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Artículos originales (todos) * Original articles (all)**

Neuroendocrine tumors.

October / November 2013

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[1]

TÍTULO / TITLE: - Somatic mutation of CDKN1B in small intestine neuroendocrine tumors.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Nat Genet. 2013 Dec;45(12):1483-6. doi: 10.1038/ng.2821. Epub 2013 Nov 3.

●● Enlace al texto completo (gratuito o de pago) 1038/ng.2821

AUTORES / AUTHORS: - Francis JM; Kiezun A; Ramos AH; Serra S; Pedamallu CS; Qian ZR; Banck MS; Kanwar R; Kulkarni AA; Karpathakis A; Manzo V; Contractor T; Philips J; Nickerson E; Pho N; Hooshmand SM; Brais LK; Lawrence MS; Pugh T; McKenna A; Sivachenko A; Cibulskis K; Carter SL; Ojesina AI; Freeman S; Jones RT; Voet D; Saksena G; Auclair D; Onofrio R; Shefler E; Sougnez C; Grimsby J; Green L; Lennon N; Meyer T; Caplin M; Chung DC; Beutler AS; Ogino S; Thirlwell C; Shivdasani R; Asa SL; Harris CR; Getz G; Kulke M; Meyerson M

INSTITUCIÓN / INSTITUTION: - 1] Broad Institute, Cambridge, Massachusetts, USA. [2] Department of Medical Oncology, Dana-Farber Cancer Institute, Boston, Massachusetts, USA. [3].

RESUMEN / SUMMARY: - The diagnosed incidence of small intestine neuroendocrine tumors (SI-NETs) is increasing, and the underlying genomic mechanisms have not yet been defined. Using exome- and genome-sequence analysis of SI-NETs, we identified recurrent somatic mutations and deletions in CDKN1B, the cyclin-dependent kinase inhibitor gene, which encodes p27. We observed frameshift mutations of CDKN1B in 14 of 180 SI-NETs, and we detected hemizygous deletions encompassing CDKN1B in 7 out of 50 SI-NETs, nominating p27 as a tumor suppressor and implicating cell cycle dysregulation in the etiology of SI-NETs.

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[2]

TÍTULO / TITLE: - Loss of DAXX and ATRX are Associated with Chromosome Instability and Reduced Survival of Patients with Pancreatic Neuroendocrine Tumors.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Gastroenterology. 2013 Oct 19. pii: S0016-5085(13)01494-7. doi: 10.1053/j.gastro.2013.10.020.

●● Enlace al texto completo (gratuito o de pago) 1053/j.gastro.2013.10.020

AUTORES / AUTHORS: - Marinoni I; Kurrer AS; Vassella E; Dettmer M; Rudolph T; Banz V; Hunger F; Pasquinelli S; Speel EJ; Perren A

INSTITUCIÓN / INSTITUTION: - Institute of Pathology, University of Bern, Switzerland.

RESUMEN / SUMMARY: - BACKGROUND & AIMS: Sporadic pancreatic neuroendocrine tumors (pNETs) are rare and genetically heterogeneous. Chromosome instability (CIN) has been detected in pNETs from patients with poor outcomes, but no specific genetic factors have been associated with CIN. Mutations in DAXX or ATRX (which both encode proteins involved in chromatin remodeling) have been detected in 40% of pNETs, in association with activation of alternative lengthening of telomeres. We investigated whether loss of DAXX or ATRX, and consequent alternative lengthening of telomeres, are related to CIN in pNETs. We also assessed whether loss of DAXX or ATRX is associated with specific phenotypes of pNETs. METHODS: We collected well-differentiated primary pNET samples from 142 patients at the University Hospital Zurich and 101 patients at the University Hospital Bern, Switzerland. Clinical follow-up data were obtained for 149 patients from general practitioners and tumor registries. The tumors were reclassified into 3 groups according to the 2010 WHO classification. Samples were analyzed by immunohistochemistrey and telomeric fluorescence in situ hybridization. We correlated loss of DAXX, or ARTX expression, and activation of alternative lengthening of telomeres with data from comparative genomic hybridization array studies, as well as with clinical and pathology features of the tumors and relapse and survival data. RESULTS: Loss of DAXX or ARTX protein and alternative lengthening of telomeres were associated with CIN in pNETs. Furthermore, loss of DAXX or ARTX correlated with tumor stage and metastasis, reduced time of relapse-free survival, and decreased time of tumor-associated survival. CONCLUSIONS: Loss of DAXX or ARTX is associated with CIN in pNETs and shorter survival times of patients. These results support the hypothesis that DAXX- and ATRX-negative tumors are a more aggressive subtype of pNET, and could lead to identification of strategies to target CIN in pancreatic tumors.

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[3]

TÍTULO / TITLE: - Are patients with autoimmune thyroid disease and autoimmune gastritis at risk for Gastric Neuroendocrine Tumours type 1?

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Clin Endocrinol (Oxf). 2013 Oct 4. doi: 10.1111/cen.12346.

●● Enlace al texto completo (gratuito o de pago) 1111/cen.12346

AUTORES / AUTHORS: - Alexandraki KI; Nikolaou A; Thomas D; Syriou V; Korkolopoulou P; Sougioultzis S; Kaltsas G

INSTITUCIÓN / INSTITUTION: - Endocrine Unit.

RESUMEN / SUMMARY: - OBJECTIVE: To investigate the prevalence of autoimmune gastritis, Enterochromaffin-like cell (ECL-cell) hyperplasia and Gastric Neuroendocrine tumours type 1 (GNET1) in patients with autoimmune thyroid disease. DESIGN: Prospective observational study in a single institutional study. PATIENTS AND MEASUREMENTS: 120 patients with autoimmune thyroid disease were consecutively recruited from the Endocrine Clinic. Upper gastrointestinal tract endoscopy (UGE) and biochemical parameters for autoimmune thyroid disease and autoimmune gastritis were assessed at recruitment and annually thereafter in patients with a mean follow-up 37.5+/-14.4months. Autoimmune gastritis was defined by the presence of anti-parietal cell antibodies (APCA) and histological confirmation after UGE. Serum gastrin and chromogranin Alpha were also measured. RESULTS: 111 patients had Hashimotos' thyroiditis and 9 Graves' disease. Autoimmune gastritis was identified in 40 (38 with Hashimotos' thyroiditis, 2 with Graves' disease) patients all of whom had increased levels of gastrin and chromogranin Alpha; Helicobacter pylori infection was histologically identified in 15/40 (37.5%). Six patients had isolated nodular ECL-cell hyperplasia and one mixed nodular and linear ECL-cell hyperplasia [7/40 (17.5%)]. Only increased gastrin (p=0.03) levels predicted the presence ECL-cell hyperplasia. A GNET1 developed in one patient with nodular ECL-cell hyperplasia after 39 months of follow-up. CONCLUSIONS: Concomitant autoimmune gastritis was found in 33.3% of patients with autoimmune thyroid disease, 17.5% of whom had ECL-cell hyperplasia that evolved to GNET1 in one (2.5%). Larger studies with longer follow-up are needed to define the incidence of GNET1 in patients with autoimmune thyroid disease and ECL-cell hyperplasia and potential implications.

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[4]

TÍTULO / TITLE: - Comparison of response evaluation in patients with gastroenteropancreatic and thoracic neuroendocrine tumors after treatment with [177Lu-DOTA0,Tyr3]octreotate.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - J Nucl Med. 2013 Oct;54(10):1689-96. doi: 10.2967/jnumed.112.117408.

●● Enlace al texto completo (gratuito o de pago) 2967/jnumed.112.117408

AUTORES / AUTHORS: - van Vliet EI; Krenning EP; Teunissen JJ; Bergsma H; Kam BL; Kwekkeboom DJ

INSTITUCIÓN / INSTITUTION: - Department of Nuclear Medicine, Erasmus MC, University Medical Center, Rotterdam, The Netherlands.

RESUMEN / SUMMARY: - Response Evaluation Criteria In Solid Tumors (RECIST) (unidimensional), Southwest Oncology Group (SWOG) solid tumor response criteria (bidimensional), and their modified variants are commonly used in the tumor response assessment after treatment of gastroenteropancreatic and thoracic neuroendocrine tumors (NETs). In the current study, RECIST, SWOG criteria, modified RECIST (mRECIST), and modified SWOG (mSWOG) criteria were compared in patients with NETs treated with [(177)Lu-DOTA(0),Tyr(3)]octreotate ((177)Lu-octreotate). METHODS: Two-hundred sixty-eight Dutch patients with NETs who had been treated with (177)Lu-octreotate between January 2000 and April 2007 were studied. CT or MR imaging scans were analyzed using RECIST, SWOG criteria, mRECIST, and mSWOG

criteria (including the tumor response class minor response [decrease of 13%-30% for mRECIST and 25%-50% for mSWOG]). The outcomes were correlated with progression-free survival (PFS) and overall survival (OS). RESULTS: Eleven patients had an unknown tumor response and were excluded. The rates of objective response (OR) (complete response + partial response [+minor response for mRECIST/mSWOG]), stable disease, and progressive disease (PD) were 28%, 49%, and 24%, respectively, according to RECIST; 25%, 49%, and 26%, respectively, according to SWOG; 44%, 33%, and 24%, respectively, according to mRECIST; and 45%, 29%, and 26%, respectively, according to mSWOG. In patients who had OR, stable disease, or PD, the median PFS was 26-30, 27-34, and 8 mo, respectively, with any of the 4 response criteria. In patients who had OR, stable disease, or PD, the median OS was 55-57, 56-74, and 11-12 mo, respectively, with any of the 4 response criteria. Subanalyses for patients who had progression before treatment start were comparable. CONCLUSION: Patients with PD as treatment outcome had significantly shorter PFS and OS than patients with an OR or stable disease with all 4 scoring systems. PFS and OS were comparable for patients with tumor regression and stable disease. The addition of the response class minor response did not improve the correlation with PFS and OS. The 4 scoring systems gave comparable results in terms of PFS and OS per categorized outcome.

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[5]

TÍTULO / TITLE: - The Effect of Extent of Surgery and Number of Lymph Node Metastases on Overall Survival in Patients with Medullary Thyroid Cancer.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - J Clin Endocrinol Metab. 2013 Nov 25.

●● Enlace al texto completo (gratuito o de pago) 1210/jc.2013-2942

AUTORES / AUTHORS: - Esfandiari NH; Hughes DT; Yin H; Banerjee M; Haymart MR

INSTITUCIÓN / INSTITUTION: - Nazanene H. Esfandiari, MD, Department of Medicine, Division of Metabolism, Endocrinology, & Diabetes, University of Michigan; David T. Hughes, MD, Department of Surgery, Division of Endocrine Surgery, University of Michigan; Huiying Yin, MS, Department of Surgery, University of Michigan; Mousumi Banerjee, PhD, Department of Biostatistics, University of Michigan; Megan R. Haymart, MD, Department of Medicine, Division of Metabolism, Endocrinology, & Diabetes and Division of Hematology/Oncology, University of Michigan.

RESUMEN / SUMMARY: - Context:Total thyroidectomy with central lymph node dissection is recommended in patients with medullary thyroid cancer (MTC). However, the relationship between disease severity and extent of resection on overall survival remains unknown.Objective:To identify the effect of surgery on overall survival in MTC patients.Methods:Using data from 2,968 patients with MTC diagnosed between 1998-2005 from the National Cancer Database, we determined the relationship between number of cervical lymph nodes metastases, tumor size, distant metastases and extent of surgery on overall survival in patients with MTC.Results:Older patient age [5.69 (3.34-9.72)], larger tumor size [2.89 (2.14-3.90)], presence of distant metastases [5.68 (4.61-6.99)], and number of positive regional lymph nodes [for >/= 16 lymph nodes, 3.40 (2.41-4.79)] were independently associated with decreased survival. Overall survival for patients with cervical lymph nodes resected and negative, cervical lymph nodes not resected, 1-5, 6-10, 11-16 and >/= 16 cervical lymph nodes metastases was

90%, 76%, 74%, 61%, 69% and 55% respectively. There was no difference in survival based on surgical intervention in patients with tumor size </= 2 cm without distant metastases. In patients with tumor size > 2.0 cm and no distant metastases, all surgical treatments resulted in a significant improvement in survival compared to no surgery (P< 0.001). In patients with distant metastases, only total thyroidectomy with regional lymph node resection resulted in a significant improvement in survival (P<0.001).Conclusions:The number of lymph node metastases should be incorporated into MTC staging. Extent of surgery in patients with MTC should be tailored to tumor size and distant metastases.

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[6]
TÍTULO / TITLE: - Neuroendocrine differentiation correlates with hormone receptor expression and decreased survival in patients with invasive breast carcinoma.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Histopathology. 2013 Oct 9. doi: 10.1111/his.12306.
●● Enlace al texto completo (gratuito o de pago) 1111/his.12306
AUTORES / AUTHORS: - Kwon SY; Bae YK; Gu MJ; Choi JE; Kang SH; Lee SJ; Kim A; Jung HR; Kang SH; Oh HK; Park JY
INSTITUCIÓN / INSTITUTION: - Department of Pathology, Keimyung University College of Medicine.
RESUMEN / SUMMARY: - AIMS: Invasive breast carcinoma (IBC) with neuroendocrine (NE) differentiation has been controversial in terms of its definition and clinical outcome. We investigated the incidence and clinical significance of NE differentiation in patients with IBC. METHODS AND RESULTS: We performed immunohistochemistry for NE markers, chromogranin-A and synaptophysin on 1428 IBC samples using tissue microarrays and classified cases with NE differentiation into two groups, focal (1-49% tumor cells positive for any NE marker) and diffuse (>/=50% tumor cells positive) groups. Fifty-nine cases (4.1%) showed NE differentiation immunohistochemically and the majority did not show typical NE morphology. Presence of NE differentiation showed a significant association with positive estrogen receptor (P = 0.001) and progesterone receptor (P = 0.008) status. Patients with NE differentiation showed worse overall survival and disease-free survival than those without NE differentiation in both univariate (P < 0.001 for both) and multivariate (OS, P = 0.004; DFS, P < 0.001) analyses. CONCLUSIONS: IBC with NE differentiation is a distinct subtype of mammary carcinoma with an aggressive clinical outcome.

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[7]
TÍTULO / TITLE: - Effects of intercellular junction protein expression on intracellular ice formation in mouse insulinoma cells.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Biophys J. 2013 Nov 5;105(9):2006-15. doi: 10.1016/j.bpj.2013.09.028.
●● Enlace al texto completo (gratuito o de pago) 1016/j.bpj.2013.09.028
AUTORES / AUTHORS: - Higgins AZ; Karlsson JO

INSTITUCIÓN / INSTITUTION: - School of Chemical, Biological and Environmental Engineering, Oregon State University, Corvallis, Oregon.

RESUMEN / SUMMARY: - The development of cryopreservation procedures for tissues has proven to be difficult in part because cells within tissue are more susceptible to intracellular ice formation (IIF) than are isolated cells. In particular, previous studies suggest that cell-cell interactions increase the likelihood of IIF by enabling propagation of ice between neighboring cells, a process thought to be mediated by gap junction channels. In this study, we investigated the effects of cell-cell interactions on IIF using three genetically modified strains of the mouse insulinoma cell line MIN6, each of which expressed key intercellular junction proteins (connexin-36, E-cadherin, and occludin) at different levels. High-speed video cryomicroscopy was used to visualize the freezing process in pairs of adherent cells, revealing that the initial IIF event in a given cell pair was correlated with a hitherto unrecognized precursor phenomenon: penetration of extracellular ice into paracellular spaces at the cell-cell interface. Such paracellular ice penetration occurred in the majority of cell pairs observed, and typically preceded and colocalized with the IIF initiation events. Paracellular ice penetration was generally not observed at temperatures >-5.65 degrees C, which is consistent with a penetration mechanism via defects in tight-junction barriers at the cell-cell interface. Although the maximum temperature of paracellular penetration was similar for all four cell strains, genetically modified cells exhibited a significantly higher frequency of ice penetration and a higher mean IIF temperature than did wild-type cells. A four-state Markov chain model was used to quantify the rate constants of the paracellular ice penetration process, the penetration-associated IIF initiation process, and the intercellular ice propagation process. In the initial stages of freezing (>-15 degrees C), junction protein expression appeared to only have a modest effect on the kinetics of propagative IIF, and even cell strains lacking the gap junction protein connexin-36 exhibited nonnegligible ice propagation rates.

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[8]

TÍTULO / TITLE: - Case 200: gastric enterochromaffinlike cell tumors in a patient with type 1 multiple endocrine neoplasia.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Radiology. 2013 Dec;269(3):940-4. doi: 10.1148/radiol.13120927.

●● Enlace al texto completo (gratuito o de pago) 1148/radiol.13120927

AUTORES / AUTHORS: - Piver D; Ronot M; Guedj N; Hentic O; Vilgrain V

INSTITUCIÓN / INSTITUTION: - From the Departments of Radiology (D.P., M.R., V.V.), Pathology (N.G.), and Pancreatology (O.H.), Hopital Beaujon, 100 bd du General Leclerc, 92118 Clichy, France; and INSERM U773, Centre de Recherche Biomedicale Bichat-Beaujon, CRB3, Paris, France (M.R., V.V.).

RESUMEN / SUMMARY: - History A 55-year-old man presented with chronic epigastric pain lasting for about 1 year and without fever or vomiting. The abdomen was soft and tender at physical examination. Laboratory tests revealed unremarkable liver function, normal hemoglobin level, and normal amylase level. White blood cell count was normal, and there was no inflammatory syndrome. The patient’s medical history included pancreatic gastrinoma resected by means of left pancreatectomy 31 years before, hyperparathyroidism treated with subtotal parathyroidectomy 24 years before,

and a slowly growing lung mass known for 9 years. Esophagogastroduodenoscopy was performed because of a suspected gastroduodenal ulcer. The results showed numerous small (<10 mm) gastric and duodenal ulcers and multiple 10-15-mm polypoid gastric masses. Contrast material-enhanced dual-phase multidetector row computed tomography (CT) of the chest and abdomen was performed with a 64-section CT scanner (LightSpeed VCT; GE Healthcare, Milwaukee, Wis). Technical parameters for CT were as follows: pitch, 0.98; section thickness and reconstruction interval, 1.25 mm; 120 kVp; and variable milliamperage determined by x-, y-, and z-axis dose modulation. After an unenhanced abdominal scan, iobitridol, a nonionic iodinated contrast agent containing 350 mg of iodine per milliliter (Xenetix 350; Guerbet, Aulnay-sousbois, France), was administered intravenously through a 16-18-gauge catheter. A 120-mL dose of the contrast agent was injected via an antecubital vein at a rate of 4 mL/sec. No oral contrast medium was administered. After preliminary unenhanced abdominal scanning, arterial and portal venous phase acquisitions were obtained 45 and 80 seconds after initiation of contrast medium injection.

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[9]

TÍTULO / TITLE: - Phylogenetic analysis of Merkel cell polyomavirus based on full-length LT and VP1 gene sequences derived from neoplastic tumours in Japanese patients.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - J Gen Virol. 2013 Oct 9. doi: 10.1099/vir.0.058149-0.

●● Enlace al texto completo (gratuito o de pago) 1099/vir.0.058149-0

AUTORES / AUTHORS: - Hashida Y; Imajoh M; Kamioka M; Taniguchi A; Kuroda N; Hayashi K; Nakajima H; Sano S; Daibata M

INSTITUCIÓN / INSTITUTION: - Kochi Medical School, Kochi University;

RESUMEN / SUMMARY: - Most Merkel cell polyomavirus (MCPyV) gene sequences have been reported from Western countries and few data are available for the virus sequences from other geographical areas, especially Asia. Thus, we performed phylogenetic analyses based on the nucleotide sequences of the full-length large T antigen (LT) and viral protein 1 (VP1) genes derived from a variety of cancers in Japanese patients and compared them with sequences from Caucasians. The LT and VP1 gene-based phylogenetic trees identified two main genetic clades. One clade comprised strains isolated from Caucasians, whereas all of the Japanese tumor-derived MCPyV strains belonged to another clade. These findings confirm that most of the MCPyV strains present in Japan form a clade, distinct from Caucasian strains.

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[10]

TÍTULO / TITLE: - Free somatostatin receptor fraction predicts the antiproliferative effect of octreotide in a neuroendocrine tumor model: implications for dose optimization.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Cancer Res. 2013 Dec 1;73(23):6865-73. doi: 10.1158/0008-5472.CAN-13-1199. Epub 2013 Sep 30.

●● Enlace al texto completo (gratuito o de pago) 1158/0008-5472.CAN-13-1199

AUTORES / AUTHORS: - Heidari P; Wehrenberg-Klee E; Habibollahi P; Yokell D; Kulke M; Mahmood U

INSTITUCIÓN / INSTITUTION: - Authors' Affiliations: Division of Nuclear Medicine and Molecular Imaging, Department of Radiology, Massachusetts General Hospital, Harvard Medical School and Department of Medical Oncology, Dana-Farber Cancer Institute, Harvard Medical School, Boston, Massachusetts.

RESUMEN / SUMMARY: - Somatostatin receptors (SSTR) are highly expressed in well-differentiated neuroendocrine tumors (NET). Octreotide, an SSTR agonist, has been used to suppress the production of vasoactive hormones and relieve symptoms of hormone hypersecretion with functional NETs. In a clinical trial, an empiric dose of octreotide treatment prolonged time to tumor progression in patients with small bowel neuroendocrine (carcinoid) tumors, irrespective of symptom status. However, there has yet to be a dose optimization study across the patient population, and methods are currently lacking to optimize dosing of octreotide therapy on an individual basis. Multiple factors such as total tumor burden, receptor expression levels, and nontarget organ metabolism/excretion may contribute to a variation in SSTR octreotide occupancy with a given dose among different patients. In this study, we report the development of an imaging method to measure surface SSTR expression and occupancy level using the PET radiotracer (68)Ga-DOTATOC. In an animal model, SSTR occupancy by octreotide was assessed quantitatively with (68)Ga-DOTATOC PET, with the finding that increased occupancy resulted in decreased tumor proliferation rate. The results suggested that quantitative SSTR imaging during octreotide therapy has the potential to determine the fractional receptor occupancy in NETs, thereby allowing octreotide dosing to be optimized readily in individual patients. Clinical trials validating this approach are warranted. Cancer Res; 73(23); 6865-73. ©2013 AACR.

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[11]

TÍTULO / TITLE: - Bedside to bench: role of muscarinic receptor activation in ultrarapid growth of colorectal cancer in a patient with pheochromocytoma.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Mayo Clin Proc. 2013 Nov;88(11):1340-6. doi: 10.1016/j.mayocp.2013.06.023. Epub 2013 Oct 4.

●● Enlace al texto completo (gratuito o de pago) 1016/j.mayocp.2013.06.023

AUTORES / AUTHORS: - von Rosenvinge EC; Cheng K; Drachenberg CB; Fowler CB; Evers DL; Xie G; Raufman JP

INSTITUCIÓN / INSTITUTION: - Division of Gastroenterology & Hepatology, University of Maryland School of Medicine, Baltimore, MD; VA Maryland Health Care System, Baltimore, MD. Electronic address: evonrose@medicine.umaryland.edu.

RESUMEN / SUMMARY: - An elderly man with long-standing, nonresectable pheochromocytoma had rapid development of rectal adenocarcinoma despite close endoscopic surveillance. We determined that the patient's colorectal cancer overexpressed muscarinic receptor subtype 3, whereas his pheochromocytoma expressed choline acetyltransferase, an enzyme required to produce acetylcholine, which is a muscarinic receptor agonist. These findings suggested that acetylcholine release from the pheochromocytoma stimulated rapid growth of the rectal neoplasm. As proof of principle, we found that culture media conditioned by pheochromocytoma

cells stimulates proliferation of a human colon cancer cell line, an effect attenuated by atropine, a muscarinic receptor inhibitor. Our observations provide both clinical and laboratory evidence that muscarinic receptor agonists promote the growth of colorectal neoplasia.

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[12]
TÍTULO / TITLE: - MEN1 Gene Mutation and Reduced Expression Are Associated with Poor Prognosis in Pulmonary Carcinoids.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - J Clin Endocrinol Metab. 2013 Nov 25.
●● Enlace al texto completo (gratuito o de pago) 1210/jc.2013-2782
AUTORES / AUTHORS: - Swarts DR; Scarpa A; Corbo V; Van Criekinge W; van Engeland M; Gatti G; Henfling ME; Papotti M; Perren A; Ramaekers FC; Speel EJ; Volante M
INSTITUCIÓN / INSTITUTION: - 1Departments of Molecular Cell Biology and.
RESUMEN / SUMMARY: - Context:MEN1 gene alterations have been implicated in lung carcinoids, but their effect on gene expression and disease outcome are unknown.Objective:To analyse MEN1 gene and expression anomalies in lung neuroendocrine neoplasms (NENs) and their correlations with clinicopathologic data and disease outcome.Design:We examined 74 lung NENs including 58 carcinoids and 16 high-grade neuroendocrine carcinomas (HGNECs) for MEN1 mutations (n=70) and allelic losses (n=69), promoter hypermethylation (n=65), and mRNA (n=74) expression. Results were correlated with disease outcome.Results:MEN1 mutations were found in 7/55 (13%) carcinoids and in 1 HGNEC, mostly associated with loss of the second allele. MEN1 decreased expression levels correlated with the presence of mutations (P=0.0060) and was also lower in HGNECs than carcinoids (P=0.0024). MEN1 methylation was not associated with mRNA expression levels. Patients with carcinoids harbouring MEN1 mutation and loss had shorter overall survival (P=0.039 and P=0.035, respectively), and low MEN1 mRNA levels correlated with distant metastasis (P=0.00010) and shorter survival (P=0.0071). In multivariate analysis, stage and MEN1 allelic loss were independent predictors of prognosis.Conclusion:Thirteen percent of pulmonary carcinoids harbour MEN1 mutation, associated with reduced mRNA expression and poor prognosis. Also in mutation-negative tumours, low MEN1 gene expression correlates with an adverse disease outcome. Hypermethylation was excluded as the underlying mechanism.

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[13]
TÍTULO / TITLE: - Whole exome sequencing is an efficient and sensitive method for detection of germline mutations in patients with phaeochromocytomas and paragangliomas.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Clin Endocrinol (Oxf). 2013 Sep 19. doi: 10.1111/cen.12331.
●● Enlace al texto completo (gratuito o de pago) 1111/cen.12331
AUTORES / AUTHORS: - McInerney-Leo AM; Marshall MS; Gardiner B; Benn DE; McFarlane J; Robinson BG; Brown MA; Leo PJ; Clifton-Bligh RJ; Duncan EL

INSTITUCIÓN / INSTITUTION: - The University of Queensland Diamantina Institute, Translational Research Institute, Princess Alexandra Hospital, Woolloongabba, Brisbane, Australia.

RESUMEN / SUMMARY: - BACKGROUND: Genetic testing is recommended when the probability of a disease-associated germline mutation exceeds 10%. Germline mutations are found in approximately 25% of individuals with phaeochromocytoma (PCC) or paraganglioma (PGL); however, genetic heterogeneity for PCC/PGL means many genes may require sequencing. A phenotype-directed iterative approach may limit costs but may also delay diagnosis, and will not detect mutations in genes not previously associated with PCC/PGL. OBJECTIVE: To assess whether whole exome sequencing (WES) was efficient and sensitive for mutation detection in PCC/PGL. METHODS: Whole exome sequencing was performed on blinded samples from eleven individuals with PCC/PGL and known mutations. Illumina TruSeq (Illumina Inc, San Diego, CA, USA) was used for exome capture of seven samples, and NimbleGen SeqCap EZ v3.0 (Roche NimbleGen Inc, Basel, Switzerland) for five samples (one sample was repeated). Massive parallel sequencing was performed on multiplexed samples. Sequencing data were called using Genome Analysis Toolkit and annotated using annovar. Data were assessed for coding variants in RET, NF1, VHL, SDHD, SDHB, SDHC, SDHA, SDHAF2, KIF1B, TMEM127, EGLN1 and MAX. Target capture of five exome capture platforms was compared. RESULTS: Six of seven mutations were detected using Illumina TruSeq exome capture. All five mutations were detected using NimbleGen SeqCap EZ v3.0 platform, including the mutation missed using Illumina TruSeq capture. Target capture for exons in known PCC/PGL genes differs substantially between platforms. Exome sequencing was inexpensive (<$A800 per sample for reagents) and rapid (results <5 weeks from sample reception). CONCLUSION: Whole exome sequencing is sensitive, rapid and efficient for detection of PCC/PGL germline mutations. However, capture platform selection is critical to maximize sensitivity.

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[14]

TÍTULO / TITLE: - The case of a patient affected by primary gliosarcoma and neuroendocrine pancreatic cancer with prolonged survival.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Tumori. 2013 May-Jun;99(3):117e-9e. doi: 10.1700/1334.14818.

●● Enlace al texto completo (gratuito o de pago) 1700/1334.14818

AUTORES / AUTHORS: - Trignani M; Taraborrelli M; Ausili Cefaro G

RESUMEN / SUMMARY: - Primary gliosarcoma (PGS) is a rare neoplasm with a poor prognosis. It is considered as a variant of glioblastoma multiforme (GBM) and as a grade IV neoplasm. There is little evidence on the optimal therapy for this disease: treatment of PGS includes surgery, radiotherapy and chemotherapy, and often the same treatment used for GBM is employed for PGS. Several studies have demonstrated that somatostatin receptors are overexpressed in gliomas; somatostatin analogues could therefore also be employed in this mixed form but to date the experience reported in the literature is unclear and there are no studies about the use of these agents in PGS. We present the case of a patient affected by both PGS and neuroendocrine pancreatic cancer. The case is interesting for the prolonged survival

and for the stabilization of disease obtained during therapy with somatostatin analogues.

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[15]

TÍTULO / TITLE: - Long-Term Outcome and Toxicity After Dose-Intensified Treatment with 131I-MIBG for Advanced Metastatic Carcinoid Tumors.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - J Nucl Med. 2013 Dec;54(12):2032-8. doi: 10.2967/jnumed.112.119313. Epub 2013 Oct 7.

●● Enlace al texto completo (gratuito o de pago) 2967/jnumed.112.119313

AUTORES / AUTHORS: - Ezziddin S; Sabet A; Logvinski T; Alkawaldeh K; Yong-Hing CJ; Ahmadzadehfar H; Grunwald F; Biersack HJ

INSTITUCIÓN / INSTITUTION: - Department of Nuclear Medicine, University Hospital, Bonn, Germany.

RESUMEN / SUMMARY: - Reported experience with systemic (131)I-metaiodobenzylguanidine ((131)I-MIBG) therapy of neuroendocrine tumors comprises different dosing schemes. The aim of this study was to assess the long-term outcome and toxicity of treatment with 11.1 GBq (300 mCi) of (131)I-MIBG per cycle. METHODS: We performed a retrospective review of 31 patients with advanced metastatic neuroendocrine tumors (20 with carcinoid tumors and 11 with other tumors) treated with (131)I-MIBG. Treatment outcome was analyzed for patients with carcinoid tumors (the most common tumors in this study), and toxicity was analyzed for the entire patient cohort (n = 31). Treatment comprised 11.1 GBq (300 mCi) per course and minimum intervals of 3 mo. The radiographic response was classified according to modified Response Evaluation Criteria in Solid Tumors. Toxicity was determined according to Common Terminology Criteria for Adverse Events (version 3.0) for all laboratory data at regular follow-up visits and during outpatient care, including complete blood counts and hepatic and renal function tests. Survival analysis was performed with the Kaplan-Meier curve method (log rank test; $P < 0.05$). RESULTS: The radiographic responses in patients with carcinoid tumors comprised a minor response in 2 patients (10%), stable disease in 16 patients (80%; median time to progression, 34 mo), and progressive disease in 2 patients (10%). The symptomatic responses in patients with functioning carcinoid tumors comprised complete resolution in 3 of the 11 evaluable symptomatic patients (27%), partial resolution in 6 patients (55%), and no significant change in 11 patients. The median overall survival in patients with carcinoid tumors was 47 mo (95% confidence interval, 32-62), and the median progression-free survival was 34 mo (95% confidence interval, 13-55). Relevant treatment toxicities were confined to transient myelosuppression of grade 3 or 4 in 15.3% (leukopenia) and 7.6% (thrombocytopenia) of applied cycles and a suspected late adverse event (3% of patients), myelodysplastic syndrome, after a cumulative administered activity of 66.6 GBq. The most frequent nonhematologic side effect was mild nausea (grade 1 or 2), which was observed in 28% of administered cycles. No hepatic or renal toxicities were noted. CONCLUSION: Dose-intensified treatment with (131)I-MIBG at a fixed dose of 11.1 GBq (300 mCi) per cycle is safe and offers effective palliation of symptoms and disease stabilization in patients with advanced carcinoid tumors. The favorable survival and limited toxicity suggest that high cycle

activities are suitable and that this modality may be used for targeted carcinoid treatment-either as an alternative or as an adjunct to other existing therapeutic options.

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[16]
TÍTULO / TITLE: - Do Merkel Cell Polyomavirus Positive Merkel Cell Carcinoma Cells Require Expression of the Viral Small T Antigen?
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - J Invest Dermatol. 2013 Nov 12. doi: 10.1038/jid.2013.486.
●● Enlace al texto completo (gratuito o de pago) 1038/jid.2013.486
AUTORES / AUTHORS: - Angermeyer S; Hesbacher S; Becker JC; Schrama D; Houben R
INSTITUCIÓN / INSTITUTION: - Department of Dermatology, University Hospital Wurzburg, Wurzburg, Germany.

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[17]
TÍTULO / TITLE: - Use of social media to conduct a cross-sectional epidemiologic and quality of life survey of patients with neuroendocrine carcinoma of the cervix: A feasibility study.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Gynecol Oncol. 2013 Oct 19. pii: S0090-8258(13)01264-X. doi: 10.1016/j.ygyno.2013.10.015.
●● Enlace al texto completo (gratuito o de pago) 1016/j.ygyno.2013.10.015
AUTORES / AUTHORS: - Zaid T; Burzawa J; Basen-Engquist K; Bodurka DC; Ramondetta LM; Brown J; Frumovitz M
INSTITUCIÓN / INSTITUTION: - Department of Gynecologic Oncology and Reproductive Medicine, The University of Texas MD Anderson Cancer Center, Houston, TX, USA.
RESUMEN / SUMMARY: - OBJECTIVE: To determine the feasibility of using social media to perform cross-sectional epidemiologic and quality-of-life research on patients with rare gynecologic tumors, we performed a survey of patients with neuroendocrine tumors of the cervix using Facebook. METHODS: After approval from our Institutional Review Board, a support group of patients with neuroendocrine tumors of the cervix was identified on Facebook. Group members were asked to complete a survey comprising 84 questions evaluating clinical presentation; treatment; recurrence; quality of life; and sexual function. RESULTS: The survey was posted for 30days, during which 57 women responded from 8 countries across 4 continents treated at 51 centers. All respondents provided a detailed clinical and tumor history. The mean age was 38.5years. The stage distribution was stage I, 36 patients (63%); II, 13 (23%); III, 2 (4%); and IV, 6 (11%). Forty-nine patients (86%) had small cell and 8 (14%) had large cell tumors. Forty-five of the respondents (79%) had completed primary therapy and were without evidence of disease. Five (9%) had recurrence, 3 (5%) had persistent disease after therapy, and 4 (7%) were still under treatment. Forty-one patients (72%) reported symptoms at time of presentation. Thirty-seven patients (65%) received multimodality primary therapy. Quality of life instruments demonstrated high scores for anxiety and a negative impact of anxiety and cancer on functional and emotional well-being. Sexual function scores did not differ significantly between respondents and the PROMIS reference population. CONCLUSIONS: Use of a social media network to

perform epidemiologic and quality of life research on patients with rare gynecologic tumors is feasible and permits such research to be conducted efficiently and rapidly.

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[18]

TÍTULO / TITLE: - Succinate-to-Fumarate Ratio as a New Metabolic Marker to Detect the Presence of SDHB/D-related Paraganglioma: Initial Experimental and Ex Vivo Findings.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Endocrinology. 2013 Nov 4.

●● Enlace al texto completo (gratuito o de pago) 1210/en.2013-1549

AUTORES / AUTHORS: - Lendvai N; Pawlosky R; Bullova P; Eisenhofer G; Patocs A; Veech RL; Pacak K

INSTITUCIÓN / INSTITUTION: - Program in Reproductive and Adult Endocrinology (N.L., P.B., K.P.), Eunice Kennedy Shriver National Institute of Child Health and Human Development, National Institutes of Health, Bethesda, Maryland 20892; Second Department of Medicine (N.L.), Semmelweis University, Budapest, Hungary; Section on Metabolic Control Analysis (R.P., R.L.V.), National Institute on Alcohol Abuse and Alcoholism, National Institutes of Health, Rockville, Maryland 20852; Department of Molecular Medicine (P.B.), Institute of Virology, Slovak Academy of Sciences, Bratislava, Slovak Republic; Institute of Clinical Chemistry and Laboratory Medicine (G.E.), University Hospital Carl Gustav Carus at the TU Dresden, Dresden, Germany; Department of Medicine IotaIotal (G.E.), University Hospital Carl Gustav Carus at the TU Dresden, Dresden, Germany; Molecular Medicine Research Group (A.P.), Hungarian Academy of Sciences and Semmelweis University, Budapest, Hungary; and Department of Laboratory Medicine Institute (A.P.), Central Isotope Laboratory, Semmelweis University, Budapest, Hungary.

RESUMEN / SUMMARY: - Pheochromocytomas (PHEOs) and paragangliomas (PGLs; extra-adrenal tumors) are rare neuroendocrine chromaffin cell tumors with a hereditary background in about 30-35%. Those caused by succinate dehydrogenase subunit B (SDHB) germline mutations are associated with a high metastatic potential and ultimately higher patient mortality. Succinate dehydrogenase converts succinate to fumarate, uniquely linking the Krebs cycle and oxidative phosphorylation. SDH mutations result in the accumulation of succinate associated with various metabolic disturbances and the shift to aerobic glycolysis in tumor tissue. In the present study, we measured succinate and fumarate levels in mouse pheochromocytoma cells and mouse tumor tissue (MTT) and in 10 apparently sporadic, 10 SDHB-, 5 SDHD-, and 2 neurofibromatosis 1-related PHEOs/PGLs and plasma samples using mass spectrometry. We found that the succinate-to-fumarate ratio was significantly higher in the SDHB- and SDHD-related PGLs than in apparently sporadic and neurofibromatosis 1-related PHEOs/PGLs ($P = .0376$). To further support our data, we silenced SDHB expression in mouse pheochromocytoma cells and MTT cells and evaluated the succinate and fumarate levels. Compared with control samples, SDHB-silenced MTT cells also showed an increase in the succinate-to-fumarate ratio (MTT cells: 2.45 vs 7.53), similar to the findings in SDHB-related PGLs. The present findings for the first time demonstrate a significantly increased succinate-to-fumarate ratio in SDHB/D-related PGLs and thus suggest this ratio may be used as a new metabolic marker for the detection of SDHB/D-related PHEOs/PGLs.

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[19]

TÍTULO / TITLE: - Merkel Cell Polyomavirus Small T Antigen Targets the NEMO Adaptor Protein To Disrupt Inflammatory Signaling.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - J Virol. 2013 Dec;87(24):13853-67. doi: 10.1128/JVI.02159-13. Epub 2013 Oct 9.

●● Enlace al texto completo (gratuito o de pago) 1128/JVI.02159-13

AUTORES / AUTHORS: - Griffiths DA; Abdul-Sada H; Knight LM; Jackson BR; Richards K; Prescott EL; Peach AH; Blair GE; Macdonald A; Whitehouse A

INSTITUCIÓN / INSTITUTION: - School of Molecular and Cellular Biology, Faculty of Biological Sciences.

RESUMEN / SUMMARY: - Merkel cell carcinoma (MCC) is a highly aggressive nonmelanoma skin cancer arising from epidermal mechanoreceptor Merkel cells. In 2008, a novel human polyomavirus, Merkel cell polyomavirus (MCPyV), was identified and is strongly implicated in MCC pathogenesis. Currently, little is known regarding the virus-host cell interactions which support virus replication and virus-induced mechanisms in cellular transformation and metastasis. Here we identify a new function of MCPyV small T antigen (ST) as an inhibitor of NF-kappaB-mediated transcription. This effect is due to an interaction between MCPyV ST and the NF-kappaB essential modulator (NEMO) adaptor protein. MCPyV ST expression inhibits IkappaB kinase alpha (IKKalpha)/IKKbeta-mediated IkappaB phosphorylation, which limits translocation of the NF-kappaB heterodimer to the nucleus. Regulation of this process involves a previously undescribed interaction between MCPyV ST and the cellular phosphatase subunits, protein phosphatase 4C (PP4C) and/or protein phosphatase 2A (PP2A) Abeta, but not PP2A Aalpha. Together, these results highlight a novel function of MCPyV ST to subvert the innate immune response, allowing establishment of early or persistent infection within the host cell.

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[20]

TÍTULO / TITLE: - Regional Lymphadenectomy Is Indicated in the Surgical Treatment of Pancreatic Neuroendocrine Tumors (PNETs).

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Ann Surg. 2013 Nov 18.

●● Enlace al texto completo (gratuito o de pago)
1097/SLA.0000000000000348

AUTORES / AUTHORS: - Hashim YM; Trinkaus KM; Linehan DC; Strasberg SS; Fields RC; Cao D; Hawkins WG

INSTITUCIÓN / INSTITUTION: - *Department of Surgery and the Alvin J. Siteman Cancer Center, Barnes-Jewish Hospital and Washington University School of Medicine, St. Louis, MO daggerDivision of Biostatistics, Washington University School of Medicine, St. Louis, MO double daggerDepartment of Pathology and Immunology, Washington University School of Medicine, St Louis, MO, and Department of Pathology, Key Laboratory of Carcinogenesis and Translational Research (Ministry of Education), Peking University Cancer Hospital, Beijing, China.

RESUMEN / SUMMARY: - OBJECTIVE:: To explore the prognostic importance and preoperative predictors of lymph node metastasis in an effort to guide surgical decision making in patients with pancreatic neuroendocrine tumors (PNETs). BACKGROUND:: PNETs are uncommon, and the natural history of the disease is not well described. As a result, there remains controversy regarding the optimal management of regional lymph nodes during resection of the primary tumor. METHODS:: A retrospective review of a prospectively maintained database of patients who underwent surgery for locoregional PNET between 1994 and 2012 was performed. Logistic regression was used to identify predictors of nodal metastasis. Overall survival and disease-free survival were calculated using Kaplan-Meier method. Results were expressed as P values and odds ratio estimates, with 95% confidence intervals. RESULTS:: One hundred thirty-six patients were identified, of whom 50 (38%) patients had nodal metastasis. The frequency of lymph node metastasis was higher for larger tumors [> 1.5 cm (odds ratio [OR] = 4.7)], tumors of the head as compared with body-tail of the pancreas (OR = 2.8), tumors with Ki-67 greater than 20% (OR = 6.7), and tumors with lymph vascular invasion (OR = 3.6) ($P < 0.05$). Median disease-free survival was lower for patients with nodal metastases (4.5 vs 14.6 years, $P < 0.0001$). CONCLUSIONS:: Lymph node metastasis is predictive of poor outcomes in patients with PNETs. Preoperative variables are not able to reliably predict patients where the probability of lymph node involvement was less than 12%. These data support inclusion of regional lymphadenectomy in patients undergoing pancreatic resections for PNET.

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[21]

TÍTULO / TITLE: - Familial history of non-medullary thyroid cancer is an independent prognostic factor for tumor recurrence in younger patients with conventional papillary thyroid carcinoma.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - J Surg Oncol. 2013 Oct 16. doi: 10.1002/jso.23447.

●● Enlace al texto completo (gratuito o de pago) 1002/jso.23447

AUTORES / AUTHORS: - Lee YM; Yoon JH; Yi O; Sung TY; Chung KW; Kim WB; Hong SJ

INSTITUCIÓN / INSTITUTION: - Department of Surgery, Asan Medical Center, University of Ulsan College of Medicine, Seoul, Korea.

RESUMEN / SUMMARY: - BACKGROUND: It is not clear whether familial non-medullary thyroid cancer (FNMTC) is more aggressive and has a poorer prognosis, than sporadic carcinoma. Therefore, the optimal clinical approach for FNMTC is yet to be established. In this study, we investigated the biological behavior and prognosis of FNMTC compared with its sporadic counterpart. METHODS: Between 1996 and 2004, 1,262 patients underwent a total thyroidectomy for conventional PTC at Asan Medical Center and 113 (9.0%) were diagnosed with FNMTC. We compared the clinico-pathologic characteristics, treatment modalities, and prognosis between familial and sporadic NMTC. RESULTS: FNMTC was significantly more multi-centric than sporadic. We also found that family history itself was an independent risk factor for recurrence. Moreover, disease-free survival in the familial group was significantly shorter than in the sporadic group in the subgroups in which age was <45 years, and in which the tumors were multi-centric, bilateral, and of N1b node status. CONCLUSION: FNMTC may be considered as a separate clinical entity with a higher rate of recurrence and

worse DFS than its sporadic counterpart. Furthermore, familial history of NMTC is an independent risk factor for recurrence, especially in younger patients with conventional PTC. J. Surg. Oncol. © 2013 Wiley Periodicals, Inc.

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[22]

TÍTULO / TITLE: - Is Intraoperative Calcitonin Monitoring Useful to Modulate the Extension of Neck Dissection in Patients With Medullary Thyroid Carcinoma?

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - World J Surg. 2013 Nov 20.

●● Enlace al texto completo (gratuito o de pago) 1007/s00268-013-2328-7

AUTORES / AUTHORS: - De Crea C; Raffaelli M; Milano V; Carrozza C; Zuppi C; Bellantone R; Lombardi CP

INSTITUCIÓN / INSTITUTION: - Istituto di Semeiotica Chirurgica, U.O. Chirurgia Endocrina e Metabolica, Universita Cattolica del Sacro Cuore, Policlinico A. Gemelli, L.go A. Gemelli 8, 00168, Rome, Italy.

RESUMEN / SUMMARY: - BACKGROUND: The extension of the compartment-oriented neck dissection at primary surgery in medullary thyroid carcinoma (MTC) is controversial. Because a <50 % decrease in intraoperative calcitonin levels (IO-CT) after total thyroidectomy plus central neck dissection (TT-CND) has been associated with residual disease, IO-CT monitoring has been proposed to predict the completeness of surgery. The goal of the present prospective study was to verify the accuracy of IO-CT monitoring. METHODS: All patients scheduled for primary surgery for suspected or proven MTC between November 2010 and January 2013 were included. Calcitonin was measured pre-incision (basal level), after tumor manipulation, at the time TT-CND was accomplished (ablation level), 10 and 30 min after ablation. A decrease >50 % with respect to the highest IO-CT level 30 min after ablation was considered predictive of cure. RESULTS: Twenty-six patients were included, and IO-CT monitoring identified 18 of 23 cured patients (true negative results) and 2 of 3 patients with persistent disease (true positive result). In 5 patients with normal basal and stimulated postoperative calcitonin levels, a decrease <50 % was observed (false positive results). In one of three patients with persistent disease a >50 % decrease in IO-CT was observed (false negative results). Specificity, sensitivity, and accuracy of IO-CT were 78.2, 66.6, and 76.9 %, respectively. CONCLUSIONS: Intraoperative calcitonin monitoring is not highly accurate in predicting the completeness of surgical resection. In the present series, relying on IO-CT would result in limited resection in about one third of the patients with residual neck disease and in unnecessary lateral neck dissection in about 20 % of the cured patients.

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[23]

TÍTULO / TITLE: - CASE 6-2013 Perioperative Management of an Adult Patient With Tetralogy of Fallot and Pheochromocytoma.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - J Cardiothorac Vasc Anesth. 2013 Dec;27(6):1399-406. doi: 10.1053/j.jvca.2013.07.009. Epub 2013 Oct 1.

●● Enlace al texto completo (gratuito o de pago) 1053/j.jvca.2013.07.009

AUTORES / AUTHORS: - Tang YK; Flora Tsang HF; Ranjan Das S; Vance ML; Kussman BD
INSTITUCIÓN / INSTITUTION: - Queen Mary Hospital, Hong Kong SAR, People's Republic of China. Electronic address: tangyeekwan@gmail.com.

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[24]
TÍTULO / TITLE: - p63 Expression in Merkel Cell Carcinoma Predicts Poorer Survival yet May Have Limited Clinical Utility.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Am J Clin Pathol. 2013 Dec;140(6):838-44. doi: 10.1309/AJCPE4PK6CTBNQJY.
●● Enlace al texto completo (gratuito o de pago) 1309/AJCPE4PK6CTBNQJY
AUTORES / AUTHORS: - Stetsenko GY; Malekirad J; Paulson KG; Iyer JG; Thibodeau RM; Nagase K; Schmidt M; Storer BE; Argenyi ZB; Nghiem P
INSTITUCIÓN / INSTITUTION: - University of Washington, 850 Republican St, Seattle, WA 98109; e-mail: pnghiem@u.washington.edu.
RESUMEN / SUMMARY: - Objectives To determine the clinical utility of p63 expression, which has been identified in several cohorts as a predictor of poorer prognosis in Merkel cell carcinoma (MCC). Methods Immunohistochemistry was used to determine p63 expression on MCC tumors from 128 patients. Results Of the patients, 33% had detectable p63 expression. p63 Positivity was associated with an increased risk of death from MCC (hazard ratio, 2.05; P = .02) in a multivariate Cox regression model considering stage at presentation, age at diagnosis, and sex. Although p63 expression correlated with diminished survival in this largest cohort reported thus far, the effect was weaker than that observed in prior studies. Indeed, within a given stage, p63 status did not predict survival in a clinically or statistically significant manner. Conclusions It remains unclear whether this test should be integrated into routine MCC patient management.

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[25]
TÍTULO / TITLE: - Anatomic and Metabolic Evaluation of Peripheral Nerve Sheath Tumors in Patients With Neurofibromatosis 1 Using Whole-Body MRI and 18F-FDG PET Fusion.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Clin Nucl Med. 2013 Oct 22.
●● Enlace al texto completo (gratuito o de pago)
1097/RLU.0b013e3182a757d3
AUTORES / AUTHORS: - Urban T; Lim R; Merker VL; Muzikansky A; Harris GJ; Kassarjian A; Bredella MA; Plotkin SR
INSTITUCIÓN / INSTITUTION: - From the *Department of Radiology, daggerDepartment of Neurology and Cancer Center, double daggerBiostatistics Center, Massachusetts General Hospital, Boston, MA; and section signCorades, S.L., Madrid, España.
RESUMEN / SUMMARY: - PURPOSE: Malignant peripheral nerve sheath tumors (MPNSTs) are the leading cause of death for patients with neurofibromatosis type 1 (NF1). Identification of hypermetabolic lesions on PET may help identify patients at risk for MPNST. The objective of this study was to identify clinical and MRI-derived

variables that predicted increased metabolic activity of neurofibromas in NF1 patients as determined by PET. METHODS: This prospective study included NF1 patients with neurofibromas of 5 cm in diameter or greater. All patients underwent whole-body MRI and F-FDG PET imaging. Tumor volume was calculated from the MR scans using a semiautomated 3-dimensional segmentation method. SUVmax's were calculated to quantify metabolic activity. Logistic regression analyses were performed to determine the relationship among SUVmax, tumor volume, location (extremity vs trunk), type (plexiform vs circumscribed), depth (superficial vs deep), patient age, and whole-body tumor burden. RESULTS: A total of 311 neurofibromas were identified in 19 NF1 patients (mean age, 38 years; range, 19-58 years). One extreme outlier was excluded from analysis. Whole-body tumor volumes ranged from 0.4 to 1182.4 mL. Fifty of 310 tumors were FDG-avid on PET (16%) with median SUVmax of 2.2 (range, 0.4-9.6). Metabolic activity (SUVmax >2.5) correlated with tumor location (deep > superficial, trunk > extremity) in tumors with PET avidity. CONCLUSIONS: In NF1 patients with neurofibromas of 5 cm or greater, the majority of internal tumors are not metabolically active on PET. Tumors with increased metabolic activity as defined by SUVmax greater than 2.5 (ie, suggestive of MPNST) are more likely to be deep and located within the trunk.

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[26]

TÍTULO / TITLE: - "Cherry Picking", a Multiple Non-anatomic Liver Resection Technique, as a Promising Option for Diffuse Liver Metastases in Patients with Neuroendocrine Tumours.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - World J Surg. 2013 Oct 8.

●● Enlace al texto completo (gratuito o de pago) 1007/s00268-013-2267-3

AUTORES / AUTHORS: - Krausch M; Raffel A; Anlauf M; Schott M; Lehwald N; Krieg A; Topp SA; Cupisti K; Knoefel WT

INSTITUCIÓN / INSTITUTION: - Department of General, Visceral and Pediatric Surgery, Heinrich-Heine-University Duesseldorf, Moorenstr. 5, 40225, Duesseldorf, Germany, krausch@med.uni-duesseldorf.de.

RESUMEN / SUMMARY: - INTRODUCTION: Liver metastases of GEP-NETs are a known major prognostic factor with a strong effect on patients' survival. To date, various treatment options are available, whereas surgery remains the only curative option. Because large liver resections often cannot be performed due to insufficient remnant liver volume, a special operative technique, "cherry picking" (multiple nonanatomic liver resections), can be used as a tissue-preserving procedure. METHODS: Of 91 patients with various GEP-NETs, 16 patients were identified with synchronous or metachronous multifocal, bilobular liver metastases (>10). All were treated with "cherry picking." Patient records were reviewed retrospectively and clinical data and pathology results were analyzed. RESULTS: Mean survival after primary tumour resection was 82.8 versus 41.2 months after liver surgery. All 16 patients are still alive. Mean recurrence-free survival after primary tumour operation was 49.8 versus 24.6 months after liver surgery. Complications of cherry picking included two postoperative biliary leakages and three small hepatic abscesses (conservative/interventional approach 25 % (n = 4), surgical approach 6.25 % (n = 1). There was no postoperative mortality. Initial hormonal symptoms (5/16 patients)

completely disappeared postoperatively in 2 patients and were significantly decreased in 3 patients. CONCLUSIONS: The tissue-preserving surgical technique “cherry picking” has developed due to improved imaging techniques and increased knowledge in liver anatomy, which has helped to make this approach safer and easier. Highly selected patients with multiple bilobular liver metastases of GEP-NET can benefit from this special surgical approach, also applicable for recurrent metastases.

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[27]

TÍTULO / TITLE: - Germline mutations and genotype-phenotype correlations in patients with apparently sporadic pheochromocytoma/paraganglioma in Korea.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Clin Genet. 2013 Oct 17. doi: 10.1111/cge.12304.

