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RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: Saif MW
INSTITUCIÓN / INSTITUTION: GI Cancers and Experimental Therapeutics, Tufts University School of Medicine, Boston, MA 02111, USA. wsaif@tuftsmedicalcenter.org
RESUMEN / SUMMARY: Adenocarcinoma ex goblet cell carcinoid is a rare neoplasm of appendiceal origin that contains features of both carcinoid tumor and adenocarcinoma. We report on a case of a 45-year-old woman, post-renal transplant who presented with ovarian metastases from this tumor. This appears to be the first report of an adenocarcinoma ex goblet cell carcinoid in a renal transplant recipient.

RESUMEN / SUMMARY: Enlace al Resumen / Link to its Summary
Medullary thyroid carcinoma (MTC) is a rare tumour which frequently occurs in the context of the multiple endocrine neoplasia syndromes, where it coexists with other usually benign tumours. The clinical picture varies and distant metastases are frequently present at diagnosis. Calcitonin levels are elevated in the presence of metastatic disease. Two MTC cases are presented, which had elevated postoperative calcitonin levels. Imaging revealed lung lesions which were originally attributed to metastatic disease from the MTC. However, at follow-up, these cases presented unusual features. The rapid increase in the lung lesions and the development of hypercalcaemia in the first patient suggested a second unrelated tumour. Biopsy of the lung lesion was compatible with lung adenocarcinoma. In the second patient, the appearance of a liver mass, although calcitonin levels remained stable, led to biopsy of the lesion: this was negative for calcitonin and compatible with metastatic lung adenocarcinoma. These MTC cases show that further malignancies may coexist with MTC and may obscure the clinical picture and influence the therapeutic decisions, especially in the case of metastatic disease. Features such as unusual imaging characteristics and the development of hypercalcaemia, never encountered in MTC outside the MEN2 syndromes, as well as ‘disproportionately’ low calcitonin levels, incompatible with extensive metastatic disease, were the factors that led to further work-up. Both the cases subsequently proved to carry an unsuspected second malignancy. It is crucial to discriminate the metastatic lesion attributed to MTC from another coexisting primary malignancy, because different therapeutic strategies are needed for each setting.

[3]

AUTORES / AUTHORS: - Kunz PL; Reidy-Lagunes D; Anthony LB; Bertino EM; Brendtro K; Chan JA; Chen H; Jensen RT; Kim MK; Klimstra DS; Kulke MH; Liu EH; Metz DC; Phan AT; Sippel RS; Strosberg JR; Yao JC
Neuroendocrine tumors are a heterogeneous group of tumors originating in various anatomic locations. The management of this disease poses a significant challenge because of the heterogeneous clinical presentations and varying degrees of aggressiveness. The recent completion of several phase 3 trials, including those evaluating octreotide, sunitinib, and everolimus, demonstrate that rigorous evaluation of novel agents in this disease is possible and can lead to practice-changing outcomes. Nevertheless, there are many aspects to the treatment of neuroendocrine tumors that remain unclear and controversial. The North American Neuroendocrine Tumor Society published a set of consensus guidelines in 2010, which provided an overview for the treatment of patients with these malignancies. Here, we present a set of consensus tables intended to complement these guidelines and serve as a quick, accessible reference for the practicing physician.

[4]

Adrenal hemorrhagic pseudocyst as the differential diagnosis of pheochromocytoma- a review of the clinical features in cases with radiographically diagnosed pheochromocytoma.

Background: Clinical diagnosis of pheochromocytoma is difficult for some adrenal tumors. Aim: Herein, we review clinical and pathological findings of 31 cases with radiographically diagnosed pheochromocytoma, including three cases of hemorrhagic pseudocysts (HPC). Materials/Subjects and Methods: Between January 1992 and December 2010, 31 patients with adrenal tumors were preoperatively diagnosed as having pheochromocytoma by radiographic imaging, and underwent adrenalectomy. Histological examination revealed HPC in 3 patients (9.7%), and pheochromocytoma in the remaining 28 patients. We reviewed and compared the clinical features, including the biochemical and radiographic features, of HPC and pheochromocytoma cases. Results: Biochemical testing showed no definitive excessive catecholamine secretion in any of the three patients with HPC and four (14.3%) of those with histologically proven pheochromocytoma. 13115 I-metaiodobenzylguanidine scintigraphy was negative in the three with HPC, but positive in all of the four with pheochromocytoma who did not have suggestive biochemical results. All HPC patients had concomitant disease or...
symptoms suggestive of pheochromocytoma, and two had received an
anticoagulant or antiplatelet agent. Laparoscopic surgery was completed in two
cases of HPC uneventfully. Conclusions: Adrenal HPC may have radiographic
characteristics similar to those of pheochromocytoma. Adrenal HPC should be
considered as a differential diagnosis of pheochromocytoma.

