Multicystic adenomatoid hamartoma of the pancreas

Brian D. Hosfield\(^a\), Britney L. Grayson\(^a\), Attila Nakeeb\(^b\), Eric A. Albright\(^b\), Troy A. Markel\(^a\)*

\(^a\) Department of Surgery, Indiana University School of Medicine, Indianapolis, IN, USA
\(^b\) Department of Pathology, Indiana University School of Medicine, Indianapolis, IN, USA

**Abstract**

Multicystic adenomatoid hamartoma is an extremely rare tumor of the pancreas, with only 4 other cases reported in the literature. We report a case of a 4-year old boy who presented with an 8 month history of abdominal pain, steatorrhea, and failure to thrive. Work-up showed severe pancreatic insufficiency and a large, multiseptated, cystic mass originating from the head of the pancreas and compressing the duodenum. The child underwent a classic pancreaticoduodenectomy with portal vein reconstruction. He tolerated the procedure well and has been seen in follow-up.

**1. Introduction**

Tumors of the pancreas are uncommon in children [1]. A review conducted by our own institution over a 20-year period included 13 cases total. Benign tumors included insulinoma and mucinous cystadenoma. Malignant tumors included pancreatic ductal adenocarcinoma, acinar tumor, papillary solid tumor, pancreatoblastoma, and rhabdomyosarcoma [2]. We report a case of a 4-year old boy with a multicystic adenomatoid pancreatic hamartoma, an extremely rare tumor with only 4 other pediatric cases reported in the literature [3–6].

**2. Case presentation**

A 4-year old boy presented with an 8-month history of abdominal pain, steatorrhea, hemaatochezia, and failure to thrive. He had no significant past medical or surgical history. There was no family history of pancreatic masses or other gastrointestinal malignancies. Labs were notable for both iron deficiency anemia and vitamin E deficiency. Stool analysis showed an increase in fecal fat and low fecal elastase. A Meckel’s scan was performed and was negative. He underwent an EGD for pancreatic enzyme stimulation testing which showed a large, ulcerated, hypervascular mass in the second portion of the duodenum (Fig. 1). Biopsy was not performed due to hypervascularity and concern for hemorrhage. Computed tomography of the abdomen and pelvis demonstrated a large, multicystic, multiseptated mass in the right upper quadrant (Fig. 2a) with marked pancreatic atrophy (Fig. 2b). Magnetic resonance cholangiopancreatography (MRCP) showed an 8.8 cm mass originating in the head of the pancreas with associated mass effect on the duodenum (Fig. 3). Laboratory findings showed normal liver and renal function. Pancreatic enzymes were normal. AFP, beta-HCG, and gastrin were within normal limits. CA 19–9 and uric acid were mildly elevated (58 Units/L, 302 Units/L, respectively).

Given the obstructing nature of the mass and severe pancreatic atrophy, he was scheduled for a classic pancreaticoduodenectomy. Provisional diagnoses included solid pseudopapillary tumor, pancreatoblastoma, and intraductal papillary mucinous neoplasm. At laparotomy, we noted a large cystic mass in the head of the pancreas without evidence of metastatic disease. There were large lymph nodes in the porta hepatitis and these were resected. The common bile duct was de-compressed and adherent to the portal vein. In dissecting the duct, an injury to the portal vein occurred which necessitated portal vein reconstruction. The anastomoses was performed in an end-to-end fashion with multiple 6–0 prolene sutures. After resection of the specimen, reconstruction was completed via a retrocolic end-to-side gastrojejunostomy, an end-to-side hepaticojejunostomy, and a two-layer pancreaticojejunostomy (Fig. 4).

Two 10 mm drains were placed. The patient tolerated the procedure well and was admitted to the ICU post-operatively for hemodynamic monitoring. His post-operative course was marked by slow return of bowel function, and erythromycin was started. He was discharged on post-op day 11. He completed his 1 month post-operative follow-up and is doing well.

The specimen was assessed by 3 different institutions. The lesion appeared to originate in the head of the pancreas and measured 9.5 × 8.5 × 8.5 cm and severely compressed the duodenum (Fig. 5). The multiloculated cystic structure contained clear mucinous fluid with a clear lining. There was an anomalous junction between the common bile duct and the pancreatobiliary ductal system. Residual pancreatic...
tissue showed atrophy from obstruction. Histopathologic analysis showed a benign appearing lesion with a mixture of bland epithelium with gastric and biliary differentiation with dense stroma (Fig. 6). All lymph nodes were negative for tumor.

