

#15#

Revisiones (todas) \*\*\* Reviews (all)

Neuroendocrine tumors.

Julio - Agosto 2013 / July - August 2013

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[1]

**TÍTULO / TITLE:** - Extrapulmonary neuroendocrine small and large cell carcinomas: a review of controversial diagnostic and therapeutic issues.

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

**REVISTA / JOURNAL:** - Hum Pathol. 2013 Jun 24. pii: S0046-8177(13)00146-9. doi: 10.1016/j.humpath.2013.03.016.

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

[1016/j.humpath.2013.03.016](#)

**AUTORES / AUTHORS:** - Volante M; Birocco N; Gatti G; Duregon E; Lorizzo K; Fazio N; Scagliotti GV; Papotti M

**INSTITUCIÓN / INSTITUTION:** - Department of Oncology, University of Turin at San Luigi Hospital, 10043 Torino, Italy. Electronic address: [marco.volante@unito.it](mailto:marco.volante@unito.it).

**RESUMEN / SUMMARY:** - Extrapulmonary neuroendocrine carcinoma (EPNEC) is a heterogeneous and rare group of high-grade neoplasms occurring in different organs. They usually share a poor prognosis, but diagnostic and therapeutic options still include several controversial issues, due to the rarity of this condition and to differences in architecture and cell size, being some cases pure small cell carcinomas, other pure large cell neuroendocrine carcinomas and some others combined/mixed neuroendocrine carcinomas with a conventional non-neuroendocrine carcinoma. In addition, the therapeutic strategy varies in different organs (surgery and/or chemotherapy and/or radiation therapy and/or targeted treatments), and clinicians and pathologists are asked to interact to reach an accurate classification of every single case, as

well as the most appropriate selection of the treatment options, even considering different time points of each EPNEC natural history. This overview highlights controversial pathological and clinical issues and summarizes possible solutions to most of such EPNEC-related problems.

[2]

**TÍTULO / TITLE:** - Evidence-based management of primary and localized Merkel cell carcinoma: a review.

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

**REVISTA / JOURNAL:** - Int J Dermatol. 2013 Jul 8. doi: 10.1111/ijd.12091.

●● Enlace al texto completo (gratis o de pago) [1111/ijd.12091](#)

**AUTORES / AUTHORS:** - Ellis DL; Davis RS

**INSTITUCIÓN / INSTITUTION:** - Department of Dermatology, Tulane University School of Medicine, New Orleans, LA, USA.

**RESUMEN / SUMMARY:** - BACKGROUND: Merkel cell carcinoma is a rare and often lethal cutaneous neuroendocrine malignancy with a tendency for early and frequent locoregional and distant metastasis and relapses. It is a tumor of the elderly and immunosuppressed, which most often appears on sun-exposed areas of the body. There is growing interest in characterization of the disease and the best approach to its management. Despite the lack of prospective randomized clinical trials, treatment is evolving. OBJECTIVE: To provide an updated review of the most current and relevant data concerning the surgical (+/- radiological) management of Merkel cell carcinoma, including the role of Mohs micrographic surgery. METHODS: Using relevant MeSH terms, we performed a review of the literature on the above subjects from 1981 to 2011. RESULTS AND CONCLUSION: For primary tumors without evidence of organ metastases, surgical excision should be the primary therapy. Owing to the high rate of local metastases, a safety margin of at least 2 cm should be considered. In situations where small, localized tumors and/or special locations necessitate a smaller safety margin, compensation by complete histological examination of the excision margins and perhaps adjuvant radiation therapy should be undertaken. The literature states that benefits of Mohs micrographic surgery (over wide local excision) include tissue conservation and identification of tumors that may require extremely wide excision margins. The majority of data to date supports the use of Mohs surgery in the treatment of Merkel cell carcinoma.

[3]

**TÍTULO / TITLE:** - Composite pheochromocytoma-ganglioneuroma of the adrenal gland: A case report with immunohistochemical study.

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

**REVISTA / JOURNAL:** - Urol Ann. 2013 Apr;5(2):115-8. doi: 10.4103/0974-7796.110011.

●● Enlace al texto completo (gratis o de pago) [4103/0974-7796.110011](https://doi.org/10.4103/0974-7796.110011)

**AUTORES / AUTHORS:** - Rao RN; Singla N; Yadav K

**INSTITUCIÓN / INSTITUTION:** - Department of Pathology, Sanjay Gandhi Postgraduate Institute of Medical Sciences, Lucknow, India.

**RESUMEN / SUMMARY:** - Composite tumors of the adrenal medulla consisting of pheochromocytoma and ganglioneuroma are rare tumors accounting for less than 3% of all sympathoadrenal tumors. These tumors display more than one line of differentiation in which normal and neoplastic chromaffin cells are capable of differentiating into ganglion cells under the influence of nerve growth factors. To the best of our knowledge, we report the second case with a composite tumor of the adrenal medulla in a normotensive patient from India.

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**TÍTULO / TITLE:** - Identification of Succinate Dehydrogenase-deficient Bladder Paragangliomas.

