May 10.

●●Enlace al texto completo (gratis o de pago) [5021/ad.2013.25.2.203](#)

AUTORES / AUTHORS: - Chun SM; Yun SJ; Lee SC; Won YH; Lee JB

INSTITUCIÓN / INSTITUTION: - Department of Dermatology, Chonnam National University Medical School, Gwangju, Korea.

RESUMEN / SUMMARY: - BACKGROUND: Merkel cell carcinoma (MCC) is an increasingly common neuroendocrine cancer of the skin. Merkel cell polyomavirus (MCPyV) is one of the causative agents of MCC. The prevalence of MCPyV in primary MCC and sun-exposed non-MCC tumors has been known to have different results depending on where it was investigated. OBJECTIVE: This study assesses the prevalence of MCPyV from primary MCC and sun-exposed non-MCC tumors in Korea. METHODS: A molecular pathology study was performed on 7 tissue specimens of MCC, 1 tissue specimen of metastatic small cell carcinoma of the lung, and 32 tissue specimens of non-MCC tumors occurring from sun-exposed areas [8 basal cell carcinomas (BCCs), 8 squamous cell carcinomas (SCCs), 8 actinic keratoses (AKs), and 8 seborrheic keratoses (SKs)]. All specimens were analyzed to determine the presence of MCPyV-DNA using both polymerase chain reaction (PCR) and real-time

quantitative PCR. Immunohistochemistry with monoclonal antibody of MCPyV large T antigen (CM2B4) was also conducted. RESULTS: Using both PCR, MCPyV sequences were detected in six of seven MCC tissue specimens (85.7%). Five (71%) of seven MCC tumors were immunoreactive for CM2B4. All five immunoreactive cases were positive for MCPyV. However, there was no association of MCPyV with BCC, SCC, AK, and SK. CONCLUSION: Our results implicate that MCPyV may contribute to the pathogenesis of primary MCC, not of non-MCC skin tumors in Korea, and the persons with MCPyV infection are unusual in Korea compared to other areas.

[193]

TÍTULO / TITLE: - Clinical usefulness of plasma chromogranin a in pancreatic neuroendocrine neoplasm.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - J Korean Med Sci. 2013 May;28(5):750-4. doi: 10.3346/jkms.2013.28.5.750. Epub 2013 May 2.

●●Enlace al texto completo (gratis o de pago)

[3346/jkms.2013.28.5.750](#)

AUTORES / AUTHORS: - Paik WH; Ryu JK; Song BJ; Kim J; Park JK; Kim YT; Yoon YB

INSTITUCIÓN / INSTITUTION: - Department of Internal Medicine and Liver Research Institute, Seoul National University College of Medicine, Seoul, Korea.

RESUMEN / SUMMARY: - Chromogranin A (CgA) is widely used as an immunohistochemical marker of neuroendocrine neoplasms and has been measurable in plasma of patients. We assessed the clinical role of plasma CgA in diagnosing pancreatic neuroendocrine neoplasm (PNEN). CgA was checked in 44 patients with pancreatic mass who underwent surgical resection from 2009 through 2011. The cutoff value for diagnosing PNEN and the relationships between CgA and clinicopathologic variables were analyzed. Twenty-six patients were PNENs and 18 patients were other pancreatic disorders. ROC analysis showed a cutoff of 60.7 ng/mL with 77% sensitivity and 56% specificity, and the area under the curve (AUC) was 0.679. Among PNEN group, the sensitivity and specificity of diagnosing metastasis were 100% and 90% respectively when CgA cutoff was 156.5 ng/mL. The AUC was 0.958. High Ki-67 index (160.8 vs 62.1 ng/mL, P = 0.001) and mitotic count (173.5 vs 74.6 ng/mL, P = 0.044) were significantly correlated with plasma CgA, but the tumor size was not. In conclusion, CgA has a little value in diagnosing PNEN. However, the high level of CgA (more than 156.5 ng/mL) can predict the metastasis. Also, plasma CgA level correlates with Ki-67 index and mitotic count which represents prognosis of PNENs.

[194]

TÍTULO / TITLE: - Genetic analysis of a Chinese Han family with multiple endocrine neoplasia type 2^a.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Indian J Biochem Biophys. 2013 Feb;50(1):26-31.

AUTORES / AUTHORS: - Guo Y; Xu H; Ren Z; Yang Y; Xiong W; Gao K; Li X; Luo Z; Deng H

INSTITUCIÓN / INSTITUTION: - Center for Experimental Medicine, The Third Xiangya Hospital, Central South University, Hunan 410013, China.

RESUMEN / SUMMARY: - Multiple endocrine neoplasia type 2 (MEN2) is an autosomal dominant disorder that can be distinguished as three different syndromes: multiple endocrine neoplasia type 2^a (MEN2A), MEN2B and familial medullary thyroid carcinoma (FMTC). This disorder is usually caused by the mutations of the rearranged during transfection protooncogene gene (RET) or the neurotrophic tyrosine kinase receptor type 1 gene (NTRK1). To investigate the genetic cause in a Chinese Han family with MEN2A and the genotype-phenotype correlations, nine members belonging to 3 generations of MEN2A family with 5 affected subjects underwent genetic analysis. Standard GTG-banded karyotype analysis and sequencing of the RET and NTRK1 genes were performed to identify the genetic cause of this family. A heterozygous mutation p.Cys634Arg in the RET gene was identified in 5 patients with MEN2A and one asymptomatic family member. The phenotype of patients was that of classic MEN2A, characterized by medullary thyroid carcinoma and pheochromocytoma. The clinical features of all cases with RET mutations varied greatly, including onset age of clinical manifestations, severity and comorbidities. Thus, this study not only identified the hereditary nature of the MEN2A in the cases, but also discovered a family member harboring the same p.Cys634Arg mutation, who was unaware of his condition. These findings may provide new insights into the cause and diagnosis of MEN2A and have implications for genetic counseling.

[195]

TÍTULO / TITLE: - A case of pancreatic neuroendocrine tumor with excessively-advanced liver metastasis treated with S-1/GEM combination chemotherapy plus the long-acting somatostatin analogue octreotide.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Nihon Shokakibyō Gakkai Zasshi. 2013 Apr;110(4):660-8.

AUTORES / AUTHORS: - Yoshida Y; Sugawara N; Minami T; Iwata N; Ikeda K; Endoh T; Sasano H

INSTITUCIÓN / INSTITUTION: - Department of Internal Medicine, Sapporo Dohoto Hospital.

RESUMEN / SUMMARY: - A 41-year-old woman who had a pancreatic tail tumor and multiple liver tumors was referred to our hospital. The results of abdominal US, CT and MRI, and histopathological and immunohistochemical findings of the liver tumor biopsy revealed a pancreatic neuroendocrine tumor with excessively-advanced liver metastasis. We treated her with S-1/gemcitabine combination chemotherapy plus long-acting somatostatin analogue octreotide, which produced tumor stabilization and good quality of life for 7 months, and survival time of 15 months. Although the tumor was diagnosed as a poorly differentiated endocrine carcinoma, this therapy was suggested to be effective in this case.

[196]

TÍTULO / TITLE: - Primary cutaneous neuroendocrine tumor (atypical carcinoid) expressing KIT and PDGFRA with myoepithelial differentiation: a case report with immunohistochemical and molecular genetic studies.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Int J Clin Exp Pathol. 2013;6(4):802-9. Epub 2013 Mar 15.

AUTORES / AUTHORS: - Terada T

INSTITUCIÓN / INSTITUTION: - Department of Pathology, Shizuoka City Shimizu Hospital, Shizuoka, Japan. piyo0111jp@yahoo.co.jp

RESUMEN / SUMMARY: - Primary cutaneous neuroendocrine tumors (NET) except for Merkel cell carcinoma have rarely been reported. Herein reported is a very unique case of primary cutaneous NET with immunohistochemical markers of myoepitheliomas. A 47-year-old woman presented a tumor measuring 0.8x0.9x0.6 cm of the face. The tumor was excised completely with wide margins. Morphologically, the tumor was located in the dermis, and the tumor was composed of epithelioid cells arranged in trabecular, sinusoidal, rosette, ribbon-like, and cord-like patterns. Focal areas show tubular formations. The tumor cells were homogenous, and their nuclei showed hyperchromasia but no apparent histological features of malignancy were seen. The stroma was very scant. No invasive features were seen. Immunohistochemically, the tumor cells were strongly positive for cytokeratin (CK) 34BE12, CD5/6, CK14, NCAM (CD56), p63, and KIT (CD117), and moderately positive for CK AE1/3, p53, chromogranin, synaptophysin, neuron-specific enolase (NSE), PDGFRA, CA19-9, and Ki-67 antigen (labeling index=23%). The tumor cells were negative for CK CAM5.2, CK7, CK8, CK18,CK19,CK20, EMA, vimentin, CEA, HMB45, S100 protein, alpha-smooth muscle antigen, desmin, CD34, GFAP, neurofilaments, CD99 (MIC2), CD45, CD57, ErbB2, TTF-1, MUC1, MUC2, MUC5AC, and MUC6. Mucins examined by d-PAS and Alcian blue techniques were negative. A genetic analysis using PCR-direct sequencing method in paraffin sections identified no mutations of KIT (exons 9, 11, 13 and 17) and PDGFRA (exons 12 and 18) genes. Imaging modalities including CT and MRI

identified no tumor in the body. The clinicians thought that the tumor was cured. She was a sailor and immediately visited other countries; therefore the follow-up could not be done.

[197]

TÍTULO / TITLE: - Neurofibromatosis type-1 with retroperitoneal stromal tumour: one case report.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Scott Med J. 2013 Feb;58(1):e37-40. doi: 10.1177/0036933012474618.

●●Enlace al texto completo (gratis o de pago)

[1177/0036933012474618](#)

AUTORES / AUTHORS: - Zong GQ; Fei Y; Wang F; Liu RM

INSTITUCIÓN / INSTITUTION: - Department of General Surgery, 81st Hospital of PLA, PLA Cancer Center, China.

RESUMEN / SUMMARY: - BACKGROUND: Neurofibromatosis with gastrointestinal stromal tumours have been reported several times, while neurofibromatosis with retroperitoneal stromal tumours are very rare. CASE DESCRIPTION: We report the case of a 44-year-old man with a long history of neurofibromatosis. He complained of severe constipation and left leg pain. The patient's examination showed prominent peripheral cutaneous neurofibromas mainly in the belly and limbs, especially a huge mass in his abdomen, no less than ten cafe-au-lait spots, four Lisch nodules of the iris. Computed tomography and magnetic resonance imaging revealed a round and lobular mass in the retroperitoneal space. It was a well-circumscribed, hypervascular mass with cystic necrosis. A surgical resection was performed, and pathology and immunohistochemistry findings were consistent with stromal tumour. The c-kit gene and platelet-derived growth factor receptor-alpha gene mutations are not observed in the specimen. CONCLUSIONS: Neurofibromatosis with retroperitoneal stromal tumour is very rare, and radiological, pathological and immunohistochemical examination may identify it. Surgical resection may be the unique method of cure for it.

[198]

TÍTULO / TITLE: - Primary hyperparathyroidism.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - J Assoc Physicians India. 2012 Sep;60:53-4.

AUTORES / AUTHORS: - Karthik R

INSTITUCIÓN / INSTITUTION: - Department of Medicine 1 and Infectious Diseases, Christian Medical College, Vellore, Tamil Nadu.

[199]

TÍTULO / TITLE: - Pathologic diagnosis of large cell neuroendocrine carcinoma of the lung in an axillary lymph node: a case report with immunohistochemical and molecular genetic studies.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Int J Clin Exp Pathol. 2013 May 15;6(6):1177-9. Print 2013.

AUTORES / AUTHORS: - Terada T

INSTITUCIÓN / INSTITUTION: - Department of Pathology, Shizuoka City Shimizu Hospital Shimizu, Shizuoka, Japan.

RESUMEN / SUMMARY: - The author herein reports a large cell neuroendocrine carcinoma (LCNEC) of the lung diagnosed in an axillary lymph node without clinical data, with an emphasis of KIT and PDGFRA. A 64-year-old woman presented with axillary and cervical lymph nodes swelling. An excisional biopsy of an axillary lymph node was performed under the clinical diagnosis of malignant lymphoma. The HE section showed a presence of large malignant cells arranged in a medullary pattern. The tumor cells had nucleoli. The HE diagnosis was large cell lymphoma or metastatic undifferentiated carcinoma, in particular large cell carcinoma of the lung. The tumor cells were positive for cytokeratins, p53 protein, thyroid transcriptional factor-1, neuron-specific enolase, synaptophysin, CD56, KIT, and PDGFRA. In contrast, they were negative for CD3, CD15, CD30, CD45, CD20, CD45RO, CEA, CA19-9, and chromogranin (Dako). Ki-67 labeling (Dako) was 100%. Therefore, a diagnosis of LCNEC of the lung was made. A molecular genetic analysis for KIT (exons 9, 11, 13, and 17) and PDGFRA (exons 12 and 18) identified no mutations. Later, a lung tumor and pleural effusion were detected, and the cytology of the effusion and sputum revealed carcinoma cells compatible with LCNEC. The patient was diagnosed as lung LCNEC, and treated by chemotherapy (cisplatin) and radiation (45 Gray). The present report is the first one with an examination of protein expression and gene mutations of KIT and PDGFRA in a metastatic focus of LCNEC of the lung.

[200]

TÍTULO / TITLE: - CARCINOID TUMOR OF THE DUODENUM: a rare tumor at an unusual site. Case series from a single institution.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Arq Gastroenterol. 2013 Mar;50(1):3-9.

AUTORES / AUTHORS: - Waisberg J; Joppert-Netto G; Vasconcellos C; Sartini GH; Miranda LS; Franco MI

RESUMEN / SUMMARY: - Context Duodenal carcinoids are extremely rare, and their characteristics and biological behavior have not been fully elucidated. Objective To analyze the clinicopathological characteristics of patients with resected duodenal carcinoids. Methods Twenty patients (12 females and 8

males) were investigated. Their average age was 66.4 +/- 5.8 years old (43 to 88 years old). The data corresponding to the clinical picture, diagnosis, treatment, and prognosis of patients with duodenal carcinoid tumors subjected to resection over a period of 18 years (1993-2011) were analyzed. Results The most common symptoms were dyspepsia (50%) and epigastric pain (45%) followed by weight loss (10%) and vomiting (5%). Carcinoid syndrome was not observed in any patient. The lesion was located on the first part of the duodenum in 15 (75%) patients, the second part in 4 (20%) patients, and the third part in 1 (5%) patient. The diagnosis of a carcinoid tumor was established through an endoscopic excision biopsy in 19 (95%) patients and an histopathological examination of the surgical specimen in 1 (5%) patient. The average tumor size was 1.1 cm +/- 0.4 cm (0.3 cm to 6.0 cm). Nineteen (95%) patients were initially treated by endoscopic resection of the duodenal lesion. One patient (5%), whose tumor was on the third part of the duodenum underwent a duodenectomy of the third and fourth duodenal parts and duodenojejunal anastomosis. The duodenal carcinoid resection margin was involved in four (20%) patients. Four (20%) patients were subjected to a partial gastrectomy to fully remove the lesion. The tumor was restricted to the submucosal layer in 16 (80%) cases, and it penetrated into the muscular layer in 4 (20%) cases. All patients exhibited positive chromogranin A, neuron-specific enolase, and/or synaptophysin immunostaining. The average duration of the follow-up period was 39.6 months (3 to 96 months). Twelve (60%) of the 20 cases in this series are alive without any evidence of active disease. Only one (5%) patient died due to liver metastases of the duodenal carcinoid. Conclusions Duodenal carcinoids are rare and indolent tumors usually associated with a benign progression. Duodenoscopy, computerized tomography, and endoscopic ultrasound should be performed to evaluate the tumor size, the level of wall invasion, and the presence of regional or distant lymphatic metastases. Endoscopic removal of tumors smaller than 1.0 cm without periampullary localization or evidence of muscular propria layer invasion assessed by histology and/or endoscopic ultrasound is recommended. The endoscopic resection with a carcinoid tumor size between 1.0 cm and 2.0 cm can be incomplete and require new endoscopic resection or even surgical removal. Duodenal carcinoid larger than 2.0 cm require full-thickness resection and concomitant lymphadenectomy.