●● Enlace al texto completo (gratuito o de pago) 1111/cge.12304

AUTORES / AUTHORS: - Kim J; Seong MW; Lee K; Choi H; Ku E; Bae J; Park S; Choi S; Kim S; Shin C; Kim S

INSTITUCIÓN / INSTITUTION: - Department of Internal medicine.

RESUMEN / SUMMARY: - The aim of our study was to assess the frequency of germline mutations and develop the genetic testing strategy in patients with apparently sporadic pheochromocytoma/paraganglioma (PPGL) in Korea. We included 53 patients diagnosed with non-syndromic PPGL without a family history of PPGLs in three referral centers from 2004 to 2011. Succinate dehydrogenase complex B (SDHB), SDHD, Von Hippel-Lindau (VHL), and rearranged during transfection (RET) genes were examined by direct sequencing and multiple ligation-dependent probe amplification. The study patients were composed of 26 men and 27 women, and mean age was 50.1 +/- 13.5 years. The frequency of germline mutations was 13.2% (7/53): RET (n = 2), VHL (n = 1), SDHB (n = 2), and SDHD (n = 2). Six of seven mutation carriers were diagnosed before the age of 50. One of two patients harboring an SDHB mutation had malignant PPGLs. One patient with multifocal head and neck paraganglioma (PGL) and pheochromocytoma (PHEO) carried a SDHD mutation. The carriers of germline mutations in patients with apparently sporadic PPGL were 13.2% in our study. We recommend genetic testing in patients below 50 years and SDHD genetic testing in patients with multifocal PPGLs. In malignant PPGLs, SDHB genetic testing may be performed.

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[28]

TÍTULO / TITLE: - In vivo 5FU-exposed human medullary thyroid carcinoma cells contain a chemoresistant CD133+ tumor-initiating cell subset.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Thyroid. 2013 Sep 27.

●● Enlace al texto completo (gratuito o de pago) 1089/thy.2013.0277

AUTORES / AUTHORS: - Kucerova L; Feketeova L; Kozovska Z; Poturnajova M; Matuskova M; Nencka R; Babal P

INSTITUCIÓN / INSTITUTION: - Cancer Research Institute, Laboratory of Molecular Oncology, Bratislava, Slovakia ; lucia.kucerova@savba.sk.

RESUMEN / SUMMARY: - The hierarchical model of solid tumor proposes the existence of rare tumor cell subpopulations with stem cell properties. Glycoprotein Prominin-1

(CD133) represents one of the cancer stem cell markers in several tumor types. The CD133+ cell subpopulation was shown to be enriched for tumor-initiating and highly chemoresistant cells in human cancer(s). We investigated whether CD133+ cells derived from human medullary thyroid carcinoma (MTC) possess tumor-initiating properties in vivo and exhibit differential responses to chemotherapeutic agents. We demonstrate that separated CD133+ cells from the human MTC cell line TT are enriched for tumor-initiating cells as demonstrated by tumor formation in vivo. Nevertheless, TT CD133+ cells do not exhibit increased chemoresistance in comparison to parental cells. However, when MTC xenotransplants were treated with the chemotherapeutic drug 5-fluorouracil (5FU) in vivo, CD133 expression increased in MTC cells. This cell line, designated FTTiv isolated from the drug-exposed xenotransplants, exhibits a significantly different response to 5FU associated with the substantial change in the expression profile of genes involved in 5FU metabolism and drug resistance. Moreover, the CD133+ tumor-initiating subpopulation derived from these drug-exposed FTTiv cells is significantly more resistant to 5FU and retains the chemoresistant properties upon FTTiv culture propagation. These data suggest that the chemoresistant phenotype and the CD133+ MTC subpopulation emerged in response to chemotherapy in vivo.

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[29]

TÍTULO / TITLE: - The unequal distribution of parathyroid neoplasms in male patients.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Am Surg. 2013 Oct;79(10):1022-5.

AUTORES / AUTHORS: - Goldner B; Lee B; Stabile BE

INSTITUCIÓN / INSTITUTION: - Harbor-UCLA Medical Center, Torrance, California, USA.

RESUMEN / SUMMARY: - There is a known lesser incidence of primary hyperparathyroidism and parathyroid neoplasms in male patients. Any difference in the anatomic distribution between males and females has not been documented. Review of our institutional experience with 125 pathologically confirmed parathyroid adenomas (119) or carcinomas (six) from 2000 through 2012 was conducted. The anatomic location was identified from operative records and the distributions between males and females were compared. Ninety-two females with parathyroid neoplasms had equal anatomic distributions between left and right sides and no significant difference between superior and inferior locations (P = 0.381). In marked contrast, tumors in 33 male patients had a significant predilection for the right side (67%, P = 0.016) and inferior position (85%, P = 0.033) and most notably the right inferior position (64%, P = 0.026). For the group as a whole, inferior adenomas were significantly more common (70%, P = 0.044). All patients had postoperative normalization of serum calcium levels. Late biochemical recurrence was noted in two patients. This is the first operatively confirmed delineation of the anatomic distributions of parathyroid neoplasms in separate sexes. Based on the unexpected findings of this study, we recommend the right inferior cervical region be explored first in males with suspected parathyroid tumors of indeterminate location.

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[30]

TÍTULO / TITLE: - Updating the management of patients with rectal neuroendocrine tumors.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Endoscopy. 2013 Dec;45(12):1039-46. doi: 10.1055/s-0033-1344794. Epub 2013 Oct 25.

●● Enlace al texto completo (gratuito o de pago) 1055/s-0033-1344794

AUTORES / AUTHORS: - de Mestier L; Brixi H; Gincul R; Ponchon T; Cadiot G
INSTITUCIÓN / INSTITUTION: - Department of Hepatogastroenterology and Digestive Oncology, University Hospital Robert-Debre, Reims, France.
RESUMEN / SUMMARY: - Rectal neuroendocrine tumors (NETs) account for approximately one-third of all digestive NETs, with an increasing incidence and good overall prognosis. Although recent guidelines have been published, endoscopic techniques have expanded substantially and the most recent reports should be taken into account for clinical practice. The objectives of this report were to review the latest advances on prognosis, pre-interventional explorations, treatment - with particular focus on endoscopy - and surveillance of well-differentiated rectal NETs, excluding poorly differentiated and metastatic tumors.

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[31]
TÍTULO / TITLE: - Preclinical Evaluation of 18F-LMI1195 for In Vivo Imaging of Pheochromocytoma in the MENX Tumor Model.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - J Nucl Med. 2013 Dec;54(12):2111-7. doi: 10.2967/jnumed.113.119966. Epub 2013 Oct 17.

●● Enlace al texto completo (gratuito o de pago) 2967/jnumed.113.119966

AUTORES / AUTHORS: - Gaertner FC; Wiedemann T; Yousefi BH; Lee M; Repokis I; Higuchi T; Nekolla SG; Yu M; Robinson S; Schwaiger M; Pellegata NS
INSTITUCIÓN / INSTITUTION: - Klinikum Rechts der Isar der Technischen Universitat Munchen, Nuklearmedizinische Klinik, Munchen, Germany.
RESUMEN / SUMMARY: - We evaluated (18)F-LMI1195 (1-(3-bromo-4-(3-(18)F-fluoro-propoxy)benzyl)guanidine), a metaiodobenzylguanidine (MIBG) analog, for the detection of pheochromocytoma in a preclinical in vivo model of endogenous neuroendocrine tumors (multiple endocrine neoplasia [MENX]). METHODS: Adrenal uptake kinetics of (18)F-LMI1195 were evaluated in healthy Wistar rats (n = 6) by dynamic PET imaging. Distribution of (18)F-LMI1195 was evaluated in tumor-bearing MENX mut/mut rats (n = 10) and control MENX wild-type rats (n = 4) by biodistribution studies and PET imaging. Biodistribution of (18)F-LMI1195 was compared with (123)I-MIBG in MENX mut/mut rats (n = 6) and correlated with histological tumor volume and norepinephrine transporter (NET) expression. Uptake specificity was evaluated by in vivo inhibition of the NET by desipramine (n = 6). Intraadrenal distribution of (18)F-LMI1195 was evaluated by autoradiography. RESULTS: (18)F-LMI1195 showed rapid tracer accumulation in adrenal glands 1 min after tracer injection. Adrenal glands of MENX mut/mut animals showed significantly higher standardized uptake value than MENX wild-type controls (maximum SUV, 10.3 +/- 2.3 vs. 6.1 +/- 0.9, P < 0.01). Adrenal uptake in MENX mut/mut rats could be inhibited by desipramine, shown by biodistribution studies (0.06 +/- 0.01 vs. 0.16 +/- 0.05 percentage injected dose, P < 0.01), PET imaging (maximum SUV, 3.8 +/- 0.8 vs. 10.3 +/- 2.3, P < 0.01), and

autoradiography. Adrenal uptake of (18)F-LMI1195 correlated with (123)I-MIBG uptake (r = 0.91), histological tumor volume (r = 0.68), and NET expression (r = 0.50). (18)F-LMI1195 showed an overall favorable distribution for tumor imaging. CONCLUSION: (18)F-LMI1195 shows high and specific accumulation in pheochromocytomas. Its favorable biodistribution makes it a promising PET tracer for tumor imaging. Further studies are warranted to evaluate its clinical value in oncologic indications.

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[32]

TÍTULO / TITLE: - Malignant peripheral nerve sheath tumor with glandular differentiation in a patient with neurofibromatosis type 1.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Am J Dermatopathol. 2013 Dec;35(8):859-63. doi: 10.1097/DAD.0b013e318284a611.

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

1097/DAD.0b013e318284a611

AUTORES / AUTHORS: - Galatian AA; Crowson AN; Fischer RJ; Yob EH; Shendrik I

INSTITUCIÓN / INSTITUTION: - *Department of Dermatology, University of Oklahoma, College of Medicine, Oklahoma City, OK; daggerOklahoma Department of Dermatology, University of Oklahoma, College of Medicine, Oklahoma City, OK; double daggerDermatopathology Section, Regional Medical Laboratory, Tulsa, OK; section signDepartment of Internal Medicine, University of Oklahoma, College of Medicine, Oklahoma City, OK; and paragraph signTulsa Cancer Institute - Skin Cancer Center, Tulsa, OK.

RESUMEN / SUMMARY: - : The authors report an unusual case of malignant peripheral nerve sheath tumor with malignant differentiation arising as a subcutaneous nodule in the thigh of a 53-year-old woman with a history significant for neurofibromatosis type 1. Peripheral nerve sheath tumors containing a glandular component, commonly referred to as glandular peripheral nerve sheath tumors, are rare neoplasms found largely in patients with neurofibromatosis type 1. These tumors are frequently malignant; recognition of metastatic potential is made based on the atypical spindle-cell component. Rarely, as in our case, the glandular component is also histologically malignant. Only 5 such tumors have been described in the literature to date. Glandular differentiation, particularly with malignant features, can be a potentially misleading feature when found as a component of malignant peripheral nerve sheath tumors and raise a wide spectrum of differential diagnoses, including metastatic Sertoli-Leydig tumors. The patient is free of disease for 22 months after wide tumor reexcision, which contrasts with previously reported devastatingly poor prognosis of these tumors.

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[33]

TÍTULO / TITLE: - The predictive value of mean platelet volume, plateletcrit and red cell distribution width in the differentiation of autoimmune gastritis patients with and without type I gastric carcinoid tumors.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Platelets. 2013 Oct 31.

●● Enlace al texto completo (gratuito o de pago) 3109/09537104.2013.821607

AUTORES / AUTHORS: - Tuzun A; Keskin O; Yakut M; Kalkan C; Soykan I

INSTITUCIÓN / INSTITUTION: - Division of Gastroenterology, Ibni Sina Hospital, Ankara University Faculty of Medicine , Ankara , Turkey.
RESUMEN / SUMMARY: - Abstract Autoimmune gastritis is an autoimmune and inflammatory condition that may predispose to gastric carcinoid tumors or adenocarcinomas. The early diagnosis of these tumors is important in order to decrease morbidity and mortality. Platelet indices such as mean platelet volume and plateletcrit levels increase in inflammatory, infectious and malign conditions. The primary aim of this study was to explore wheter platelet indices and red cell distribution width have any predictive role in the discrimination of autoimmune gastritis patients with and without gastric carcinoid tumors. Also secondary aim of this study was to investigate whether any changes exist betwenn autoimmune gastritis and functional dyspepsia patients by means of platelet indices. Plateletcrit (0.22 +/- 0.06 vs. 0.20 +/- 0.03%, p < 0.001) and red cell distribution width (16.11 +/- 3.04 vs. 13.41 +/- 0.95%, p < 0.001) were significantly higher in autoimmune gastritis patients compared to control group. Receiver operating curve analysis suggested that optimum plateletcrit cut-off point was 0.20% (AUC: 0.646), and 13.95% as the cut off value for red cell distribution width (AUC: 0.860). Although plateletcrit (0.22 +/- 0.06 vs. 0.21 +/- 0.04%, p = 0.220) and mean platelet volume (8.94 +/- 1.44 vs. 8.68 +/- 0.89 fl, p = 0.265) were higher in autoimmune gastritis patients without carcinoid tumor compared to patients with carcinoid tumors, these parameters were not statistically significant. Changes in plateletcrit and red cell distribution width values may be used as a marker in the discrimination of autoimmune gastritis and fucntional dyspepsia patients but not useful in patients with gastric carcinoid tumor type I.

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[34]
TÍTULO / TITLE: - An expression signature of the angiogenic response in gastrointestinal neuroendocrine tumours: correlation with tumour phenotype and survival outcomes.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Br J Cancer. 2013 Nov 14. doi: 10.1038/bjc.2013.682.
●● Enlace al texto completo (gratuito o de pago) 1038/bjc.2013.682
AUTORES / AUTHORS: - Pinato DJ; Tan TM; Toussi ST; Ramachandran R; Martin N; Meeran K; Ngo N; Dina R; Sharma R
INSTITUCIÓN / INSTITUTION: - Division of Experimental Medicine, Imperial College London, Hammersmith Hospital, Du Cane Road, W120HS London, UK.
RESUMEN / SUMMARY: - Background:Gastroenteropancreatic neuroendocrine tumours (GEP-NETs) are heterogeneous with respect to biological behaviour and prognosis. As angiogenesis is a renowned pathogenic hallmark as well as a therapeutic target, we aimed to investigate the prognostic and clinico-pathological role of tissue markers of hypoxia and angiogenesis in GEP-NETs.Methods:Tissue microarray (TMA) blocks were constructed with 86 tumours diagnosed from 1988 to 2010. Tissue microarray sections were immunostained for hypoxia inducible factor 1alpha (Hif-1alpha), vascular endothelial growth factor-A (VEGF-A), carbonic anhydrase IX (Ca-IX) and somatostatin receptors (SSTR) 1-5, Ki-67 and CD31. Biomarker expression was correlated with clinico-pathological variables and tested for survival prediction using Kaplan-Meier and Cox regression methods.Results:Eighty-six consecutive cases were included: 51% male, median age 51 (range 16-82), 68% presenting with a pancreatic

primary, 95% well differentiated, 51% metastatic. Higher grading (P=0.03), advanced stage (P<0.001), high Hif-1alpha and low SSTR-2 expression (P=0.03) predicted for shorter overall survival (OS) on univariate analyses. Stage, SSTR-2 and Hif-1alpha expression were confirmed as multivariate predictors of OS. Median OS for patients with SSTR-2+/Hif-1alpha-tumours was not reached after median follow up of 8.8 years, whereas SSTR-2-/Hif-1alpha+ GEP-NETs had a median survival of only 4.2 years (P=0.006).Conclusion:We have identified a coherent expression signature by immunohistochemistry that can be used for patient stratification and to optimise treatment decisions in GEP-NETs independently from stage and grading. Tumours with preserved SSTR-2 and low Hif-1alpha expression have an indolent phenotype and may be offered less aggressive management and less stringent follow up.British Journal of Cancer advance online publication, 14 November 2013; doi:10.1038/bjc.2013.682 www.bjcancer.com.

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[35]

TÍTULO / TITLE: - Novel insights into pancreatic beta-cell glucolipotoxicity from real-time functional analysis of mitochondrial energy metabolism in INS-1E insulinoma cells.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Biochem J. 2013 Dec 15;456(3):417-26. doi: 10.1042/BJ20131002.

●● Enlace al texto completo (gratuito o de pago) 1042/BJ20131002

AUTORES / AUTHORS: - Barlow J; Affourtit C

INSTITUCIÓN / INSTITUTION: - *School of Biomedical and Healthcare Sciences, Plymouth University, Drake Circus, Plymouth PL4 8AA, U.K.

RESUMEN / SUMMARY: - High circulating glucose and non-esterified (free) fatty acid levels can cause pancreatic beta-cell failure. The molecular mechanisms of this beta-cell glucolipotoxicity are yet to be established conclusively. In the present paper we report on the involvement of mitochondrial dysfunction in fatty-acid-induced beta-cell failure. We have used state-of-the-art extracellular flux technology to functionally probe mitochondrial energy metabolism in intact INS-1E insulinoma cells in real-time. We show that 24-h palmitate exposure at high glucose attenuates the glucose-sensitivity of mitochondrial respiration and lowers coupling efficiency of glucose-stimulated oxidative phosphorylation. These mitochondrial defects coincide with an increased level of ROS (reactive oxygen species), impaired GSIS (glucose-stimulated insulin secretion) and decreased cell viability. Palmitate lowers absolute glucose-stimulated respiration coupled to ATP synthesis, but does not affect mitochondrial proton leak. Palmitate is not toxic when administered at low glucose unless fatty acid beta-oxidation is inhibited. Palmitoleate, on the other hand, does not affect mitochondrial respiration, ROS levels, GSIS or cell viability. Although palmitoleate protects against the palmitate-induced ROS increase and cell viability loss, it does not protect against respiratory and insulin secretory defects. We conclude that mitochondrial dysfunction contributes to fatty-acid-induced GSIS impairment, and that glucolipotoxic cell viability and GSIS phenotypes are mechanistically distinct.

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[36]

TÍTULO / TITLE: - Gastric inhibitory polypeptide receptor (GIPR) is a promising target for imaging and therapy in neuroendocrine tumors.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Surgery. 2013 Dec;154(6):1206-14. doi: 10.1016/j.surg.2013.04.052.
●● Enlace al texto completo (gratuito o de pago) 1016/j.surg.2013.04.052
AUTORES / AUTHORS: - Sherman SK; Carr JC; Wang D; O'Dorisio MS; O'Dorisio TM; Howe JR
INSTITUCIÓN / INSTITUTION: - Department of Surgery, University of Iowa Carver College of Medicine, Iowa City, IA.
RESUMEN / SUMMARY: - BACKGROUND: Ligands binding the somatostatin receptor type 2 (SSTR2) are useful for imaging and treatment of neuroendocrine tumors (NETs), but not all tumors express high levels of these receptors. The aim of this study was to evaluate gene expression of new therapeutic targets in NETs relative to SSTR2. METHODS: RNA was extracted from 103 primary small bowel and pancreatic NETs, matched normal tissue, and 123 metastases. Expression of 12 candidate genes was measured by quantitative polymerase chain reaction normalized to internal controls; candidate gene expression was compared with SSTR2. RESULTS: Relative to normal tissue, primary NET expression of SSTR2, GPR98, BRS3, GIPR, GRM1, and OPRK1 were increased by 3, 8, 13, 13, 17, and 20-fold, respectively. Similar changes were found in metastases. Although most candidate genes showed lesser absolute expressions than SSTR2, absolute GIPR expression was closest to SSTR2 (mean dCT 3.6 vs 2.7, P = .01). Absolute OPRK1 and OXTR expression varied greatly by primary tumor type and was close to SSTR2 in small bowel NETs but not pancreatic NETs. CONCLUSION: Compared with the current treatment standard SSTR2, GIPR has only somewhat lesser absolute gene expression in tumor tissue but much lesser expression in normal tissue, making it a promising new target for NET imaging and therapy.

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[37]
TÍTULO / TITLE: - Molecular genetics of paragangliomas of the skull base and head and neck region: implications for medical and surgical management.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - J Neurosurg. 2013 Nov 15.
●● Enlace al texto completo (gratuito o de pago) 3171/2013.10.JNS13659
AUTORES / AUTHORS: - Hussain I; Husain Q; Baredes S; Eloy JA; Jyung RW; Liu JK
INSTITUCIÓN / INSTITUTION: - Departments of Neurological Surgery and.
RESUMEN / SUMMARY: - Paragangliomas are rare, slow-growing tumors that frequently arise in the head and neck, with the carotid bodies and temporal bone of the skull base being the most common sites. These neoplasms are histologically similar to pheochromocytomas that form in the adrenal medulla and are divided into sympathetic and parasympathetic subtypes based on functionality. Skull base and head and neck region paragangliomas (SHN-PGs) are almost always derived from parasympathetic tissue and rarely secrete catecholamines. However, they can cause significant morbidity by mass effect on various cranial nerves and major blood vessels. While surgery for SHN-PG can be curative, postoperative deficits and recurrences make these lesions challenging to manage. Multiple familial syndromes predisposing individuals to development of paragangliomas have been identified, all involving

mutations in the succinate dehydrogenase complex of mitochondria. Mutations in this enzyme lead to a state of “pseudohypoxia” that upregulates various angiogenic, survival, and proliferation factors. Moreover, familial paraganglioma syndromes are among the rare inherited diseases in which genomic imprinting occurs. Recent advances in gene arrays and transcriptome/exome sequencing have identified an alternate mutation in sporadic SHN-PG, which regulates proto-oncogenic pathways independent of pseudohypoxia-induced factors. Collectively these findings demonstrate that paragangliomas of the skull base and head and neck region have a distinct genetic signature from sympathetic-based paragangliomas occurring below the neck, such as pheochromocytomas. Paragangliomas serve as a unique model of primarily surgically treated neoplasms whose future will be altered by the elucidation of their genomic complexities. In this review, the authors present an analysis of the molecular genetics of SHN-PG and provide future directions in patient care and the development of novel therapies.

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[38]

TÍTULO / TITLE: - Circulating plasma and platelet 5-hydroxytryptamine in carcinoid heart disease: a pilot study.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - J Heart Valve Dis. 2013 May;22(3):400-7.

AUTORES / AUTHORS: - Bhattacharyya S; Jagroop A; Gujral DM; Hayward C; Toumpanakis C; Caplin M; Mikhailidis DP; Davar J

INSTITUCIÓN / INSTITUTION: - Department of Cardiology, Royal Free Hospital, London, UK.

RESUMEN / SUMMARY: - BACKGROUND AND AIM OF THE STUDY: Carcinoid heart disease (CaHD) is a rare form of valvular heart disease in patients with carcinoid syndrome (CS). The role of 5-hydroxytryptamine (5-HT) in the pathogenesis of CaHD is unclear. The study aim was to evaluate the association between platelet 5-HT (5-HTplt) and plasma 5-HT (5-HTpls) and valvular dysfunction. METHODS: Twelve patients with CaHD, 18 with CS and 10 ‘normal’ subjects were recruited. Patients with CaHD underwent cardiac catheterization and echocardiography. 5-HTplt and 5-HTpls was sampled in blood from the femoral vein, right and left ventricle, and left antecubital fossa vein. RESULTS: Levels of 5-HTpls and 5-HTplt were significantly higher in patients with CaHD (median 5-HTpls 325 nmol/l and 5-HTplt 18.9 nmol/10(9) platelets) and CS (median 5-HTpls 155 nmol/l and 5-HTplt 16.4 nmol/10(9) platelets) when compared to healthy controls (median 5-HTpls 9 nmol/l and 5-HTplt 3.7 nmol/10(9) platelets; $p < 0.0001$ and $p = 0.003$, respectively). There was a significant increase in 5-HTplt and 5-HTpls between the femoral vein and right heart ($p = 0.007$ and $p = 0.0002$, respectively). There was no significant difference in 5-HTplt or 5-HTpls between the right and left side of the heart, irrespective of the presence of a patent foramen ovale or of left-sided CaHD. CONCLUSION: Plasma and platelet 5-HT levels are elevated in patients with CaHD. Despite exposure to similar levels of intracardiac plasma and platelet 5-HT, the development of valve dysfunction is heterogeneous. This suggests that individual heart valves have a susceptibility to the development of valvular dysfunction which is not related solely to plasma or platelet 5-HT levels.

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[39]
TÍTULO / TITLE: - Lymphoma as a Second Malignancy in a Patient With Neuroendocrine Tumor: Mimicking Dedifferentiation on Dual-Tracer PET/CT With 68Ga-DOTANOC and 18F-FDG.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Clin Nucl Med. 2013 Nov 7.
●● Enlace al texto completo (gratuito o de pago)
1097/RLU.0b013e31828e98c5
AUTORES / AUTHORS: - Jain S; Sharma P; Dhull VS; Bal C; Kumar R
INSTITUCIÓN / INSTITUTION: - From the Department of Nuclear Medicine, All India Institute of Medical Sciences, New Delhi, India.
RESUMEN / SUMMARY: - Neuroendocrine tumors (NETs) are rare tumors which express somatostatin receptors (SSTRs). We here present a case of a 50-year-old female patient with metastatic bronchial carcinoid. She underwent Ga-DOTANOC PET/CT and F-FDG PET/CT which suggested a diagnosis of poorly differentiated NET. Biopsy of the lesion, however, revealed a second malignancy in the form of diffuse large B-cell lymphoma. Thus, very rarely, other primary tumors can mimic NETs on dual-tracer PET/CT, and biopsy is advised in doubtful cases.

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[40]
TÍTULO / TITLE: - Predictive Value of 68Ga-DOTANOC PET/CT in Patients With Suspicion of Neuroendocrine Tumors: Is Its Routine Use Justified?
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Clin Nucl Med. 2013 Oct 22.
●● Enlace al texto completo (gratuito o de pago)
1097/RLU.0000000000000257
AUTORES / AUTHORS: - Sharma P; Arora S; Mukherjee A; Pal S; Sahni P; Garg P; Khadgawat R; Thulkar S; Bal C; Kumar R
INSTITUCIÓN / INSTITUTION: - From the Departments of *Nuclear Medicine, daggerSurgical Gastroenterology, double daggerGastroenterology and Human Nutrition, section signEndocrinology and Metabolism, and paragraph signRadiodiagnosis, All India Institute of Medical Sciences, New Delhi, India.
RESUMEN / SUMMARY: - OBJECTIVE: The objective of this study was to evaluate the predictive value of Ga-DOTANOC PET/CT in patients with suspected neuroendocrine tumor (NET). METHODS: Data of 164 patients (mean age, 42.5 +/- 17.3 years; 54.8% male) who underwent Ga-DOTANOC PET/CT for suspected NET were retrospectively analyzed. Neuroendocrine tumor was suspected based on clinical features (n = 94) and/or raised biochemical markers (n = 83, serum chromogranin A, gastrin, serum/urinary catecholamines, insulin/C-peptide, and 5-hydroxytrytophan/5-hydroxyindoleacetic acid) and/or imaging findings (n = 93). PET/CT images were reviewed by 2 experienced nuclear medicine physicians, and any nonphysiological Ga-DOTANOC uptake was taken as positive for NET. Histopathology (n = 55) and clinical/imaging follow-up (n = 109; median, 11 months) was used as reference standard. RESULTS: Based on the reference standard, 97 of 164 patients had NET. Ga-DOTANOC PET/CT was positive for NET in 101 and negative in 63 patients. Primary tumor was demonstrated in 90 patients (commonest site-pancreas) and metastasis in 30 (commonest site-liver). PET/CT was true positive in 92 patients, true

negative in 58, false positive in 9, and false negative in 5. The overall sensitivity was 94.8%, specificity was 86.5%, positive predictive value was 91%, negative predictive value was 92%, and accuracy was 91.4%. The accuracy of PET-CT in patients with clinical features of NET was 90.4%, with raised biochemical markers was 86.7%, and with imaging findings suggestive of NET was 93.5%. No difference was seen in the accuracy in patients with or without clinical symptoms (P = 0.794), raised versus those with normal/unknown biochemical markers (P = 0.094), and suggestive imaging versus those with negative/unavailable imaging (P = 0.420). CONCLUSIONS: Ga-DOTANOC PET-CT shows high positive and negative predictive values in patients with suspected NET and can be routinely used for this purpose.

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[41]

TÍTULO / TITLE: - Neuroendocrine tumors: insights into innovative therapeutic options and rational development of targeted therapies.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Drug Discov Today. 2013 Oct 27. pii: S1359-6446(13)00382-6. doi: 10.1016/j.drudis.2013.10.015.

●● Enlace al texto completo (gratuito o de pago) 1016/j.drudis.2013.10.015

AUTORES / AUTHORS: - Barbieri F; Albertelli M; Grillo F; Mohamed A; Saveanu A; Barlier A; Ferone D; Florio T

INSTITUCIÓN / INSTITUTION: - Department of Internal Medicine and Center of Excellence for Biomedical Research (CEBR), University of Genova, viale Benedetto XV, 2-16132 Genova, Italy.

RESUMEN / SUMMARY: - Neuroendocrine tumors (NETs) are heterogeneous neoplasms with respect to molecular characteristics and clinical outcome. Although slow-growing, NETs are often late diagnosed, already showing invasion of adjacent tissues and metastases. Precise knowledge of NET biological and molecular features has opened the door to the identification of novel pharmacological targets. Therapeutic options include somatostatin analogs, alone or in combination with interferon-alpha, multi-targeted tyrosine kinase inhibitors (e.g. sunitinib) or mammalian target of rapamycin (mTOR) inhibitors (e.g. everolimus). Antiangiogenic approaches and anti insulin-like growth factor receptor (IGFR) compounds have been also proposed as combination therapies with the aforementioned compounds. This review will focus on recent studies that have improved therapeutic strategies in NETs, discussing management challenges such as drug resistance development as well as focusing on the need for predictive biomarkers to design distinct drug combinations and optimize pharmacological control.

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[42]

TÍTULO / TITLE: - The Presence of SDHB Mutations Should Modify Surgical Indications for Carotid Body Paragangliomas.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Ann Surg. 2013 Oct 28.

●● Enlace al texto completo (gratuito o de pago) 1097/SLA.0000000000000283

AUTORES / AUTHORS: - Ellis RJ; Patel D; Prodanov T; Nilubol N; Pacak K; Kebebew E

INSTITUCIÓN / INSTITUTION: - *Endocrine Oncology Branch, National Cancer Institute, National Institutes of Health, Bethesda, MD; daggerPerelman School of Medicine at the University of Pennsylvania, Philadelphia, PA; and double daggerProgram in Reproductive and Adult Endocrinology, Eunice Kennedy Shriver National Institute of Child Health and Human Development, National Institutes of Health, Bethesda, MD.

RESUMEN / SUMMARY: - OBJECTIVE:: The aim of this study was to determine whether the genetic background of the disease should be incorporated into treatment decision making. BACKGROUND:: Carotid body paragangliomas are rare tumors that often affect patients with genetic mutations of the succinate dehydrogenase complex (SDHx). Despite growing evidence that germ line genetic mutations alter the aggressiveness of paragangliomas, treatment decisions are currently based only on clinical symptoms and tumor size in patients with carotid body paragangliomas. METHODS:: Retrospective analysis of 34 patients with carotid body paragangliomas who underwent genetic testing and surgical treatment. Recurrence was defined by the return of locoregional disease and/or development of distant metastases. Clinical characteristics and genetic testing results were analyzed as predictors of patient outcomes. RESULTS:: Thirty-four patients underwent 41 primary carotid body paraganglioma resections (median follow-up time of 42 months, range: 1-293). Overall survival was 91.2%. Twelve patients had germ line mutations in SDHB, 17 in SDHD, and 5 carried no known mutation. Surgical resection of larger tumors was associated with higher operative complications (odds ratio: 5.4, P = 0.05). Tumor size at resection was significantly smaller in patients with SDHB mutations than in patients with non-SDHB mutations (2.1 vs 3.3 cm, P = 0.02). Patients with a mutation in the SDHB gene also had significantly worse disease-free survival compared with patients without an SDHB gene mutation (P = 0.03). CONCLUSIONS:: Mutations in the SDHB gene are associated with worse disease-free survival after resection in patients with carotid body paragangliomas despite earlier intervention. This suggests that a more aggressive surgical approach is warranted in patients with SDHB mutations.

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[43]

TÍTULO / TITLE: - Clinical and Prognostic Features of Rectal Neuroendocrine Tumors.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Neuroendocrinology. 2013 Nov 5.

●● Enlace al texto completo (gratuito o de pago) 1159/000355612

AUTORES / AUTHORS: - Weinstock B; Ward SC; Harpaz N; Warner RR; Itzkowitz S; Kim MK

INSTITUCIÓN / INSTITUTION: - Division of Gastroenterology, Departments of Medicine, Mount Sinai School of Medicine, New York, N.Y., USA.

RESUMEN / SUMMARY: - Background: Rectal neuroendocrine tumors (NETs) are among the most common NETs. The aim was to validate European Neuroendocrine Tumor Society (ENETS)/North American Neuroendocrine Tumor Society (NANETS) staging and grading systems with regard to clinical outcomes. Methods: A comprehensive database was constructed from existing databases of the Mount Sinai Division of Gastrointestinal Pathology and the Carcinoid Cancer Foundation. Analysis was performed on 141 patients identified with rectal NETs seen at Mount Sinai Hospital between 1972 and 2011. Results: The median age was 52.7 years; 43% were males. Average tumor size was 0.88 cm. NETs <1 cm accounted for 75.6% of the tumors.

Stage I, II, III and IV accounted for 79.4, 2.8, 5.0 and 12.8% of the tumors, respectively. G1 tumors accounted for 88.1%, G2 8.3% and G3 3.6%. Of G1 tumors, 94.6% were stage I and 5.4% were stage IV. The median survival time for all 141 patients was 6.8 years (range, 0.8-34.7 years). The overall 5-year survival rate was 84.4%. The 5-year survival rates for patients in stages I-IV were 92.7, 75.0, 42.9 and 33.2%, respectively. The 5-year survival rates for patients with G1-G3 tumors were 87.7, 47.6 and 33.3%, respectively. Univariate analysis of increased survival showed significance for lower stage, lower grade, smaller size, absence of symptoms and endoscopically treated tumors. Multivariate analysis showed that stage alone was statistically significant as the strongest predictor of survival. Conclusion: The results of our study validated ENETS/NANETS guidelines for staging and grading of rectal NETs in the US setting of a tertiary referral center. Staging according to ENETS/NANETS guidelines should be used in the treatment algorithm rather than size alone.

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[44]

TÍTULO / TITLE: - Appendiceal Neuroendocrine Tumors (Carcinoid of the Appendix) in Childhood: A Clinical Report From the Italian Trep Project.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - J Pediatr Gastroenterol Nutr. 2013 Oct 17.

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

1097/MPG.0000000000000217

AUTORES / AUTHORS: - Virgone C; Cecchetto G; Alaggio R; Ferrari A; Bisogno G; Conte M; Inserra A; Fagnani AM; Indolfi P; Salfi N; Dall'igna P

INSTITUCIÓN / INSTITUTION: - *Pediatric Surgery, Department of Women's and Children's Health, University-Hospital of Padua, Padua, Italy daggerPathology Unit, Department of Medical and Diagnostic Sciences and Special Therapies, University of Padua, Padua, Italy double daggerHematology Oncology, IRCCS Istituto Nazionale dei Tumori, Milan, Italy section signHematology Oncology, Department of Women's and Children's Health, University-Hospital of Padua, Padua, Italy ||Hematology Oncology, Giannina Gaslini Children's Hospital, Genoa, Italy paragraph signPediatric Surgery, Bambino Gesu Children's Hospital, Rome, Italy #Pediatric Surgery, IRCCS Ca' Granda Ospedale Maggiore Policlinico, Milan, Italy **Hematology Oncology, Department of Pediatrics, II University, Naples, Italy daggerdaggerPathology Unit, Sant'Orsola-Malpighi Hospital, Bologna, Italy.

RESUMEN / SUMMARY: - BACKGROUND:: Neuroendocrine tumors (NETs) of the appendix are slow-growing tumors and, although rare, they are the most common G-I epithelial tumors in childhood and adolescence. The treatment and the follow-up screenings have not been standardized. Above all, though tumor size is considered the main prognostic variable to define the aggressiveness of approach, it remains to be established a precise cut-off. METHODS:: 113 patients under 18 years of age with a diagnosis of appendiceal NETs were registered as of 1 January 2000 until 30 May 2013 within the TREP Project (Rare Tumors in Pediatric Age), an Italian multi-institutional network dedicated to very rare tumors in children and adolescents. The recommendations of the TREP study included imaging and laboratory investigations. The treatment after appendectomy was decided on the basis of histology, tumor size and imaging: primary re-excision (PRE) was not recommended in completely excised tumors, regardless of tumor size and invasiveness. RESULTS:: 113/113 had a

diagnosis of well differentiated NETs: in 108/113 the tumor was smaller than 2 cm and in 5 larger than 2. Excision margins were free in 111/113 patients. In 3/113 a PRE was performed: in 1 residual tumor was detected. 113/113 patients are alive in complete remission (median follow-up of 41 months). CONCLUSIONS:: Reported data and our experience showed that no relapse or death occurred in children and adolescents affected by appendiceal NETs. Appendectomy alone should be considered curative for most patients and a more aggressive surgical approach is deserved in those cases with incompletely excised tumors.

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[45]

TÍTULO / TITLE: - Peptide Receptor Radionuclide Therapy With 177Lu DOTATATE in a Case of Recurrent Carotid Body Paraganglioma With Spinal Metastases.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Clin Nucl Med. 2013 Nov 7.

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

1097/RLU.0000000000000273

AUTORES / AUTHORS: - Gupta SK; Singla S; Karunanithi S; Damle N; Bal C

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

RESUMEN / SUMMARY: - Paragangliomas are rare benign neuroendocrine tumors, and 80% of all paragangliomas are either carotid body tumors or glomus jugulare tumors. We present a case of recurrent unresectable carotid body paraganglioma with nodal and T7 vertebral metastases in a 30-year-old man 6 years postsurgery detected with Ga DOTANOC PET/CT and was administered with peptide receptor radionuclide therapy using Lu DOTATATE. After 5 cycles of Lu DOTATATE (total cumulative activity of 750 mCi [27 GBq]), significant response at the primary site on Ga DOTANOC PET/CT and complete disappearance of nodal and T7 vertebral metastases were noted.

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[46]

TÍTULO / TITLE: - Synergistic cooperation between sunitinib and cisplatin promotes apoptotic cell death in human medullary thyroid cancer.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - J Clin Endocrinol Metab. 2013 Nov 25.

●● Enlace al texto completo (gratuito o de pago) 1210/jc.2013-2574

AUTORES / AUTHORS: - Lopergolo A; Nicolini V; Favini E; Dal Bo L; Tortoreto M; Cominetti D; Folini M; Perego P; Castiglioni V; Scanziani E; Grazia Borrello M; Zaffaroni N; Cassinelli G; Lanzi C

INSTITUCIÓN / INSTITUTION: - Molecular Pharmacology Unit, Department of Experimental Oncology and Molecular Medicine, Fondazione IRCCS Istituto Nazionale dei Tumori, Milan, Italy (AL, VN, EF, LD, MT, DC, MF, PP, NZ, GC, CL); Department of Veterinary Sciences and Public Health, Universita degli Studi di Milano, Milan, Italy and Mouse & Animal Pathology Lab, Fondazione Filarete, Milan, Italy (VC, ES); Molecular Mechanisms Unit, Department of Experimental Oncology and Molecular Medicine, Fondazione IRCCS Istituto Nazionale dei Tumori, Milan, Italy (MGB).

RESUMEN / SUMMARY: - Context:Tyrosine kinase inhibitors (TKI) represent a new treatment option for patients with advanced medullary thyroid cancer (MTC). However, cures have not been achieved with current available agents used in monotherapy.Objective:Since RET has been shown to negatively regulate CD95 death receptor activation in preclinical models of RET-dependent MTC, we investigated the potential of the combination approach with the RET targeting TKI sunitinib and cisplatin to enhance apoptosis activation through the extrinsic pathway.Design:The effects of sunitinib and cisplatin were examined in human MTC cell lines harboring oncogenic RET mutations. Experiments were designed to determine drug effects on RET signaling, cell growth, apoptosis, autophagy, tumor growth in mice, and to investigate the mechanisms of the drug interaction.Results:Sunitinib and cisplatin synergistically inhibited the growth of MZ-CRC-1 cells harboring the RET M918T activating mutation. The combination enhanced apoptosis activation through CD95-mediated, caspase-8 dependent, pathway. Moreover, sunitinib induced a severe perturbation of the autophagic flux characterized by autophagosome accumulation and a remarkable lysosomal dysfunction which was further enhanced, with lysosomal leakage induction, by cisplatin. Administration of the drug combination to mice xenografted with MZ-CRC-1 cells improved the antitumor efficacy, as compared to single agent treatments, inducing complete responses in 30% of treated mice, a significant increase in caspase-3 activation ($P < 0.01$ vs cisplatin, $P < 0.0005$ vs sunitinib) and apoptosis in tumor cells.Conclusions:Addition of cisplatin to sunitinib potentiates apoptotic cell death and has promising preclinical activity in MTCs harboring the RET M918T oncogene.

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[47]

TÍTULO / TITLE: - Malignant paraganglioma presenting with hemorrhagic stroke in a child.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Pediatrics. 2013 Dec;132(6):e1709-14. doi: 10.1542/peds.2013-0492. Epub 2013 Nov 25.

●● Enlace al texto completo (gratuito o de pago) 1542/peds.2013-0492

AUTORES / AUTHORS: - Luiz HV; da Silva TN; Pereira BD; Santos JG; Goncalves D; Manita I; Portugal J

INSTITUCIÓN / INSTITUTION: - Department of Endocrinology and Diabetology, Garcia de Orta Hospital, Avenida Torrado da Silva, 2801-951 Almada, Portugal. hvaraluiz@gmail.com.

RESUMEN / SUMMARY: - Sympathetic paragangliomas are rare catecholamine-secreting tumors of extra-adrenal origin, and their diagnosis in children is even more infrequent. They usually manifest as hypertension, palpitations, headache, sweating, and pallor. Malignant paragangliomas are identified by the presence of metastasis. Hemorrhagic stroke in the pediatric population is a life-threatening condition with several etiologies. We report here the case of a 12-year-old boy with malignant sympathetic paraganglioma presenting with hemorrhagic stroke. Severe hypertension was found and the patient evolved into a coma. Brain computed tomography scan showed right thalamus hemorrhage with intraventricular extension. After clinical improvement, further investigation revealed elevated catecholamine and metanephrine levels, and 2 abdominal tumors were identified by computed tomography. Resection of both lesions was performed, and histologic findings were consistent with

paraganglioma. Multiple metastatic involvement of bones and soft tissues appeared several years later. Genetic testing identified a mutation in succinate dehydrogenase subunit B gene, with paternal transmission. (131)I-metaiodobenzylguanidine therapy was performed 3 times with no tumoral response. Our patient is alive, with adequate quality of life, 25 years after initial diagnosis. To our knowledge, this is the first pediatric case of paraganglioma presenting with hemorrhagic stroke. Intracerebral hemorrhage was probably caused by severe hypertension due to paraganglioma. Therefore, we expand the recognized clinical spectrum of the disease. Physicians evaluating children with hemorrhagic stroke, particularly if hypertension is a main symptom, should consider the possibility of catecholamine-secreting tumors. Metastatic disease is associated with succinate dehydrogenase subunit B mutations and, although some patients have poor prognosis, progression can be indolent.

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[48]

TÍTULO / TITLE: - Autophagy sensitivity of neuroendocrine lung tumor cells.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Int J Oncol. 2013 Dec;43(6):2031-8. doi: 10.3892/ijo.2013.2136. Epub 2013 Oct 11.

●● Enlace al texto completo (gratuito o de pago) 3892/ijo.2013.2136

AUTORES / AUTHORS: - Hong SK; Kim JH; Starenki D; Park JI

INSTITUCIÓN / INSTITUTION: - Department of Biochemistry, Medical College of Wisconsin, Milwaukee, WI 53226, USA.

RESUMEN / SUMMARY: - Neuroendocrine (NE) phenotypes characterize a spectrum of lung tumors, including low-grade typical and intermediate-grade atypical carcinoid, high-grade large-cell NE carcinoma and small cell lung carcinoma. Currently, no effective treatments are available to cure NE lung tumors, demanding identification of biological features specific to these tumors. Here, we report that autophagy has an important role for NE lung tumor cell proliferation and survival. We found that the expression levels of the autophagy marker LC3 are relatively high in a panel of lung tumor cell lines expressing high levels of neuron-specific enolase (NSE), a key NE marker in lung tumors. In response to bafilomycin A1 and chloroquine, NE lung tumor cells exhibited cytotoxicity whereas non-NE lung tumor cells exhibited cytostasis, indicating a distinct role of autophagy for NE lung tumor cell survival. Intriguingly, in certain NE lung tumor cell lines, the levels of processed LC3 (LC3-II) were inversely correlated with AKT activity. When AKT activity was inhibited using AKTi or MK2206, the levels of LC3-II and SQSTM1/p62 were increased. In contrast, torin 1, rapamycin or mTOR knockdown increased p62 levels, suggesting that these two pathways have opposing effects on autophagy in certain NE lung tumors. Moreover, inhibition of one pathway resulted in reduced activity of the other, suggesting that these two pathways crosstalk in the tumors. These results suggest that NE lung tumor cells share a common feature of autophagy and are more sensitive to autophagy inhibition than non-NE lung tumor cells.

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[49]

TÍTULO / TITLE: - Potent antitumor activity of the novel HSP90 inhibitors AUY922 and HSP990 in neuroendocrine carcinoid cells.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Int J Oncol. 2013 Dec;43(6):1824-32. doi: 10.3892/ijo.2013.2130. Epub 2013 Oct 4.

●● Enlace al texto completo (gratuito o de pago) 3892/ijo.2013.2130

AUTORES / AUTHORS: - Zitzmann K; Ailer G; Vlotides G; Spoettl G; Maurer J; Goke B; Beuschlein F; Auernhammer CJ
INSTITUCIÓN / INSTITUTION: - Department of Internal Medicine II Campus Grosshadern, University-Hospital, Ludwig-Maximilians-University of Munich, D81377 Munich, Germany.
RESUMEN / SUMMARY: - The heat shock protein (HSP) 90 chaperone machine involved in numerous oncogenic signaling pathways is overexpressed in cancer cells and is currently being evaluated for anticancer therapy. Neuroendocrine tumors (NETs) of the gastroenteropancreatic (GEP) system comprise a heterogeneous group of tumors with increasing incidence and poor prognosis. Here, we report the antiproliferative effects of the HSP90 inhibitors AUY922 and HSP990 in neuroendocrine tumor cells. Treatment of human pancreatic BON1, bronchopulmonary NCI-H727 and midgut carcinoid GOT1 neuroendocrine tumor cells with increasing concentrations of AUY922 and HSP990 dose-dependently suppressed cell viability. Significant effects on neuroendocrine cell viability were observed with inhibitor concentrations as low as 5 nM. Inhibition of cell viability was associated with the induction of apoptosis as demonstrated by an increase in sub-G1 events and PARP cleavage. HSP90 inhibition was associated with decreased neuroendocrine ErbB and IGF-I receptor expression, decreased Erk and Akt phospho-rylation and the induction of HSP70 expression. These findings provide evidence that targeted inhibition of upregulated HSP90 activity could be useful for the treatment of aggressive neuroendocrine tumors resistant to conventional therapy.

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[50]
TÍTULO / TITLE: - Dandy-Walker Malformation, Papillary Thyroid Carcinoma, and SDHD-Associated Paraganglioma Syndrome.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - J Clin Endocrinol Metab. 2013 Oct 23.

●● Enlace al texto completo (gratuito o de pago) 1210/jc.2013-3015

AUTORES / AUTHORS: - Huguet I; Walker L; Karavitaki N; Byrne J; Grossman AB
INSTITUCIÓN / INSTITUTION: - Oxford Centre for Diabetes, Endocrinology, and Metabolism (I.H., N.K., A.B.G.), Churchill Hospital, University of Oxford, Oxford OX3 7LE, United Kingdom; Department of Clinical Genetics (L.W.), Churchill Hospital, Oxford OX3 7LE, United Kingdom; and Department of Neuroradiology (J.B.), Oxford Radcliffe Hospital, Oxford OX3 9DU, United Kingdom.
RESUMEN / SUMMARY: - Abstract Not Available.

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[51]
TÍTULO / TITLE: - Mosaicism in HIF2A-related polycythaemia-paraganglioma syndrome.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - J Clin Endocrinol Metab. 2013 Nov 25.

●● Enlace al texto completo (gratuito o de pago) 1210/jc.2013-2600

AUTORES / AUTHORS: - Buffet A; Smati S; Mansuy L; Menara M; Lebras M; Heymann MF; Simian C; Favier J; Murat A; Cariou B; Gimenez-Roqueplo AP

INSTITUCIÓN / INSTITUTION: - 1 Assistance Publique-Hopitaux de Paris, Hopital europeen Georges Pompidou, Service de Genetique, Paris, France;

RESUMEN / SUMMARY: - Context:HIF2A germline mutations were known to cause congenital polycythaemia. Recently HIF2A somatic mutations were found in several patients with polycythaemia and paraganglioma, pheochromocytoma or somatostatinoma suggesting the occurrence of de novo post-zygotic HIF2A mutation that has not been demonstrated clearly.Patients:Patient 1 is a woman suffering from polycythaemia diagnosed at the age of 16. She was operated on for a pheochromocytoma at 45 years and for two abdominal paragangliomas at 59 years. She was also diagnosed with somatostatinoma. Patient 2 is a young boy suffered from polycythaemia since infancy. He underwent surgery for a non-functional adrenal paraganglioma at the age of nine.Methods:We sequenced by Sanger and next generation sequencing the HIF2A gene in DNA extracted from tumors, leucocytes and buccal cells.Results:In patient 1, we identified a somatic HIF2A mutation (c.1586T>C; p.Leu529Pro) in DNA extracted from both paragangliomas. The mutation was detected as a somatic mosaic in DNA extracted from somatostatinoma and was absent from germline DNA. In patient 2, we found a HIF2A heterozygous mutation (c.1625T>C; p.Leu542Pro) in the paraganglioma but the mutation was also present as a mosaic in leucocyte DNA and in DNA extracted from buccal cells (3.3% and 8.96% of sequencing reads, respectively). Both mutations disrupt the hydroxylation domain of the HIF2alpha protein.Conclusions:Our study shows that HIF2A-related tumors are caused by postzygotic mutations occurring in early developmental stage. Potential germline mosaicism should be considered during the familial genetic counselling when an individual has been diagnosed with HIF2A-related polycythaemia-paraganglioma syndrome.

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[52]

TÍTULO / TITLE: - Penetrance of functioning and non-functioning pancreatic neuroendocrine tumors in multiple endocrine neoplasia type 1 in the second decade of life.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - J Clin Endocrinol Metab. 2013 Oct 31.

●● Enlace al texto completo (gratuito o de pago) 1210/jc.2013-1768

AUTORES / AUTHORS: - Goncalves TD; Toledo RA; Sekiya T; Matuguma SE; Maluf Filho F; Rocha MS; Siqueira SA; Glezer A; Bronstein MD; Pereira Ricardo Jureidini MA; Bacchella T; Machado MC; Toledo SP; Lourenco DM Jr

INSTITUCIÓN / INSTITUTION: - Endocrine Genetics Unit (LIM-25) (T.D.G., R.A.T.*, T.S., S.P.A.T., D.M.L.J.), Endocrinology Division (T.D.G., R.A.T., T.S., A.G., M.D.B., M.A.A.P., S.P.A.T., D.M.L.J.), Endoscopy Division (S.E.M., F.M.F.), Radiology Division (M.S.R.), Pathology Division (S.A.C.S.), Department of Gastroenterology, Surgical Division (T.B., M.C.C.M.), Hospital das Clinicas, University of Sao Paulo School of Medicine; Department of Gastroenterology, Surgical Division (T.B., R.J.) and Endocrine Oncology Division, Cancer Institute of Sao Paulo (D.M.L.J.), Sao Paulo, Brazil.; * R.A.T is now at Division of Hematology and Medical Oncology, Department of Medicine,

Cancer Therapy and Research Center at the University of Texas Health Science Center, 7703 Floyd Curl Dr, MC 7880, San Antonio, TX 78229-3900, USA.
RESUMEN / SUMMARY: - Context:Data are scarce on penetrance of multiple endocrine neoplasia type 1 (MEN1)-related non-functioning pancreatic neuroendocrine tumors (NF-PETs) and insulinomas in young MEN1 patients. A potential positive correlation between tumor size and malignancy (2-3cm, 18%; >3cm, 43%) has greatly influenced the management of MEN1 adults with NF-PETs.Objective:To estimate the penetrance of NF-PETs, insulinomas and gastrinomas in young MEN1 carriers.Design:The data were obtained from a screening program (1996-2012) involving 113 MEN1 patients (tertiary academic reference center).Patients:Nineteen MEN1 patients (aged 12-20y; 16 patients aged 15-20y and 3 patients aged 12-14y) were screened for NF-PETs, insulinomas and gastrinomas.Methods:MRI/CT and endoscopic US (EUS) were performed on 10 MEN1 carriers, MRI/CT was performed on five patients, and four other patients underwent an EUS.Results:The overall penetrance of PETs during the second decade of life was 42% (8/19). All eight PET patients had NF-PETs, and half of those tumors were multicentric. One-fifth of the screened patients (21%; 4/19) harbored at least one large tumor (>2.0cm). Insulinoma was detected in two NF-PET patients (11%) at the initial screening; gastrinoma was not present in any cases. Six of the 11 (54%) screened patients aged 15-20y who underwent an EUS had NF-PETs. Potential false-positive EUS results were excluded based on EUS-guided biopsy results, the reproducibility of the NF-PET findings or the observation of increased tumor size during follow-up. Distal pancreatectomy and the nodule enucleation of pancreatic head tumors were conducted on three patients with large tumors (>2.0cm; T2N0M0) that were classified as grade 1 neuroendocrine tumors (Ki-67<2%).Conclusions:Our data demonstrated high penetrance of NF-PETs in 15-20-y-old MEN1 patients. The high percentage of the patients presenting consensus criteria for surgery for NF-PET alone or NF-PET/insulinoma suggests a potential benefit for the periodic surveillance of these tumors in this age group.