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

**REVISTA / JOURNAL:** - Am J Surg Pathol. 2013 Jun 20.

●● Enlace al texto completo (gratis o de pago) [1097/PAS.0b013e318293d83c](https://doi.org/10.1097/PAS.0b013e318293d83c)

**AUTORES / AUTHORS:** - Mason EF; Sadow PM; Wagner AJ; Remillard SP; Flood TA; Belanger EC; Hornick JL; Barletta JA

**INSTITUCIÓN / INSTITUTION:** - \*Department of Pathology, Brigham and Women's Hospital, Harvard Medical School daggerDepartment of Pathology, Massachusetts General Hospital, Harvard Medical School double daggerDana-Farber Cancer Institute, Center for Sarcoma and Bone Oncology, Harvard Medical School, Boston, MA section signDepartment of Pathology, The Ottawa Hospital, Ottawa, ON, Canada.

**RESUMEN / SUMMARY:** - A significant number of patients with paragangliomas harbor germline mutations in one of the succinate dehydrogenase (SDH) genes (SDHA, B, C, or D). Tumors with mutations in SDH genes can be identified using immunohistochemistry. Loss of SDHB staining is seen in tumors with a mutation in any one of the SDH genes, whereas loss of both SDHB and SDHA expression is seen only in the context of an SDHA mutation. Identifying an SDH-deficient tumor can be prognostically significant, as tumors with SDHB mutations are more likely to pursue a malignant course. Although the rate of SDH deficiency in paragangliomas in general is known to be approximately 30%, there are only rare reports of SDH-deficient bladder paragangliomas. Therefore, the aim of this study was to determine the rate of SDH deficiency in bladder paragangliomas. Eleven cases of bladder paragangliomas were identified. Hematoxylin and eosin-stained slides of all tumors were reviewed, and immunohistochemical analysis for SDHB and SDHA was performed. For cases with loss of SDHA expression by immunohistochemistry, mutation

analysis of the SDHA gene was performed. Loss of SDHB staining was seen in 3 (27%) cases (2 with loss of SDHB only, 1 with loss of SDHB and SDHA). Patients with SDH-deficient tumors were younger than those with tumors with intact SDH expression (mean age at presentation 39 y and 58 y, respectively). Of the 2 patients with SDHB-deficient and SDHA-intact tumors, one was found to have a germline SDHB mutation, and the other had a family history of a malignant paraganglioma. Both patients developed metastatic disease. The one patient with a tumor that was deficient for both SDHB and SDHA had no family history of paragangliomas and no evidence of metastatic disease. Sequencing of this tumor revealed a deleterious heterozygous single-base pair substitution in exon 10 of SDHA (c.1340 A>G; p.His447Arg) in both the tumor and normal tissue, indicative of a germline SDHA mutation, and a deleterious single-base pair substitution in exon 5 of SDHA (c.484 A>T; p.Arg162\*) in 1 allele of the tumor only. No patients with intact SDH expression had a family history of paragangliomas; 1 had a synchronous paraganglioma, but none developed metastatic disease. A significant subset of bladder paragangliomas is SDH deficient. It is essential to identify SDH-deficient tumors, as the presence of an SDH mutation has prognostic implications and is important in guiding genetic counseling.

[4]

**TÍTULO / TITLE:** - Diagnostic accuracy of endoscopic ultrasound in pancreatic neuroendocrine tumors: A systematic review and meta analysis.

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

**REVISTA / JOURNAL:** - World J Gastroenterol. 2013 Jun 21;19(23):3678-84. doi: 10.3748/wjg.v19.i23.3678.

●● [Enlace al texto completo \(gratis o de pago\) 3748/wjg.v19.i23.3678](#)

**AUTORES / AUTHORS:** - Puli SR; Kalva N; Bechtold ML; Pamulaparthi SR; Cashman MD; Estes NC; Pearl RH; Volmar FH; Dillon S; Shekleton MF; Forcione D

**INSTITUCIÓN / INSTITUTION:** - Srinivas R Puli, Smitha R Pamulaparthi, Micheal D Cashman, Fritz-Henry Volmar, Sonu Dillon, Michael F Shekleton, Division of Gastroenterology and Hepatology, University of Illinois Peoria Campus, OSF Saint Francis Medical Center, Peoria, IL 61637, United States.

**RESUMEN / SUMMARY:** - AIM: To detect pancreatic neuroendocrine tumors (PNETs) has been varied. This study is undertaken to evaluate the accuracy of endoscopic ultrasound (EUS) in detecting PNETs. METHODS: Only EUS studies confirmed by surgery or appropriate follow-up were selected. Articles were searched in Medline, Ovid journals, Medline nonindexed citations, and Cochrane Central Register of Controlled Trials and Database of Systematic Reviews. Pooling was conducted by both fixed and random effects model). RESULTS: Initial search identified 2610 reference articles, of these 140 relevant articles were selected and reviewed. Data was extracted from 13 studies (n =

456) which met the inclusion criteria. Pooled sensitivity of EUS in detecting a PNETs was 87.2% (95%CI: 82.2-91.2). EUS had a pooled specificity of 98.0% (95%CI: 94.3-99.6). The positive likelihood ratio of EUS was 11.1 (95%CI: 5.34-22.8) and negative likelihood ratio was 0.17 (95%CI: 0.13-0.24). The diagnostic odds ratio, the odds of having anatomic PNETs in positive as compared to negative EUS studies was 94.7 (95%CI: 37.9-236.1). Begg-Mazumdar bias indicator for publication bias gave a Kendall's tau value of 0.31 (P = 0.16), indication no publication bias. The P for chi(2) heterogeneity for all the pooled accuracy estimates was > 0.10. CONCLUSION: EUS has excellent sensitivity and specificity to detect PNETs. EUS should be strongly considered for evaluation of PNETs.