[201]

TÍTULO / TITLE: - Small cell neuroendocrine carcinoma of rectum with associated paraneoplastic syndrome: a case report.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - P R Health Sci J. 2013 Mar;32(1):51-3.

AUTORES / AUTHORS: - Vergeli-Rojas JA; Santiago-Caraballo DL; Caceres-Perkins W; Magno-Pagatzartundua P; Toro DH

INSTITUCIÓN / INSTITUTION: - Department of Gastroenterology, Veterans Affairs Caribbean Healthcare System, San Juan, Puerto Rico.

RESUMEN / SUMMARY: - Neuroendocrine carcinomas of the colon and rectum comprise fewer than 1% of all colorectal cancers. These aggressive tumors generally have a poor prognosis compared to that associated with colorectal adenocarcinoma. We describe herein the case of a 68-year-old female presenting with a bleeding rectal mass involving the anal canal, which case was associated with hyponatremia due to inappropriate serum levels of antidiuretic hormone. The histopathological examination was consistent with a small-cell neuroendocrine tumor. She was treated with combination chemotherapy and radiation therapy. The Syndrome of Inappropriate Antidiuretic Hormone (SIADH) was managed with vasopressin antagonists. After the completion of therapy, endoscopic ultrasound revealed evidence of residual disease, for which she underwent an abdominoperineal resection (APR). The patient died 4 months later of disease progression. To our knowledge, this is the first report of a small-cell neuroendocrine tumor involving the rectum and anal canal that presented with the paraneoplastic syndrome, SIADH.

[202]

TÍTULO / TITLE: - PANCREATIC SPLENOSIS MIMICKING NEUROENDOCRINE TUMORS: microhistological diagnosis by endoscopic ultrasound guided fine needle aspiration.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Arq Gastroenterol. 2013 Mar;50(1):10-4.

AUTORES / AUTHORS: - Ardengh JC; Lopes CV; Kemp R; Lima-Filho ER; Venco F; Santos JS

RESUMEN / SUMMARY: - Context Pancreatic splenosis is a benign condition which can mimic a pancreatic neoplasm. Objective To describe the role of the endoscopic ultrasound-guided fine-needle aspiration (EUS-FNA) of pancreatic nodules suspicious for pancreatic splenosis. Method From 1997 to 2011, patients with pancreatic solid tumors suspicious for splenosis by computed tomography and/or magnetic resonance imaging were referred to EUS-FNA. Those cases with pancreatic splenosis confirmed by EUS-FNA or surgery were included. Endosonographic findings and clinicopathologic features were also analysed. Results A total of 2,060 patients with pancreatic solid tumors underwent EUS-FNA. Fourteen (0.6%) cases with pancreatic splenosis were found. After applying exclusion criteria, 11 patients were selected. Most patients were male (7), young (mean age: 42 years) and asymptomatic (8). Endoscopic ultrasound imaging alone suspected pancreatic splenosis in 6 cases, and neuroendocrine tumors in 5 cases. Pancreatic splenosis was found most

commonly in the tail, was round, hypoechoic, with homogeneous pattern, regular borders, and with scintigraphy negative for somatostatin receptors. The average diameter of these nodules identified by endoscopic ultrasound was 2.15 cm. Microhistology obtained by EUS-FNA confirmed the diagnosis in 9/10 patients. Conclusion Pancreatic splenosis can be diagnosed by EUS-FNA. Microhistology prevents unnecessary surgeries, and reassures asymptomatic patients with hypoechoic, homogeneous, and well circumscribed pancreatic nodules.

[203]

TÍTULO / TITLE: - Primary neuroendocrine tumor of the breast: imaging features.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Korean J Radiol. 2013 May;14(3):395-9. doi: 10.3348/kjr.2013.14.3.395. Epub 2013 May 2.

●●Enlace al texto completo (gratis o de pago) [3348/kjr.2013.14.3.395](#)

AUTORES / AUTHORS: - Chang ED; Kim MK; Kim JS; Whang IY

INSTITUCIÓN / INSTITUTION: - Department of Clinical Pathology, Uijeongbu St. Mary's Hospital, College of Medicine, The Catholic University of Korea, Uijeongbu 480-717, Korea.

RESUMEN / SUMMARY: - Focal neuroendocrine differentiation can be found in diverse histological types of breast tumors. However, the term, neuroendocrine breast tumor, indicates the diffuse expression of neuroendocrine markers in more than 50% of the tumor cell population. The imaging features of neuroendocrine breast tumor have not been accurately described due to extreme rarity of this tumor type. We present a case of a pathologically confirmed, primary neuroendocrine breast tumor in a 42-year-old woman, with imaging findings difficult to be differentiated from that of invasive ductal carcinoma.

[204]

TÍTULO / TITLE: - Pharmacokinetics and pharmacodynamics of sunitinib for the treatment of advanced pancreatic neuroendocrine tumors.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Expert Opin Drug Metab Toxicol. 2013 Jun;9(6):777-88. doi: 10.1517/17425255.2013.791281. Epub 2013 Apr 16.

●●Enlace al texto completo (gratis o de pago)

[1517/17425255.2013.791281](#)

AUTORES / AUTHORS: - Bisht S; Feldmann G; Brossart P

INSTITUCIÓN / INSTITUTION: - University Hospital of Bonn, Department of Internal Medicine 3, Bonn, Germany savita.Bisht@ukb.uni-bonn.de.

RESUMEN / SUMMARY: - Introduction: Despite being the second most common malignancy of the pancreas, pancreatic neuroendocrine tumors (PNET) have long been understudied due to their low incidence and heterogeneous clinical presentation. Emerging data from a Phase III trial demonstrates improved progression-free survival of patients with advanced PNET on treatment with sunitinib. Areas covered: This article reviews the role of sunitinib, a multitargeted tyrosine kinase inhibitor with potent antiangiogenic and antitumor effects, in the clinical management of PNET. Furthermore, the authors also discuss the pharmacokinetics and pharmacodynamics as well as other clinically relevant aspects regarding sunitinib. Expert opinion: A recent Phase III clinical trial of sunitinib demonstrated significant improvement of progression-free survival in patients with advanced or metastatic well-differentiated PNET that led to its approval in several countries, including Europe and United States. This marks a significant step forward in the clinical management of this disease and spurs hopes to further improve overall survival in this once difficult-to-treat set of patients in the coming years. Fields of future interest will include evaluation of combinatorial regimens, including conventional cytotoxic agents as well as additional targeted drugs in order to overcome resistance to sunitinib.

[205]

TÍTULO / TITLE: - RET codon 618 mutations in Saudi families with multiple endocrine neoplasia Type 2^a and familial medullary thyroid carcinoma.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Ann Saudi Med. 2013 Mar-Apr;33(2):155-8. doi: 10.5144/0256-4947.2013.155.

●●Enlace al texto completo (gratis o de pago) [5144/0256-4947.2013.155](#)

AUTORES / AUTHORS: - Qari F

INSTITUCIÓN / INSTITUTION: - Endocrine, King Abdulaziz University Hospital KAUH, PO Box 13042, Jeddah 21943, Saudi Arabia. faiza_qari@yahoo.com

RESUMEN / SUMMARY: - BACKGROUND AND OBJECTIVES: Certain diseases such as multiple endocrine neoplasia (MEN) 2^a, MEN 2B, familial and sporadic medullary thyroid carcinoma (MTC) and renal dysgenesis are related to abnormalities of the RET protein. Our aim was to evaluate the frequency of RET mutation in 10 Saudi families with MEN type 2^a and familial MTC. DESIGN AND SETTING: A cross-sectional prospective study of patients followed up at King Abdulaziz University Hospital and King Abdulaziz Medical City, Jeddah, between March 2001 and March 2011. PATIENTS AND METHODS: Genomic DNA was isolated from peripheral blood leukocytes of all subjects by standard procedures. Exons 10, 11, 13, 14 and 16 of the RET proto-oncogene were analyzed by single-strand conformation polymorphism, direct DNA sequencing and/or restriction enzyme analysis. RESULTS: We screened 79 subjects for the RET mutation. Of which 43 subjects had hereditary

MTC were enrolled in this study. MEN type 2^a was identified in 25 subjects; MTC was diagnosed in all 25 subjects (100%), pheochromocytoma in 13 subjects (52%) and hyperparathyroidism in 4 subjects (16%). The most frequent genotype in patients with MEN 2^a syndrome was a codon 618 mutation (46.6%), followed by a codon 634 mutation (44.2%). Among the 5 families with MEN 2^a, 3 had a mutation at codon 634, whereas 2 had a mutation at codon 618.
CONCLUSION: The most frequent RET proto-oncogene mutation in our series was in codon 618 (exon 10).

[206]

TÍTULO / TITLE: - Primary neuroendocrine carcinoma of the breast.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - J Coll Physicians Surg Pak. 2013 Apr;23(4):282-4. doi: 04.2013/JCPSP.282284.

AUTORES / AUTHORS: - Rehman A

INSTITUCIÓN / INSTITUTION: - Department of General Surgery, PAEC General Hospital, Islamabad. surgeonarehman@yahoo.com

RESUMEN / SUMMARY: - Primary neuroendocrine carcinoma of the breast (NECB) is an extremely rare variant of breast cancer having aggressive clinicopathological behaviour and poor prognosis. A 62 years old woman presented with a painless lump in the left breast. Microscopic and immunohistochemical evaluation of the core-tissue biopsy and of the mastectomy specimen revealed moderately-differentiated neuroendocrine carcinoma of the breast. She was labeled as a case of primary neuroendocrine carcinoma of the breast after an infallible exclusion of any concomitant lesion elsewhere in the body. Modified radical mastectomy with level II axillary clearance, chemoradiotherapy and Famoxifen have led to an uneventful 5-year survival till the last follow-up.

[207]

TÍTULO / TITLE: - Primary neuroendocrine carcinoma of the thymus.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Niger Med J. 2013 Jan;54(1):68-71. doi: 10.4103/0300-1652.108904.

●●Enlace al texto completo (gratis o de pago) [4103/0300-1652.108904](https://doi.org/10.4103/0300-1652.108904)

AUTORES / AUTHORS: - Gaude GS; Hattiholi V; Malur PR; Hattiholi J

INSTITUCIÓN / INSTITUTION: - Department of Pulmonary Medicine, J. N. Medical College, Belgaum, Karnataka, India.

RESUMEN / SUMMARY: - Primary neuroendocrine tumors of the thymus, previously known as carcinoid tumors of the thymus, are unusual and rare tumors, and prognosis for these patients has been difficult to predict. We

hereby report a case of primary neuroendocrine tumor of the thymus that had an aggressive and fatal course in spite of surgical resection and adjuvant chemotherapy. These tumors must be regarded as a malignant neoplasm that is prone to metastasize to distant sites, even after total excision.

[208]

TÍTULO / TITLE: - Double valve replacement due to dysfunction secondary to carcinoid tumor.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Arq Bras Cardiol. 2013 Mar;100(3):e32-4.

AUTORES / AUTHORS: - Pego-Fernandes PM; Laurino AM; Ferronato Dde S; Costa FP; Anbar R; Jatene FB

INSTITUCIÓN / INSTITUTION: - Hospital Sirio-Libanés, Sao Paulo, SP, Brazil.
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[209]

TÍTULO / TITLE: - Sunitinib malate as first-line treatment for an advanced, poorly differentiated pancreatic neuroendocrine tumor.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Future Oncol. 2013 Jun;9(6):909-13. doi: 10.2217/fon.13.48.

●●Enlace al texto completo (gratis o de pago) [2217/fon.13.48](#)

AUTORES / AUTHORS: - Lin LZ; Li P; Chen HR; Pang LJ

INSTITUCIÓN / INSTITUTION: - Department of Oncology, The First Affiliated Hospital of Guangzhou University of Traditional Chinese Medicine, Guangzhou, 510407, China.

RESUMEN / SUMMARY: - Pancreatic neuroendocrine tumors (PNETs) are rare, accounting for approximately 2% of primary malignant tumors of the pancreas. Compared with common pancreatic ductal adenocarcinomas, they grow more slowly, are less invasive and have a better prognosis. At present, surgery is the preferred method of treatment of PNETs, and offers the only chance of a cure. However, owing to the occult onset of PNETs, once diagnosed they are often inoperable when the diagnosis is established, and the optimal treatment of patients with inoperable liver metastases remains uncertain. In recent years, targeted drug therapies have emerged and have proved effective in prolonging progression-free survival in patients with advanced well-differentiated PNETs, but hardly any progress has been made in the treatment of poorly differentiated PNETs. In the patient described in this report, who had a poorly differentiated PNET with multiple hepatic metastases and had refused cytotoxic chemotherapy, oral sunitinib malate treatment for 22 months with regular

follow-ups proved tolerable and effective in significantly reducing the size of the intrahepatic masses.

[210]

TÍTULO / TITLE: - En bloc resection of a pheochromocytoma metastatic to the spine for local tumor control and for treatment of chronic catecholamine release and related hypertension.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - J Neurosurg Spine. 2013 Jun;18(6):611-6. doi: 10.3171/2013.3.SPINE12966. Epub 2013 Apr 19.

●●Enlace al texto completo (gratis o de pago)

[3171/2013.3.SPINE12966](#)

AUTORES / AUTHORS: - Kaloostian PE; Zadnik PL; Awad AJ; McCarthy E; Wolinsky JP; Sciubba DM

INSTITUCIÓN / INSTITUTION: - Departments of Neurosurgery and.

RESUMEN / SUMMARY: - Resection of metastatic pheochromocytomas may be complicated by transient postoperative neurological deficits due to hypotension. The authors report the first case of en bloc excision of a spinal pheochromocytoma with associated long-term hypertensive management off all medication. Interestingly, this is the first case of transient hypotension following en bloc resection of pheochromocytoma associated with temporary hypotension-associated neurological decline that resolved completely after correction of hypotension postoperatively. A 23-year-old man with a prior adrenalectomy for pheochromocytoma presented with focal thoracic pain. He had a known T-10 vertebral body lesion for which he received chemotherapy and radiation therapy. Imaging demonstrated increased destruction of the T-10 vertebral body, which was concerning for tumor growth. The patient underwent angiographic embolization followed by single-stage posterior en bloc vertebrectomy with placement of a cage and posterior instrumentation and fusion without event. However, approximately 24 hours after surgery, the patient's systolic blood pressure was consistently no higher than 70 mm Hg. During this time, he began suffering from severe bilateral lower-extremity weakness. His systolic blood pressure increased with dopamine, and his strength immediately improved. The patient's oral regimen of adrenergic blockade was stopped, and he recovered without event. Since that time, the patient has been symptom free and requires no antihypertensive medication. The role of en bloc resection for metastatic lesions of the spine is controversial but may be warranted in cases of metastatic pheochromocytoma. En bloc resection avoids intralesional tumor resection and thus may help prevent complications of hypertensive crisis associated with hormonal secretion and extensive blood loss, which are not uncommon with pheochromocytoma resection surgeries. Additionally, the role of en bloc spondylectomy in this

setting may allow for metabolic treatment as patients with actively secreting tumors may no longer require antiadrenergic medications.

