# Cystic Pancreatic Mass in a Child

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#### **Case Presentation**

A 16-year-old boy presented to the emergency department complaining of vague abdominal pain following a motor vehicle collision. Based on initial findings on contrast-enhanced computed tomography (**CT**, **Figure 1A**), additional workup included ultrasound (**US**, **Figure 1B**) and MRI (**MRI**, **Figure 1C**) through the abdomen.



**FIGURE 1.** Axial contrast-enhanced CT image (A) demonstrates a cystic mass with a few internal septations within the head of the pancreas. Transverse US image (B) shows the mass with heterogeneous internal echogenicity. Axial T1 fat-suppressed postcontrast MR image (C) reveals the cystic mass with enhancing solid components posteriorly (enhancing regions were hypointense on precontrast T1 sequences, not shown).

## **Key Imaging Findings**

Cystic pancreatic mass in a child

## **Differential Diagnosis**

Solid psuedopapillary tumor (SPT) Pancreatic pseudocyst Pancreatoblastoma

## Discussion

Given the history of trauma in the case presented, along with the imaging findings on CT, the initial differential diagnosis included a pancreatic pseudocyst vs. solid pseudopapillary tumor (SPT). Based on the patient's age, pancreatoblastoma was considered unlikely. Follow-up imaging and cytological evaluation revealed the cystic mass to have intrinsic solid components with internal hemorrhage.

The overall occurrence of primary pancreatic neoplasms in the pediatric population is relatively rare, comprising only 0.1% of pancreatic tumors from all age groups.<sup>1</sup> Various imaging modalities have proven useful in the evaluation of such lesions, with cytological evaluation providing definitive diagnosis.

### Solid pseudopapillary tumor

Solid pseudopapillary tumor (SPT) is rare in the pediatric population, typically presenting as a heterogeneous and encapsulated mass with a combination of cystic, solid, and hemorrhagic components.<sup>2</sup> These tumors can occur throughout the pancreas, with a predilection for the pancreatic head and tail. This tumor is seen predominantly in adolescent girls (91%),3 and overall is more common in African-Americans and Asians.<sup>4</sup> Patients are often asymptomatic, with the tumor being discovered incidentally. A usual presenting symptom is vague abdominal pain, likely secondary to mass effect.

Imaging aids significantly in the diagnosis of SPT. This entity typically appears as a solid mass with internal heterogeneity and often an enhancing capsule on CT. Calcifications can be infrequently associated with these lesions.<sup>1,3</sup> Ultrasound typically demonstrates solid and cystic components, with only some demonstrating internal vascularity upon sonographic interrogation.<sup>1</sup>

MRI provides more distinctive features, such as a fibrous hypointense rim and fluid-fluid levels. Internal hemorrhage can be identified on the T1weighted images with enhancement of solid components and the fibrous capsule.<sup>1</sup> The lesions are well-circumscribed and are usually large upon initial evaluation, ranging 8-10 cm.<sup>3</sup>

Histological evaluation reveals polygonal cells in sheets and cords, with interspersed pseudopapillary structures and pseudorosettes.<sup>3</sup> Although typically of low malignant potential, spread beyond the pancreas can occur in up to 15% of patients. Because of the risk of metastases, surgical excision, often with a Whipple procedure or distal pancreatectomy depending on tumor location, is recommended.<sup>5</sup>

#### Pancreatic pseudocyst

Pancreatic pseudocyst is a common postinflammatory entity primarily comprised of amylase-rich fluid. These lesions are most often seen following a bout of pancreatitis or traumatic injury, usually appearing 4-6 weeks following the insult.<sup>6</sup> Pseudocysts typically present as a unilocular peripancreatic lesion that is round or oval. Additional imaging clues include peripancreatic inflammatory changes, or findings of chronic pancreatic insult, such as atrophy or calcifications. On MR imaging, the lesions show bright fluid signal on T2-weighted sequences often with internal debris.<sup>7,8</sup>

If pancreatic pseudocysts are  $\leq 6$  cm and asymptomatic, the usual course of action is to perform follow-up imaging with conservative management. If there is concern or uncertainty of the etiology, cyst aspiration under ultrasound guidance can be done for cytological analysis. If symptomatic, treatment may involve image-guided, surgical, or endoscopic decompression for larger lesions.