[5]

**TÍTULO / TITLE:** - Laparoscopic antrectomy for retained antrum in type 1 gastric carcinoid: a case report and review of literature.

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

**REVISTA / JOURNAL:** - Pancreas. 2013 Aug;42(6):1040-2. doi: 10.1097/MPA.0b013e31827e2d3b.

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

[1097/MPA.0b013e31827e2d3b](#)

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[6]

**TÍTULO / TITLE:** - Primary neuroendocrine carcinoma of the appendix: a case report and review of the literature.

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

**REVISTA / JOURNAL:** - Anticancer Res. 2013 Jun;33(6):2635-8.

**AUTORES / AUTHORS:** - Tomioka K; Fukoe Y; Lee Y; Lee M; Wada Y; Aoki T; Murakami M

**INSTITUCIÓN / INSTITUTION:** - Department of Gastroenterological Surgery, Shiroyama Hospital, Ota, Gunma, Japan.

**RESUMEN / SUMMARY:** - AIM: We report on a rare case of appendiceal primary neuroendocrine carcinoma (NEC) and discuss three cases previously described. CASE REPORT: A 58-year-old woman presented with acute abdominal pain and a low-grade fever. She was diagnosed with acute appendicitis and underwent laparoscopic appendectomy. Pathological examination of the resected specimen revealed NEC. Immunohistochemical analyses were positive for synaptophysin, chromogranin, and CD-56. The

tumour was high grade and the Ki-67 index was >20%. Primary NEC of the appendix is extremely rare. To our knowledge, this is the first case report of an appendiceal NEC that meets the 2010 diagnostic criteria of the World Health Organization. CONCLUSION: Due to its non-specific clinical presentation, NEC is often misdiagnosed as appendicitis; however, it can advance rapidly and carries a very poor prognosis, despite chemotherapy. In the future, a treatment protocol for immunohistochemical analyses should be established to improve the survival rate.

[7]

**TÍTULO / TITLE:** - Renal hybrid oncocytic/chromophobe tumors - A review.

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

**REVISTA / JOURNAL:** - Histol Histopathol. 2013 Jun 6.

**AUTORES / AUTHORS:** - Hes O; Petersson F; Kuroda N; Hora M; Michal M

**INSTITUCIÓN / INSTITUTION:** - Department of Pathology, Charles University in Prague, Faculty of Medicine in Plzen, Czech Republic. [hes@medima.cz](mailto:hes@medima.cz).

**RESUMEN / SUMMARY:** - Hybrid oncocytic/chromophobe tumors (HOCT) occur in three clinico-pathologic situations; (1) sporadically, (2) in association with renal oncocytomatosis and (3) in patients with Birt-Hogg-Dube syndrome (BHD). There are no specific clinical symptoms in patients with sporadic or HOCT associated with oncocytosis/oncocytomatosis. HOCT in patients with BHD are usually encountered on characteristic BHD clinicopathologic background. Sporadic HOCT are composed of neoplastic cells with eosinophilic oncocytic cytoplasm. Tumors are usually arranged in a solid-alveolar pattern. Some neoplastic cells may have a perinuclear halo, no raisinoid nuclei are present. HOCT occurring in patients with oncocytomatosis are morphologically identical to sporadic HOCT. HOCT in BHD frequently display 3 morphologic patterns, either in isolation or in combination; (1) An admixture of areas typical of RO and CHRCC, respectively, (2) Scattered chromophobe cells in the background of a typical RO, (3) Large eosinophilic cells with intracytoplasmic vacuoles. The immunohistochemical profiles of HOCT in all clinicopathologic and morphologic groups differ slightly. The majority of tumors express parvalbumin, antimitochondrial antigen and CK 7. CD117 is invariably positive. HOCT show significant molecular genetic heterogeneity. The highest degree of variability in numerical chromosomal changes is present in sporadic HOCT. HOCT in the setting of oncocytomatosis have revealed a lesser degree of variability in the chromosomal numerical aberrations. HOCT in patients with BHD display FLCN gene mutations, which are absent in the other groups. HOCT (all three clinicopathologic groups) seem to behave indolently, as no evidence of aggressive behavior has been documented. However, no report with follow up longer than 10 years has been published.

[8]

**TÍTULO / TITLE:** - Somatostatin receptor PET/CT in neuroendocrine tumours: update on systematic review and meta-analysis.