[211]

TÍTULO / TITLE: - Transurethral resection of bladder tumour (TURBT) as an optional treatment method on pheochromocytoma of the urinary bladder.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Can Urol Assoc J. 2013 Jan-Feb;7(1-2):E130-4. doi: 10.5489/cuaj.255.

●●Enlace al texto completo (gratis o de pago) [5489/cuaj.255](#)

AUTORES / AUTHORS: - Ahn SG; Jang H; Han DS; Lee JU; Yuk SM

INSTITUCIÓN / INSTITUTION: - Departments of Hospital Urology, Daejeon St. Mary's Hospital, The Catholic University of Korea, Daejeon, Korea;

RESUMEN / SUMMARY: - Pheochromocytoma of the urinary bladder is rare. We have experienced a case of unexpected pheochromocytoma of the urinary bladder in a 45-year-old female. An ultrasonographic, computed tomography scan and cystoscopic examination showed a submucosal bladder mass. After transurethral resection of bladder tumour was performed, the bladder mass was confirmed as pheochromocytoma by a pathologist. After surgery, the patient underwent a subsequent pelvic magnetic resonance imaging, positron emission tomography and I(131)-methyl iodobenzylguanidine (I(131)-MIBG). An image study showed no residual tumour sites and no lymphatic metastasis. The patient has had no tumour recurrence and no voiding symptoms 3 years after the surgery.

[212]

TÍTULO / TITLE: - Glucagon-Like Peptide-1 Receptor Imaging with [Lys (40) (Ahx-HYNIC- (99 m) Tc/EDDA)NH₂]-Exendin-4 for the Diagnosis of Recurrence or Dissemination of Medullary Thyroid Cancer: A Preliminary Report.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Int J Endocrinol. 2013;2013:384508. doi: 10.1155/2013/384508. Epub 2013 Mar 31.

●●Enlace al texto completo (gratis o de pago) [1155/2013/384508](#)

AUTORES / AUTHORS: - Pach D; Sowa-Staszczak A; Jabrocka-Hybel A; Stefanska A; Tomaszuk M; Mikolajczak R; Janota B; Trofimiuk-Muldner M; Przybylik-Mazurek E; Hubalewska-Dydejczyk A

INSTITUCIÓN / INSTITUTION: - Department of Endocrinology, Jagiellonian University Medical College, Kopernika 17, 31-501 Krakow, Poland.

RESUMEN / SUMMARY: - Introduction. Epidemiological studies on medullary thyroid cancer (MTC) have shown that neither a change in stage at diagnosis nor improvement in survival has occurred during the past 30 years. In patients with detectable serum calcitonin and no clinically apparent disease, a careful

search for local recurrence, and nodal or distant metastases, should be performed. Conventional imaging modalities will not show any disease until basal serum calcitonin is at least 150 pg/mL. The objective of the study was to present the first experience with labelled glucagon-like peptide-1 (GLP-1) analogue [Lys(40)(Ahx-HYNIC-(99m)Tc/EDDA)NH₂]-exendin-4 in the visualisation of MTC in humans. Material and Method. Four patients aged 22-74 years (two with sporadic and two with MEN2 syndrome-related disseminated MTC) were enrolled in the study. In all patients, GLP-1 receptor imaging was performed. Results. High-quality images were obtained in all patients. All previously known MTC lesions have been confirmed in GLP-1 scintigraphy. Moreover, one additional liver lesion was detected in sporadic MTC male patient. Conclusions. GLP-1 receptor imaging with [Lys(40)(Ahx-HYNIC-(99m)Tc/EDDA)NH₂]-exendin-4 is able to detect MTC lesions. GLP-1 scintigraphy can serve as a confirmatory test in MTC patients, in whom other imaging procedures are inconsistent.

[213]

TÍTULO / TITLE: - p.Arg82Leu von Hippel-Lindau (VHL) Gene Mutation among Three Members of a Family with Familial Bilateral Pheochromocytoma in India: Molecular Analysis and In Silico Characterization.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - PLoS One. 2013 Apr 23;8(4):e61908. doi: 10.1371/journal.pone.0061908. Print 2013.

●●Enlace al texto completo (gratis o de pago)

1371/journal.pone.0061908

AUTORES / AUTHORS: - John AM; C GP; Ebenazer A; Seshadri MS; Nair A; Rajaratnam S; Pai R

INSTITUCIÓN / INSTITUTION: - Department of Endocrinology, Christian Medical College, Vellore, Tamil Nadu, India.

RESUMEN / SUMMARY: - Various missense mutations in the VHL gene have been reported among patients with familial bilateral pheochromocytoma. However, the p.Arg82Leu mutation in the VHL gene described here among patients with familial bilateral pheochromocytoma, has never been reported previously in a germline configuration. Interestingly, long-term follow-up of these patients indicated that the mutation might have had little impact on the normal function of the VHL gene, since all of them have remained asymptomatic. We further attempted to correlate this information with the results obtained by in silico analysis of this mutation using SIFT, PhD-SNP SVM profile, MutPred, PolyPhen2, and SNPs&GO prediction tools. To gain, new mechanistic insight into the structural effect, we mapped the mutation on to 3D structure (PDB ID 1LM8). Further, we analyzed the structural level changes in time scale level with respect to native and mutant protein complexes by using 12 ns molecular dynamics simulation method. Though these methods predict the mutation to

have a pathogenic potential, it remains to be seen if these patients will eventually develop symptomatic disease.

[214]

TÍTULO / TITLE: - Digestive neuroendocrine tumors (DNET): the era of targeted therapies.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Clin Res Hepatol Gastroenterol. 2013 Apr;37(2):134-41. doi: 10.1016/j.clinre.2012.09.010. Epub 2013 Apr 3.

●●Enlace al texto completo (gratis o de pago)

[1016/j.clinre.2012.09.010](#)

AUTORES / AUTHORS: - Boussaha T; Rougier P; Taieb J; Lepere C

INSTITUCIÓN / INSTITUTION: - Service d'hepatogastroenterologie, Groupement Hospitalier Pitie-Salpetriere, Paris, France.

RESUMEN / SUMMARY: - Neuroendocrine tumors (NETs) are a heterogeneous group of malignancies. Therapeutic options depend on location of the primitive tumor, its expandability, its hormonal symptoms and its differentiation. Though relatively rare, with an increasing incidence and a high prevalence digestive neuroendocrine tumors (DNETs) are ranked just behind colorectal cancer as the most common digestive cancers in developed countries. Three main therapeutic axes have been individualized in the field of well-differentiated DNETs (corresponding to grades 1 and 2 of new WHO classification 2010), firstly, antitumor activity of somatostatin analogs, particularly in slowly progressive metastatic DNETs with limited hepatic invasion, secondly, targeting angiogenesis in these hypervascular tumors and thirdly targeting the mTOR pathway involved in DNETs carcinogenesis. As a consequence of two major randomized phase III trials in 2011, sunitinib and everolimus have been considered as new therapeutic options for well-differentiated, advanced and progressive pancreatic NETs. For everolimus, another phase III study, although non-significant with the chosen criteria, showed effectiveness notably against small intestine NETs. These targeted therapies are new therapeutic weapons in management of well-differentiated DNETs, but its exact role in care strategy, in comparison with other treatments (somatostatin analogs, chemo-embolization, chemotherapy..) deserves to be precise in the future.

[215]

TÍTULO / TITLE: - Malignant pheochromocytoma and paraganglioma: future considerations for therapy.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Q J Nucl Med Mol Imaging. 2013 Apr 18.

AUTORES / AUTHORS: - Buzzoni R; Pusceddu S; Damato A; Meroni E; Cumali A; Milione M; Mazzaferro V; De Braud F; Spreafico C; Maccauro M; Zaffaroni N; Castellani MR

INSTITUCIÓN / INSTITUTION: - Day Hospital Unit, Fondazione IRCCS Istituto Nazionale dei Tumori, Milan, Italy - roberto.buzzoni@istitutotumori.mi.it.

RESUMEN / SUMMARY: - Pheochromocytoma and paraganglioma are rare neuroendocrine tumors. Knowledge about such neoplasms ameliorated in the last 10-15 years with the discovery of increasing number of germ line mutations even in apparently sporadic cases. Seemingly, genetic tests are going to be an integral part of diagnostic procedures. Standard therapies (advanced surgery, radiometabolic therapy, chemotherapy and radiotherapy) have revealed suboptimal results in tumor size reduction and survival. Currently, there is no standard therapeutic protocol and thus some patients end up with overtreatment while others are undertreated. An effective molecular target therapy aiming at permanent control of these highly complex neoplasms should be the aim of future efforts. In clinical setting investigatory trials with multiple drug therapies targeting a variety of different parallel pathways should be available. Successful management requires a multidisciplinary teamwork.

[216]

TÍTULO / TITLE: - Clinicopathologic assay of 15 tumor resections in a family with neurofibromatosis type 2.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - J Neurol Surg B Skull Base. 2012 Apr;73(2):90-103. doi: 10.1055/s-0032-1301394.

●●Enlace al texto completo (gratis o de pago) [1055/s-0032-1301394](https://doi.org/10.1055/s-0032-1301394)

AUTORES / AUTHORS: - Di Maio S; Mrak G; Juric-Sekhar G; Born D; Mantovani A; Sekhar LN

INSTITUCIÓN / INSTITUTION: - Department of Neurological Surgery, University of Washington, Harborview Medical Center, Seattle, Washington.

RESUMEN / SUMMARY: - The objective of this study is the management of multiple family members with multiple neurofibromatosis type 2 (NF2) related tumors of the skull base that can be challenging, on purely technical, decision-making, and ethical levels. These issues are addressed in this manuscript based on an experience treating an unique large family with NF2. A retrospective chart review was performed, reviewing clinical, radiological, surgical, and pathological data. A unique family of 17 siblings, whose father was the proband as a sporadic mutation is reported. Over a 4-month period, five of eight affected siblings underwent 12 procedures for resection of 15 different NF2-related tumors. This single family experience of NF2-related skull base tumors underscores the importance of preservation of function and quality of life as the major determinants of treatment success.

[217]

TÍTULO / TITLE: - Hormonal assessment in clinically silent adrenal pheochromocytoma.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Can Urol Assoc J. 2013 Mar-Apr;7(3-4):86. doi: 10.5489/cuaj.260.

●●Enlace al texto completo (gratis o de pago) [5489/cuaj.260](#)

AUTORES / AUTHORS: - Shakiba B

INSTITUCIÓN / INSTITUTION: - Department of Urology, Mashhad University of Medical Sciences, Islamic Republic of Iran.

[218]

TÍTULO / TITLE: - Clinical, dermoscopic and histological features of a Merkel cell carcinoma of the hand.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - J Dermatol Case Rep. 2013 Mar 30;7(1):15-7. doi: 10.3315/jdcr.2013.1129. Print 2013 Mar 30.

●●Enlace al texto completo (gratis o de pago) [3315/jdcr.2013.1129](#)

AUTORES / AUTHORS: - Scalvenzi M; Palmisano F; Ilardi G; Varricchio S; Costa C

INSTITUCIÓN / INSTITUTION: - Department of Dermatology, Federico II University, Naples, Italy.

RESUMEN / SUMMARY: - BACKGROUND: Merkel cell carcinoma (MCC) is a rare and typically aggressive form of skin cancer. The benign appearance of the tumor usually on exposed skin parts, contrasting with its extensive microscopic invasion, can delay timely diagnosis. MAIN OBSERVATIONS: We report a case of a 71-year-old man with a slowly progressive nodule on the left hand. CONCLUSION: At the dermoscopic examination, the presence of a polymorphous vascular pattern, including milky-red areas may constitute an additional clinical clue to accurately diagnose this rare tumor. Clinical, dermoscopic and histological features are discussed.

[219]

TÍTULO / TITLE: - Germline genetic variation modulates tumor progression and metastasis in a mouse model of neuroendocrine prostate carcinoma.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - PLoS One. 2013 Apr 19;8(4):e61848. doi: 10.1371/journal.pone.0061848. Print 2013.

●●Enlace al texto completo (gratis o de pago)

[1371/journal.pone.0061848](#)

AUTORES / AUTHORS: - Patel SJ; Molinolo AA; Gutkind S; Crawford NP

INSTITUCIÓN / INSTITUTION: - Metastasis Genetics Section, Cancer Genetics Branch, National Human Genome Research Institute, National Institutes of Health, Bethesda, Maryland, United States of America.

RESUMEN / SUMMARY: - Neuroendocrine (NE) differentiation has gained increased attention as a prostate cancer (PC) prognostic marker. The aim of

this study is to determine whether host germline genetic variation influences tumor progression and metastasis in C57BL/6-Tg(TRAMP)8247Ng/J (TRAMP) mouse model of aggressive NEPC. TRAMP mice were crossed to the eight progenitor strains of the Collaborative Cross recombinant inbred panel to address this. Tumor growth and metastasis burden were quantified in heterozygous transgene positive F1 male mice at 30 weeks of age. Compared to wild-type C57BL/6J-Tg(TRAMP)824Ng/J males, TRAMP x CAST/EiJ, TRAMP x NOD/ShiLtJ and TRAMP x NZO/HILtJ F1 males displayed significant increases in tumor growth. Conversely, TRAMP x WSB/EiJ and TRAMP x PWK/PhJ F1 males displayed significant reductions in tumor growth. Interestingly, despite reduced tumor burden, TRAMP x WSB/EiJ males had an increased nodal metastasis burden. Patterns of distant pulmonary metastasis tended to follow the same patterns as that of local dissemination in each of the strains. All tumors and metastases displayed positive staining for NE markers, synaptophysin, and FOXA2. These experiments conclusively demonstrate that the introduction of germline variation by breeding modulates tumor growth, local metastasis burden, and distant metastasis frequency in this model of NEPC. These strains will be useful as model systems to facilitate the identification of germline modifier genes that promote the development of aggressive forms of PC.

[220]

TÍTULO / TITLE: - Anesthetic management of a pregnant woman undergoing laparoscopic surgery for pheochromocytoma -A case report-

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Korean J Anesthesiol. 2013 Apr;64(4):373-5. doi: 10.4097/kjae.2013.64.4.373. Epub 2013 Apr 22.

●●Enlace al texto completo (gratis o de pago) [4097/kjae.2013.64.4.373](#)

AUTORES / AUTHORS: - Doo AR; Kim D; Cha KN; Han YJ; Kim DC

INSTITUCIÓN / INSTITUTION: - Department of Anesthesiology and Pain Medicine, Chonbuk National University Medical School, Jeonju, Korea.