[5]
TITULO / TITLE: - Insulinoma: only in adults? - case reports and literature review.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary

Resumen / Summary: Insulinomas first presenting as refractory seizure
disorders are well documented in adulthood but rarely found in children. Only a
few cases of childhood insulinoma have been reported so far. We report on two
adolescents with hyperinsulinaemic hypoglycaemia, initially misdiagnosed as
epilepsy and migraine accompagnée, and compare those to other cases
published. Localization of insulinoma was challenging and, in one patient,
angiography with selective arterial calcium stimulation and hepatic venous
sampling in addition to CT and MRI was necessary. In these patients, long-
term recovery was achieved by laparoscopic distal pancreatic resection in one and
by conventional enucleation in the pancreatic head in the second patient. In
contrast to adults, macrosomy and a decrease in school performance were the
main symptoms and, during fasting, impaired cognitive function occurred after
a relatively short period and at a higher glucose threshold or lower
insulin/glucose ratio, respectively. Neuroglycopenic signs may be attributed to
behaviour abnormalities or seizure disorders but in children and adolescents
may already be caused by insulinoma. In these cases, timely diagnosis as well
as tumour resection ensure long-term cure.

[6]
TITULO / TITLE: - Intracranial malignant triton tumor in a patient with
neurofibromatosis type 1: case report and review of the literature.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
REVISTA / JOURNAL: - Brain Tumor Pathol. 2013 Apr 30.

Resumen / Summary: Intracranial malignant triton tumor in a patient with
neurofibromatosis type 1: case report and review of the literature.
AUTORES / AUTHORS: - Smith RE; Kebriaei MA; Gard AP; McComb RD; Bridge JA; Lennarson PJ
INSTITUCIÓN / INSTITUTION: - Division of Neurosurgery, Department of Surgery, University of Nebraska Medical Center, 11364 Gold St, Omaha, NE, 68144, USA, rsmithe@unmc.edu.
RESUMEN / SUMMARY: - We report the fourth case of an intracranial malignant triton tumor not associated with a cranial nerve in a 26-year-old male with a clinical history of neurofibromatosis type 1. The patient was found unresponsive and displayed confusion, lethargy, hyperreflexia, and dysconjugate eye movements upon arrival at the emergency room. MRI revealed a large bifrontal mass. Biopsy demonstrated a high-grade spindle cell tumor with focal areas of rhabdomyoblasts that stained positive for desmin, myogenin, and muscle-specific actin. Electron microscopy showed skeletal muscle differentiation. Based on the clinical history of NF1 and the pathologic results, a diagnosis of malignant triton tumor was made. The differential diagnosis, immunohistochemistry, molecular genetics, and treatment of malignant triton tumor are reviewed.

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RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Then C; Nam-Apostolopoulos YC; Seissler J; Lechner A
INSTITUCIÓN / INSTITUTION: - Diabetes Center, Clinic and Polyclinic IV, “Ludwig-Maximilians” University. Munich, Germany. cornelia.then@med.uni-muenchen.de.
RESUMEN / SUMMARY: - CONTEXT: Non-insulinoma pancreatogenous hypoglycemia is a rare cause of spontaneous hypoglycemia in adults. The ideal diagnostic and therapeutic approach is still controversial, not least because most reported cases lack long-term follow-up. CASE REPORT: We describe the case of a 58-year-old woman, who was diagnosed with idiopathic non-insulinoma pancreatogenous hypoglycemia in 2001. After resection of 75% of the distal pancreas, she initially experienced no additional hypoglycemic episodes and did not suffer from diabetes mellitus. However, after one month, recurrent hypoglycemia occurred. After resection of the larger part of the remaining pancreatic tissue, the patient suffered from hypoglycemic as well as hyperglycemic episodes. Octreotide and diazoxide were not successful in preventing the hypoglycemic attacks, whereas continuous insulin therapy with an insulin pump helped to stabilize the blood glucose level temporarily. Finally, all remaining pancreatic tissue had to be removed. CONCLUSION: This long-term follow-up of non-insulinoma pancreatogenous hypoglycemia treatment in
an adult patient indicates that lateral pancreatectomy may not be sufficient for permanent blood glucose control and emphasizes the need of follow-up data after subtotal pancreatectomy.