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

**REVISTA / JOURNAL:** - Eur J Nucl Med Mol Imaging. 2013 Jul 20.

●● Enlace al texto completo (gratis o de pago) [1007/s00259-013-2482-](#)

**Z**

**AUTORES / AUTHORS:** - Geijer H; Breimer LH

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**RESUMEN / SUMMARY:** - **PURPOSE:** Neuroendocrine tumours (NET) are uncommon and may be localized in many different places in the body. Traditional imaging has mainly been performed with CT and somatostatin receptor scintigraphy (SRS). Recently, it has become possible to use somatostatin receptor PET/CT (SMSR PET) instead, which might improve diagnostic quality. To evaluate the diagnostic quality of SMSR PET we performed a meta-analysis as an update of a previous study published in 2012. **METHODS:** A literature search was performed searching MEDLINE, Embase and five other databases with a combination of the expressions "PET", "positron emission tomography", "neuroendocrine" and "NET". The search was updated to 31 December 2012. Studies were selected which evaluated the sensitivity and specificity of SMSR PET for NET in the thorax or abdomen with a study size of at least eight patients. The methodological quality of the included studies was evaluated with QUADAS-2. **RESULTS:** Eight studies fulfilled the inclusion criteria and were selected for final analysis, and 14 articles from a previous meta-analysis were added for a total of 22 articles. A total of 2,105 patients were included in the studies, an increase from 567 in the previous meta-analysis. The pooled sensitivity was 93 % (95 % CI 91 - 94 %) and specificity 96 % (95 % CI 95 - 98 %). The area under the summary ROC curve was 0.98 (95 % CI 0.95 - 1.0). In the previous meta-analysis the pooled sensitivity was 93 % (95 % CI 91 - 95 %) and specificity 91 % (95 % CI 82 - 97 %). **CONCLUSION:** SMSR PET has good diagnostic performance for evaluation of NET in the thorax and abdomen, better than SRS which has been the previous standard method. This meta-analysis gives further support for switching to SMSR PET.

[9]

**TÍTULO / TITLE:** - Quality of life, resource utilisation and health economics assessment in advanced neuroendocrine tumours: a systematic review.

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

**REVISTA / JOURNAL:** - Eur J Cancer Care (Engl). 2013 Jul 29. doi: 10.1111/ecc.12085.

●● Enlace al texto completo (gratis o de pago) [1111/ecc.12085](#)

**AUTORES / AUTHORS:** - Chau I; Casciano R; Willet J; Wang X; Yao JC

**INSTITUCIÓN / INSTITUTION:** - The Royal Marsden Hospital, London, UK.

**RESUMEN / SUMMARY:** - Neuroendocrine tumours (NET) are often diagnosed at an advanced stage when the prognosis is poor for patients, who often experience diminished quality of life (QoL). As new treatments for NET become available, it is important to characterise the associated outcomes, costs and QoL. A comprehensive search was performed to systematically review available data in advanced NET regarding cost of illness/resource utilisation, economic studies/health technology assessment and QoL. Four rounds of sequential review narrowed the search results to 22 relevant studies. Most focused on surgical procedures and diagnostic tools and contained limited information on the costs and consequences of medical therapies. Multiple tools are used to assess health-related QoL in NET, but few analyses have been conducted to assess the comparative impact of available treatment alternatives on QoL. Limitations include English language and the focus on advanced NET; ongoing terminology and classification changes prevented pooled statistical analyses. This systematic review suggests a lack of comparative economic and outcomes data associated with NET treatments. Further research on disease costs, resource utilisation and QoL for patients with advanced NET is warranted.

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[10]

**TÍTULO / TITLE:** - Factors Associated with Lymph Node Metastasis in Radically Resected Rectal Carcinoids: a Systematic Review and Meta-analysis.

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

**REVISTA / JOURNAL:** - J Gastrointest Surg. 2013 Jul 2.

●● Enlace al texto completo (gratis o de pago) [1007/s11605-013-2249-](#)

[7](#)

**AUTORES / AUTHORS:** - Zhou X; Xie H; Xie L; Li J; Fu W

**INSTITUCIÓN / INSTITUTION:** - Department of General Surgery, Peking University Third Hospital, Beijing, 100191, China.

**RESUMEN / SUMMARY:** - BACKGROUND: Although various guidelines regarding neuroendocrine tumors were released, treatment for rectal neuroendocrine tumors with size between 1 and 2 cm has not been explicitly elucidated. The determinant factor of the choice between endoscopic resection and radical surgery is whether lymph node involvement exists. AIM: This study aims to explore factors associated with lymph node involvement in rectal neuroendocrine tumors by conducting a meta-analysis. METHODS: A broad literature research of Pubmed, Embase&Medline, and The Cochrane Library was performed, and systematic review and meta-analysis about factors associated with lymph node involvement were conducted. RESULTS: Seven studies were included in this meta-analysis. Tumor size > 1 cm (odds ratio (OR) 6.72, 95 % confidence interval (CI) [3.23, 14.02]), depth of invasion (OR 5.06, 95 % CI [2.30, 11.10]), venous invasion (OR 5.92, 95 % CI [2.21, 15.87]), and

central depression (OR 3.00, 95 % CI [1.07, 8.43]) were significantly associated with lymph node involvement. CONCLUSION: The available clinical evidence suggests that tumor size > 1 cm, invasion of muscularis propria, venous invasion, and central depression could be risk factors of lymph node involvement, while other factors reported by few studies need further research.

[11]

**TÍTULO / TITLE:** - Merkel cell carcinoma in elderly: case report and review of the literature.

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

**REVISTA / JOURNAL:** - Aging Clin Exp Res. 2013 May;25(2):211-4. doi: 10.1007/s40520-013-0020-2. Epub 2013 Apr 9.

●● Enlace al texto completo (gratis o de pago) [1007/s40520-013-0020-](#)

[2](#)

**AUTORES / AUTHORS:** - Ultori C; Cimetti L; Stefanoni P; Pellegrini R; Rapazzini P; Capella C

**INSTITUCIÓN / INSTITUTION:** - Unita Operativa di Geriatria, Azienda Ospedaliera, Ospedale di Circolo e Fondazione Macchi, Varese, Italy.

