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Solid pseudopapillary neoplasm of the pancreas in pediatric patients: A case report and institutional case series

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ABSTRACT

Solid pseudopapillary neoplasm (SPN) of the pancreas is a rare tumor presenting in adolescent and young adult females. A previously healthy 13 year-old female presented to our institution with abdominal pain and emesis. Imaging revealed a pancreatic cystic mass. Endoscopic ultrasound (EUS) with fine needle biopsy suggested SPN. Pathologic evaluation following resection revealed immunohistochemical (IHC) staining positive for β-catenin and α-1-antitrypsin despite extensive necrosis. We discuss this patient as well as our institutional series of SPN of the pancreas, describing the evaluation, management, and histopathology of this rare tumor.

Solid pseudopapillary neoplasm (SPN) of the pancreas is an uncommon tumor of the exocrine pancreas, occurring predominantly in young females [1,2]. Unlike other forms of pancreatic neoplasm, SPN of the pancreas demonstrates an excellent long-term prognosis with adequate resection [3,4]. With accurate preoperative diagnosis, surgical planning and intraoperative decision making are enhanced.

We present a case of SPN of the pancreas in which the diagnosis was suggested after fine-needle aspiration cytology on endoscopic ultrasound. Definitive diagnosis was made from immunohistochemical (IHC) staining, which allowed for appropriate surgical planning and adequate resection. Without IHC staining, the extensive necrosis on endoscopic biopsy would have resulted in an otherwise indeterminate diagnosis, demonstrating the importance of this histologic evaluation. We also present a single-institutional case series of patients undergoing resection for SPN of the pancreas, revealing additional pediatric patients for whom immunochemistry was valuable in establishing their diagnosis.

1. Case report

A 13-year old previously healthy female presented to our emergency department with a three day history of abdominal pain, emesis, and malaise. Physical exam revealed mild right-sided abdominal tenderness. A limited right lower quadrant abdominal ultrasound was unremarkable, therefore a computed tomography (CT) scan was obtained demonstrating a predominantly cystic structure arising from the duodenum and abutting the pancreatic head, without biliary or pancreatic ductal dilation (Fig. 1).

Carcinoembryonic antigen (CEA) and cancer antigen (CA) 19-9 were not elevated. Magnetic resonance imaging (MRI) further characterized the lesion as a peripherally enhancing round lesion insinuated between the pancreatic head and second portion of the duodenum, (Fig. 2) suggestive of either a duplication cyst with internal hemorrhage or a solid pancreatic head mass. She then underwent endoscopic ultrasonography (EUS) which revealed that this cystic mass was heterogenous in nature and without evidence of internal vascular flow (Fig. 3). Fine needle aspiration revealed monomorphic necrotic cells

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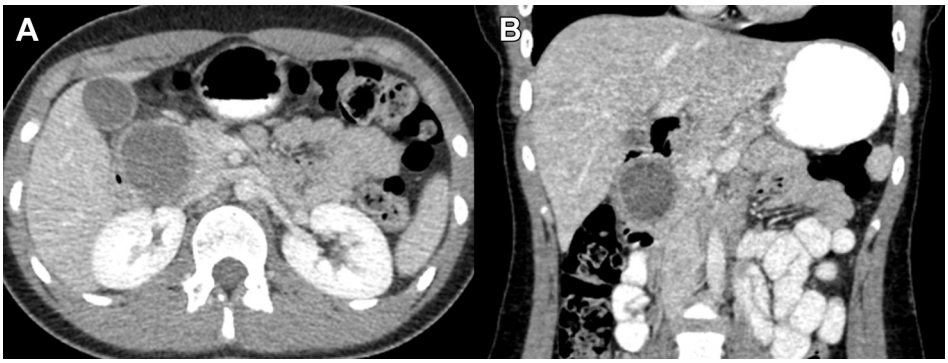

Fig. 1. Axial (A) and coronal (B) computed tomographic images of the intra-abdominal cystic mass at diagnosis.

that on immunohistochemistry were positive for vimentin and β-catenin and weakly reactive for synaptophysin and chromogranin.

Given these findings, the patient underwent exploratory laparotomy via a bilateral subcostal incision. A 3 cm mass arising from the head of the pancreas and adherent to the second portion of the duodenum was identified (Fig. 4). A pancreaticoduodenectomy was performed for complete resection. The pancreatic duct was not dilated, measuring approximately 2–3 mm. Visceral reconstruction was achieved with hepaticojejunostomy, pancreaticojejunostomy, and gastrojejunostomy.

