

Case Report

Solid pseudopapillary neoplasm, a rare pancreatic tumor-case report

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ABSTRACT

The uncommon cystic exocrine pancreatic tumor known as solid pseudopapillary epithelial neoplasm (SPEN) most frequently affects young females. We provide a case study of a 17-year-old female who experienced intermittent vomiting and epigastric pain. Her laboratory results were within normal ranges, and a clinical abdominal exam revealed pain in the region of epigastrium. A 40×3×31 mm iso-hyperdense minimally enhancing solid mass with well-defined borders was detected on her CECT scan. Nothing calcified, neither localized lymphadenopathy nor fat stranding. It was just next to the splenic vein, which was completely opaque. Her histopathology revealed a solid pseudopapillary pancreatic tumor after she underwent a spleen preserving distal pancreatectomy. Beta-catenin and vimentin IHC assays were significantly positive, confirming the diagnosis and she was on a regular follow-up. As of now, the patient is asymptomatic and has not experienced a recurrence. In contrast to other forms of pancreas tumors, SPEN is treatable with early identification and full surgical resection.

Keywords: Pancreas, Pseudopapillary neoplasm, Distal pancreatectomy

INTRODUCTION

Solid pseudopapillary tumors (SPT) of the pancreas are a rare type of exocrine pancreatic neoplasm that typically affects young girls between their second and fourth decades of life.¹ The world health organization (WHO) gave it the official designation "SPT" and classed it as a borderline malignant tumor with uncertain biological behaviour in 1996.² Most young females in their 30s are affected by it. It may or may not have symptoms. SPTs are frequently big tumors that can affect any area of the pancreas.^{3,4}

CASE REPORT

A 17-year-old female presented to the emergency room complaining of lower abdominal and epigastric pain that had persisted for 10 days. It was subtle in its onset, advancing in character, and radiating to the back. There were no causes of aggravation or relief for the pain, which was dull and agonizing in character. Vomiting that occurred on and off was non-bilious, non-projectile, and

accompanied by pain. The patient had no known comorbidities and no prior surgical history. The history of menstrual cycles was uneventful.

General examination was within normal ranges. An abdominal exam indicated a soft abdomen and some slight epigastric discomfort. No lump or organomegaly that could be felt. Bowel sounds could be heard and were normal.

The results of the tests were within acceptable bounds.

A well-defined iso-hyperdense solid mass in the distal body measuring 40×3×31 mm was detected by CECT scan. Nothing calcified, neither localized lymphadenopathy nor fat stranding. It is opaque and next to the splenic vein. The liver was healthy with no metastasis. The remainder of the abdominal organs were healthy.

The patient underwent a successful spleen-preserving distal pancreatectomy.

Intraoperative findings were mass of tumour measuring around 5 cm was found in the pancreas' body and tail regions. Additionally, there was evidence that the posterior tumor mass abutted the splenic vein. There was no local lymph node enlargement.

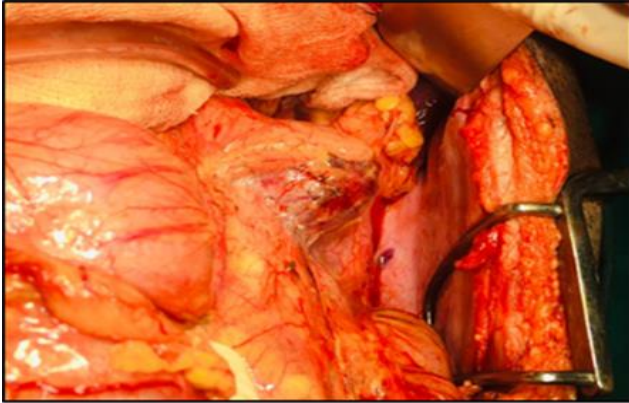


Figure 1: Tumour mass at the tail of pancreas.

A single monolayered sheet and clusters of epithelial cells were the only things to be found in the intraoperatively collected peritoneal fluid. There were also no cancerous cells to be found.