**RESUMEN / SUMMARY:** - Merkel cell carcinoma (MCC) is a rare neuroendocrine tumour of the skin, characterised by an aggressive clinical course. The incidence of this rare neoplasia is rapidly increasing. Herein we report our experience with a patient who developed a MCC of the inguinal region.

[12]

**TÍTULO / TITLE:** - Giant merkel cell carcinoma of the lower limb: case report and review of the literature.

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

**REVISTA / JOURNAL:** - J Cutan Med Surg. 2013;17(0):1-5.

**AUTORES / AUTHORS:** - Marchesi A; Camillo Parodi P; Brioschi M; Sileo G; Marchesi M; Vaienti L

**RESUMEN / SUMMARY:** - Background:Merkel cell carcinoma (MCC) is a rare cutaneous neuroendocrine malignancy that usually grows rapidly at the head and neck. Giant forms at the lower limbs are rarely reported and usually affect patients in the eighth decade or older.Methods:We report the case of a 60-year-old man who presented with a giant MCC on his right thigh. We managed this case by applying the 2012 updated guidelines and reviewed all cases of giant MCC of the lower limbs reported in the literature.Results:At the 4-month follow-up, the patient showed complete remission.Conclusion:Giant forms of MCC are still treated as typical cases of MCC, when these patients show a very poor prognosis. In young and adult people, such as our case, wide surgical excisions, sentinel lymph node biopsy in clinically negative node cases,

radiotherapy of the regional drain area, and a strict follow-up should be routinely performed to improve patients' survival.

[13]

**TÍTULO / TITLE:** - Neuroendocrine tumor in the liver of a patient with isolated polycystic liver disease: A case report and review of the literature.

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

**REVISTA / JOURNAL:** - Oncol Lett. 2013 May;5(5):1664-1666. Epub 2013 Mar 6.

●● [Enlace al texto completo \(gratis o de pago\) 3892/ol.2013.1233](#)

**AUTORES / AUTHORS:** - Koutsampasopoulos K; Antoniadou E; Zoutis S; Iacovidis G; Burova O; Taplidis A

**INSTITUCIÓN / INSTITUTION:** - Department of Internal Medicine, General Hospital of Naoussa, Thessaloniki, Greece.

**RESUMEN / SUMMARY:** - Neuroendocrine tumors (NETs) frequently metastasize to the liver, but it is rare to find them there as primary tumors. Isolated polycystic liver disease (PCLD) is a rare autosomal dominant disease. There is no known association between polycystic liver disease and neuroendocrine or other tumors. We report a case of a 64-year-old female with a past medical history of isolated PCLD who presented with increasing abdominal pain over a two-week period. Our patient underwent open surgical biopsy one month after presentation. The histological examination and immunohistochemical findings suggested an intermediate grade neuroendocrine tumor. A 24-h delayed whole-body scintigraphy technique was utilized for the identification and localization of neuroendocrine tumors via the administration of In-111-labeled OctreoScan; however, no extrahepatic accumulation was observed. No previous studies in the literature describe a patient with PCLD and a primary or metastatic neuroendocrine tumor of the liver.

[14]

**TÍTULO / TITLE:** - Pancreatic schwannoma: A case report and review of literature.

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

**REVISTA / JOURNAL:** - J Med Assoc Thai. 2013 Jan;96(1):112-6.

**AUTORES / AUTHORS:** - Poosawang W; Kiatkungwankai P

**INSTITUCIÓN / INSTITUTION:** - Department of Surgery, Samutsakhon Hospital, Samutsakhon, Thailand. [boon8281@hotmail.com](mailto:boon8281@hotmail.com)

**RESUMEN / SUMMARY:** - Pancreatic schwannoma is an extremely rare neoplasm, derived from Schwann cells that line the nerve sheaths. It is also referred to as neurilemmoma. The authors report a case of a 46-year-old Thai female who presented with dyspepsia, weight loss and epigastric mass. An examination by ultrasonography and computed tomography (CT) scan revealed a septated cystic tumor in the pancreatic head, 5.8x5.5x5.3 cm in size. Pancreaticoduodenectomy was performed to remove this tumor. A microscopic

examination identified proliferating spindle cells that are consistent with neurilemmoma (schwannoma). No complications were found after the operation. At 18-month follow-up, the patient remains asymptomatic and has no signs of recurrence.

[15]

**TÍTULO / TITLE:** - Large-cell neuroendocrine carcinoma of lung with epidermal growth factor receptor (EGFR) gene mutation and co-expression of adenocarcinoma markers: a case report and review of the literature.

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

**REVISTA / JOURNAL:** - Multidiscip Respir Med. 2013 Jul 18;8(1):47. doi: 10.1186/2049-6958-8-47.

●● Enlace al texto completo (gratis o de pago) [1186/2049-6958-8-47](#)

**AUTORES / AUTHORS:** - Sakai Y; Yamasaki T; Kusakabe Y; Kasai D; Kotani Y; Nishimura Y; Itoh T

**INSTITUCIÓN / INSTITUTION:** - Department of Diagnostic Pathology, Kobe University Hospital, 7-5-2 Kusunoki-cho, Chuo-ku, Kobe-shi, Hyogo 650-0017, Japan. [sakaiyasuhiro@gaia.eonet.ne.jp](mailto:sakaiyasuhiro@gaia.eonet.ne.jp).