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[53]
TÍTULO / TITLE: - Clinical outcomes of rectal neuroendocrine tumors </= 10 mm following endoscopic resection.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Endoscopy. 2013 Dec;45(12):1018-23. doi: 10.1055/s-0033-1344860. Epub 2013 Nov 28.
●● Enlace al texto completo (gratuito o de pago) 1055/s-0033-1344860
AUTORES / AUTHORS: - Kim GU; Kim KJ; Hong SM; Yu ES; Yang DH; Jung KW; Ye BD; Byeon JS; Myung SJ; Yang SK; Kim JH
INSTITUCIÓN / INSTITUTION: - Department of Gastroenterology, University of Ulsan College of Medicine, Asan Medical Center, Seoul, Korea.
RESUMEN / SUMMARY: - Background and study aims: This study was conducted to determine the clinical outcome of rectal neuroendocrine tumors (NETs) </= 10 mm following conventional endoscopic resection. Patients and methods: A total of 107 patients who underwent conventional endoscopic treatment for rectal NETs (median size 5.0 mm [range 1.0 - 10.0]) were followed up for a median of 31 months (range 13 - 121). The following data were analyzed: lesion characteristics, clinical outcomes, and histological features determined using tissue microarray analysis (TMA), including the

mitotic count and the Ki-67 index. Results: En bloc removal was achieved for all tumors, and the complete resection rate was 49.5 % (53 /107). Resection margin status was indeterminate in 37 patients (34.6 %) and positive in 17 (15.9 %). Rectal NETs in 71 patients demonstrated a score of </= 2 % on the Ki-67 index and < 2 for mitotic count on TMA. In another 28 tumors that did not undergo TMA, the mitotic count was 0 - 1 per 10 high-power fields. Neither recurrence nor metastasis was noted during the follow-up period following resection. Conclusions: Rectal NETs (</= 10 mm in size) appear to demonstrate benign behavior based on the mitotic count and the Ki-67 index. These results suggest that the outcome of rectal NETs (</= 10 mm in size) following conventional endoscopic resection might be comparatively excellent, regardless of the resection margin status. However, long term follow-up data are required to confirm this.

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[54]

TÍTULO / TITLE: - Role of Preoperative Adrenergic Blockade with Doxazosin on Hemodynamic Control during the Surgical Treatment of Pheochromocytoma: A Retrospective Study of 48 Cases.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Am Surg. 2013 Nov;79(11):1196-202.

AUTORES / AUTHORS: - Conzo G; Musella M; Corcione F; Depalma M; Stanzione F; Della-Pietra C; Palazzo A; Napolitano S; Pasquali D; Milone M; Agostino-Sinisi A; Ferraro F; Santini L

INSTITUCIÓN / INSTITUTION: - Department of Anesthesiologic, Surgical and Emergency Science, VII Division of General Surgery Second University of Naples, Naples, Italy.

RESUMEN / SUMMARY: - Authors evaluated the effects of selective adrenergic blockade by means of doxazosin on blood pressure in 48 patients operated on for pheochromocytoma by a multicenter retrospective study. Age, tumor size, surgical approach, and operative time were analyzed as predictive factors of intraoperative hypertensive crises. Forty-eight patients underwent adrenalectomy-four open surgery and 44 laparoscopic surgery-for pheochromocytoma of adrenal glands from 1998 to 2008 after preoperative administration of doxazosin. Perioperative cardiovascular status modifications and surgical medium- and long-term outcomes were analyzed. There was no mortality, conversion rate was 4.5 per cent, and morbidity rate was 8.3 per cent. Intraoperative hypertensive crises (180/90 mmHg or higher) were observed in 14.5 per cent (seven of 48) of patients and were treated pharmacologically with no aftermath. None of the examined variables influenced the occurrence of intraoperative hypertensive episodes. Postoperative hypotension (lower than 90/60 mmHg) was observed in four of 48 patients (8.3%) and was treated by crystalloids and hydrocortisone. In the surgical treatment of pheochromocytoma, the preoperative adrenergic blockade by doxazosin does not prevent intraoperative hypertensive crises. Nevertheless, in our series, they were of short duration and were not associated with major cardiovascular complications. Perioperative hemodynamic instability was managed by preoperative pharmacological treatment, allowing low morbidity.

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[55]

TÍTULO / TITLE: - Prognostic factors in a multicentre study of 247 atypical pulmonary carcinoids.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Eur J Cardiothorac Surg. 2013 Oct 6.

●● Enlace al texto completo (gratuito o de pago) 1093/ejcts/ezt470

AUTORES / AUTHORS: - Daddi N; Schiavon M; Filosso PL; Cardillo G; Ambrogi MC; De Palma A; Luzzi L; Bandiera A; Casali C; Ruffato A; De Angelis V; Andriolo LG; Guerrera F; Carleo F; Davini F; Urbani M; Mattioli S; Morandi U; Zannini P; Gotti G; Loizzi M; Puma F; Mussi A; Ricci A; Oliaro A; Rea F

INSTITUCIÓN / INSTITUTION: - Thoracic Surgery Unit, Perugia University School of Medicine, Perugia, Italy.

RESUMEN / SUMMARY: - OBJECTIVES: To analyse clinical and biomolecular prognostic factors associated with the surgical approach and the outcome of 247 patients affected by primary atypical carcinoids (ACs) of the lung in a multi-institutional experience. METHODS: We retrospectively evaluated clinical data and pathological tissue samples collected from 247 patients of 10 Thoracic Surgery Units from different geographical areas of our country. All patients were divided into four groups according to surgical procedure: sub-lobar resections (SURG1), lobar resections (SURG2), tracheobronchoplastic procedures (SURG3) and pneumonectomies (SURG4). Overall survival analysis was performed using the Kaplan-Meier method and log-rank test. Survival was calculated from the date of surgery to the last date of follow-up or death. The parameters evaluated included age, gender, smoking habits, laterality, type of surgery, 7th edition of TNM staging, mitosis Ki-67 (MIB1), multifocal forms, tumourlets, type of lymphadenectomy and neo/adjuvant therapy. For multivariate analysis, a Cox regression model was used with a forward stepwise selection of covariates. RESULTS: Two hundred and forty-seven patients (124 females and 123 males; range 10-84, median 60 years) underwent surgical resection for AC in the last 30 years as follows: n = 38 patients in SURG1, 181 in SURG2, 15 in SURG3 and 14 in SURG4. A smoking history was present in 136 of 247 (55%) patients. The median follow-up period was 98.7 (range 11.2-369.9) months. The overall survival probability analysis of the AC was 86.7% at 5 years, 72.4% at 10 years, 64.4% at 15 years and 58.1% at 20 years. Neuroendocrine multicentric forms were detected in 12 of 247 patients (4.8%; 1 of 12 pts) during the follow-up (range 11.2-200.4, median 98.7 months) and 33.4% had recurrence of disease. There were no significant differences between gender, tumour location and type of surgery at the multivariate analysis. Age [P < 0.001, hazard ratio (HR) 0.60; confidence interval (CI) 0.32-1.12], smoking habits (P = 0.002; HR 0.43, 95% CI 0.23-0.80) and lymph nodal metastatic involvement (P = 0.008; HR 0.46, 95% CI 0.26-0.82) were all significant at multivariate analysis. CONCLUSIONS: ACs of the lung are malignant neuroendocrine tumours with a worst outcome in patients over 70 years and in smokers. With the exception of pneumonectomy, the extent of resection does not seem to affect survival and should be accompanied preferably by lymphadenectomy. Pathological staging, along with a mitotic index more than Ki-67 (MIB1), appears to be the most significant prognostic factor at the univariate analysis.

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[56]

TÍTULO / TITLE: - Capsaicin induces cytotoxicity in pancreatic neuroendocrine tumor cells via mitochondrial action.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Cell Signal. 2014 Jan;26(1):41-8. doi: 10.1016/j.cellsig.2013.09.014. Epub 2013 Sep 27.

●● Enlace al texto completo (gratuito o de pago) 1016/j.cellsig.2013.09.014

AUTORES / AUTHORS: - Skrzypski M; Sassek M; Abdelmessih S; Mergler S; Grotzinger C; Metzke D; Wojciechowicz T; Nowak KW; Strowski MZ

INSTITUCIÓN / INSTITUTION: - Department of Hepatology and Gastroenterology & the Interdisciplinary Centre of Metabolism: Endocrinology, Diabetes and Metabolism, Charite-University Medicine Berlin, 13353 Berlin, Germany; Department of Animal Physiology and Biochemistry, Poznan University of Life Sciences, 60-637 Poznan, Poland. Electronic address: marek.skrzypski@charite.de.

RESUMEN / SUMMARY: - Capsaicin (CAP), the pungent ingredient of chili peppers, inhibits growth of various solid cancers via TRPV1 as well as TRPV1-independent mechanisms. Recently, we showed that TRPV1 regulates intracellular calcium level and chromogranin A secretion in pancreatic neuroendocrine tumor (NET) cells. In the present study, we characterize the role of the TRPV1 agonist - CAP - in controlling proliferation and apoptosis of pancreatic BON and QGP-1 NET cells. We demonstrate that CAP reduces viability and proliferation, and stimulates apoptotic death of NET cells. CAP causes mitochondrial membrane potential loss, inhibits ATP synthesis and reduces mitochondrial Bcl-2 protein production. In addition, CAP increases cytochrome c and cleaved caspase 3 levels in cytoplasm. CAP reduces reactive oxygen species (ROS) generation. The antioxidant N-acetyl-l-cysteine (NAC) acts synergistically with CAP to reduce ROS generation, without affecting CAP-induced toxicity. TRPV1 protein reduction by 75% reduction fails to attenuate CAP-induced cytotoxicity. In summary, these results suggest that CAP induces cytotoxicity by disturbing mitochondrial potential, and inhibits ATP synthesis in NET cells. Stimulation of ROS generation by CAP appears to be a secondary effect, not related to CAP-induced cytotoxicity. These results justify further evaluation of CAP in modulating pancreatic NETs in vivo.

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[57]

TÍTULO / TITLE: - Amylase alpha-1A (AMY1A): A Novel Immunohistochemical Marker to Differentiate Chromophobe Renal Cell Carcinoma From Benign Oncocytoma.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Am J Surg Pathol. 2013 Dec;37(12):1824-30. doi: 10.1097/PAS.0000000000000108.

●● Enlace al texto completo (gratuito o de pago) 1097/PAS.0000000000000108

AUTORES / AUTHORS: - Jain S; Roy S; Amin M; Acquafondata M; Yin M; Laframboise W; Bastacky S; Pantanowitz L; Dhir R; Parwani A

INSTITUCIÓN / INSTITUTION: - *Department of Pathology, University of Pittsburgh Medical Center, Pittsburgh, PA daggerDepartment of Pathology, East Carolina University, Greenville, NC.

RESUMEN / SUMMARY: - Chromophobe renal cell carcinoma (ChRCC) and oncocytoma present with a perplexing overlap of morphologic and immunohistochemical features. ChRCC have deletions in the 1p21.1 region including the amylase alpha-1A gene (AMY1A). No such deletions are found in oncocytoma. Instead, oncocytomas shared other deletions on chromosome 1: 1p31.3, 1q25.2, and

1q44. We performed AMY1A immunostaining on 75 oncocytomas (57 tissue microarray [TMA] cores, 18 whole slides) and 54 ChRCCs (20 TMA cores, 34 whole slides). Staining was assessed using the H-score method. The intensity was graded as follows: no staining=0, weak=1, moderate=2, and strong=3. The AMY1A immunostain preferentially stained the distal tubules and collecting ducts of normal kidney. All oncocytomas (100%) expressed AMY1A with an H-score that varied from 100 to 300 (mean 205). Mild to moderate heterogeneity in staining intensity was noted within a given oncocytoma. For oncocytomas, 87% (65/75) cases had H-scores of at least 120 with a mean score of 221. Notably, the 13% (10/75) of oncocytoma cases that had an H-score of 100 were derived from the TMA. A total of 87% (47/54) of the ChRCC cases were negative for the AMY1A immunostain. Of the ChRCC cases, 4% (2/54) showed very weak cytoplasmic staining (H-score of 70 each), which was less than the lowest H-score of oncocytoma cases. All 5 cases of ChRCC, which showed an H-score of 100 or more, were referred to as eosinophilic variants of ChRCC. Three of these 5 cases showed a very nondescript, diffuse staining of the cytoplasm. Two of these 5 cases showed an H-score of 130. We think that as the staining pattern of these 2 cases is similar to that of oncocytoma, they should be put in a category of renal oncocytic neoplasms favoring oncocytoma. This result shows that AMY1A staining could be very helpful in further classifying even a subset of the eosinophilic variants of ChRCC. The difference between ChRCC and oncocytoma was statistically significant (chi test, P<0.0001). All cases of clear cell RCC and papillary RCC were negative for AMY1A expression. Overall, sensitivity and specificity of AMY1A staining for oncocytoma was 100% (95% confidence interval, 0.95-1.00) and 96.75% (95% confidence interval, 0.93-0.99), respectively. Similarly, the sensitivity and specificity for distinguishing oncocytoma from ChRCC was 100% (95% confidence interval, 0.95-1.00) and 90.74% (95% confidence interval, 0.80-0.97), respectively. These data show that the novel marker AMY1A can be of great diagnostic utility when trying to differentiate ChRCC (classic and eosinophilic variant) and oncocytoma.

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[58]

TÍTULO / TITLE: - Overexpression of Membrane Proteins in Primary and Metastatic Gastrointestinal Neuroendocrine Tumors.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Ann Surg Oncol. 2013 Oct 10.

●● Enlace al texto completo (gratuito o de pago) 1245/s10434-013-3318-6

AUTORES / AUTHORS: - Carr JC; Sherman SK; Wang D; Dahdaleh FS; Bellizzi AM; O'Dorisio MS; O'Dorisio TM; Howe JR

INSTITUCIÓN / INSTITUTION: - Department of Surgery, University of Iowa Carver College of Medicine, Iowa City, IA, USA.

RESUMEN / SUMMARY: - BACKGROUND: Small bowel and pancreatic neuroendocrine tumors (SBNETs and PNETs) are rare tumors whose incidence is increasing. Drugs targeting the somatostatin receptor are beneficial in these tumors. To identify additional cell-surface targets, we recently found receptors and membrane proteins with gene expression significantly different from adjacent normal tissues in a small number of primary SBNETs and PNETs. We set out to validate these expression differences in a large group of primary neuroendocrine tumors and to determine whether they are present in corresponding liver and lymph node metastases. METHODS: Primary

SBNETs and PNETs, normal tissue, nodal, and liver metastases were collected and mRNA expression of six target genes was determined by quantitative PCR. Expression was normalized to GAPDH and POLR2A internal controls, and differences as compared to normal tissue were assessed by Welch's t test. RESULTS: Gene expression was determined in 45 primary PNETs with 20 nodal and 17 liver metastases, and 51 SBNETs with 50 nodal and 29 liver metastases. Compared to normal tissue, the oxytocin receptor (OXTR) showed significant overexpression in both primary and metastatic SBNETs and PNETs. Significant overexpression was observed for MUC13 and MEP1B in PNET primary tumors, and for GPR113 in primary SBNETs and their metastases. SCTR and ADORA1 were significantly underexpressed in PNETs and their metastases. OXTR protein expression was confirmed by immunohistochemistry. CONCLUSIONS: OXTR is significantly overexpressed relative to normal tissue in primary SBNETs and PNETs, and this overexpression is present in their liver and lymph node metastases, making OXTR a promising target for imaging and therapeutic interventions.

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[59]

TÍTULO / TITLE: - Grading of well-differentiated pancreatic neuroendocrine tumors is improved by the inclusion of both Ki67 proliferative index and mitotic rate.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Am J Surg Pathol. 2013 Nov;37(11):1671-7. doi: 10.1097/PAS.0000000000000089.

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

1097/PAS.0000000000000089

AUTORES / AUTHORS: - McCall CM; Shi C; Cornish TC; Klimstra DS; Tang LH; Basturk O; Mun LJ; Ellison TA; Wolfgang CL; Choti MA; Schulick RD; Edil BH; Hruban RH

INSTITUCIÓN / INSTITUTION: - Departments of *Pathology section signSurgery, The Sol Goldman Pancreatic Cancer Research Center, The Johns Hopkins University School of Medicine, Baltimore, MD daggerDepartment of Pathology, Microbiology and Immunology, Vanderbilt University School of Medicine, Nashville, TN double daggerDepartment of Pathology, Memorial Sloan-Kettering Cancer Center, New York, NY parallelDepartment of Surgery, University of Colorado Anschutz Medical Campus, Aurora, CO.

RESUMEN / SUMMARY: - The grading system for pancreatic neuroendocrine tumors (PanNETs) adopted in 2010 by the World Health Organization (WHO) mandates the use of both mitotic rate and Ki67/MIB-1 index in defining the proliferative rate and assigning the grade. In cases when these measures are not concordant for grade, it is recommended to assign the higher grade, but specific data justifying this approach do not exist. Thus, we counted mitotic figures and immunolabeled, using the Ki67 antibody, 297 WHO mitotic grade 1 and 2 PanNETs surgically resected at a single institution. We quantified the Ki67 proliferative index by marking at least 500 cells in "hot spots" and by using digital image analysis software to count each marked positive/negative cell and then compared the results with histologic features and overall survival. Of 264 WHO mitotic grade 1 PanNETs, 33% were WHO grade 2 by Ki67 proliferative index. Compared with concordant grade 1 tumors, grade-discordant tumors were more likely to have metastases to lymph node (56% vs. 34%) (P<0.01)

and to distant sites (46% vs. 12%) (P<0.01). Discordant mitotic grade 1 PanNETs also showed statistically significantly more infiltrative growth patterns, perineural invasion, and small vessel invasion. Overall survival was significantly different (P<0.01), with discordant mitotic grade 1 tumors showing a median survival of 12 years compared with 16.7 years for concordant grade 1 tumors. Conversely, mitotic grade 1/Ki67 grade 2 PanNETs showed few significant differences from tumors that were mitotic grade 2 and either Ki67 grade 1 or 2. Our data demonstrate that mitotic rate and Ki67-based grades of PanNETs are often discordant, and when the Ki67 grade is greater than the mitotic grade, clinical outcomes and histopathologic features are significantly worse than concordant grade 1 tumors. Patients with discordant mitotic grade 1/Ki67 grade 2 tumors have shorter overall survival and larger tumors with more metastases and more aggressive histologic features. These data strongly suggest that Ki67 labeling be performed on all PanNETs in addition to mitotic rate determination to define more accurately tumor grade and prognosis.

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[60]
TÍTULO / TITLE: - Merkel Cell Polyomavirus Positive Merkel Cell Carcinoma Requires Viral Small T Antigen For Cell Proliferation.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - J Invest Dermatol. 2013 Nov 12. doi: 10.1038/jid.2013.483.
●● Enlace al texto completo (gratuito o de pago) 1038/jid.2013.483
AUTORES / AUTHORS: - Shuda M; Chang Y; Moore PS
INSTITUCIÓN / INSTITUTION: - Cancer Virology Program, University of Pittsburgh, Pittsburgh, Pennsylvania, USA.

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[61]
TÍTULO / TITLE: - Adenomatous polyposis coli gene involvement in ileal enterochromaffin cell neuroendocrine neoplasms.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Hum Pathol. 2013 Dec;44(12):2736-42. doi: 10.1016/j.humpath.2013.06.019. Epub 2013 Oct 16.
●● Enlace al texto completo (gratuito o de pago) 1016/j.humpath.2013.06.019
AUTORES / AUTHORS: - Bottarelli L; Azzoni C; Pizzi S; D'Adda T; Silini EM; Bordi C; Rindi G
INSTITUCIÓN / INSTITUTION: - Centre for Molecular and Translational Oncology (COMT), Department of Biomedical, Biotechnological and Translational Sciences, Unit of Pathological Anatomy University and University Hospital of Parma, 43126 Parma, Italy.
RESUMEN / SUMMARY: - The adenomatous polyposis coli gene is a key tumor suppressor gene. Alterations in this gene have been found in most sporadic colon cancers; associated with familial adenomatous polyposis; and found in neoplasms of other organs, such as the liver, stomach, lung, breast, and cerebellar medulloblastoma. In the heterogeneous group of neuroendocrine neoplasms of the gastrointestinal tract, the involvement of adenomatous polyposis coli is debated, and only occasional reports found adenomatous polyposis coli alterations in foregut and midgut neuroendocrine neoplasms, with adenomatous polyposis coli mutations only in the latter. To elucidate

the penetrance of adenomatous polyposis coli alterations in ileal neuroendocrine neoplasms, we performed DNA fragment analysis (loss of heterozygosity for 5q22-23 and 5q23) and sequencing on the mutation cluster region of the adenomatous polyposis coli gene on 30 ileal enterochromaffin cell neuroendocrine neoplasms. Adenomatous polyposis coli gene mutations were detected in 23% of cases (7/30); in particular, 57% were missense and 14%, nonsense/frameshift, all novel and different from those reported in colorectal or other cancers. Loss of heterozygosity analysis demonstrated a deletion frequency of 15% (4/27). No association was found with features of tumor progression. Our observations support the involvement of somatic adenomatous polyposis coli alterations in tumorigenesis of ileal enterochromaffin cell neuroendocrine neoplasms; the mechanisms of adenomatous polyposis coli gene inactivation appear to be different from those reported in other tumor types.

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[62]

TÍTULO / TITLE: - Metabolic expressivity of human genetic variants: NMR metabotyping of MEN1 pathogenic mutants.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - J Pharm Biomed Anal. 2013 Oct 16. pii: S0731-7085(13)00459-7. doi: 10.1016/j.jpba.2013.09.029.

●● Enlace al texto completo (gratuito o de pago) 1016/j.jpba.2013.09.029

AUTORES / AUTHORS: - Blaise BJ; Lopez C; Vercherat C; Lacheretz-Bernigaud A; Bayet-Robert M; Rezig L; Scoazec JY; Calender A; Emsley L; Elena-Herrmann B; Cordier-Bussat M

INSTITUCIÓN / INSTITUTION: - Universite de Lyon, Institut des Sciences Analytiques CNRS/ENS Lyon/UCB Lyon 1, Centre de RMN a Tres Hauts Champs, 5 rue de la Doua, 69100 Villeurbanne, France.

RESUMEN / SUMMARY: - Functional consequences of mutations in predisposition genes for familial cancer syndromes remain often elusive, especially when the corresponding gene products play pleiotropic functions and interact with numerous partners. Understanding the consequences of these genetic alterations requires access to their functional effects at the phenotypic level. Nuclear magnetic resonance (NMR) has emerged as a promising functional genomics probe, through its ability to monitor the consequences of genetic variations at the biochemical level. Here, we determine by NMR the metabolic perturbations associated with different disease-related mutations in the MEN1 gene, responsible for the multiple endocrine neoplasia syndrome, type 1 (MEN1), an example of hereditary cancer. The MEN1 gene encodes the Menin protein. Based on a cellular model that allows exogenous overexpression of either the wild type (WT) Menin protein or disease-related variant forms, we evaluate the feasibility of using metabolic profiles to discriminate cells with WT versus variant Menin overexpression. High-resolution magic angle spinning (HRMAS) NMR of whole cells allows to determine the metabolic features associated with overexpression of WT Menin as compared to the one of six different missense variants observed in MEN1 patients. We then identify several statistically significant individual metabolites associated with the metabolic signature of pathogenic versus WT variants. Whether such a metabolic phenotyping approach using cell lines could be exploited as a functional test in a human genetic cancer syndrome is further discussed.

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[63]

TÍTULO / TITLE: - A looming danger: pheochromocytoma.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Am J Med. 2013 Dec;126(12):1054-6. doi: 10.1016/j.amjmed.2013.08.023. Epub 2013 Oct 14.

●● Enlace al texto completo (gratuito o de pago) 1016/j.amjmed.2013.08.023

AUTORES / AUTHORS: - Li Y; Spiler IJ; Fahey T 3rd; Akbar G; Pattan V; Jessani N; Hossain M; Yousif A

INSTITUCIÓN / INSTITUTION: - Department of Medicine, Raritan Bay Medical Center, Perth Amboy, NJ. Electronic address: yli@rbmc.org.

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[64]

TÍTULO / TITLE: - Multimodality therapy for large cell neuroendocrine carcinoma of the thymus.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Ann Thorac Surg. 2013 Oct;96(4):e85-7. doi: 10.1016/j.athoracsur.2013.04.107.

●● Enlace al texto completo (gratuito o de pago) 1016/j.athoracsur.2013.04.107

AUTORES / AUTHORS: - Ose N; Inoue M; Morii E; Shintani Y; Sawabata N; Okumura M

INSTITUCIÓN / INSTITUTION: - Department of General Thoracic Surgery, Toneyama National Hospital, Osaka, Japan. Electronic address: naokoose@toneyama.go.jp.

RESUMEN / SUMMARY: - A case of large cell neuroendocrine carcinoma of the thymus successfully treated with chemoradiation, followed by extended resection under cardiopulmonary bypass, is reported. A 44-year-old man diagnosed with thymic large cell neuroendocrine carcinoma received induction chemoradiation therapy (3 cycles of cisplatin/etoposide and 45 Gy of hyperfractionated radiation) because of invasion to the aortic arch and pulmonary trunk. After radiographic partial response was noted, a radical resection under cardiopulmonary bypass was performed. Pathologic examination revealed no viable cells in the tumor. The patient is alive 3 years later, without recurrence. Aggressive multimodality therapy could be an option for thymic large cell neuroendocrine carcinoma.

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[65]

TÍTULO / TITLE: - Evaluation of somatostatin receptor subtype expression in human neuroendocrine tumors using two sets of new monoclonal antibodies.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Regul Pept. 2013 Nov 10;187:35-41. doi: 10.1016/j.regpep.2013.10.007. Epub 2013 Nov 1.

●● Enlace al texto completo (gratuito o de pago) 1016/j.regpep.2013.10.007

AUTORES / AUTHORS: - Lambertini C; Barzaghi-Rinaudo P; D'Amato L; Schulz S; Nuciforo P; Schmid HA

INSTITUCIÓN / INSTITUTION: - Novartis Pharma AG, CH-4057 Basel, Switzerland. Electronic address: chiara.lambertini@novartis.com.

RESUMEN / SUMMARY: - INTRODUCTION: The expression and reliable detection of somatostatin receptor subtypes (SSTR1-5) is a prerequisite for the successful use of somatostatin analogs in neuroendocrine tumors (NETs). Two sets of monoclonal antibodies (mAbs) against human SSTR1, 2A, 3 and 5 have recently been developed by two independent laboratories using rabbit and mouse hybridomas. Our aim was to evaluate the usefulness of both sets of mAbs for detection of SSTRs in NET samples as they are routinely collected in clinical practice. METHODS: Mouse and rabbit mAbs were characterized in SSTR1, 2A, 3 and 5-transfected HEK293 cells and human archival samples of pancreatic tissue and NET. Comparative analysis of mAbs was also conducted by immunostaining of a tissue microarray composed of 75 cores of NET. RESULTS: Immunohistochemical analysis of HEK293 cells showed that both rabbit and mouse mAbs specifically detect their cognate receptor subtype, with mild cytoplasmic cross-reactivity observed for rabbit mAbs. Both sets of mAbs labeled normal pancreatic islets and showed similar patterns of immunoreactivity in NET controls. Direct comparison of mAb sets using a NET tissue microarray revealed strong correlation between rabbit and mouse mAbs against SSTR1 and 5, and moderate correlation for SSTR3. The rabbit mAb against SSTR2A showed higher affinity for its cognate receptor than the corresponding mouse mAb, resulting in a more reliable detection of this SSTR. CONCLUSIONS: mAbs from both sets are reliable tools for the detection of SSTR1, 3 and 5, whereas the rabbit mAb against SSTR2A is recommended for use in routine clinical testing due to its superior binding affinity.

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[66]

TÍTULO / TITLE: - Vandetanib for the Treatment of Medullary Thyroid Carcinoma.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Ann Pharmacother. 2013 Nov 14.

●● Enlace al texto completo (gratuito o de pago) 1177/1060028013512791

AUTORES / AUTHORS: - Cooper MR; Yi SY; Alghamdi W; Shaheen DJ; Steinberg M

INSTITUCIÓN / INSTITUTION: - MCPHS University, Manchester, NH, USA.

RESUMEN / SUMMARY: - OBJECTIVE: To review the place in therapy of vandetanib for medullary thyroid carcinoma (MTC). DATA SOURCES: Literature searches were performed in Ovid MEDLINE, EMBASE, and Google Scholar using the search terms ZD6474 OR vandetanib OR Caprelsa combined with medullary thyroid carcinoma. STUDY SELECTION AND DATA EXTRACTION: Two phase 2 trials and 1 phase 3 trial were identified. DATA SYNTHESIS: Vandetanib is approved for the treatment of unresectable, locally advanced or metastatic MTC in patients with symptomatic or progressive disease. In the phase 3 randomized, double-blind, placebo-controlled trial, vandetanib 300 mg daily (n = 231) was compared with placebo (n = 100). Vandetanib-treated patients experienced a significant improvement in progression-free survival (PFS; hazard ratio [HR] = 0.46; 95% CI = 0.31-0.69; P < .001). No difference in overall survival (OS) was seen at the time of publication. Most adverse effects were grade 1 or 2 and managed by dose interruptions or reductions. The most common grade ¾ adverse effects were diarrhea, hypertension, QT prolongation, fatigue, and rash. Because of the potential for QT prolongation, torsades de pointes, and sudden death, vandetanib is restricted via a Risk Evaluations and Mitigation Strategy program. CONCLUSIONS: Vandetanib prolongs PFS but has not been shown to improve OS. Vandetanib can be considered for patients with unresectable locoregional disease. It is

a first-line option for patients with unresectable symptomatic distant metastases as well as an option for advanced disseminated symptomatic metastatic disease. Vandetanib is expected to be an important addition to the formulary of health plans that provide prescription drug benefits.

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[67]

TÍTULO / TITLE: - Next-generation sequencing for the genetic screening of pheochromocytomas and paragangliomas: riding the new wave, but with caution.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Clin Endocrinol (Oxf). 2013 Oct 29. doi: 10.1111/cen.12357.

●● Enlace al texto completo (gratuito o de pago) 1111/cen.12357

AUTORES / AUTHORS: - Toledo RA; Dahia PL

INSTITUCIÓN / INSTITUTION: - Division of Hematology and Medical Oncology, Dept. Medicine, Cancer Therapy and Research Center, University of Texas Health Science Center at San Antonio.

RESUMEN / SUMMARY: - Pheochromocytomas (PHEO) and Paragangliomas (PGL) are catecholamine-secreting tumors derived from chromaffin cells of the adrenal medulla or sympathetic paraganglia, respectively, which carry high genetic and allelic heterogeneity 1 . More than one-third of all PHEO/PGLs have a pathogenic germline mutation in one of several susceptibility genes and genetic testing is now recommended for all patients. Furthermore, somatic driver mutations were recently recognized as frequent events in these tumors 2-5 . In all, 16 different PCC/PGL-related genes have been identified (VHL, RET, NF1, SDHA, SDHB, SDHC, SDHD, SDHAF2, MAX, TMEM127, HIF2A/EPAS1, KIF1B, PDH2, FH and HRAS), involving a total of 20,791-30,078 coding nucleotides spanned by 141-217 exons, depending on the gene isoform analyzed. This has become a laborious and costly process for genetic laboratories.

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[68]

TÍTULO / TITLE: - The activation of the WNT signalling pathway is a hallmark in Neurofibromatosis type 1 tumorigenesis.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Clin Cancer Res. 2013 Nov 11.

●● Enlace al texto completo (gratuito o de pago) 1158/1078-0432.CCR-13-0780

AUTORES / AUTHORS: - Luscan A; Shackleford G; Masliah-Planchon J; Laurendeau I; Ortonne N; Varin J; Lallemand F; Leroy K; Dumaine V; Hivelin M; Borderie D; De Raedt T; Valeyrie-Allanore L; Larousserie F; Terris B; Lantieri L; Vidaud M; Vidaud D; Wolkenstein P; Parfait B; Bieche I; Massaad C; Pasmant E

INSTITUCIÓN / INSTITUTION: - UMR745, INSERM.

RESUMEN / SUMMARY: - PURPOSE: The hallmark of neurofibromatosis type 1 (NF1) is the onset of dermal or plexiform neurofibromas, mainly composed of Schwann cells. Plexiform neurofibromas can transform into malignant peripheral nerve sheath tumors (MPNSTs) that are resistant to therapies. Experimental design: The aim of this study was to identify an additional pathway in the NF1-tumorigenesis. We focused our work on Wnt signalling that is highly implicated in cancer, mainly in regulating the

proliferation of cancer stem cells. We quantified mRNAs of 89 Wnt pathway genes in 57 NF1-associated tumors including dermal and plexiform neurofibromas and MPNSTs. Expression of two major stem cell marker genes and five major epithelial-mesenchymal transition marker genes was also assessed. The expression of significantly deregulated Wnt genes was then studied in normal human Schwann cells, fibroblasts, endothelial cells, and mast cells and in seven MPNST cell lines. RESULTS: The expression of nine Wnt genes was significantly deregulated in plexiform neurofibromas in comparison with dermal neurofibromas. Twenty Wnt genes showed altered expression in MPNST biopsies and cell lines. Immunohistochemical studies confirmed the Wnt pathway activation in NF1-associated MPNSTs. We then confirmed that the knock-down of NF1 in Schwann cells but not in epithelial cells provoked the activation of Wnt pathway by functional transfection assays. Furthermore, we showed that the protein expression of active beta-catenin was increased in NF1-silenced cell lines. Wnt pathway activation was strongly associated to both cancer stem cell reservoir and Schwann-mesenchymal transition. CONCLUSION: We highlighted the implication of Wnt pathway in NF1-associated tumorigenesis.

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[69]

TÍTULO / TITLE: - The carboxyl terminus and pore-forming domain properties specific to Cx37 are necessary for Cx37 mediated suppression of insulinoma cell proliferation.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Am J Physiol Cell Physiol. 2013 Oct 16.

●● Enlace al texto completo (gratuito o de pago) 1152/ajpcell.00159.2013

AUTORES / AUTHORS: - Nelson TK; Sorgen PL; Burt JM

INSTITUCIÓN / INSTITUTION: - University of Arizona.

RESUMEN / SUMMARY: - Connexin 37 (Cx37) suppresses cell proliferation when expressed in rat insulinoma (Rin) cells, an effect also manifest in vivo during vascular development and in response to tissue injury. Mutant forms of Cx37 with non-functional channels but normally localized, wild-type carboxyl termini are not growth suppressive. Here we determined whether the carboxyl terminal (CT) domain is required for Cx37-mediated growth suppression and whether the Cx37 pore-forming domain can be replaced with the Cx43 pore-forming domain and still retain growth suppressive properties. We show that despite forming functional gap junction channels and hemichannels, Cx37 with residues subsequent to 273 replaced with a V5-epitope tag (Cx37-273tr*V5) had no effect on the proliferation of Rin cells, did not facilitate G1 cell cycle arrest with serum deprivation, and did not prolong cell cycle time comparably to the wild-type protein. The chimera Cx43*CT37, comprising the pore forming domain of Cx43 and CT of Cx37, also did not suppress proliferation, despite forming functional gap junctions with a permselective profile similar to wild-type Cx37. Differences in channel behavior of both Cx37-273tr*V5 and Cx43*CT37 relative to their wild-type counterparts and failure of the Cx37-CT to interact as the Cx43-CT does with the Cx43 cytoplasmic loop suggest that the Cx37-CT and pore-forming domains are both essential to growth suppression by Cx37.

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[70]

TÍTULO / TITLE: - Endoscopic resection for duodenal carcinoid tumors: A multicenter, retrospective study.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - J Gastroenterol Hepatol. 2013 Oct 3. doi: 10.1111/jgh.12390.
●● Enlace al texto completo (gratuito o de pago) 1111/jgh.12390
AUTORES / AUTHORS: - Kim GH; Kim JI; Jeon SW; Moon JS; Chung IK; Jee SR; Kim HU; Seo GS; Baik GH; Lee YC
INSTITUCIÓN / INSTITUTION: - Department of Internal Medicine, Pusan National University School of Medicine and Biomedical Research Institute, Pusan National University Hospital, Busan.
RESUMEN / SUMMARY: - BACKGROUND AND AIM: Gastrointestinal carcinoid tumors <10 mm in diameter and limited to the submucosal layer demonstrate a low frequency of lymph node and distant metastasis, and are suitable for endoscopic treatment. The aim of this study was to assess the efficacy, safety, and long-term prognosis of endoscopic resections for the treatment of duodenal carcinoid tumors. METHODS: This study included a total of 41 duodenal carcinoid tumors in 38 patients between January 2006 and December 2011. The indications for endoscopic resection were lesions </=10 mm in diameter, confined to the submucosal layer, and without lymph node or distant metastasis. Endoscopic resection was accomplished using endoscopic mucosal resection (EMR), EMR with a ligation device (EMR-L), EMR after circumferential precutting (EMR-P), or endoscopic submucosal dissection (ESD). RESULTS: EMR was performed in 18 tumors, EMR-L in 16, EMR-P in 3, and ESD in 4. En bloc resection was performed in 39 tumors (95%), and endoscopic complete resection was achieved in 40 (98%); pathologic complete resection was achieved in 17 tumors (41%). The endoscopic complete resection rate did not differ according to the resection method, but the pathologic complete resection rate was higher for ESD than for EMR and EMR-L. Intraprocedural bleeding was noted in 5 cases, with no occurrence of perforation. Recurrence was not observed during the mean follow-up period of 17 months (range, 1-53 months). CONCLUSION: Endoscopic resection appears to be a safe and effective treatment for duodenal carcinoid tumors measuring </=10 mm in diameter and confined to the submucosal layer.

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[71]
TÍTULO / TITLE: - When to worry about incidental renal and adrenal masses.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - J Fam Pract. 2013 Sep;62(9):476-83.
AUTORES / AUTHORS: - Higgins JC; Arnold MJ
INSTITUCIÓN / INSTITUTION: - Family Medicine Department, Naval Hospital, Jacksonville, FL, USA. Email: James.Higgins@med.navy.mil.
RESUMEN / SUMMARY: - Greater use of imaging has led to a corresponding rise in the detection of renal and adrenal incidentalomas-and left many primary care physicians unsure of what to do about the masses they've found.

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[72]
TÍTULO / TITLE: - Clinical Suspicion of Bilateral Carotid Body Paraganglioma and an Unexpected Histologic Diagnosis.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Ann Vasc Surg. 2013 Oct 3. pii: S0890-5096(13)00347-6. doi: 10.1016/j.avsg.2013.05.005.

●● Enlace al texto completo (gratuito o de pago) 1016/j.avsg.2013.05.005

AUTORES / AUTHORS: - Bozzani A; Arici V; Ragni F; Sagrada PF
INSTITUCIÓN / INSTITUTION: - Division of Vascular Surgery, Foundation I.R.C.C.S. Policlinico San Matteo, Pavia, Italy. Electronic address: a.bozzani@smatteo.pv.it.
RESUMEN / SUMMARY: - Carotid body tumor (CBT) is the most common of the head and neck paragangliomas (PGLs). Conversely, synovial sarcomas are usually located around knee and ankle joint and rare variants occur in the oral cavity. A 68-year-old man presented with a left voluminous painless cervical mass. The diagnosis of CBT of type III Shamblin was suspected. The cervical mass was removed en bloc. Unexpectedly, pathologic examination showed monophasic synovial sarcoma. Excision of PGLs remains the therapy of choice, especially to make a correct histologic diagnosis.

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[73]
TÍTULO / TITLE: - Illuminating somatostatin analog action at neuroendocrine tumor receptors.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Trends Pharmacol Sci. 2013 Dec;34(12):676-88. doi: 10.1016/j.tips.2013.10.001. Epub 2013 Oct 31.

●● Enlace al texto completo (gratuito o de pago) 1016/j.tips.2013.10.001

AUTORES / AUTHORS: - Reubi JC; Schonbrunn A
INSTITUCIÓN / INSTITUTION: - Cell Biology and Experimental Cancer Research, Institute of Pathology, University of Berne, Berne, Switzerland. Electronic address: reubi@pathology.unibe.ch.
RESUMEN / SUMMARY: - Somatostatin analogs for the diagnosis and therapy of neuroendocrine tumors (NETs) have been used in clinical applications for more than two decades. Five somatostatin receptor subtypes have been identified and molecular mechanisms of somatostatin receptor signaling and regulation have been elucidated. These advances increased understanding of the biological role of each somatostatin receptor subtype, their distribution in NETs, as well as agonist-specific regulation of receptor signaling, internalization, and phosphorylation, particularly for the sst2 receptor subtype, which is the primary target of current somatostatin analog therapy for NETs. Various hypotheses exist to explain differences in patient responsiveness to somatostatin analog inhibition of tumor secretion and growth as well as differences in the development of tumor resistance to therapy. In addition, we now have a better understanding of the action of both first generation (octreotide, lanreotide, Octreoscan) and second generation (pasireotide) FDA-approved somatostatin analogs, including the biased agonistic character of some agonists. The increased understanding of somatostatin receptor pharmacology provides new opportunities to design more sophisticated assays to aid the future development of somatostatin analogs with increased efficacy.

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[74]

TÍTULO / TITLE: - Changes in signaling pathways induced by vandetanib in a human medullary thyroid carcinoma model, as analyzed by Reverse Phase Protein Array.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Thyroid. 2013 Nov 20.
●● Enlace al texto completo (gratuito o de pago) 1089/thy.2013.0514
AUTORES / AUTHORS: - Broutin S; Commo F; De Koning L; Marty-Prouvost B; Lacroix L; Talbot M; Caillou B; Dubois T; Ryan AJ; Dupuy C; Schlumberger M; Bidart JM
INSTITUCIÓN / INSTITUTION: - Institut Gustave-Roussy, Departement de Biologie et Pathologie Medicales, 114 rue Edouard Vaillant, Villejuif, France, 94805, 0033142114057, 0031142114401 ; broutin@igr.fr.
RESUMEN / SUMMARY: - Background: Medullary thyroid carcinoma (MTC) is a rare tumor that is caused by activating mutations in the proto-oncogene RET. Vandetanib, a tyrosine-kinase inhibitor, has been recently approved to treat adult patients with metastatic MTC. The aim of this study was to investigate changes in signaling pathways induced by vandetanib treatment in preclinical MTC models, using the reverse-phase protein array method (RPPA). Methods: The human TT cell line was used to assess in vitro and in vivo activity of vandetanib. Protein extracts from TT cells or TT xenografted mice, treated by increasing concentrations of vandetanib for different periods of time, were probed with a set of 12 antibodies representing major signaling pathways, using RPPA. Results were validated using two distinct protein detection methods, western immunoblotting and immunohistochemistry. Results: Vandetanib displays antiproliferative and antiangiogenic activities and inhibits RET auto-phosphorylation. The MAPK and AKT pathways were the two major signaling pathways inhibited by vandetanib. Interestingly, phosphorylated levels of NFkappaB-p65 were significantly increased by vandetanib. Comparable results were obtained in both the in vitro and in vivo approaches as well as for the protein detection methods. However, some discrepancies were observed between RPPA and western immunoblotting, possibly due to lack of specificity of the primary antibodies used. Conclusions: Overall, our results confirmed the interest of RPPA for screening global changes induced in signaling pathways by kinase inhibitors. MAPK and AKT were identified as the main pathways involved in vandetanib response in MTC models. Our results also suggest alternative routes for controlling the disease and provide a rationale for the development of therapeutic combinations based on the comprehensive identification of molecular events induced by inhibitors.

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[75]
TÍTULO / TITLE: - Sporadic nonfunctioning pancreatic neuroendocrine tumors: Prognostic significance of incidental diagnosis.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Surgery. 2013 Nov 12. pii: S0039-6060(13)00465-0. doi: 10.1016/j.surg.2013.08.007.
●● Enlace al texto completo (gratuito o de pago) 1016/j.surg.2013.08.007
AUTORES / AUTHORS: - Birnbaum DJ; Gaujoux S; Cherif R; Dokmak S; Fuks D; Couvelard A; Vullierme MP; Ronot M; Ruszniewski P; Belghiti J; Sauvanet A
INSTITUCIÓN / INSTITUTION: - Department of Hepato-Pancreato-Biliary Surgery - Pole des Maladies de l'Appareil Digestif (PMAD), AP-HP, hopital Beaujon, Clichy, France.

RESUMEN / SUMMARY: - BACKGROUND: Sporadic nonfunctioning pancreatic neuroendocrine tumors (NF-PNETs) are increasingly diagnosed as incidentalomas, and their resection is usually recommended. The prognostic significance of this diagnosis feature is poorly studied, and management of these tumors remains controversial. Clinical, pathologic characteristics and outcome of resected incidentally diagnosed NF-PNET (Inc) were compared with resected symptomatic NF-PNET (Symp) to better assess their biologic behavior and tailor their management. METHODS: From 1994 to 2010, 108 patients underwent resection for sporadic nonmetastatic NF-PNET. Diagnosis was considered as incidental in patients with no abdominal symptoms or symptoms unlikely to be related to tumor mass. Patients with Inc were compared with patients with Symp, regarding demographics, postoperative course, pathology, and disease-free survival (DFS). RESULTS: Of the 108 patients, 65 (61%) had incidentally diagnosed tumors. Pancreas-sparing pancreatectomies (enucleation/central pancreatectomy) were performed more frequently in Inc (62% vs 30%, P = .001). Inc tumors were more frequently <20 mm (65% vs 42%, P = .019), staged T1 (62% vs 33%, P = .0001), node negative (85% vs 60%; P = .005), and grade 1 (66% vs 33%, P = .0001). One postoperative death occurred in the Inc group, and postoperative morbidity was similar between the two groups (60% vs 65%, P = .59). DFS was substantially better in the Inc group (5-year DFS = 92% vs 82%, P = .0016). CONCLUSION: Incidentally diagnosed NF-PNETs are associated with less aggressive features compared with symptomatic lesions but cannot always be considered to be benign. Operative resection remains recommended for most. Incidentally diagnosed NF-PNET may be good candidates for pancreas-sparing pancreatectomies.

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[76]

TÍTULO / TITLE: - Clinical value of technetium-99m-labeled octreotide scintigraphy in local recurrent or metastatic medullary thyroid cancers: a comparison of lesions with 18F-FDG-PET and MIBI images.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Nucl Med Commun. 2013 Dec;34(12):1190-5. doi: 10.1097/MNM.0000000000000006.

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

1097/MNM.0000000000000006

AUTORES / AUTHORS: - Sager S; Kabasakal L; Ocak M; Maecke H; Uslu L; Halac M; Asa S; Sager G; Onsel C; Kanmaz B

INSTITUCIÓN / INSTITUTION: - aDepartment of Nuclear Medicine, Cerrahpasa Medical Faculty, Istanbul University bDepartment of Pediatrics, Fatih Sultan Mehmet Education and Research Hospital, Istanbul, Turkey cDepartment of Nuclear Medicine, University Hospital Freiburg, Freiburg, Germany.

RESUMEN / SUMMARY: - AIM: Various studies have been conducted for determining the most optimal method for the early diagnosis of local recurrent or distant metastatic thyroid cancers. The aim of this study was to evaluate the clinical utility of technetium-99m (Tc-99m)-labeled octreotide derivatives in the detection of recurrence or distant metastases in medullary thyroid cancer patients and to compare the lesions with those detected using F-fluorodeoxyglucose (F-FDG)-PET and Tc-99m MIBI studies in the same patient group. PATIENTS AND METHODS: Sixteen medullary thyroid cancer patients [two male and 14 female; mean age 52.0+/-14.1 years (range 13-72 years)]

were included in this study. All patients underwent a whole-body scan 1 and 4 h after injection with octreotide derivatives and single photon emission computed tomography images were taken of the sites suspicious for metastasis. The lesions seen in Tc-99m HYNIC octreotide studies were compared with those seen in F-FDG-PET and Tc-99m MIBI studies. RESULTS: Among the Tc-99m-labeled octreotide scintigraphy studies, nine were evaluated as true positive (56.2%) and one was evaluated as false positive (6.2%); six were false negative (37.5%). In 16 patients, the total number of lesions seen on octreotide scintigraphy was 21. Thirteen of the 16 patients underwent F-FDG-PET imaging. Of the 13 patients studied, 10 showed true-positive (76.9%) and three showed false-negative (23.1%) results. The total number of lesions seen on F-FDG-PET was 23. The Tc-99m MIBI study yielded positive results in seven of 16 patients (43.7%) and negative results in nine patients (56.3%). The total number of lesions on Tc-99m MIBI was 12. CONCLUSION: The Tc-99m-labeled somatostatin receptor scintigraphy analogs HYNIC-tyrosine octreotide and HYNIC-TATE are useful imaging alternatives in somatostatin receptor-expressing thyroid cancers. Radiolabeling using these analogs is easy and they are readily available for routine use.

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[77]

TÍTULO / TITLE: - Evaluation of Ki-67 index in EUS-FNA specimens for the assessment of malignancy risk in pancreatic neuroendocrine tumors.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Endoscopy. 2013 Nov 11.

●● Enlace al texto completo (gratuito o de pago) 1055/s-0033-1344958

AUTORES / AUTHORS: - Hasegawa T; Yamao K; Hijioka S; Bhatia V; Mizuno N; Hara K; Imaoka H; Niwa Y; Tajika M; Kondo S; Tanaka T; Shimizu Y; Kinoshita T; Kohsaki T; Nishimori I; Iwasaki S; Saibara T; Hosoda W; Yatabe Y

INSTITUCIÓN / INSTITUTION: - Department of Gastroenterology, Aichi Cancer Center Hospital, Nagoya, Japan.

RESUMEN / SUMMARY: - Background and study aim: Malignancy in pancreatic neuroendocrine tumors (PNETs) is graded by assessing the resected specimens according to the World Health Organization (WHO) 2010 criteria. The feasibility of such grading using endoscopic ultrasound-guided fine-needle aspiration (EUS-FNA) specimens remains unclear. The aim of this study was to ascertain the optimal method of measuring the Ki-67 index in EUS-FNA specimens, using resected specimens as the criterion standard. Patients and methods: A total of 58 consecutive patients diagnosed with PNETs between March 1998 and May 2011 were included. The study measured intratumoral Ki-67 index heterogeneity, concordance rates of PNET grading by EUS-FNA with grade of the resected tumor, optimal method of measuring the Ki-67 index in EUS-FNA specimens, and survival analysis based on EUS-FNA specimen grading. Results: Intratumoral dispersion of Ki-67 index in resected specimens was 0.033 for Grade 1 and 0.782 for Grade 2 tumors ($P < 0.001$). Concordance rates for WHO classification between EUS-FNA and resected specimens were 74.0 % using the mean Ki-67 index in EUS-FNA specimens and 77.8 % using the highest Ki-67 index. The concordance rate rose to 90 % when EUS-FNA samples with less than 2000 tumor cells were excluded (26 % of EUS-FNA cases). The Kaplan-Meier survival curves were significantly stratified by the EUS-FNA grading of PNETs with 5-year survival rates of 100 %, 58.3 %, and 0 %, for Grade 1, Grade 2, and neuroendocrine carcinoma (NEC)

tumors, respectively. Conclusions: Grading of PNETs by the highest Ki-67 index in EUS-FNA specimens with adequate cellularity has a high concordance with grading of resected specimens, and can predict long term patient survival with high accuracy.

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[78]
TÍTULO / TITLE: - Paragangliomas: update on differential diagnostic considerations, composite tumors, and recent genetic developments.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Semin Diagn Pathol. 2013 Aug;30(3):207-23. doi: 10.1053/j.semdp.2013.06.006.
●● Enlace al texto completo (gratuito o de pago) 1053/j.semdp.2013.06.006
AUTORES / AUTHORS: - Papathomas TG; de Krijger RR; Tischler AS
INSTITUCIÓN / INSTITUTION: - Department of Pathology, Josephine Nefkens Institute, Erasmus MC-University Medical Center, P.O. Box 2040, 3000 CA, Rotterdam, The Netherlands. Electronic address: t.papathomas@erasmusmc.nl.
RESUMEN / SUMMARY: - Recent developments in molecular genetics have expanded the spectrum of disorders associated with pheochromocytomas (PCCs) and extra-adrenal paragangliomas (PGLs) and have increased the roles of pathologists in helping to guide patient care. At least 30% of these tumors are now known to be hereditary, and germline mutations of at least 10 genes are known to cause the tumors to develop. Genotype-phenotype correlations have been identified, including differences in tumor distribution, catecholamine production, and risk of metastasis, and types of tumors not previously associated with PCC/PGL are now considered in the spectrum of hereditary disease. Important new findings are that mutations of succinate dehydrogenase genes SDHA, SDHB, SDHC, SDHD, and SDHAF2 (collectively "SDHx") are responsible for a large percentage of hereditary PCC/PGL and that SDHB mutations are strongly correlated with extra-adrenal tumor location, metastasis, and poor prognosis. Further, gastrointestinal stromal tumors and renal tumors are now associated with SDHx mutations. A PCC or PGL caused by any of the hereditary susceptibility genes can present as a solitary, apparently sporadic, tumor, and substantial numbers of patients presenting with apparently sporadic tumors harbor occult germline mutations of susceptibility genes. Current roles of pathologists are differential diagnosis of primary tumors and metastases, identification of clues to occult hereditary disease, and triaging of patients for optimal genetic testing by immunohistochemical staining of tumor tissue for the loss of SDHB and SDHA protein. Diagnostic pitfalls are posed by morphological variants of PCC/PGL, unusual anatomic sites of occurrence, and coexisting neuroendocrine tumors of other types in some hereditary syndromes. These pitfalls can be avoided by judicious use of appropriate immunohistochemical stains. Aside from loss of staining for SDHB, criteria for predicting risk of metastasis are still controversial, and "malignancy" is diagnosed only after metastases have occurred. All PCCs/PGLs are considered to pose some risk of metastasis, and long-term follow-up is advised.

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[79]
TÍTULO / TITLE: - Preoperative protective stenting of the internal carotid artery in the management of complex head and neck paragangliomas: long-term results.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Audiol Neurootol. 2013;18(6):345-52. doi: 10.1159/000354158. Epub 2013 Oct 4.
●● Enlace al texto completo (gratuito o de pago) 1159/000354158
AUTORES / AUTHORS: - Piazza P; Di Lella F; Bacciu A; Di Trapani G; Ait Mimoune H; Sanna M
INSTITUCIÓN / INSTITUTION: - Gruppo Otologico, Piacenza, Italy.
RESUMEN / SUMMARY: - Objectives: To investigate the long-term results of preoperative stenting of the internal carotid artery (ICA) in complex head and neck paragangliomas (HNP) as well as to report on indications and technical details of the procedure. Method: A comprehensive retrospective review of patients affected by HNP, consecutively operated on and preoperatively treated with stenting of the ICA in a quaternary referral skull base center, was performed. Results: Nineteen patients affected by complex HNP were identified, on whom 21 preoperative stenting procedures were performed. The mean follow-up period after stent insertion was 53.8 months; the patients' age ranged from 33 to 56 years. Fourteen patients were affected by tympanojugular paragangliomas, 4 by vagal paragangliomas and 1 by bilateral carotid body tumors. Five patients presented with recurrent tumors, while 7 presented with multiple or bilateral HNP. There were no complications associated with endovascular procedures. Total tumor removal was accomplished in 52.4% of the cases with 1 recurrence. An advanced stage was the main factor conditioning total removal. Clinical control was obtained in 80% of the patients with residual disease. Total tumor removal from and around the ICA was obtained in 95.2% of the cases. Long-term stent patency was evident in 20 of 21 cases. Conclusions: Preoperative stenting of the ICA represents a safe and effective procedure in selected cases, obviating the need for balloon occlusion or bypass procedures and reducing the risk of intraoperative vascular injury. © 2013 S. Karger AG, Basel.