A solid mass measuring about 4 cm in length and well-circumscribed with central cystic regions filled with haemorrhagic fluid was seen during a gross examination of the pancreas. There was healthy pancreatic tissue in the region surrounding the tumor.

An inspection under the microscope revealed a tumor with solid and cystic components. Cells are organized in cords, sheets, and papillary structures with pseudo-rosette development in the solid component. Haemorrhage and haemosiderin-loaded cyst macrophages were visible in the cystic component. The tumor's perimeter contained healthy pancreatic tissue.

After surgery, patient made a good recovery. Since the pseudopapillary neoplasm was solid, a R₀ resection was performed, and regular follow-up was indicated. Patient has not had a recurrence and is currently symptom-free.



Figure 2: Gross appearance of tumour (cut open section).



Figure 3: Gross appearance of resected tumour mass.

IHC showed results consistent with solid pseudopapillary epithelial neoplasm with following markers:

Table 1: showing tumour marker study.

Marker	Clone	Results
Synaptophysin	SNP88	Positive (weak to focal)
Beta catenin	EP35	Positive
Chromogranin A	EP38	Negative
PCK	AE1/AE3	Positive (moderate to weak)
Vimentin	V9	Positive

DISCUSSION

The unusual and unexplained solid pseudopapillary tumour of the pancreas (SPT) was initially identified by Frantz in 1959.³ In addition to these names, it is also known as Frantz's tumor, solid and papillary tumor, papillary cystic tumor, solid-cystic tumor, solid-cystic and papillary epithelial neoplasm, benign or malignant papillary tumour of the pancreas, papillary epithelial neoplasm of the pancreas in a child, and papillary adenocarcinoma of the pancreas.³ The tumour's origin is still uncertain. It is an uncommon tumor that frequently affects teenage girls. In 1999, 452 cases of SPT were documented in English-language publications globally, according to Lam et al male to female ratio is 1:9.5, and it has low-grade malignant potential with 15% invasion or metastasis.^{4,5}

The WHO changed the name of this tumor to SPT in 1996 to reflect the international histologic categorization of exocrine pancreas tumors. Its genesis is unknown, however pluripotent cells are thought to be its source. However, it is hypothesized that SPTs may originate from genital ridge/ovarian anlage-related cells that were attached to the pancreatic tissue during early embryogenesis because of the female predominance of

SPTs and the known close proximity of the genital ridges to the pancreatic anlage during embryogenesis.⁶

The majority of patients report vague symptoms, which may include stomach pain or a visible, non-tender abdominal tumor.⁷ The abdominal mass may also cause other symptoms like nausea, vomiting, abdominal pain, or early satiety. These general problems could result in a tumor diagnosis that is made too late.⁸ It's uncommon to see obstructive jaundice.⁹ Exocrine or endocrine symptoms of pancreatic insufficiency are typically absent.¹⁰

Despite the fact that the majority of SPTs are benign, malignancy can develop in 15% of instances and show up as metastases or invasion of nearby structures.¹¹ Liver and peritoneum are metastatic sites that occur most frequently.¹² Most SPTs are found in the pancreas' body and tail.¹³ Due of its slow growth and lack of symptoms, it can develop to huge sizes. The tumors in the largest series had a mean diameter of 6.08 cm and ranged in size from 0.5 to 34.5 cm.¹⁴

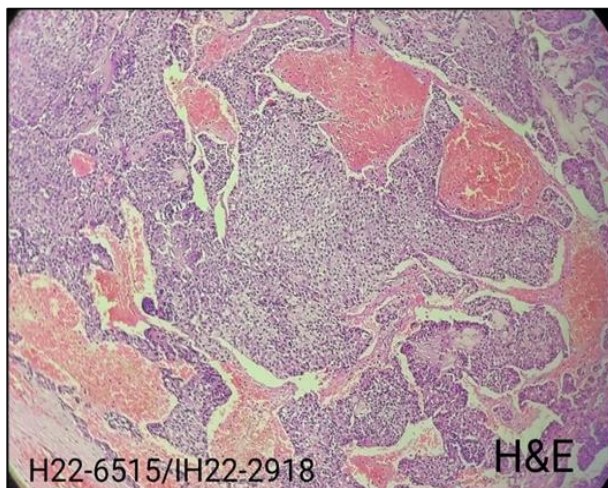


Figure 4: Microscopic image of the tumour section.