**RESUMEN / SUMMARY:** - PURPOSE: A high rate of response to treatment with epidermal growth factor receptor tyrosine kinase inhibitor (EGFR-TKI) has been observed in certain patients (women, of East Asian ethnicity, with non-smoking history and adenocarcinoma histology) with mutations in exons 18 to 21 of the tyrosine kinase domain of EGFR. Some cases of high-grade neuroendocrine carcinoma of the lung harboring mutations have been sporadically reported. METHODS: We describe the case of a 78-year-old woman with large-cell neuroendocrine carcinoma of the lung, with mutation in exon 21 L858R and co-expression of adenocarcinoma markers. RESULTS: A mass (3.0 cm in diameter) was identified in the inferior lobe of the left lung, accompanied by metastases into ipsilateral mediastinal lymph nodes and elevations of serum pro-gastrin-releasing peptide and carcinoembryonic antigen. Initial transbronchial brushing cytology suggested high-grade neuroendocrine carcinoma favoring small-cell carcinoma in poorly smeared and degenerated preparations, and revealed exon 21 L858R mutation. Re-enlargement of the cancer and bone metastases was observed after chemotherapy, and further testing suggested large-cell neuroendocrine carcinoma with immunoreactivity to markers of primary lung adenocarcinoma and L858R mutation. High-grade neuroendocrine carcinoma with mutations in the tyrosine kinase domain of EGFR may be associated with adenocarcinoma, as reviewed from the literature and may also apply to our case. CONCLUSIONS: EGFR-TKI could provide better quality of life and survival in patients with advanced or relapsed high-grade neuroendocrine carcinoma with EGFR gene mutations. Further studies in this respect are warranted.

[16]

**TÍTULO / TITLE:** - Guidelines for biomarker testing in gastroenteropancreatic neuroendocrine neoplasms: a national consensus of the Spanish Society of Pathology and the Spanish Society of Medical Oncology.

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

**REVISTA / JOURNAL:** - Clin Transl Oncol. 2013 Jun 8.

●● Enlace al texto completo (gratis o de pago) [1007/s12094-013-1062-](#)

[9](#)

**AUTORES / AUTHORS:** - Garcia-Carbonero R; Vilardell F; Jimenez-Fonseca P; Gonzalez-Campora R; Gonzalez E; Cuatrecasas M; Capdevila J; Aranda I; Barriuso J; Matias-Guiu X

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**RESUMEN / SUMMARY:** - The annual incidence of neuroendocrine tumours in the Caucasian population ranges from 2.5 to 5 new cases per 100,000 inhabitants. Gastroenteropancreatic neuroendocrine tumours is a family of neoplasms widely variable in terms of anatomical location, hormone composition, clinical syndromes they cause and in their biological behaviour. This high complexity and clinical heterogeneity, together with the known difficulty of predicting their behaviour from their pathological features, are reflected in the many classifications that have been developed over the years in this field. This article reviews the main tissue and clinical biomarkers and makes recommendations for their use in medical practice. This document represents a consensus reached jointly by the Spanish Society of Medical Oncology (SEOM) and the Spanish Society of Pathology (SEAP).

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[17]

**TÍTULO / TITLE:** - Neuroendocrine small cell rectal cancer metastasizing to the liver: a unique treatment strategy, case report, and review of the literature.

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

**REVISTA / JOURNAL:** - World J Surg Oncol. 2013 Jul 11;11:153. doi: 10.1186/1477-7819-11-153.

●● Enlace al texto completo (gratis o de pago) [1186/1477-7819-11-153](#)

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**RESUMEN / SUMMARY:** - We describe the treatment of a 46-year-old Saudi man with advanced stage liver metastatic neuroendocrine rectal cancer. The patient presented with a large liver lesion and rectal bleeding. He was cachectic, with a firm tender mass 20 mm above the anal verge. Computed tomography (CT) showed a mass 9.5 x 13 cm in size in the right hemi-liver, abutting the middle

hepatic vein. The patient refused treatment, and consulted another hospital. After 3 months, he presented with the same symptoms in addition to delirium. Colonoscopy showed an ulcerating anorectal mass, from which a biopsy was taken. Repeat CT showed an increase in the size of the liver lesion to 17 cm and no change in the pelvis. The final histopathology report identified anaplastic small cell carcinoma. The patient underwent extended right liver resection followed by abdominoperineal resection, then 13 cycles of chemotherapy and monthly somatostatin injections. At the most recent follow-up, the patient had been disease-free for 48 months. Surgical resection (R0) of the primary and secondary tumor, followed by platinum-based chemotherapy can result in good survival in cases of small cell carcinoma with large liver metastasis, irrespective of whether the primary or secondary tumor is resected first.

[18]

**TÍTULO / TITLE:** - Comparison of metaiodobenzylguanidine scintigraphy with positron emission tomography in the diagnostic work-up of pheochromocytoma and paraganglioma: a systematic review.