[8]

TÍTULO / TITLE: - Gallium-68 DOTANOC imaging in paraganglioma/pheochromocytoma: presentation of sample cases and review of the literature.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Lopci E; Zanoni L; Fanti S; Ambrosini V; Castellani MR; Aktolun C; Chiti A
INSTITUCIÓN / INSTITUTION: - Nuclear Medicine Department Humanitas Clinical and Research Center Rozzano (MI), Italy - egesta.lopci@gmail.com.
RESUMEN / SUMMARY: - Gallium-68 DOTANOC is a high affinity somatostatin receptor ligand, first introduced in 2005 for imaging neuroendocrine tumors. Due to its technically simple production, broad availability, favourable biodistribution and advantageous dosimetry, although not approved yet in all European countries, gallium-68 DOTANOC has rapidly gained acceptance in the diagnostic and therapeutic work-flow of different types of neuroendocrine tumors. Principal indications in clinical practice in countries where it is officially approved include diagnosis and staging, restaging after treatment, identification of sites of unknown primary and selection of patients with neuroendocrine tumors eligible for therapy with somatostatin analogues.

[9]

TÍTULO / TITLE: - Pheochromocytoma of the urinary bladder: a systematic review of the contemporary literature.
RESUMEN / SUMMARY: - Enlace al Resumen / Link to its Summary
AUTORES / AUTHORS: - Beilan J; Lawton A; Hajdenberg J; Rosser CJ
RESUMEN / SUMMARY: - BACKGROUND: Pheochromocytoma (paraganglioma) of the urinary bladder is a rare tumor. Herein we sought to review the contemporary literature on pheochromocytomas of the urinary bladder in order to further illustrate the presentation, treatment options and outcomes of patients diagnosed with these tumors. METHODS: A comprehensive review of the current literature was conducted according to the PRISMA guidelines by accessing the NCBI PubMed database and using the search terms “paraganglioma, pheochromocytoma, bladder.” This search resulted in the identification of 186 articles published between January 1980 and April 2012 of which 80 articles were ultimately included in our analysis. RESULTS: Pheochromocytomas usually occurred in young adult Caucasians (mean age, 43.3 years; range, 11--84 years). According to the literature, the most common
symptoms and signs of pheochromocytomas of the urinary bladder were hypertension, headache, and hematuria. Of the 77 cases that commented on catecholamine production, 65 patients had biochemically functional tumors. Approximately 20% of patients were treated by transurethral resection alone, 70% by partial cystectomy and 10% by radical cystectomy. The 75 patients with follow-up information had a mean follow-up of 35 months. At the time of last follow-up, 15 (14.2%) had disease recurrence, 10 (9.4%) had metastasis, and 65 (61.3%) were alive. CONCLUSIONS: Pheochromocytomas of the urinary bladder tend to be functional and occur mostly in young adult Caucasians. Patients with localized tumors have an extremely favorable prognosis and may be managed by less aggressive modalities, whereas patients with metastatic disease have a significant reduction in survival rates despite aggressive treatment.

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

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- Enlace al texto completo (gratuito o de pago) 1136/bcr-2013-008778

AUTORES / AUTHORS: - Furrukh M; Qureshi A; Saparamadu A; Kumar S
INSTITUCIÓN / INSTITUTION: - Department of Oncology, Sultan Qaboos University Hospital, Muscat, Oman.
RESUMEN / SUMMARY: - A 58-year-old woman presented to a tertiary care centre with signs and symptoms of acute cholecystitis, cholelithiasis and diagnoses of a high-grade neuroendocrine tumour of the gallbladder primarily with peritoneal and liver metastases. She had a liver abscess secondary to Salmonella and Enterococcus fecalis that was drained and treated with appropriate antibiotics. Interestingly, the serum chromogranin A levels were within normal limits, but carcinoembryonic antigen was elevated, which helped evaluate responses and pick progression. She was treated with 10 cycles of palliative chemotherapy when malignancy associated complications started to recur, that is, cholangitis, worsening pain, cachexia, intestinal obstruction, etc leading to chemotherapy delays. Her disease progressed during these times with rapid deterioration of performance status. She died of septic complications postlaparotomy for intestinal obstruction. Her progression-free survival remained for 8 months with subjective and objective improvements, and her overall survival remained at 13 months. We describe the course of her illness and give a brief review of the literature.