3. Discussion

Due to the rarity of pancreatic tumors in children, there is little available data in the literature and developing an evidence-based approach to treatment remains challenging. Incidence of pediatric pancreatic tumors is estimated at 0.018 per 100,000 [7]. The differential for pediatric pancreatic tumors is broad and is comprised of epithelial tumors, non-epithelial tumors, pancreatoblastomas, endocrine tumors, solid pseudopapillary (Frantz) tumors, acinar cell carcinomas, sarcomas, and lymphomas [8]. Symptoms can vary based on the size and location of the tumor. Given the rare nature of these tumors and their significant heterogeneity in children, our understanding remains poor and there has been significant effort to develop prospective studies to better understand this disease process [9].

Four additional cases of multicystic adenomatoid hamartoma of the pancreas are reported in the literature [3–6]. Variations in patient presentation, tumor size and location are summarized in Table 1. Three out of five cases were male, two were female. Age ranged from 34 weeks to 4 years. Symptoms varied and included abdominal pain, distention, and steatorrhea. While other pancreatic tumors are associated with genetic syndromes such as Beckwith-Wiedemann syndrome [10], none of the patients were known to have a hereditary syndrome. Two out of the five patients had laboratory values consistent with pancreatitis. Our patient had a mildly elevated CA 19–9, other tumor markers were normal. It is not clear if elevated CA 19–9 is associated with multicystic adenomatoid hamartoma, as markers were only available in one other case report and were normal. Size of the tumor varied from 3 to 14 cm. Three out of the five tumors were located in the head of the pancreas, one was located in the tail, and the remaining tumor was diffusely present throughout the pancreas. Three of the five patients underwent a Whipple operation, while the remaining underwent local resection. While adjuvant therapy is often required for many pancreatic tumors such as pancreatoblastomas [11,12], the reported patients with

![Fig. 1. EGD demonstrating ulcerated and hypervascular mass invading second portion of duodenum.](image1)

![Fig. 2. (a) CT Abdomen and Pelvis axial demonstrating large multiseptated cystic mass in the right-upper quadrant. (b) coronal section showing accompanying atrophy in the tail of the pancreas.](image2)

![Fig. 3. Abdominal MRCP T2 weighted image demonstrating large cystic mass with compression of duodenum, which is seen wrapping around the mass. Pancreatic duct is not visualized.](image3)
multicystic adenomatoid hamartoma did not undergo any type of adjuvant chemotherapy or radiation. Only one patient did not survive (34 week old female) who had a prolonged hospital course due to pulmonary disease, metabolic issues, and hepatic dysfunction. The patient ultimately passed away 3 months after surgery.

The most distinguishing feature of our case was the significant amount of time from development of symptoms to diagnosis. The child first presented with encopresis and steatorrhea. This was likely due to both the large size of the tumor and the location in the head of the pancreas, causing pancreatic atrophy and resultant exocrine insufficiency. He remains on pancreatic enzymes at this time. Fortunately there was enough remnant gland remaining after resection, and the child does not currently have any needed for insulin or evidence of endocrine insufficiency.

4. Conclusion

Multicystic adenomatoid pancreatic hamartoma is an extremely rare benign tumor and should be in the differential diagnosis of cystic pancreatic masses in children. Patients can present with abdominal

Fig. 4. Illustration demonstrating classic pancreaticoduodenectomy before and after specimen removal.

Fig. 5. Gross specimen showing multiseptated, multicystic mass compressing the duodenum.

Fig. 6. Left: Low-power field demonstrating large cysts lined with ductal epithelium. Right: Low-power field showing differentiated gastric epithelium surrounded by dense stroma (hematoxylin and eosin x10).
pain, pancreatic exocrine insufficiency, and pancreatitis. Treatment of choice includes surgical resection, with tumor location determining type of surgery performed.

Patient consent

Consent to publish the case report was not obtained because this report does not contain any personal information that could lead to the identification of the patient.

Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

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Conflicts of interest

The authors declare that there are no conflicts of interest regarding the publication of this paper.

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Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.epsc.2019.101258.

References