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

**REVISTA / JOURNAL:** - Q J Nucl Med Mol Imaging. 2013 Jun;57(2):122-33.

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**RESUMEN / SUMMARY:** - Aim: The aim of this paper was to systematically review published data about the comparison of radiolabelled metaiodobenzylguanidine (MIBG) scintigraphy and positron emission tomography (PET) with different radiopharmaceuticals in patients with pheochromocytoma and paraganglioma (Pheo/PGL). Methods: A comprehensive literature search of studies published in PubMed/MEDLINE and Embase databases through September 2012 and regarding MIBG scintigraphy and PET imaging with different radiopharmaceuticals in patients with Pheo/PGL was carried out. Results: Twenty-eight studies comprising 852 patients who underwent both MIBG scintigraphy and PET or PET/CT with different radiopharmaceuticals were included and discussed. Three studies evaluated carbon-11-hydroxyephedrine ([<sup>11</sup>C]HED) as PET radiopharmaceutical, nine studies fluorine-18-dopamine ([<sup>18</sup>F]DA), eight studies fluorine-18-dihydroxyphenylalanine ([<sup>18</sup>F]DOPA), twelve studies fluorine-18-fluorodeoxyglucose ([<sup>18</sup>F]FDG) and five studies gallium-68-somatostatin analogues. Conclusions: Despite the heterogeneity of the studies included in the analysis, it can be concluded that the diagnostic performance of PET with various agents is clearly superior to that of MIBG scintigraphy in patients with Pheo/PGL, mainly for familial, extra-adrenal and metastatic diseases; however, MIBG maintains a unique role in selecting patients suitable for <sup>131</sup>I-MBG therapy. Further larger prospective studies comparing MIBG and different PET tracers in patients with Pheo/PGL as well as a cost-effectiveness analysis of the two techniques are needed.

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[19]

**TÍTULO / TITLE:** - Locally-advanced primary neuroendocrine carcinoma of the breast: case report and review of the literature.

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

**REVISTA / JOURNAL:** - World J Surg Oncol. 2013 Jun 5;11:128. doi: 10.1186/1477-7819-11-128.

●● Enlace al texto completo (gratis o de pago) [1186/1477-7819-11-128](#)

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**RESUMEN / SUMMARY:** - BACKGROUND: Primary neuroendocrine carcinoma of the breast is a heterogeneous group of rare tumors with positive immunoreactivity to neuroendocrine markers in at least 50% of cells. Diagnosis also requires that other primary sites be ruled out and that the same tumor show histological evidence of a breast in situ component. Primary neuroendocrine carcinoma of the breast rarely presents as locally advanced disease and less frequently with such widespread metastatic disease as described herein. The review accompanying this case report is the first to provide an overview of all the cases of primary neuroendocrine carcinoma of the breast published in the literature and encompasses detailed information regarding epidemiology, histogenesis, clinical and histologic diagnosis criteria, classification, surgical and adjuvant treatment, as well as prognosis. We also provide recommendations for common clinical and histologic pitfalls associated with this tumor. CASE PRESENTATION: We describe a case of a 51-year-old Hispanic woman initially diagnosed with locally-advanced invasive ductal carcinoma that did not respond to neoadjuvant treatment. After undergoing modified radical mastectomy the final surgical pathology showed evidence of alveolar-type primary neuroendocrine carcinoma of the breast. The patient was treated with cisplatin/etoposide followed by paclitaxel/carboplatinum. Thirteen months after surgery the patient is alive, but developed pulmonary, bone, and hepatic metastasis. CONCLUSION: The breast in situ component of primary neuroendocrine carcinoma of the breast may prevail on a core biopsy samples increasing the probability of underdiagnosing this tumor preoperatively. Being aware of the existence of this disease allows for timely diagnosis and management. Optimal treatment requires simultaneous consideration of both the neuroendocrine and breast in situ tumor features.

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[20]

**TÍTULO / TITLE:** - Von Hippel-Lindau and myotonic dystrophy of Steinert along with pancreatic neuroendocrine tumor and renal clear cell carcinoma neoplasm: Case report and review of the literature.

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

**REVISTA / JOURNAL:** - Int J Surg Case Rep. 2013;4(8):648-50. doi: 10.1016/j.ijscr.2013.03.004. Epub 2013 Mar 29.

●● Enlace al texto completo (gratis o de pago) [1016/j.ijscr.2013.03.004](http://1016/j.ijscr.2013.03.004)

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**RESUMEN / SUMMARY:** - INTRODUCTION: Myotonic dystrophy of Steinert, DM1, is the most common adult muscular dystrophy and generally is not associated to development on multiple site neoplasm. Von Hippel-Lindau (VHL) disease is a dominantly inherited familial cancer syndrome that is associated to tumors such as hemangioblastoma of the retina or central nervous system, clear-cell renal carcinoma (RCC) and endocrine tumors, most commonly pheochromocytoma and non-secretory pancreatic islet cell cancers. No data exist in literature describing the coexistence of both DM1 and VHL.

PRESENTATION OF CASE: Herein we report a case of renal and pancreatic neoplasm in a young adult female affected by DM1 and VHL simultaneously.