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[80]
TÍTULO / TITLE: - Benign whole body tumor volume is a risk factor for malignant peripheral nerve sheath tumors in neurofibromatosis type 1.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - J Neurooncol. 2013 Oct 29.
●● Enlace al texto completo (gratuito o de pago) 1007/s11060-013-1293-1
AUTORES / AUTHORS: - Nguyen R; Jett K; Harris GJ; Cai W; Friedman JM; Mautner VF
INSTITUCIÓN / INSTITUTION: - Department of Pediatrics, University of Maryland, 22 South Greene St, N5W70, Baltimore, MD, 21201, USA, rosi.nguyen@gmail.com.
RESUMEN / SUMMARY: - The purpose of this study is to determine whether benign whole body tumor volume of plexiform neurofibromas (PNs) is a risk factor for malignant peripheral nerve sheath tumors (MPNST) in individuals with neurofibromatosis type 1 (NF1). Thirty-one NF1 patients with MPNSTs and 62 age- and sex-matched NF1 patients without MPNSTs, who had undergone whole body magnetic resonance imaging (MRI) were analyzed for benign whole body tumor volume. Mann-Whitney U test, Wilcoxon signed ranks test, Fisher's exact test (two-tailed), and logistic regression analysis were used for statistical analysis. Sixteen percent of all patients with MPNST did not have internal PN. The median whole body

benign tumor volume in patients with PN was 352.0 mL among the MPNST group and 3.8 mL in the comparison group ($p < 0.001$). When the patients were stratified by age as younger or older than 30 years (median age of MPNST diagnosis), the median benign whole body tumor volume was 693.0 mL in MPNST patients and 0.0 mL in control patients younger than 30 years ($p < 0.001$). The mean number of PNs in MPNST patients was 2.8 (range 0-13, median 2.0) and 1.4 (range 0-13, median 1.0) in patients without MPNST ($p = 0.001$). The risk of MPNST development increased 0.2 % with each additional mL of benign PN volume (adjusted odds ratio [OR] = 1.002, 95 % confidence interval [CI] 1.001-1.003, $p = 0.005$) and was higher in patients younger than 30 years (adjusted OR = 1.007, 95 % CI 1.002-1.012, $p = 0.003$). Higher numbers of PNs, larger whole body benign tumor volume, and younger age are important risk factors for MPNST. We identified a subgroup of patients with MPNST without internal PN on MRI and the lack of correlation of MPNST development with tumor burden in older patients. These findings may alter our belief that all MPNSTs arise from pre-existing PNs and suggest that surveillance MRI based on clinical suspicion may be warranted in older patients, respectively.

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[81]
TÍTULO / TITLE: - Schwannoma and nerve abscess of leprosy: differential diagnosis.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Lepr Rev. 2013 Jun;84(2):141-4.
AUTORES / AUTHORS: - Lima CM; Da Costa PC; Carneiro L; De Oliveira ML
INSTITUCIÓN / INSTITUTION: - Dermatology Service, Federal University of Rio de Janeiro (HUCFF/UFRJ), Brazil.

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[82]
TÍTULO / TITLE: - Chronic mTOR activation promotes cell survival in Merkel cell carcinoma.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Cancer Lett. 2013 Nov 19. pii: S0304-3835(13)00807-0. doi: 10.1016/j.canlet.2013.11.005.
●● Enlace al texto completo (gratuito o de pago) 1016/j.canlet.2013.11.005
AUTORES / AUTHORS: - Lin Z; McDermott A; Shao L; Kannan A; Morgan M; Stack BC Jr; Moreno M; Davis DA; Cornelius LA; Gao L
INSTITUCIÓN / INSTITUTION: - Department of Dermatology, University of Arkansas for Medical Sciences, Little Rock, AR 72205, United States; Cancer Center Union Hospital, Tongji Medical College, Huazhong University of Science and Technology, Wuhan 430000, China.
RESUMEN / SUMMARY: - Merkel cell carcinoma (MCC) is an aggressive skin cancer with rising incidence. In this study, we demonstrate that mTOR activation and suppressed autophagy is common in MCCs. mTOR inhibition in two primary human MCC cell lines induces autophagy and cell death that is independent of caspase activation but can be attenuated by autophagy inhibition. This is the first study to evaluate mTOR and autophagy in MCC. Our data suggests a potential role of autophagic cell death upon mTOR inhibition and thus uncovers a previously

underappreciated role of mTOR signaling and cell survival, and merits further studies for potential therapeutic targets.

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[83]
TÍTULO / TITLE: - Merkel cell carcinoma: diagnosis, management, and outcomes.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Plast Reconstr Surg. 2013 Nov;132(5):894e-5e. doi: 10.1097/PRS.0b013e3182a4c681.
●● Enlace al texto completo (gratuito o de pago)
1097/PRS.0b013e3182a4c681
AUTORES / AUTHORS: - Koljonen V

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[84]
TÍTULO / TITLE: - Prospective Study to Compare Peri-operative Hemodynamic Alterations following Preparation for Pheochromocytoma Surgery by Phenoxybenzamine or Prazosin.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - World J Surg. 2013 Nov 14.
●● Enlace al texto completo (gratuito o de pago) 1007/s00268-013-2325-x
AUTORES / AUTHORS: - Agrawal R; Mishra SK; Bhatia E; Mishra A; Chand G; Agarwal G; Agarwal A; Verma AK
INSTITUCIÓN / INSTITUTION: - Department of Endocrine Surgery, Sanjay Gandhi Post Graduate Institute of Medical Sciences, Raibareli Road, Lucknow, India.
RESUMEN / SUMMARY: - BACKGROUND: Prospective studies comparing the efficacy of selective versus nonselective alpha blockers for preoperative preparation of pheochromocytoma (PCC) are lacking. In this prospective nonrandomized study, we compared the outcome of preoperative preparation with phenoxybenzamine (PBZ) and prazosin (PRZ) in terms of perioperative hemodynamic alterations. METHODS: The study was conducted at a tertiary referral center from July 2010 to December 2012. Thirty-two patients with PCC underwent operation after adequate preparation with PBZ (n = 15) or PRZ (n = 17). Five pediatric and adolescent patients were excluded because of different hemodynamics in this population. Perioperative monitoring was done for pulse rate (PR) and blood pressure (BP) alterations, occurrence of arrhythmias, and time taken to achieve hemodynamic stability. Groups were compared with the Mann-Whitney test, Student's t test, and the chi 2 test as applicable. RESULTS: Patients in the two groups were similar in age, gender, 24 h urinary metanephrine and normetanephrine levels, and type of procedure. Patients prepared with PRZ had significantly more intraoperative episodes of transient hypertension (systolic BP >/= 160 mmHg) and hypertensive urgency (BP > 180/110 mmHg) (p 0.02, 0.03, respectively). More patients receiving PRZ suffered from transient severe hypertension (SBP >/= 220 mmHg) (p 0.03). The PRZ group also had more median maximum SBP (233 mmHg vs PBZ 181.5 mmHg) (p = 0.01) and lesser median minimum SBP (71 mmHg vs PBZ 78 mmHg) (p 0.03). No significant differences were found between the study groups for changes in PR, postoperative BP alterations, occurrence of arrhythmias, and time taken to achieve hemodynamic stability.

CONCLUSIONS: PBZ was found superior to PRZ in having fewer intraoperative hemodynamic fluctuations.

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[85]
TÍTULO / TITLE: - Neuroendocrine carcinomas: Optimal surgery of peritoneal metastases (and associated intra-abdominal metastases).
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Surgery. 2013 Sep 28. pii: S0039-6060(13)00275-4. doi: 10.1016/j.surg.2013.05.030.
●● Enlace al texto completo (gratuito o de pago) 1016/j.surg.2013.05.030
AUTORES / AUTHORS: - Elias D; David A; Sourrouille I; Honore C; Goere D; Dumont F; Stoclin A; Baudin E
INSTITUCIÓN / INSTITUTION: - Department of Surgery, Gustave Roussy, Cancer Campus, Villejuif, France. Electronic address: elias@igr.fr.
RESUMEN / SUMMARY: - AIM: To report the results of complete cytoreductive surgery (CCRS) of peritoneal metastases from neuroendocrine tumor (NET) and to compare patients treated with or without hyperthermic intraperitoneal chemotherapy (HIPEC). BACKGROUND: Aggressive management of peritoneal metastases from NET (in most of the cases associated with other types of metastases) has not been addressed in the literature, but these metastases affect overall survival. PATIENTS AND METHODS: From 1994 to 2012, 41 patients underwent CCRS, with HIPEC (n = 28) from 1994 to 2007 but without HIPEC (n = 13) from 2008 to 2012. Liver metastases were treated during the same operative procedure in 66% of the patients. RESULTS: Mortality was 2% and morbidity 56%. Overall survival at 5 and 10 years was 69% and 52%, respectively, and disease-free survival at 5 and 10 years was 17% and 6%, respectively. At 5 years, peritoneal metastases and liver metastases recurred in 47% and in 66% of cases, respectively. Overall survival was not different between patients treated with or without HIPEC, but disease-free survival was greater in the HIPEC group (P = .018), mainly because of fewer lung and bone metastases. CONCLUSION: CCRS of peritoneal metastases from a NET is feasible in most of the patients and seems to increase survival rates. We were unable to determine whether adding HIPEC had a positive or a negative impact.

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[86]
TÍTULO / TITLE: - Long-term results of endoscopic resection for type I gastric neuroendocrine tumors.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - J Surg Oncol. 2013 Oct 25. doi: 10.1002/jso.23477.
●● Enlace al texto completo (gratuito o de pago) 1002/jso.23477
AUTORES / AUTHORS: - Uygun A; Kadayifci A; Polat Z; Yilmaz K; Gunal A; Demir H; Bagci S
INSTITUCIÓN / INSTITUTION: - Division of Gastroenterology, Gulhane Military Medical Academy, Ankara, Turkey.
RESUMEN / SUMMARY: - BACKGROUND: A number of different therapies, including endoscopic resection, have been suggested for the treatment of Type 1 gastric neuroendocrine tumors (NETs). The current study aimed to determine the long-term

efficacy of endoscopic resection for Type 1 gastric NETs. METHODS: Twenty-two patients (from 1999 to 2012) with Type 1 gastric NETs were included in the study. All patients were treated with endoscopic resection and received regular followed-up appointments at a tertiary referral center. RESULTS: All patients were initially diagnosed with hypergastrinemia, atrophic gastritis and intestinal metaplasia. Polyps' diameters were >1 cm in 4 patients, and between 0.5 and 1 cm in 18 patients. All detectable lesions were successfully resected. One patient required surgery due to gastric perforation during endoscopic mucosal resection. Recurrence was detected in four patients (18%) and endoscopic resection was performed again. Local or distant metastasis was not observed in any patient during follow-up. Median follow-up time was 7 years, with a maximum of 14 years. Seventeen patients (78%) completed a 5-year follow-up period, and overall disease-free survival rate was 100%. CONCLUSIONS: Long-term follow-ups with 22 patients suggest that endoscopic resection of Type 1 gastric NETs is a safe and effective treatment option with a relatively low recurrence rate. J. Surg. Oncol. © 2013 Wiley Periodicals, Inc.

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[87]
TÍTULO / TITLE: - Thyroid hemiagenesis associated with medullary or papillary carcinoma: Two cases report.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Head Neck. 2013 Sep 30. doi: 10.1002/hed.23501.
●● Enlace al texto completo (gratuito o de pago) 1002/hed.23501
AUTORES / AUTHORS: - Wang J; Gao MM; Song C
INSTITUCIÓN / INSTITUTION: - Department of Head and Neck Surgery, Institute of Minimally Invasive Surgery of Zhejiang University, Sir Run Run Shaw Hospital, Zhejiang University School of Medicine, Hangzhou, Zhejiang, 310016, China.
RESUMEN / SUMMARY: - Background. Thyroid hemiagenesis (TH) is a rare congenital anomaly in which one thyroid lobe fails to develop. Cooccurrence of hemiagenesis and thyroid carcinoma is extremely rare. Here we report two cases of TH with carcinoma. Methods. The first patient was referred with a left thyroid mass and absent right lobe. The frozen section examination considered medullary thyroid carcinoma (MTC), so left thyroid lobectomy plus neck dissection was performed. Another patient was referred with a right thyroid mass and absent left lobe. Fine needle aspiration biopsy was suspicious of papillary carcinoma. The patient underwent right thyroid lobectomy plus neck dissection. Results. The operative findings confirmed hemiagenesis of the right lobe and MTC in the left lobe for the first case, and hemiagenesis of the left lobe and papillary carcinoma in the right lobe for the second case. Conclusions. Our case represents the first reported case of association between TH and MTC. Head Neck, 2013.

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[88]
TÍTULO / TITLE: - Multifocal and microscopic chromophobe renal cell carcinomatous lesions associated with 'capsulomas' without FCLN gene abnormality.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Pathol Int. 2013 Oct;63(10):510-5. doi: 10.1111/pin.12099. Epub 2013 Oct 18.

●● Enlace al texto completo (gratuito o de pago) 1111/pin.12099

AUTORES / AUTHORS: - Sugimoto K; Takasawa A; Ichimiya S; Murata M; Kimura H; Aoyama T; Gille JJ; Kuroda N; Shimizu H; Hasegawa T; Sawada N; Furuya M; Nagashima Y

INSTITUCIÓN / INSTITUTION: - Department of Pathology, Sapporo Medical University School of Medicine, Sapporo, Japan.

RESUMEN / SUMMARY: - Chromophobe renal cell carcinoma (RCC) accounts for approximately 5% of renal epithelial neoplasms. Multiple and/or bilateral chromophobe RCCs in an individual are generally rare but frequently occur in patients with Birt-Hogg-Dube syndrome (BHDS) and in patients with tuberous sclerosis complex (TSC). The responsible genes in both BHDS and TSC act as tumor suppressors. Therefore, it seems that some genetic backgrounds are required for the generation and progression of multiple chromophobe RCCs. Here, we report a case of multiple and bilateral chromophobe RCCs along with several small-sized capsular angiomyolipomas known as 'capsulomas' in a 39-year-old woman who had neither a particular medical history nor specific gene mutation. There has been no report of sporadic multiple chromophobe RCCs and 'capsulomas' developing in a patient without genetic features, having potential for novel genetic variation.

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[89]

TÍTULO / TITLE: - Primary cutaneous neuroendocrine carcinoma, Merkel cell carcinoma. Case series 1991-2012.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Acta Otorrinolaringol Esp. 2013 November - December;64(6):396-402. doi: 10.1016/j.otorri.2013.06.003. Epub 2013 Oct 4.

●● Enlace al texto completo (gratuito o de pago) 1016/j.otorri.2013.06.003

AUTORES / AUTHORS: - Campillo R; Gil-Carcedo E; Alonso D; Vallejo LA; Onate JM; Gil-Carcedo LM

INSTITUCIÓN / INSTITUTION: - Servicio de Cirugia Plastica, Hospital Universitario Rio Hortega, Universidad de Valladolid, Valladolid, España.

RESUMEN / SUMMARY: - INTRODUCTION AND OBJECTIVES: Merkel cell carcinoma was first described by Toker in 1972. It is an uncommon, primary neuroendocrine skin carcinoma which appears in the dermoepidermic area, grows fast, is very aggressive and has a poor prognosis. The aim of this work is to highlight the importance of this tumour, which develops mainly in the skin of the head and neck area, and whose prevalence has increased in recent years. MATERIAL AND METHOD: We gathered data on 16 patients suffering cutaneous neuroendocrine carcinoma treated at our hospital between September 12, 1991 and July 13, 2012. We indicated the age and gender of patients. We described the area where the tumour was located, indicating the size in millimetres, according to the major axis of the lesion. RESULTS: Most of the patients studied were over 70 years old, except for one who was 55. The highest frequency of cases appeared among patients aged over 80 years. In the cases studied, when the tumour appeared in the head and neck region (10/16), its location could be nasal-lateronasal, cheek-malar, upper eyelid, frontal or mandibular. The major axis of the lesion ranged between 7 and 35mm. Unlike with epidermoid or basocellular carcinomas, recurrence and ganglionar metastases were common. Immunohistochemical (CK20) tests are essential for a correct diagnosis. Treatment is

usually surgical and occasionally followed by radiotherapy and chemotherapy. CONCLUSION: This carcinoma is not a very common skin tumour. It appears in old age, in the head and neck region in 50% of cases and often leads to exitus.

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[90]

TÍTULO / TITLE: - Diffusion-weighted MRI: usefulness for differentiating intrapancreatic accessory spleen and small hypervascular neuroendocrine tumor of the pancreas.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Acta Radiol. 2013 Nov 20.

●● Enlace al texto completo (gratuito o de pago) 1177/0284185113513760

AUTORES / AUTHORS: - Kang BK; Kim JH; Byun JH; Lee SS; Kim HJ; Kim SY; Lee MG

INSTITUCIÓN / INSTITUTION: - Department of Radiology and Research Institute of Radiology, University of Ulsan College of Medicine, Asan Medical Center, Seoul, Republic of Korea.

RESUMEN / SUMMARY: - BACKGROUND: Image findings of intrapancreatic accessory spleen (IPAS) can closely resemble those of neuroendocrine tumor (NET) of the pancreas. PURPOSE: To investigate the usefulness of diffusion-weighted imaging (DWI) for differentiating IPAS from small (</=3 cm) hypervascular NET of the pancreas. MATERIAL AND METHODS: The visually assessed signal intensity of pancreatic lesions compared with the spleen on DWI (b value of 1000 s/mm2) and the apparent diffusion coefficient (ADC) values were compared in 25 patients with IPAS and 31 patients with small hypervascular NET. Two blinded radiologists independently rated their confidence in differentiating the two conditions and compared the diagnostic performance of contrast-enhanced magnetic resonance imaging (CE-MRI) alone with that of combined CE-MRI and DWI. RESULTS: The isointensity of the pancreatic lesions compared with the spleen on DWI was more frequently observed in IPAS than in NET (92% vs. 12.9%, P < 0.001). The mean ADC value was significantly lower in IPAS than in NET (0.90 x 10-3 mm2/s vs. 1.44 x 10-3 mm2/s, P < 0.001). The sensitivity and specificity of ADC quantification for differentiating the two conditions when using 1.07 x 10-3 mm2/s as the cut-off value were 96% and 93.5%, respectively. For both readers, the area under the receiver operating characteristic curve and accuracy in differentiating the two conditions of combined CE-MRI and DWI were significantly greater than those of CE-MRI alone (P </= 0.039). CONCLUSION: Visual assessment of DWI and ADC quantification were useful in differentiating IPAS from small hypervascular NET of the pancreas.

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[91]

TÍTULO / TITLE: - Transient interhemispheric disconnection in a case of insulinoma-induced hypoglycemic encephalopathy.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - J Neurol Sci. 2013 Dec 15;335(1-2):233-7. doi: 10.1016/j.jns.2013.09.025. Epub 2013 Sep 25.

●● Enlace al texto completo (gratuito o de pago) 1016/j.jns.2013.09.025

AUTORES / AUTHORS: - Yamashita C; Shigeto H; Maeda N; Kawaguchi M; Uryu M; Motomura S; Kira J

INSTITUCIÓN / INSTITUTION: - Department of Neurology, Neurological Institute, Graduate School of Medical Sciences, Kyushu University, Japan.
RESUMEN / SUMMARY: - We report a case of a 22-year-old male who was transferred to our hospital in a comatose state following successive seizures. Low blood glucose had been detected upon his arrival at the previous hospital. He became responsive 12days after the onset of coma. Upon regaining consciousness he exhibited severe dysarthria and several interhemispheric disconnection signs such as intermanual conflict, left-hand dysgraphia, left hemispatial neglect confined to the right hand, impaired interhemispheric transfer, and unilateral constructional apraxia of the right hand. Brain MRI disclosed T2-weighted and diffusion-weighted hyperintense lesions with reduced apparent diffusion coefficients in the bilateral centrum semiovale, splenium of the corpus callosum, right posterior limb of the internal capsule, and bilateral middle cerebellar peduncles. As the MRI findings vanished, his interhemispheric disconnection signs gradually resolved. Abdominal imaging studies revealed a pancreatic tumor, which was later endocrinologically diagnosed as an insulinoma. This is an extremely rare report of interhemispheric disconnection signs due to hypoglycemic encephalopathy. The lesions in the bilateral centrum semiovale likely contributed to the interhemispheric disconnection signs.

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[92]
TÍTULO / TITLE: - Differentiation of oncocytoma from chromophobe renal cell carcinoma (RCC): can novel molecular biomarkers help solve an old problem?
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - J Clin Pathol. 2013 Oct 29. doi: 10.1136/jclinpath-2013-201895.
●● Enlace al texto completo (gratuito o de pago) 1136/jclinpath-2013-201895
AUTORES / AUTHORS: - Ng KL; Rajandram R; Morais C; Yap NY; Samaratunga H; Gobe GC; Wood ST
INSTITUCIÓN / INSTITUTION: - Centre for Kidney Disease Research, School of Medicine, The University of Queensland, Translational Research Institute, Brisbane, Australia.
RESUMEN / SUMMARY: - Standard treatment of renal neoplasms remains surgical resection, and nephrectomy for localised renal cell carcinoma (RCC) still has the best chance of cure with excellent long-term results. For smaller renal masses, especially stage T1a tumours less than 4 cm, nephron-sparing surgery is often employed. However, small incidentally detected renal masses pose an important diagnostic dilemma as a proportion of them may be benign and could be managed conservatively. Renal oncocytoma is one such lesion that may pose little risk to a patient if managed with routine surveillance rather than surgery. Additionally, lower-risk RCC, such as small chromophobe RCC, may be managed in a similar way, although with more caution than the renal oncocytomas (RO). The ability to differentiate ROs from chromophobe RCCs, and from other RCCs with a greater chance of metastasis, would guide the physician and patient towards the most appropriate management, whether nephron-sparing surgical resection or conservative surveillance. Consistent accurate diagnosis of ROs is likely to remain elusive until modern molecular biomarkers are identified and applied routinely. This review focuses on the differentiation of renal oncocytomas and chromophobe RCCs. It summarises the history, epidemiology and

clinical presentation of the renal neoplasms, explains the diagnostic dilemma, and describes the value, or not, of current molecular markers that are in development to assist in diagnosis of the renal neoplasms.

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[93]

TÍTULO / TITLE: - Predicting aggressive behavior in nonfunctioning pancreatic neuroendocrine tumors.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Surgery. 2013 Oct;154(4):785-91; discussion 791-3. doi: 10.1016/j.surg.2013.07.004.

●● Enlace al texto completo (gratuito o de pago) 1016/j.surg.2013.07.004

AUTORES / AUTHORS: - Cherenfant J; Stocker SJ; Gage MK; Du H; Thurow TA; Odeleye M; Schimpke SW; Kaul KL; Hall CR; Lamzabi I; Gattuso P; Winchester DJ; Marsh RW; Roggin KK; Bentrem DJ; Baker MS; Prinz RA; Talamonti MS

INSTITUCIÓN / INSTITUTION: - NorthShore University HealthSystems, Evanston, IL. Electronic address: jovenel1@yahoo.com.

RESUMEN / SUMMARY: - PURPOSE: The biologic potential of nonfunctioning pancreatic neuroendocrine tumors (PNETs) is highly variable and difficult to predict before resection. This study was conducted to identify clinical and pathologic factors associated with malignant behavior and death in patients diagnosed with PNETs. METHODS: We used International Classification of Diseases 9th edition codes to identify patients who underwent pancreatectomy for PNETs from 1998 to 2011 in the databases of 4 institutions. Functioning PNETs were excluded. Multivariate regression Cox proportional models were constructed to identify clinical and pathologic factors associated with distant metastasis and survival. RESULTS: The study included 128 patients-57 females and 71 males. The age (mean +/- standard deviation) was 55 +/- 14 years. The body mass index was 28 +/- 5 kg/m(2). Eighty-nine (70%) patients presented with symptoms, and 39 (30%) had tumors discovered incidentally. The tumor size was 3.3 +/- 2 cm with 56 (44%) of the tumors measuring </=2 cm. Seventy-three (57%) patients had grade 1 histology tumors, 37 (29%) had grade 2, and 18 (14%) had grade 3. Peripancreatic lymph node involvement was present in 31 patients (24%), absent in 75 (59%), and unknown in 22 (17%). Distant metastasis occurred in 18 patients (14%). There were 12 deaths, including 1 perioperative, 8 disease related, and 3 of unknown cause. With a median follow-up of 33 months, the overall 5-year survival was 75%. Multivariate Cox regression analysis identified age >55 (hazard ratio [HR], 5.89; 95% confidence interval [CI], 1.64-20.58), grade 3 histology (HR, 6.08; 95% CI, 1.32-30.2), and distant metastasis (HR, 8.79; 95% CI, 2.67-28.9) as risk factors associated with death (P < .05). Gender, race, body mass index, clinical symptoms, lymphovascular and perineural invasion, and tumor size were not related to metastasis or survival (P > .05). Three patients with tumors </=2 cm developed distant metastasis resulting in 2 disease-related deaths. CONCLUSION: Age >55 years, grade 3 histology, and distant metastasis predict a greater risk of death from nonfunctioning PNETs. Resection or short-term surveillance should be considered regardless of tumor size.

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[94]

TÍTULO / TITLE: - Surgical resection of carotid body paragangliomas: 10 years of experience.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Am J Surg. 2013 Oct 10. pii: S0002-9610(13)00497-2. doi: 10.1016/j.amjsurg.2013.06.002.
●● Enlace al texto completo (gratuito o de pago) 1016/j.amjsurg.2013.06.002
AUTORES / AUTHORS: - Amato B; Bianco T; Compagna R; Siano M; Esposito G; Buffone G; Serra R; de Franciscis S
INSTITUCIÓN / INSTITUTION: - Department of General, Geriatric, Oncologic Surgery and Advanced Technologies.
RESUMEN / SUMMARY: - BACKGROUND: Carotid body tumors (CBTs) are relatively rare neoplasms, and even if they are considered predominantly benign, there is an indication for early surgical removal. The objective of this study was to conduct a review of the surgical management of CBTs. METHODS: A retrospective study identified 34 cases (12 men and 19 women) of tumors in patients who had undergone surgical resection of pathologically confirmed CBTs over a period of 10 years from 2001 to 2011 in 2 academic departments of general surgery in Italy. RESULTS: In our series, 10 CBTs (31%) were Shamblin class I, 13 (41%) were class II, and 9 tumors (27%) were class III. Two patients (6%) had transient cerebral ischemia immediately after operation. One patient (3%) died of postoperative cerebral ischemia after surgery for internal carotid artery thrombosis. CONCLUSIONS: The experience of this casuistry shows that the procedure is relatively low risk for Shamblin I and II classes, whereas there is an increasing risk of neurovascular complications for Shamblin III class.

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[95]
TÍTULO / TITLE: - BAI3, CDX2 and VIL1: a panel of three antibodies to distinguish small cell from large cell neuroendocrine lung carcinomas.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Histopathology. 2013 Nov 25. doi: 10.1111/his.12278.
●● Enlace al texto completo (gratuito o de pago) 1111/his.12278
AUTORES / AUTHORS: - Bari MF; Brown H; Nicholson AG; Kerr KM; Gosney JR; Wallace WA; Soomro I; Muller S; Peat D; Moore JD; Ward LA; Freidin MB; Lim E; Vatish M; Snead DR
INSTITUCIÓN / INSTITUTION: - Department of Pathology, Dow International Medical College, Karachi, Pakistan; Department of Pathology, University Hospitals Coventry and Warwickshire NHS Trust, Coventry, UK; Divisions of Reproduction and Metabolic and Vascular Health, Warwick Medical School, Coventry, UK.
RESUMEN / SUMMARY: - AIMS: Discriminating small-cell lung carcinoma (SCLC) from large-cell neuroendocrine carcinoma (LCNEC) rests on morphological criteria, and reproducibility has been shown to be poor. We aimed to identify immunohistochemical markers to assist this diagnosis. METHODS AND RESULTS: Gene expression profiling on laser captured frozen tumour samples from eight SCLC and eight LCNEC tumours identified a total of 888 differentially expressed genes (DEGs), 23 of which were validated by qRT-PCR. Antibodies to four selected gene products were then evaluated as immunohistochemical markers on a cohort of 173 formalin-fixed paraffin-embedded (FFPE) SCLC/LCNEC tumour samples, including 26 indeterminate tumours without a consensus diagnosis. Three markers, CDX2, VIL1 and BAI3, gave significantly different

results in the two tumour types (P < 0.0001): CDX2 and VIL1 in combination (either marker positive) showed sensitivity and specificity of 81% for LCNEC while BAI3 showed 89% sensitivity and 75% specificity for SCLC. Of the 26 indeterminate tumours 15 (58%) showed an immunophenotype suggesting either SCLC or LCNEC, eight (31%) showed staining of both tumour types, and three (11%) were negative for all markers. CONCLUSION: A panel of three markers, BAI3, CDX2 and VIL1, is a useful adjunct in the diagnosis of these tumour types.

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[96]

TÍTULO / TITLE: - Primary Neuroendocrine Carcinoma in Oral Cavity: Two Case Reports and Review of the Literature.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - J Oral Maxillofac Surg. 2013 Nov 8. pii: S0278-2391(13)01108-7. doi: 10.1016/j.joms.2013.08.020.

●● Enlace al texto completo (gratuito o de pago) 1016/j.joms.2013.08.020

AUTORES / AUTHORS: - Wu BZ; Gao Y; Yi B

INSTITUCIÓN / INSTITUTION: - Resident, Department of Oral and Maxillofacial Surgery, Peking University School and Hospital of Stomatology, Beijing, China.

RESUMEN / SUMMARY: - Neuroendocrine carcinoma (NEC) is a tumor that occurs in different locations, particularly the lungs and larynx. The oral cavity is a rare site for a primary NEC. This report describes 2 cases of primary NEC in the oral cavity. Case 1 occurred in the anterior mandibular gingiva in a 25-year-old woman and presented with a special histologic appearance. This patient showed no evidence of recurrence 13 months after marginal resection of the anterior mandible. Case 2 was a primary NEC with some foci of squamous cell differentiation arising in the right buccal region in a 38-year-old woman. This patient showed no evidence of disease 8 months after tumor resection and postoperative iodine-125 brachytherapy. To the best of the authors' knowledge, case 1 is the youngest patient with NEC reported in the oral cavity to date in the English-language literature, and case 2 is the first report of a primary NEC in the buccal region.

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[97]

TÍTULO / TITLE: - Intestinal neurilemmoma.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Cir Esp. 2013 Oct 21. pii: S0009-739X(13)00278-9. doi: 10.1016/j.ciresp.2013.04.023.

●● Enlace al texto completo (gratuito o de pago) 1016/j.ciresp.2013.04.023

AUTORES / AUTHORS: - Borges Sandrino RS; Ramon Musibay E; Escobar Rojas I; Santiesteban Pupo WE

INSTITUCIÓN / INSTITUTION: - Servicio de Cirugia General, Hospital General Docente Dr. Carlos J. Finlay, La Habana, Cuba. Electronic address: rborges@infomed.sld.cu.

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[98]

TÍTULO / TITLE: - SPECT and 18F-FDG PET/CT Imaging of Multiple Paragangliomas and a Growth Hormone-Producing Pituitary Adenoma as Phenotypes From a Novel Succinate Dehydrogenase Subunit D Mutation.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Clin Nucl Med. 2013 Oct 22.

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

1097/RLU.0000000000000235

AUTORES / AUTHORS: - Skoura E; Datseris IE; Xekouki P; Tolis G; Stratakis CA

INSTITUCIÓN / INSTITUTION: - From the *Department of Nuclear Medicine, Evangelismos General Hospital, Athens, Greece; daggerSection on Endocrinology and Genetics, Program on Developmental Endocrinology and Genetics, National Institute of Child Health and Human Development, National Institutes of Health, Bethesda, MD; and double daggerDivision of Endocrinology and Metabolism, Hippocrateion General Hospital, Athens, Greece.

RESUMEN / SUMMARY: - Mutations in the subunits B, C, D, and recently in A of the succinate dehydrogenase have been associated with the development of paragangliomas. We report the case of a 37-year-old man presented with multiple paragangliomas and a growth hormone-producing pituitary adenoma, with a novel succinate dehydrogenase subunit D mutation as the genetic analysis revealed. We present the similarities and the differences of the findings in patient imaging with either methods of SPECT (I-MIBG and In-pentetreotide) or PET/CT with F-FDG. This case revealed that F-FDG PET/CT detected more lesions and was superior compared with the other methods.

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[99]

TÍTULO / TITLE: - Spectrum of pulmonary neuroendocrine proliferations and neoplasms.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Radiographics. 2013 Oct;33(6):1631-49. doi: 10.1148/rg.336135506.

●● Enlace al texto completo (gratuito o de pago) 1148/rg.336135506

AUTORES / AUTHORS: - Benson RE; Rosado-de-Christenson ML; Martinez-Jimenez S; Kunin JR; Pettavel PP

INSTITUCIÓN / INSTITUTION: - Departments of Radiology and Pathology, Saint Luke's Hospital of Kansas City, 4401 Wornall Rd, Kansas City, MO 64111.

RESUMEN / SUMMARY: - Neuroendocrine neoplasms are ubiquitous tumors found throughout the body, most commonly in the gastrointestinal tract followed by the thorax. Neuroendocrine cells occur normally in the bronchial and bronchiolar epithelium and may be solitary or may occur in clusters. Although neuroendocrine cell proliferations may be found in association with chronic lung disease, a broad range of neuroendocrine proliferations and neoplasms may occur and exhibit variable biologic behavior. Diffuse idiopathic neuroendocrine cell hyperplasia (DIPNECH) is a diffuse idiopathic form of neuroendocrine cell hyperplasia and is considered a preinvasive lesion that may give rise to carcinoid tumors. Patients with DIPNECH are typically older women who may be asymptomatic or may present with chronic respiratory symptoms. DIPNECH manifests as multifocal bilateral pulmonary micronodules on expiratory high-resolution computed tomographic (CT) images; the air trapping is secondary to

constrictive bronchiolitis. Carcinoid tumors are low-grade malignant neoplasms that typically affect symptomatic children and young adults. Carcinoids manifest as well-defined pulmonary nodules or masses that are often closely related to central bronchi. They may exhibit intrinsic calcification and contrast material enhancement at CT, and patients with carcinoids may have postobstructive atelectasis and pneumonia. Although typical carcinoids are indolent neoplasms and patients have a good prognosis, atypical carcinoids are aggressive malignancies with a propensity for metastasis. Both are optimally treated with complete surgical excision. Large cell neuroendocrine carcinoma and small cell lung cancer are highly aggressive neuroendocrine malignancies that usually affect elderly smokers. These tumors manifest with large peripheral or central pulmonary masses. Local invasion, intrathoracic lymphadenopathy, and distant metastases are frequent at presentation. As a result, affected patients may not be candidates for surgical resection, are often treated with chemotherapy with or without radiation, and have a poor prognosis.

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[100]

TÍTULO / TITLE: - Prognostic factors in neuroendocrine tumours of the lung: a single-centre experience.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Eur J Cardiothorac Surg. 2013 Oct 3.

●● Enlace al texto completo (gratuito o de pago) 1093/ejcts/ezt442

AUTORES / AUTHORS: - Filosso PL; Ruffini E; Di Gangi S; Guerrera F; Bora G; Ciccone G; Galassi C; Solidoro P; Lyberis P; Oliaro A; Sandri A

INSTITUCIÓN / INSTITUTION: - Department of Thoracic Surgery, University of Torino, Torino, Italy.

RESUMEN / SUMMARY: - OBJECTIVES: To assess the independent prognostic role of histological subtypes, tumour size and lymph nodal involvement upon survival in lung neuroendocrine tumours (NETs). METHODS: A retrospective search of the database of the Department of Thoracic Surgery (Turin, Italy) identified 157 patients operated on for a newly diagnosed NET between January 1995 and December 2011. Multivariable Cox models were used to analyse predictors of overall survival and progression-free survival. RESULTS: According to histology, 71 (45.2%) were typical carcinoids (TCs), 35 (22.3%) atypical carcinoids (ACs), 37 (23.6%) large-cell neuroendocrine carcinomas (LCNCs) and 14 (8.9%) small-cell lung carcinomas (SCLCs). After a median follow-up time of 6.5 years, 60 patients died and 73 had a recurrence or died. The overall 5-, 10- and 15-year survival rates were 64%, 53% and 46%, respectively. Older age, histology (ACs, LCNCs and SCLCs vs TCs) and lymph nodal involvement were confirmed to be independent negative prognostic factors in the multivariable models for overall survival and progression-free survival. CONCLUSIONS: Tumour histology and lymph nodal involvement are definitively the predominant and relevant factors influencing survival. ACs showed an intermediate prognosis between TCs and poorly differentiated NETs.

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[101]

TÍTULO / TITLE: - Current update on medullary thyroid carcinoma.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - AJR Am J Roentgenol. 2013 Dec;201(6):W867-76. doi: 10.2214/AJR.12.10370.

●● Enlace al texto completo (gratuito o de pago) 2214/AJR.12.10370

AUTORES / AUTHORS: - Ganeshan D; Paulson E; Duran C; Cabanillas ME; Busaidy NL; Charnsangavej C

INSTITUCIÓN / INSTITUTION: - 1 Department of Diagnostic Imaging, Unit 1473, The University of Texas M. D. Anderson Cancer Center, 1515 Holcombe Blvd, Houston, TX 77030-4009.

RESUMEN / SUMMARY: - OBJECTIVE. This article will review the multimodality imaging spectrum of medullary thyroid carcinoma (MTC) with an emphasis on anatomic and functional imaging. Recent advances in the molecular cytogenetics of this tumor and the impact on diagnosis, prognosis, and development of novel targeted therapy will be discussed. CONCLUSION. MTC is a neuroendocrine tumor with unique clinicopathologic and radiologic features compared with other thyroid malignancies. Imaging plays an important role in the optimal management of this malignancy.

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[102]

TÍTULO / TITLE: - Chromophobe Renal Cell Carcinoma: Multiphase MDCT Enhancement Patterns and Morphologic Features.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - AJR Am J Roentgenol. 2013 Dec;201(6):1268-76. doi: 10.2214/AJR.13.10813.

●● Enlace al texto completo (gratuito o de pago) 2214/AJR.13.10813

AUTORES / AUTHORS: - Raman SP; Johnson PT; Allaf ME; Netto G; Fishman EK

INSTITUCIÓN / INSTITUTION: - 1 Department of Radiology, Johns Hopkins University, JHOC 3251, 601 N Caroline St, Baltimore, MD 21287.

RESUMEN / SUMMARY: - OBJECTIVE. The purpose of this investigation is to retrospectively describe morphologic features, enhancement characteristics, and clinical outcomes in a series of pathologically proven chromophobe renal cell carcinomas (RCCs). MATERIALS AND METHODS. Thirty-five patients who were imaged at a single institution between 2005 and 2012 with pathologically proven chromophobe RCC were identified, all of whom underwent preoperative renal protocol CT (unenhanced, arterial, venous, and delayed images). The morphologic characteristics of each tumor (e.g., necrosis, tumor composition, and calcification), as well as attenuation values (in Hounsfield units) of the tumor, aorta, inferior vena cava, and kidney were evaluated by a board-certified radiologist. In addition, information regarding patient demographics and survival was obtained by a separate radiologist from the electronic medical record. RESULTS. Sixty percent of the patients were men, with a mean age of 60.2 years. Forty-six percent of cases were incidentally identified, without patient symptoms. None of the patients had evidence of distant metastatic disease, either on initial staging CT or over the course of follow-up (mean, 2.0 years). Mean maximal tumor diameter was 5.24 cm. Forty-six percent of tumors were homogeneous, 85% of lesions were either completely solid or mostly solid, 14% showed calcifications, and 34% showed a central scar or necrosis. Mean maximum attenuation values were 87.9 HU (arterial phase), 83.9 HU (venous phase), and 60.6 HU (delayed phase), with an average delayed washout of 31%. Tumor-to-cortex ratios for the three enhanced phases were 0.59, 0.48, and 0.50, respectively. CONCLUSION.

Chromophobe RCCs were found to have a wider variability of CT features than previously reported, although they do have a greater propensity for homogeneity and the presence of a central scar or necrosis. Their enhancement characteristics fall in between those of clear cell and papillary RCC, although there is considerable overlap.

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[103]
TÍTULO / TITLE: - A Rare Case of Sudden Death Due to Hypotension during Cesarean Section in a Woman Suffering from Pheochromocytoma and Neurofibromatosis.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - J Forensic Sci. 2013 Nov;58(6):1636-9. doi: 10.1111/1556-4029.12279. Epub 2013 Oct 3.
●● Enlace al texto completo (gratuito o de pago) 1111/1556-4029.12279
AUTORES / AUTHORS: - Cecchi R; Frati P; Capri O; Cipolloni L
INSTITUCIÓN / INSTITUTION: - Department of Anatomical Histological Legal Medical and Orthopaedic Sciences, Faculty of Medicine and Pharmacology, Sapienza University of Rome, Rome, Italy.
RESUMEN / SUMMARY: - Sudden death following acute hypotension due to an undiagnosed pheochromocytoma (PHEO) is a rare event. Moreover, histopathology of the myocardium in such cases is rarely reported. We present a case of a woman who died during delivery. A 37-year-old parturient, who was 38 weeks pregnant, suffering from neurofibromatosis underwent a cesarean section following peridural anesthesia. Acute hypotension, acute intra-operative pulmonary edema and supraventricular paroxysmal tachyarrhythmia occurred during delivery, followed by death. The autopsy revealed the presence of a PHEO, confirmed immunohistochemically with chromogranin-A (CgA), CD20 antibody (L26), anti-Keratocan antibody (KER-1) and neuron-specific enolase (NSE), and a PHEO-induced cardiomyopathy. The physiopathology of both stress-induced cardiomyopathy and PHEO-induced cardiomyopathy, as well as the role of anesthesia in provoking the death, are discussed. The association of an undiagnosed PHEO with neurofibromatosis as the cause of sudden death in pregnancy is an obstetric urgency that raises forensic pathology issues.

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[104]
TÍTULO / TITLE: - Dual Inhibition of PI3K and mTOR Signaling Pathways Decreases Human Pancreatic Neuroendocrine Tumor Metastatic Progression.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Pancreas. 2013 Nov 20.
●● Enlace al texto completo (gratuito o de pago)
1097/MPA.0b013e3182a44ab4
AUTORES / AUTHORS: - Djukom C; Porro LJ; Mrazek A; Townsend CM Jr; Hellmich MR; Chao C
INSTITUCIÓN / INSTITUTION: - From the Department of Surgery, University of Texas Medical Branch, Galveston, TX.
RESUMEN / SUMMARY: - OBJECTIVES: Patients with advanced pancreatic neuroendocrine tumors have limited therapeutic options. Everolimus (RAD001), an inhibitor of the mammalian target of rapamycin (mTOR) pathway, has been shown to

increase progression-free survival, but not overall survival, indicating a need to identify additional therapeutic targets. Inhibition of mTOR complex 1 by RAD001 may induce upstream AKT upregulation. We hypothesized that dual inhibition of AKT along with mTOR will overcome the limited activity of RAD001 alone. METHODS: The BON cell line has been used as a model to study pancreatic neuroendocrine tumor cell biology. Western blots and cell growth assays were performed with mTOR inhibitor RAD001 (50 nM), mitogen-activated protein kinase inhibitor PD0325901 (50 nM), PI3K (phosphatidylinositol 3-kinase) inhibitor LY294002 (25 muM), or vehicle control. Nude mice were treated daily for 6 weeks with RAD001 (oral gavage) and with LY29400 (subcutaneous) 1 week after intrasplenic injection of BON cells. RESULTS: Cellular proliferation was most attenuated with the combination therapy of LY29400 and RAD001. Similarly, the volume of liver metastasis was lowest in the group treated with both LY29400 (100 mg/kg per week, subcutaneous) and RAD001 (2.5 mg/kg per day) compared with that in the vehicle group (P = 0.04). CONCLUSION: The combination therapy of LY29400 and RAD001 decreased the cell growth in vitro and progression of liver metastasis in vivo compared with vehicle or with single-drug therapy.

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[105]

TÍTULO / TITLE: - CD8+ lymphocytes and apoptosis in typical and atypical medullary carcinomas of the breast.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Immunol Lett. 2013 Nov-Dec;156(1-2):123-6. doi: 10.1016/j.imlet.2013.10.001. Epub 2013 Oct 11.

●● Enlace al texto completo (gratuito o de pago) 1016/j.imlet.2013.10.001

AUTORES / AUTHORS: - Nurlaila I; Telisinghe PU; Ramasamy R

INSTITUCIÓN / INSTITUTION: - Institute of Health Sciences, Universiti Brunei Darussalam, Gadong, Brunei Darussalam.

RESUMEN / SUMMARY: - Medullary breast carcinoma (MBC) is a form of ductal invasive carcinoma (DIC) characterized by an abundant infiltration of the tumour by lymphocytes. MBC has been classified histologically into typical medullary carcinoma (TMC) and atypical medullary carcinoma (AMC), with TMC having a better prognosis than AMC and other DIC. The distribution of CD8+ lymphocytes within tumour nests and lymphocyte tracts, and apoptosis in lymphocytes and tumour cells within tumour nests, were studied in archived formalin fixed and paraffin embedded tissues of TMC and AMC. CD8+ lymphocytes tend to accumulate along the margins of lymphocyte tracts that adjoin tumour nests. There were significantly more CD8+ lymphocytes within tumour nests of TMC than AMC. TMC also tended to have more CD8+ lymphocytes within lymphocyte tracts than AMC. Apoptosis of lymphocytes in contact with tumour cells and of tumour cells in contact with lymphocytes was observed in both AMC and TMC within tumour nests but differences in the proportions of apoptotic tumour cells and lymphocytes between the two tumour types could not be established. The findings are consistent with CD8+ cytotoxic lymphocyte-mediated immunity contributing to the more favourable prognosis for TMC compared to AMC.

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[106]

TÍTULO / TITLE: - Thymic neuroendocrine carcinoma producing ectopic adrenocorticotropic hormone and Cushing's syndrome.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Ann Thorac Surg. 2013 Oct;96(4):e81-3. doi: 10.1016/j.athoracsur.2013.04.140.

●● Enlace al texto completo (gratuito o de pago)
1016/j.athoracsur.2013.04.140
AUTORES / AUTHORS: - Dixon JL; Borgaonkar SP; Patel AK; Reznik SI; Smythe WR; Rascoe PA
INSTITUCIÓN / INSTITUTION: - Scott & White Memorial Hospital & Clinic, Texas A&M Health Science Center, Temple, Texas.
RESUMEN / SUMMARY: - Neuroendocrine carcinoma of the thymus, previously termed thymic carcinoid, is a rare clinical entity. Rarer still are such cases presenting with endocrinopathies. We report a case of thymic neuroendocrine carcinoma presenting with ectopic adrenocorticotroic hormone production and resultant Cushing's syndrome.

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[107]
TÍTULO / TITLE: - Extra-appendiceal Neuroendocrine Neoplasms in Children - Data from the GPOH-MET 97 Study.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Klin Padiatr. 2013 Nov;225(6):315-319. Epub 2013 Oct 24.

●● Enlace al texto completo (gratuito o de pago) 1055/s-0033-1355353
AUTORES / AUTHORS: - Redlich A; Wechsung K; Boxberger N; Leuschner I; Vorwerk P
INSTITUCIÓN / INSTITUTION: - Pediatric Oncology, GPOH-MET Registry, Otto-von-Guericke University Magdeburg.
RESUMEN / SUMMARY: - Neuroendocrine neoplasms (NEN) in children are rare. In Germany, children with NEN of the gastroenteropancreatic system are prospectively registered since 1997. The objective of this study was to evaluate diagnostics, treatment and outcome in children with extra-appendiceal NEN.Clinical data of 39 patients with NEN registered in the GPOH-MET 97 trial from 1997 to 2012 were analyzed. Children with NEN of the appendix were excluded.14 patients with pancreatic, 12 patients with bronchial, 6 patients with gastrointestinal, 2 patients with nasopharyngeal and 5 patients with NEN of unknown primary were registered. About half of the patients had localized disease and rather low grade tumors, including all bronchial NEN, 5 of 14 pancreatic and 2 of 6 gastrointestinal tumors. Metastatic disease and high grade tumors were stated in cases with nasopharyngeal tumors, NEN of unknown -primary and in part of pancreatic and gastrointestinal NEN. Complete surgical resection was performed in patients with localized NEN with an overall survival of 100%. In contrast, overall survival in metastatic disease was 26%.Outcome in children with low grade NEN and localized disease is excellent. Management of high grade tumors and metastatic disease remains challenging. Establishing international registries is inevitable for further improvements.

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[108]

TÍTULO / TITLE: - Laparoscopic transduodenal local resection of periampullary neuroendocrine tumor: A case report.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - World J Gastroenterol. 2013 Oct 21;19(39):6693-8. doi: 10.3748/wjg.v19.i39.6693.
●● Enlace al texto completo (gratuito o de pago) 3748/wjg.v19.i39.6693
AUTORES / AUTHORS: - Zhang RC; Xu XW; Wu D; Zhou YC; Ajoodhea H; Chen K; Mou YP
INSTITUCIÓN / INSTITUTION: - Ren-Chao Zhang, Xiao-Wu Xu, Di Wu, Yu-Cheng Zhou, Harsha Ajoodhea, Ke Chen, Yi-Ping Mou, Department of General Surgery, Sir Run Run Shaw Hospital, School of Medicine, Zhejiang University, Hangzhou 310016, Zhejiang Province, China.
RESUMEN / SUMMARY: - Studies on laparoscopic transduodenal local resection have not been readily available. Only three cases have been reported in the English-language literature. We describe herein a case of 25-year-old woman with periampullary neuroendocrine tumor (NET). Endoscopic ultrasonography revealed a duodenal papilla mass originated from the submucosa and close to the ampulla. The periampullary tumor was successfully managed with laparoscopic transduodenal local resection without any procedure-related complications. Pathological examination showed a NET (Grade 2) with negative margin. The patient was followed up for six months without signs of recurrence. This case suggests that laparoscopic transduodenal local resection is a feasible procedure in selected patients with periampullary tumor.

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[109]
TÍTULO / TITLE: - Collision of extensive exocrine and neuroendocrine neoplasms in multiple endocrine neoplasia type 1 revealed by cytogenetic analysis of loss of heterozygosity: A case report.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Pathol Int. 2013 Sep;63(9):469-75. doi: 10.1111/pin.12088. Epub 2013 Aug 28.
●● Enlace al texto completo (gratuito o de pago) 1111/pin.12088
AUTORES / AUTHORS: - Moriyoshi K; Minamiguchi S; Miyagawa-Hayashino A; Fujimoto M; Kawaguchi M; Haga H
INSTITUCIÓN / INSTITUTION: - Department of Diagnostic Pathology, Kyoto University Hospital, Kyoto, Japan.
RESUMEN / SUMMARY: - The combination of exocrine and neuroendocrine neoplasms is rarely found in the pancreas. These combined lesions vary from a clonal tumor with mixed differentiation to the incidental co-existence of two or more independent tumors, but the differential diagnosis is sometimes difficult. Here we report a case of multiple endocrine neoplasia type 1 (MEN1) with extensive ductal and neuroendocrine neoplastic changes. These two types of tumors admixed markedly in some parts, which made it difficult to determine the pathological diagnosis based on histological findings. Cytogenetic analysis showed that loss of heterozygosity (LOH) of the MEN1 locus exists in neuroendocrine but not in exocrine neoplasms, indicating that independent mechanisms of tumorigenesis may occur in these two types of tumors. This case shows the usefulness of cytogenetic analysis for the diagnosis of combined

tumors of the pancreas. Extensive exocrine neoplastic change, including pancreatic intraepithelial neoplasia (PanIN) in virtually all pancreatic ducts and a focus of intraductal papillary mucinous neoplasm (IPMN) with focal invasion, was a distinguishing feature of the present case. The possible association of ductal tumorigenesis and a MEN1 background is discussed.

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[110]
TÍTULO / TITLE: - Analytical and preanalytical validation of a new mass spectrometric serum 5-hydroxyindoleacetic acid assay as neuroendocrine tumor marker.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Clin Chim Acta. 2013 Nov 8;428C:38-43. doi: 10.1016/j.cca.2013.10.025.
●● Enlace al texto completo (gratuito o de pago) 1016/j.cca.2013.10.025
AUTORES / AUTHORS: - Tohmola N; Itkonen O; Sane T; Markkanen H; Joenvaara S; Renkonen R; Hamalainen E
INSTITUCIÓN / INSTITUTION: - Haartman Institute, University of Helsinki, Helsinki, Finland. Electronic address: niina.tohmola@helsinki.fi.
RESUMEN / SUMMARY: - BACKGROUND: Serum 5-hydroxyindoleacetic acid (5-HIAA) could replace the determination of 24-h urinary 5-HIAA for diagnosis and follow-up of neuroendocrine tumors (NETs). We developed and validated a straightforward liquid chromatography tandem mass spectrometry (LC-MS/MS) assay for serum 5-HIAA. METHODS: We used serum samples from healthy volunteers (n=136) and patients suspected or followed for NET (n=129). Samples were spiked with 5-HIAA-D2, extracted and quantified by LC-MS/MS. We studied the effects of sample storage, sample device, a meal and diurnal variation on serum 5-HIAA. Furthermore, we established a reference range for serum 5-HIAA and compared our assay with a urinary 5-HIAA HPLC assay and a commercial plasma chromogranin A (CgA) immunoassay. RESULTS: Our LC-MS/MS assay is sensitive (LOQ 5nmol/L), has a wide assay range (5-10,000nmol/L) and short analysis time (7min). 5-HIAA in serum is stable for several days in various temperatures and during five freeze-thaw cycles. We found no diurnal variation (p>/=0.20) and a meal had no effect on serum 5-HIAA (p=0.89). We suggest an upper reference limit of 123nmol/L for serum 5-HIAA. The area under curve (AUC) in receiver operator characteristics (ROC) analysis was 0.83 for urinary 5-HIAA, 0.81 for serum 5-HIAA and 0.76 for CgA, respectively. CONCLUSIONS: The LC-MS/MS assay for serum 5-HIAA discriminates between healthy individuals and patients with NET and is well suited for the diagnosis and follow-up of NETs.

