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Neuroendocrine tumor of the pancreas causing biliary obstruction in a 12 year-old girl: A case report and literature review☆

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ABSTRACT

Pancreatic tumors are uncommon in children and rarely result in biliary obstruction. A previously well 12-year old female presented with a one-week history of fatigue, pruritis, and painless jaundice. Abdominal ultrasound demonstrated a mass in the pancreatic head associated with dilation of the common bile duct. Further workup included abdominal MRI, CT and endoscopic retrograde pancreaticogram (ERCP) with biliary stenting. Octreotide scan did not reveal uptake in the pancreatic tumor. Percutaneous biopsies were consistent with a grade 2 pancreatic neuroendocrine tumor (NET). Preoperative imaging demonstrated involvement of the portal vein. The patient was brought the operating room for a pancreaticoduodenectomy and portal vein resection. Final pathology revealed a T3N1M0 pancreatic NET. The patient recovered uneventfully.

Pancreatic tumors are exceedingly rare in children and adolescents. Neuroendocrine tumors (NET) account for approximately 25% of pediatric pancreatic masses [1,2]. NET arise from endocrine tissues of the pancreas and are classified as either non-functioning or functioning. Functioning NETs secrete various peptide hormones, which act at distant sites and can result in a variety of clinical syndromes. Pancreatic NETs can be benign or malignant, however the majority of non-functioning tumors are malignant. Pancreatic tumors in children rarely present with biliary obstruction, owing to the expansive rather than infiltrative growth patterns of the common pediatric pancreatic neoplasms [3,4].

1. Case report

A 12 year-old female was referred to our center after presenting to her local emergency department with a one-week history of fatigue, pruritis, and acholic stools. She was visibly jaundiced. An abdominal ultrasound at her local hospital suggested a mass within the head of the pancreas. Her laboratory tests were: total bilirubin 136 umol/L, direct bilirubin 84.6 umol/L, alkaline phosphatase 543 U/L, alanine aminotransferase 172 U/L, and aspartate aminotransferase 75 U/L. Serum lipase was within normal limits. Following these investigations, the patient was transferred to our institution for further workup in hospital.

Magnetic resonance imaging (MRI) of the abdomen demonstrated extensive dilation throughout the intra and extrahepatic biliary tree (Fig. 1). The common bile duct (CBD) measured 2.5 cm with an abrupt transition at the pancreatic head. Within the pancreatic head there was a 3 cm solid, smoothly marginated mass. Computed tomography (CT) of the abdomen (Fig. 2) was also performed to better assess the surrounding vasculature. The lesion was found to be in direct contact with the portal vein (PV) with a narrowed segment, suspicious for tumor infiltration. Both the superior mesenteric artery (SMA) and superior mesenteric vein (SMV) appeared to be uninvolved. There also appeared to be 5 × 3 mm calculus within the CBD. Because of the biliary obstruction and possible CBD stone, the patient underwent endoscopic retrograde pancreaticochoangiogram (ERCP). This identified a tight stricture, 1 cm from the ampulla, for a distance of 5 mm. No stone was identified and a plastic stent was placed for biliary drainage. ERCP

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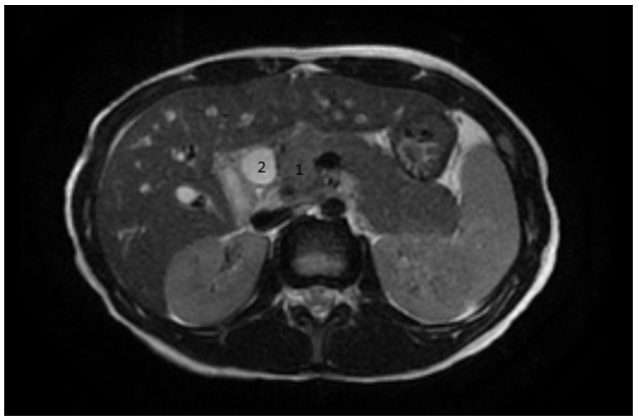

Fig. 1. T2-weighted magnetic resonance imaging (MRI) demonstrating a mass in head of pancreas (1) causing massive dilatation of the common bile duct (2).

was arranged prior to obtaining a tissue diagnosis because the patient was highly symptomatic with pruritis.

Tumor markers were sent in an attempt to determine the etiology of this mass. Serum carbohydrate antigen 19-9 (CA19-9) was mildly elevated at 38 U/ml. Serum carbohydrate antigen 125 (CA-125), β-human chorionic gonadotropin (β-HCG), alpha-fetoprotein (AFP) and Immunoglobulin 4 (IgG4) levels were within normal limits. Furthermore, serum gastrin, insulin and chromogranin A were also non-elevated. Twenty-four hour urine collection revealed normal levels of vanillylmandelic acid (VMA), homovanillic acid (HVA), and 5-hydroxyindole acetic acid (5-HIAA). A bone marrow aspirate was also normal, making the diagnosis of leukemia unlikely. CT chest was also normal.