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


**AUTORES / AUTHORS:** - Hu J; Wu J; Cai L; Jiang L; Lang Z; Qu G; Liu H; Yao W; Yu G

**INSTITUCIÓN / INSTITUTION:** - Department of Pathology, Affiliated Yantai Yuhuangding Hospital, Medical College of Qingdao University, No,20, Yuhuangding East Road, Yantai 264000, China. caili200403@yahoo.com.cn

**RESUMEN / SUMMARY:** - Composite pheochromocytoma/paraganglioma is a rare tumor with elements of pheochromocytoma/paraganglioma and neurogenic tumor. Most were located in the adrenal glands, and extra-adrenal composite pheochromocytoma is extremely rare. Only 4 cases in the retroperitoneum have been described in the online database PUBMED. Here, we report a case of retroperitoneal extra-adrenal composite pheochromocytoma and review the related literature. VIRTUAL SLIDES: The virtual slide(s) for this article can be found here: http://www.diagnosticpathology.diagnomx.eu/vs/1700539911908679.

[12] **TÍTULO / TITLE:** - Carcinoid tumor of the lung with massive ossification: report of a case showing the evidence of osteomimicry and review of the literature.

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


**AUTORES / AUTHORS:** - Tsubochi H; Endo S; Oda Y; Dobashi Y

**INSTITUCIÓN / INSTITUTION:** - Department of Thoracic Surgery, Jichi Medical University Saitama, Japan.

**RESUMEN / SUMMARY:** - Carcinoid tumor is one of the commonly encountered primary pulmonary neoplasms. Although it has been known to be accompanied by calcification and/or ossification, presentation with a large ossified mass is rare. We describe here the case of a 29-year-old female with the radiological finding of a single bony nodular lesion. Pathological examination of the surgically resected specimen led to the diagnosis of carcinoid tumor of the lung with massive ossification. Although histological features showed the tumor of low grade malignancy, subcarinal and right hilar lymph nodes were found to be positive for metastasis. Further immunohistochemical analysis revealed that the tumor cells expressed the osteogenic inducer protein, bone morphogenic protein-2 [BMP-2] and osteoblastic marker protein, osteocalcin. We interpreted this to mean that the carcinoid tumor cells had acquired an osteoblastic phenotype and had subsequently developed marked intratumoral ossification.
The relevant literature is reviewed and possible mechanisms of tumor-related osteogenesis are discussed.


**Resumen / Summary:** Paraganglioma (also known as extra-adrenal pheochromocytoma) is a rare neuroendocrine neoplasm observed in patients of all ages, with an estimated incidence of 1 per 300,000 population. It has long been recognized that some cases are familial. The majority of these tumors are benign, and the only absolute criterion for malignancy is the presence of metastases at sites where chromaffin tissue is not usually found. Some tumors show gross local invasion and recurrence, which may indeed kill the patient, but this does not necessarily correlate with metastatic potential. Here, we report a case of vertebral metastatic paraganglioma that occurred 19 months after the patient had undergone partial cystectomy for urinary bladder paraganglioma. We believe this to be a rarely reported bone metastasis of paraganglioma arising originally within the urinary bladder. In this report, we also provide a summary of the general characteristics of this disease, together with progress in diagnosis, treatment, and prognosis.

[14] **Título / Title:** Duodenal somatostatinoma: a case report and review of the literature.

**Resumen / Summary:** Introduction: About 70% of well-differentiated endocrine tumors arise from the gastrointestinal tract. Duodenal well-differentiated endocrine tumors account for only 2.6% of all neuroendocrine tumors. Following the first two case reports of somatostatin-secreting tumors in...
1977, fewer than 200 cases of somatostatinoma have been reported. These tumors of the duodenum are usually silent and asymptomatic, but can cause gastrointestinal symptoms. Depending on the localization of the tumor, multiple surgical procedures can be performed, ranging from local resection to pancreaticoduodenectomy. CASE PRESENTATION: Here, we report a case of a submucosal duodenal mass in a 42-year-old Turkish White man presenting with nausea, vomiting, fatigue and abdominal pain. The treatment decision of pancreaticoduodenectomy made preoperatively was later altered to intraoperative removal via local resection with sphincteroplasty. CONCLUSION: Tumors of the periampullary region are considered highly malignant, and the Whipple operation is usually the only procedural treatment. In the current case, we decided not to perform pancreaticoduodenectomy but to excise the mass intraoperatively, and consequently avoided unnecessary resection of the pancreas and anastomosis to undilated hepatic and pancreatic ducts. This protective strategy prevented duodenum- and pancreas-related morbidity.