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[111]
TÍTULO / TITLE: - Calcific malignant pheochromocytoma.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - QJM. 2013 Nov 21.
●● Enlace al texto completo (gratuito o de pago) 1093/qjmed/hct218
AUTORES / AUTHORS: - Chen CT; Chu HC
INSTITUCIÓN / INSTITUTION: - Department of Internal Medicine, Tri-Service General Hospital, National Defense Medical Center, Taipei, Taiwan.

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[112]

TÍTULO / TITLE: - F-FDG PET/CT for detection of malignant peripheral nerve sheath tumours in neurofibromatosis type 1: tumour-to-liver ratio is superior to an SUV cut-off.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Eur Radiol. 2013 Oct 5.

●● Enlace al texto completo (gratuito o de pago) 1007/s00330-013-3020-x

AUTORES / AUTHORS: - Salamon J; Veldhoen S; Apostolova I; Bannas P; Yamamura J; Herrmann J; Friedrich RE; Adam G; Mautner VF; Derlin T

INSTITUCIÓN / INSTITUTION: - Department of Diagnostic and Interventional Radiology, University Medical Centre Hamburg-Eppendorf, Hamburg, Germany, j.salamon@uke.de.

RESUMEN / SUMMARY: - OBJECTIVES: To evaluate the usefulness of normalising intra-tumour tracer accumulation on 18F-fluorodeoxyglucose (FDG) positron emission tomography/computed tomography (PET/CT) to reference tissue uptake for characterisation of peripheral nerve sheath tumours (PNSTs) in neurofibromatosis type 1 (NF1) compared with the established maximum standardised uptake value (SUVmax) cut-off of >3.5. METHODS: Forty-nine patients underwent FDG PET/CT. Intra-tumour tracer uptake (SUVmax) was normalised to three different reference tissues (tumour-to-liver, tumour-to-muscle and tumour-to-fat ratios). Receiver operating characteristic (ROC) analyses were used out to assess the diagnostic performance. Histopathology and follow-up served as the reference standard. RESULTS: Intra-tumour tracer uptake correlated significantly with liver uptake (r s = 0.58, P = 0.016). On ROC analysis, the optimum threshold for tumour-to-liver ratio was >2.6 (AUC = 0.9735). Both the SUVmax cut-off value of >3.5 and a tumour-to-liver ratio >2.6 provided a sensitivity of 100 %, but specificity was significantly higher for the latter (90.3 % vs 79.8 %; P = 0.013). CONCLUSIONS: In patients with NF1, quantitative 18F-FDG PET imaging may identify malignant change in neurofibromas with high accuracy. Specificity could be significantly increased by using the tumour-to-liver ratio. The authors recommend further evaluation of a tumour-to-liver ratio cut-off value of >2.6 for diagnostic intervention planning. KEY POINTS: * 18 F-FDG PET/CT is used for detecting malignancy in PNSTs in NF1 patients * An SUV max cut-off value may give false-positive results for benign plexiform neurofibromas * Specificity can be significantly increased using a tumour-to-liver ratio.

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[113]

TÍTULO / TITLE: - Neuroendocrine tumor of the extrahepatic bile duct: a tumor in an unusual site visualized by cholangioscopy.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Endoscopy. 2013 Oct;45 Suppl 2:E338-9. doi: 10.1055/s-0032-1326453. Epub 2013 Oct 22.

●● Enlace al texto completo (gratuito o de pago) 1055/s-0032-1326453

AUTORES / AUTHORS: - De Luca L; Tommasoni S; de Leone A; Bianchi ML; de Nictolis M; Baroncini D

INSTITUCIÓN / INSTITUTION: - Gastroenterology and Digestive Endoscopy Unit, San Salvatore Hospital, Pesaro, Italy.

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[114]

TÍTULO / TITLE: - Gut neuroendocrine tumor blood qPCR fingerprint assay: characteristics and reproducibility.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Clin Chem. Acceso gratuito al texto completo a partir de 1 año de la fecha de publicación.

●● Enlace a la Editora de la Revista http://www.journals.uchicago.edu/

●● Cita: Clinical Infectious Diseases: <> Lab Med. 2013 Oct 14:1-11. doi: 10.1515/cclm-2013-0496.

●● Enlace al texto completo (gratuito o de pago) 1515/cclm-2013-0496

AUTORES / AUTHORS: - Modlin IM; Drozdov I; Kidd M

RESUMEN / SUMMARY: - Abstract Background: We have developed a PCR-based tool that measures a 51-gene panel for identification of gastroenteropancreatic (GEP) neuroendocrine neoplasms (NENs) in peripheral blood. This manuscript assesses the robustness (performance metrics) of this tool with a specific focus on the effects of individual parameters including collection, storage, acid suppressive medication [proton pump inhibitor (PPI)], age, sex, race and food on accuracy. Methods: Performance metrics were evaluated using a gold standard (mRNA derived from three individual human neuroendocrine tumor cell lines) and clinical samples using qPCR. Results: One hundred percent of the 51 transcripts were amplified in the gold standard (NEN cell line-derived mRNA) (CQ<35, average efficiency 1.94). The inter- and intra-assay variations were 1%-2%. In clinical samples, 50 of 51 targets (98%) were amplified. The inter- and intra-assay reproducibility ranged between 0.4% and 1.2%. The coefficient of variation (CV) was 5.3%. Expression of the reference gene, ALG9, was robust [low variation, low M-value, high (99.5%) PCR efficiency] and unaffected by sample processing. Test meals, long-term PPI use (>1 year), age, sex and ethnicity had no effect on the signature. Expression of two genes, ALP2 and CD59 correlated strongly with RNA integrity (R=0.72, p<0.001) and could be used to assess storage and processing. Conclusions: The 51 marker gene signature was robust and reproducible, exhibiting acceptable inter- and intra-assay metrics (<5%). Feeding, PPI intake, age, sex and ethnicity do not affect the signature. Expression levels of APLP2 and CD59 are effective surrogate markers of proper sample collection and processing.

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[115]

TÍTULO / TITLE: - Ga-DOTA-TOC-PET/CT detects heart metastases from ileal neuroendocrine tumors.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Endocrine. 2013 Nov 23.

●● Enlace al texto completo (gratuito o de pago) 1007/s12020-013-0108-7

AUTORES / AUTHORS: - Calissendorff J; Sundin A; Falhammar H

INSTITUCIÓN / INSTITUTION: - Endocrine Section, VO Internmedicin, Department of Clinical Science and Education, Karolinska Institutet, Sodersjukhuset, Sjukhusbacken 10, 118 83, Stockholm, Sweden, jan.calissendorff@sodersjukhuset.se.

RESUMEN / SUMMARY: - Metastases from ileal neuroendocrine tumors (NETs) to the myocardium are rare and generally seen in patients with widespread metastatic NET

disease. The objectives of this investigation were to describe the frequency of intracardiac metastases in ileal NET patients examined by 68Ga-DOTA-TOC-PET/CT and to describe the cases in detail. All 68Ga-DOTA-TOC-PET/CT examinations performed at the Karolinska University Hospital since 2010 until April 2012 were reviewed. In all, 128 out of 337 examinations were in patients with ileal NETs. Four patients had seven myocardiac metastases, yielding a frequency of 4.3 % in patients with ileal NETs. One patient had cardiac surgery while three were treated with somatostatin analogs. The cardiac metastases did not affect the patients' activity of daily life. 68Ga-DOTA-TOC-PET/CT is an established imaging modality in identifying cardiac metastases in ileal NETs. Prospective studies are needed to confirm the true clinical value of 68Ga-DOTA-TOC-PET/CT in detecting cardiac metastases in both ileal and non-ileal NETs.

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[116]

TÍTULO / TITLE: - Tissue microarray analysis as a screening tool for neuroendocrine carcinoma of the breast.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - APMIS. 2013 Nov 28. doi: 10.1111/apm.12198.

●● Enlace al texto completo (gratuito o de pago) 1111/apm.12198

AUTORES / AUTHORS: - Brask JB; Talman ML; Wielenga VT

INSTITUCIÓN / INSTITUTION: - The Department of Pathology, Rigshospitalet, Copenhagen, Denmark.

RESUMEN / SUMMARY: - Neuroendocrine carcinoma of the breast (NCB) is a fairly recent diagnostic entity added by WHO in 2003. Since then, studies have indicated that NCB potentially displays a worse prognosis than invasive ductal carcinoma. However, due to a lack of standard use of immunohistochemical staining for neuroendocrine markers and the fact that NCB may only show slight neuroendocrine morphology that can easily be overlooked, NCB is often underdiagnosed. Consequently, there is a need for fast and reliable detection method for NCB. Here, we take a first step toward finding an easy way of identifying NCB by investigating the usefulness of tissue microarray (TMA) analysis as a screening tool. We present our findings with regard to sensitivity and specificity compared with whole-mount sections. The material consists of 240 cases of breast cancer divided into 20 TMA blocks that were all immunohistochemically stained for the neuroendocrine markers chromogranin A and synaptophysin. Cases positive in more than 50% of the tumor cells were accepted in accordance with WHO (2003) standards of NCB. Sensitivity and specificity for TMA sections vs whole-mount sections were found to be 100% and 97.8%, respectively, suggesting that TMA analysis is a reliable method for NCB detection.

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[117]

TÍTULO / TITLE: - Biological correlation of (1)(8)F-FDG uptake on PET in pulmonary neuroendocrine tumors.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Anticancer Res. 2013 Oct;33(10):4219-28.

AUTORES / AUTHORS: - Kaira K; Murakami H; Endo M; Ohde Y; Naito T; Kondo H; Nakajima T; Yamamoto N; Takahashi T

INSTITUCIÓN / INSTITUTION: - Division of Thoracic Oncology, Shizuoka Cancer Center, 1007 Shimonagakubo Nagaizumi-cho Sunto-gun, Shizuoka, 411-8777, Japan. kkaira1970@yahoo.co.jp.

RESUMEN / SUMMARY: - BACKGROUND: It is widely recognized that pulmonary neuroendocrine tumors (PNET) include a spectrum that ranges from low-grade typical carcinoid (TC) and atypical carcinoid (AC) to high-grade large cell neuroendocrine carcinoma (LCNEC) and small cell lung carcinoma (SCLC). However, little is known about the usefulness of 2-[(18)F]-fluoro-2-deoxy-D-glucose ((18)F-FDG) positron-emission tomography (PET) in such tumors. We therefore, conducted a study including the analysis of the underlying biology of (18)F-FDG uptake. MATERIALS AND METHODS: Thirty-four patients with early-stage PNETs who underwent (18)F-FDG PET before treatment were included in this study. Tumor sections were stained by immunohistochemistry for glucose transporter-1 (Glut1 and Glut3), hypoxia-inducible factor-1 alpha (HIF-1alpha), hexokinase-I, vascular endothelial growth factor (VEGF), microvessel density (MVD) determined by CD34 and (Akt)/mammalian target of rapamycin (mTOR) signaling pathway. RESULTS: (18)F-FDG uptake correlated significantly with Glut1, HIF-1alpha, VEGF and CD34 expression. Uptake of (18)F-FDG tended to increase from low-grade to high-grade PNETs. Tumor metabolic activity was a useful marker for predicting postoperative prognosis in patients with early-stage PNETs. CONCLUSION: The amount of (18)F-FDG uptake is determined by the presence of glucose metabolism, hypoxia and angiogenesis.

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[118]

TÍTULO / TITLE: - Dual Tracer Functional Imaging of Gastroenteropancreatic Neuroendocrine Tumors Using 68Ga-DOTA-NOC PET-CT and 18F-FDG PET-CT: Competitive or Complimentary?

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Clin Nucl Med. 2013 Nov 7.

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

1097/RLU.0b013e31827a216b

AUTORES / AUTHORS: - Naswa N; Sharma P; Gupta SK; Karunanithi S; Reddy RM; Patnecha M; Lata S; Kumar R; Malhotra A; Bal C

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

RESUMEN / SUMMARY: - OBJECTIVE: This study aimed to compare the diagnostic performance of Ga-DOTANOC PET/CT with F-FDG PET/CT in the patients with gastroenteropancreatic neuroendocrine tumors (GEP-NETs). PATIENTS AND METHODS: Data of 51 patients with definite histological diagnosis of GEP-NET who underwent both Ga-DOTA-NOC PET-CT and F-FDG PET-CT within a span of 15 days were selected for this retrospective analysis. Sensitivity, specificity, and predictive values were calculated for Ga-DOTA-NOC PET-CT and F-FDG PET-CT, and results were compared both on patientwise and regionwise analysis. RESULTS: Ga-DOTA-NOC PET-CT is superior to F-FDG PET-CT on patientwise analysis ($P < 0.0001$). On regionwise analysis, Ga-DOTA-NOC PET-CT is superior to F-FDG PET-CT only for lymph node metastases ($P < 0.003$). Although Ga-DOTA-NOC PET-CT detected more liver and skeletal lesions compared with F-FDG PET-CT, the difference was not statistically significant. In addition, the results of combined imaging helped in selecting

candidates who would undergo the appropriate mode of treatment, whether octreotide therapy or conventional chemotherapy CONCLUSIONS: Ga-DOTA-NOC PET-CT seems to be superior to F-FDG PET-CT for imaging GEP-NETs. However, their role seems to be complementary because combination of Ga-DOTA-NOC PET-CT and F-FDG PET-CT in such patients helps demonstrate the total disease burden and segregate them to proper therapeutic groups.

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[119]

TÍTULO / TITLE: - The impact of 18F-FDG-PET/CT on Merkel cell carcinoma management: a retrospective study of 66 scans from a single institution.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Nucl Med Commun. 2013 Nov 14.

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

1097/MNM.0000000000000039

AUTORES / AUTHORS: - George A; Girault S; Testard A; Delva R; Soulie P; Couturier OF; Morel O

INSTITUCIÓN / INSTITUTION: - aDepartments of Nuclear Medicine and Medical Oncology, Comprehensive Cancer Center ICO Paul Papin, Angers, bDepartment of Nuclear Medicine, University Hospital, Angers, France.

RESUMEN / SUMMARY: - PURPOSE: Merkel cell carcinomas (MCC) are neuroendocrine skin tumours frequently responsible for lymph node recurrence and metastatic disease and for which optimal management remains to be defined. The objective of this study was to evaluate the impact of F-fluorodeoxyglucose (F-FDG)-PET/computed tomography (CT) on the staging and treatment of MCC patients. MATERIALS AND METHODS: Twenty-three patients with a histologic diagnosis of MCC explored by F-FDG-PET/CT between 2004 and 2012 were retrospectively included in the study. The detection of new lesions and the change in tumour staging and treatment were evaluated. For each patient, the F-FDG-PET/CT results were compared with histological, clinical and imaging data. RESULTS: Sixty-six F-FDG-PET/CT scans were performed at initial presentation (n=18), during subsequent monitoring (n=34) or during evaluation of chemotherapy response (n=14). The sensitivity, specificity and positive and negative predictive values of the F-FDG-PET/CT were 97, 89, 94 and 94%, respectively. Two false-positive results (lymphadenitis) and one false-negative result (regional metastatic lymph nodes) were accounted for. Lesions not detected clinically or by conventional imaging techniques were found in 44% of the 52 F-FDG-PET/CTs performed at initial presentation and subsequent monitoring, with, respectively, 50 and 41% of scans identifying new lesions. At initial presentation, F-FDG-PET/CT led to a change in tumour staging in 39% of patients. Patient management was modified by F-FDG-PET/CT results in one-third of patients (33% of patients at initial presentation, 32% during subsequent monitoring and 36% during evaluation of chemotherapy response). F-FDG-PET/CT incidentally detected four additional histologically confirmed cancers. CONCLUSION: This retrospective study confirms the important impact of F-FDG-PET/CT on the management of MCC patients.

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[120]

TÍTULO / TITLE: - Recurrence of cardiomyopathy by recurrent pheochromocytoma.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Int J Cardiol. 2013 Oct 30;169(2):e31-2. doi: 10.1016/j.ijcard.2013.08.099. Epub 2013 Sep 9.
●● Enlace al texto completo (gratuito o de pago) 1016/j.ijcard.2013.08.099
AUTORES / AUTHORS: - Baskok FA; Senturk B; Ozeke O; Aras D
INSTITUCIÓN / INSTITUTION: - Turkiye Yuksek Ihtisas Hospital, Department of Cardiology, Ankara, Turkey. Electronic address: firdevs_bsk@hotmail.com.

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[121]
TÍTULO / TITLE: - Synchronous Ileocecal Neuroendocrine Tumor and Carotid Chemodectoma: Diagnosis by 111In Pentetreotide SPECT/CT.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Clin Nucl Med. 2013 Oct 3.
●● Enlace al texto completo (gratuito o de pago)
1097/RLU.0b013e3182a20d78
AUTORES / AUTHORS: - Wisotzki C; Jacobsen F; Salamon J; Derlin T
INSTITUCIÓN / INSTITUTION: - From the *Department of Nuclear Medicine, daggerInstitute of Pathology, and double daggerDepartment of Diagnostic and Interventional Radiology, University Medical Center Hamburg-Eppendorf, Hamburg, Germany.
RESUMEN / SUMMARY: - We report a case of carotid chemodectoma diagnosed by In pentetreotide SPECT/CT. A 72-year-old woman with an ileocecal neuroendocrine tumor underwent whole-body In pentetreotide scintigraphy for exclusion of distant metastases. Planar scintigraphy demonstrated marked tracer uptake in the ileocecal region and intense focal tracer accumulation within the neck. SPECT/CT demonstrated a space-occupying lesion at the carotid bifurcation. Histopathological evaluation revealed carotid chemodectoma. SPECT/CT is a valuable tool for the evaluation and precise anatomical localization of tracer uptake. In addition, other benign or malignant pathologies accumulating In pentetreotide may mimic neuroendocrine tumor metastases and should be considered particularly in unusual localizations.

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[122]
TÍTULO / TITLE: - The Relationship Between Palisaded Encapsulated Neuroma and the Mucocutaneous Neuroma Seen in Multiple Endocrine Neoplasia 2b Syndrome: A Histopathologic and Immunohistochemical Study.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Am J Dermatopathol. 2013 Nov 15.
●● Enlace al texto completo (gratuito o de pago)
1097/DAD.0000000000000021
AUTORES / AUTHORS: - Misago N; Toda S; Narisawa Y
INSTITUCIÓN / INSTITUTION: - *Division of Dermatology, Department of Internal Medicine, Saga University, Saga, Japan; and daggerDepartment of Pathology, Faculty of Medicine, Saga University, Saga, Japan.
RESUMEN / SUMMARY: - : A relationship between the palisaded encapsulated neuroma (PEN) and the mucocutaneous neuroma seen in multiple endocrine neoplasia

(MEN) 2b syndrome has been noted. We experienced a case of multiple mucocutaneous neuromas including both MEN 2b type neuromas and PENs. We evaluated the histopathologic and immunohistochemical features of 48 lesions in this patient. The lesions were histopathologically classified into 3 groups: (1) MEN 2b type neuroma (18 lesions), (2) PEN (22 lesions), and (3) an intermediate form of the 2 conditions (8 lesions). The intermediate form was classified into 2 subtypes: 1 type characterized by PEN nodules made up of assembled neuroma fascicles neighboring MEN 2b type neuroma fascicles and the other type characterized by more broad nerve fascicles than those seen in typical MEN 2b type neuroma. The idea that PEN is a progressive form of MEN 2b type neuroma may be speculative. Instead, the present study suggests that the observation of hybrid MEN 2b type neuroma/PEN in association with MEN 2b type neuroma and PEN may be a characteristic finding in cases of multiple mucocutaneous neuromas. The view that MEN 2b type neuroma and PEN lie within a spectrum of the same disease entity may be an overstatement; however, the present study suggests that PEN is basically a neural hamartoma/benign neoplasm, like MEN 2b type neuroma, and that there is a close relationship between the 2 conditions in terms of their histogenesis.

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[123]

TÍTULO / TITLE: - Merkel Cell Carcinoma (Primary Neuroendocrine Carcinoma of Skin) Mimicking Basal Cell Carcinoma With Review of Different Histopathologic Features.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Am J Dermatopathol. 2013 Nov 15.

●● Enlace al texto completo (gratuito o de pago) 1097/DAD.0b013e3182a67f6f

AUTORES / AUTHORS: - Succaria F; Radfar A; Bhawan J

INSTITUCIÓN / INSTITUTION: - *Dermatopathology Section, Department of Dermatology, Boston University School of Medicine, Boston, MA; and daggerMiraca Life Sciences, Newton, MA.

RESUMEN / SUMMARY: - : Merkel cell carcinoma (MCC) is a rare but highly aggressive malignancy, which often has typical histopathologic and immunohistochemical (IHC) features. Sometimes the diagnosis is missed because of atypical histological or aberrant IHC findings. A case of MCC that showed irregular lobules of basaloid cells with keratotic areas on the initial shave biopsy is being reported. IHC showed positive staining for high-molecular weight cytokeratin but negative staining for cytokeratin 20, findings consistent with basal cell carcinoma. Subsequent excision specimen showed histopathologic features more typical of MCC. IHC still was negative for cytokeratin 20 but positive for synaptophysin. Review of the literature shows other examples of MCC with basal cell carcinoma-like features. Various other histopathologic differentiations of MCC include those that demonstrate squamous cell and eccrine carcinoma features and those that show melanocytic, lymphomatous, sarcomatous, muscular, and atypical fibroxanthoma-like features. Different histopathologic patterns and mimics of MCC are reviewed to help prevent dermatopathologists from misdiagnosing this aggressive tumor.

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[124]

TÍTULO / TITLE: - Stress-related cardiomyopathy, ventricular dysfunction, artery thrombosis: a hidden pheochromocytoma.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Am J Emerg Med. 2013 Oct 8. pii: S0735-6757(13)00636-0. doi: 10.1016/j.ajem.2013.09.034.
●● Enlace al texto completo (gratuito o de pago) 1016/j.ajem.2013.09.034
AUTORES / AUTHORS: - Battimelli A; Polito MV; Di Maio M; Poto S; Pierro L; Caggiano D; Piscione F
INSTITUCIÓN / INSTITUTION: - School of Medicine, Department of Medicine and Surgery, University of Salerno, Salerno, Italy. Electronic address: spunzilla @hotmail.it.

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[125]
TÍTULO / TITLE: - Transoral robotic surgery for atypical carcinoid tumor of the larynx.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - J Craniofac Surg. 2013 Nov;24(6):1996-9. doi: 10.1097/SCS.0b013e3182a28c2c.
●● Enlace al texto completo (gratuito o de pago)
1097/SCS.0b013e3182a28c2c
AUTORES / AUTHORS: - Muderris T; Bercin S; Sevil E; Acar B; Kiris M
INSTITUCIÓN / INSTITUTION: - From the *Department of Otorhinolaryngology, Head and Neck Surgery, Ataturk Training and Research Hospital; daggerDepartment of Otorhinolaryngology, Head and Neck Surgery, Yildirim Beyazit University Medical Faculty; double daggerDepartment of Otorhinolaryngology, Head and Neck Surgery, Ataturk Training and Research Hospital; and section signDepartment of Otorhinolaryngology, Head and Neck Surgery, Kecioren Training and Research Hospital, Ankara, Turkey.
RESUMEN / SUMMARY: - In recent years, transoral robotic surgery has been introduced as an efficient and a reliable method for excision of selected oral cavity, tongue base, and supraglottic tumors in otolaryngology. In this case report, a 39-year-old woman with a history of hoarseness and dysphagia for approximately 6 months is presented. The patient was diagnosed with atypical carcinoid tumor on the laryngeal aspect of the epiglottis, and excision of the tumor was performed through transoral robotic surgery using the robotic da Vinci surgical system, a 0-degree three-dimensional endoscope, 5-mm microinstruments compatible with the da Vinci robot, and a Feyh-Kastenbauer/Weinstein-O'Malley retractor. The mass was removed completely, and no complications occurred. The patient recovered without a need for tracheotomy. Findings of the 1-year clinical follow-up revealed no locoregional recurrence or distant metastasis. This case shows, once again, that transoral robotic surgery could be used safely and effectively regardless of pathologic diagnosis in the supraglottic region tumors.

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[126]
TÍTULO / TITLE: - Neuroendocrine tumors of the gynecologic tract: select topics.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Semin Diagn Pathol. 2013 Aug;30(3):224-33. doi: 10.1053/j.semdp.2013.06.007.

●● Enlace al texto completo (gratuito o de pago) 1053/j.semdp.2013.06.007

AUTORES / AUTHORS: - Rouzbahman M; Clarke B

INSTITUCIÓN / INSTITUTION: - Department of Pathology, Laboratory Medicine Program, University Health Network, Toronto, Ontario, Canada; Department of Laboratory Medicine and Pathobiology, University of Toronto, Toronto, Canada.

RESUMEN / SUMMARY: - Tumors of the diffuse neuroendocrine cell system (DNES) may arise in any component of the gynecologic tract, including the vulva, vagina, cervix, endometrium, and ovary. Overall such tumors in the gynecologic tract are rare, constituting only 2% of gynecologic cancers, comprising a spectrum of tumors of variable biologic potential. Due to the rarity of such tumors, pathologists experience may be limited and these may present diagnostic challenges. Currently the nomenclature employed is still that of the pulmonary classification systems, carcinoid, atypical carcinoid, small and large cell neuroendocrine carcinoma that broadly correlates to low/grade 1, intermediate/grade 2, and high grade/grade 3 of the WHO gastroenteropancreatic neuroendocrine tumors classification. Furthermore in keeping with the lung, proliferative rate is assessed based on mitotic index rather than Ki-67 staining. In this review we cover select neuroendocrine tumors of the gynecologic tract.

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[127]

TÍTULO / TITLE: - Gastroenteropancreatic neuroendocrine neoplasms: historical context and current issues.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Semin Diagn Pathol. 2013 Aug;30(3):186-96. doi: 10.1053/j.semdp.2013.06.005.

●● Enlace al texto completo (gratuito o de pago) 1053/j.semdp.2013.06.005

AUTORES / AUTHORS: - Yang Z; Tang LH; Klimstra DS

INSTITUCIÓN / INSTITUTION: - Department of Pathology, Penn State Milton S. Hershey Medical Center, Hershey, Pennsylvania.

RESUMEN / SUMMARY: - The digestive organs contain a large number of neuroendocrine cells as part of the diffuse neuroendocrine system. Neuroendocrine tumors can occur in every digestive organ. It has long been recognized that this is a diverse group of tumors with very different clinical outcomes; however, well-recognized prognostic parameters had been elusive until recently. Over the years, there have been several different classification schemes, each with different strengths and weaknesses. In an effort to standardize the classification and grading criteria for gastroenteropancreatic neuroendocrine tumors, the current World Health Organization classification includes a histologic grade based on proliferative rate (mitotic rate and Ki67 index) and a TNM stage that varies from organ to organ. The prognostic value of both the grade and stage has been validated in multiple studies. However, several issues remain, including the lack of standardized methods to assess proliferative rate, potential discrepancies between the mitotic count and the Ki67 index; intratumoral heterogeneity in proliferative rate; and the need for refinement in proliferative cut-points to define the grades. More studies are needed to further improve the classification of neuroendocrine tumors, thus guiding optimal treatment for these tumors.

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[128]

TÍTULO / TITLE: - Thyroid neoplasms of follicular cell derivation: a simplified approach.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Semin Diagn Pathol. 2013 Aug;30(3):178-85. doi: 10.1053/j.semdp.2013.06.004.

●● Enlace al texto completo (gratuito o de pago) 1053/j.semdp.2013.06.004

AUTORES / AUTHORS: - Asa SL; Mete O

INSTITUCIÓN / INSTITUTION: - Department of Pathology, Laboratory Medicine Program, University Health Network, Toronto, Ontario, Canada M5G 2C4; Department of Laboratory Medicine and Pathobiology, University of Toronto, Toronto, Ontario, Canada. Electronic address: sylvia.asa@uhn.ca.

RESUMEN / SUMMARY: - Thyroid tumors of follicular cell derivation are increasing in incidence. These lesions exhibit a spectrum of morphologic and behavioral features that provide the opportunity to understand malignant transformation and progression. Molecular data suggest that the thyroid undergoes a series of genetic alterations that account for the development of the various types of thyroid carcinoma. Our understanding of these tumors has progressed dramatically over the past 50 years and the classification has become complex and cumbersome. We provide a practical approach to clinical diagnosis and propose a simplified classification of these common neoplasms.

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[129]

TÍTULO / TITLE: - Usefulness of Negative and Weak-Diffuse Pattern of SDHB Immunostaining in Assessment of SDH Mutations in Paragangliomas and Pheochromocytomas.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Endocr Pathol. 2013 Dec;24(4):199-205. doi: 10.1007/s12022-013-9269-4.

●● Enlace al texto completo (gratuito o de pago) 1007/s12022-013-9269-4

AUTORES / AUTHORS: - Castelblanco E; Santacana M; Valls J; de Cubas A; Cascon A; Robledo M; Matias-Guiu X

INSTITUCIÓN / INSTITUTION: - Department of Pathology and Molecular Genetics, and Research Laboratory, Arnau de Vilanova Universitary Hospital, University of Lleida IRBLLEIDA, Lleida, España.

RESUMEN / SUMMARY: - This is a confirmatory study about usefulness of SDHB and SDHA immunostaining in assessment of SDH mutations in paragangliomas and pheochromocytomas. Paraganglioma/pheochromocytoma syndrome (PGL/PCC syndrome) consists of different entities, associated with germline mutations in five different genes: SDHD, SDHAF2, SDHC, SDHA and SDHB. It has been suggested that negative immunostaining of SDHB can be taken as an indicator of the presence of a mutation in one of the five SDH genes. We have performed SDHB and SDHA immunohistochemical staining in a series of paragangliomas and pheochromocytomas from 64 patients. The patients had been previously checked for mutations in SDHD, SDHC and SDHB, but also for mutation in RET and VHL. All 14 patients with SDH mutations (9 with SDHB and 5 with SDHD mutations) exhibited negative or weak-diffuse SDHB staining pattern in tumour tissue, whereas cells of the 23 RET mutated

and 8 VHL mutated tumours showed a positive SDHB immunostaining. Sixteen of the patients that did not exhibit a mutation in any gene showed positive SDHB immunostaining in tumour tissue, while only three of the patients without mutation exhibited negative staining. All patients exhibited positive pattern of SDHA immunostaining. The results confirm the value of SDHB immunohistochemical status in assessment of germline mutations in PGL/PCC syndrome.

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[130]

TÍTULO / TITLE: - Vandetanib in advanced medullary thyroid cancer: review of adverse event management strategies.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Adv Ther. 2013 Nov;30(11):945-66. doi: 10.1007/s12325-013-0069-5. Epub 2013 Nov 19.

●● Enlace al texto completo (gratuito o de pago) 1007/s12325-013-0069-5

AUTORES / AUTHORS: - Grande E; Kreissl MC; Filetti S; Newbold K; Reinisch W; Robert C; Schlumberger M; Tolstrup LK; Zamorano JL; Capdevila J

INSTITUCIÓN / INSTITUTION: - Servicio de Oncologia Medica, Hospital Ramon y Cajal, Carretera de Colmenar Viejo Km. 9.100, 28034, Madrid, España, egrande@oncologiahrc.com.

RESUMEN / SUMMARY: - INTRODUCTION: Vandetanib has recently demonstrated clinically meaningful benefits in patients with unresectable, locally advanced or metastatic medullary thyroid cancer (MTC). Given the potential for long-term vandetanib therapy in this setting, in addition to treatment for disease-related symptoms, effective management of related adverse events (AEs) is vital to ensure patient compliance and maximize clinical benefit with vandetanib therapy. METHODS: This expert meeting-based review aims to summarize published data on AEs associated with vandetanib therapy and to provide clinicians with specific practical guidance on education, monitoring, and management of toxicities induced in patients treated with vandetanib in advanced and metastatic MTC. The content of this review is based on the expert discussions from a multidisciplinary meeting held in October 2012. RESULTS: Characteristics, frequency, and risk data are outlined for a number of dermatological, cardiovascular, gastrointestinal, and general AEs related to vandetanib treatment. Preventive strategies, practical treatment suggestions, and points for clinical consideration are provided. CONCLUSIONS: Good patient and team communication is necessary for the prevention, early detection, and management of AEs of vandetanib. Physicians, nurses, and other healthcare providers play a critical role in providing AE management and patient support to optimize outcomes with vandetanib in MTC.

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[131]

TÍTULO / TITLE: - Elevated Ki-67 labeling index in ‘synchronous liver metastases’ of well differentiated enteropancreatic neuroendocrine tumor.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Pathol Int. 2013 Nov;63(11):532-8. doi: 10.1111/pin.12108.

●● Enlace al texto completo (gratuito o de pago) 1111/pin.12108

AUTORES / AUTHORS: - Zen Y; Heaton N

INSTITUCIÓN / INSTITUTION: - Histopathology Section, King's College London School of Medicine at King's College Hospital, London, UK.
RESUMEN / SUMMARY: - There is no consensus as to whether or not metastatic nodules in the liver should be biopsied for tumor grading in cases of neuroendocrine tumors with 'synchronous liver metastasis'. In this study, we compared the Ki-67 labeling index between the primary tumor and synchronous liver metastasis in 30 patients, who had received simultaneous resections. Examined tumors were of the small bowel (n = 18) or pancreas (n = 12), and G1 or G2 in primary histologic grade. In 20 patients (67%), the Ki-67 index was similar between the primary tumor and liver metastasis, but 10 (33%) showed an elevation of 3.4-14.4% in the liver, which increased the tumor grade in 4 cases. The Ki-67 elevation in the liver was more common in G2 than G1 neoplasms (P = 0.002). The size, but not number, of liver metastases was significantly larger in patients with an elevated Ki-67 index (P = 0.006). Using 40 mm as a provisional cutoff for the greatest diameter of liver metastases, the positive predictive value of this discriminator for elevated Ki-67 was 56%, and the negative predictive value was 93%. In conclusion, synchronous liver metastases can yield a higher Ki-67 labeling index than primary neuroendocrine tumours, particularly when the secondary is greater than 40 mm.

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[132]
TÍTULO / TITLE: - Ewing's Sarcoma/Primitive Neuroectodermal Tumor With Neuroendocrine Differentiation: Report of an Unusual Lung Tumor.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Int J Surg Pathol. 2013 Oct 17.
●● Enlace al texto completo (gratuito o de pago) 1177/1066896913502227
AUTORES / AUTHORS: - Barroca H; Souto Moura C; Lopes JM; Lisboa S; Teixeira MR; Damasceno M; Bastos P; Sobrinho Simoes M
RESUMEN / SUMMARY: - Ewing's sarcoma/primitive neuroectodermal tumor (PNET) has been the subject of recent reports describing morphologic variants (adamantinoma-like, large cell, spindle cell, sclerosing, clear cell, and vascular-like) of the most classic form, as well as cases displaying unusual morphologic differentiation and atypical immunohistochemical features. We report a case of an uncommon lung tumor in a 20-year-old female, morphologically and molecularly consistent with an Ewing's sarcoma/PNET tumor with foci of squamous differentiation, and peculiar expression of vimentin, high-molecular-weight keratins, p63, synaptophysin, and chromogranin. This case raises a challenging differential diagnostic problem with therapeutic implications: Should the patient be treated following the protocols for Ewing's sarcoma/PNET tumors or as for lung carcinoma with neuroendocrine features? The patient we report here was treated with neoadjuvant chemotherapy for Ewing's sarcoma/PNET according to Euro Ewing 99 study protocol followed by surgery and has no evidence of disease 15 months after the initial diagnosis. This highlights the importance of achieving the correct diagnosis of these atypical tumors using all clinical, morphological, and ancillary methods available to allow for their correct and timely treatment.

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[133]

TÍTULO / TITLE: - First Experience With Image-guided Resection of Paraganglioma.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Clin Nucl Med. 2013 Nov 7.
●● Enlace al texto completo (gratuito o de pago)
1097/RLU.0000000000000239
AUTORES / AUTHORS: - Einspieler I; Novotny A; Okur A; Essler M; Martignoni ME
INSTITUCIÓN / INSTITUTION: - From the Departments of *Nuclear Medicine and daggerSurgery, Klinikum rechts der Isar, Technische Universitat Munchen, Munich, Germany.
RESUMEN / SUMMARY: - A 32-year-old male patient showed 2 focal uptakes of I-MIBG next to the left renal vein in a diagnostic scan, corresponding to paragangliomas. An operation was indicated, and to guide resection during surgery we used the freehand SPECT system. In the operating room, using freehand SPECT, both lesions were found. The system was of additional value in planning the operative access to the region of interest and in determining the depth of 1 lesion for precise and more rapid extirpation. Furthermore, it confirmed no residues in the operating field after resection of the tumors.

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[134]
TÍTULO / TITLE: - 68Ga-Dotatate Avid Medullary Thyroid Cancer With Occult Liver Metastases.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Clin Nucl Med. 2013 Oct 3.
●● Enlace al texto completo (gratuito o de pago)
1097/RLU.0b013e3182a755e8
AUTORES / AUTHORS: - Sounness BD; Schembri GP
INSTITUCIÓN / INSTITUTION: - From the Royal North Shore Hospital, St Leonards, NSW, Australia.
RESUMEN / SUMMARY: - A 50-year-old male patient with a history of medullary thyroid cancer and extensive previous investigation including FDG PET 18 months earlier presented for a Ga-dotatate scan. A single area of abnormal uptake was identified in the right ninth rib. An MRI scan and bone scan confirmed the bony metastasis; however, the MRI of the liver demonstrated multiple liver metastases not apparent on the PET study. These remained occult despite coregistration of the MRI and PET data. The occult nature of these lesions may have been due to a similar degree of uptake in the liver and metastases.

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[135]
TÍTULO / TITLE: - 68Ga DOTATATE PET/CT in a Rare Coexistence of Pituitary Macroadenoma and Multiple Paragangliomas.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Clin Nucl Med. 2013 Oct 22.
●● Enlace al texto completo (gratuito o de pago)
1097/RLU.0b013e3182a77b78
AUTORES / AUTHORS: - Parghane RV; Agrawal K; Mittal BR; Shukla J; Bhattacharya A; Mukherjee KK

INSTITUCIÓN / INSTITUTION: - From the Departments of *Nuclear Medicine & PET, and daggerNeurosurgery, Postgraduate Institute of Medical Education and Research, Chandigarh, India.

RESUMEN / SUMMARY: - The coexistence of a pituitary neoplasm and pheochromocytoma is a rare condition, which may be another undefined variant of Multiple endocrine neoplasia (MEN) syndrome. Moreover, the coexistence of pituitary macroadenoma and multiple paragangliomas is more uncommon and only few authors have reported these findings. We are reporting the use of Ga DOTATATE PET/CT in a rare case of coexisting pituitary macroadenoma and multiple paragangliomas.

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[136]

TÍTULO / TITLE: - Autoimmune retinopathy associated with carcinoid tumour of the small bowel.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - J Clin Neurosci. 2013 Oct 11. pii: S0967-5868(13)00399-8. doi: 10.1016/j.jocn.2013.07.002.

●● Enlace al texto completo (gratuito o de pago) 1016/j.jocn.2013.07.002

AUTORES / AUTHORS: - Ogra S; Sharp D; Danesh-Meyer H

INSTITUCIÓN / INSTITUTION: - Department of Ophthalmology, Faculty of Medical and Health Sciences, The University of Auckland, Private Bag 92019, Auckland, New Zealand. Electronic address: s.ogra@auckland.ac.nz.

RESUMEN / SUMMARY: - Cancer associated retinopathy (CAR) is an immune mediated paraneoplastic condition associated with vision loss. It has been associated with a variety of systemic malignancies. The primary clinical presentation is rapid, progressive vision loss. Rod and cone dysfunction can cause other associated symptoms, such as nyctalopia. Electrophysiological testing and detection of anti-retinal antibodies are used to confirm the diagnosis. To our knowledge we describe the first patient with CAR associated with a carcinoid tumour of the gastrointestinal system. Auto-antibodies against alpha enolase and carbonic anhydrase II were detected with western blotting. Electroretinogram findings were consistent with rod and cone dysfunction.

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[137]

TÍTULO / TITLE: - Pancreatic neuroendocrine tumors.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Curr Probl Surg. 2013 Nov;50(11):509-45. doi: 10.1067/j.cpsurg.2013.08.001.

●● Enlace al texto completo (gratuito o de pago) 1067/j.cpsurg.2013.08.001

AUTORES / AUTHORS: - Krampitz GW; Norton JA

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[138]

TÍTULO / TITLE: - Non-pheochromocytoma (PCC)/paraganglioma (PGL) tumors in patients with succinate dehydrogenase-related PCC-PGL syndromes: a clinicopathological and molecular analysis.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Eur J Endocrinol. 2013 Nov 22;170(1):1-12. doi: 10.1530/EJE-13-0623. Print 2014.

●● Enlace al texto completo (gratuito o de pago) 1530/EJE-13-0623

AUTORES / AUTHORS: - Papathomas TG; Gaal J; Corssmit EP; Oudijk L; Korpershoek E; Heimdal K; Bayley JP; Morreau H; van Dooren M; Papaspyrou K; Schreiner T; Hansen T; Andresen PA; Restuccia DF; van Kessel I; van Leenders GJ; Kros JM; Looijenga LH; Hofland LJ; Mann W; van Nederveen FH; Mete O; Asa SL; de Krijger RR; Dinjens WN

INSTITUCIÓN / INSTITUTION: - Department of Pathology, Josephine Nefkens Institute, Erasmus MC, University Medical Center, PO Box 2040, 3000 CA Rotterdam, The Netherlands.

RESUMEN / SUMMARY: - OBJECTIVE: Although the succinate dehydrogenase (SDH)-related tumor spectrum has been recently expanded, there are only rare reports of non-pheochromocytoma/paraganglioma tumors in SDHx-mutated patients. Therefore, questions still remain unresolved concerning the aforementioned tumors with regard to their pathogenesis, clinicopathological phenotype, and even causal relatedness to SDHx mutations. Absence of SDHB expression in tumors derived from tissues susceptible to SDH deficiency is not fully elucidated. DESIGN AND METHODS: Three unrelated SDHD patients, two with pituitary adenoma (PA) and one with papillary thyroid carcinoma (PTC), and three SDHB patients affected by renal cell carcinomas (RCCs) were identified from four European centers. SDHA/SDHB immunohistochemistry (IHC), SDHx mutation analysis, and loss of heterozygosity analysis of the involved SDHx gene were performed on all tumors. A cohort of 348 tumors of unknown SDHx mutational status, including renal tumors, PTCs, PAs, neuroblastic tumors, seminomas, and adenomatoid tumors, was investigated by SDHB IHC. RESULTS: Of the six index patients, all RCCs and one PA displayed SDHB immunonegativity in contrast to the other PA and PTC. All immunonegative tumors demonstrated loss of the WT allele, indicating bi-allelic inactivation of the germline mutated gene. Of 348 tumors, one clear cell RCC exhibited partial loss of SDHB expression. CONCLUSIONS: These findings strengthen the etiological association of SDHx genes with pituitary neoplasia and provide evidence against a link between PTC and SDHx mutations. Somatic deletions seem to constitute the second hit in SDHB-related renal neoplasia, while SDHx alterations do not appear to be primary drivers in sporadic tumorigenesis from tissues affected by SDH deficiency.

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[139]

TÍTULO / TITLE: - Long term prognosis of patients with pediatric pheochromocytoma.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Endocr Relat Cancer. 2013 Oct 29.

●● Enlace al texto completo (gratuito o de pago) 1530/ERC-13-0415

AUTORES / AUTHORS: - Bausch B; Wellner U; Bausch D; Schiavi F; Barontini M; Sanso G; Walz MK; Peczkowska M; Weryha G; Dall'igna P; Cecchetto G; Bisogno G; Moeller L; Bockenhauer D; Patocs A; Racz K; Zabolotnyi D; Yaremchuk S; Dzivite-Krisane I; Castinetti F; Taieb D; Malinoc A; von Dobschuetz E; Roessler J; Schmid KW; Opocher G; Eng C; Neumann HP

INSTITUCIÓN / INSTITUTION: - B Bausch, Innere Medizin 2, Universitatsklinikum Freiburg, Freiburg, 79106, Germany.

RESUMEN / SUMMARY: - A third of patients with paraganglial tumors, pheochromocytoma and paraganglioma, carry germline mutations in one of the susceptibility genes, RET, VHL, NF1, SDHAF2, SDHA, SDHB, SDHC, SDHD, TMEM127 and MAX. Despite increasing importance, data for long-term prognosis are scarce in pediatric presentations. The European-American-Pheochromocytoma-Paraganglioma-Registry, with a total of 2001 patients with confirmed paraganglial tumors, was the platform for this study. Molecular genetic and phenotypic classification and assessment of gene-specific long-term outcome with second and/or malignant paraganglial tumors and life expectancy was performed in patients diagnosed <18 years. Of 177 eligible registrants, 80% had mutations, 49% VHL, 15% SDHB, 10% SDHD, 4% NF1 and one patient each in RET, SDHA and SDHC. A second primary paraganglial tumor developed in 38% with increasing frequency over time, reaching 50% at 30 years after initial diagnosis. Their prevalence was associated with hereditary disease (p=0.001), particularly in VHL and SDHD mutation carriers (VHL vs others, p=0.001, SDHD vs others, p=0.042). A total of 16 (9%) patients with hereditary disease had malignant tumors, 10 at initial diagnosis and another 6 during follow-up. The highest prevalence was associated with SDHB (SDHB vs others, p<0.001). Eight patients died (5%), all of whom had germline mutations. Mean life expectancy was 62 years with hereditary disease. Hereditary disease and the underlying germline mutation define the long-term prognosis of pediatric patients in terms of prevalence and time of second primaries, malignant transformation and survival. Based on these data, gene-adjusted, specific surveillance guidelines can help effective preventive medicine.

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[140]
TÍTULO / TITLE: - How smart is peptide receptor radionuclide therapy of neuroendocrine tumors especially in the salvage setting? The clinician's perspective.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Eur J Nucl Med Mol Imaging. 2013 Nov 6.
●● Enlace al texto completo (gratuito o de pago) 1007/s00259-013-2593-6
AUTORES / AUTHORS: - Prasad V; Brenner W; Modlin IM
INSTITUCIÓN / INSTITUTION: - Department of Nuclear Medicine, Campus Virchow-Klinikum, Charite University Hospital, Berlin, Germany, vikas.prasad@charite.de.

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[141]
TÍTULO / TITLE: - A founder SDHB mutation in Portuguese paraganglioma patients.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Endocr Relat Cancer. 2013 Nov 4;20(6):L23-6. doi: 10.1530/ERC-12-0399. Print 2013.
●● Enlace al texto completo (gratuito o de pago) 1530/ERC-12-0399
AUTORES / AUTHORS: - Martins RG; Nunes JB; Maximo V; Soares P; Peixoto J; Catarino T; Rito T; Soares P; Pereira L; Sobrinho-Simoes M; Santos AP; Couto J; Henrique R; Matos-Loureiro J; Dias P; Torres I; Lima J
INSTITUCIÓN / INSTITUTION: - IPATIMUP (Institute of Pathology and Molecular Immunology of the University of Porto), Rua Dr Roberto Frias s/n, 4200-465, Porto, Portugal Medical Faculty of the University of Porto, Porto, Portugal Department of Endocrinology, Portuguese Oncology Institute, Porto, Portugal Department of

Pathology, Hospital S. Joao, Porto, Portugal Department of Pathology, Portuguese Oncology Institute, Porto, Portugal.

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[142]
TÍTULO / TITLE: - Peptide receptor radionuclide therapy for neuroendocrine tumours: standardized and randomized, or personalized?
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Eur J Nucl Med Mol Imaging. 2013 Nov 13.
●● Enlace al texto completo (gratuito o de pago) 1007/s00259-013-2621-6
AUTORES / AUTHORS: - Hofman MS; Hicks RJ
INSTITUCIÓN / INSTITUTION: - Molecular Imaging, Centre for Cancer Imaging, Peter MacCallum Cancer Centre, Melbourne, Australia, michael.hofman@petermac.org.

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[143]
TÍTULO / TITLE: - Gastric granular cell tumour clinically mimicking carcinoid tumour treated by endoscopic submucosal dissection.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - ANZ J Surg. 2013 Oct 31. doi: 10.1111/ans.12355.
●● Enlace al texto completo (gratuito o de pago) 1111/ans.12355
AUTORES / AUTHORS: - Min KW; Lee KG; Han H; Jang SM; Paik SS
INSTITUCIÓN / INSTITUTION: - Department of Pathology, Konkuk University School of Medicine, Seoul, Korea.

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[144]
TÍTULO / TITLE: - Treatment with tandem [Y]DOTA-TATE and [Lu]DOTA-TATE of neuroendocrine tumours refractory to conventional therapy.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Eur J Nucl Med Mol Imaging. 2013 Nov 14.
●● Enlace al texto completo (gratuito o de pago) 1007/s00259-013-2578-5
AUTORES / AUTHORS: - Seregni E; Maccauro M; Chiesa C; Mariani L; Pascali C; Mazzaferro V; De Braud F; Buzzoni R; Milione M; Lorenzoni A; Bogni A; Coliva A; Vullo SL; Bombardieri E
INSTITUCIÓN / INSTITUTION: - Nuclear Medicine, Fondazione IRCCS Istituto Nazionale Tumori, Via G Venezian 1, Milan, Italy, ettore.seregni@istitutotumori.mi.it.
RESUMEN / SUMMARY: - PURPOSE: Peptide receptor radionuclide therapy (PRRT) with radiolabelled somatostatin analogues has been demonstrated to be an effective therapeutic option in patients with disseminated neuroendocrine tumours (NET). Treatment with tandem [90Y]DOTA-TATE and [177Lu]DOTA-TATE may improve the efficacy of PRRT without increasing the toxicity. In a phase II study we evaluated the feasibility of combined PPRT with a high-energy beta emitter (90Y) and a medium-energy beta/gamma emitter (177Lu) in patients with metastatic NET refractory to conventional therapy. METHODS: A group of 26 patients with metastatic NET were treated with four therapeutic cycles of alternating [177Lu]DOTA-TATE (5.55 GBq) and [90Y]DOTA-TATE (2.6 GBq). A dosimetric evaluation was carried out after administration of [177Lu]DOTA-TATE to calculate the absorbed doses in healthy

organs. The acute and long-term toxicities of repeated treatment were analysed. PRRT efficacy was evaluated according to RECIST. RESULTS: Administration of tandem [90Y]DOTA-TATE and [177Lu]DOTA-TATE induced objective responses in 42.3 % of patients with metastatic NET with a median progression-free survival longer than 24 months. Of patients with pretreatment carcinoid syndrome, 90 % showed a symptomatic response or a reduction in tumour-associated pain. The cumulative biologically effective doses (BED) were below the toxicity limit in the majority of patients, in the absence of renal function impairment CONCLUSION: The results of our study indicates that combined [90Y]DOTA-TATE and [177Lu]DOTA-TATE therapy is a feasible and effective therapeutic option in NET refractory to conventional therapy. Furthermore, the absence of kidney damage and the evaluated cumulative BEDs suggest that increasing the number of tandem administrations is an interesting approach.

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[145]
TÍTULO / TITLE: - Radiolabelled somatostatin analogue treatment in gastroenteropancreatic neuroendocrine tumours: factors associated with response and suggestions for therapeutic sequence: response to comments by Ezziddin et al.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Eur J Nucl Med Mol Imaging. 2014 Jan;41(1):176-7. doi: 10.1007/s00259-013-2603-8.
●● Enlace al texto completo (gratuito o de pago) 1007/s00259-013-2603-8
AUTORES / AUTHORS: - Campana D; Delle Fave G; Falconi M; Tommassetti P
INSTITUCIÓN / INSTITUTION: - Department of Medical and Surgical Sciences, S. Orsola-Malpighi Hospital, University of Bologna, Bologna, Italy, davide.campana@unibo.it.

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[146]
TÍTULO / TITLE: - Comment on Campana et al.: 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. 2014 Jan;41(1):174-5. doi: 10.1007/s00259-013-2599-0. Epub 2013 Nov 6.
●● Enlace al texto completo (gratuito o de pago) 1007/s00259-013-2599-0
AUTORES / AUTHORS: - Ezziddin S; Sabet A; Yong-Hing CJ; Biersack HJ
INSTITUCIÓN / INSTITUTION: - Department of Nuclear Medicine, University Hospital Bonn, Sigmund-Freud-Str. 25, 53105, Bonn, Germany, samer.ezziddin@ukb.uni-bonn.de.

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[147]
TÍTULO / TITLE: - Clinical Outcomes for Neuroendocrine Tumors of the Duodenum and Ampulla of Vater:A Population-Based Study.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - J Gastrointest Surg. 2013 Oct 10.

●● Enlace al texto completo (gratuito o de pago) 1007/s11605-013-2365-4

AUTORES / AUTHORS: - Randle RW; Ahmed S; Newman NA; Clark CJ

INSTITUCIÓN / INSTITUTION: - Department of General Surgery, Wake Forest Baptist Health, Medical Center Blvd, Winston-Salem, NC, 27157, USA.

RESUMEN / SUMMARY: - BACKGROUND: Previous case series report that neuroendocrine tumors (NETs) of the ampulla of Vater have worse overall survival (OS) than NETs in the duodenum. We aimed to compare the OS of patients with ampullary NETs to patients with duodenal NETs. METHODS: This retrospective comparative cohort study used the Surveillance, Epidemiology, and End Results (SEER) registry from 1988 to 2009. OS was evaluated using Kaplan-Meier estimates and Cox proportional hazard regression. RESULTS: Ampullary NETs (n = 120) were larger (median size 18 vs. 10 mm, p < 0.001), higher grade (poorly and undifferentiated tumor 42 % vs. 12 %, p < 0.001), higher SEER historic stage (distant metastasis 16 % vs. 7 %, p < 0.001), and more often resected (78 % vs. 60 %, p < 0.001) than duodenal NETs (n = 1,360). Median OS was significantly worse for patients with ampullary NETs than with duodenal NETs (98 vs. 143 months, p = 0.037). Local resection was performed for 50.5 % of the resected ampullary NETs and resulted in similar OS compared to locally resected duodenal NETs (HR 1.37, 95 % CI 0.76-2.48, p = 0.291). CONCLUSIONS: While ampullary NETs are more advanced at presentation and have worse OS than duodenal NETs, long-term survival is possible with proximal small bowel NETs. For locally resected NETs, OS is similar between ampullary and duodenal NETs.