A well-circumscribed, encapsulated tumor with heterogeneous cystic and hemorrhagic degeneration is shown on a CT scan. Other characteristics could include central calcification, peripheral artery augmentation, and major pancreatic duct enlargement.⁷ A heterogeneous solid and cystic mass can be seen on ultrasound. The heterogeneity of the mass may be brought on by tumour hemorrhage.⁵ On T1-weighted images and on T2-weighted images, MRI typically reveals a well-circumscribed lesion with heterogeneous low or high signal intensity. Both T1 and T2 weighted scans often show the tumor capsule as a hypo-dense rim.^{17,18}

The Frantz tumors are epithelial, according to immunohistology investigations, however well-documented instances of cytokeratin and vimentin expression raise the possibility that these tumors are mesenchymal in nature.⁶

Additionally, neoplastic cells are typically positive for the neuroendocrine markers CD56, 1-antitrypsin, Leu-M1, and Ki-M1P while mainly negative for the neuroendocrine markers NSE, synaptophysin, and chromogranin.¹⁵ Therefore, any phenotypic relationship with the pancreatic cell lining is diminished by the lack of a distinctive immunoreactivity pattern.¹⁹

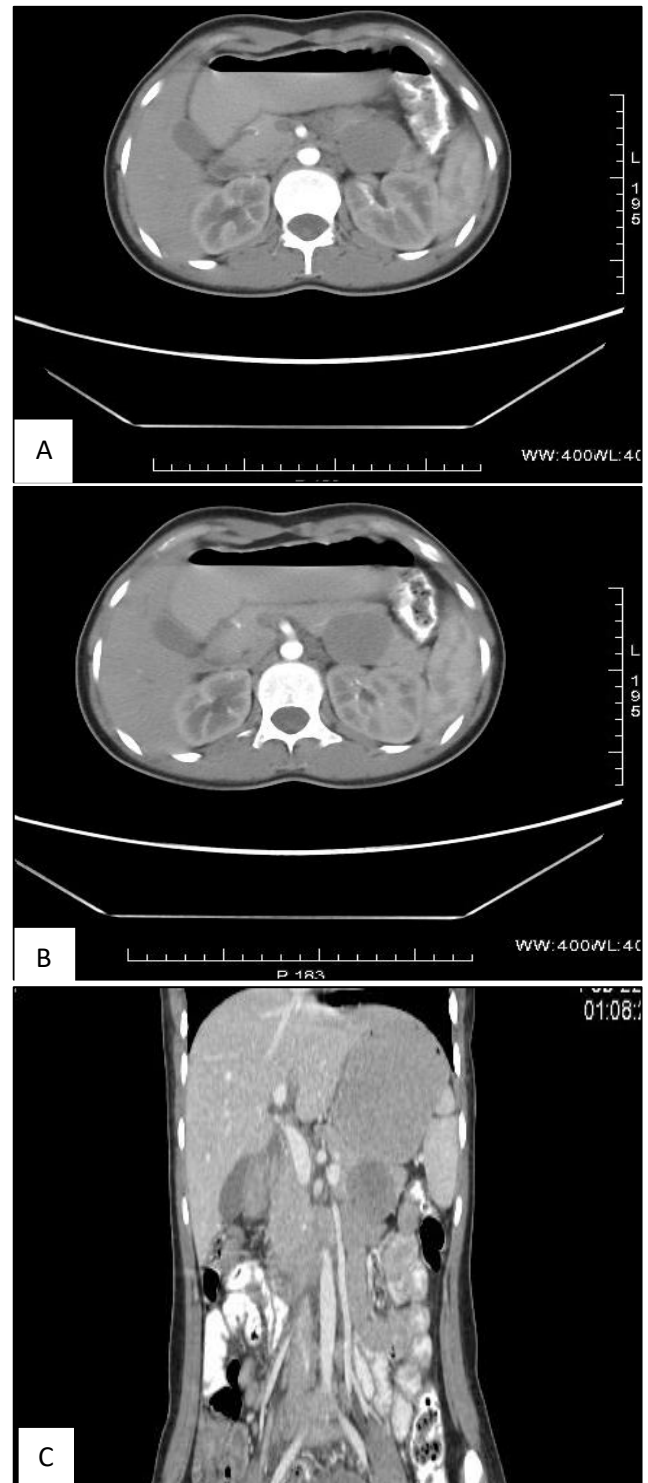


Figure 5 (A-C): CT scan images of the tumour mass on pancreas.

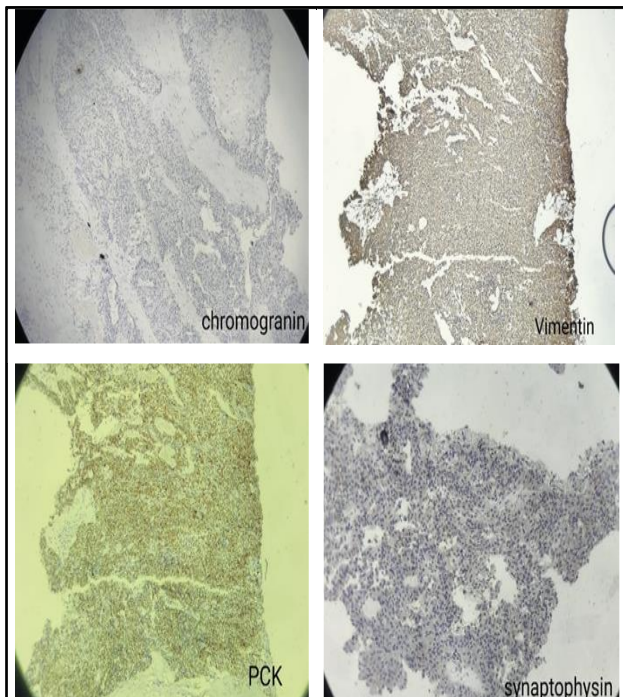


Figure 6: Microscopic views of IHC markers uptake (chromogranin, vimentin, PCK, synaptophysin).