RESUMEN / SUMMARY: - Pheochromocytoma is a rare catecholamine producing tumor. Anesthetic management for the resection of pheochromocytoma is hard and challenging issue to anesthesiologist, because of its potentially lethal cardiovascular complications. It becomes more complicated when the patient is pregnant. Clinicians must keep the safety of both mother and fetus in mind. The timing of surgery for pheochromocytoma in pregnancy is very important for the maternal and fetal safety and depends on the gestational age when diagnosis is made, clinical response to medical treatment, the surgical accessibility of the tumor, and the presence of fetal distress. We report anesthetic experience of a laparoscopic resection for pheochromocytoma in 25th week gestational woman.

[221]

TÍTULO / TITLE: - Prospective evaluation of biweekly streptozotocin in 19 dogs with insulinoma.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - J Vet Intern Med. 2013 May;27(3):483-90. doi: 10.1111/jvim.12086. Epub 2013 Apr 18.

●●Enlace al texto completo (gratis o de pago) 1111/jvim.12086

AUTORES / AUTHORS: - Northrup NC; Rassnick KM; Gieger TL; Kosarek CE; McFadden CW; Rosenberg MP

INSTITUCIÓN / INSTITUTION: - Department of Small Animal Medicine and Surgery, University of Georgia, College of Veterinary Medicine, Athens, GA.

RESUMEN / SUMMARY: - BACKGROUND: Administration of streptozotocin (STZ) at a 21-day interval has been described in dogs with stage II and III insulinoma. Myelosuppression was not observed, suggesting the possibility of increasing dose intensity by decreasing the interval between doses. OBJECTIVE: To describe the tolerability of a biweekly STZ protocol. A secondary objective was to describe the outcome of dogs treated with this protocol. ANIMALS: Nineteen dogs with residual local, metastatic, or recurrent insulinoma. METHODS: After surgery for insulinoma, or at the time of recurrence, dogs were treated with a previously described STZ and saline diuresis protocol. Treatments were administered every 14 days. All dogs received antiemetic treatment. Adverse events (AEs) were recorded and graded. Outcome endpoints assessed were progression-free survival (PFS) and survival. RESULTS: None of the dogs experienced neutropenia or thrombocytopenia. Mild to moderate gastrointestinal toxicity was the most common AE. Diabetes mellitus was observed in 8 dogs and, in 6, resulted in euthanasia or death. Two dogs developed nephrotoxicity manifested as Fanconi syndrome in 1 and nephrogenic diabetes insipidus in the other. Six dogs developed increased alanine amino transferase activity. Hypoglycemia at the end of the STZ infusion, resulted in collapse in 1 dog and a generalized seizure in another. The median overall PFS and survival time were 196 and 308 days, respectively. CONCLUSIONS AND CLINICAL IMPORTANCE: Streptozotocin can be safely administered to dogs with insulinoma, but serious AEs are possible. Additional investigation is required to better define the role of STZ in managing dogs with insulinoma.

[222]

TÍTULO / TITLE: - An unusual combination of extra-adrenal pheochromocytoma and arteriovenous malformation of the ureter in a young adult.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - British Medical J (BMJ). Acceso gratuito al texto completo.

●●Enlace a la Editora de la Revista <http://bmj.com/search.dtl>

●●Cita: British Medical J. (BMJ): <> Case Rep. 2013 Apr 23;2013. pii: bcr2013009491. doi: 10.1136/bcr-2013-009491.

●●Enlace al texto completo (gratis o de pago) 1136/bcr-2013-009491

AUTORES / AUTHORS: - Khawaja A; Aziz W; Nazim SM; Abbas F

INSTITUCIÓN / INSTITUTION: - Medical College, The Aga Khan University Hospital, Karachi, Sindh, Pakistan.

RESUMEN / SUMMARY: - We present a case of a 24-year-old gentleman who presented with painless pan haematuria for 2 weeks. During the workup, he was diagnosed to have a retrocaval mass after a CT scan while cystoscopy revealed a polypoidal pulsating lesion in the left ureter. After surgical manipulation of the retrocaval mass, the blood pressure of the patient raised to 260/130 mm Hg. It was completely resected and diagnosed as extra-adrenal pheochromocytoma (paraganglioma) after histopathology. The lesion in the ureter was completely excised and fulgurated and diagnosed as an arteriovenous malformation. To the best of our knowledge, this is the first patient to be presented in the literature with this unusual combination.

[223]

TÍTULO / TITLE: - Late anastomotic perforation following surgery for gastric neuroendocrine tumor complicated by perforated duodenal ulcer: a case report.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - J Biomed Res. 2013 Mar;27(2):159-62. doi: 10.7555/JBR.27.20110109. Epub 2012 Mar 29.

●●Enlace al texto completo (gratis o de pago) 7555/JBR.27.20110109

AUTORES / AUTHORS: - Han J; He Z

INSTITUCIÓN / INSTITUTION: - Department of General Surgery, the Second Affiliated Hospital of Nanjing Medical University, Nanjing, Jiangsu 210011, China.

RESUMEN / SUMMARY: - Neuroendocrine tumors (NETs) are a group of neoplasms that are characterized by the secretion of a variety of hormones and diverse clinical syndromes. NETs are considered to be rare, but the incidence of NETs has increased rapidly in recent years. NETs provide a clinical challenge for physicians because they comprise a heterogeneous group of malignancies with a wide range of morphological, functional, and behavioral characteristics. Subtotal gastrectomy with Billroth II reconstruction is the mainstay of therapy in the management of gastric NETs complicated by perforated duodenal ulcer. Late perforation of anastomotic stoma as a long-term complication has been rarely reported. Here, we report a case of anastomotic perforation 5 years after subtotal gastrectomy due to perforated duodenal ulcer and gastric NETs.

[224]

TÍTULO / TITLE: - Multiple endocrine neoplasia syndromes.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - J Infus Nurs. 2013 May-Jun;36(3):160-6. doi: 10.1097/NAN.0b013e318288a118.

●●Enlace al texto completo (gratis o de pago)

[1097/NAN.0b013e318288a118](https://doi.org/10.1097/NAN.0b013e318288a118)

AUTORES / AUTHORS: - Clark P

INSTITUCIÓN / INSTITUTION: - Accredo Health Group, Inc.

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RESUMEN / SUMMARY: - Multiple endocrine neoplasia (MEN) is a term used to describe a group of hereditary carcinoma syndromes. Patients carrying a characteristic autosomal dominant gene aberration exhibit various endocrine carcinomas, as well as other anatomical abnormalities. Unfortunately, familial endocrine carcinoma patients are too often unrecognized by primary care providers, resulting in delayed diagnosis and treatment, with profound consequences related to morbidity and mortality. This article will introduce the various MEN syndromes and the infusion nurse's role in the care of these individuals and their families.

[225]

TÍTULO / TITLE: - Paraneoplastic hypercalcemia in a dog with thyroid carcinoma.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Can Vet J. 2012 Oct;53(10):1101-4.

AUTORES / AUTHORS: - Lane AE; Wyatt KM

INSTITUCIÓN / INSTITUTION: - Perth Veterinary Oncology, 305 Selby Street North, Osborne Park, Washington, 6017 USA. amy@pvo.com.au

RESUMEN / SUMMARY: - This case report describes a dog with thyroid carcinoma and paraneoplastic hypercalcemia. Following thyroidectomy the dog became hypocalcemic and required supplementation with calcitriol and calcium carbonate. During the following 2 years, attempts to reduce the supplementation resulted in hypocalcemia. The dog died from renal failure with no evidence of thyroid carcinoma.

[226]

TÍTULO / TITLE: - Cancer: Neuroendocrine tumours and liver transplantation.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Nat Rev Endocrinol. 2013 Jun;9(6):316. doi:

10.1038/nrendo.2013.85. Epub 2013 Apr 16.

●●Enlace al texto completo (gratis o de pago) [1038/nrendo.2013.85](https://doi.org/10.1038/nrendo.2013.85)

[227]

TÍTULO / TITLE: - Virtual neck exploration for parathyroid adenomas: a first step toward minimally invasive image-guided surgery.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - JAMA. Acceso gratuito al texto completo.

●●Enlace a la Editora de la Revista <http://jama.ama-assn.org/search.dtl>

●●Cita: JAMA: <> Surg. 2013 Mar;148(3):232-8; discussion 238.

AUTORES / AUTHORS: - D'Agostino J; Wall J; Soler L; Vix M; Duh QY; Marescaux J

INSTITUCIÓN / INSTITUTION: - IRCAD/Institut Hospitalo Universitaire, Strasbourg, France. jacques.marescaux@ircad.fr

RESUMEN / SUMMARY: - **OBJECTIVE:** To evaluate the performance of 3-dimensional (3D) virtual neck exploration (VNE) as a modality for preoperative localization of parathyroid adenomas in primary hyperparathyroidism and assess the feasibility of using augmented reality to guide parathyroidectomy as a step toward minimally invasive imageguided surgery. **DESIGN:** Enhanced 3D rendering methods can be used to transform computed tomographic scan images into a model for 3D VNE. In addition to a standard imaging modality, 3D VNE was performed in all patients and used to preoperatively plan minimally invasive parathyroidectomy. All preoperative localization studies were analyzed for their sensitivity, specificity, positive predictive value, and negative predictive value for the correct side of the adenoma(s) (lateralization) and the correct quadrant of the neck (localization). The 3D VNE model was used to generate intraoperative augmented reality in 3 cases. **SETTING:** Tertiary care center. **PATIENTS:** A total of 114 consecutive patients with primary hyperparathyroidism were included from January 8, 2008, through July 26, 2011. **RESULTS:** The accuracy of 3D VNE in lateralization and localization was 77.2% and 64.9%, respectively. Virtual neck exploration had superior sensitivity to ultrasonography (P.001), sestamibi scanning (P=.07), and standard computed tomography (P.001). Use of the 3D model for intraoperative augmented reality was feasible. **CONCLUSIONS:** 3-Dimensional VNE is an excellent tool in preoperative localization of parathyroid adenomas with sensitivity, specificity, and diagnostic accuracy commensurate with accepted first-line imaging modalities. The added value of 3D VNE includes enhanced preoperative planning and intraoperative augmented reality to enable less-invasive image-guided surgery.

[228]

TÍTULO / TITLE: - Integrated DNA-based/biochemical screening for early diagnosis of multiple endocrine neoplasia type 2^a (MEN2A).

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - J Biomed Res. 2013 Mar;27(2):145-50. doi: 10.7555/JBR.27.20120121. Epub 2013 Feb 20.

●●Enlace al texto completo (gratuito o de pago) [7555/JBR.27.20120121](http://dx.doi.org/10.7555/JBR.27.20120121)

AUTORES / AUTHORS: - Cui Q; Wang W; Fu Z; Shao X; Zhang Z; Zhang M; Ju X; Wang K; Chen J; Zhou H

INSTITUCIÓN / INSTITUTION: - Department of Geriatrics, Department of Cadres, Tong Ling People's Hospital, Tongling, Anhui 244000, China;

RESUMEN / SUMMARY: - Multiple endocrine neoplasia type 2^a (MEN2A), a subtype of MEN2, is characterized by medullary thyroid cancer, pheochromocytoma, and primary hyperparathyroidism. A Han Chinese pedigree with MEN2A was investigated following confirmation of the proband's diagnosis by pathological findings and DNA/biochemical screening. DNA samples from 4 other family members were collected and exon 5, 8, 10, 11, 13, 16 and 18 of the RET proto-oncogene were sequenced and then analyzed. A missense mutation of TGG (Trp) to TGC (Cys) at codon 634 (the classic MEN2A mutation) in exon 11 of the RET gene was detected in 3 family members, including the proband. Sequencing data were compared with the human gene mutation database. Elevated serum calcitonin level was detected initially; medullary thyroid carcinoma was revealed in the 3 cases and adrenal pheochromocytoma was also found in the proband. Elective operations were successfully performed on the adrenal and thyroid glands because of pheochromocytoma and medullary thyroid carcinoma. Our case study confirms that integrated DNA-based/biochemical screening is crucial for early diagnosis of MEN2A and is helpful in the screening of their relatives. In addition, DNA-based screening may occasionally uncover a previously unknown RET sequence.

[229]

TÍTULO / TITLE: - Intestinal ischaemia associated with carcinoid tumor: a case report with review of the pathogenesis.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Prague Med Rep. 2013;114(1):43-7.

AUTORES / AUTHORS: - Yener O

INSTITUCIÓN / INSTITUTION: - Department of Surgery, Goztepe Training and Research Hospital, Istanbul, Turkey.

RESUMEN / SUMMARY: - Carcinoid tumors are rare, slow-growing neuroendocrine neoplasms that are often indolent and may not become clinically apparent until there is a metastatic spread or evidence of carcinoid syndrome. A 44-year-old man presented to our clinic department with a history of previous left colon cancer operation, chronic crampy left lower quadrant pain, mass and severe anemia. A MR scan was obtained which demonstrated a calcified mesenteric mass 12x8x10 cm diameter with surrounding left colon mesenteric infiltration. The liver was normal. A case of ischaemic ileal necrosis is reported. It was associated with elastic vascular sclerosis produced by mesenteric metastases of an ileal carcinoid tumor. It is postulated that intestinal ischaemia may be of more importance in the production of abdominal pain by carcinoid tumors than has been generally accepted, and that it is the result of

functional and structural changes in and around the mesenteric blood vessels, caused by substances secreted by the carcinoid tumor.

[230]

TÍTULO / TITLE: - A case of ascending colon neuroendocrine carcinoma drained through a fistula of the descending part of the duodenum.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Nihon Shokakibyō Gakkai Zasshi. 2013 Apr;110(4):622-9.

AUTORES / AUTHORS: - Yoshida H; Takahashi H; Ukai K; Sugimura M; Iwabuchi M; Mano Y; Tadokoro K; Suzuki H

INSTITUCIÓN / INSTITUTION: - Department of Gastroenterology, Sendai Medical Center.

RESUMEN / SUMMARY: - A 70-year-old woman was admitted for investigation of an abdominal tumor. Abdominal CT revealed an ascending colonic mass measuring 10x10cm, with evidence of liver and lung metastasis. Colonoscopy revealed a cancerous lesion with a central ulcer in the ascending colon. Upper gastrointestinal endoscopy revealed an ulcerative lesion in the descending part of the duodenum. Histologically, the tumor showed features of neuroendocrine carcinoma. The patient died of the primary cancer two and a half months after admission. Autopsy revealed a fistula connecting the ascending colonic mass with the ulcerative lesion in the duodenum.

[231]

TÍTULO / TITLE: - A case of normotensive pheochromocytoma with management dilemma.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Indian J Endocrinol Metab. 2012 Dec;16(Suppl 2):S371-2. doi: 10.4103/2230-8210.104097.

●●Enlace al texto completo (gratis o de pago) 4103/2230-8210.104097

AUTORES / AUTHORS: - Roy M; Sengupta N; Sahana PK; Giri D; Das C

INSTITUCIÓN / INSTITUTION: - Department of Endocrinology, N.R.S Medical College & Hospital, Kolkata, India.

