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A Rare Case of Large Solid Pseudopapillary Tumor in a Child

Authors' Contribution:

Study Design A

Data Collection B

Statistical Analysis C

Data Interpretation D

Manuscript Preparation E

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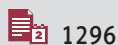
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None declared

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Conflict of interest:

Patient: Female, 15-year-old
Final Diagnosis: Solid pseudopapillary tumor
Symptoms: Abdominal pain
Medication: —
Clinical Procedure: —
Specialty: Radiology

Objective: Rare disease**Background:** Solid pseudopapillary tumor (SPT) is a rare disease with low-grade malignancy potential of the pancreas. SPT accounts from 0.13% to 2.7% of all exocrine pancreatic tumors. We report an unusual case with a large solid pseudopapillary tumor that was diagnosed and treated in our pediatric hospital.**Case Report:** A 15-year-old girl was incidentally found to have an abdominal mass on ultrasound examination. Computed tomography (CT) scans showed a well-defined tumor that was raised in part of the tail and body of the pancreas. The tumor size at greatest diameter was 18.2 cm. A mass excision was performed to remove the whole tumor, and the histopathological findings confirmed SPT without evidence of malignancy.**Conclusions:** SPT is a rare disease that mostly affects young female patients. The clinical symptoms of this disease are un-specific. The prognosis of SPT is good, even in case of distant metastasis. Close follow-up is required to detect metastasis so the appropriate treatment method can be chosen.**MeSH Keywords:** Case Reports • Pancreatic Neoplasms • Tomography, X-Ray ComputedFull-text PDF: <https://www.amjcaserep.com/abstract/index/idArt/923990>

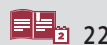
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Background

Solid pseudopapillary tumor (SPT) is one of the rarest types, accounting for 0.13% to 2.7% of all exocrine pancreatic tumors [1,2], usually affecting young women, with a reported male/female ratio of 7–11: 1 [3,4]. Before being named SPT by the World Health Organization in 1996 [5], SPT was known as Franz's tumor [6] or Hammoudi tumor [7].

Solid pseudopapillary tumors are frequently incidentally found during radiological and surgical procedures. They present as an abdominal mass without specific clinical presentation, such as vague abdominal pain or abdominal discomfort.

SPT is considered a tumor with low-grade malignant potential. The incidence of malignant transformation of SPT is around 15%. Total surgical resection is the first-line treatment option, with a favorable long-term outcome.

Here, we report an unusual case with a large solid pseudopapillary tumor that was diagnosed and treated in our pediatric hospital.

Case Report

Clinical history

A 15-year-old female patient presented at Ho Chi Minh City Children's Hospital #2 in November 2019. She reported recurrent abdominal pain over the past 6 months but she had never received an abdominal examination before. The tumor was incidentally discovered on ultrasound examination. On physical examination, the specific symptoms were unremarkable: the abdomen was soft and no other abdominal signs were found. Laboratory investigations were unremarkable. All the tests for serum amylase and serum lipase levels, as well as tumor markers such as carcinoembryonic antigen (CEA), carbohydrate antigen (CA19-9), and α -fetoprotein (AFP), were within the normal range. No evidence of abnormalities in liver function or pancreatic insufficiency were found.

Imaging findings

The computed tomography (CT) scan showed a heterogeneous well-defined tumor 18.2 cm in diameter. The tumor was raised in the tail and body of the pancreas, and consisted of solid, cystic, and calcifications components (Figure 1). The Wirsung's duct was normal. The tumor did not invade the spleen parenchyma or the adjacent anatomical structure.

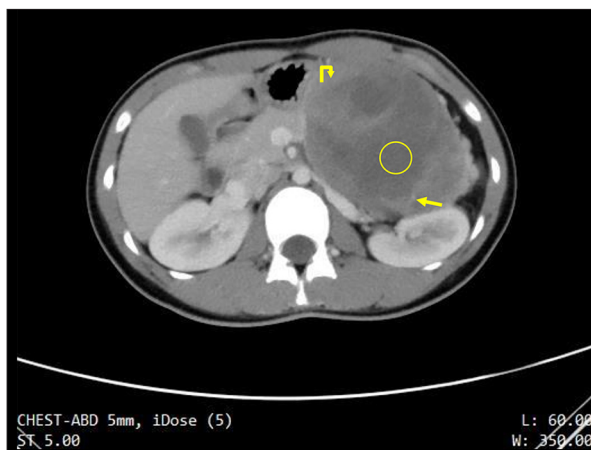


Figure 1. Abdominal computed tomography (CT) scan showing a large heterogeneous mass that was partly solid and cystic and partly calcified soft tissue.

Surgical and histopathological findings

After consultation with our Multidisciplinary Tumor Board, we decided upon total excision of the tumor 10 days later. The tumor size was confirmed to be 10×16×18 cm, with a well-defined border. The tumor covered the tail and body of the pancreas, and was slightly adherent to the spleen and splenic vein, and to the angle of Treitz and duodenal segment D4.

Histopathological findings confirmed a solid pseudopapillary tumor of the pancreas, with no evidence of a malignancy.