Complete tumour excision is the recommended course of treatment for SPT (R0). Surgical technique is based on the position and size of the tumor.¹⁶ Distal pancreatectomy is the most frequent surgery because the majority of SPTs are found in the tail and/or body of the pancreas.²⁰ SPT of the head or uncinate process of the pancreas occurs in around one-third of patients and is treated with pancreatoduodenectomy. Central pancreatectomy may be used to treat pancreatic tumors that are in the body or neck and do not involve any vessels.^{21,22} Even in the presence of local invasion or metastasis, surgical resection is necessary.²³ The viability and advantages of splenic preservation following distal pancreatectomy have been shown in numerous studies, including a decreased lifetime risk of post-splenectomy sepsis and malignancies.²⁴

It is recommended to do a spleen-preserving laparoscopic distal pancreatectomy for benign or low-grade malignant tumors like SPT that are found in the body or tail of the pancreas. The secure separation of the body and tail pancreatic parenchyma from the splenic arteries is the most crucial technical part of this technique. Laparoscopy might make this procedure simpler to complete than open surgery.²⁵

CONCLUSION

Finally, pancreatic pseudopapillary tumors are extremely uncommon neoplasms with the potential to be malignant. They might exhibit an abdominal lump, stomach pain, or, in rare cases, jaundice. Long-term survival is provided by prompt resection after diagnosis.

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