RESUMEN / SUMMARY: - INTRODUCTION: We report an unusual case of normotensive pheochromocytoma detected incidentally, presenting a pre-operative management problem. CASE NOTE: A 40-year-old lady with vague abdominal symptoms was initially discovered with a left adrenal incidentaloma by ultrasound abdomen, which was also revealed in computed tomography (CT). After exclusion of all the causes with possible necessary investigations, pheochromocytoma was confirmed with elevated 24 hour urinary metanephrine and normetanephrine. Her blood pressure was in low to normotensive range all throughout. She was attempted to be prepared with combined alpha and beta blockade but could not tolerate this regimen due to symptomatic hypotension. Subsequently, surgical preparation was planned cautiously with alpha-

adenergic blockade only. With intensive monitoring, she underwent uneventful left adrenalectomy, and surgical pathology was consistent with pheochromocytoma. CONCLUSION: This case illustrates an unusual presentation of normotensive pheochromocytoma as adrenal incidentaloma. Pre-operative preparation in these patients can be achieved with alpha-adrenergic blockade, adequate hydration, and liberal salt intake.

[232]

- CASTELLANO -

TÍTULO / TITLE: Mesanenin Buyuk Hucreli Noroendokrin Karsinomu; Olgu Sunumu.

TÍTULO / TITLE: - Large Cell Neuroendocrine Carcinoma of Urinary Bladder; Case Presentation.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Turk Patoloji Derg. 2013;29(2):138-142. doi: 10.5146/tjpath.2013.01165.

●●Enlace al texto completo (gratis o de pago) [5146/tjpath.2013.01165](#)

AUTORES / AUTHORS: - Sari A; Ermete M; Sadullahoglu C; Bal K; Bolukbasi A

INSTITUCIÓN / INSTITUTION: - Department of Pathology, Izmir Atatürk Training and Research Hospital, IZMIR, TURKEY.

RESUMEN / SUMMARY: - Large cell neuroendocrine tumor of the urinary bladder is very rare. It is a type of neuroendocrine carcinoma that is morphologically different from small cell carcinoma. This manuscript describes a 67-year-old man who presented with hematuria. Ultrasonographic and computer tomography revealed a 5 cm mass in right posterolateral wall of the bladder that invaded perivesical tissue and he subsequently underwent transurethral resection. Microscopic examination showed a tumor with a sheet-like and trabecular growth pattern comprising necrotic areas which infiltrated the muscularis propria. Tumoral cells had coarse chromatin, prominent nucleoli, moderate amount of cytoplasm and immunohistochemically stained strongly positive with synaptophysin, chromogranin and CD56. There are only few case reports of large cell neuroendocrine tumor of the urinary bladder so the biological behavior and the treatment protocol of these tumors are still obscure. Appropriate management protocols and prognostic estimation could be achieved by the increased number of cases being reported. Therefore in a case of a poorly differentiated tumor in bladder, although rare, it is important to consider large cell neuroendocrine carcinoma in differential diagnosis.

[233]

TÍTULO / TITLE: - Pancreatic insulinoma combined with glucagon positive cell: A case report.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - World J Gastrointest Surg. 2013 Mar 27;5(3):68-72. doi: 10.4240/wjgs.v5.i3.68.

●●Enlace al texto completo (gratis o de pago) 4240/wjgs.v5.i3.68

AUTORES / AUTHORS: - Yamashita S; Tanaka N; Takahashi M; Nagai M; Furuya T; Suzuki Y; Nomura Y

INSTITUCIÓN / INSTITUTION: - Suguru Yamashita, Nobutaka Tanaka, Michiro Takahashi, Motoki Nagai, Takatoshi Furuya, Yukihiro Nomura, Department of Surgery, Asahi General Hospital, Chiba 289-2511, Japan.

RESUMEN / SUMMARY: - We present a 70-year-old man who was referred for surgery with uncontrollable hypoglycemia. Ultrasonography and abdominal contrast computed tomography revealed a hypervascular tumor of 1 cm in diameter in the pancreatic tail. With a diagnosis of insulinoma, we performed a distal pancreatectomy. The patient showed a good postoperative course without any complications. The patient's early morning fasting hypoglycemia disappeared. The respective levels of C-peptide and insulin dropped from 14.9 ng/mL and 4860 mU/mL preoperatively to 5.3 ng/mL and 553 mU/mL after surgery. A histopathological examination demonstrated that the tumor was a pancreatic neuroendocrine tumor, grade 1. Immunostaining was negative for insulin and positive for CD56, chromogranin A, synaptophysin and glucagon. These findings suggested that the tumor was clinically an insulinoma but histopathologically a glucagonoma. Among all insulinoma cases reported between 1985 and 2010, only 5 cases were associated with independent glucagonoma. In this report, we characterize and discuss this rare type of insulinoma by describing the case we experienced in detail.

[234]

TÍTULO / TITLE: - Mesenteric paraganglioma: Report of a case.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - World J Gastrointest Surg. 2013 Mar 27;5(3):62-7. doi: 10.4240/wjgs.v5.i3.62.

●●Enlace al texto completo (gratis o de pago) 4240/wjgs.v5.i3.62

AUTORES / AUTHORS: - Fujita T; Kamiya K; Takahashi Y; Miyazaki S; Iino I; Kikuchi H; Hiramatsu Y; Ohta M; Baba S; Konno H

INSTITUCIÓN / INSTITUTION: - Takeshi Fujita, Kinji Kamiya, Yoshiaki Takahashi, Shinichiro Miyazaki, Ichirota Iino, Hirotohi Kikuchi, Yoshihiro Hiramatsu, Manabu Ohta, Hiroyuki Konno, Second Department of Surgery, Hamamatsu University School of Medicine, Shizuoka 431-3192, Japan.

RESUMEN / SUMMARY: - We report a rare case of paraganglioma that developed in the mesentery of terminal ileum. A 78-year-old woman complained of right-sided abdominal pain. Abdominal computed tomography revealed a solid heterogeneously enhanced mass in the right lower abdomen. The tumor was laparoscopically excised. The mesenteric tumor was well circumscribed, ovoid, and encapsulated and measured 3 cm x 1.5 cm x 1.5 cm. Histological

examination showed a cellular neoplasm comprised of nests and groups of tumor cells separated by fibrovascular connective tissue, giving a characteristic nested Zellballen pattern. Immunohistochemically, the tumor cells were positive for chromogranin, synaptophysin, CD56, and vimentin and negative for cytokeratins, SMA, CD34, CD117/c-kit and S100. On the basis of histologic and immunohistochemical features, a diagnosis of mesenteric paraganglioma was made. The operative and postoperative courses were unremarkable, and the patient was discharged on postoperative day 7. She was doing well 1 year after the surgery with no signs of recurrence. Extra-adrenal paragangliomas most commonly develop adjacent to the aorta, particularly the area corresponding to the organ of Zuckerkandl. Mesenteric paraganglioma, as in our case, is extremely rare; only 11 cases have been reported in the literature. We herein discuss the clinical findings of these cases.

[235]

TÍTULO / TITLE: - Locally advanced paraganglioma of the urinary bladder: a case report.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - BMC Res Notes. 2013 Apr 18;6:156. doi: 10.1186/1756-0500-6-156.

●●Enlace al texto completo (gratis o de pago) [1186/1756-0500-6-156](#)

AUTORES / AUTHORS: - Beilan J; Lawton A; Hajdenberg J; Rosser CJ

INSTITUCIÓN / INSTITUTION: - Section of Urologic Oncology, MD Anderson Cancer Center Orlando, Orlando, FL 32806, USA.

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RESUMEN / SUMMARY: - BACKGROUND: Paraganglioma of the urinary bladder is a rare tumor. Herein we sought to describe a case of locally advanced paraganglioma of the urinary bladder managed by partial cystectomy and extended pelvic lymph node dissection. CASE PRESENTATION: The case of a 43-year old Haitian male with locally advanced paraganglioma of the urinary bladder is presented in detail. Through surgical extirpation, our patient was rendered disease-free. Eighteen months later the patient is doing well without symptoms but is noted to have subcentimeter bilateral pulmonary nodules and retroperitoneal lymph nodes. No further therapy has been initiated at this time. CONCLUSIONS: Patients with localized tumors have an extremely favorable prognosis and may be managed by less aggressive modalities, whereas patients with metastatic disease have a significant reduced survival rate despite aggressive treatment.

[236]

TÍTULO / TITLE: - Targeting VEGF-VEGFR Pathway by Sunitinib in Peripheral Primitive Neuroectodermal Tumor, Paraganglioma and Epithelioid Hemangioendothelioma: Three Case Reports.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Case Rep Oncol. 2013 Feb 16;6(1):90-7. doi: 10.1159/000348429. Print 2013 Jan.

●●Enlace al texto completo (gratis o de pago) [1159/000348429](#)

AUTORES / AUTHORS: - Prochilo T; Savelli G; Bertocchi P; Abeni C; Rota L; Rizzi A; Zaniboni A

INSTITUCIÓN / INSTITUTION: - UO Oncologia, Fondazione Poliambulanza, Brescia, Mantua, Italy.

RESUMEN / SUMMARY: - Sunitinib malate (Sutent™; Pfizer Inc., New York, N.Y., USA) is a small molecule kinase inhibitor with activity against a number of tyrosine kinase receptors, including vascular endothelial growth factor receptors, stem-cell factor receptor, and platelet-derived growth factor receptors alpha and beta. Sunitinib, registered for the treatment of renal cell carcinoma and gastrointestinal stromal tumors, has recently been approved for the treatment of patients with advanced pancreatic neuroendocrine tumors. Peripheral primitive neuroectodermal tumor (pPNET), paraganglioma (PGL) and epithelioid hemangioendothelioma (EHE) are rare tumors in which there is an overexpression of pro-angiogenic factors and in which a high intratumoral microvessel density is a significant poor prognostic factor. On the basis of this preclinical rationale and the lack of effective treatments in pre-treated advanced stages of these rare diseases, we report our interesting experience of pPNET, PGL and EHE treatment with sunitinib.

[237]

TÍTULO / TITLE: - A case of the large cell neuroendocrine carcinoma of the urinary bladder.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Case Rep Med. 2013;2013:804136. doi: 10.1155/2013/804136. Epub 2013 Apr 4.

●●Enlace al texto completo (gratis o de pago) [1155/2013/804136](#)

AUTORES / AUTHORS: - Hata S; Tasaki Y

INSTITUCIÓN / INSTITUTION: - Department of Urology, National Hospital Organization Beppu Medical Center, 1473 Uchikamado, Beppu City, Oita 874-0011, Japan.

RESUMEN / SUMMARY: - Large cell neuroendocrine carcinoma (LCNEC) of the urinary bladder is very rare. Definite treatment strategy has not been established and prognosis of the disease is not clear yet. We report a case of primary LCNEC of the urinary bladder here with some review of the literature. The patient was a 84-year-old man. He underwent transurethral resection of bladder tumor (TURBT). Histological examination revealed a rosette arrangement of the tumor cells by HE staining and immunohistochemical study revealed positive CD 56, synaptophysin, and chromogranin A (LCNEC). After

TURBT, he has no sign of recurrence for 8 months. We have to strictly observe the progress because LCNEC is very aggressive.

[238]

TÍTULO / TITLE: - A case of synchronous presentation of primary non-small cell lung carcinoma and pheochromocytoma.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Tuberc Respir Dis (Seoul). 2013 Apr;74(4):181-6. doi: 10.4046/trd.2013.74.4.181. Epub 2013 Apr 30.

●●Enlace al texto completo (gratis o de pago) [4046/trd.2013.74.4.181](#)

AUTORES / AUTHORS: - Han JW; Kim CH; Jang J; Lee HG; Chung DC; Choi JE; Kim K; Lim AL; Song WJ; Song YK; Woo H; Hyun IG; Shin MK; Lee YS; Shin HS

INSTITUCIÓN / INSTITUTION: - Department of Internal Medicine, Hallym University College of Medicine, Seoul, Korea.

RESUMEN / SUMMARY: - We report a rare synchronous presentation of primary lung cancer and adrenal pheochromocytoma. A 59-year-old woman was diagnosed with right upper lobe non-small cell lung carcinoma measuring 2.8 cm and a right adrenal gland mass measuring 3.5 cm, which displayed increased metabolic activity on (18)F-fluorodeoxyglucose positron emission tomography-computed tomography. The adrenal lesion was revealed to be asymptomatic. The patient underwent right adrenalectomy and histological examination revealed a pheochromocytoma. Ten days later, right upper lobectomy was performed for lung cancer. This case indicates that incidental adrenal lesions found in cases of resectable primary lung cancer should be investigated.

[239]

TÍTULO / TITLE: - Strumal carcinoid of the ovary: report of two cases.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Acta Clin Croat. 2012 Dec;51(4):649-53.

AUTORES / AUTHORS: - Lenicek T; Tomas D; Soljadic-Vranes H; Kraljevic Z; Klaric P; Kos M; Kos M

INSTITUCIÓN / INSTITUTION: - Ljudevit Jurak University Department of Pathology, Sestre milosrdnice University Hospital Center, Zagreb, Croatia.

RESUMEN / SUMMARY: - Primary carcinoid tumors of the ovary account for 5% of ovarian teratomas. They are frequently components of mature cystic teratomas or, less commonly, mucinous cystadenomas. Most tumors are seen in peri- or postmenopausal women with symptoms of enlarging mass, or are incidental findings. Microscopically, there are four major variants of ovarian teratomas of carcinoid type: insular, trabecular, strumal and mucinous. One-third of patients with the insular type of carcinoids have symptoms of the carcinoid syndrome. Strumal carcinoid is an unusual form of ovarian teratoma composed of an

intimate admixture of thyroid and carcinoid tissues that vary in their relative proportions. Two patients with ovarian tumors showing typical morphology of primary ovarian strumal carcinoid are described.

[240]

TÍTULO / TITLE: - Evaluation of the WHO 2010 Grading and AJCC/UICC Staging Systems in Prognostic Behavior of Intestinal Neuroendocrine Tumors.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - PLoS One. 2013 Apr 19;8(4):e61538. doi: 10.1371/journal.pone.0061538. Print 2013.

●●Enlace al texto completo (gratis o de pago)

[1371/journal.pone.0061538](#)

AUTORES / AUTHORS: - Araujo PB; Cheng S; Mete O; Serra S; Morin E; Asa SL; Ezzat S

INSTITUCIÓN / INSTITUTION: - Department of Medicine, University Health Network, Toronto, Ontario, Canada.

RESUMEN / SUMMARY: - BACKGROUND: The increasing incidence and heterogeneous behavior of intestinal neuroendocrine tumors (iNETs) pose a clinicopathological challenge. Our goal was to describe the prognostic value of the new WHO 2010 grading and the AJCC/UICC TNM staging systems for iNETs. Moreover, outcomes of patients treated with somatostatin analogs were assessed. METHODS: We collected epidemiological and clinicopathological data from 93 patients with histologically proven iNETs including progression and survival outcomes. The WHO 2010 grading and the AJCC/UICC TNM staging systems were applied for all cases. RECIST criteria were used to define progression. Kaplan-Meier analyses for progression free survival (PFS) and overall survival (OS) were performed. RESULTS: Mean follow-up was 58.6 months (4-213 months). WHO 2010 grading yielded PFS and disease-specific OS of 125.0 and 165.8 months for grade 1 (G1), 100.0 and 144.2 months for G2 and 15.0 and 15.8 months for G3 tumors ($p = 0.004$ and $p = 0.001$). Using AJCC staging, patients with stage I and II tumors had no progression and no deaths. Stage III and IV patients demonstrated PFS of 138.4 and 84.7 months ($p = 0.003$) and disease-specific OS of 210.0 and 112.8 months ($p = 0.017$). AJCC staging also provided informative PFS (91.2 vs. 50.0 months, $p = 0.004$) and OS (112.3 vs. 80.0 months, $p = 0.005$) measures with somatostatin analog use in stage IV patients. CONCLUSION: Our findings underscore the complementarity of WHO 2010 and AJCC classifications in providing better estimates of iNETS disease outcomes and extend the evidence for somatostatin analog benefit in patients with metastatic disease.