Follow-up

No postoperative complications occurred and no additional treatment was administered. After 3 months of follow-up, no signs of tumor recurrence were observed, with no evidence of exocrine or endocrine pancreatic insufficiency.

Discussion

Solid pseudopapillary tumor is a very rare disease that mostly affects young female patients in their 20s and 30s. SPT is often incidentally diagnosed on imaging examination in both adults and children. SPTs most frequently arise from the tail or head of the pancreas [8]. Since the first case report was published in 1959 by Frantz [9], multiple case reports and studies have been published on diagnosis and treatment (laparoscopic or open surgical) for SPT. However, to the best of our knowledge, this is the first case of such a large tumor (18.2 cm in diameter) in a child. Branco [10] presented the case of a 17-year-old found to have as SPT 14 cm in diameter. Mahida reported 8 pediatric cases of SPT, with the largest tumor being 14 cm in diameter [11].

Our case is a 15-year-old female patient in whom the tumor was incidentally diagnosed on ultrasound examination. A large review showed that the average age of patients with SPT is 21.97 years and that the male-to-female ratio is 1: 9.8 [8]. In another systematic review of 292 patients diagnosed with SPT of the pancreas, the average patient age was 23.9 years, with a male-to-female ratio of 1: 9.4 [12].

The clinical presentation of this disease is usually nonspecific. Vague gastrointestinal symptoms are reported, such as upper-abdominal fullness or discomfort, epigastric or abdominal pain in the left upper quadrant, depending on an enlarged and often palpable abdominal mass [13]. Some patients present without any symptoms, and the tumor is found incidentally by imaging or abdominal examination. Commonly, patients have no signs of exocrine, insufficiency of pancreatic endocrine, abnormal function of the liver, or cholestasis. All the test results for serum tumor markers are also within the normal range [4,8]. In our case, the patient had experienced recurrent abdominal pain for more than the past 6 months, but she had not previously received an abdominal examination. The blood tests for tumor markers before surgery, such as AFP, CEA, and CA 19-9, were considered normal. The clinical features and laboratory findings in our case were consistent with the literature.

SPTs can arise from all parts of the pancreas, but SPT raising in the tail is more common [14]. Finding a SPT invading the adjacent organs to the pancreas, such as the duodenum or the spleen, is rare. Differential diagnosis is required between SPT and other tumors, such as adrenal tumor, endocrine tumor of the pancreas, cyst of the liver or hepatic tumor, or a pseudocyst, when the tumors raise in the head, body, or tail of the pancreas [15].

Abdominal imaging (ultrasound, computed tomography) shows a heterogeneous, well-encapsulated area with cystic and solid components, and the displacement of nearby structures by a well-defined border. Calcification of components at the periphery of the tumor can occur (as seen in our case) and contrast enhancement in the center of the tumor suggests hemorrhagic necrosis [16]. However, CT scans have some limitations for capturing certain imaging characteristics of the tissue, such as cystic degeneration, hemorrhage, or a capsule, compared to magnetic resonance imaging (MRI). These findings might, as presented in the histopathology results, be indicative of a specific lesion such as SPT of the pancreas. Thus, MRI might help further describe these findings, and can be useful in distinguishing SPT from complex cystic masses in the pancreas [17]. Despite the developments in imaging technologies, the diagnosis of

SPT before surgery remains difficult due to radiological similarities among cystic lesions. In some reports, the authors performed fine-needle aspiration (FNA) biopsy preoperatively under endosonography for diagnosis of the tumor. However, this might not be widely accepted because it is unreliable for confirmation and the potential exists for tumor invasion [18,19]. In our case, we did not perform fine-needle aspiration biopsy before surgery.

Regarding management of SPT, total surgical resection is the first-line treatment option when SPT is diagnosed. The SPT is usually covered by a pseudocapsule and frequently presents as a benign or low-grade malignant tumor. A surgical procedure – distal pancreatectomy, Whipple operation, pylorus preserving pancreato-duodenectomy, or enucleation – is selected depending on the location of the tumor within the pancreas. Many studies showed that less aggressive surgery can be used for surgical management of SPT [20].

In our case, the patient underwent a total excision to remove the whole tumor. No postoperative complications occurred and no additional therapy was provided. Due to the favorable resectability of SPTs, adjuvant treatment is indicated only for a small group of patients. The effectiveness of chemoradiotherapy or chemotherapy in treatment for patients with SPT is also unclear. Adjuvant therapy, such as radiotherapy and chemotherapy, for unresectable cases has been documented in several previous reports, with favorable outcomes [21,22].

Conclusions

This case report describes a large SPT of the pancreas that was diagnosed and successfully treated in our pediatric hospital. SPTs mostly affect young female patients. The clinical presentation of this disease is usually nonspecific. The prognosis of SPT is good, even in case of metastasis. Although surgical resection is a curative method, it is important to closely follow the patient to detect metastasis and indicate the appropriate treatment method.

Institution where work was done

Ho Chi Minh City Children's Hospital #2, Ho Chi Minh City, Vietnam.

Conflict of interests

None.

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