On pathological examination, the mass was an encapsulated $3 \times 3 \times 2.5$ cm lesion arising from the head of the pancreas. The cut surface showed yellow brown necrotic tissue with gritty yellow calcified material. Microscopically, the lesion had a large amount of necrosis with focal areas of neoplastic cells, characterized by small to medium sized cells mostly in sheets. Within the necrotic areas, ghosts of papilliform structures were present (Fig. 5). On IHC, the tumor was positive for β-catenin and α-1-antitrypsin. Testing for CD10 was indeterminate, and the tumor was not tested for e-cadherin.

The patient's postoperative recovery was complicated by prolonged gastroparesis. She received total parenteral nutrition for two weeks and subsequent nasojejunal feeding which was discontinued once she tolerated oral intake. This ultimately resolved and her postoperative course was otherwise uncomplicated. Final histopathology demonstrated this lesion to be a solid pseudopapillary neoplasm of the pancreas with extensive necrosis and negative margins. Follow-up imaging has demonstrated no evidence of recurrence at 1 year from resection. Given complete resection, adjuvant therapy was not indicated.

Following this case, a review of medical and pathological records at our institution, revealed eight patients between January 1995 and December 2014 who were treated for SPN of the pancreas (Table 1). Three of the patients demonstrated intralesional necrosis on histopathology. All eight patients were female and none required adjuvant therapy following resection. With a median follow up time of 5 years (range 1–5 years), there were no recurrences. There was one death that was unrelated to the diagnosis or treatment of SPN of the pancreas.

2. Discussion

SPNs of the pancreas are neoplasms of low malignant potential found predominantly in young females [1,2]. They account for 1–2% of all exocrine pancreatic tumors [5], but 52–71% of pancreatic tumors in children and adolescents [6,7]. The etiology is unknown but suspected to be genetically distinct from pancreatic ductal neoplasms [2]. Despite their preferential association with young women, there are no reports suggesting an association with endocrine disturbances [8]. These tumors do not demonstrate a preferential localization within the pancreas. These neoplasms are often found incidentally on routine physical examination or in patients who present with abdominal pain [9,10]. As in our patient and our institutional review, they are not often associated with pancreatic ductal dilation [11,12].

The differential diagnosis for SPN includes pancreatic pseudocysts, duplication cysts, and other neoplasms of the pancreas including lymphoma and cystic neoplasms [13,14]. Pathology often reveals lobulated solid areas with zones of hemorrhage and necrosis, and cystic spaces filled with necrotic debris [9,15]. IHC is useful for diagnosis of SPN of the pancreas [9]. Positive markers for α-1-antitrypsin, α-1-antichymotrypsin, phospholipase A2, CD 10, and CD 56 are suggestive of pancreatic lesions [16]. A combination

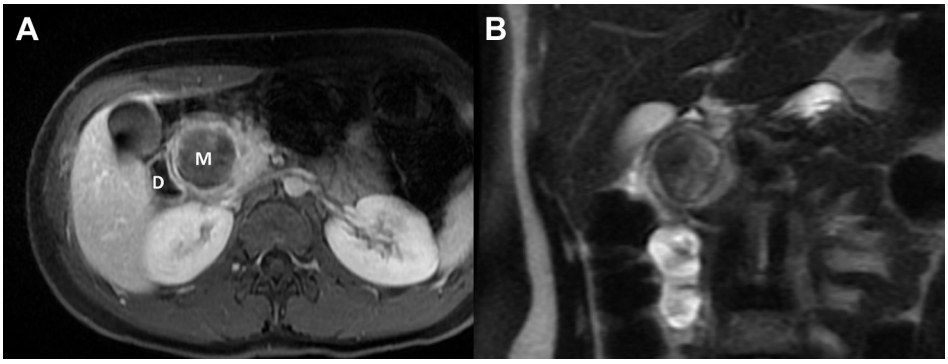

Fig. 2. A) Axial T2-weighted image of the cystic mass (M) with delineation of the plane between the mass and the duodenum (D). B) Coronal T2-weighted image of the mass, demonstrating its heterogeneity and encapsulation.