DISCUSSION: DM1 is due to an unstable trinucleotide (CTG) expansion in the 30 untranslated region of the dystrophin myotonia-protein kinase (DMPK) gene, located on chromosome 19q13.3. Several molecular mechanisms thought to be determining the classical DM phenotype have been shown. VHL disease is characterized by marked phenotypic variability and the most common tumors are hemangioblastomas of the retina or central nervous system, clear-cell renal carcinoma (RCC) and endocrine tumors, most commonly pheochromocytoma and non-secretory pancreatic islet cell cancers. The pancreatic manifestations seen in patients with VHL disease are divided into 2 categories: pancreatic neuroendocrine tumor (PNET) as solid tumors, and cystic lesions, including a simple cyst and serous cystadenoma. The surgical approach for these cystic lesions is to consider as golden standard. Blansfield has proposed 3 criteria to predict metastatic disease of PNET in patients with VHL disease: (1) tumor size greater than or equal to 3cm; (2) presence of a mutation in exon 3; and (3) tumor doubling time less than 500d. If the patient has none of these criteria the patient could be followed with physical examination and radiological surveillance on a 2/3 years base.(4) If the patient has 1 criterion, the patient should be followed more closely every 6 months to 1 year. If the patient has 2 or 3 criteria, the patient should be considered for surgery given the high risk of future malignancy. Our patient owned only one criterion but in presence of a second malignant tumor. Our hypothesis for this rare findings is that both DM and VHL might be derived from genetic aberration and these might be linked to a major cancer susceptibility. As far as we know this is the first confirmed case of RCC and neuroendocrine pancreatic cancer occurring concurrently with VHL and, at the same time, DM1. According to this case report and the literature data a VHL should be ruled out in the presence of RCC presenting along with pancreatic cysts/tumor. CONCLUSION: As far as we know this is the first confirmed case of RCC and neuroendocrine pancreatic cancer occurring

concurrently with VHL and, at the same time, DM1. Our hypothesis for the unusual findings is that both DM and VHL derived from genetic aberration and these are linked to a major cancer susceptibility.

[21]

**TÍTULO / TITLE:** - Rare large cell neuroendocrine tumor of the endometrium: A case report and review of the literature.

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

**REVISTA / JOURNAL:** - Int J Surg Case Rep. 2013;4(8):651-5. doi: 10.1016/j.ijscr.2013.04.027. Epub 2013 May 3.

●● Enlace al texto completo (gratis o de pago) [1016/j.ijscr.2013.04.027](http://1016/j.ijscr.2013.04.027)

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**RESUMEN / SUMMARY:** - INTRODUCTION: Large cell neuroendocrine carcinoma (LCNEC) of the endometrium is a rare malignancy with an aggressive course. Although data is limited to case reports, the prognosis appears to be poor, similar to other type II uterine cancers. A total of 12 cases of LCNEC of the uterus have been published to date. PRESENTATION OF CASE: A 71 year-old woman presented with postmenopausal vaginal bleeding. Endometrial biopsy was non-diagnostic for LCNEC. She underwent surgical debulking and staging of a 22cm endometrial tumor with omental metastasis and positive lymph nodes. Her final FIGO stage was IVB. DISCUSSION: We summarize all prior case reports of LCNEC of the endometrium and discuss the definition, presentation, imaging and surgical management. The pathology with immunohistochemical review, adjuvant therapy and prognosis of LCNEC of the endometrium are also reviewed. CONCLUSION: Pathologic findings and immunohistochemistry are essential in making a diagnosis of LCNEC of the endometrium. Primary debulking and surgical staging is typically performed, but if a diagnosis of LCNEC can be made preoperatively with immunohistochemistry, surgeons should consider neoadjuvant chemotherapy due to its high grade histology and aggressive course. Otherwise adjuvant chemotherapy is usually given. Even with early stage disease, the prognosis seems poor. Due to the rarity of this aggressive malignancy, more data is needed to establish incidence.

[22]

**TÍTULO / TITLE:** - Merkel cell carcinoma of left groin: a case report and literature review.

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

**REVISTA / JOURNAL:** - Case Rep Oncol Med. 2013;2013:431743. doi: 10.1155/2013/431743. Epub 2013 May 21.

●● Enlace al texto completo (gratis o de pago) [1155/2013/431743](#)

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**RESUMEN / SUMMARY:** - Merkel cell carcinoma (MCC) is an uncommon highly aggressive skin malignancy with an increased tendency to recur locally, invade regional lymph nodes, and metastasize distally to lung, liver, brain, bone, and skin. The sun-exposed skin of head and neck is the most frequent site of involvement (55%). We report the case of a 63-year-old Caucasian male patient who presented with a recurrent left inguinal mass for the third time after surgical resection with safe margins and no postoperative radio- or chemotherapy. The presented mass was excised, and pathological diagnosis revealed recurrent MCC. The patient underwent postoperative radiation therapy, and 6 months later, he developed a right groin mass which was resected and pathological diagnosis confirmed metastatic MCC. Six months later, patient developed an oropharyngeal mass which was unresectable, and pathological biopsy confirmed metastatic MCC. Patient was offered palliative radio- and chemotherapy. In this paper, we also present a brief literature review on MCC.

[23]

**TÍTULO / TITLE:** - Brief S2k guidelines—Merkel cell carcinoma.

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

**REVISTA / JOURNAL:** - J Dtsch Dermatol Ges. 2013 Jun;11 Suppl 3:29-36, 31-8. doi: 10.1111/ddg.12015\_6.

●● Enlace al texto completo (gratis o de pago) [1111/ddg.12015\\_6](#)

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