

Solid Pseudopapillary Tumor of the Pancreas Presenting as an Inflammatory Mass in an 11-Year-Old Child

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Background	An 11-year-old prepubertal girl presented with a systemic inflammatory response and was found to have a solid pseudopapillary tumor of the pancreatic head. She underwent successful pylorus-sparing pancreaticoduodenectomy (PD).
Summary	PD is a very uncommon surgical procedure in children largely because pancreatic masses are extremely rare at an early age. Our patient was a prepubertal 11-year-old girl who presented with a systemic inflammatory response and found to have a pancreatic mass on imaging. The mass was initially believed to be inflammatory, but biopsy confirmed a solid pseudopapillary tumor. She successfully underwent pylorus preserving PD for removal of her tumor. Our case report underscores the variability in the presentation of this mass in pediatric patients and expands the literature on this rare malignancy in children. It additionally confirms that PD is a safe and curative procedure for these patients.
Conclusion	Solid pseudopapillary tumors are rare pancreatic malignancies, especially in the pediatric population. We present a case of a young girl diagnosed with this tumor in the pancreatic head who underwent successful surgical resection. It highlights the complexity of this rare tumor and the role of surgical cure.
Keywords	Solid pseudopapillary tumor, pancreaticoduodenectomy

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Case Description

Despite being the established approach for most pancreatic tumors of the head for adults, pancreaticoduodenectomy (PD) remains an uncommon surgical procedure for pediatric patients. This is largely due to the fact that pancreatic masses are extremely rare at an early age, with an estimated age-adjusted annual incidence of pancreatic tumors in 0- to 19-year olds of 0.191 per million population.^{1,2} We herein present a case of an 11-year-old girl with solid pseudopapillary tumor (SPT) of the pancreatic head who successfully underwent pylorus preserving PD for removal of her tumor.

An 11-year-old female, with a medical history significant for appendectomy due to perforated appendicitis, presented with persistent, generalized abdominal pain, distention, and emesis. She was febrile and tachycardic, and she had severe mid-abdominal tenderness and a leukocytosis of 12,000.

CT imaging revealed a 5.7 x 5.4 cm peripherally-enhancing pancreatic head mass, with central hypodensity and mass effect to adjacent duodenum (Figure 1).



Figure 1. A: Axial and B: coronal computed tomographic (CT) images of the intra-abdominal mass with adjacent inflammatory changes.

The CT scan also showed fat stranding and edema surrounding the mass, consistent with marked inflammatory changes. Possible etiologies included malignancy, abscess, infected choledochocoele, or infected pancreatic pseudocyst. She subsequently underwent magnetic resonance cholangiopancreatography (MRCP), which revealed a complex cystic mass extending from the pancreatic head, measuring 5.7 cm with internal bright heterogeneous material, peripheral rim enhancement and significant retroperitoneal edema. Endoscopic retrograde cholangiopancreatography (ERCP) with fine needle aspiration (FNA) confirmed the diagnosis of SPT of the pancreas.

Based on the prominent inflammatory changes on cross-sectional imaging, as well as her profound systemic inflammatory response, the patient was treated with two weeks of antibiotics and surgical management was deferred for six weeks. Four weeks after initial presentation, repeat magnetic resonance imaging (MRI) showed the surrounding fat stranding, edema resolution, and a decreased mass (from 5.7 cm to 4.2 cm). After medical optimization, the patient underwent a pylorus-preserving PD (Figure 2).



Figure 2. Gross specimen of pylorus-sparing pancreaticoduodenectomy with a mass in pancreatic head.

She tolerated the procedure well and was discharged home after seven days following an uncomplicated postoperative course. Final pathology confirmed the diagnosis of a non-invasive SPT with negative resection margins. The patient recovered without complication and will undergo surveillance imaging six months after surgery.

Discussion

Due to the rareness of pancreatic tumors in the pediatric population, there is a paucity of data on the topic. Most understanding of these tumors comes from case studies from large tertiary institutions, which pool data on all types of pediatric pancreatic masses. Our case features a previously healthy young girl who presented with severe abdominal pain, a profound systemic inflammatory response, and an inflammatory mass within the pancreatic head. While infectious etiologies were considered, FNA of the mass confirmed SPT of the pancreas.