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[148]

TÍTULO / TITLE: - Are targeted therapies a consideration in poorly differentiated neuroendocrine tumors?

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Oncologist. 2013;18(11):1239. doi: 10.1634/theoncologist.2013-0206.

●● Enlace al texto completo (gratuito o de pago) 1634/theoncologist.2013-0206

AUTORES / AUTHORS: - Sorscher S

INSTITUCIÓN / INSTITUTION: - Department of Oncology, Washington University School of Medicine, St. Louis, Missouri, USA.

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[149]

TÍTULO / TITLE: - Nonfunctional Pancreatic Neuroendocrine Tumors <2 cm on Preoperative Imaging are Associated with a Low Incidence of Nodal Metastasis and an Excellent Overall Survival.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - J Gastrointest Surg. 2013 Dec;17(12):2105-13. doi: 10.1007/s11605-013-2360-9. Epub 2013 Oct 8.

●● Enlace al texto completo (gratuito o de pago) 1007/s11605-013-2360-9

AUTORES / AUTHORS: - Toste PA; Kadera BE; Tatishchev SF; Dawson DW; Clerkin BM; Muthusamy R; Watson R; Tomlinson JS; Hines OJ; Reber HA; Donahue TR

INSTITUCIÓN / INSTITUTION: - Division of General Surgery, Department of Surgery, David Geffen School of Medicine at University of California, Los Angeles (UCLA), Los Angeles, CA, USA, ptoste@mednet.ucla.edu.

RESUMEN / SUMMARY: - BACKGROUND: The optimal surgical management of small nonfunctional pancreatic neuroendocrine tumors (NF-PNETs) remains controversial. We sought to identify (1) clinicopathologic factors associated with survival in NF-PNETs and (2) preoperative tumor characteristics that can be used to determine which lesions require resection and lymph node (LN) harvest. METHODS: The records of all 116 patients who underwent resection for NF-PNETs between 1989 and 2012 were reviewed retrospectively. Preoperative factors, operative data, pathology, surgical morbidity, and survival were analyzed. RESULTS: The overall 5- and 10-year survival rates were 83.9 and 72.8 %, respectively. Negative LNs (p = 0.005), G1 or G2 histology (p = 0.033), and age <60 years (p = 0.002) correlated with better survival on multivariate analysis. The 10-year survival rate was 86.6 % for LN-negative patients (n = 73) and 34.1 % for LN-positive patients (n = 32). Tumor size >/=2 cm on preoperative imaging predicted nodal positivity with a sensitivity of 93.8 %. Positive LNs were found in 38.5 % of tumors >/=2 cm compared to only 7.4 % of tumors <2 cm. CONCLUSIONS: LN status, a marker of systemic disease, was a highly significant predictor of survival in this series. Tumor size on preoperative imaging was predictive of nodal disease. Thus, it is reasonable to consider parenchyma-sparing resection or even close observation for NF-PNETs <2 cm.

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[150]

TÍTULO / TITLE: - Somatostatin-based radiopeptide therapy with [Lu-DOTA]-TOC versus [Y-DOTA]-TOC in neuroendocrine tumours.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Eur J Nucl Med Mol Imaging. 2013 Oct 2.

●● Enlace al texto completo (gratuito o de pago) 1007/s00259-013-2559-8

AUTORES / AUTHORS: - Romer A; Seiler D; Marincek N; Brunner P; Koller MT; Ng QK; Maecke HR; Muller-Brand J; Rochlitz C; Briel M; Schindler C; Walter MA

INSTITUCIÓN / INSTITUTION: - Institute of Nuclear Medicine, University Hospital Basel, Basel, Switzerland.

RESUMEN / SUMMARY: - PURPOSE: Somatostatin-based radiopeptide treatment is generally performed using the beta-emitting radionuclides 90Y or 177Lu. The present study aimed at comparing benefits and harms of both therapeutic approaches. METHODS: In a comparative cohort study, patients with advanced neuroendocrine tumours underwent repeated cycles of [90Y-DOTA]-TOC or [177Lu-DOTA]-TOC until progression of disease or permanent adverse events. Multivariable Cox regression and competing risks regression were employed to examine predictors of survival and adverse events for both treatment groups. RESULTS: Overall, 910 patients underwent 1,804 cycles of [90Y-DOTA]-TOC and 141 patients underwent 259 cycles of [177Lu-DOTA]-TOC. The median survival after [177Lu-DOTA]-TOC and after [90Y-DOTA]-TOC was comparable (45.5 months versus 35.9 months, hazard ratio 0.91, 95 % confidence interval 0.63-1.30, p = 0.49). Subgroup analyses revealed a significantly longer survival for [177Lu-DOTA]-TOC over [90Y-DOTA]-TOC in patients with low tumour uptake, solitary lesions and extra-hepatic lesions. The rate of severe transient haematotoxicities was lower after [177Lu-DOTA]-TOC treatment (1.4 vs 10.1 %, p =

0.001), while the rate of severe permanent renal toxicities was similar in both treatment groups (9.2 vs 7.8 %, p = 0.32). CONCLUSION: The present results revealed no difference in median overall survival after [177Lu-DOTA]-TOC and [90Y-DOTA]-TOC. Furthermore, [177Lu-DOTA]-TOC was less haematotoxic than [90Y-DOTA]-TOC.

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[151]

TÍTULO / TITLE: - Cardiorespiratory crisis at the end of pregnancy: a case of pheochromocytoma.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Middle East J Anesthesiol. 2013 Jun;22(2):195-202.

AUTORES / AUTHORS: - Haddad S; Al-Raiy B; Madkhali A; Al-Qahtani S; Al-Sultan M; Arabi Y

INSTITUCIÓN / INSTITUTION: - Intensive Care Department, MC 1425, Surgical Intensive Care Unit, King Abdulaziz Medical City, Riyadh, Kingdom of Saudi Arabia. haddads55@yahoo.com

RESUMEN / SUMMARY: - Pheochromocytoma during pregnancy is extremely rare. Its clinical manifestation includes hypertension with various clinical presentations, possibly resembling those of pregnancy-induced hypertension. The real challenge for clinicians is differentiating pheochromocytoma from other causes of hypertension (preeclampsia, gestational hypertension, and pre-existing or essential hypertension), from other cause of pulmonary edema (preeclampsia, peripartum cardiomyopathy, stress or Takotsubo cardiomyopathy, pre-existing cardiac disease [mitral stenosis], and high doses betamimetics), and from other causes of cardiovascular collapse (pulmonary embolism, and amniotic fluid embolism). Although, several cases of pheochromocytoma during pregnancy have been published, fetal and maternal mortalities due to undiagnosed cases are still reported. We report a case of a patient whose delivery by cesarean section was complicated by severe hemodynamic instability resulting in a cardiac arrest. Later on, pheochromocytoma was suspected based on computed tomography (CT) scan findings. Diagnosis was confirmed with special biochemical investigations that showed markedly elevated catecholamines in urine and metanephrines in serum, and later by histopathology of the excised left adrenal mass. This case illustrates the difficulty of diagnosing pheochromocytoma in pregnancy and raises the awareness to when this rare disease should be suspected.

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[152]

TÍTULO / TITLE: - Netazepide, a gastrin receptor antagonist, normalises tumour biomarkers and causes regression of type 1 gastric neuroendocrine tumours in a nonrandomised trial of patients with chronic atrophic gastritis.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - PLoS One. 2013 Oct 1;8(10):e76462. doi: 10.1371/journal.pone.0076462.

●● Enlace al texto completo (gratuito o de pago) 1371/journal.pone.0076462

AUTORES / AUTHORS: - Moore AR; Boyce M; Steele IA; Campbell F; Varro A; Pritchard DM

INSTITUCIÓN / INSTITUTION: - Department of Gastroenterology, Institute of Translational Medicine, University of Liverpool, Liverpool, United Kingdom ;

Department of Cellular and Molecular Physiology, Institute of Translational Medicine, University of Liverpool, Liverpool, United Kingdom.

RESUMEN / SUMMARY: - INTRODUCTION: Autoimmune chronic atrophic gastritis (CAG) causes hypochlorhydria and hypergastrinaemia, which can lead to enterochromaffin-like (ECL) cell hyperplasia and gastric neuroendocrine tumours (type 1 gastric NETs). Most behave indolently, but some larger tumours metastasise. Antrectomy, which removes the source of the hypergastrinaemia, usually causes tumour regression. Non-clinical and healthy-subject studies have shown that netazepide (YF476) is a potent, highly selective and orally-active gastrin/CCK-2 receptor antagonist. Also, it is effective in animal models of ECL-cell tumours induced by hypergastrinaemia. AIM: To assess the effect of netazepide on tumour biomarkers, number and size in patients with type I gastric NETs. METHODS: We studied 8 patients with multiple tumours and raised circulating gastrin and chromogranin A (CgA) concentrations in an open trial of oral netazepide for 12 weeks, with follow-up 12 weeks later. At 0, 6, 12 and 24 weeks, we carried out gastroscopy, counted and measured tumours, and took biopsies to assess abundances of several ECL-cell constituents. At 0, 3, 6, 9, 12 and 24 weeks, we measured circulating gastrin and CgA and assessed safety and tolerability. RESULTS: Netazepide was safe and well tolerated. Abundances of CgA ($p<0.05$), histidine decarboxylase ($p<0.05$) and matrix metalloproteinase-7($p<0.10$) were reduced at 6 and 12 weeks, but were raised again at follow-up. Likewise, plasma CgA was reduced at 3 weeks ($p<0.01$), remained so until 12 weeks, but was raised again at follow-up. Tumours were fewer and the size of the largest one was smaller ($p<0.05$) at 12 weeks, and remained so at follow-up. Serum gastrin was unaffected. CONCLUSION: The reduction in abundances, plasma CgA, and tumour number and size by netazepide show that type 1 NETs are gastrin-dependent tumours. Failure of netazepide to increase serum gastrin further is consistent with achlorhydria. Netazepide is a potential new treatment for type 1 NETs. Longer, controlled trials are justified. TRIAL REGISTRATION: European Union EudraCT database 2007-002916-24 https://www.clinicaltrialsregister.eu/ctr-search/search?query=2007-002916-24Clini calTrials.gov NCT01339169 http://clinicaltrials.gov/ct2/show/NCT01339169?term=yf476&rank=5.

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[153]

TÍTULO / TITLE: - Diagnostic accuracy of Ga-DOTANOC PET/CT imaging in pheochromocytoma.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Eur J Nucl Med Mol Imaging. 2013 Oct 25.

●● Enlace al texto completo (gratuito o de pago) 1007/s00259-013-2598-1

AUTORES / AUTHORS: - Sharma P; Dhull VS; Arora S; Gupta P; Kumar R; Durgapal P; Malhotra A; Chumber S; Ammini AC; Kumar R; Bal C

INSTITUCIÓN / INSTITUTION: - Department of Nuclear Medicine, All India Institute of Medical Sciences, Ansari Nagar, New Delhi, 110029, India.

RESUMEN / SUMMARY: - PURPOSE: The purpose of the present study was to evaluate the diagnostic accuracy of 68Ga-DOTANOC positron emission tomography (PET)/CT in patients with suspicion of pheochromocytoma. METHODS: Data of 62 patients [age 34.3 +/- 16.1 years, 14 with multiple endocrine neoplasia type 2 (MEN2)] with clinical/biochemical suspicion of pheochromocytoma and suspicious adrenal lesion on contrast CT (n = 70), who had undergone 68Ga-DOTANOC PET/CT, were

retrospectively analyzed. PET/CT images were analyzed visually as well as semiquantitatively, with measurement of maximum standardized uptake value (SUVmax), SUVmean, SUVmax/SUVliver, and SUVmean/SUVliver. Results of PET/CT were compared with 131I-metaiodobenzylguanidine (MIBG) imaging, which was available in 40 patients (45 lesions). Histopathology and/or imaging/clinical/biochemical follow-up (minimum 6 months) was used as reference standard. RESULTS: The sensitivity, specificity, and accuracy of 68Ga-DOTANOC PET/CT was 90.4, 85, and 88.7 %, respectively, on patient-based analysis and 92, 85, and 90 %, respectively, on lesion-based analysis. 68Ga-DOTANOC PET/CT showed 100 % accuracy in patients with MEN2 syndrome and malignant pheochromocytoma. On direct comparison, lesion-based accuracy of 68Ga-DOTANOC PET/CT for pheochromocytoma was significantly higher than 131I-MIBG imaging (91.1 vs 66.6 %, p = 0.035). SUVmax was higher for pheochromocytomas than other adrenal lesions (p = 0.005), MEN2-associated vs sporadic pheochromocytoma (p = 0.012), but no difference was seen between benign vs malignant pheochromocytoma (p = 0.269). CONCLUSION: 68Ga-DOTANOC PET/CT shows high diagnostic accuracy in patients with suspicion of pheochromocytoma and is superior to 131I-MIBG imaging for this purpose. Best results of 68Ga-DOTANOC PET/CT are seen in patients with MEN2-associated and malignant pheochromocytoma.

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[154]

TÍTULO / TITLE: - Altered MENIN expression disrupts the MAFA differentiation pathway in insulinoma.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Endocr Relat Cancer. 2013 Oct 24;20(6):833-48. doi: 10.1530/ERC-13-0164. Print 2013 Dec.

●● Enlace al texto completo (gratuito o de pago) 1530/ERC-13-0164

AUTORES / AUTHORS: - Hamze Z; Vercherat C; Bernigaud-Lacheretz A; Bazzi W; Bonnavion R; Lu J; Calender A; Pouponnot C; Bertolino P; Roche C; Stein R; Scoazec JY; Zhang CX; Cordier-Bussat M

INSTITUCIÓN / INSTITUTION: - INSERM U1052/CNRS UMR5286/Universite de Lyon, Lyon1 UMR-S1052, Cancer Research Center of Lyon, Lyon F-69008, France Service de Genetique Moleculaire et Clinique, Hospices Civils de Lyon, Hopital Edouard Herriot, Lyon F-69437, France UMR 3347/CNRS, U1021/INSERM, Institut Curie, Orsay F-91405, France Service Central d'Anatomie et Cytologie Pathologiques, Hospices Civils de Lyon, Hopital Edouard Herriot, Lyon F-69437, France Department of Molecular Physiology and Biophysics, Vanderbilt University Medical Center, Nashville, Tennessee 37232, USA.

RESUMEN / SUMMARY: - The protein MENIN is the product of the multiple endocrine neoplasia type I (MEN1) gene. Altered MENIN expression is one of the few events that are clearly associated with foregut neuroendocrine tumours (NETs), classical oncogenes or tumour suppressors being not involved. One of the current challenges is to understand how alteration of MENIN expression contributes to the development of these tumours. We hypothesised that MENIN might regulate factors maintaining endocrine-differentiated functions. We chose the insulinoma model, a paradigmatic example of well-differentiated pancreatic NETs, to study whether MENIN interferes with the expression of v-MAF musculoaponeurotic fibrosarcoma oncogene homologue A

(MAFA), a master glucose-dependent transcription factor in differentiated beta-cells. Immunohistochemical analysis of a series of human insulinomas revealed a correlated decrease in both MENIN and MAFA. Decreased MAFA expression resulting from targeted Men1 ablation was also consistently observed in mouse insulinomas. In vitro analyses using insulinoma cell lines showed that MENIN regulated MAFA protein and mRNA levels, and bound to Mafa promoter sequences. MENIN knockdown concomitantly decreased mRNA expression of both Mafa and beta-cell differentiation markers (Ins1/2, Gck, Slc2a2 and Pdx1) and, in parallel, increased the proliferation rate of tumours as measured by bromodeoxyuridine incorporation. Interestingly, MAFA knockdown alone also increased proliferation rate but did not affect the expression of candidate proliferation genes regulated by MENIN. Finally, MENIN variants with missense mutations detected in patients with MEN1 lost the WT MENIN properties to regulate MAFA. Together, our findings unveil a previously unsuspected MENIN/MAFA connection regarding control of the beta-cell differentiation/proliferation balance, which could contribute to tumorigenesis.

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[155]

TÍTULO / TITLE: - Merkel cell carcinoma of the hand and upper extremity: Current trends and outcomes.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - J Plast Reconstr Aesthet Surg. 2013 Oct 2. pii: S1748-6815(13)00554-8. doi: 10.1016/j.bjps.2013.09.030.

●● Enlace al texto completo (gratuito o de pago) 1016/j.bjps.2013.09.030

AUTORES / AUTHORS: - Soltani AM; Allan BJ; Best MJ; Panthaki ZJ; Thaller SR

INSTITUCIÓN / INSTITUTION: - Department of Surgery, Division of Plastic, Aesthetic, and Reconstructive Surgery, University of Miami, Miller School of Medicine, USA. Electronic address: asoltani8@gmail.com.

RESUMEN / SUMMARY: - Merkel cell carcinomas represent an uncommon yet aggressive skin cancer. We sought to identify changes in incidence and predictors of outcomes and survival of patients with Merkel cell carcinomas of the hand and upper extremity. The Surveillance, Epidemiology and End Results database was used to identify all patients with Merkel cell carcinomas of the skin located specifically in the hand and upper extremity. Overall, 5211 cases were identified in the period from 1986 through 2009. The age-adjusted incidence of Merkel cell carcinoma of the hand and upper extremity increased from 0.02 cases per 100,000 in 1986 to 0.14 cases per 100,000 in 2009. The mean age of these patients was 75 years and positive regional lymph nodes were identified in 33%. Rate of metastasis was 4.1%. Overall survival for the study cohort was 49%. Multivariate analysis identified tumor size >5 cm, positive regional lymph nodes and metastasis at diagnosis as independent predictors of mortality. The incidence of Merkel cell carcinomas has increased substantially over the study period. Overall survival for Merkel cell carcinomas of the body and hand and upper extremity is related to tumor size and extent of disease at time of clinical presentation. Merkel cell carcinomas of the hand and upper extremity tend to be diagnosed at an earlier stage with lower rates of regional and systemic spread, and subsequently have a higher overall survival rate.

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[156]
TÍTULO / TITLE: - Thyroid Carcinoma Metastases to Axillary Lymph Nodes: Report of Two Rare Cases of Papillary and Medullary Thyroid Carcinoma and a Review of the Literature.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Endocr Pract. 2013 Nov 18:1-10.
●● Enlace al texto completo (gratuito o de pago) 4158/EP13339.CR
AUTORES / AUTHORS: - Cummings AL; Goldfarb M
INSTITUCIÓN / INSTITUTION: - Division of Breast/Soft Tissue and Endocrine Surgery, Keck School of Medicine of the University of Southern California, 1510 San Pablo Street, Los Angeles, 90033 CA, USAM.
RESUMEN / SUMMARY: - Background: Axillary lymph nodes (ALNs) are a rare manifestation of thyroid carcinoma; only 16 cases are in the published literature. This study adds two additional patients, one differentiated (DTC) papillary (PTC) and one medullary (MTC) carcinoma, and reviews the limited information on this topic.Methods / Case Presentation: (1) A 56-year-old female diagnosed in 2004 with stage IV PTC (lung and rib metastases) underwent total thyroidectomy (TTx) and received radioiodine and antineoplastics for progression in the lung, liver and chest wall (2008-2011). In 2012, screening mammography detected multiple axillary masses corresponding to ALNs on magnetic resonance imaging. After fine needle biopsy demonstrated metastatic PTC, the patient underwent right ALN dissection and is currently with stable disease. (2) A 59-year-old male diagnosed in 2011 with stage III MTC underwent TTx and bilateral modified LN dissection for cervical LN metastases. Three months later, a positron emission tomography scan revealed hypermetabolic ALNs confirmed by excisional biopsy as metastatic MTC. A completion left ALN dissection and supraclavicular LN excision was performed and the patient is currently with stable diseaseResults: Sixteen reports of ALN metastases from thyroid cancer exist in the literature: 11 PTC, 2 mucoepidermoid carcinoma variants, and 1 each of follicular, MTC, and poorly differentiated mucinproducing adenocarcinoma. This study reports the second case of MTC metastatic to ALNs.Conclusion: Thyroid cancer ALN metastases are rare representations of distant metastatic disease. Complete surgical resection remains the standard of care for all MTC metastases and for DTC patients with local symptoms or otherwise stable disease that can tolerate the operation.

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[157]
TÍTULO / TITLE: - Chromophobe renal cell carcinoma: a morphologic and immunohistochemical study of 45 cases.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Ann Diagn Pathol. 2013 Dec;17(6):508-13. doi: 10.1016/j.anndiagpath.2013.06.005. Epub 2013 Oct 2.
●● Enlace al texto completo (gratuito o de pago)
1016/j.anndiagpath.2013.06.005
AUTORES / AUTHORS: - Din NU; Fatima S; Ahmad Z
INSTITUCIÓN / INSTITUTION: - Histopathology, Department of Pathology and Microbiology, Aga Khan University Hospital, Karachi, Pakistan. Electronic address: nd176@yahoo.com.

RESUMEN / SUMMARY: - The aim of this study was to evaluate the morphological spectrum of chromophobe renal cell carcinoma (CRCC) and diagnostic utility of a panel of three immunohistochemical stains. All cases of CRCC reported between 2002 and 2012 in the Section of Histopathology, Aga Khan University Hospital, were retrieved. A total of 45 cases were identified. Slides were reviewed and immunohistochemical stains (CK7, CD117, and vimentin) were performed. Ages ranged from 18 to 90years (mean, 48.5years). Male-to-female ratio was 0.8:1. The tumor was located in the left kidney in 24 patients and the right kidney in 20 patients. The tumor size ranged from 3.5 to 22cm (mean 10cm). Histologically, 4 were classic, 22 were eosinophilic, 16 were mixed, and 3 were sarcomatoid type. Morphologic patterns included broad alveolar, solid, nested, tubular, tubulocystic, trabecular, papillary, and microglandular. Binucleation and perinuclear halos were seen in all cases. Nuclear grooves and pseudoinclusions were seen in 17 and 6 cases, respectively. Multinucleated cells were seen in 19 cases. Mitoses ranged from 1 to 11/10 HPFs (mean 3/10 HPFs). Hyalinized stroma was seen in 38 cases and calcification in 26 cases. Necrosis was seen in 18 cases. Palisading of smaller cells around the broad alveolar pattern was noted in 5 cases. The Furhman's nuclear grade was I (11), II (26), III (5), and IV (3). Hale's colloidal iron was positive in all cases. Immunohistochemical stain CK7 and CD117 were positive in 100% and 95.5% of cases respectively. Vimentin was negative in all cases, except in the sarcomatoid areas of 3 cases. In conclusion, chromophobe renal cell carcinoma has certain unique morphological features and immunohistochemical profile which help to distinguish it from conventional renal cell carcinoma and oncocytoma. We identified nuclear pseudoinclusions, microglandular pattern and palisading of smaller cells, which have not been reported earlier.

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[158]

TÍTULO / TITLE: - A case of multiple endocrine neoplasia 2B with probable ectopic adrenocorticotropic hormone-secreting liver metastasis from medullary thyroid carcinoma.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - J UOEH. 2013 Sep 1;35(3):193-9.

AUTORES / AUTHORS: - Kurozumi A; Okada Y; Arao T; Nakamoto Y; Togashi K; Tanaka Y

INSTITUCIÓN / INSTITUTION: - First Department of Internal Medicine, School of Medicine, University of Occupational and Environmental Health, Japan.

RESUMEN / SUMMARY: - A 31 year old woman was diagnosed with multiple endocrine neoplasia (MEN) 2B at 10 years old. Dark pigmentation gradually developed on her skin and her serum adrenocorticotropic hormone (ACTH) was high, suggesting concurrent ectopic ACTH syndrome (EAS). Corticotropin-releasing hormone (CRH) loading test ruled out Cushing's disease and supported the diagnosis of EAS. Multiple low attenuation mass in the liver was observed in a computed tomography (CT) scan, and was suspected as ectopic ACTH-secreting metastatic tumor from medullary thyroid carcinoma (MTC). ACTH production by MTC is relatively rare, particularly in patients with MEN; patients with ectopic ACTH-secreting liver metastatic tumor from MTC in MEN 2B have never been reported previously.

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[159]
TÍTULO / TITLE: - Global DNA methylation patterns in small intestinal neuroendocrine tumors (SI-NETs).
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Endocr Relat Cancer. 2013 Nov 5.
●● Enlace al texto completo (gratuito o de pago) 1530/ERC-13-0481
AUTORES / AUTHORS: - Delgado Verdugo A; Crona J; Starker LF; Stalberg P; Akerstrom G; Westin G; Hellman P; Bjorklund P
INSTITUCIÓN / INSTITUTION: - A Delgado Verdugo, Department of Surgical Sciences, Uppsala University, Uppsala, 75185, Sweden.
RESUMEN / SUMMARY: - Small intestinal neuroendocrine tumors (SI-NETs) are rare hormone producing tumors and are often diagnosed at advanced stage. The genetic and epigenetic background of SI-NETs are poorly understood, but several reports have indicated chromosomal losses at 18.21-qter and 11q22-q23. The aim of this study was to characterize CpG DNA methylation status of primary SI-NETs and the corresponding lymph node metastases. We used the commercially available HumanMethylation27 Beadchip array (Illumina), which covers 27578 CpG sites spanning over 14495 genes, and analyzed a discovery cohort of 10 primary SI-NETs with matched metastases. Messenger- mRNA, were determined for selected genes in a 47 tumors. In comparison to the primary tumors, the metastases showed 2697 statistically significant differentially genes. Metastases were generally less methylated than primary tumors. The relative mRNA expression level of the differentially methylated genes AXL, CRMP1, FGF5, and APOBEC3C largely reflected the methylation status. MAPK4, RUNX3, TP73, CCND1, CHFR, AHRR, and Rb1 known to be hypermethylated in other cancer types, displayed overall high methylation level (beta-value >/= 0.9). Methylation (beta -value >0,7) at 18q21-qter and 11q22-q23 were detected in genes SETBP1, ELAC1, MBD1, MAPK4, TCEB3C and ARVC1, MMP8, BTG4, APOA1, FAM89B, HSPB1, respectively. Furthermore unsupervised clustering of the tumors identified three distinct clusters, one with a highly malignant behavior. Our data supports involvement of CpG DNA methylation in metastatic progression of SI-NETs and this could present a possibility to identify more aggressive tumors based on DNA methylation.

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[160]
TÍTULO / TITLE: - Surgery for Giant Primary Neuroendocrine Carcinoma of the Liver.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - J Gastrointest Surg. 2013 Oct 22.
●● Enlace al texto completo (gratuito o de pago) 1007/s11605-013-2346-7
AUTORES / AUTHORS: - Sotiropoulos GC; Charalampoudis P; Delladetsima I; Stamopoulos P; Dourakis S; Kouraklis G
INSTITUCIÓN / INSTITUTION: - 2nd Department of Propedeutic Surgery, University of Athens Medical School, Athens, Greece, georgios.sotiropoulos@uni-due.de.
RESUMEN / SUMMARY: - Liver resection for primary hepatic neuroendocrine carcinoma (phNEC) has only scarcely been reported in the literature. We herein report on a 19-year-old female with a solitary 27 x 13-cm-big phNEC, which was initially considered as hemangioma. An extended right hepatectomy (segments V-VIII, partially IVa) was performed. Resection margins were free of tumor (R0 resection). Ki67 expression was 35 %. Postoperative course was uneventful, and the patient was discharged on the

seventh postoperative day. Two years after surgery, the patient remains disease-free and in good general condition. Large series and longer follow-up studies are required for the better understanding on this rare tumor entity.

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[161]

TÍTULO / TITLE: - Palliative bypass for small bowel carcinoid with mesenteric mass and vascular encasement.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - ANZ J Surg. 2013 Oct 31. doi: 10.1111/ans.12333.

●● Enlace al texto completo (gratuito o de pago) 1111/ans.12333

AUTORES / AUTHORS: - Nathan AD; Chandrasegaram MD; Neo EL; Dolan PM; Tan CP; Chen JW; Worthley CS

INSTITUCIÓN / INSTITUTION: - Hepatobiliary Unit, Royal Adelaide Hospital, Adelaide, South Australia, Australia.

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[162]

TÍTULO / TITLE: - A miRNA signature associated with human metastatic medullary thyroid carcinoma.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Endocr Relat Cancer. 2013 Oct 14;20(6):809-23. doi: 10.1530/ERC-13-0357. Print 2013.

●● Enlace al texto completo (gratuito o de pago) 1530/ERC-13-0357

AUTORES / AUTHORS: - Santarpia L; Calin GA; Adam L; Ye L; Fusco A; Giunti S; Thaller C; Paladini L; Zhang X; Jimenez C; Trimarchi F; El-Naggar AK; Gagel RF

INSTITUCIÓN / INSTITUTION: - Departments of Endocrine Neoplasia and Hormonal Disorders Experimental Therapeutics Urology, The University of Texas M.D. Anderson Cancer Center, Houston, Texas, USA Department of Oncology, The University of Naples, Naples, Italy Department of Pathology, Centro Oncologico Fiorentino, Sesto Fiorentino, Florence, Italy Verna and Marrs McLean Department of Biochemistry and Molecular Biology Baylor College of Medicine, Houston, Texas, USA Department of Oncology, Istituto Toscano Tumori, Hospital of Prato, Prato, Italy Department of Gynecologic Oncology, Center for RNA Interference and Non-coding RNAs, The University of Texas MD Anderson Cancer Center, Houston, Texas, USA Department of Endocrinology, University of Messina, Messina, Italy Department of Pathology, The University of Texas M.D. Anderson Cancer Center, Houston, Texas, USA Department of Internal Medicine, The University of Texas M.D. Anderson Cancer Center, Houston, Texas, USA.

RESUMEN / SUMMARY: - MicroRNAs (miRNAs) represent a class of small, non-coding RNAs that control gene expression by targeting mRNA and triggering either translational repression or RNA degradation. The objective of our study was to evaluate the involvement of miRNAs in human medullary thyroid carcinoma (MTC) and to identify the markers of metastatic cells and aggressive tumour behaviour. Using matched primary and metastatic tumour samples, we identified a subset of miRNAs aberrantly regulated in metastatic MTC. Deregulated miRNAs were confirmed by quantitative real-time PCR and validated by in situ hybridisation on a large independent set of primary and metastatic MTC samples. Our results uncovered ten miRNAs that

were significantly expressed and deregulated in metastatic tumours: miR-10a, miR-200b/-200c, miR-7 and miR-29c were down-regulated and miR-130a, miR-138, miR-193a-3p, miR-373 and miR-498 were up-regulated. Bioinformatic approaches revealed potential miRNA targets and signals involved in metastatic MTC pathways. Migration, proliferation and invasion assays were performed in cell lines treated with miR-200 antagomirs to ascertain a direct role for this miRNA in MTC tumourigenesis. We show that the members of miR-200 family regulate the expression of E-cadherin by directly targeting ZEB1 and ZEB2 mRNA and through the enhanced expression of tumour growth factor beta (TGFbeta)-2 and TGFbeta-1. Overall, the treated cells shifted to a mesenchymal phenotype, thereby acquiring an aggressive phenotype with increased motility and invasion. Our data identify a robust miRNA signature associated with metastatic MTC and distinct biological processes, e.g., TGFbeta signalling pathway, providing new potential insights into the mechanisms of MTC metastasis.

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[163]
TÍTULO / TITLE: - The role of Cdk5 in neuroendocrine thyroid cancer.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Cancer Cell. 2013 Oct 14;24(4):499-511. doi: 10.1016/j.ccr.2013.08.027.
●● Enlace al texto completo (gratuito o de pago) 1016/j.ccr.2013.08.027
AUTORES / AUTHORS: - Pozo K; Castro-Rivera E; Tan C; Plattner F; Schwach G; Siegl V; Meyer D; Guo A; Gundara J; Mettlach G; Richer E; Guevara JA; Ning L; Gupta A; Hao G; Tsai LH; Sun X; Antich P; Sidhu S; Robinson BG; Chen H; Nwariaku FE; Pfragner R; Richardson JA; Bibb JA
INSTITUCIÓN / INSTITUTION: - Department of Psychiatry, University of Texas Southwestern Medical Center, Dallas, TX 75390, USA.
RESUMEN / SUMMARY: - Medullary thyroid carcinoma (MTC) is a neuroendocrine cancer that originates from calcitonin-secreting parafollicular cells, or C cells. We found that Cdk5 and its cofactors p35 and p25 are highly expressed in human MTC and that Cdk5 activity promotes MTC proliferation. A conditional MTC mouse model was generated and corroborated the role of aberrant Cdk5 activation in MTC. C cell-specific overexpression of p25 caused rapid C cell hyperplasia leading to lethal MTC, which was arrested by repressing p25 overexpression. A comparative phosphoproteomic screen between proliferating and arrested MTC identified the retinoblastoma protein (Rb) as a crucial Cdk5 downstream target. Prevention of Rb phosphorylation at Ser807/Ser811 attenuated MTC proliferation. These findings implicate Cdk5 signaling via Rb as critical to MTC tumorigenesis and progression.

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[164]
TÍTULO / TITLE: - Carcinoid syndrome caused by a serotonin secreting pituitary tumor.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Eur J Endocrinol. 2013 Oct 30.
●● Enlace al texto completo (gratuito o de pago) 1530/EJE-13-0622
AUTORES / AUTHORS: - Lynggaard LA; Nielsen EH; Laurberg P
INSTITUCIÓN / INSTITUTION: - L Lynggaard, Department of Endocrinology and Medicine, Aalborg University Hospital, Aalborg, Denmark.

RESUMEN / SUMMARY: - NEUROENDOCRINE TUMOURS ARE MOST FREQUENTLY LOCATED IN THE GASTROINTESTINAL ORGAN SYSTEM OR IN THE LUNGS, BUT THEY MAY OCCASIONALLY BE FOUND IN OTHER ORGANS.CASE: We describe a 56-year old woman suffering from a cardinoid syndrome caused by a large serotonin secreting pituitary tumour. She had for years suffered from episodes of palpitations, dyspnoea and flushing. Cardiac disease had been suspected, which delayed the diagnosis, until blood tests revealed elevated serotonin and chromogranin A in plasma. Somatostatin receptor (SSR) scintigraphy showed a single positive focus in the region of the pituitary gland and MRI a corresponding intra- and suprasellar heterogeneous mass. After pre-treatment with octreotide leading to symptomatic improvement, the patient underwent trans-cranial surgery with removal of the tumour. This led to clinical improvement and to a normalisation of SSR scintigraphy, as well as serotonin and chromogranin A levels. CONCLUSION: To our knowledge, this is the first reported case of a serotonin secreting tumour with a primary location in the pituitary.

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[165]

TÍTULO / TITLE: - Detection of tumor progression in optic pathway glioma with and without neurofibromatosis type 1.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Neuro Oncol. 2013 Nov;15(11):1560-7. doi: 10.1093/neuonc/not120. Epub 2013 Oct 6.

●● Enlace al texto completo (gratuito o de pago) 1093/neuonc/not120

AUTORES / AUTHORS: - Kelly JP; Weiss AH

INSTITUCIÓN / INSTITUTION: - Corresponding Author: Avery H. Weiss, MD, Division of Ophthalmology, OA.9.220, Seattle Children's Hospital, 4800 Sand Point Way NE, Seattle, WA 98105. avery.weiss@seattlechildrens.org.

RESUMEN / SUMMARY: - Background We wanted to determine the sensitivity and specificity of serial changes in visual acuity and visual evoked potentials (VEPs) to detect radiological progression of tumor volume in children with optic pathway gliomas. Methods From a retrospective review of a cohort of 69 patients, 54 patients met inclusion criteria (31 with primary chemotherapy, 4 with primary radiotherapy, and 19 with stable tumor volume and no treatment). Age at presentation ranged from 0.3 to 13 years. Patients were serially followed by MRI, age-corrected visual acuity in log minimum angle of resolution (logMAR), and pattern VEP. Longitudinal data averaged 7.9 years (range 0.5-16 y). Visual assessments were aligned with MRI data within 6-month intervals. Tumor progression was defined by 25% or greater increase in volume. Results Visual acuity in the better eye had poor sensitivity and specificity for detecting tumor volume progression (0.5 and 0.5, respectively). Visual acuity in the worse eye showed worse sensitivity and specificity because false positives (visual decline without tumor progression) were more frequent than true positives (visual decline with tumor progression). VEPs showed slightly better sensitivity and specificity (0.69 and 0.58, respectively). In patients with stable tumors, visual acuity fluctuated +/- 0.55 logMAR (SD = 0.15) between examinations. VEP amplitude fluctuated -0.74 to 0.48 log units (SD = 0.19) between examinations. Conclusions Serial changes in visual function do not reliably detect tumor progression. Conversely, tumor progression does

not reliably indicate decreased visual function. Objective visual function and serial MRIs are complementary in management of optic pathway gliomas.

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[166]

TÍTULO / TITLE: - Management and clinical outcomes of type I gastric carcinoid patients: Retrospective, multicenter study in Japan.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Dig Endosc. 2013 Nov 5. doi: 10.1111/den.12197.

●● Enlace al texto completo (gratuito o de pago) 1111/den.12197

AUTORES / AUTHORS: - Sato Y; Imamura H; Kaizaki Y; Koizumi W; Ishido K; Kurahara K; Suzuki H; Fujisaki J; Hirakawa K; Hosokawa O; Ito M; Kaminishi M; Furuta T; Chiba T; Haruma K

INSTITUCIÓN / INSTITUTION: - Department of Gastroenterology, Niigata University Graduate School of Medical and Dental Sciences, Niigata, Japan.

RESUMEN / SUMMARY: - BACKGROUND AND AIM: Type I gastric carcinoids (TIGC) are associated with chronic atrophic gastritis (CAG) with hypergastrinemia and hyperplasia of enterochromaffin-like cells. Several treatment options are currently available for these tumors including total gastrectomy, partial resection, antrectomy, endoscopic resection and endoscopic surveillance. The present study evaluated different treatment approaches and clinical outcomes of patients with TIGC in Japan. METHODS: Between 1991 and 2011, 82 patients with TIGC were identified at multicenter institutions in Japan. Patient demographics, tumor size, depth of invasion, vessel involvement, treatment approach, Helicobacter pylori infection, serum gastrin level, recurrence-free survival (RFS) and disease-specific survival (DSS) were analyzed. RESULTS: Median age of all patients at the time of diagnosis was 56 years (range, 24-79 years). There were 44 males and 38females. Patients underwent endoscopic surveillance (n = 25), endoscopic resection (n = 41) or surgical resection (n = 16). Intramucosal invasion was found in 19 patients, submucosal invasion in 44 patients and muscularis propria invasion in one patient. Tumor diameter was </=10 mm in 71 patients, 11-20 mm in five patients and >/=21 mm in five patients. None of the patients showed rapidly growing tumors, local recurrence or metastasis. The median (range) follow-up period was 7 (0-20) years. RFS was 97.6% and DSS was 100% in all the patients. CONCLUSION: The prognosis of TIGC patients treated by different modalities in Japan is favorable regardless of the generational change of management for TIGC.

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[167]

TÍTULO / TITLE: - The effect of resveratrol in combination with irradiation and chemotherapy : Study using Merkel cell carcinoma cell lines.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Strahlenther Onkol. 2013 Nov 8.

●● Enlace al texto completo (gratuito o de pago) 1007/s00066-013-0445-8

AUTORES / AUTHORS: - Heiduschka G; Lill C; Seemann R; Brunner M; Schmid R; Houben R; Bigenzahn J; Thurnher D

INSTITUCIÓN / INSTITUTION: - Department of Otorhinolaryngology, Head and Neck Surgery, Medical University of Vienna, Waehringer Guertel 18-20, 1090, Vienna, Austria.

RESUMEN / SUMMARY: - BACKGROUND AND PURPOSE: Merkel cell carcinoma (MCC) is a rare, but highly malignant tumor of the skin. In case of systemic disease, possible therapeutic options include irradiation or chemotherapy. The aim of this study was to evaluate whether the flavonoid resveratrol enhances the effect of radiotherapy or chemotherapy in MCC cell lines. MATERIALS AND METHODS: The two MCC cell lines MCC13 and MCC26 were treated with increasing doses of resveratrol. Combination experiments were conducted with cisplatin and etoposide. Colony forming assays were performed after sequential irradiation with 1, 2, 3, 4, 6, and 8 Gy and apoptosis was assessed with flow cytometry. Expression of cancer drug targets was analyzed by real-time PCR array. RESULTS: Resveratrol is cytotoxic in MCC cell lines. Cell growth is inhibited by induction of apoptosis. The combination with cisplatin and etoposide resulted in a partially synergistic inhibition of cell proliferation. Resveratrol and irradiation led to a synergistic reduction in colony formation compared to irradiation alone. Evaluation of gene expression did not show significant difference between the cell lines. CONCLUSION: Due to its radiosensitizing effect, resveratrol seems to be a promising agent in combination with radiation therapy. The amount of chemosensitizing depends on the cell lines tested.

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[168]
TÍTULO / TITLE: - Sonographic diagnosis of pheochromocytoma in childhood and adolescence.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Ultraschall Med. 2013 Oct;34(5):413-6.
AUTORES / AUTHORS: - Gerdemann C; Deeg KH

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[169]
TÍTULO / TITLE: - Impact of cinacalcet hydrochloride in clinical management of primary hyperparathyroidism in multiple endocrine neoplasia type 1.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Minerva Endocrinol. 2013 Dec;38(4):389-94.
AUTORES / AUTHORS: - Del Prete M; Marotta V; Ramundo V; Marciello F; Di Sarno A; Esposito R; Carratu AC; De Luca Di Roseto C; Di Somma C; Colao A; Faggiano A
INSTITUCIÓN / INSTITUTION: - Department of Molecular and Clinical Endocrinology and Oncology, “Federico II” University, Naples, Italy - michidelpre@gmail.com.
RESUMEN / SUMMARY: - Aim: Primary hyperparathyroidism (PHPT) is one of main cause of morbidity in patients with multiple endocrine neoplasia type 1 (MEN1). Medical therapy with cinacalcet-hydrochloride may modify the therapeutic strategy of MEN1 related PHPT. We present an experience with cinacalcet-hydrochloride in two patients with MEN1 PHPT. Methods: The study included two MEN1 patients belonging to the same family (a 50-year-old woman and her daughter aged 20 years) with PHPT secondary to multiple involvement of parathyroid glands and other MEN1 related tumors. As both patients refused to undergo parathyroid surgery, we decided to start medical treatment with cinacalcet at the dose of 30 mg/day, which was the first treatment for the youngest patient, while the oldest had already been treated with partial parathyroidectomy. Serum concentrations of PTH, calcium and phosphorus, 24-h urine calcium-to-creatinine ratio and renal-threshold-phosphate concentration were

evaluated before and after therapy. Results: Serum calcium and PTH levels were normalized after 1 and 6 months of therapy, respectively, and 60 and 54 months after the beginning of cinacalcet remained normal. Hypercalciuria, hypophosphoremia and renal-threshold-phosphate normalized during therapy with cinacalcet. At ultrasonography, parathyroid nodular lesion remained unchanged. Cinacalcet was well tolerated without occurrence of side effects. Conclusion: Cinacalcet seems to be highly effective in controlling PHPT in patients with MEN1 either in naive patients or in those with postsurgical recurrence. If cinacalcet will be confirmed to ensure a long-time control of PHPT or even to prevent the development and progression of PHPT, this may led to modify the therapeutic strategy of MEN1 PHPT.

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[170]

TÍTULO / TITLE: - Prognostic impact of histology in patients with cervical squamous cell carcinoma, adenocarcinoma and small cell neuroendocrine carcinoma.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Asian Pac J Cancer Prev. 2013;14(9):5355-60.

AUTORES / AUTHORS: - Intaraphet S; Kasatpibal N; Siriaunkgul S; Sogaard M; Patumanond J; Khunamornpong S; Chandacham A; Suprasert P

INSTITUCIÓN / INSTITUTION: - Boromarajonnani College of Nursing, Khon Kaen, Khon Kaen, Thailand E-mail : yrai250@hotmail.com.

RESUMEN / SUMMARY: - Background: Clarifying the prognostic impact of histological type is an essential issue that may influence the treatment and follow-up planning of newly diagnosed cervical cancer cases. This study aimed to evaluate the prognostic impact of histological type on survival and mortality in patients with cervical squamous cell carcinoma (SCC), adenocarcinoma (ADC) and small cell neuroendocrine carcinoma (SNEC). Materials and Methods: All patients with cervical cancer diagnosed and treated at Chiang Mai University Hospital between January 1995 and October 2011 were eligible. We included all patients with SNEC and a random weighted sample of patients with SCC and ADC. We used competing-risks regression analysis to evaluate the association between histological type and cancer-specific survival and mortality. Results: Of all 2,108 patients, 1,632 (77.4%) had SCC, 346 (16.4%) had ADC and 130 (6.2%) had SNEC. Overall, five-year cancer-specific survival was 60.0%, 54.7%, and 48.4% in patients with SCC, ADC and SNEC, respectively. After adjusting for other clinical and pathological factors, patients with SNEC and ADC had higher risk of cancer-related death compared with SCC patients (hazard ratio [HR] 2.6; 95% CI, 1.9-3.5 and HR 1.3; 95% CI, 1.1-1.5, respectively). Patients with SNEC were younger and had higher risk of cancer-related death in both early and advanced stages compared with SCC patients (HR 4.9; 95% CI, 2.7-9.1 and HR 2.5; 95% CI, 1.7-3.5, respectively). Those with advanced-stage ADC had a greater risk of cancer-related death (HR 1.4; 95% CI, 1.2-1.7) compared with those with advanced-stage SCC, while no significant difference was observed in patients with early stage lesions. Conclusion: Histological type is an important prognostic factor among patients with cervical cancer in Thailand. Though patients with SNEC were younger and more often had a diagnosis of early stage compared with ADC and SCC, SNEC was associated with poorest survival. ADC was associated with poorer survival compared with SCC in advanced stages, while no difference was observed at early stages. Further tailored treatment-

strategies and follow-up planning among patients with different histological types should be considered.

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[171]
TÍTULO / TITLE: - Goblet cell carcinoid tumor of the appendix.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - JBR-BTR. 2013 Jul-Aug;96(4):216-7.
AUTORES / AUTHORS: - De Keyzer B; Crolla D; Ovreeide P; Crevits I
INSTITUCIÓN / INSTITUTION: - Department of Radiology, Heilig Hart Ziekenhuis Roeselare-Menen, Belgium.

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[172]
TÍTULO / TITLE: - Emerging therapies and latest development in the treatment of unresectable pancreatic neuroendocrine tumors: an update for clinicians.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Therap Adv Gastroenterol. 2013 Nov;6(6):474-90. doi: 10.1177/1756283X13498808.
●● Enlace al texto completo (gratuito o de pago) 1177_1756283X13498808 [pii
●● Enlace al texto completo (gratuito o de pago) 1177/1756283X13498808
AUTORES / AUTHORS: - Sharma J; Duque M; Saif MW
INSTITUCIÓN / INSTITUTION: - Tufts University School of Medicine, Tufts Medical Center, Boston, MA, USA.
RESUMEN / SUMMARY: - Pancreatic neuroendocrine tumors (pNETs) differ in their clinical behavior, presentation and prognosis based on their initial histological features and disease stage. While small resectable tumors can be treated surgically, metastatic and locally advanced disease carries a significant mortality and treatment options have been limited in terms of their efficacy. Streptozocin-based regimens were the only agents available before but recent advances have improved the armamentarium to treat pNETs. Newer chemotherapeutic agents such as temozolomide, somatostatin analogs and targeted therapies including everolimus and sunitinib are now available to treat these tumors. Several combination regimens with targeted therapies and newer agents such as pazopanib are being developed and tested in ongoing trials.

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[173]
TÍTULO / TITLE: - Cutaneous squamous cell carcinoma in a patient with neurofibromatosis type 1: A case report.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Oncol Lett. 2013 Oct;6(4):878-880. Epub 2013 Jul 25.
●● Enlace al texto completo (gratuito o de pago) 3892/ol.2013.1490
AUTORES / AUTHORS: - Ishida M; Okabe H
INSTITUCIÓN / INSTITUTION: - Department of Clinical Laboratory Medicine and Division of Diagnostic Pathology, Shiga University of Medical Science, Otsu, Shiga, Japan.
RESUMEN / SUMMARY: - Neurofibromatosis type 1 (NF1) is an autosomal dominant inherited disease that is characterized by the presence of multiple neurofibromas, cafe-au-lait spots and iris hamartomas. It is well established that the incidence of tumors in patients with NF1 is high compared with the normal population and that the majority of

the tumors are non-epithelial neoplasms, including neurofibromas, malignant peripheral nerve sheath tumors, gliomas and leukemia. Studies have suggested that patients with NF1 also have a significantly higher risk of certain types of carcinomas. However, the occurrence of cutaneous squamous cell carcinoma (SCC) in a patient with NF1 is extremely rare. The present study describes the second documented case of a cutaneous SCC adjacent to a neurofibroma of the forehead with histopathological analyses in a patient with NF1. An 80-year-old female with NF1 presented with a rapidly growing skin tumor of the forehead. Histopathological study of the resected forehead tumor demonstrated that there were two tumorous lesions. One was an invasive SCC and the other was a neurofibroma. The lesions were adjacent, but no continuity was present. NF1 is caused by inactivating mutations in the NF1 gene and loss of heterozygosity of this gene has been reported in neurofibromas, malignant peripheral nerve sheath tumors, gliomas and pheochromocytomas in patients with NF1. However, the genetic mechanism of carcinoma development in patients with NF1 is not well understood. Studies have suggested the role of the NF1 and/or the BRCA gene in the occurrence of breast cancer. Additional studies are required to elucidate these mechanisms.

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[174]

TÍTULO / TITLE: - Antioxidant effect of mogrosides against oxidative stress induced by palmitic acid in mouse insulinoma NIT-1 cells.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Braz J Med Biol Res. 2013 Nov 18;0:0.

●● Enlace al texto completo (gratuito o de pago) 1590/1414-431X20133163

AUTORES / AUTHORS: - Xu Q; Chen SY; Deng LD; Feng LP; Huang LZ; Yu RR

INSTITUCIÓN / INSTITUTION: - Guilin Medical University, Department of Pharmacy, Guilin, China.

RESUMEN / SUMMARY: - Excessive oxidative stress in pancreatic beta cells, caused by glucose and fatty acids, is associated with the pathogenesis of type 2 diabetes. Mogrosides have shown antioxidant and antidiabetic activities in animal models of diabetes, but the underlying mechanisms remain unclear. This study evaluated the antioxidant effect of mogrosides on insulinoma cells under oxidative stress caused by palmitic acid, and investigated the underlying molecular mechanisms. Mouse insulinoma NIT-1 cells were cultured in medium containing 0.75 mM palmitic acid, mimicking oxidative stress. The effects of 1 mM mogrosides were determined with the dichlorodihydrofluorescein diacetate assay for intracellular reactive oxygen species (ROS) and FITC-Annexin V/PI assay for cell apoptosis. Expression of glucose transporter-2 (GLUT2) and pyruvate kinase was determined by semi-quantitative reverse-transcription polymerase chain reaction. Palmitic acid significantly increased intracellular ROS concentration 2-fold ($P<0.05$), and decreased expression of GLUT2 (by 60%, $P<0.05$) and pyruvate kinase (by 80%, $P<0.05$) mRNAs in NIT-1 cells. Compared with palmitic acid, co-treatment with 1 mM mogrosides for 48 h significantly reduced intracellular ROS concentration and restored mRNA expression levels of GLUT2 and pyruvate kinase. However, mogrosides did not reverse palmitic acid-induced apoptosis in NIT-1 cells. Our results indicate that mogrosides might exert their antioxidant effect by reducing intracellular ROS and regulating expression of genes involved in glucose metabolism. Further research is needed to achieve a better

understanding of the signaling pathway involved in the antioxidant effect of mogrosides.

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[175]

TÍTULO / TITLE: - A case of presumed choroidal metastasis from carcinoid tumor treated by photodynamic therapy with verteporfin.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Clin Ophthalmol. 2013;7:2003-6. doi: 10.2147/OPTH.S51196. Epub 2013 Oct 9.

●● Enlace al texto completo (gratuito o de pago) 2147/OPTH.S51196

AUTORES / AUTHORS: - Kawakami S; Wakabayashi Y; Goto H

INSTITUCIÓN / INSTITUTION: - Department of Ophthalmology, Tokyo Medical University, Tokyo, Japan.

RESUMEN / SUMMARY: - We report a case of metastatic choroidal carcinoid tumor with favorable outcome after photodynamic therapy. A 75-year-old woman was presumptively diagnosed with bilateral choroidal metastases from carcinoid tumor. Although the tumor in the right eye showed a tendency toward rapid expansion and required aggressive treatment to preserve vision, the size was still small and we hesitated to use external-beam radiotherapy because of the risk of radiation retinopathy. Consequently, photodynamic therapy was performed on the right eye, resulting in drastic reduction of the size and height of the choroidal tumor. Good visual acuity was maintained after photodynamic therapy. Photodynamic therapy may be an effective treatment for choroidal metastasis from carcinoid tumor.

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[176]

TÍTULO / TITLE: - Renal medullary cancer in a patient with sickle cell trait.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Case Rep Oncol Med. 2013;2013:129813. doi: 10.1155/2013/129813. Epub 2013 Oct 7.

●● Enlace al texto completo (gratuito o de pago) 1155/2013/129813

AUTORES / AUTHORS: - Alappan N; Marak CP; Chopra A; Joy PS; Dorokhova O; Guddati AK

INSTITUCIÓN / INSTITUTION: - Division of Pulmonary and Critical Care Medicine, Montefiore Hospital, Albert Einstein College of Medicine, Yeshiva University, New York, NY, USA.

RESUMEN / SUMMARY: - Renal medullary cancer is a rare malignancy almost exclusively seen in young patients of African ethnicity. These patients often present with the cardinal symptoms of hematuria, flank pain, and an abdominal mass, and this malignancy has been associated with patients carrying sickle cell trait. It is estimated that 300 million people worldwide carry sickle cell trait, and the presence of hematuria in these patients should be treated as a harbinger of a possible malignancy. Notably, this tumor mostly develops on the right side of the body. Patients often present with it at an advanced stage and the prognosis is poor. Therefore, a high index of suspicion in a patient of African descent presenting with a right sided abdominal mass and hematuria may assist in an early diagnosis. Current chemotherapy options are very limited, and early detection may provide a chance for surgical resection. It may also

provide a bigger time frame for the initiation of novel chemotherapy regimens in patients who fail current chemotherapy regimens.

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[177]

TÍTULO / TITLE: - Added value of fused somatostatin receptor imaging/magnetic resonance imaging in a rare case of paraganglioma of the urinary bladder.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Rev Esp Med Nucl. Acceso gratuito al texto completo a partir de los 2 años de la fecha de publicación.

