Title

A Rare Case of Chronic Diarrhea in a Pediatric Patient

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monitoring, which precluded him from undergoing CT with IV contrast or MRI studies. Abdominal computed tomography with oral contrast did not reveal any lesion. In an attempt to localize the suspected Insulinoma, he had celiac angiogram and portal vein sampling, which were inconclusive. He had a PET scan with dotatate that showed increased amount activity in the uncinate process of the pancreas. He was diagnosed with insulinoma and placed on monthly lanreotide injections, however continued to have severe hypoglycemia episodes. Due to persistent refractory hypoglycemia, patient underwent distal pancreatectomy and splenectomy and the histological findings were consistent with nesidioblastosis. Patient developed hyperglycemia in the post-operative course which was controlled with a carb consistent diet. This case demonstrates that differentiation between insulinoma and nesidioblastosis is very challenging, and in most cases, the diagnosis is made post operatively based on histologic findings.

Tumor Biology
ENDOCRINE NEOPLASIA CASE REPORTS

A Rare Case of Chronic Diarrhea in a Pediatric Patient

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Background: VIPoma, also known as Watery Diarrhea-Hypokalemia- achlorhydria (WDHA) Syndrome is a rare manifestation of multiple endocrine neoplasia syndrome type 1 (MEN1). Vasoactive intestinal peptide, part of the secretin-glucagon family, may be overexcreted in tumors associated with MEN1 and results in diarrhea that persists while fasting, resulting in massive secretion of water and electrolytes. First-line treatment is surgical resection.
Clinical Case: We present a 13-year-old male with a past medical history of chronic diarrhea for four years who was transferred from an outside hospital for severe diarrhea and associated electrolyte derangements, including hypokalemia of <1.0 mmol/L (3.5-5.0 mmol/L), sodium of 120 (135-145 mmol/L), and chloride of 84 mmol/L (101-110 mmol/L). Family history was significant for pancreatic, breast, thyroid, stomach, parathyroid, and uterine cancer, as well as hyperparathyroidism and nephrolithiasis. The patient had been admitted to the hospital before for a similar episode of acute on chronic diarrhea but was treated for infectious diarrhea during that admission. CT enterography was obtained during this hospitalization and it revealed multiple solid and heterogeneous appearing pancreatic masses in the head and tail of the pancreas. Work-up was significant for pancreatic polypeptide of 1,523 pg/mL (92-752 pg/mL) and vasoactive intestinal peptide of 1,105 pg/mL (<75 pg/mL). Pancreatic biopsy revealed a grade 2 pancreatic neuroendocrine tumor. Genetic testing revealed a known pathogenic mutation in the menin gene p.R526 (c. 1579>T, pArg527). A pylorus-preserving total pancreatectomy, duodenectomy, cholecystectomy, and splenectomy was performed and surgical pathology revealed a well-differentiated grade 1 neuroendocrine tumor in the head of the pancreas and a well-differentiated grade 2 neuroendocrine tumor in the tail of the pancreas. Since the surgery, the patient currently has no signs of other neuroendocrine tumors associated with MEN1 but continues to follow-up for regular screening for other tumors associated with MEN1.
Clinical Lessons: 1. VIPoma, also known as Watery Diarrhea- Hypokalemia- Achlorhydria Syndrome, is characterized by secretory diarrhea that persists while fasting. 2. VIPoma should be considered in patients with a history of chronic diarrhea and a significant family history of neuroendocrine tumors.

Tumor Biology
ENDOCRINE NEOPLASIA CASE REPORTS

A Rare Presentation of Multiple Endocrine Neoplasia Type 1

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Background: Multiple endocrine neoplasia type 1 (MEN1) is a rare, autosomal dominant inherited syndrome caused by mutations in the MEN1 tumor suppressor gene with a reported incidence of 2 in 100,000.
Clinical Case: A 26-year-old Caucasian female was seen for surveillance screening given positive familial mutation in the MEN1 gene. She had a significant family history of pathogenic MEN1 in her son, brother, father and paternal aunt. On presentation, she denied any history of headache, nipple discharge, kidney stones, fractures, heart burn, abdominal pain, diarrhea, hypoglycemia, flushing, lightheadedness. She was on hormonal IUD for contraception and had not had menstrual cycles for the last 6 months. Vital signs and physical examination were unremarkable. Her initial evaluation included a normal calcium of 9.8mg/dL (RR:8.6-10.6mg/dL), PTH of 70pg/ml (RR:12-88pg/ml), 25OH-vitamin-D of 11ng/ml (RR:20-80ng/ml). She had normal gastrin, chromogranin A, glucagon, and vasoactive intestinal peptide levels. Prolactin was 17.6ng/ml (RR:3.3-26.7 ng/ml). MRI pituitary showed a 6mm lesion representing a Rathke's cleft cyst, not compressing surrounding structures.
Two months following the initial encounter, she presented with abdominal pain, nausea, vomiting. She was found to have an obstructing renal stone requiring stent placement. Calcium was 10.1mg/dl. She was also noted to have an incidental pancreatic tail mass of 4.2cm on CT abdomen. She underwent laparoscopic robotic distal spleno-pancreatectomy. Surgical pathology showed a well differentiated 3.5cm neuroendocrine tumor with negative margins, which stained positively for insulin. She had not reported symptoms of weight gain or any episodes suspicious for sympathoadrenal activation or neuroglycopenia, and was again confirmed after surgery.
Patient was re-admitted to the hospital for right-sided flank pain and was found to have a new 6mm obstructing calculus with moderate hydronephrosis of the right kidney. Corrected calcium level was found to be mildly elevated at 11.1mg/dL. Repeat PTH was stable at 81pg/ml, and urinary calcium was elevated at 447.3mg/24h (RR:50-250mg/24h).