[241]

TÍTULO / TITLE: - Multiple endocrine neoplasia 2B: delayed presentation, rapid diagnosis.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - British Medical J (BMJ). Acceso gratuito al texto completo.

●●Enlace a la Editora de la Revista <http://bmj.com/search.dtl>

●●Cita: British Medical J. (BMJ): <> Case Rep. 2013 May 2;2013. pii: bcr2013009185. doi: 10.1136/bcr-2013-009185.

●●Enlace al texto completo (gratis o de pago) 1136/bcr-2013-009185

AUTORES / AUTHORS: - Balachandran K; Kamalanathan S; Gopalakrishnan S; Murugananadham K

INSTITUCIÓN / INSTITUTION: - Department of Endocrinology, Jawaharlal Institute of Post Graduate Medical Education & Research, Puducherry, India.

RESUMEN / SUMMARY: - Multiple endocrine neoplasia (MEN) refers to the synchronous or metachronous development of tumours in two or more endocrine organs. MEN 2B is associated with medullary thyroid carcinoma and pheochromocytoma along with classic morphological features such as marfanoid habitus and mucosal neuromas. Dominantly inherited germline mutations involving the Rearranged during Transfection (RET) proto-oncogene are responsible. Affected patients usually present in childhood with thyroid mass or gastrointestinal symptoms. We describe the case of a 28-year-old man who presented to us with metastatic medullary thyroid carcinoma. He lacked the classic marfanoid habitus, but had mucosal neuromas and thickened corneal nerves. Whole-body metaiodobenzyl guanidine scan (MIBG) showed tracer uptake in adrenal and thyroid-confirming pheochromocytoma and medullary thyroid carcinoma. This case exemplifies the late presentation of multiple endocrine neoplasia 2B and emphasises the need to screen all cases of medullary thyroid carcinoma for pheochromocytoma.

[242]

TÍTULO / TITLE: - Effects of pre-operative administration of medetomidine on plasma insulin and glucose concentrations in healthy dogs and dogs with insulinoma.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Vet Anaesth Analg. 2013 May 29. doi: 10.1111/vaa.12047.

●●Enlace al texto completo (gratis o de pago) 1111/vaa.12047

AUTORES / AUTHORS: - Guedes AG; Rude EP

INSTITUCIÓN / INSTITUTION: - Veterinary Clinical Sciences Department, College of Veterinary Medicine, University of Minnesota, St. Paul, MN, USA.

RESUMEN / SUMMARY: - OBJECTIVE: To investigate the effect of medetomidine on plasma glucose and insulin concentrations in dogs with insulinoma and in healthy dogs undergoing anesthesia and surgery. ANIMALS: Twenty-five dogs with insulinoma and 26 healthy dogs. METHODS: In dogs with insulinoma, medetomidine (5 µg kg⁻¹) was randomly included (n = 12) or omitted (n = 13)

from the pre-anesthetic medication protocol, which typically contained an opioid and an anticholinergic. Healthy dogs received medetomidine (5 µg kg⁻¹ ; n = 13) or acepromazine (0.04 mg kg⁻¹ ; n = 13) plus an opioid (morphine 0.5 mg kg⁻¹) and an anticholinergic (atropine 0.04 mg kg⁻¹) as pre-anesthetic medications. Pre-anesthetic medications were given intramuscularly. Plasma glucose and insulin concentrations were measured before (sample 1) and 30 minutes after pre-anesthetic medication (sample 2), and at the end of surgery in dogs with insulinoma or at 2 hours of anesthesia in healthy dogs (sample 3). Glucose requirement to maintain intra-operative normoglycemia in dogs with insulinoma was quantified and compared. Data were analyzed with anova and Bonferroni post-test, t-tests or chi-square tests as appropriate with p < 0.05 considered significant. Data are shown as mean +/- SD. RESULTS: Medetomidine significantly decreased plasma insulin concentrations and increased plasma glucose concentrations in healthy dogs and those with insulinoma. These variables did not change significantly in the dogs not receiving medetomidine. In the dogs with insulinoma, intra-operative glucose administration rate was significantly less in the animals that received medetomidine compared to those that did not. CONCLUSIONS: Pre-anesthetic administration of medetomidine significantly suppressed insulin secretion and increased plasma glucose concentration in dogs with insulinoma and in healthy dogs undergoing anesthesia and surgery. CLINICAL RELEVANCE: These findings support the judicious use of medetomidine at low doses as an adjunct to the anesthetic management of dogs with insulinoma.

[243]

TÍTULO / TITLE: - Identification and characterization of poorly differentiated invasive carcinomas in a mouse model of pancreatic neuroendocrine tumorigenesis.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - PLoS One. 2013 May 14;8(5):e64472. doi: 10.1371/journal.pone.0064472. Print 2013.

●●Enlace al texto completo (gratis o de pago)

1371/journal.pone.0064472

AUTORES / AUTHORS: - Hunter KE; Quick ML; Sadanandam A; Hanahan D; Joyce JA

INSTITUCIÓN / INSTITUTION: - Cancer Biology and Genetics Program, Memorial Sloan-Kettering Cancer Center, New York, New York, United States of America ; Louis V. Gerstner Jr. Graduate School of Biomedical Sciences, Memorial Sloan-Kettering Cancer Center, New York, New York, United States of America.

RESUMEN / SUMMARY: - Pancreatic neuroendocrine tumors (PanNETs) are a relatively rare but clinically challenging tumor type. In particular, high grade, poorly-differentiated PanNETs have the worst patient prognosis, and the

underlying mechanisms of disease are poorly understood. In this study we have identified and characterized a previously undescribed class of poorly differentiated PanNETs in the RIP1-Tag2 mouse model. We found that while the majority of tumors in the RIP1-Tag2 model are well-differentiated insulinomas, a subset of tumors had lost multiple markers of beta-cell differentiation and were highly invasive, leading us to term them poorly differentiated invasive carcinomas (PDICs). In addition, we found that these tumors exhibited a high mitotic index, resembling poorly differentiated (PD)-PanNETs in human patients. Interestingly, we identified expression of Id1, an inhibitor of DNA binding gene, and a regulator of differentiation, specifically in PDIC tumor cells by histological analysis. The identification of PDICs in this mouse model provides a unique opportunity to study the pathology and molecular characteristics of PD-PanNETs.

[244]

TÍTULO / TITLE: - Efficacy of nasopancreatic stenting prior to laparoscopic enucleation of pancreatic neuroendocrine tumor.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Asian J Endosc Surg. 2013 May;6(2):140-2. doi: 10.1111/ases.12006.

●●Enlace al texto completo (gratis o de pago) [1111/ases.12006](#)

AUTORES / AUTHORS: - Misawa T; Imazu H; Fujiwara Y; Kitamura H; Tsutsui N; Ito R; Shiba H; Futagawa Y; Wakiyama S; Ishida Y; Yanaga K

INSTITUCIÓN / INSTITUTION: - Department of Surgery, The Jikei University School of Medicine, Tokyo, Japan. take-misawa@jikei.ac.jp

RESUMEN / SUMMARY: - We report a patient who underwent laparoscopic enucleation for a nonfunctioning pancreatic neuroendocrine tumor. The patient was a 55-year-old man who had a 12- x 11-mm tumor close to the main pancreatic duct (MPD) in the pancreatic body. To avoid and detect injury to the main pancreatic duct during operation, a nasopancreatic drainage stent (NPDS) was endoscopically placed prior to the operation. According to the NPDS, the relation between the tumor and MPD was easily identified by laparoscopic ultrasonography during enucleation, thus enabling the resecting line to be determined. Moreover, after enucleation, pancreatography through the NPDS was able to clarify the absence of injury to the MPD. The NPDS was removed postoperatively, and the patient was discharged uneventfully on postoperative day 8. Preoperative placement of the NPDS seems to be a useful option for performing safe laparoscopic enucleation of pancreatic neuroendocrine tumor, especially when the lesion is located close to the MPD.

[245]

TÍTULO / TITLE: - eComment. Surgical management of carcinoid tumours of the lung: sublobar resection versus lobectomy.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Interact Cardiovasc Thorac Surg. 2013 Jun;16(6):864. doi: 10.1093/icvts/ivt176.

●●Enlace al texto completo (gratis o de pago) [1093/icvts/ivt176](#)

AUTORES / AUTHORS: - Wurtz AJ; Hysi I; Benhamed L

INSTITUCIÓN / INSTITUTION: - Cardiac and Thoracic Surgery Division, Lille University Teaching Hospital, CHU Lille, Lille, France.

[246]

TÍTULO / TITLE: - Postoperative radiotherapy for residual tumor of primary mediastinal carcinoid teratoma.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - J Thorac Dis. 2013 Apr;5(2):E61-4. doi: 10.3978/j.issn.2072-1439.2012.11.11.

●●Enlace al texto completo (gratis o de pago) [3978/j.issn.2072-1439.2012.11.11](#)

AUTORES / AUTHORS: - Tu L; Sun L; Zhou Y; Gong Y; Xue J; Gao J; Lu Y; Wang Y

INSTITUCIÓN / INSTITUTION: - Department of Thoracic Oncology, Cancer Center, West China Hospital, Sichuan University, Chengdu, China;

RESUMEN / SUMMARY: - A 36-year-old woman had presented with dry cough for 2 months. Thoracic computed tomography (CT) scan showed a 12 cm x 8 cm x 5 cm mass in the anterior mediastinum. Due to intimately involving the aortic arch, tumor was removed incompletely. Residual tumor remained approximate 2 cm x 3 cm x 4 cm. Histologic diagnosis was a mature cystic teratoma containing a carcinoid. Subsequently, radiotherapy (RT) was administered on residual tumor for a total dose of 50 Gy at 2 Gy/d fraction in 25 fractions. At 2-year follow-up, the patient had stable disease. In conclusion, adjuvant radiotherapy with 50 Gy is an effective approach for residual tumor of mediastinal carcinoid teratoma.

[247]

TÍTULO / TITLE: - Pure bronchoplastic resections of the bronchus without pulmonary resection for endobronchial carcinoid tumours.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Interact Cardiovasc Thorac Surg. 2013 Apr 28.

●●Enlace al texto completo (gratis o de pago) [1093/icvts/ivt154](#)

AUTORES / AUTHORS: - Nowak K; Karenovics W; Nicholson AG; Jordan S; Dusmet M

INSTITUCIÓN / INSTITUTION: - Department of Thoracic Surgery, Royal Brompton Hospital, London, UK.

RESUMEN / SUMMARY: - OBJECTIVES Bronchopulmonary carcinoid tumours are relatively uncommon primary lung neoplasms. A small proportion of these

lesions are predominantly endobronchial and do not extend beyond the bronchial wall. Endoscopic resection can be performed, but carries around a one in three risk of local recurrence and, therefore, mandates long-term surveillance. An alternative is complete surgical resection via bronchoplastic resection. We present our experience of surgical resection in patients with endobronchial carcinoids. **METHODS** From 2000 to 2010, 13 patients (age 45 +/- 16 years, 10 males) underwent pure bronchoplastic resection, including systematic nodal dissection, for endobronchial carcinoid tumours, without the resection of lung parenchyma. **RESULTS** There was no significant operative morbidity or mortality. This is a retrospective review of a consecutive case series. The last follow-up for all patients was obtained in 2011. The mean maximum tumour size was 18 +/- 8 mm. No lymph node invasion was observed. The median follow-up was 6.3 +/- 3.3 years, with no regional recurrence. In 1 case, a tumourlet was identified at 5 years in the contralateral airway and viewed as a metachronous new lesion. **CONCLUSIONS** Bronchial sleeve resection is a safe procedure for suitably located endobronchial carcinoid tumours. Endoscopic resection should be reserved for patients who decline, or are unfit, for surgery.

[248]

TÍTULO / TITLE: - Hyperaccumulation of F-FDG in order to differentiate solid pseudopapillary tumors from adenocarcinomas and from neuroendocrine pancreatic tumors and review of the literature.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Hell J Nucl Med. 2013 May 20. pii: s002449910084.

●●Enlace al texto completo (gratis o de pago) [1967/s002449910084](#)

AUTORES / AUTHORS: - Guan ZW; Xu BX; Wang RM; Sun L; Tian JH

INSTITUCIÓN / INSTITUTION: - Fuxing Road 28, Beijing, 100853, China.

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RESUMEN / SUMMARY: - Solid pseudopapillary tumors (SPT) are rare, unique pancreatic tumors with benign entity and low malignant potential. Limited information is available in the literature reporting their accumulation of fluorine-18 fluoro deoxyglucose (18F-FDG) using positron emission tomography/computed tomography (PET/CT). The aim of this retrospective study was to define the uptake-accumulation of 18F-FDG PET/CT in a comparatively large cohort of SPT, and to compare their uptake with the uptake of 18F-FDG in pancreatic ductal adenocarcinomas (PAC) and neuroendocrine tumors (PNET). Between June 2007 and January 2013, 18 pathologically proven SPT were identified from the total of patients studied by PET/CT in our Center, including 13 women and 5 men, aging from 23 to 56 years old (mean age, 38.5 years). Malignant SPT was histologically classified using the WHO criteria. Eighty-six PAC patients and 28 PNET patients were also identified and included in this study for comparison. Positron emission tomography results

were considered as positive if focal accumulation of 18F-FDG exceeded the surrounding normal pancreatic tissue. Regions of interest were drawn on the pancreatic lesions, and the maximal standardized uptake values (SUVmax values) were calculated. The mean values of SUVmax were compared with independent-samples t test or with the nonparametric Mann-Whitney U method. Correlation of SUVmax values and tumor size were analyzed in cases of SPT. Receiver operating characteristics (ROC curve) were used to study the efficiency of SUV values for the differential diagnosis between SPT versus (vs) PAC and SPT vs PNET. A value of $P < 0.05$ was considered statistically significant. All SPT cases were 18F-FDG-PET positive, with SUVmax values ranging from 3.5-18.3. The SUVmax values of SPT had poor correlation with tumor size, and no significant difference by gender and age. Areas under the curve ROC were 0.619 and 0.526, respectively for the differentiation of SPT from PNET and PAC tumors. Five SPT tumors were malignant, and exhibited relatively low 18F-FDG uptake (SUVmax range, 3.0-4.5) except a tumor after recurrence (SUVmax 17.7). Images of CT were of low dose and thus were not evaluated. In conclusion, our results suggest that SPT benign or malignant are consistently hyperaccumulating 18F-FDG above SUVmax 3. Differentiation from PAC and PNET if only based on the higher SUVmax values was not possible but if based on lower SUVmax, of ≤ 2.6 (in 14%) and ≤ 2.5 (in 21,4%) of PAC and PNET, respectively, these pancreatic tumors could be differentiated from SPT.