●● Enlace a la Editora de la Revista http://db.doyma.es/

●● Cita: Revista Española de Medicina Nuclear: <> Imagen Mol. 2013 Oct 1. pii: S2253-654X(13)00128-5. doi: 10.1016/j.remn.2013.08.001.

●● Enlace al texto completo (gratuito o de pago) 1016/j.remn.2013.08.001

AUTORES / AUTHORS: - Treglia G; Ceriani L; Merlo E; Ruberto T; Paone G; Giovanella L

INSTITUCIÓN / INSTITUTION: - Department of Nuclear Medicine and PET/CT Centre, Oncology Institute of Southern Switzerland, Bellinzona, Switzerland. Electronic address: giorgiomednuc@libero.it.

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[178]

TÍTULO / TITLE: - Pheochromocytoma Management, Outcomes and the Role of Cortical Preservation.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Indian J Pediatr. 2013 Nov 8.

●● Enlace al texto completo (gratuito o de pago) 1007/s12098-013-1283-5

AUTORES / AUTHORS: - Gupta A; Agarwala S; Tandon N; Srinivas M; Bajpai M; Gupta DK; Gupta AK; Bal C; Kumar R; Bhatnagar V

INSTITUCIÓN / INSTITUTION: - Department of Pediatric Surgery, All India Institute of Medical Sciences, New Delhi, 110029, India.

RESUMEN / SUMMARY: - OBJECTIVES: To evaluate the management and outcome of children with pheochromocytoma and determine the role of cortex preservation in cases of bilateral disease. METHODS: Retrospective review of children, below 12 y of age, with pheochromocytoma managed between November 2003 and December 2012 was done. RESULTS: Twelve patients, nine boys and three girls with median age 9 y were enroled. Eleven (92 %) had adrenal tumors and in one it was extra-adrenal. Five (42 %) had bilateral disease. Ten presented with hypertension, one with headache and one with abdominal pain and fever. All were stabilized pre-operatively with alpha and beta blockers and volume expansion. Six children with unilateral disease underwent total adrenalectomy. Out of five with bilateral disease, one child underwent bilateral total adrenalectomy and was later started on hormone replacement. Remaining four underwent total adrenalectomy on one side and partial on the other side. Post-operatively all became symptom free and normotensive and were off medications within 1 mo. Two children developed recurrence 1 mo post-operatively, one with an initial unilateral pheochromocytoma and one with paraganglionoma. At the last follow up, 10 out of 12(83 %) were disease free while two with recurrence are still awaiting surgery. CONCLUSIONS: Surgical resection of pheochromocytoma is effective treatment to achieve cure and prolong survival. Cortex preservation should be done in bilateral disease as risk of recurrence in such cases seems to be of lesser significance

as compared to the morbidity and mortality of adrenal insufficiency and consequent lifelong hormone replacement.

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[179]

TÍTULO / TITLE: - Severe hypersensitivity pneumonitis associated with everolimus therapy for neuroendocrine tumour: a case report.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - BMC Res Notes. 2013 Nov 18;6(1):471.

●● Enlace al texto completo (gratuito o de pago) 1186/1756-0500-6-471

AUTORES / AUTHORS: - Sibertin-Blanc C; Norguet E; Duluc M; Louis G; Seitz JF; Dahan L

RESUMEN / SUMMARY: - BACKGROUND: Novel therapeutic agents are currently being investigated for neuroendocrine tumour treatment. CASE PRESENTATION: We report here on the case of a patient presenting with hypersensitivity pneumonitis while being treated with everolimus, a mammalian target of rapamycin (mTOR) inhibitor. CONCLUSION: Side effects of everolimus should be familiar to clinicians, including nonspecialists, and be monitored carefully to allow for prompt management.

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[180]

TÍTULO / TITLE: - Merkel cell carcinoma of the head and neck: challenges in diagnosis and therapy.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - J Skin Cancer. 2013;2013:427984. doi: 10.1155/2013/427984. Epub 2013 Sep 10.

●● Enlace al texto completo (gratuito o de pago) 1155/2013/427984

AUTORES / AUTHORS: - Miles BA

INSTITUCIÓN / INSTITUTION: - Otolaryngology Head and Neck Surgery, Icahn School of Medicine at Mount Sinai, New York, NY 10029, USA ; Oral and Maxillofacial Surgery, Icahn School of Medicine at Mount Sinai, New York, NY 10029, USA.

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[181]

TÍTULO / TITLE: - Solid neuroendocrine carcinoma of the breast.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - J Coll Physicians Surg Pak. 2013 Nov;23(10):820-2. doi: 11.2013/JCPSP.820822.

AUTORES / AUTHORS: - Abbasi NZ; Zahur Z; Sheikh AS; Khan AA; Ali F; Memon KH; Moizuddin SS; Loya A; Barkat N

INSTITUCIÓN / INSTITUTION: - Department of Oncology, Institute of Nuclear Medicine, Oncology and Radiotherapy (INOR), Abbottabad.

RESUMEN / SUMMARY: - Primary neuroendocrine carcinoma of the breast is a rare disease that accounts for less than 5% of all cancers arising from the breast. The tumour cells stain positively for chromogranin and synaptophysin. This report describes the occurrence of infiltrating ductal carcinoma of breast with neuroendocrine differentiated tumour in 37 year old female. Early small cell neuroendocrine cancer of the breast that is treated with surgery and adjuvant chemotherapy shows an increased disease-free survival. She is planned for anthracycline/cyclophosphamide based chemotherapy followed by etoposide/platinum based chemotherapy. As her tumour

showed ER/PR positivity, she will be given hormonal therapy subsequently, however, more extensive review is required to define a standard treatment protocol for this rare neoplasm.

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[182]

TÍTULO / TITLE: - Treatment of ampullary neuroendocrine tumor by endoscopic snare papillectomy.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Am J Case Rep. 2013 Oct 25;14:439-43. doi: 10.12659/AJCR.889601.

●● Enlace al texto completo (gratuito o de pago) 12659/AJCR.889601

AUTORES / AUTHORS: - Odabasi M; Yildiz KM; Cengiz E; Hasan AH; Gunay E; Ozkan E; Aktekin A; Kaya B; Muftuoglu TM

INSTITUCIÓN / INSTITUTION: - Department of Surgery, Haydarpasa Education and Research Hospital, Istanbul,Turkey.

RESUMEN / SUMMARY: - Patient: Female, 45 Final Diagnosis: Neuroendocrine tumor Symptoms: Abdominal pain Medication: - Clinical Procedure: - Specialty: Gastroenterology and Hepatology. OBJECTIVE: Unusual setting of medical care. BACKGROUND: Neuroendocrine tumor of the ampulla of Vater is extremely rare and is generally a low-grade endocrine cell tumor. The merits of radical vs. local resection remain uncertain. CASE REPORT: A 45-year-old female patient presented with abdominal pain lasting for 2 months. Papilla that was tumor-like macroscopically was seen in the second part of the duodenum in endoscopic retrograde cholangiopancreatography. Biopsy was histologically confirmed as a low-grade neuroendocrine tumor. No lymphadenopathy or visceral metastasis was found on an abdominal CT scan, In-111 octreotide scan, and EUS. The ampulla was removed by endoscopic snare papillectomy. All margins of resection were negative for tumor. CONCLUSIONS: Endoscopic snare papillectomy may be the first step in the management of neuroendocrine tumors of the ampulla of Vater in high-risk surgical candidates and selected patients such as those with a well differentiated, low-grade, small tumor without regional/ distant metastasis. However, it can also be used in younger patients who wish to avoid surgical resection.

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[183]

TÍTULO / TITLE: - Combination of Capecitabine and Oxaliplatin is an Effective Treatment Option for Advanced Neuroendocrine Tumors.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Rare Tumors. 2013 Sep 24;5(3):e35. doi: 10.4081/rt.2013.e35.

●● Enlace al texto completo (gratuito o de pago) 4081/rt.2013.e35

AUTORES / AUTHORS: - Ferrarotto R; Testa L; Riechelmann RP; Sahade M; Siqueira LT; Costa FP; Hoff PM

INSTITUCIÓN / INSTITUTION: - Hospital Sirio Libanes Universidade de Sao Paulo , Brazil.

RESUMEN / SUMMARY: - The role of chemotherapy in well differentiated neuroendocrine tumors (NET) has been questioned. It was recently demonstrated that everolimus and sunitinib have activity in low and intermediate grade pancreatic NET. The aim of this study was to evaluate the activity of capecitabine and oxaliplatin

(CapOx) combination in treating NET in an unselected population. In this regard, we retrospectively evaluated 24 patients diagnosed with metastatic NET treated with CapOx at two Brazilian institutes that are reference centers in cancer care. Tumor response was measured by RECIST criteria. Median age at diagnosis was 56 years, 71% had ECOG 0 or 1, the majority of tumors were primary from pancreas (67%) followed by lung (17%), and 29% were functional. According to WHO classification criteria, 25% were grade 1, 37.5% grade 2 and 37.5% grade 3. Most patients received CapOx as second-line therapy, with a median of 6 cycles. Twenty-nine percent of patients had partial response by RECIST criteria. No association was observed between response rate and tumor grade, primary site or line of CapOx. The median time to progression was 9.8 months and median time to treatment failure was 12.1 months. Seventy-five percent of patients are alive at the time of this analysis; therefore, median overall survival was not reached. The CapOx combination was shown to be active in an unselected population with metastatic NET and may be a good platform for the incorporation of the newer molecular targeted agents being investigated for the treatment of this entity.

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[184]

TÍTULO / TITLE: - A role for radiotherapy in the management of advanced medullary thyroid carcinoma: the mayo clinic experience.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Rare Tumors. 2013 Jul 12;5(3):e37. doi: 10.4081/rt.2013.e37.

●● Enlace al texto completo (gratuito o de pago) 4081/rt.2013.e37

AUTORES / AUTHORS: - Call JA; Caudill JS; McIver B; Foote RL

INSTITUCIÓN / INSTITUTION: - Department of Radiation Oncology, Mayo Clinic , Rochester, MN, USA.

RESUMEN / SUMMARY: - Outcomes of external beam radiotherapy (EBRT) in advanced medullary thyroid carcinoma (MTC) are largely unknown. Retrospective review of data from patients with MTC, diagnosed from June 1, 1970, through December 31, 2007. Overall survival and locoregional tumor control rates were calculated. Seventeen patients had adjuvant or palliative EBRT delivered to 41 sites. Six patients initially had adjuvant EBRT (median, 60.80 Gy); none had relapse in the treated area. Five patients with locoregional recurrence after surgery were treated (median, 59.40 Gy), and durable disease control was achieved in 3. Twelve patients received palliative EBRT to 29 sites of metastatic disease (median, 30.00 Gy), which provided sustained symptom relief at 45% of sites. Five- and ten-year overall survival rates were 44% and 19%, respectively. Adjuvant EBRT may be most effective for prevention of locoregional recurrence. EBRT may provide sustained control of advanced, metastatic disease in select patients.

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[185]

TÍTULO / TITLE: - Missense mutation in the MEN1 gene discovered through whole exome sequencing co-segregates with familial hyperparathyroidism.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Genet Res (Camb). 2013 Aug;95(4):114-20. doi: 10.1017/S0016672313000141.

●● Enlace al texto completo (gratuito o de pago) 1017/S0016672313000141

AUTORES / AUTHORS: - Isakov O; Rinella ES; Olchovsky D; Shimon I; Ostrer H; Shomron N; Friedman E
INSTITUCIÓN / INSTITUTION: - Department of Cell and Developmental Biology, The Sackler School of Medicine, Tel Aviv University, Tel-Aviv, Israel.
RESUMEN / SUMMARY: - Summary Familial isolated hyperparathyroidism (FIHP) can be encountered in the context of multiple endocrine neoplasia type 1 (MEN1), hyperparathyroidism and jaw tumour syndrome (HPT-JT) and in familial hypocalciuric hypercalcaemia (FHH). In these syndromes, germline mutations in the relevant genes (MEN1, HPRT2 and CaSR, respectively) are detected. In some FIHP cases, the causative gene is still elusive. The objective of this study is to define the genetic basis of FIHP in a Georgian Jewish family with FIHP using whole exome capture and sequencing. DNA extracted from two sibs and one offspring from a single family all affected with multiglandular hyperparathyroidism was subjected to whole exome capturing and sequencing using the Roche NimbleGen V2 chip and the Illumina HiSeq2000 sequencing platform. Genetic variants were detected and annotated using a combination of the Genome Analysis Tool Kit and in-house scripts. Subsequent confirmation of the mutations and co-segregation analyses were carried out by Sanger sequencing in additional affected and unaffected family members. Whole exome capture and sequencing revealed the collection of variations common to the three-sequenced patients, including a very rare previously described missense mutation (c.T1021C: p.W341R) in the MEN1 gene. The p.W341R mutation in the MEN1 gene showed complete co-segregation in the family. Whole exome capture and sequencing led to the discovery of a missense mutation in the MEN1 gene and ruling out of the additional candidates in a single experiment. The limited expressivity of this mutation may imply a specific genotype-phenotype correlation for this mutation.

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[186]
TÍTULO / TITLE: - Ectopic Cushing syndrome associated with thymic carcinoid tumor as the first presentation of MEN1 syndrome-report of a family with MEN1 gene mutation.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Fam Cancer. 2013 Nov 12.
●● Enlace al texto completo (gratuito o de pago) 1007/s10689-013-9692-1
AUTORES / AUTHORS: - Hasani-Ranjbar S; Rahmanian M; Ebrahim-Habibi A; Soltani A; Soltanzade A; Mahrampour E; Amoli MM
INSTITUCIÓN / INSTITUTION: - Obesity and Eating Habits Research Center, Endocrinology and Metabolism Cellular and Molecular Science Institute, Endocrinology and Metabolism Research Institute, Tehran University of Medical Sciences, Tehran, Iran.
RESUMEN / SUMMARY: - Multiple endocrine neoplasia type 1(MEN1) is an autosomal dominant syndrome. Although thymic carcinoid tumor is recognized as a part of MEN1 syndrome but functioning thymic carcinoid tumor as the first presentation of the MEN1 seems to be very rare. In this report, we present a 29-year-old male who developed ectopic Cushing syndrome secondary to thymic carcinoid tumor and was diagnosed as MEN1 syndrome 2 years later. Further evaluation revealed the presence of carcinoid tumor and other MEN 1 manifestations in several other member of family. Genetic evaluation showed presence of a previously reported mutation in exon 10(R527X) of MEN1 gene in these patients. This presentation showed that thymic neuroendocrine

tumor could be the first manifestation of the MEN1 syndrome and it might be diagnosed as a dominant manifestation of this syndrome in a family. We suggest biochemical or genetic screening for MEN-1 syndrome for patients with thymic carcinoid.

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[187]

TÍTULO / TITLE: - Analysis of risk factors for recurrence after curative resection of well-differentiated pancreatic neuroendocrine tumors based on the new grading classification.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - J Hepatobiliary Pancreat Sci. 2013 Oct 20. doi: 10.1002/jhbp.47.

●● Enlace al texto completo (gratuito o de pago) 1002/jhbp.47

AUTORES / AUTHORS: - Tsutsumi K; Ohtsuka T; Fujino M; Nakashima H; Aishima S; Ueda J; Takahata S; Nakamura M; Oda Y; Tanaka M

INSTITUCIÓN / INSTITUTION: - Department of Surgery and Oncology, Graduate School of Medical Sciences, Kyushu University, 3-1-1 Maidashi, Fukuoka, 812-8582, Japan.

RESUMEN / SUMMARY: - BACKGROUND: It is difficult to predict the malignant potential of pancreatic neuroendocrine tumors (PNETs) precisely. This study investigated the validity of a new grading system adopted by the World Health Organization 2010 classification to determine risk factors for recurrence of PNETs. METHODS: Data of 70 patients with PNETs who underwent curative resection were retrospectively examined by uni- and multivariate analyses. Histopathological findings were re-reviewed by experienced pathologists. NET G1 was defined as mitotic count <2 per 10 high power fields (HPF) and/or </=2% Ki67 index, and NET G2 as 2-20 mitosis per 10 HPF and/or 3-20% Ki67 index. RESULTS: There were 58 patients with NET G1 and 12 with NET G2. Incidence of recurrence was 11.4%. Univariate analysis demonstrated significant risk factors for recurrence including NET G2 of histological grade (P = 0.0089), male gender (P = 0.0333), tumor size >/= 20 mm (P = 0.0117), lymph node metastasis (P = 0.0004), liver metastasis (P < 0.0001), lymphatic invasion (P = 0.046), and neural invasion (P = 0.0002). By multivariate analysis, histological grade (hazard ratio; 59.76, P = 0.0022) and neural invasion (hazard ratio; 147.49, P = 0.0016) were significantly associated with recurrence of PNETs. CONCLUSIONS: This study confirmed the prognostic relevance of the new grading classification and that evaluation of perineural invasion and histological grade should be considered as prognostic predictors in well-differentiated PNETs (NET G1 and G2).

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[188]

TÍTULO / TITLE: - A Case of Small Cell Cancer of the Breast in a Male with Synchronous Stage IV Non-Small Cell Lung Carcinoma.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Rare Tumors. 2013 Sep 24;5(3):e52. doi: 10.4081/rt.2013.e52.

●● Enlace al texto completo (gratuito o de pago) 4081/rt.2013.e52

AUTORES / AUTHORS: - Matt L; Limjoco T; Sehgal R

INSTITUCIÓN / INSTITUTION: - Department of Hematology/Oncology, Edwards Comprehensive Cancer Center, Cabell Huntington Hospital , Huntington, WV.

RESUMEN / SUMMARY: - Extrapulmonary small cell carcinomas (EPSCC) are extremely rare. Most reports indicate success with therapy directed at the tumor as if it was pulmonary small cell carcinoma Primary small cell carcinoma of the breast is an

uncommon form of EPSCC. Differentiating between a primary small cell carcinoma of the breast from metastatic disease to the breast is very important. According to the literature, there have been approximately 70 cases reported worldwide. Of these cases, only two cases are documented in men. Prognosis is varied and depends on stage of disease at presentation. A combination of surgery, chemotherapy and/or radiation is required to adequately treat patients with small cell carcinoma of the breast. We present a case of a male patient diagnosed with stage IV non-small cell lung carcinoma first and then subsequently diagnosed with a concurrent small cell carcinoma of the breast responding to treatment with concurrent chemotherapy and radiation.

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[189]

TÍTULO / TITLE: - Identification of candidate serum proteins for classifying well-differentiated small intestinal neuroendocrine tumors.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - PLoS One. 2013 Nov 25;8(11):e81712. doi: 10.1371/journal.pone.0081712.

●● Enlace al texto completo (gratuito o de pago) 1371/journal.pone.0081712

AUTORES / AUTHORS: - Darmanis S; Cui T; Drobin K; Li SC; Oberg K; Nilsson P; Schwenk JM; Giandomenico V

INSTITUCIÓN / INSTITUTION: - Department of Immunology, Genetics, and Pathology, Uppsala University, Uppsala, Sweden ; Science for Life Laboratory, Uppsala, Sweden.

RESUMEN / SUMMARY: - BACKGROUND: Patients with well-differentiated small intestine neuroendocrine tumors (WD-SI-NETs) are most often diagnosed at a metastatic stage of disease, which reduces possibilities for a curative treatment. Thus new approaches for earlier detection and improved monitoring of the disease are required. MATERIALS AND METHODS: Suspension bead arrays targeting 124 unique proteins with antibodies from the Human Protein Atlas were used to profile biotinylated serum samples. Discoveries from a cohort of 77 individuals were followed up in a cohort of 132 individuals both including healthy controls as well as patients with untreated primary WD-SI-NETs, lymph node metastases and liver metastases. RESULTS: A set of 20 antibodies suggested promising proteins for further verification based on technically verified statistical significance. Proceeding, we assessed the classification performance in an independent cohort of patient serum, achieving, classification accuracy of up to 85% with different subsets of antibodies in respective pairwise group comparisons. The protein profiles of nine targets, namely IGFBP2, IGF1, SHKBP1, ETS1, IL1alpha, STX2, MAML3, EGR3 and XIAP were verified as significant contributors to tumor classification. CONCLUSIONS: We propose new potential protein biomarker candidates for classifying WD-SI-NETs at different stage of disease. Further evaluation of these proteins in larger sample sets and with alternative approaches is needed in order to further improve our understanding of their functional relation to WD-SI-NETs and their eventual use in diagnostics.

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[190]

TÍTULO / TITLE: - The VHL gene is epigenetically inactivated in pheochromocytomas and abdominal paragangliomas.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Epigenetics. 2013 Oct 22;8(12).

AUTORES / AUTHORS: - Andreasson A; Kiss NB; Caramuta S; Sulaiman L; Svahn F; Backdahl M; Hoog A; Juhlin CC; Larsson C
INSTITUCIÓN / INSTITUTION: - Department of Oncology-Pathology; Karolinska Institutet; Karolinska University Hospital; Stockholm, Sweden.
RESUMEN / SUMMARY: - Pheochromocytoma (PCC) and abdominal paraganglioma (PGL) are neuroendocrine tumors that present with clinical symptoms related to increased catecholamine levels. About a third of the cases are associated with constitutional mutations in pre-disposing genes, of which some may also be somatically mutated in sporadic cases. However, little is known about inactivating epigenetic events through promoter methylation in these genes. Using bisulphite pyrosequencing we assessed the methylation density of 11 PCC/PGL disease genes in 96 tumors (83 PCCs and 13 PGLs) and 34 normal adrenal references. Gene expression levels were determined by quantitative RT-PCR. Both tumors and normal adrenal samples exhibited low methylation index (MetI) in the EGLN1 (PDH2), MAX, MEN1, NF1, SDHB, SDHC, SDHD, SDHAF2 (SDH5), and TMEM127 promoters, not exceeding 10% in any of the samples investigated. Aberrant RET promoter methylation was observed in two cases only. For the VHL gene we found increased MetI in tumors as compared with normal adrenals (57% vs. 27%; P<0.001), in malignant vs. benign tumors (63% vs. 55%; P<0.05), and in PGL vs. PCC (66% vs. 55%; P<0.0005). Decreased expression of the VHL gene was observed in all tumors compared with normal adrenals (P<0.001). VHL MetI and gene expressions were inversely correlated (R = -0.359, P<0.0001). Our results show that the VHL gene promoter has increased methylation compared with normal adrenals (MetI>50%) in approximately 75% of PCCs and PGLs investigated, highlighting the role of VHL in the development of these tumors.

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[191]
TÍTULO / TITLE: - Response of merkel cell polyomavirus-positive merkel cell carcinoma xenografts to a survivin inhibitor.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - PLoS One. 2013 Nov 18;8(11):e80543. doi: 10.1371/journal.pone.0080543.
●● Enlace al texto completo (gratuito o de pago) 1371/journal.pone.0080543
AUTORES / AUTHORS: - Dresang LR; Guastafierro A; Arora R; Normolle D; Chang Y; Moore PS
INSTITUCIÓN / INSTITUTION: - Cancer Virology Program, University of Pittsburgh Cancer Institute, Pittsburgh, Pennsylvania, United States of America.
RESUMEN / SUMMARY: - Merkel cell carcinoma (MCC) is a neuroendocrine skin cancer associated with high mortality. Merkel cell polyomavirus (MCV), discovered in 2008, is associated with ~80% of MCC. The MCV large tumor (LT) oncoprotein upregulates the cellular oncoprotein survivin through its conserved retinoblastoma protein-binding motif. We confirm here that YM155, a survivin suppressor, is cytotoxic to MCV-positive MCC cells in vitro at nanomolar levels. Mouse survival was significantly improved for NOD-Scid-Gamma mice treated with YM155 in a dose and duration dependent manner for 3 of 4 MCV-positive MCC xenografts. One MCV-positive MCC xenograft (MS-1) failed to significantly respond to YM155, which corresponds with in vitro dose-response activity. Combination treatment of YM155 with other chemotherapeutics resulted in additive but not synergistic cell killing of MCC cell

lines in vitro. These results suggest that survivin targeting is a promising therapeutic approach for most but not all MCV-positive MCCs.

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[192]

TÍTULO / TITLE: - Combination of irinotecan and a platinum agent for poorly differentiated neuroendocrine carcinomas.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Rare Tumors. 2013 Sep 4;5(3):e39. doi: 10.4081/rt.2013.e39.

●● Enlace al texto completo (gratuito o de pago) 4081/rt.2013.e39

AUTORES / AUTHORS: - Ramella Munhoz R; de Mendonca Rego JF; de Celis Ferrari AR; Ignez Braghiroli M; Mendonca Bariani G; Marcelo Hoff P; Perego Costa F; Eduardo Flesch Pfiffer T; Riechelmann R

INSTITUCIÓN / INSTITUTION: - Centro de Oncologia, Hospital Sirio Libanes , Sao Paulo.

RESUMEN / SUMMARY: - Extrapulmonary poorly differentiated neuroendocrine carcinoma (PDNEC) is a rare and highly aggressive neoplasm for which the optimal chemotherapy remains unclear. The objective of this study was to evaluate the outcomes of patients with PDNEC treated with cisplatin and irinotecan (IP) and perform a review of the literature. From 2008 to 2012, patients with advanced PDNEC (Ki67>/=20%) who received the IP combination were selected for analysis. Radiologic responses were determined through Response Evaluation Criteria In Solid Tumors criteria. Twenty-eight patients were included. The median age at diagnosis was 57 years and the most common presentation was pancreatic PDNEC. Twenty-five patients (89%) received chemotherapy with cisplatin and irinotecan and three received carboplatin and irinotecan. Forty-six percent of the patients achieved objective response and the median time to tumor progression was 3.7 months. The median overall survival was 11.7 months. Thirteen patients (46%) had treatment interruptions or dose reductions due to grade ¾ toxicity. This retrospective cohort of advanced extrapulmonary PDNEC patients suggests that the IP combination is feasible and resulted in similar response rate and median survival to other treatments previously reported.

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[193]

TÍTULO / TITLE: - Extra-adrenal retroperitoneal paraganglioma in a dog.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - J Vet Diagn Invest. 2013 Nov;25(6):803-6. doi: 10.1177/1040638713506579. Epub 2013 Oct 8.

●● Enlace al texto completo (gratuito o de pago) 1177/1040638713506579

AUTORES / AUTHORS: - Ilha MR; Styer EL

INSTITUCIÓN / INSTITUTION: - 1Marcia R. S. Ilha, Tifton Veterinary Diagnostic and Investigational Laboratory, University of Georgia, 43 Brighton Road, Tifton, GA 31793. milha@uga.edu.

RESUMEN / SUMMARY: - An extra-adrenal retroperitoneal paraganglioma was observed in a 10.5-year-old male Boxer dog. Additionally, the dog had an aortic base tumor, multiple thyroid adenomas, multiple testicular interstitial cell tumors, bilateral nodular adrenal cortical hyperplasia, and parathyroid gland hyperplasia. The hypothesis that the retroperitoneal mass represents a primary extra-adrenal paraganglioma rather than metastatic mass from the aortic body tumor is considered.

Either primary or metastatic extra-adrenal retroperitoneal paragangliomas are rarely reported in dogs.

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[194]

TÍTULO / TITLE: - Recurrence of a carcinoid tumor of the ovary 13 years after the primary surgery: A case report.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Oncol Lett. 2013 Nov;6(5):1241-1244. Epub 2013 Aug 16.

●● Enlace al texto completo (gratuito o de pago) 3892/ol.2013.1530

AUTORES / AUTHORS: - Amano Y; Mandai M; Baba T; Hamanishi J; Yoshioka Y; Matsumura N; Konishi I

INSTITUCIÓN / INSTITUTION: - Department of Gynecology and Obstetrics, Graduate School of Medicine, Kyoto University, Kyoto 606-8507, Japan.

RESUMEN / SUMMARY: - The current study presents the case of a patient with a recurrent carcinoid tumor of the ovary, 13-years after the primary surgery. The primary surgery consisted of a total abdominal hysterectomy and bilateral salpingo-oophorectomy for a left ovarian tumor at 54 years old. Pathologically, the tumor was diagnosed as a carcinoid tumor of the ovary. Following the primary treatment, the patient was admitted to a cardiologist due to carcinoid-induced heart failure. At 67 years old, the patient was referred to Kyoto University Hospital with a solitary mass 8 cm in diameter and located in the paraaortic area, which was detected by routine ultrasonography and subsequent computed tomography (CT) scans. Urinary 5-hydroxyindole acetate (5-HIAA), a serotonin degradation metabolite, was present at elevated levels. With a diagnosis of a recurrent carcinoid tumor, the patient underwent a tumor resection. The pathological diagnosis was that of lymph node metastasis of the trabecular carcinoid. Post-operatively, the 5-HIAA levels returned to normal. Carcinoid tumors occasionally recur following surgery due to borderline malignant potential. Due to the slow growing nature of these tumors, in specific cases, recurrence occurs following a long interval. Therefore, a relatively long follow-up period is required.

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[195]

TÍTULO / TITLE: - Neuroendocrine tumours of the head and neck: anatomical, functional and molecular imaging and contemporary management.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Cancer Imaging. 2013 Oct 4;13(3):407-22. doi: 10.1102/1470-7330.2013.0034.

●● Enlace al texto completo (gratuito o de pago) 1102/1470-7330.2013.0034

AUTORES / AUTHORS: - Subedi N; Prestwich R; Chowdhury F; Patel C; Scarsbrook A

INSTITUCIÓN / INSTITUTION: - Department of Radiology, Leeds Teaching Hospitals NHS Trust, Leeds, UK.

RESUMEN / SUMMARY: - Neuroendocrine tumours (NETs) of the head and neck are rare neoplasms and can be of epithelial or non-epithelial differentiation. Although the natural history of NETs is variable, it is crucial to establish an early diagnosis of these tumours as they can be potentially curable. Conventional anatomical imaging and functional imaging using radionuclide scintigraphy and positron emission tomography/computed tomography can be complementary for the diagnosis, staging and monitoring of treatment response. This article describes and illustrates the

imaging features of head and neck NETs, discusses the potential future role of novel positron-emitting tracers that are emerging into clinical practice and reviews contemporary management of these tumours. Familiarity with the choice of imaging techniques and the variety of imaging patterns and treatment options should help guide radiologists in the management of this rare but important subgroup of head and neck neoplasms.

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[196]

TÍTULO / TITLE: - Expression of aldo-keto reductase family 1 member C3 (AKR1C3) in neuroendocrine tumors & adenocarcinomas of pancreas, gastrointestinal tract, and lung.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Int J Clin Exp Pathol. 2013 Oct 15;6(11):2419-29.

AUTORES / AUTHORS: - Chang TS; Lin HK; Rogers KA; Brame LS; Yeh MM; Yang Q; Fung KM

INSTITUCIÓN / INSTITUTION: - Department of Pathology, University of Oklahoma Health Sciences Center Oklahoma City, OK.

RESUMEN / SUMMARY: - Human aldo-keto reductase family 1 member C3 (AKR1C3) was initially identified as an enzyme in reducing 5alpha-dihydrotestosterone (5alpha-DHT) to 5alpha-androstane-3alpha, 17beta-diol (3alpha-diol) and oxidizing 3alpha-diol to androsterone. It was subsequently demonstrated to possess ketosteroid reductase activity in metabolizing other steroids including estrogen and progesterone, 11-ketoprostaglandin reductase activity in metabolizing prostaglandins, and dihydrodiol dehydrogenase x (DDx) activity in metabolizing xenobiotics. AKR1C3 was demonstrated in sex hormone-dependent tissues including testis, breast, endometrium, and prostate; in sex hormone-independent tissues including kidney and urothelium. Our previous study described the expression of AKR1C3 in squamous cell carcinoma and adenocarcinoma but not in small cell carcinoma. In this report, we studied the expression of AKR1C3 in normal tissue, adenocarcinomas (43 cases) and neuroendocrine (NE) tumors (40 cases) arising from the aerodigestive tract and pancreas. We demonstrated wide expression of AKR1C3 in superficially located mucosal cells, but not in NE cells. AKR1C3-positive immunoreactivity was detected in 38 cases (88.4%) of adenocarcinoma, but only in 7 cases (17.5%) of NE tumors in all cases. All NE tumors arising from the pancreas and appendix and most tumors from the colon and lung were negative. The highest ratio of positive AKR1C3 in NE tumors was found in tumors arising from the small intestine (50%). These results raise the question of AKR1C3's role in the biology of normal mucosal epithelia and tumors. In addition, AKR1C3 may be a useful adjunct marker for the exclusion of the NE phenotype in diagnostic pathology.

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[197]

TÍTULO / TITLE: - Adrenal incidentaloma: A case of pheochromocytoma with sub-clinical Cushing's syndrome.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Indian J Endocrinol Metab. 2013 Oct;17(Suppl 1):S246-8. doi: 10.4103/2230-8210.119587.

●● Enlace al texto completo (gratuito o de pago) 4103/2230-8210.119587

AUTORES / AUTHORS: - Goyal A; Panchani R; Varma T; Bhalla S; Tripathi S

INSTITUCIÓN / INSTITUTION: - Department of Endocrinology, Sir Ganga Ram Hospital, New Delhi, India.

RESUMEN / SUMMARY: - Adrenal incidentalomas (AIs) are a cluster of different pathologies, but AIs with dual functional aspects are very rare. We report a case of AI with the evidence of both pheochromocytoma and sub-clinical Cushing's syndrome. A 42-year-old female patient presented with the history of abdominal pain. Abdominal computed tomography revealed right adrenal mass suggestive of pheochromocytoma. On endocrine evaluation, she admitted history of intermittent headache and palpitations for 4 years and was on treatment for hypertension and diabetes. There were no signs and symptoms suggestive of Cushing's syndrome. The laboratory data demonstrated 10 times raised 24-h urinary fractionated metanephrines with non-suppressible serum cortisol after 2-day low-dose dexamethasone suppression test. She underwent right-sided adrenalectomy with subsequent resolution of both pheochromocytoma and hypercortisolism. Patient was discharged in good clinical condition.

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[198]

TÍTULO / TITLE: - Unexpected pheochromocytoma presenting as a pancreatic tumor: A case report.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Oncol Lett. 2013 Sep;6(3):833-834. Epub 2013 Jul 8.

●● Enlace al texto completo (gratuito o de pago) 3892/ol.2013.1447

AUTORES / AUTHORS: - Huang YH; Liaw WJ; Kuo CP; Wu ZF; Cherng CH; Yu JC; Horng HC; Huang ST

INSTITUCIÓN / INSTITUTION: - Department of Anesthesiology, Tri-Service General Hospital and National Defense Medical Center, Taipei 11490, Taiwan, R.O.C.

RESUMEN / SUMMARY: - A 54-year-old female presented with a large pancreatic tumor of the tail during a regular physical examination. The patient underwent surgical intervention and the surgeon identified that the tumor originated from the retroperitoneal region. Markedly severe hemodynamic fluctuations occurred during the manipulation of the tumor and continued to occur subsequent to the tumor being removed. The vital signs were adequately managed and the surgery was successful without complications. The patient was discharged without any sequelae days later. The pathology report indicated a diagnosis of pheochromocytoma. Unexpected pheochromocytoma may lead to a fatal hypertensive crisis with catastrophic sequelae during surgery. The peri-operative management of pheochromocytoma remains a complicated challenge that requires intensive pre-operative preparation and vigilant peri-operative care. For surgeons and anesthesiologists who may encounter an unexpected hypertensive crisis during abdominal tumor surgery, undiagnosed pheochromocytoma should always be considered.

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[199]

TÍTULO / TITLE: - Unsuspected paraganglioma of the urinary bladder with intraoperative hypertensive crises: A case report.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Exp Ther Med. 2013 Oct;6(4):1067-1069. Epub 2013 Aug 1.

●● Enlace al texto completo (gratuito o de pago) 3892/etm.2013.1242

AUTORES / AUTHORS: - Li S; Lui S; Li F; Yue Q; Huang X; Gong Q

INSTITUCIÓN / INSTITUTION: - Department of Radiology, Huaxi MR Research Center (HMRRC), West China Hospital of Sichuan University, Chengdu, Sichuan 610041; ; Department of Radiology, The First People's Hospital of Zunyi, Zunyi, Guizhou 653002, P.R. China.

RESUMEN / SUMMARY: - Paraganglioma of the urinary bladder is rare, accounting for <0.05% of all bladder tumors. Common clinical findings in patients with bladder paraganglioma include hematuria and intermittent hypertension during urination, along with generalized symptoms due to increased levels of catecholamines. Although unsuspected bladder paraganglioma may result in intraoperative hypertensive crises, these may be avoided if characteristic imaging signs are observed. The present study reports a case in which a patient with unsuspected paraganglioma experienced a severe hypertensive episode during cystoscopic tumor resection. Although this case had typical computed tomographic characteristics of the bladder paraganglioma, the possibility of the paraganglioma pre-operatively was not taken into account.

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[200]

TÍTULO / TITLE: - Maxillary neurilemmoma-Rarest of the rare tumour: Report of 2 cases.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Int J Surg Case Rep. 2013;4(11):1044-7. doi: 10.1016/j.ijscr.2013.09.006. Epub 2013 Sep 21.

●● Enlace al texto completo (gratuito o de pago) 1016/j.ijscr.2013.09.006

AUTORES / AUTHORS: - Verma A; Banerjee K; Verma A; Singh S; Rao J; Om P

INSTITUCIÓN / INSTITUTION: - S.M.S. Medical College and Hospital, Jaipur 403002, Rajasthan, India. Electronic address: ankit.doc2004@gmail.com.

RESUMEN / SUMMARY: - INTRODUCTION: Intraoral neurilemmomas (schwannoma) are rare, even rarer are intraosseous ones, and the rarest are the maxillary neurilemmomas. Going by the literature only 5 cases of maxillary neurilemomas are reported till now. Neurilemmomas are benign tumours of nerve sheath origin. Approximately 30% arise in head and neck region, of these 1-12% arise intraorally mainly involving tongue. PRESENTATION OF CASE: Here we report two cases of maxillary neurilemmoma, one in a 9 year old girl and second one in a 27 year old female, both involving the lateral surface of maxilla on right side. Both the patients presented with a long standing history of swelling which was increasing gradually. 9-Year-old girl also had 1 lesion in the temporal region on right side and the 27-year-old patient had associated erosion of the soft palate. Diagnosis was made on the basis of histopathology and immunohistochemistry. DISCUSSION: Neurilemmomas are slow growing benign tumour of the nerve sheath origin arising from the Schwann cells. Their aetiology is not known. Most common complaint is that of a gradually increasing swelling followed by pain and paresthesias. Surgery remains the treatment of choice with close follow up. CONCLUSION: Maxillary neurilemmomas are rarest of the rare tumour which closely mimic benign odontogenic cysts and tumours, and should be kept in the differential diagnosis of these lesions. Knowledge of the radiologic and clinical behaviour of these tumours is extremely important for prompt diagnosis and treatment.

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[201]

TÍTULO / TITLE: - Neuroendocrine Dysplasia Combined in a Tubular Adenoma of Rectum: A Case Report.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Korean J Pathol. 2013 Oct;47(5):495-498. Epub 2013 Oct 25.
●● Enlace al texto completo (gratuito o de pago)
4132/KoreanJPathol.2013.47.5.495
AUTORES / AUTHORS: - Lee SY; Hwang DY; Hwang TS; Kim WS; Lim SD; Kim WY; Kim SH; Han HS
INSTITUCIÓN / INSTITUTION: - Department of Pathology, Konkuk University Medical Center, Konkuk University School of Medicine, Seoul, Korea.

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[202]
TÍTULO / TITLE: - Metastatic Merkel cell carcinoma (MCC) of pancreas and breast: a unique case.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - World J Surg Oncol. 2013 Oct 7;11(1):261. doi: 10.1186/1477-7819-11-261.
●● Enlace al texto completo (gratuito o de pago) 1186/1477-7819-11-261
AUTORES / AUTHORS: - Vernadakis S; Moris D; Bankfalvi A; Makris N; Sotiropoulos GC
INSTITUCIÓN / INSTITUTION: - Department of General, Visceral and Transplantation Surgery, University Hospital Essen, Hufelandstr, 55, Esse 45122, Germany. svernadakis@yahoo.com.
RESUMEN / SUMMARY: - Merkel cell carcinoma (MCC) is a rare potentially fatal skin tumor affecting older and immunosuppressed individuals. It is highly malignant with high rates of metastasis and poor survival.We present a case of a 67-year-old woman with a palpable mass in the upper abdomen. An abdominal CT revealed a mass in the tail of the pancreas. Two weeks before, lumpectomy of a 3.5 cm tumor of the left breast had been performed. Histology showed a primary neuroendocrine carcinoma of the mammary gland. The patient's medical history was significant for a 0.7 x 0.9 cm MCC removed from her left forearm 2.5 years ago. There was no evidence of vascular involvement or peritoneal disease and by all criteria was resectable. A somatostatin receptor scintigraphy showed an enhanced uptake in the pancreatic tail region. The tumor was immunohistochemically strong staining for synaptophysin and CD56. The diagnosis of a metastatic-MCC in the tail of the pancreas was made. Further histological investigation of the prior removed neuroendocrine breast tumor and the MCC of the left forearm confirmed neuroendocrine origin and identical histology to the previously resected MCC of the left forearm. In this article, we aim to highlight that MCC has the potential to spread even in unusual organs, such as pancreas or breast, and therefore a diligent follow-up should be applied in patients with MCC.

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[203]
TÍTULO / TITLE: - Renal medullary carcinoma response to chemotherapy: a referral center experience in Brazil.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Rare Tumors. 2013 Aug 20;5(3):e44. doi: 10.4081/rt.2013.e44.
●● Enlace al texto completo (gratuito o de pago) 4081/rt.2013.e44
AUTORES / AUTHORS: - Maroja Silvino MC; Venchiarutti Moniz CM; Munhoz Piotto GH; Siqueira S; Galapo Kann A; Dzik C

INSTITUCIÓN / INSTITUTION: - Instituto do Cancer de Sao Paulo, Faculdade de Medicina da Universidade de Sao Paulo.

RESUMEN / SUMMARY: - Renal medullary carcinoma (RMC) is rare, accounting for less than 1% of all renal neoplasms. Case reports suggest RMC is highly aggressive, poorly responsive to chemotherapy, often metastatic at diagnosis, affects young men with sickle cell trait, and median overall survival (mOS) is less than 12 months. We report the epidemiological characteristics, treatments performed, response rate to each treatment and mOS of five patients with RMC. All patients had sickle cell trait, four were male, three had metastatic disease at diagnosis and mean age at diagnosis was 25 years. Non-metastatic patients were submitted to nephrectomy. Two patients had partial response to first line chemotherapy including cisplatin and gemcitabine. There was no response to sunitinib or second line chemo - therapy; mOS was 6 months. Due to its rarity, case series are the only evidence available to discuss the treatment for RMC. In our experience, only cisplatin and gemcitabine based regimen offered response.

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[204]

TÍTULO / TITLE: - Ultrasound-aided diagnosis of an insulinoma in a cat.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Tierarztl Prax Ausg K Kleintiere Heimtiere. 2013;41(5):338-42.

AUTORES / AUTHORS: - Schaub S; Wigger A

INSTITUCIÓN / INSTITUTION: - Sebastian Schaub, Tierarztliche Praxis Dres. Schaub, Waldstrasse 23, 07745 Jena, Germany, Email: sebastian.schaub@vetmed.uni-giessen.de.

RESUMEN / SUMMARY: - A 15-year old, neutered female, domestic shorthaired cat was presented for evaluation of a 3-month history of paroxysmal falling over and trembling. In laboratory work the cat displayed a mild hypoglycemia. Ultrasound revealed a nodule in the left pancreatic lobe and surgical excision was performed. The histological diagnosis was an insulinoma. To the authors knowledge this is the first ultrasound description of an insulinoma in a cat. Up to date the cat has a survival time of 32 months without recurrence of symptoms.

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[205]

TÍTULO / TITLE: - Prognostic impact of p16 and p21 on gastroenteropancreatic neuroendocrine tumors.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Oncol Lett. 2013 Dec;6(6):1641-1645. Epub 2013 Oct 9.

●● Enlace al texto completo (gratuito o de pago) 3892/ol.2013.1610

AUTORES / AUTHORS: - Liu S; Chang Y; Ma J; Li X; Li X; Fan J; Huang R; Duan G; Sun X

INSTITUCIÓN / INSTITUTION: - Department of Epidemiology, College of Public Health of Zhengzhou University, Zhengzhou, Henan 450001, P.R. China ; Henan Cancer Research and Control Office, Henan Cancer Hospital, Zhengzhou, Henan 450008, P.R. China.

RESUMEN / SUMMARY: - Aberrant expression of the cell cycle kinase inhibitors, p16 and p21, has been associated with poor prognosis in a number of human malignancies. These proteins may also be involved in the development and progression of gastroenteropancreatic neuroendocrine tumors (GEP-NETs). The

present study aimed to investigate protein levels of p16 and p21 in GEP-NETs and to evaluate their clinical significance. p16 and p21 protein expression was tested immunohistochemically in the tissue samples of 68 GEP-NETs. The association between expression and clinicopathological characteristics and overall survival was assessed. Low expression of p16 (no positive nuclear staining) was found in 37 (54%) cases and high p21 expression (>/=5% positive nuclear staining) was detected in 23 (34%) cases. Low p16 protein levels indicated a poorer prognosis for patients graded as G2 subgroup in the univariate analysis (relative risk, 4.4; 95% CI, 1.8-10.6). No significant correlation was found between the expression of p21 and any of the clinicopathological variables. The present study indicates a prognostic relevance for p16 immunoreactivity. Low levels of p16 protein were associated with a shorter survival in the G2 subgroup of GEP-NETs. p21 protein expression was not identified to be useful as a predictive indicator in GEP-NETs.

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[206]

TÍTULO / TITLE: - Merkel Cell Carcinoma of the Eyelid: Management and Prognosis.

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: <> Ophthalmol. 2013 Nov 28. doi: 10.1001/jamaophthalmol.2013.6077.

●● Enlace al texto completo (gratuito o de pago) 1001/jamaophthalmol.2013.6077

AUTORES / AUTHORS: - Herbert HM; Sun MT; Selva D; Fernando B; Saleh GM; Beaconsfield M; Collin R; Uddin J; Meligonis G; Leatherbarrow B; Ataullah S; Irion L; McLean CJ; Huilgol SC; Davis G; Sullivan TJ

INSTITUCIÓN / INSTITUTION: - Moorfields Eye Hospital, London, England.

RESUMEN / SUMMARY: - IMPORTANCE The literature on Merkel cell carcinoma (MCC) of the eyelid remains scarce, and there has yet to be a study using the most up-to-date TNM staging system for this rare but aggressive tumor. OBJECTIVE To analyze the TNM stage, management, and outcomes of patients with MCC of the eyelid. DESIGN, SETTING, AND PARTICIPANTS Retrospective case series of 21 patients from 5 tertiary referral centers in the United Kingdom and Australia with primary MCC of the eyelid presenting at a median age of 77 years, with median follow-up of 54 months. Tumors were staged according to the American Joint Committee on Cancer, 7th edition, TNM criteria for eyelid carcinoma and MCC. MAIN OUTCOMES AND MEASURES TNM stage, treatment modalities, and clinical outcome. RESULTS The eyelid carcinoma TNM stages were T2aN0M0 for 5 patients, T2bN0M0 for 7 patients, T3aN0M0 for 4 patients, T3bN0M0 for 3 patients, T2bN1M0 for 1 patient, and T3aN1M0 for 1 patient. The MCC TNM stages were T1N0M0 for 12 patients, T2N0M0 for 7 patients, T1N1M0 for 1 patient, and T2N1M0 for 1 patient. One patient had a sentinel lymph node biopsy, and 8 patients underwent head/neck imaging. Eighteen patients underwent a wide local excision, 12 with a paraffin section and 6 with a frozen section. Two patients underwent Mohs surgery, 1 of whom required an orbital exenteration. Twelve patients (57%) received adjuvant radiotherapy, and 2 patients received chemotherapy. The local recurrence rate was 10%, the regional nodal recurrence rate was 10%, and the distant metastatic recurrence rate was 19%. The lowest T category tumor metastasizing to both regional nodes and distant locations

was a T2a (eyelid TNM)/T1 (Merkel TNM) tumor measuring 8 mm. Two patients with T3a (eyelid TNM)/T2 (Merkel TNM) tumors died of metastatic MCC. CONCLUSIONS AND RELEVANCE The majority of patients with MCC of the eyelid present with localized eyelid disease of T category T2 (eyelid TNM)/T1 (Merkel TNM). A wide local excision with margin control remains the mainstay of treatment, whereas the use of radiotherapy is institution specific. Tumors with a low T category are associated with regional nodal and distant metastatic disease. It may therefore be reasonable to consider a sentinel lymph node biopsy or strict regional lymph node surveillance for all MCCs of the eyelid, regardless of T category or size.

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[207]

TÍTULO / TITLE: - Are the (18)F-FDG positron emission tomography/computed tomography findings in bronchopulmonary carcinoid tumors different than expected?

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Hell J Nucl Med. 2013 Sep-Dec;16(3):213-7.

AUTORES / AUTHORS: - Alpay L; Lacin T; Kanbur S; Kiral H; Ersoz E; Bayram S; Dogruyol T; Baysungur V; Yalcinkaya I

INSTITUCIÓN / INSTITUTION: - Sureyyapasa Training and Research Hospital, Department of Thoracic Surgery Basibuyuk Mah, Maltepe, 34844, Istanbul, Turkey. leventalpay@yahoo.com.

RESUMEN / SUMMARY: - Bronchopulmonary carcinoid tumors (BPCT) are known as low malignity tumors. Different surgical methods are therapeutically used, ranging from simple excision of the mass to large regional resections. Also, the role of positron emission tomography in the diagnosis and staging of BPCT is controversial as false negative results has been reported in literature. Our aim was to study the diagnostic value of fluorine-18 fluorodeoxyglucose positron emission tomography/computed tomography ((18)F-FDG PET/CT) and the therapeutic effect of specific surgical treatment on BPCT. We studied retrospectively from 2005 to 2011 75 cases of BPCT. Preoperative investigations included computerized tomography (CT), bronchoscopy and (18)F-FDG PET. Statistical comparisons were performed based on tumor type, extent of the resection and the standardized uptake value (SUV). Fifty six cases were typical, 15 atypical and 4 oncocytic (a subtype of typical carcinoid). Of these patients, 27 (17 with typical, 8 with atypical and 2 with oncocytic carcinoid) had undergone a (18)F-FDG PET scan. Operatory mortality was 0%, while the 7 years survival rate amounted to 97.5%. No recurrences were seen. Mean SUV was 5.28 for typical and 5.08 for atypical BPCT. The oncocytic type exhibited a particularly high SUV. In conclusion, our study, contrary to the findings of others, showed that the (18)F-FDG uptake of BPCT was similar to that of malignant diseases. Aggressive surgical treatment resulted in a very good prognosis for these carcinoid tumors.

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[208]

TÍTULO / TITLE: - Bone metastasis from a neuroendocrine tumor detected by 99m-technetium-hydrazinonicotinyl-Tyr3-octreotide single-photon emission computed tomography/computed tomography.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Indian J Nucl Med. 2013 Jul;28(3):187-8. doi: 10.4103/0972-3919.119520.

●● Enlace al texto completo (gratuito o de pago) 4103/0972-3919.119520

AUTORES / AUTHORS: - Kamaleshwaran KK; Subramanian PV; Natarajan S; Mohanan V; Shinto AS
INSTITUCIÓN / INSTITUTION: - Department of Nuclear Medicine and PET/CT, Kovai Medical Centre and Hospital Limited, Coimbatore, Tamil Nadu, India.

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[209]
TÍTULO / TITLE: - Cancer: Biochemical diagnosis of phaeochromocytomas and paragangliomas requires supine blood sampling.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Nat Rev Endocrinol. 2013 Dec;9(12):692. doi: 10.1038/nrendo.2013.195. Epub 2013 Oct 1.

●● Enlace al texto completo (gratuito o de pago) 1038/nrendo.2013.195

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[210]
TÍTULO / TITLE: - Pancreatic paraganglioma: An extremely rare entity and crucial role of immunohistochemistry for diagnosis.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Indian J Endocrinol Metab. 2013 Sep;17(5):917-9. doi: 10.4103/2230-8210.117217.

●● Enlace al texto completo (gratuito o de pago) 4103/2230-8210.117217

AUTORES / AUTHORS: - Borgohain M; Gogoi G; Das D; Biswas M
INSTITUCIÓN / INSTITUTION: - Department of Pathology, Assam Medical College and Hospital, Dibrugarh, Assam, India.
RESUMEN / SUMMARY: - Paragangliomas are rare neuroendocrine neoplasms arising in extra-adrenal chromaffin cells of autonomic nervous system and histologically akin to chemodectomas. They are rare, affecting about 1 in 2,000,000 population. It is a generic term applied to tumors of paraganglia regardless of the location. In rare instances, paragangliomas present around and involve the pancreas, thereby mimicking any one of the more common primary pancreatic lesions. Pancreatic paraganglioma is an extremely rare tumor. It grows slowly, so radical resection is recommended to achieve curability with good prognosis. These neoplasms present considerable diagnostic difficulty not only for the clinician and radiologist but also for the pathologist. Here, we report a case of a 55-year-old woman who presented with a left-sided abdominal swelling for 3 months duration, initially having clinical suspicion of an ovarian tumor. The radiological imaging revealed a lesion in the tail of pancreas with a differential diagnosis of pancreatic carcinoma and metastatic tumor. Only after exploratory laparotomy, the diagnosis was made as a rare case of pancreatic paraganglioma on the basis of histological examination and immunohistochemistry.

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[211]
TÍTULO / TITLE: - Synchronous bilateral bronchial carcinoid diagnosed with combined dual tracer (F-FDG and Ga-DOTATOC) PET/CT scans.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Rev Esp Med Nucl. Acceso gratuito al texto completo a partir de los 2 años de la fecha de publicación.

●● Enlace a la Editora de la Revista http://db.doyma.es/

●● Cita: Revista Española de Medicina Nuclear: <> Imagen Mol. 2013 Oct 16. pii: S2253-654X(13)00160-1. doi: 10.1016/j.remn.2013.08.007.

●● Enlace al texto completo (gratuito o de pago) 1016/j.remn.2013.08.007

AUTORES / AUTHORS: - Paci M; Lococo F; Rapicetta C; Roncali M; Cavazza A; Treglia G; Sgarbi G

INSTITUCIÓN / INSTITUTION: - Unit of Thoracic Surgery, IRCCS Arcispedale Santa Maria Nuova, Reggio Emilia, Italy.

