

Large Solid Pseudopapillary Tumor Pancreas with Extensive Splenoportal Thrombosis and Malignant Transformation—A Rare Entity

Thara Pratap¹ Dhanya Jacob¹ Abhishek Yadav² Muhammed Jasim Abdul Jalal³ Iona Leekha Mathew⁴

¹Department of Radiology, VPS Lakeshore Hospital, Kochi, Kerala, India

²Comprehensive Liver Care Institute, VPS Lakeshore Hospital, Kochi, Kerala, India

³Department of Internal Medicine and Rheumatology, VPS Lakeshore Hospital, Kochi, Kerala, India

⁴Department of Pathology, VPS Lakeshore Hospital, Kochi, Kerala, India

Address for correspondence Muhammed Jasim Abdul Jalal, MBBS, DNB, MNAMS, MRCP, Department of Internal Medicine and Rheumatology, VPS Lakeshore Hospital, Nettoor.P.O., Maradu, NH 47-Byepass, Kochi 682040, Kerala, India
(e-mail: jasimabduljalal@yahoo.com).

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Abstract

Keywords

- solid pseudopapillary tumor
- papillary epithelial neoplasm
- papillary cystic neoplasm
- solid and papillary neoplasm
- low-grade papillary neoplasm
- Frantz or Hamoudi tumor

Solid pseudopapillary tumor (SPT) is an uncommon low-grade malignant cystic exocrine neoplasm of the pancreas that typically affects young women. The tumor has an excellent prognosis after surgical resection. Vascular involvement is usually not seen at presentation. Local recurrence and metastasis are also rare. Herein, we present a case of large, locally advanced SPT in a young female with splenoportal extension and malignant transformation.

Introduction

Solid pseudopapillary tumor (SPT) is a rare pancreatic neoplasm occurring in females in the second and third decades. The mean age of presentation is 24 years and one-third occur in adolescents.¹ It is also known as papillary epithelial neoplasm, papillary cystic neoplasm, solid and papillary neoplasm, low-grade papillary neoplasm, and Frantz or

Hamoudi tumor.² The first published description of a solid pseudopapillary neoplasm (SPN) was by Frantz in 1959. Hamoudi and colleagues detailed the electron microscopic appearance of the tumor later.

The tumor often presents with abdominal pain or as a palpable mass. SPT is a potentially malignant lesion.³ Surgery is often curative, and the 5-year survival is 95 to 97%.^{4,5} However, SPT has an unpredictable behavior and metastasis

or local invasion has been reported in up to 10 to 15% of cases.⁴ Prognosis is poor when the tumor occurs in male patients, when there is local invasion into adjacent structures, vascular infiltration, cellular atypia, or capsular invasion on histopathology, and when it is unresectable or is metastatic tumor.²

Case Report

A 17-year-old female patient presented with abdominal distension, intermittent abdominal pain, loss of appetite, occasional episodes of vomiting, and weight loss of 3 months duration. She also had an episode of hematemesis and melena.

On clinical examination, the patient was icteric and the abdomen was distended. A mass was felt in the right hypochondrium and epigastric region.

Laboratory parameters were as follows: hemoglobin 8.4; total leukocyte count $4,290 \times 10^3/\mu\text{L}$; platelet count $1,90,000 \times 10^3/\mu\text{L}$; bilirubin total/direct $3.6/0.6 \text{ mg/dL}$; aspartate aminotransferase 25 U/L; alanine aminotransferase 24 U/L; and alkaline phosphatase 102 U/L.

Ultrasound showed a lobulated exophytic mass in the region of pancreatic head extending into the main portal vein and its branches with luminal expansion (**►Fig. 1**).