SPTs account for approximately 2 to 3 percent of all pancreatic tumors, regardless of age.³ Though pancreatic neoplasms are exceedingly rare in children, SPTs are one of the most common histologies of such lesions, accounting for 8 to 44 percent.³ SPTs occur most commonly during the third decade of life, but up to 22 percent are noted to occur in individuals <19 years old.⁴ They disproportionately affect women at an overall ratio of 10:1, though this gender discrepancy is less prominent in children.^{3,5} With regards to pediatric patients, the three largest series of SPTs report a median age at presentation of 13–15 years.^{3,6,7} Though no series includes demographic information on pubertal status, it is reasonable to deduce that such a tumor is exceedingly rare in prepubertal patients such as ours.

This patient's presentation of abdominal pain and emesis is consistent with prior literature, which reports abdominal symptoms to be the most common presenting complaints (67–73 percent).^{3,7–9} Although our patient did not have a palpable mass on exam, some series indicate that children are more likely than adults to present with a palpable abdominal mass.¹⁰ Fever, which was present in our patient, is a fairly uncommon presenting symptom (4–17 percent).^{8,9} Further, our literature review suggests that our patient represents the first reported case of a SPT presenting as an inflammatory mass in a child.

The etiology of our patient's mass was initially thought to be infectious given her clinical presentation, which ultimately provoked an ERCP with FNA to obtain a definitive diagnosis. However, imaging is frequently sufficient for diagnosis in many patients, making further preoperative workup unnecessary. Pediatric cases series have reported rates of preoperative FNA as low as 17 percent.⁶ Though the radiologic features of this tumor have been extensively described, findings may be elusive, and further diagnostics may be required.

PD is considered the mainstay of surgical treatment for tumors of the pancreatic head; however, it is rarely performed in children. Mortality and morbidity rates following PD in adult patients range from 1 to 3 percent and 30 to 40 percent, respectively.^{11,12} Similarly, morbidity rates following PD in children are high, ranging from 29 to 50 percent. Reported complications include intraabdominal abscess, pancreatic fistula, pancreatic leak, and chyle leak.^{9,11–14} Nevertheless, PD has proven to be safe for children in several series, including those as young as 3–4 years

old, with SPT appearing to be one of the more common indications for such treatment.^{3,6–8,12–14} Our patient is one of the youngest reported to undergo PD for a pancreatic SPT and possibly the youngest to undergo PD for SPT in the United States. The postoperative course was uncomplicated, and she remains without issue after five months of follow-up.

Our patient underwent a pylorus-sparing PD, consistent with prior literature on this topic.^{9,13} Pylorus preservation remains critical when performing PD for pediatric patients given the desire to avoid hindering future growth and development. The outcome for patients with SPT is excellent, with 2- and 5-year survival estimated to be 97 percent and 95 percent, respectively.⁴ Overall, these tumors tend to be indolent, slow-growing, and have a low risk of malignant potential.³ Age under 13.5 years at diagnosis, tumor rupture and metastases have been identified as risk factors for recurrence.¹⁵ Surgical resection remains the mainstay of treatment and is indicated even in the presence of local invasion and metastatic disease.⁷

Conclusion

Complete resection with pylorus-preserving PD was performed in a young girl with SPT, and no complications occurred. Consistent with prior reports, PD appears to be a safe and curative option for pediatric patients with SPT of the pancreatic head. In addition, our case highlights the fact that, although rare, SPT should be considered a potential diagnosis for children presenting with a primary inflammatory mass of the pancreas. If the exact etiology remains unclear based on imaging, ERCP with FNA can help make the diagnosis and guide definitive treatment.

Lessons Learned

The presentation of solid pseudopapillary tumors is variable and rare in the pediatric population. That said, it is always most appropriate to expect the unexpected while searching for the unexpected in the case of a pediatric patient with the unusual presentation of a mass in the head of the pancreas. In general, the prognosis for these patients is generally very good, and pancreaticoduodenectomy is a safe and well-tolerated form of curative treatment.

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