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[212]

TÍTULO / TITLE: - Role of radiotherapy for pancreatobiliary neuroendocrine tumors.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Radiat Oncol J. 2013 Sep;31(3):125-30. doi: 10.3857/roj.2013.31.3.125. Epub 2013 Sep 30.

●● Enlace al texto completo (gratuito o de pago) 3857/roj.2013.31.3.125

AUTORES / AUTHORS: - Lee J; Choi J; Choi C; Seong J

INSTITUCIÓN / INSTITUTION: - Department of Radiation Oncology, Yonsei Cancer Center, Yonsei University College of Medicine, Seoul, Korea.

RESUMEN / SUMMARY: - PURPOSE: We investigated the role of radiotherapy (RT) for pancreatobiliary neuroendocrine tumors (PB-NETs). MATERIALS AND METHODS: We identified 9 patients with PB-NETs who received RT between January 2005 and March 2012. Of these 9 patients, 4 were diagnosed with NETs in the pancreas and 5 were diagnosed with NETs in the gallbladder. All patients received RT to the primary tumor or resection bed with a median total irradiation dose of 50.4 Gy, with or without chemotherapy. RESULTS: The tumor response rate and tumor control rate in the RT field were 60% and 100 %, respectively. All 4 patients who underwent surgery had no evidence of disease in the RT field. Of the 5 patients who received RT to the primary gross tumor, 1 had complete response, 2 had partial response, and 2 had stable disease in the RT field. The median time to progression was 11 months. Of the 9 patients, four patients had no progression, and 5 patients had progression of disease (locoregional, 2; distant, 2; locoregional/distant, 1). Of the 4 patients without progression, 3 were treated with RT in adjuvant or neoadjuvant setting, and one received RT to primary tumor. One patient experienced radiation-induced duodenitis at 3 months after concurrent chemoradiation without treatment-related mortality. CONCLUSION: RT can yield local control for advanced PB-NETs. RT should be considered an essential part of multimodality treatment in management of advanced PB-NETs.

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[213]

TÍTULO / TITLE: - Chromogranin a and its fragments as regulators of small intestinal neuroendocrine neoplasm proliferation.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - PLoS One. 2013 Nov 19;8(11):e81111. doi: 10.1371/journal.pone.0081111.

●● Enlace al texto completo (gratuito o de pago) 1371/journal.pone.0081111

AUTORES / AUTHORS: - Giovinazzo F; Schimmack S; Svejda B; Alaimo D; Pfragner R; Modlin I; Kidd M

INSTITUCIÓN / INSTITUTION: - Department of Surgery, Yale University School of Medicine, New Haven, Connecticut, United States of America ; Laboratory of Translational Surgery-LURM, University of Verona, Verona, Italy.

RESUMEN / SUMMARY: - INTRODUCTION: Chromogranin A is a neuroendocrine secretory product and its loss is a feature of malignant NEN de-differentiation. We hypothesized that chromogranin A fragments were differentially expressed during NEN metastasis and played a role in the regulation of NEN proliferation. METHODS: Chromogranin A mRNA (PCR) and protein (ELISA/western blot) were studied in 10 normal human mucosa, 5 enterochromaffin cell preparations, 26 small intestinal NEN primaries and 9 liver metastases. Cell viability (WST-1 assay), proliferation (bromodeoxyuridine ELISA) and expression of AKT/AKT-P (CASE ELISA/western blot) in response to chromogranin A silencing, inhibition of prohormone convertase and mTOR inhibition (RAD001/AKT antisense) as well as different chromogranin A fragments were examined in 4 SI-NEN cell lines. RESULTS: Chromogranin A mRNA and protein levels were increased (37-340 fold, $p<0.0001$) in small intestinal NENs compared to normal enterochromaffin cells. Western blot identified chromogranin A-associated processing bands including vasostatin in small intestinal NENs as well as up-regulated expression of prohormone convertase in metastases. Proliferation in small intestinal NEN cell lines was decreased by silencing chromogranin A as well as by inhibition of prohormone convertase ($p<0.05$). This inhibition also decreased secretion of chromogranin A ($p<0.05$) and 5-HT ($p<0.05$) as well as expression of vasostatin. Metastatic small intestinal NEN cell lines were stimulated (50-80%, $p<0.05$) and AKT phosphorylated (Ser473: $p<0.05$) by vasostatin I, which was completely reversed by RAD001 ($p<0.01$) and AKT antisense ($p<0.05$) while chromostatin inhibited proliferation (~50%, $p<0.05$). CONCLUSION: Chromogranin A was differentially regulated in primary and metastatic small intestinal NENs and cell lines. Chromogranin A fragments regulated metastatic small intestinal NEN proliferation via the AKT pathway indicating that CgA plays a far more complex role in the biology of these tumors than previously considered.

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[214]

TÍTULO / TITLE: - Aberrant Promoter Hypermethylation of RASSF Family Members in Merkel Cell Carcinoma.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Cancers (Basel). 2013 Nov 18;5(4):1566-1576.

●● Enlace al texto completo (gratuito o de pago) 3390/cancers5041566

AUTORES / AUTHORS: - Richter AM; Haag T; Walesch S; Herrmann-Trost P; Marsch WC; Kutzner H; Helmbold P; Dammann RH

INSTITUCIÓN / INSTITUTION: - Institute for Genetics, University of Giessen, Giessen D-35392, Germany. Reinhard.Dammann@gen.bio.uni-giessen.de.

RESUMEN / SUMMARY: - Merkel cell carcinoma (MCC) is one of the most aggressive cancers of the skin. RASSFs are a family of tumor suppressors that are frequently inactivated by promoter hypermethylation in various cancers. We studied CpG island promoter hypermethylation in MCC of RASSF2, RASSF5A, RASSF5C and RASSF10 by combined bisulfite restriction analysis (COBRA) in MCC samples and control tissue. We found RASSF2 to be methylated in three out of 43 (7%), RASSF5A in 17 out of 39 (44%, but also 43% in normal tissue), RASSF5C in two out of 26 (8%) and RASSF10 in 19 out of 84 (23%) of the cancer samples. No correlation between the methylation status of the analyzed RASSFs or between RASSF methylation and MCC characteristics (primary versus metastatic, Merkel cell polyoma virus infection, age, sex) was found. Our results show that RASSF2, RASSF5C and RASSF10 are

aberrantly hypermethylated in MCC to a varying degree and this might contribute to Merkel cell carcinogenesis.

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[215]

TÍTULO / TITLE: - Successful endoscopic mucosal resection of a low esophageal carcinoid tumor.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Clin Endosc. 2013 Sep;46(5):576-8. doi: 10.5946/ce.2013.46.5.576. Epub 2013 Sep 30.

●● Enlace al texto completo (gratuito o de pago) 5946/ce.2013.46.5.576

AUTORES / AUTHORS: - Lim CS; Park SJ; Park MI; Moon W; Kim HH; Lee JS; Kim BJ; Ku DY

INSTITUCIÓN / INSTITUTION: - Department of Internal Medicine, Kosin University College of Medicine, Busan, Korea.

RESUMEN / SUMMARY: - Esophageal carcinoid tumors remain some of the rarest of all carcinoid tumors, with only several cases previously reported in the literature. The endoscopic mucosal resection of selected carcinoid tumors has been shown to be a valid, safe, and effective method of treatment. Endoscopic ultrasonography is the technique of choice to select patients eligible for endoscopic resection. Here, we report successful endoscopic mucosal resection of a low esophageal carcinoid tumor and review the relevant literature. The present case is the first reported case of esophageal carcinoid tumor in Korea.

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[216]

TÍTULO / TITLE: - Resection of a malignant paraganglioma located behind the retrohepatic segment of the inferior vena cava.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - BMC Surg. 2013 Oct 29;13(1):49.

●● Enlace al texto completo (gratuito o de pago) 1186/1471-2482-13-49

AUTORES / AUTHORS: - Jia C; Wang X; Dai C; Bu X; Peng S; Xu F; Xu Y; Zhao Y

RESUMEN / SUMMARY: - BACKGROUND: Resection of a retrocaval paraganglioma is technically challenging due to limited tumor accessibility and proximity to the vena cava. CASE PRESENTATION: A large, malignant paraganglioma was found behind the retrohepatic segment of the inferior vena cava of a 60-year-old male. During resection of this rare paraganglioma, the left lateral lobe of the liver, a portion of the caudate lobe of the liver, and the gallbladder were also removed. Unfortunately, the patient died six months after surgery due to hepatic metastasis. CONCLUSION: This case demonstrates that a partial hepatectomy may be necessary to improve tumor accessibility during resection of a retrocaval paraganglioma, particularly if the tumor is proximal to the vena cava. Furthermore, palliative treatments may help prevent tumor recurrence and metastasis of malignant paragangliomas.

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[217]

TÍTULO / TITLE: - Concomitant elective resection of pheochromocytoma and repair of aortic abdominal aneurysm.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - JRSM Cardiovasc Dis. 2013 Aug 14;2:2048004013493403. doi: 10.1177/2048004013493403.

●● Enlace al texto completo (gratuito o de pago) 1177_2048004013493403 [pii
●● Enlace al texto completo (gratuito o de pago) 1177/2048004013493403
AUTORES / AUTHORS: - Matadial C; Giquel J; Martinez-Ruiz R
INSTITUCIÓN / INSTITUTION: - Department of Clinical Anaesthesiology, University of Miami, Miller School of Medicine, Veteran Hospital, USA.
RESUMEN / SUMMARY: - Perioperative management of a patient with ischemic heart disease with coexisting abdominal aortic aneurysm and pheochromocytoma creates a difficult management dilemma, and surgical intervention in these patients carries a significant risk. The state of catecholamine excess and various other coexisting factors can lead to simultaneous occurrence of abdominal aortic aneurysm and pheocromocytoma. The purpose of this report is to present an integrated approach to the management of concomitant abdominal aortic aneurysm and pheochromocytoma, where a combined surgical approach in addressing these two lesions was preferable due to patient comorbidities and surgical implications without significant complication.

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[218]
TÍTULO / TITLE: - Laparoscopic phrenectomy for a diaphragmatic neurilemmoma.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - J Res Med Sci. 2013 Jun;18(6):522-5.
AUTORES / AUTHORS: - Liu K; Zhang M; Liang X; Cai X
INSTITUCIÓN / INSTITUTION: - Department of Hepatobiliary Surgery, Ocean University of China, Qingdao 266071, China.
RESUMEN / SUMMARY: - Diaphragmatic neurilemmoma (schwannoma) is an extremely rare tumor that is often discovered incidentally. Even when diagnosed, patients are commonly advised only to attend regular follow-up appointments as conventional tumorectomy is enormously invasive and confers relatively few benefits. Here, we report a unique case of a diaphragmatic neurilemmoma with concomitant symptomatic cholecystolithiasis, who was treated successfully by pure laparoscopy. The entire operation lasted 65 min, and patient was discharged uneventfully on the 3(rd) post-operative day. The follow-up for 29 months has shown with no recurrence or symptoms.

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[219]
TÍTULO / TITLE: - Investigation of factors potentially influencing calcitonin levels in the screening and follow-up for medullary thyroid carcinoma: a cautionary note.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - BMC Clin Pathol. 2013 Nov 4;13(1):27.
●● Enlace al texto completo (gratuito o de pago) 1186/1472-6890-13-27
AUTORES / AUTHORS: - Guesgen C; Willms A; Zwad A; Waldeck S; Wieler H; Schwab R
RESUMEN / SUMMARY: - BACKGROUND: The malignant transformation of thyroid C cells is associated with an increase in human calcitonin (hCT), which can thus be helpful in the early diagnosis of medullary thyroid carcinoma (MTC). For this reason, hCT levels should be determined in all patients with nodular goitre. Hashimoto's thyroiditis, nodular goitre and proton pump inhibitor (PPI) therapy are factors reported to influence basal serum hCT concentrations. The diagnostic role of mildly to moderately increased hCT levels is thus a matter of debate. In this study, we attempt to clarify the role of the aforementioned factors. METHODS: From 2008 to 2009, we

collected data from 493 patients who were divided into five groups. We assessed whether there were significant differences in hCT levels between patients with Hashimoto's thyroiditis, patients with nodular goitre, patients with PPI therapy, and healthy control subjects. In addition, we investigated whether a delayed analysis of blood samples has an effect on serum hCT concentrations. RESULTS: Immunoradiometric assays (Calcitonin IRMA magnum, MEDIPAN) revealed that the time of analysis did not play a role when low levels were measured. Delayed analysis, however, carried the risk of false low results when serum hCT concentrations were elevated. Men had significantly higher serum hCT levels than women. The serum hCT concentrations of patients with Hashimoto's thyroiditis and nodular goitre were not significantly different from those of control subjects. Likewise, PPI therapy did not lead to a significant increase in serum hCT concentrations regardless of the presence or absence of nodular goitre. CONCLUSIONS: Increases in serum hCT levels are not necessarily attributable to Hashimoto's thyroiditis, nodular goitre or the regular use of PPIs and always require further diagnostic attention.

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[220]

TÍTULO / TITLE: - Copy number alterations in small intestinal neuroendocrine tumors determined by array comparative genomic hybridization.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - BMC Cancer. 2013 Oct 29;13(1):505.

●● Enlace al texto completo (gratuito o de pago) 1186/1471-2407-13-505

AUTORES / AUTHORS: - Hashemi J; Fotouhi O; Sulaiman L; Kjellman M; Hoog A; Zedenius J; Larsson C

RESUMEN / SUMMARY: - BACKGROUND: Small intestinal neuroendocrine tumors (SI-NETs) are typically slow-growing tumors that have metastasized already at the time of diagnosis. The purpose of the present study was to further refine and define regions of recurrent copy number (CN) alterations (CNA) in SI-NETs. METHODS: Genome-wide CNAs was determined by applying array CGH (a-CGH) on SI-NETs including 18 primary tumors and 12 metastases. Quantitative PCR analysis (qPCR) was used to confirm CNAs detected by a-CGH as well as to detect CNAs in an extended panel of SI-NETs. Unsupervised hierarchical clustering was used to detect tumor groups with similar patterns of chromosomal alterations based on recurrent regions of CN loss or gain. The log rank test was used to calculate overall survival. Mann—Whitney U test or Fisher's exact test were used to evaluate associations between tumor groups and recurrent CNAs or clinical parameters. RESULTS: The most frequent abnormality was loss of chromosome 18 observed in 70% of the cases. CN losses were also frequently found of chromosomes 11 (23%), 16 (20%), and 9 (20%), with regions of recurrent CN loss identified in 11q23.1-qter, 16q12.2-qter, 9pter-p13.2 and 9p13.1-11.2. Gains were most frequently detected in chromosomes 14 (43%), 20 (37%), 4 (27%), and 5 (23%) with recurrent regions of CN gain located to 14q11.2, 14q32.2-32.31, 20pter-p11.21, 20q11.1-11.21, 20q12-qter, 4 and 5. qPCR analysis confirmed most CNAs detected by a-CGH as well as revealed CNAs in an extended panel of SI-NETs. Unsupervised hierarchical clustering of recurrent regions of CNAs revealed two separate tumor groups and 5 chromosomal clusters. Loss of chromosomes 18, 16 and 11 and again of chromosome 20 were found in both tumor groups. Tumor group II was enriched for alterations in chromosome cluster-d, including gain of chromosomes 4, 5, 7, 14 and gain of 20 in chromosome cluster-b. Gain in 20pter-p11.21 was associated with short

survival. Statistically significant differences were observed between primary tumors and metastases for loss of 16q and gain of 7. CONCLUSION: Our results revealed recurrent CNAs in several candidate regions with a potential role in SI-NET development. Distinct genetic alterations and pathways are involved in tumorigenesis of SI-NETs.

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[221]

TÍTULO / TITLE: - Malignant head/neck paragangliomas. Comparative Study.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Eur Ann Otorhinolaryngol Head Neck Dis. 2013 Nov 14. pii: S1879-7296(13)00127-0. doi: 10.1016/j.anorl.2013.05.003.

●● Enlace al texto completo (gratuito o de pago) 1016/j.anorl.2013.05.003

AUTORES / AUTHORS: - Mediouni A; Ammari S; Wassef M; Gimenez-Roqueplo AP; Laredo JD; Duet M; Tran Ba Huy P; Oker N

INSTITUCIÓN / INSTITUTION: - Service de medecine nucleaire, hopital Lariboisiere (Assistance Publique-Hopitaux de Paris), 2, rue Ambroise-Pare, 75010 Paris, France.

RESUMEN / SUMMARY: - BACKGROUND: The objective of this study was to report 11 cases of malignant head and neck paraganglioma and to compare their epidemiological, clinical, and genetic characteristics, their natural history and their treatment with those of a series of 131 benign paragangliomas. PATIENTS AND METHODS: Retrospective analysis of 142 patients with head and neck paraganglioma managed between 2001 and 2008. Age at the time of diagnosis, gender, primary tumour site, presence of other non-head/neck paragangliomas and/or metastases diagnosed by imaging (CT, MRI, Octreoscan or 18F-FDG PET), histology, urinary catecholamine and metanephrine levels, family history, and genetic test results were recorded. RESULTS: This series comprised 131 benign head and neck paragangliomas, mostly observed in women with a mean age at diagnosis of 45 years and a predominance of tympanojugular sites (followed by carotid and vagal sites) with 5% of secreting tumours and 20% of multifocal tumours. Eleven patients (7.7%) with a 1:1 sex ratio presented criteria of malignancy. These patients, with a lower mean age (38 years), predominantly presented carotid lesions with a higher rate of secreting and multifocal tumours, 27% and 46% respectively. The main sites of metastases were bone and lymph nodes. No tympanic paragangliomas were observed. CONCLUSIONS: Malignant paragangliomas are mainly observed in young patients with multifocal tumours, particularly carotid tumours, and are predominantly related to subunit SDH-B mutation. The work-up in these high-risk patients must include whole body scintigraphy and spine MRI. Malignancy is not necessarily associated with a poor short-term prognosis due to the slow course of the disease.

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[222]

TÍTULO / TITLE: - Single institutional series of neuroendocrine tumors managed in the Australian Capital Territory.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Asia Pac J Clin Oncol. 2013 Oct 24. doi: 10.1111/ajco.12121.

●● Enlace al texto completo (gratuito o de pago) 1111/ajco.12121

AUTORES / AUTHORS: - Malik L; Chua YJ; Butt NS; Yip D

INSTITUCIÓN / INSTITUTION: - Medical Oncology Unit, The Canberra Hospital, Garran, Australian Capital Territory, Australia.

RESUMEN / SUMMARY: - AIMS: Retrospective review of neuroendocrine tumors (NETs) treated within the Australian Capital Territory to describe the local epidemiology and assess prognostic clinicopathological factors. METHODS: Patients with histologically proven non-pulmonary low to intermediate grade NETs were identified from our hospital clinical database. Data were analyzed according to epidemiological, clinical and histopathological characteristics. RESULTS: Of the 107 included patients, the most common primary tumor site was jejunum/ileum (32%), followed by rectum (22%) and pancreas (11.2%). In total, 32% had distant metastases at presentation, most commonly in the liver. Most patients were symptomatic at diagnosis, while 22.4% of cases were found incidentally. Second malignancies, in particular of gastrointestinal origin, were diagnosed in 33.6%. Surgical debulking was the most common treatment (59.8%) while 18% had multimodality therapy. With a median follow-up of 25 months from diagnosis, about 78% of patients are still alive. Median time to first relapse was 15 months and the 5-year survival rate was 80% for NETs of jejunum/ileum. Univariate survival analysis revealed tumor location, high Ki67 index, raised plasma chromogranin A, and urine 5-hydroxyindoleacetic acid upon diagnosis to be associated with shorter 5-year survival. CONCLUSION: The epidemiologic characteristics and long-term outcome in our series are comparable to other reported studies. This analysis presents some important prognostic factors which could be used for risk stratification in patients with NETs.

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[223]

TÍTULO / TITLE: - Mixed Medullary-Follicular Carcinoma of the Thyroid.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Case Rep Endocrinol. 2013;2013:571692. Epub 2013 Oct 29.

●● Enlace al texto completo (gratuito o de pago) 1155/2013/571692

AUTORES / AUTHORS: - Tohidi M; Pourbehi G; Bahmanyar M; Eghbali SS; Kalantar Hormozi M; Nabipour I

INSTITUCIÓN / INSTITUTION: - The Persian Gulf Marine Medicine Biotechnology Research Center, Department of Endocrinology, Bushehr University of Medical Sciences, Bushehr, Iran.

RESUMEN / SUMMARY: - Introduction. Mixed medullary-follicular thyroid carcinoma is an uncommon tumor that consists of both follicular and parafollicular cells. Case. We report a 43-year-old woman with a palpable mass in the right side of the neck. Fine needle aspiration suggested a diagnosis of high grade anaplastic carcinoma that has been associated with papillary features. Total thyroidectomy was done in which histopathological examination showed diagnosis of medullary carcinoma. Immunohistochemical staining was positive for chromogranin, calcitonin, and thyroglobulin in tumoral cells. Conclusion. Mixed medullary-follicular thyroid carcinoma is a rare tumor. Diagnosis of these tumors with fine needle aspiration is very difficult and may lead to misdiagnosis. It is necessary to correlate the cytological finding with serum calcitonin and thyroglobulin. Also immunostaining for calcitonin and thyroglobulin confirms diagnosis.

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[224]

TÍTULO / TITLE: - Tenascin-C downregulates wnt inhibitor dickkopf-1, promoting tumorigenesis in a neuroendocrine tumor model.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Cell Rep. 2013 Oct 31;5(2):482-92. doi: 10.1016/j.celrep.2013.09.014. Epub 2013 Oct 17.

●● Enlace al texto completo (gratuito o de pago) 1016/j.celrep.2013.09.014

AUTORES / AUTHORS: - Saupe F; Schwenzer A; Jia Y; Gasser I; Spenle C; Langlois B; Kammerer M; Lefebvre O; Hlushchuk R; Rupp T; Marko M; van der Heyden M; Cremel G; Arnold C; Klein A; Simon-Assmann P; Djonov V; Neuville-Mechine A; Esposito I; Slotta-Huspenina J; Janssen KP; de Wever O; Christofori G; Hussenet T; Orend G

INSTITUCIÓN / INSTITUTION: - Inserm U1109, MN3T Team, The Microenvironmental Niche in Tumorigenesis and Targeted Therapy, 3 Avenue Moliere, 67200 Strasbourg, France; Universite de Strasbourg, 67000 Strasbourg, France; LabEx Medalis, Universite de Strasbourg, 67000 Strasbourg, France; Federation de Medecine Translationnelle de Strasbourg (FMTS), 67000 Strasbourg, France.

RESUMEN / SUMMARY: - The extracellular matrix molecule tenascin-C (TNC) is a major component of the cancer-specific matrix, and high TNC expression is linked to poor prognosis in several cancers. To provide a comprehensive understanding of TNC's functions in cancer, we established an immune-competent transgenic mouse model of pancreatic beta-cell carcinogenesis with varying levels of TNC expression and compared stochastic neuroendocrine tumor formation in abundance or absence of TNC. We show that TNC promotes tumor cell survival, the angiogenic switch, more and leaky vessels, carcinoma progression, and lung micrometastasis. TNC downregulates Dickkopf-1 (DKK1) promoter activity through the blocking of actin stress fiber formation, activates Wnt signaling, and induces Wnt target genes in tumor and endothelial cells. Our results implicate DKK1 downregulation as an important mechanism underlying TNC-enhanced tumor progression through the provision of a proangiogenic tumor microenvironment.

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[225]

TÍTULO / TITLE: - Small cell neuroendocrine carcinoma of the paranasal sinus.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Natl J Maxillofac Surg. 2013 Jan;4(1):111-3. doi: 10.4103/0975-5950.117818.

●● Enlace al texto completo (gratuito o de pago) 4103/0975-5950.117818

AUTORES / AUTHORS: - Krishnamurthy A; Ravi P; Vijayalakshmi R; Majhi U

INSTITUCIÓN / INSTITUTION: - Department of Surgical Oncology, Cancer Institute (WIA), Adyar, Chennai, Tamil Nadu, India.

RESUMEN / SUMMARY: - Small cell neuroendocrine carcinoma (SNEC) is an uncommon tumor. This tumor usually occurs in the lungs, the extra-pulmonary form accounts for only about 4% of all cases. Primary SNEC of the paranasal sinuses is extremely rare; only about 76 cases have been reported in literature. Unfortunately due to the rarity of this neoplasm, there are no specific recommendations pertaining to the management, treatment options are generally extrapolated from similar tumors of pulmonary origin. While Surgery was used in the past, upfront chemoradiation now seems to be evolving as the treatment of choice. We report a case of sinonasal SNEC who had undergone definitive concurrent chemoradiation and is currently disease-free for close to 2 years. The clinical presentation, imaging studies, histopathological diagnosis with immunohistochemistry correlation, management protocols, and a brief review of literature of this rare tumor is discussed.

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[226]

TÍTULO / TITLE: - Simultaneous pheochromocytoma and third-degree atrioventricular block in 2 dogs.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - J Vet Emerg Crit Care (San Antonio). 2013 Sep 19. doi: 10.1111/vec.12101.

●● Enlace al texto completo (gratuito o de pago) 1111/vec.12101

AUTORES / AUTHORS: - Mak G; Allen J

INSTITUCIÓN / INSTITUTION: - VCA California Animal Hospital Veterinary Specialty Group, Los Angeles, CA, 90025.

RESUMEN / SUMMARY: - OBJECTIVE: To describe the occurrence of pheochromocytoma with third-degree atrioventricular (AV) block in 2 dogs. CASE SERIES SUMMARY: Two dogs were referred for echocardiograms and further diagnostic and therapeutic treatment for third-degree AV block. Abdominal ultrasound of 1 dog revealed an adrenal mass, while that of the other dog revealed bilateral adrenal masses. While undergoing treatment, 1 dog experienced cardiac arrest and could not be revived with cardiopulmonary resuscitation, and the other dog was humanely euthanized. Histological findings of the masses were consistent with pheochromocytomas. NEW OR UNIQUE INFORMATION PROVIDED: To the authors' knowledge, this is the first report with histologic evidence of pheochromocytoma and clinical presentation of third-degree AV block in dogs. In human literature, simultaneous presentation of both disease states is rare and has been infrequently reported.

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[227]

TÍTULO / TITLE: - Multifocal thoracic chordoma mimicking a paraganglioma.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - J Cancer Res Ther. 2013 Jul-Sep;9(3):497-9. doi: 10.4103/0973-1482.119312.

●● Enlace al texto completo (gratuito o de pago) 4103/0973-1482.119312

AUTORES / AUTHORS: - Conzo G; Gambardella C; Pasquali D; Ciancia G; Avenia N; Pietra CD; Napolitano S; Palazzo A; Mauriello C; Parmeggiani D; Pettinato G; Napolitano V; Santini L

INSTITUCIÓN / INSTITUTION: - Department of Anaesthesiologic, Surgical and Emergency Science, VII Division of General and Endocrine Surgery, Second University of Naples, Naples, Italy.

RESUMEN / SUMMARY: - Chordoma of thoracic vertebras is a very rare locally invasive neoplasm with low grade malignancy arising from embryonic notochordal remnants. Radical surgery remains the cornerstone of the treatment. We describe a case of multifocal T1-T2 chordoma, without bone and disc involvement, incidentally misdiagnosed as a paraganglioma, occurring in a 47-year-old male asymptomatic patient. Neoplasm was radically removed by an endocrine surgeon through a right extended cervicotomy. A preoperative reliable diagnosis of chordoma, as in the reported case, is often difficult. Radical surgery can provide a favorable outcome but, given the high rates of local recurrence of this neoplasm, a strict and careful follow-up is recommended. Although very rare, chordoma should be suggested in the differential diagnosis of the paravertebral cervical masses of unknown origin. Spine surgeon

consultation and a FNB should be routinely included in the multidisciplinary preoperative work-up of these neoplasms.

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[228]

TÍTULO / TITLE: - Rare testicular tumor discovered by assault: an unusual presentation of a primary testicular neuroendocrine tumor grade 2.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Case Rep Pathol. 2013;2013:709352. doi: 10.1155/2013/709352. Epub 2013 Sep 19.

●● Enlace al texto completo (gratuito o de pago) 1155/2013/709352

AUTORES / AUTHORS: - Epperson JR; Pope NM; Abuzeid MJ

INSTITUCIÓN / INSTITUTION: - Department of Pathology and Laboratory Services, San Antonio Military Medical Center, 3551 Roger Brooke Dr, Fort Sam Houston, San Antonio, TX 78234, USA.

RESUMEN / SUMMARY: - Testicular neuroendocrine tumors (NET) or carcinoid tumors are rare neoplasms which represent 1% of all testicular tumors and can be divided into 3 subgroups: pure primary testicular NET, primary testicular NET associated with a teratoma, and NET metastases to the testis. We report an unusual presentation of a primary testicular neuroendocrine tumor in a 39-year-old male who presented after a physical altercation during a soccer game. Histology showed a diffuse infiltrating tumor with extensive involvement of the tunica albuginea and tunica vaginalis. Immunohistochemical expression of CD56, synaptophysin, and chromogranin A was strongly positive in the tumor cells. Foci of tumor cell necrosis and occasional mitotic figures as well as extensive lymph-vascular invasion were also identified. A review of the literature reveals differing opinions on the prognostic significance of primary tumor size, mitotic index, tumor necrosis, and nuclear atypia. In our patient, the increased mitotic rate (3-5 mitotic figures per 10 hpf and a Ki-67 index of 5%), foci of necrosis, and mild to moderate nuclear atypia warranted a diagnosis of neuroendocrine tumor grade 2, formerly atypical carcinoid. Long term surveillance in these patients is essential as metastasis occurs in up to 15% of cases. At the 6-month followup, the patient remains symptom free.

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[229]

TÍTULO / TITLE: - Ischemic stroke as a presenting feature of VIPoma due to MEN 1 syndrome.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Indian J Endocrinol Metab. 2013 Oct;17(Suppl 1):S215-8. doi: 10.4103/2230-8210.119576.

●● Enlace al texto completo (gratuito o de pago) 4103/2230-8210.119576

AUTORES / AUTHORS: - Maheshwari RR; Desai M; Rao VP; Palanki RR; Namburi RP; Reddy KT; Reddy AP

INSTITUCIÓN / INSTITUTION: - Department of Endocrinology and Metabolism, Narayana Medical College and Hospital, Nellore, Andhra Pradesh, India.

RESUMEN / SUMMARY: - INTRODUCTION: Presentation of the ischemic stroke due to vasoactive intestinal peptide producing tumor (VIPoma) or Verner Morrison syndrome is rare. This is first of its kind case which we are reporting here which was later turned out to be multiple endocrine neoplasia type 1 (MEN 1) syndrome with diagnosis of primary hyperparathyroidism in the same patient in follow-up. DESCRIPTION OF THE

CASE: A 13-year-old girl presented to our emergency department with features of disorientation, weakness of left sided extremities. She had watery high volume diarrhea and related dehydration with renal failure. Blood chemistry was suggestive of hypokalemia with metabolic acidosis. Patient had flushing on her face during this episode of illness. Magnetic resonance imaging (MRI) of brain suggested venous infarct. Computed tomography (CT) scan of abdomen done with high index of suspicion was suggestive of mass in tail of pancreas mostly a VIPoma. Patient was operated for the tumor after which there was no recurrence of diarrhea. Biopsy of tumor was consistent with VIPoma with chomogranin A positivity. Patient improved of her stroke episode with time. On follow-up she is diagnosed to have primary hyperparathyroidism with hypercalcemia due to left inferior parathyroid adenoma which improved with intravenous (IV) zolindronic acid therapy and now she is planned to undergo parathyroidectomy. CONCLUSION: VIPoma is a rare tumor but is well-described with MEN 1. Stroke as a presenting feature of VIPoma is first reported with this case.

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[230]

TÍTULO / TITLE: - Bronchial carcinoid in college freshman with persistent focal wheeze.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - J Am Assoc Nurse Pract. 2013 Dec;25(12):641-3. doi: 10.1002/2327-6924.12059. Epub 2013 Aug 22.

●● Enlace al texto completo (gratuito o de pago) 1002/2327-6924.12059

AUTORES / AUTHORS: - Holzer R; Rosen D

INSTITUCIÓN / INSTITUTION: - Division of Respiratory Diseases, Children's Hospital Boston, Boston, Massachusetts.

RESUMEN / SUMMARY: - PURPOSE: To bring attention to a rare diagnosis in the pediatric population that is in the differential diagnosis for not well-controlled asthma. DATA SOURCES: Case presentation. CONCLUSIONS: Pulmonary carcinoid tumors are rare and usually present late in adolescence. Most of these tumors are located in the proximal airways and symptoms may be similar to those of asthma including cough, wheeze, chest pain, or recurrent pneumonia. IMPLICATIONS FOR PRACTICE: Bronchial carcinoid should be in the differential diagnosis for adolescents with difficult to control asthma, who have symptoms including chronic cough and focal wheeze. Referral to a pulmonary specialist should be considered to help work up the differential diagnoses.

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[231]

TÍTULO / TITLE: - Merkel cell polyomavirus and extrapulmonary small cell carcinoma.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Oncol Lett. 2013 Oct;6(4):1049-1052. Epub 2013 Jul 23.

●● Enlace al texto completo (gratuito o de pago) 3892/ol.2013.1483

AUTORES / AUTHORS: - Hourdequin KC; Lefferts JA; Brennick JB; Ernstoff MS; Tsongalis GJ; Pipas JM

INSTITUCIÓN / INSTITUTION: - Department of Medicine, Section of Hematology/Oncology, Geisel School of Medicine at Dartmouth, Hanover, NH, USA ; Dartmouth Hitchcock Medical Center and Norris Cotton Cancer Center, Lebanon, NH, USA.

RESUMEN / SUMMARY: - The Merkel cell polyomavirus (MCV) is involved in the development of up to 100% of Merkel cell cancer (MCC) cases. Early studies have

reported that the virus was infrequently detected in other small cell or neuroendocrine lung carcinomas, which share histological features with MCC. The present study investigated the presence of MCV in cases of extrapulmonary small cell carcinoma (ESCC), which also shares histological features with MCC. A total of 25 cases of ESCC that were diagnosed between 2004 and 2009 were identified at The Dartmouth Hitchcock Medical Center. Archived tissue was available for testing in 16 of these cases. A total of 11 tissue specimens of MCC were used as positive controls. DNA that was extracted from the archived tissue was subjected to five separate quantitative (q)PCR assays for the detection of four MCV genomic targets. MCV DNA was detected in 3/16 (19%) of the ESCCs and in all 11 MCCs. In the three MCV-positive ESCCs, the viral target was only detected by either one or two of the PCR assays. In 8/11 MCV-positive MCCs, the DNA tested positive by either three or all four assays and the remaining three MCCs tested positive by either one or two assays. The beta-globin endogenous control was detected in all the samples that were tested. Although MCC and ESCC share numerous histological features, MCV is detected at a lower frequency in ESCC. The possible role for MCV in the etiology of ESCC remains uncertain and may account for the rare cases of ESCC with no other identifiable etiology. The failure of other assays to detect MCV may be due to sequence variability in the MCV genome.

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[232]

TÍTULO / TITLE: - Malignant catatonia mimicking pheochromocytoma.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Case Rep Endocrinol. 2013;2013:815821. doi: 10.1155/2013/815821. Epub 2013 Oct 22.

●● Enlace al texto completo (gratuito o de pago) 1155/2013/815821

AUTORES / AUTHORS: - Wong S; Hughes B; Pudek M; Li D

INSTITUCIÓN / INSTITUTION: - Department of Pathology and Laboratory Medicine, University of British Columbia, 855 West 12th Avenue, Vancouver, BC, Canada V5Z 1M9.

RESUMEN / SUMMARY: - Malignant catatonia is an unusual and highly fatal neuropsychiatric condition which can present with clinical and biochemical manifestations similar to those of pheochromocytoma. Differentiating between the two diseases is essential as management options greatly diverge. We describe a case of malignant catatonia in a 20-year-old male who presented with concurrent psychotic symptoms and autonomic instability, with markedly increased 24-hour urinary levels of norepinephrine at 1752 nmol/day (normal, 89-470 nmol/day), epinephrine at 1045 nmol/day (normal, <160 nmol/day), and dopamine at 7.9 mu mol/day (normal, 0.4-3.3 mu mol/day). The patient was treated with multiple sessions of electroconvulsive therapy, which led to complete clinical resolution. Repeat urine collections within weeks of this presenting event revealed normalization or near normalization of his catecholamine and metanephrine levels. Malignant catatonia should be considered in the differential diagnosis of the hypercatecholamine state, particularly in a patient who also exhibits concurrent catatonic features.

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[233]

TÍTULO / TITLE: - Neuroendocrine tumor of the small intestine.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Clin Res Hepatol Gastroenterol. 2013 Oct 28. pii: S2210-7401(13)00188-5. doi: 10.1016/j.clinre.2013.09.001.

●● Enlace al texto completo (gratuito o de pago) 1016/j.clinre.2013.09.001

AUTORES / AUTHORS: - Arbache A; Kara M; Mouhadi SE; Arrive L

INSTITUCIÓN / INSTITUTION: - Department of Radiology, Saint-Antoine Hospital, AP-HP, Universite Pierre-et-Marie-Curie, Paris-VI, 184, rue du Faubourg-Saint-Antoine, 75012 Paris, France.

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[234]

TÍTULO / TITLE: - Carcinoid tumour presenting as recurrent pneumonia.

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 Nov 20;2013. pii: bcr2013202203. doi: 10.1136/bcr-2013-202203.

●● Enlace al texto completo (gratuito o de pago) 1136/bcr-2013-202203

AUTORES / AUTHORS: - Dharmagunawardena R; Lipman M; Cleverley J; Cash C

INSTITUCIÓN / INSTITUTION: - Department of Thoracic Medicine, Royal Free Hospital, London, UK.

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[235]

TÍTULO / TITLE: - Paraganglioma as a rare cause of left ventricular thrombus in the setting of preserved ejection fraction: discussing the literature.

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 Nov 19;2013. pii: bcr2013202001. doi: 10.1136/bcr-2013-202001.

●● Enlace al texto completo (gratuito o de pago) 1136/bcr-2013-202001

AUTORES / AUTHORS: - Shafiq A; Nguyen P; Hudson MP; Rabbani B

INSTITUCIÓN / INSTITUTION: - Henry Ford Hospital, Detroit, Michigan, USA.

RESUMEN / SUMMARY: - Paragangliomas and pheochromocytomas are catecholamine-secreting tumours which if remain undiagnosed may cause severe morbidity and mortality. In rare circumstances these tumours can cause left ventricular (LV) thrombi to form by inducing cardiomyopathy and subsequent embolic complications. After a thorough literature review, six previous cases were found that presented the formation of an LV thrombus in the setting of a pheochromocytoma or paraganglioma. A majority of these cases were associated with significant wall motion abnormalities and their cardiac ejection fraction (EF) was compromised. This is a rare case of a patient developing LV thrombi in the setting of a paraganglioma with normal cardiac EF. We present this case to compare the similarities and differences of our case with previously reported cases and emphasise the importance of suspecting these LV thrombi in patients with these neuroendocrine tumours.

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[236]

TÍTULO / TITLE: - Chemometric evaluation of urinary steroid hormone levels as potential biomarkers of neuroendocrine tumors.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Molecules. 2013 Oct 16;18(10):12857-76. doi: 10.3390/molecules181012857.

●● Enlace al texto completo (gratuito o de pago) 3390/molecules181012857

AUTORES / AUTHORS: - Plenis A; Miekus N; Oledzka I; Baczek T; Lewczuk A; Wozniak Z; Koszalka P; Seroczynska B; Skokowski J

INSTITUCIÓN / INSTITUTION: - Department of Pharmaceutical Chemistry, Medical University of Gdansk, Hallera 107, Gdansk 80-416, Poland. aplenis@gumed.edu.pl.

RESUMEN / SUMMARY: - Neuroendocrine tumors (NETs) are uncommon tumors which can secrete specific hormone products such as peptides, biogenic amines and hormones. So far, the diagnosis of NETs has been difficult because most NET markers are not specific for a given tumor and none of the NET markers can be used to fulfil the criteria of high specificity and high sensitivity for the screening procedure. However, by combining the measurements of different NET markers, they become highly sensitive and specific diagnostic tests. The aim of the work was to identify whether urinary steroid hormones can be identified as potential new biomarkers of NETs, which could be used as prognostic and clinical course monitoring factors. Thus, a rapid and sensitive reversed-phase high-performance liquid chromatographic method (RP-HPLC) with UV detection has been developed for the determination of cortisol, cortisone, corticosterone, testosterone, epitestosterone and progesterone in human urine. The method has been validated for accuracy, precision, selectivity, linearity, recovery and stability. The limits of detection and quantification were 0.5 and 1 ng mL-1 for each steroid hormone, respectively. Linearity was confirmed within a range of 1-300 ng mL-1 with a correlation coefficient greater than 0.9995 for all analytes. The described method was successfully applied for the quantification of six endogenous steroid levels in human urine. Studies were performed on 20 healthy volunteers and 19 patients with NETs. Next, for better understanding of tumor biology in NETs and for checking whether steroid hormones can be used as potential biomarkers of NETs, a chemometric analysis of urinary steroid hormone levels in both data sets was performed.

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[237]

TÍTULO / TITLE: - Unexpected small urinary bladder pheochromocytoma: a nonspecific presentation.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Case Rep Urol. 2013;2013:496547. doi: 10.1155/2013/496547. Epub 2013 Sep 22.

●● Enlace al texto completo (gratuito o de pago) 1155/2013/496547

AUTORES / AUTHORS: - Mallat F; Hmida W; Slama A; Mosbah F

INSTITUCIÓN / INSTITUTION: - Department of Urology, Hospital of Sahloul, 4054 Sousse, Tunisia.

RESUMEN / SUMMARY: - Objectives. Pheochromocytoma of the urinary bladder is an extremely rare tumor that typically presents with a hypertensive crisis during micturition. Preoperatively, it may be misdiagnosed due to nonspecific symptomatology, physical, and radiologic findings. Method. We report a case of unsuspected small pheochromocytoma which was incidentally found by CT scan and confirmed by the histological aspects after transurethral resection in a 63-year-old woman. Here, we have described the clinical presentation, physical findings, laboratory investigations, and treatment provided in our case. We have also included

radiological images and histopathology slides with input from both radiologists and pathologists. Surgical management and postoperative follow-up are discussed, as are details of previous published data. Results. After undergoing surgical treatment (transurethral resection), our patient is asymptomatic, with complete resolution of her pathology. Conclusion. Diagnosis is difficult before histopathological examination and should be considered in patients with no risk factors for usual bladder tumor. Our purpose is to raise clinician's awareness for this condition so that they will be more likely to diagnose it. This will facilitate prompt diagnosis and treatment and especially prevent complications due to pheochromocytoma which may be severe.

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[238]

TÍTULO / TITLE: - Insulinoma presenting with cardiac arrest and cardiomyopathy.

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 Oct 23;2013. pii: bcr2013009193. doi: 10.1136/bcr-2013-009193.

●● Enlace al texto completo (gratuito o de pago) 1136/bcr-2013-009193

AUTORES / AUTHORS: - Thirumalai A; Levander XA; Mookherjee S; White AA

INSTITUCIÓN / INSTITUTION: - Internal Medicine Residency Program, University of Washington, Seattle, Washington, USA.

RESUMEN / SUMMARY: - A 33-year-old woman presented with ventricular fibrillation cardiac arrest and was found to have a blood glucose of 1.83 mmol/L. Cardiac catheterisation revealed a dilated left ventricle with an ejection fraction (EF) of 26% and angiographically normal coronary arteries. Continuous dextrose infusion was required to treat hypoglycaemia, which prompted consideration of insulinoma as a possible cause for her cardiomyopathy. Whipple's triad was demonstrated; a 72 h fast provided biochemical evidence of insulinoma, and imaging localised a tumour in her pancreas. The tumour was resected and pathology confirmed insulinoma; pancreaticoduodenectomy cured her hypoglycaemia. No alternate cause of cardiomyopathy was found and 4 months after surgery her EF improved to 41%. High insulin levels can close cardiac KATP channels associated with dilated cardiomyopathy; the catecholamine surge from hypoglycaemia may also contribute to ventricular remodelling. Hypoglycaemia can cause QT segment prolongation, and may have precipitated fibrillation in this patient's arrhythmia-prone myocardium.

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[239]

TÍTULO / TITLE: - Undiagnosed bladder pheochromocytoma: An anesthetic challenge.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - J Anaesthesiol Clin Pharmacol. 2013 Oct;29(4):574-6. doi: 10.4103/0970-9185.119165.

●● Enlace al texto completo (gratuito o de pago) 4103/0970-9185.119165

AUTORES / AUTHORS: - Khatavkar SS; Raje DM; Doshi CM; Mohite SN; Dhande AV

INSTITUCIÓN / INSTITUTION: - Department of Anaesthesia, Padmashree Dr. D. Y. Patil Medical College and Hospital, Navi Mumbai, Maharashtra, India.

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[240]

TÍTULO / TITLE: - A Drug Repositioning Approach Identifies Tricyclic Antidepressants as Inhibitors of Small Cell Lung Cancer and Other Neuroendocrine Tumors.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Cancer Discov. 2013 Sep 26.
●● Enlace al texto completo (gratuito o de pago) 1158/2159-8290.CD-13-0183
AUTORES / AUTHORS: - Jahchan NS; Dudley JT; Mazur PK; Flores N; Yang D; Palmerton A; Zmoos AF; Vaka D; Tran KQ; Zhou M; Krasinska K; Riess JW; Neal JW; Khatri P; Park KS; Butte AJ; Sage J
INSTITUCIÓN / INSTITUTION: - Departments of 1Pediatrics, 2Genetics, and 5Medicine-Oncology, 3Vincent Coates Mass Spectrometry Laboratory, Stanford University, Stanford; and 4Division of Hematology/Oncology, Department of Internal Medicine, University of California Davis Cancer Center, University of California Davis School of Medicine, Sacramento, California.
RESUMEN / SUMMARY: - Small cell lung cancer (SCLC) is an aggressive neuroendocrine subtype of lung cancer with high mortality. We used a systematic drug repositioning bioinformatics approach querying a large compendium of gene expression profiles to identify candidate U.S. Food and Drug Administration (FDA)-approved drugs to treat SCLC. We found that tricyclic antidepressants and related molecules potently induce apoptosis in both chemonaive and chemoresistant SCLC cells in culture, in mouse and human SCLC tumors transplanted into immunocompromised mice, and in endogenous tumors from a mouse model for human SCLC. The candidate drugs activate stress pathways and induce cell death in SCLC cells, at least in part by disrupting autocrine survival signals involving neurotransmitters and their G protein-coupled receptors. The candidate drugs inhibit the growth of other neuroendocrine tumors, including pancreatic neuroendocrine tumors and Merkel cell carcinoma. These experiments identify novel targeted strategies that can be rapidly evaluated in patients with neuroendocrine tumors through the repurposing of approved drugs.

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[241]
TÍTULO / TITLE: - Can DCE-MRI Explain the Heterogeneity in Radiopeptide Uptake Imaged by SPECT in a Pancreatic Neuroendocrine Tumor Model?
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - PLoS One. 2013 Oct 8;8(10):e77076. doi: 10.1371/journal.pone.0077076.
●● Enlace al texto completo (gratuito o de pago) 1371/journal.pone.0077076
AUTORES / AUTHORS: - Bol K; Haeck JC; Groen HC; Niessen WJ; Bernsen MR; de Jong M; Veenland JF
INSTITUCIÓN / INSTITUTION: - Biomedical Imaging Group Rotterdam, Departments of Radiology and Medical Informatics, Erasmus Medical Center, Rotterdam, The Netherlands ; Department of Radiology, Erasmus Medical Center, Rotterdam, The Netherlands ; Department of Nuclear Medicine, Erasmus Medical Center, Rotterdam, The Netherlands.
RESUMEN / SUMMARY: - Although efficient delivery and distribution of treatment agents over the whole tumor is essential for successful tumor treatment, the distribution of most of these agents cannot be visualized. However, with single-photon emission computed tomography (SPECT), both delivery and uptake of radiolabeled peptides can be visualized in a neuroendocrine tumor model overexpressing somatostatin

receptors. A heterogeneous peptide uptake is often observed in these tumors. We hypothesized that peptide distribution in the tumor is spatially related to tumor perfusion, vessel density and permeability, as imaged and quantified by DCE-MRI in a neuroendocrine tumor model. Four subcutaneous CA20948 tumor-bearing Lewis rats were injected with the somatostatin-analog (111)In-DTPA-Octreotide (50 MBq). SPECT-CT and MRI scans were acquired and MRI was spatially registered to SPECT-CT. DCE-MRI was analyzed using semi-quantitative and quantitative methods. Correlation between SPECT and DCE-MRI was investigated with 1) Spearman's rank correlation coefficient; 2) SPECT uptake values grouped into deciles with corresponding median DCE-MRI parametric values and vice versa; and 3) linear regression analysis for median parameter values in combined datasets. In all tumors, areas with low peptide uptake correlated with low perfusion/density/ /permeability for all DCE-MRI-derived parameters. Combining all datasets, highest linear regression was found between peptide uptake and semi-quantitative parameters ($R(2)>0.7$). The average correlation coefficient between SPECT and DCE-MRI-derived parameters ranged from 0.52-0.56 ($p<0.05$) for parameters primarily associated with exchange between blood and extracellular extravascular space. For these parameters a linear relation with peptide uptake was observed. In conclusion, the 'exchange-related' DCE-MRI-derived parameters seemed to predict peptide uptake better than the 'contrast amount- related' parameters. Consequently, fast and efficient diffusion through the vessel wall into tissue is an important factor for peptide delivery. DCE-MRI helps to elucidate the relation between vascular characteristics, peptide delivery and treatment efficacy, and may form a basis to predict targeting efficiency.

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[242]

TÍTULO / TITLE: - Collision renal cell papillary and medullary carcinoma in a 66-year-old man.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Oncology (Williston Park). 2013 Sep;27(9):893, 896, 898.

AUTORES / AUTHORS: - Lam ET; Kessler ER; Flaig TW; La Rosa FG

INSTITUCIÓN / INSTITUTION: - Division of Medical Oncology, School of Medicine, University of Colorado Denver, Anschutz Medical Campus, Aurora, Colorado, USA.

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[243]

TÍTULO / TITLE: - Gastric somatostatinoma: an extremely rare cause of upper gastrointestinal bleeding.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Clin Endosc. 2013 Sep;46(5):582-5. doi: 10.5946/ce.2013.46.5.582. Epub 2013 Sep 30.

●● Enlace al texto completo (gratuito o de pago) 5946/ce.2013.46.5.582

AUTORES / AUTHORS: - Prachayakul V; Aswakul P; Deesomsak M; Pongpaibul A

INSTITUCIÓN / INSTITUTION: - Division of Gastroenterology, Department of Internal Medicine, Siriraj Hospital, Mahidol University Faculty of Medicine, Bangkok, Thailand.

RESUMEN / SUMMARY: - A 49-year-old woman presented with chronic abdominal discomfort, significant weight loss, and chronic intermittent diarrhea. She suddenly developed massive upper gastrointestinal bleeding and was referred for further treatment. Endoscopy indicated a large mass in the upper gastric body with antral and duodenal bulb involvement. Endosonography showed a large well-defined isoechoic

gastric subepithelial mass with multiple intra-abdominal and peripancreatic lymphadenopathy, suspected to be malignant on the basis of fine needle aspiration cytology. The tumor was surgically removed, and histopathology showed typical characteristics of a neuroendocrine tumor. On the basis of immunohistochemical staining, somatostatinoma, a rare neuroendocrine tumor, was diagnosed. Gastrointestinal bleeding is a rare presentation and the stomach is an uncommon tumor location.

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[244]

TÍTULO / TITLE: - Carotid and vagal body paragangliomas.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Transl Med UniSa. 2013 May 6;6:11-5.

AUTORES / AUTHORS: - Del Guercio L; Narese D; Ferrara D; Butrico L; Padricelli A; Porcellini M

INSTITUCIÓN / INSTITUTION: - Vascular Surgery, Federico II University of Naples, Italy.

RESUMEN / SUMMARY: - Between 1972 and 2012, 25 patients presenting 32 paragangliomas of the neck were observed. Tumor locations included the carotid body (CBTs) in 21 patients and the vagus nerve in 4. Four patients had bilateral CBT and one a bilateral vagal tumor; a metachronous bilateral jugulare paraganglioma was diagnosed in one patient with bilateral CBT Shamblin type III. Five patients presented CBTs type II and three type III. Preoperative embolization was performed in 5 CBTs, with no significant difference in blood loss. Twenty-nine paragangliomas were resected (with three internal carotid artery resection): there were no cerebrovascular accident or perioperative death. Nine patients (36%) had cranial nerve palsy prior to surgery and a postoperative nerve dysfunction occurred in four other tumors (16%). Persistent nerve deficits occurred in 3 patients (12%). No evidence of malignancy was shown, intraoperatively or during a postoperative follow-up period (9 months to 18 years; mean: 8 years).

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[245]

TÍTULO / TITLE: - The role of the immune response in merkel cell carcinoma.

RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

REVISTA / JOURNAL: - Cancers (Basel). 2013 Feb 28;5(1):234-54. doi: 10.3390/cancers5010234.

●● Enlace al texto completo (gratuito o de pago) 3390/cancers5010234

AUTORES / AUTHORS: - Triozzi PL; Fernandez AP

INSTITUCIÓN / INSTITUTION: - Taussig Cancer Institute, Cleveland Clinic Foundation, 9500 Euclid Avenue, Cleveland, OH 44195, USA. triozzp@ccf.org.

RESUMEN / SUMMARY: - Merkel cell carcinoma (MCC) is an aggressive neuroendocrine skin cancer. The Merkel cell polyomavirus (MCPyV) is implicated in its pathogenesis. Immune mechanisms are also implicated. Patients who are immunosuppressed have an increased risk. There is evidence that high intratumoral T-cell counts and immune transcripts are associated with favorable survival. Spontaneous regressions implicate immune effector mechanisms. Immunogenicity is also supported by observation of autoimmune paraneoplastic syndromes. Case reports suggest that immune modulation, including reduction of immune suppression, can result in tumor regression. The relationships between MCPyV infection, the immune response, and clinical outcome, however, remain poorly understood. Circulating

antibodies against MCPyV antigens are present in most individuals. MCPyV-reactive T cells have been detected in both MCC patients and control subjects. High intratumoral T-cell counts are also associated with favorable survival in MCPyV-negative MCC. That the immune system plays a central role in preventing and controlling MCC is supported by several observations. MCCs often develop, however, despite the presence of humoral and cellular immune responses. A better understanding on how MCPyV and MCC evade the immune response will be necessary to develop effective immunotherapies.