This patient's case was then reviewed in multi-disciplinary tumor board rounds with radiology, pediatric medical oncology, pathology, and pediatric surgery. A joint decision was made to proceed with a surgical biopsy, as the group felt that percutaneous biopsies were less likely to result in adequate tissue for pathologic interpretation and had a theoretical potential for seeding the tumor through the needle tract. She was brought to the operating room for laparoscopic biopsies of the pancreatic head. Tissue samples sent to pathology for frozen sections revealed only normal pancreatic tissue. As a result, interventional radiology was consulted to perform ultrasound-guided, percutaneous biopsies while the patient was under the same general anesthetic. These specimens demonstrated a grade 2 NET (2 mitoses per 10 high power fields, Ki-67 index 12%). Unfortunately, the patient bled post-biopsy and underwent angiographic embolization of

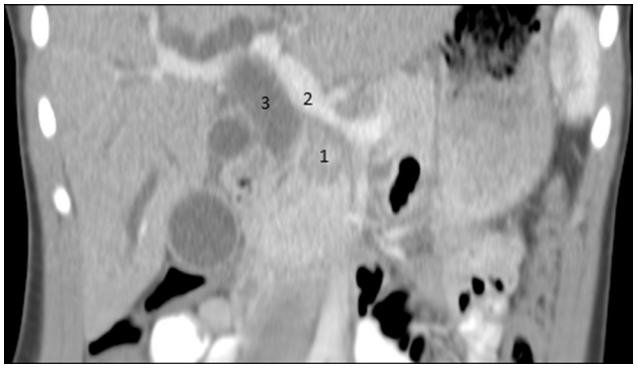

Fig. 2. Coronal slice from computed tomography (CT) scan of the abdomen: pancreatic mass (1), portal vein (2), and dilated common bile duct (3).

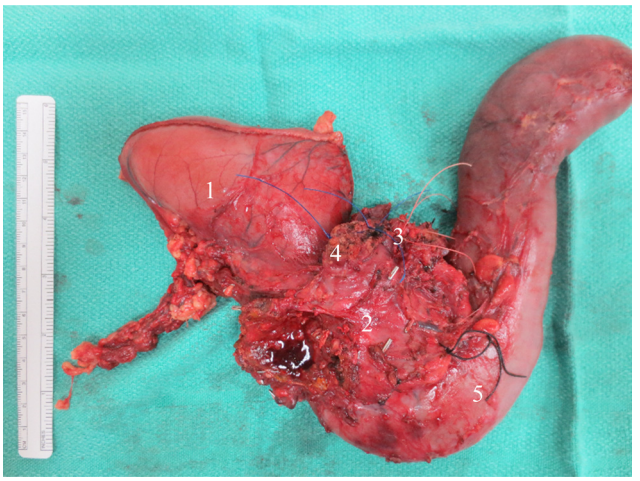

Fig. 3. Posterior view of specimen: gastric antrum (1), pancreatic tumor (2), bile duct (3), pancreatic neck margin (4), and duodenum (5).

branches of the pancreaticoduodenal artery. She tolerated this well and eventually recovered.

An octreotide scintigraphic scan was then performed. This revealed no significant octreotide uptake within the known pancreatic tumor, and no evidence of octreotide-positive metastatic disease. Although not available in our center, but commonly utilized in Europe, a 68Ga DOTATOC/68Ga DOTATATE PET scan would have been a good alternative imaging test for preoperative evaluation. If positive, this would have provided an imaging modality for postoperative surveillance.

The patient was then reviewed in multi-disciplinary neuroendocrine tumor board rounds. In conjunction with our adult hepatopancreaticobiliary surgeons, a decision was made to take the patient to the operating room for a pancreaticoduodenectomy (Whipple's procedure) with a plan for possible PV resection. The mass was visible in the pancreatic head, with firm nodes along the uncinate margin. The nodal tissue along the hepatic artery and posterior to the SMA was cleared and sent with the specimen. She underwent a standard pancreaticoduodenectomy with an end-to-side pancreaticojejunostomy, end-to-side hepaticojejunostomy, and a loop gastrojejunostomy. A frozen section of the pancreatic resection margin was sent to pathology before reconstruction, which was negative for malignancy (Fig. 3). A short segment (5 mm) of the PV was resected, due to significant tumor adherence, and reconstructed using an end-to-end anastomosis (Fig. 4). Total clamp time was 25 min. We elected to perform a primary anastamosis of the PV rather than a vein graft due to the short segment of resection, and the fact that it came together under no tension. On final pathology the PV was not infiltrated with tumor and therefore the dense adherence was likely desmoplastic reaction.

Final pathology demonstrated a T3N1M0 grade 2 (World Health Organization Classification) pancreatic NET with negative resection margins. The tumor extended beyond the pancreas but did not involve the celiac axis or SMA. Lymphovascular and perineural invasion were present. The Ki-67 index ranged from 3% to 20%. Five out of nineteen lymph nodes were positive. She was again presented at our neuroendocrine tumor board. Given her complete resection and the non-functionality of her tumor, no further treatment was recommended. Adjuvant treatment of completely resected (R0) pancreatic neuroendocrine tumors is somewhat controversial, however after consultation with international experts and a through literature review, it was felt that the potential