Contrast-enhanced computed tomography (CECT) was done for a better characterization of the lesion, which showed a large lobulated mass measuring $22 \times 19 \times 18 \text{ cm}$ involving

the head and body of pancreas with exophytic extension, displacing and compressing the adjacent bowel loops. There was no atrophy or duct dilatation of distal pancreas. A plain CT scan showed curvilinear calcification in the periphery of mass (**►Figs. 2–4**). Post-contrast study showed heterogeneous enhancement with tumor vascularity from branches of the superior mesenteric artery. There was no significant necrosis or cystic changes. Contiguous intravascular tumor extension was noted with tumor expanding the main portal vein to almost three times its size and into the entire intrahepatic portal vein radicles. Distally there was extension to the splenic vein up to the splenic hilum and inferiorly to the superior mesenteric vein for a length of approximately 6 cm. Dilated intra-tumoral with peritumoral venous collaterals were noted (**►Fig. 5**) with thrombosis of a few collaterals in the mesentery and omentum. There was associated mild intrahepatic biliary radicle dilatation. Minimal free fluid was present. The possible differentials considered were SPT, neuroendocrine tumor, and leiomyoma/leiomyosarcoma as these are the tumors which present with intravascular extension.

Ultrasound-guided Trucut biopsy from the tumor was suggestive of SPN. Radiological features favored a locally aggressive SPEN in view of intravenous extension; however, the biopsy had not revealed any features of malignancy.

The patient was taken up for surgery, as the tumor was locally advanced and portal hypertension as evidenced by

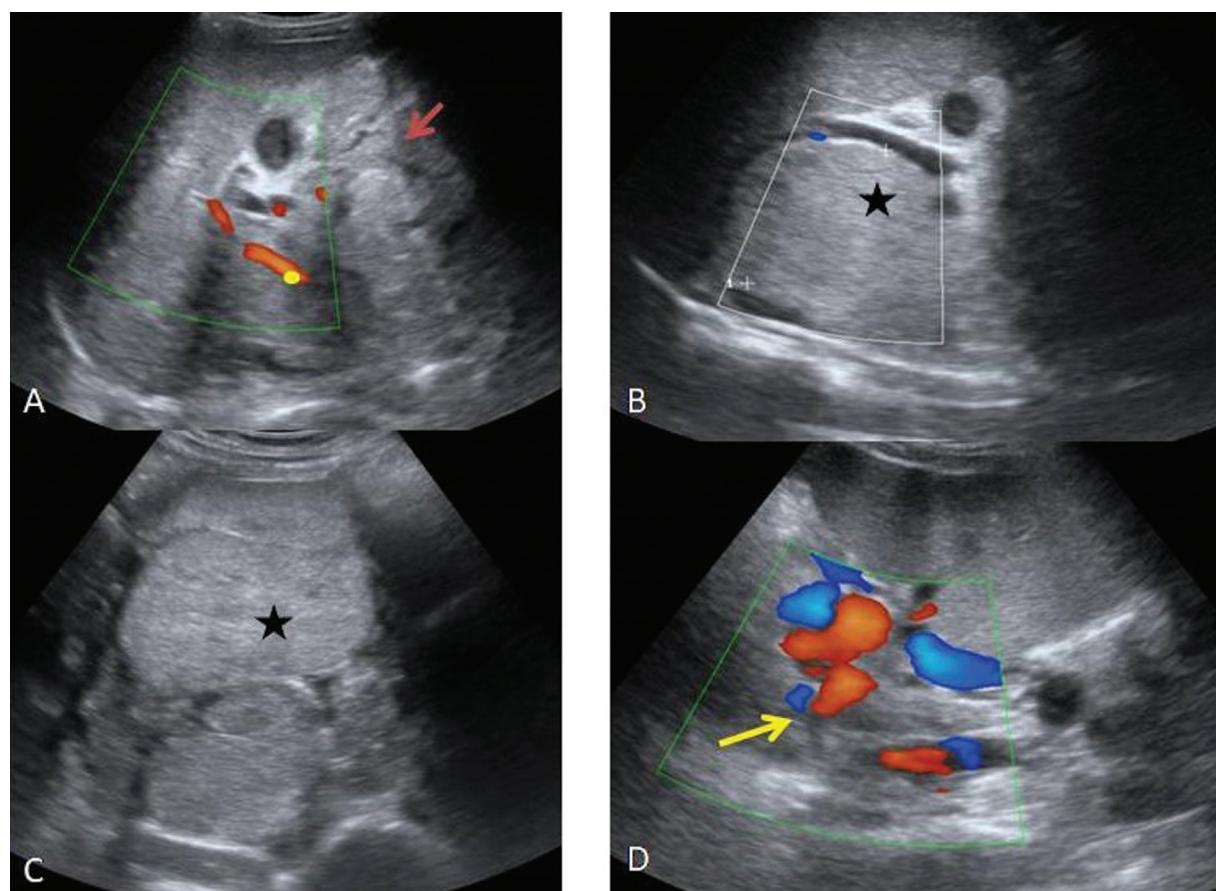


Fig. 1 (A–D) Ultrasound of abdomen showing a large lobulated isoechoic mass in the pancreatic head region (arrow) with intravenous extension and expansion of the main portal vein (asterisk) and its branches. Vascularity noted within the tumor thrombus (dot in A) with multiple periportal collaterals (arrow in D) also seen.

