A total pancreaticoduodenectomy was performed. The pathology revealed 7 grade 2 well-differentiated neuroendocrine tumors (6 in pancreas and 1 in duodenum) and 18 of 22 positive lymph nodes. Post-surgically she experienced multiple admissions due to surgical complications and the expected endocrine and exocrine dysfunction. Eight weeks after surgery gastrin remained elevated [652 pg/ml (13 - 115 pg/ml)]. The surgical management of gastrinomas in MEN1 remains without consensus. Due to the multifocality of gastrinomas associated with MEN1 and the high probability of persistent hypergastrinemia, surgery is not usually recommended. However, most guidelines are in favor of surgical excision for tumors > 2 cm to decrease their associated risk for liver metastasis and improve long-term survival. In this young patient with a 3.2 cm tumor, a surgical approach was favored to improve prognosis. Persistent hypersecretion may occur, as in our patient, due to residual hypertrophy of gastric parietal cells, although the possibility of remaining tumors still exists, especially in view of high lymph node positivity. The best available evidence-based treatment alternatives were offered to our patient; unfortunately, both the disease itself and surgical treatment options impose high morbidity and decreased quality of life. More studies reporting on the long-term outcomes after surgical resection of gastrinomas in MEN1 are needed to identify predictors to help recognize patients which benefit from a surgical approach.

Tumor Biology

ENDOCRINE NEOPLASIA CASE REPORTS

The Diagnosis and Management of a Rare ACTH-Producing Neuroendocrine Tumor Causing Cushing's Syndrome

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A 57-year-old woman presented with a three-month history of worsening lethargy, weight gain, central adiposity, proximal muscle weakness, and hypokalemic hypertension. Initial investigations were consistent with ACTH-dependent Cushing's Syndrome. 24-hour urine cortisol: 711nmol/day (normal<275), cortisol after 1 mg dexamethasone suppression test 960nmol/L (normal<50)), 2-day dexamethasone suppression cortisol: 575 and ACTH: 19 pmol/L. MRI sella was normal. She required several hospital admissions for worsening proximal muscle weakness and falls. She was started on ketoconazole and spironolactone. Basal insulin was started for dysglycemia. Petrosal vein sampling indicated ectopic ACTH production. CT scan of chest/abdomen/pelvis showed no intrathoracic or intra-abdominal abnormality. Thyroid ultrasound and whole-body octreotide scan showed no abnormalities. A Gallium 68-dotatate PET scan showed focal tracer uptake in the pancreatic tail suspicious for a well-differentiated neuroendocrine tumor (NET). MRI pancreas showed a slightly truncated pancreatic tail with no discrete lesion. We proceeded with Endoscopic Ultrasound (EUS) which revealed an irregular hypoechoic mass in the pancreatic tail. EUS–guided fine-needle biopsy (EUS-FNB) using a 22G SharkCore needle, revealed a neuroendocrine neoplasm staining positively for ACTH. She underwent distal pancreatectomy with pathology showing a 1.4cm well-differentiated NET, staining positive for ACTH with a Ki67 index of 10%. Postoperatively, her ketoconazole, spironolactone, and insulin were discontinued. Her dysglycemia, hypertension and other clinical symptoms gradually improved. Immediately post-operatively, her AM cortisol was 403 and her 24-hour urine cortisol was elevated 959nmol/day. Repeat AM cortisol and 24-hour urine cortisol one-month post-surgery were 392 nmol/L and 72 nmol/day respectively, confirming biochemical remission. **Discussion:** ACTH-producing pancreatic NETs are rare, accounting for less than 15% of reported cases of EAS. They are often aggressive, with 78.7% of reported cases having distant metastasis at diagnosis (1). They are associated with significant morbidity and mortality. Diagnosis and management can be difficult and require multiple investigations and multidisciplinary team involvement. In our case, we utilized endoscopic ultrasound to ensure a correct preoperative localization. We discuss the systematic approach for diagnosis and the management of an ACTH-producing pancreatic NET. **References:** 1. Maragliano R, Vanoli A, Albarello L, Milione M, Basturk O, et al. ACTH-secreting pancreatic neoplasms associated with Cushing syndrome: clinicopathologic study of 11 cases and review of the literature. Am J Surg Pathol. 2015 Mar;39(3):374-82. doi: 10.1097/PAS.0000000000000340. PMID: 25353285.

Tumor Biology

ENDOCRINE NEOPLASIA CASE REPORTS

Thymoma and Not Just Thymic Carcinoid Can Be Associated With Multiple Endocrine Neoplasia Type 1

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Background: Multiple endocrine neoplasia type 1 (MEN1) is an inherited tumor syndrome with autosomal dominant inheritance. Thymic neuroendocrine tumors (NETs) are known manifestations of MEN1 occurring in 2-8% of patients. However, thymomas, a type of thymic epithelial tumors, have only been described in rare case reports. They markedly differ from thymic NETs in their natural history, morphology, prognosis, and therapeutic options. Here we present a case of an aggressive, recurrent thymoma associated with MEN1.

Case Report: A 58-year-old Caucasian female with a family history of MEN1 was diagnosed with a prolactinoma at age 15 when she presented with irregular menses, galactorrhea, headaches and visual field defects. She was referred to our institution for further evaluation where genetic testing confirmed the diagnosis of MEN1. Throughout the years, she