[249]

TÍTULO / TITLE: - The epidemiology of neuroendocrine tumors in taiwan: a nation-wide cancer registry-based study.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - PLoS One. 2013 Apr 22;8(4):e62487. doi: 10.1371/journal.pone.0062487. Print 2013.

●●Enlace al texto completo (gratis o de pago)

[1371/journal.pone.0062487](#)

AUTORES / AUTHORS: - Tsai HJ; Wu CC; Tsai CR; Lin SF; Chen LT; Chang JS

INSTITUCIÓN / INSTITUTION: - National Institute of Cancer Research, National Health Research Institutes, Tainan, Taiwan.

RESUMEN / SUMMARY: - BACKGROUND: The epidemiology of neuroendocrine tumors (NETs) is not well illustrated, particularly for Asian countries.

METHODS: The age-standardized incidence rates and observed survival rates of NETs diagnosed in Taiwan from January 1, 1996 to December 31, 2008 were calculated using data of the Taiwan Cancer Registry (TCR) and compared to those of the Norwegian Registry of Cancer (NRC) and the US Surveillance, Epidemiology, and End Results (SEER) program. RESULTS: During the study period, a total of 2,187 NET cases were diagnosed in Taiwan, with 62% males and a mean age of 57.9 years-old. The age-standardized incidence rate of

NETs increased from 0.30 per 100,000 in 1996 to 1.51 per 100,000 in 2008. The most common primary sites were rectum (25.4%), lung and bronchus (20%) and stomach (7.4%). The 5-year observed survival was 50.4% for all NETs (43.4% for men and 61.8% for women, $P < 0.0001$). The best 5-year observed survivals for NETs by sites were rectum (80.9%), appendix (75.7%), and breast (64.8%). CONCLUSIONS: Compared to the data of Norway and the US, the age-standardized incidence rate of NETs in Taiwan is lower and the major primary sites are different, whereas the long-term outcome is similar. More studies on the pathogenesis of NETs are warranted to devise preventive strategies and improve treatment outcomes for NETs.

[250]

TÍTULO / TITLE: - Endoscopic mucosal resection with circumferential mucosal incision of duodenal carcinoid tumors.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - World J Gastrointest Endosc. 2013 Apr 16;5(4):197-200. doi: 10.4253/wjge.v5.i4.197.

●●Enlace al texto completo (gratis o de pago) [4253/wjge.v5.i4.197](#)

AUTORES / AUTHORS: - Otaki Y; Homma K; Nawata Y; Imaizumi K; Arai S

INSTITUCIÓN / INSTITUTION: - Yuzo Otaki, Kiyoaki Homma, Yoshitakata Nawata, Department of Therapeutic Endoscopy, Nihonkai General Hospital, Yamagata 998-0828, Japan.

RESUMEN / SUMMARY: - Duodenal carcinoids are a rare form of neuroendocrine tumors, and tend to invade the submucosa during the early stage. Endoscopic treatment is generally recommended for duodenal carcinoids less than 10 mm in diameter. Although a few reports have described the use of endoscopic resection of duodenal carcinoids, there are no published studies on endoscopic mucosal resection with circumferential mucosal incision (EMR-CMI). We performed EMR-CMI for 5 cases of duodenal carcinoids in the duodenal bulb. The mean tumor diameter was 4.6 +/- 1.8 mm. Although all of the tumors were located in the submucosa, R0 resection was performed without complication in each case. EMR-CMI may thus be a safe and effective treatment for duodenal carcinoids less than 10 mm in diameter.

[251]

TÍTULO / TITLE: - Endoscopic mucosal resection of an oesophageal carcinoid tumour.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - British Medical J (BMJ). Acceso gratuito al texto completo.

●●Enlace a la Editora de la Revista <http://bmj.com/search.dtl>

●●Cita: British Medical J. (BMJ): <> Case Rep. 2013 Apr 3;2013. pii: bcr2012008288. doi: 10.1136/bcr-2012-008288.

●●Enlace al texto completo (gratuito o de pago) 1136/bcr-2012-008288

AUTORES / AUTHORS: - Eccles JK; McManus DT; Mainie I

INSTITUCIÓN / INSTITUTION: - Department of Gastroenterology, Belfast City Hospital, Belfast, Antrim, UK. johnkeccles@doctors.org.uk

[252]

TÍTULO / TITLE: - Analysis of loss of heterozygosity effect on thyroid tumor with oxyphilia cell locus in familial non medullary thyroid carcinoma in Iranian families.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Indian J Hum Genet. 2012 Sep;18(3):340-3. doi: 10.4103/0971-6866.107989.

●●Enlace al texto completo (gratuito o de pago) 4103/0971-6866.107989

AUTORES / AUTHORS: - Shirazi HA; Hedayati M; Daneshpour MS; Shafiee A; Azizi F

INSTITUCIÓN / INSTITUTION: - Cellular Molecular and Endocrine Research Center, Research Institute for Endocrine Sciences, Shahid Beheshti University of Medical Sciences, Tehran, Iran.

RESUMEN / SUMMARY: - MATERIAL AND METHODS: 22 nuclear families (78 persons including 12 patients) with papillary and follicular tumors were selected in a period of six months from Milad hospital. Five microsatellite markers (D19S413, D19S391, D19S916, D19S568, D19S865) on 19p13.2 were selected for genetic analysis. Genomic DNAs was extracted; PCR and polyacrylamide gel electrophoresis method were used for variation detection. RESULTS: The results show that 5.4% of the follicular carcinomas and 17.9% of the papillary carcinomas presented LOH at recognition sites. LOH of Papillary carcinoma detected about 13.9% and follicular carcinoma 7.2% in this study. The frequency of informative cases was not similar for each marker: D19S413 (41.1%)[1], D19S391 (12.5%), D19S916 (10.7%), D19S568 (1.8%) and D19S865 (3.6%). Loss of heterozygosity in D19S413 predicts the relation between variation in this region and the disease. DISCUSSION: Our findings showed an average of 13.9% LOH in FNMTTC cases. Among the five major microsatellites, D19S413 was the most informative for LOH analysis of FNMTTC.

[253]

TÍTULO / TITLE: - eComment. The definition of neuroendocrine tumour and sublobar resection.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Interact Cardiovasc Thorac Surg. 2013 Jun;16(6):863. doi: 10.1093/icvts/ivt170.

●●Enlace al texto completo (gratuito o de pago) 1093/icvts/ivt170

AUTORES / AUTHORS: - Bolukbas S; Eberlein M; Schirren J

INSTITUCIÓN / INSTITUTION: - Department of Thoracic Surgery, Dr. Horst Schmidt Klinik, Wiesbaden, Germany.

[254]

TÍTULO / TITLE: - First successful transoral robotic resection of a laryngeal paraganglioma.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - J Otolaryngol Head Neck Surg. 2012 Dec 1;41(6):E54-7.

AUTORES / AUTHORS: - Tulin Kayhan F; Hakan Kaya K; Altintas A; Firat P; Sayin I

[255]

TÍTULO / TITLE: - The evolution in the use of MIBG scintigraphy in pheochromocytomas and paragangliomas.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Hormones (Athens). 2013 Jan;12(1):58-68.

AUTORES / AUTHORS: - Rufini V; Treglia G; Perotti G; Giordano A

INSTITUCIÓN / INSTITUTION: - Institute of Nuclear Medicine, Universita Cattolica del Sacro Cuore, Rome, Italy.

RESUMEN / SUMMARY: - Radioiodinated metaiodobenzylguanidine (MIBG) was developed in the late 1970's, at the Michigan University Medical Center, for imaging of the adrenal medulla and its diseases. Soon after, MIBG was shown to depict a wide range of tumors of neural crest origin other than pheochromocytomas/paragangliomas (Pheo/PGL) with the result that its use rapidly spread to many countries. After more than 30 years of clinical application, MIBG continues to be the most widespread radiopharmaceutical for the functional imaging of Pheo/PGL in spite of the emergent role of PET agents for detection of these tumors. In this paper we review the evolution in the use of MIBG over more than 30 years of experimental and clinical applications, with particular focus on the uptake mechanisms, pharmacokinetics, biodistribution and drug interaction as well as on clinical studies in Pheo/PGL also in comparison to other gamma-emitters tracers and PET radiopharmaceuticals.

[256]

TÍTULO / TITLE: - Primary large cell neuroendocrine carcinoma of the ureter.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Int J Clin Exp Pathol. 2013;6(4):729-36. Epub 2013 Mar 15.

AUTORES / AUTHORS: - Oshiro H; Odagaki Y; Iobe H; Ozu C; Takizawa I; Nagai T; Matsubayashi J; Inagaki A; Miyake S; Nagao T

INSTITUCIÓN / INSTITUTION: - Department of Pathology, Tokyo Medical University Hospital, Tokyo, Japan. oshiroh@yokohama-cu.ac.jp

RESUMEN / SUMMARY: - Large cell neuroendocrine carcinoma (LCNEC) is the rarest type of urinary tract malignancy. Herein, we report a case of LCNEC that arose in the ureter of a 78-year-old Japanese man with a history of ascending colon cancer that had been excised by a right hemicolectomy. Left-sided hydronephrosis associated with the ureteral tumor was discovered during follow-up. A left nephroureterectomy combined with a partial resection of the urinary bladder was performed because atypical cells were detected using voided urine cytology. A histopathological examination revealed that the ureteral tumor contained large atypical epithelial cells of neuroendocrine morphology without a urothelial carcinomatous component. The neoplastic cells were immunohistochemically positive for synaptophysin, chromogranin A, CD56, and cytokeratins, but they were negative for uroplakin III and thyroid transcription factor-1. The Ki-67 labeling index of the neoplastic cells was 50%. Transmission electron microscopy demonstrated the presence of numerous dense granules in the cytoplasm of the neoplastic cells. The ureteral lesion was finally classified as stage III, pT3 cN0 cM0. The patient's postoperative course was uneventful without chemoradiotherapy, and LCNEC did not recur in the subsequent nine months. This case demonstrates that LCNEC can occur in the ureter, which normally does not contain neuroendocrine cells in the urothelium.

[257]

TÍTULO / TITLE: - Advanced medullary thyroid cancer: pathophysiology and management.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Cancer Manag Res. 2013 May 8;5:57-66. doi: 10.2147/CMAR.S33105. Print 2013.

●●Enlace al texto completo (gratis o de pago) [2147/CMAR.S33105](#)

AUTORES / AUTHORS: - Ferreira CV; Siqueira DR; Ceolin L; Maia AL

INSTITUCIÓN / INSTITUTION: - Thyroid Section, Endocrine Division, Hospital de Clinicas de Porto Alegre, Porto Alegre, Brazil.

RESUMEN / SUMMARY: - Medullary thyroid carcinoma (MTC) is a rare malignant tumor originating from thyroid parafollicular C cells. This tumor accounts for 3%-4% of thyroid gland neoplasias. MTC may occur sporadically or be inherited. Hereditary MTC appears as part of the multiple endocrine neoplasia syndrome type 2^a or 2B, or familial medullary thyroid cancer. Germ-line mutations of the RET proto-oncogene cause hereditary forms of cancer, whereas somatic mutations can be present in sporadic forms of the disease. The RET gene encodes a receptor tyrosine kinase involved in the activation of intracellular signaling pathways leading to proliferation, growth, differentiation, migration, and survival. Nowadays, early diagnosis of MTC followed by total thyroidectomy offers the only possibility of cure. Based on the knowledge of the pathogenic mechanisms of MTC, new drugs have been developed in an attempt to control metastatic disease. Of these, small-molecule tyrosine kinase inhibitors

represent one of the most promising agents for MTC treatment, and clinical trials have shown encouraging results. Hopefully, the cumulative knowledge about the targets of action of these drugs and about the tyrosine kinase inhibitor-associated side effects will help in choosing the best therapeutic approach to enhance their benefits.

[258]

TÍTULO / TITLE: - Adrenal artery pseudoaneurysm in pheochromocytoma presenting with catastrophic retroperitoneal haemorrhage.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Can Urol Assoc J. 2013 Mar-Apr;7(3-4):E254-6. doi: 10.5489/cuaj.541.

●●Enlace al texto completo (gratis o de pago) [5489/cuaj.541](#)

AUTORES / AUTHORS: - Kumar S; Nanjappa B; Kumar S; Prasad S; Pushkarna A; Singh SK

INSTITUCIÓN / INSTITUTION: - Department of Urology, Postgraduate Institute of Medical Education and Research, Chandigarh, India;

RESUMEN / SUMMARY: - Spontaneous rupture of adrenal pheochromocytoma is an extremely rare condition which presents as an abdominal catastrophe. Unrecognized, this transformation can rapidly lead to death. We report a case of a 63-year-old male who presented with hemorrhagic shock secondary to ruptured adrenal pheochromocytoma. The clinical course is notable for immediate transarterial catheter embolization for control of bleeding, followed by optimization and elective adrenalectomy. High mortality is associated with an operative intervention in the face of an unrecognized pheochromocytoma. This reinforces the need for maintaining a high index of suspicion in the setting of a suprarenal mass despite hypotension.

[259]

TÍTULO / TITLE: - Malignant peripheral nerve sheath tumor of non-neurofibromatosis type I metastasized to the cerebrospinal axis.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - J Korean Neurosurg Soc. 2013 Mar;53(3):190-3. doi: 10.3340/jkns.2013.53.3.190. Epub 2013 Mar 31.

●●Enlace al texto completo (gratis o de pago) [3340/jkns.2013.53.3.190](#)

AUTORES / AUTHORS: - Park MK; Sung JK; Nam KH; Kim KT

INSTITUCIÓN / INSTITUTION: - Department of Neurosurgery, Kyungpook National University Hospital, Daegu, Korea.

RESUMEN / SUMMARY: - A malignant peripheral nerve sheath tumor (MPNST) is a type of sarcoma that arises from peripheral nerves or cells of the associated nerve sheath. This tumor most commonly metastasizes to the lung and metastases to the spinal cord and brain are very rare. We describe a case of young patient with spinal cord and brain metastases resulting from MPNST. An

18-year-old man presented with a 6-month history of low back pain and radiating pain to his anterior thigh. Magnetic resonance imaging showed a paraspinal mass that extended from the central space of L2 to right psoas muscle through the right L2-3 foraminal space. The patient underwent surgery and the result of the histopathologic study was diagnostic for MPNST. Six months after surgery, follow-up images revealed multiple spinal cord and brain metastases. The patient was managed with chemotherapy, but died several months later. Despite complete surgical excision, the MPNST progressed rapidly and aggressively. Thus, patients with MPNST should be followed carefully to identify local recurrence or metastasis as early as possible.

[260]

- CASTELLANO -

TÍTULO / TITLE: Tumores neuroendocrinos do colon e reto: experiencia do Instituto Nacional do Cancer no Brasil.

TÍTULO / TITLE: - Colon and rectum neuroendocrine tumors: experience of the National Cancer Institute in Brazil.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Arq Bras Cir Dig. 2013 Mar;26(1):36-39.

AUTORES / AUTHORS: - Cesar D; Zanatto RM; Silva MV; Golcalves R; Mello EL; Jesus JP

INSTITUCIÓN / INSTITUTION: - Departamento de Cirurgia Abdomino-Pelvica, Instituto Nacional do Cancer, Rio de Janeiro, RJ, Brasil.