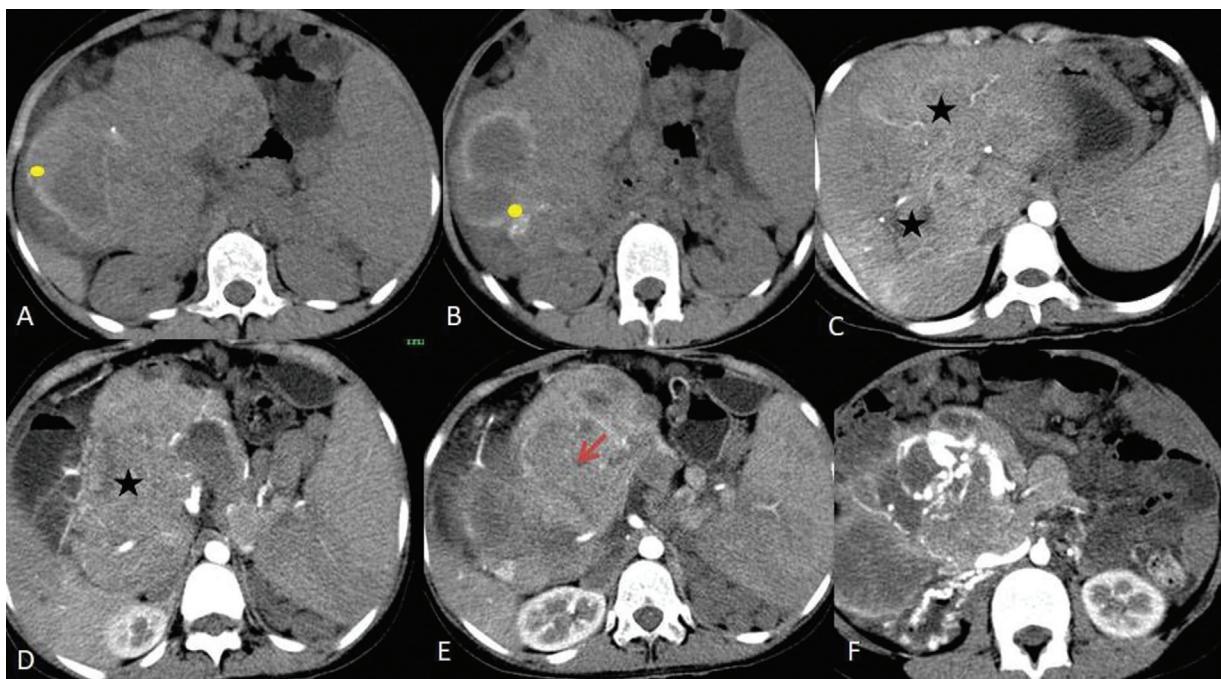


Fig. 2 (A, B) Contrast-enhanced computed tomography plain and (C–F) arterial phase axial sections showing peripheral curvilinear calcifications (dot) in the mass with heterogeneous enhancement in the pancreatic head region (arrows). Mass shows extension into the main portal vein and intrahepatic branches (asterisk).