### Pancreatoblastoma

An unlikely diagnosis given the patient's age, pancreatoblastoma is the most common pancreatic tumor of young children,<sup>9</sup> although the incidence is rare, accounting for approximately 0.2% of all pancreatic tumors. It is primarily seen in children 1-8 years (mean: 5 years-old) with slight male predilection. They have been described as either arising from the ventral or dorsal anlage anatomically, with microscopic evaluation demonstrating resemblance to incompletely differentiated acini of the fetal pancreas. A congenital form of pancreatoblastoma has been associated with Beckwith-Wiedemann syndrome. Typical presentation is an asymptomatic, large and slow-growing mass with symptoms arising secondary to mass effect.<sup>10</sup>

These lesions can appear on CT as a multiloculated mass with enhancing septae, which are usually located near the pancreatic head. Ultrasound demonstrates a heterogeneous mass with multiple cystic spaces.<sup>9</sup> MR imaging will often demonstrate a lesion with intermediate T1 and increased T2 signal intensity.<sup>10</sup> Definitive diagnosis is made with tissue sampling.

Surgical excision is the treatment of choice for pancreatoblastoma. In the rare instance of metastatic disease, chemotherapy and radiation are utilized. Tumors located in the body or tail carry a poorer prognosis.<sup>9</sup>

### Diagnosis

Solid pseudopapillary tumor (SPT) of the pancreas; the patient underwent surgical excision without complication.

## Summary

Primary differential considerations for a cystic pancreatic lesion in a child in the setting of prior trauma or inflammation include pancreatic pseudocyst vs. neoplasm. Primary pancreatic neoplasms are rare in the pediatric population, with solid pseudopapillary tumors most commonly seen in adolescent girls. Imaging characteristics include a heterogeneous mass containing solid and cystic components with internal hemorrhage and a surrounding fibrous capsule. Lesions are often discovered incidentally or as a result of exploring vague abdominal pain. The tumor has low malignant potential; however, it is treated with surgical excision when possible due to the potential for metastases. Imaging and tissue sampling are imperative in differentiating this neoplasm from pancreatic masses with a similar appearance yet different prognosis and management.

#### REFERENCES

1. Ahmed TS, Chavhan GB, Navarro OM, et al. Imaging features of pancreatic tumors in children: 13-year experience at a pediatric tertiary hospital. Pediatric Radiol 2013;43:1435-1443.

2. Cantisani V, Koenraad JM, Levy A, et al. MR imaging features of solid pseudopapillary tumor of the pancreas in adult and pediatric patients. Am J Roentgenol 2003;181:394-401.

3. Rebhandl W, Felberbauer FX, Puig S, et al. Solid-pseudopapillary tumor of the pancreas (Frantz tumor) in children: report of four cases and review of the literature. J Surg Oncol 2001;76:289-296.

4. Coleman KM, Doherty MC, Bigler SA. Solid-pseudopapillary tumor of the pancreas. Radio-Graphics 2003;23:1644-1648.

5. Hernandez JM, Centeno BA, Kelley ST. Solid pseudopapillary tumors of the pancreas: case presentation and review of the literature. Am Surg 2007;73:290-293.

6. Kim YH, Saini S, Sahani D, et al. Imaging diagnosis

of cystic pancreatic lesions: pseudocyst versus nonpseudocyst. RadioGraphics 2005;25:671-685

7. Sahani DV, Kadavigere R, Saokar A, et al. Cystic pancreatic lesions: a simple imaging-based classification system for guiding management. Radio-Graphics 2005;25:1471-1484.

8. Macari M, Finn ME, Bennett GL, et al. Differentiating pancreatic cystic neoplasms from pancreatic pseudocysts at MR imaging: value of perceived internal debris. Radiology 2009;251:77-84.

9. Low G, Panu A, Millo N, et al. Multimodality imaging of neoplastic and non-neoplastic solid lesions of the pancreas. RadioGraphics 2011;31:993-1015.

10. Montemarano H, Lonergan GJ, Bulas DL, et al. Pancreatoblastoma: imaging findings in 10 patients and review of the literature. Radiology 2000;214:476-482.