RESUMEN / SUMMARY: - BACKGROUND: Neuroendocrine tumors (NETs) are rare, comprising nearly 0.49% of all malignancies. The majority occurs in the gastrointestinal tract. AIM: To analyze the demographic factors, clinicopathologic features, treatment employed, prognostic factors and the oncologic results related to colorectal NETs. METHODS: Between the period from 1996 to 2010 174 patients were treated. From these, 34 were localized in the colon and rectum. Demographic factors, stage, therapeutics and its results were analyzed. All patients were followed for more than three years with image exams, urinary 5-hydroxyindolacetic acid (5-HIAA), serum chromogranin A and prostatic acid phosphatase. RESULTS: The median age was 54,4 years (22-76), the majority was female (64,7%). Out of the 12 patients with colon NETs, one (8.3%) patient was classified as Stage IA; one (8.3%) as Stage IB; three (25%) as Stage IIIB and seven (58.4%) as Stage IV. Out of the 22 patients with rectum NETs, six (27.3%) were classified as Stage IA; four (18.2%) as IB; three (13.6 %) as IIIA; one (4.5%) as IIIB and eight (36.4%) as IV. Of rectal NETs, nine (41%) were treated with endoscopic resection, six (27.2%) underwent conventional surgical treatment and six (27.2%) were treated with chemotherapy. Eleven patients with colon NETs (91.6%) were surgically treated, seven of them with palliative surgery, one (8.4%) was treated with endoscopic resection and no patient was submitted to chemotherapy. After an

average follow-up of 55 months, 19 (55%) patients were alive. Analyzing the overall survival was obtained an average overall survival of 29 months in Stage IA, 62 months in IB, 12 months in IIIA, 31 months in IIIB and 39 months in IV. CONCLUSION: The treatment of colon and rectal NETs is complex, because it depends of the individuality of each patient. With adequate management, the prognosis can be favorable with long survival, but it is related to the tumor differentiation degree, efficacy of the chosen treatment and to the patient adhesion to the follow-up after treatment.

[261]

TÍTULO / TITLE: - Multiple endocrine neoplasia type 1.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Indian J Endocrinol Metab. 2012 Dec;16(Suppl 2):S272-4. doi: 10.4103/2230-8210.104058.

●●Enlace al texto completo (gratis o de pago) 4103/2230-8210.104058

AUTORES / AUTHORS: - Thakker RV

INSTITUCIÓN / INSTITUTION: - Nuffield Department of Medicine, Oxford Centre for Diabetes, Endocrinology and Metabolism, University of Oxford, Headington, Oxford, OX3 7LJ, UK.

RESUMEN / SUMMARY: - Multiple endocrine neoplasia type 1 (MEN1) is characterized by the occurrence of parathyroid, pancreatic islet and anterior pituitary tumors. Some patients may also develop carcinoid tumors, adrenocortical tumors, facial angiofibromas, collagenomas, and lipomas. MEN1 is an autosomal-dominant disorder, due to mutations in the tumor suppressor gene MEN1, which encodes a 610 amino acid protein, menin. Thus, the finding of MEN1 in a patient has important implications for family members because first-degree relatives have a 50% risk of developing the disease and can often be identified by MEN1 mutational analysis. Patients with MEN1 have a decreased life-expectancy and the outcomes of current treatments, which are generally similar to that for the respective tumors occurring in non-MEN1 patients, are not as successful because of multiple tumors, which may be larger, more aggressive, and resistant to treatment, and the concurrence of metastases. The prognosis for MEN1 patients might be improved by pre-symptomatic tumor detection and undertaking treatment specific for MEN1-tumors. Thus, it is recommended that MEN1 patients and their families should be cared for by multi-disciplinary teams comprising relevant specialists with experience in the diagnosis and treatment of patients with endocrine tumors.

[262]

TÍTULO / TITLE: - Glucose stimulation induces dynamic change of mitochondrial morphology to promote insulin secretion in the insulinoma cell line INS-1E.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - PLoS One. 2013;8(4):e60810. doi: 10.1371/journal.pone.0060810. Epub 2013 Apr 2.

●●Enlace al texto completo (gratis o de pago)

[1371/journal.pone.0060810](https://doi.org/10.1371/journal.pone.0060810)

AUTORES / AUTHORS: - Jhun BS; Lee H; Jin ZG; Yoon Y

INSTITUCIÓN / INSTITUTION: - Center for Translational Medicine, Department of Medicine, Thomas Jefferson University, Philadelphia, PA, USA.

RESUMEN / SUMMARY: - Fission and fusion of mitochondrial tubules are the major processes regulating mitochondrial morphology. However, the physiological significance of mitochondrial shape change is poorly understood. Glucose-stimulated insulin secretion (GSIS) in pancreatic beta-cells requires mitochondrial ATP production which evokes Ca(2+) influx through plasma membrane depolarization, triggering insulin vesicle exocytosis. Therefore, GSIS reflects mitochondrial function and can be used for evaluating functional changes associated with morphological alterations of mitochondria. Using the insulin-secreting cell line INS-1E, we found that glucose stimulation induced rapid mitochondrial shortening and recovery. Inhibition of mitochondrial fission through expression of the dominant-negative mutant DLP1-K38A eliminated this dynamic mitochondrial shape change and, importantly, blocked GSIS. We found that abolishing mitochondrial morphology change in glucose stimulation increased the mitochondrial inner membrane proton leak, and thus significantly diminished the mitochondrial ATP producing capacity in response to glucose stimulation. These results demonstrate that dynamic change of mitochondrial morphology is a previously unrecognized component for metabolism-secretion coupling of pancreatic beta-cells by participating in efficient ATP production in response to elevated glucose levels.

[263]

TÍTULO / TITLE: - Primary adenocarcinoma of the lung with a synchronous carcinoid tumour of the small intestine.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - JRSM Short Rep. 2013 Mar;4(3):18. doi: 10.1177/2042533313476416. Epub 2013 Mar 6.

●●Enlace al texto completo (gratis o de pago)

[1177/2042533313476416](https://doi.org/10.1177/2042533313476416)

AUTORES / AUTHORS: - Loewenthal L; Thompson M; Elkin SL

INSTITUCIÓN / INSTITUTION: - Department of Chest & Allergy, Imperial College Healthcare NHS Trust, St Mary's Hospital, Praed Street, London W1 1NY, UK.

[264]

TÍTULO / TITLE: - Merkel cell carcinoma: the past, the present, and the future.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - J Skin Cancer. 2013;2013:929364. doi: 10.1155/2013/929364. Epub 2013 Apr 16.

●●Enlace al texto completo (gratis o de pago) [1155/2013/929364](https://doi.org/10.1155/2013/929364)

AUTORES / AUTHORS: - Erovic I; Erovic BM

INSTITUCIÓN / INSTITUTION: - Medical University of Vienna, 1090 Vienna, Austria.

RESUMEN / SUMMARY: - Since the first description of the Merkel cell carcinoma by Cyril Toker in 1972, the number of studies has significantly increased over the last 4 decades. In this review, we will illustrate the historical background of the Merkel cell carcinoma beginning with the 19th century, the first description of the Merkel cell to the finding of the CK20 as a highly specific diagnostic marker and finally to the recently detected Merkel cell polyomavirus (MCPyV). Moreover, we will highlight the beginning of adjuvant therapeutic regimens with radiotherapy and chemotherapy and discuss the diagnostic work-up including imaging and histology of patients with Merkel cell carcinoma. Another very rapidly growing and interesting field of research is the development of patients' specific and tailored targeted therapy, in particular in patients with distant metastatic disease.

[265]

TÍTULO / TITLE: - Serum biomarkers for neurofibromatosis type 1 and early detection of malignant peripheral nerve-sheath tumors.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - BMC Med. 2013 Apr 23;11:109. doi: 10.1186/1741-7015-11-109.

●●Enlace al texto completo (gratis o de pago) [1186/1741-7015-11-109](https://doi.org/10.1186/1741-7015-11-109)

AUTORES / AUTHORS: - Park SJ; Sawitzki B; Kluwe L; Mautner VF; Holtkamp N; Kurtz A

INSTITUCIÓN / INSTITUTION: - Berlin-Brandenburg Center for Regenerative Therapies, Charite - Universitätsmedizin Berlin, Augustenburger Platz 1, Berlin, 13353, Germany. andreas.kurtz@charite.de.

RESUMEN / SUMMARY: - BACKGROUND: Neurofibromatosis type 1 (NF1) is a hereditary tumor syndrome characterized by the development of benign nerve-sheath tumors, which transform to malignant peripheral nerve-sheath tumors (MPNST) in about 8 to 13% of patients with NF1. MPNST are invasive sarcomas with extremely poor prognosis, and their development may correlate with internal tumor load of patients with NF1. Because early identification of patients with NF1 at risk for developing MPNST should improve their clinical outcome, the aim of this study was to identify serum biomarkers for tumor progression in NF1, and to analyze their correlation with tumor type and internal tumor load. METHODS: We selected candidate biomarkers for NF1 by manually mining published data sources, and conducted a systematic screen of 56 candidate serum biomarkers using customized antibody arrays. Serum from 104 patients with NF1 with and without MPNST, and from 41 healthy control

subjects, was analyzed. Statistical analysis was performed using the non-parametric Mann-Whitney U-test, followed by Bonferroni correction. RESULTS: Our analysis identified four markers (epidermal growth factor receptor, interferon-gamma, interleukin-6, and tumor necrosis factor-alpha) for which significantly different serum concentrations were seen in patients with NF1 compared with healthy controls. Two markers (insulin-like growth factor binding protein 1 (IGFBP1) and regulated upon activation, normal T-cell expressed and secreted (RANTES)) showed significantly higher concentrations in patients with NF1 and MPNST compared with patients with NF1 without MPNST. A correlation with internal tumor load was found for IGFBP1. CONCLUSION: Our study identified two serum markers with potential for early detection of patients with NF1 at risk for developing MPNST, and four markers that could distinguish between patients with NF1 and healthy subjects. Such markers may be useful as diagnostic tools to support the diagnosis of NF1 and for timely identification of MPNST. Moreover, the data suggest that there is a systemic increase in inflammatory cytokines independently of tumor load in patients with NF1.

[266]

TÍTULO / TITLE: - Paraganglioma of the thyroid gland: cytologists' enigma.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - British Medical J (BMJ). Acceso gratuito al texto completo.

●●Enlace a la Editora de la Revista <http://bmj.com/search.dtl>

●●Cita: British Medical J. (BMJ): <> Case Rep. 2013 May 22;2013. pii: bcr2013009518. doi: 10.1136/bcr-2013-009518.

●●Enlace al texto completo (gratis o de pago) 1136/bcr-2013-009518

AUTORES / AUTHORS: - Akhtar K; Sen Ray P; Ahmad SS; Sherwani RK

INSTITUCIÓN / INSTITUTION: - Department of Pathology, Jawaharlal Nehru Medical College, AMU, Aligarh, Uttar Pradesh, India.

RESUMEN / SUMMARY: - Paraganglioma is a neuroendocrine tumour derived from extra-adrenal cells of the neural crest paraganglia of the autonomic nervous system. These rare neoplasms comprise of around 0.012% of head and neck tumours. Paraganglioma arising in the thyroid gland is exceptionally uncommon and can present as a diagnostic challenge on fine-needle aspiration cytology (FNAC). We report a case of primary thyroid paraganglioma in a 19-year-old woman who presented with a solitary thyroid nodule without palpable cervical lymphadenopathy. FNAC from the lesion caused diagnostic dilemma by mimicking follicular neoplasm and C-cell-derived thyroid tumours; final diagnosis was established by histopathology and immunohistochemistry. The main purpose of this case report is to discuss the differential diagnosis and emphasise on the need of immune markers in the diagnosis of thyroid paraganglioma. In view of the uncertain malignant potential of these tumours, a long-term follow-up is recommended.

[267]

TÍTULO / TITLE: - Efficacy of temsirolimus in metastatic chromophobe renal cell carcinoma.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - BMC Urol. 2013 May 21;13(1):26.

●●Enlace al texto completo (gratis o de pago) [1186/1471-2490-13-26](#)

AUTORES / AUTHORS: - Venugopal B; Ansari J; Aitchison M; Tho LM; Campbell R; Jones RJ

RESUMEN / SUMMARY: - BACKGROUND: Renal cell carcinoma (RCC) is a histopathologically and molecularly heterogeneous disease with the chromophobe subtype (chRCC) accounting for approximately 5% of all cases. The median overall survival of advanced RCC has improved significantly since the advent of tyrosine kinase inhibitors and mammalian target of rapamycin (mTOR) inhibitors. However, high-quality evidence for the use of new generation tyrosine kinase inhibitors in patients with advanced chRCC is lacking. Few published case reports have highlighted the use of temsirolimus in chRCC. CASE PRESENTATION: Here, we report the case of a 36-year-old Caucasian woman with metastatic chRCC with predominantly skeletal metastases who was refractory to sunitinib who demonstrated a durable clinical response to temsirolimus lasting 20 months. We review the available evidence pertaining to the use of new generation molecularly targeted agents, in particular mTOR inhibitors in chRCC and discuss their emerging role in the management of this disease which would aid the oncologists faced with the challenge of treating this rare type of RCC. CONCLUSION: Conducting randomised clinical trials in this rarer sub-group of patients would be challenging and our case report and the evidence reviewed would guide the physicians to make informed decision regarding the management of these patients.

[268]

TÍTULO / TITLE: - Synchronous occurrence of advanced neuroendocrine carcinoma and tubular adenocarcinoma of the rectum.

RESUMEN / SUMMARY: - [Enlace al Resumen / Link to its Summary](#)

REVISTA / JOURNAL: - Case Rep Gastroenterol. 2013 Mar 9;7(1):117-21. doi: 10.1159/000350252. Print 2013 Jan.

●●Enlace al texto completo (gratis o de pago) [1159/000350252](#)

AUTORES / AUTHORS: - Nakayama Y; Torigoe T; Minagawa N; Uehara T; Yamaguchi K

INSTITUCIÓN / INSTITUTION: - Department of Surgery 1, School of Medicine, Kitakyushu, Japan ; Department of Gastroenterological and General Surgery, Wakamatsu Hospital of University of Occupational and Environmental Health, Kitakyushu, Japan.

RESUMEN / SUMMARY: - This report presents a rare case with the synchronous occurrence of advanced neuroendocrine carcinoma (NEC) and tubular adenocarcinoma of the rectum. A 52-year-old Japanese male presented with general fatigue and bloody stool. Endoscopic examination showed an ulcerated lesion of the lower rectum. The pathological diagnosis of biopsy specimens from this lesion indicated moderately differentiated adenocarcinoma. He was referred to the surgical outpatient clinic with advanced rectal cancer. Barium enema indicated two lesions in the upper and lower rectum. Computed tomography revealed multiple hepatic metastases. A low anterior resection was performed with lymph node dissection. The resected specimen indicated an elevated lesion with ulceration in the upper rectum and an ulcerated lesion in the lower rectum. Histopathological and immunohistochemical analyses revealed NEC from the upper rectum and moderately differentiated tubular adenocarcinoma from the lower rectum. These two lesions were completely separated from each other. Therefore, this case demonstrates the synchronous occurrence of advanced NEC and tubular adenocarcinoma in the rectum.