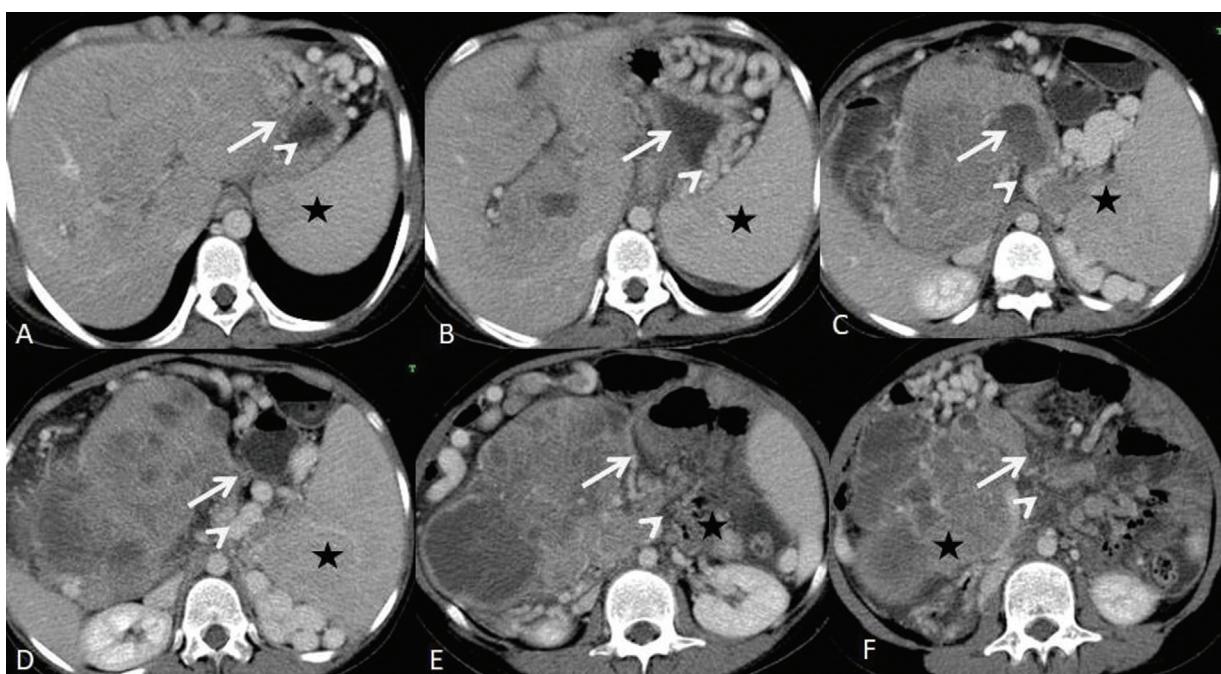


Fig. 3 (A–F) Contrast-enhanced computed tomography in hepatic venous phase scans showing large heterogeneous tumor (arrow) with splenomegaly (arrowhead) and collaterals in the peri-splenic and peritumoral region (asterisk).

multiple collaterals was seen. Whipple's pancreaticoduodenectomy was done, and intravascular thrombus was removed to the possible extent. The final histopathological diagnosis was SPN with foci of high-grade malignant transformation and lympho-vascular emboli with metastatic tumor in one peripancreatic lymph node. Immunohistochemistry showed positivity for synaptophysin and vimentin, and β -catenin confirming SPT (**►Figs. 6 and 7**).

The patient had postoperative collections, which was drained, developed septicemia and succumbed to the disease.

Discussion

SPT is an uncommon tumor and constitutes 1 to 2% of exocrine pancreatic tumors. Ninety percent of cases occur in females. SPT has a low-grade malignant potential and can



Fig. 4 (A–C) Coronal contrast-enhanced computed tomography in portal venous phase scans showing the large tumor (arrow) with splenomegaly, extension to the main portal vein with associated splenomegaly.



Fig. 5 (A) Contrast-enhanced computed tomography angiography and venography (B) with maximum intensity projection reconstruction showing tumor neovascularity from branches of superior mesenteric artery in the first image and spleno-portal collaterals due to portal hypertension in the second image.

be locally aggressive.² According to the 2000 World Health Organization classification system, aggressive SPT should be classified as solid pseudopapillary carcinomas.

The tumor commonly involves the pancreatic head (34–40%) or tail region (24–36%). Large SPT will typically displace the adjacent structures rather than invading them. These tumors exhibit a soft consistency which rarely causes obstruction of the common bile duct or main pancreatic duct, even if located in the pancreatic head.⁶

Mean tumor size at presentation described is 6 to 8 cm, though variable. Most of the tumors are moderate sized at presentation and present as palpable masses. About 15% of the patients are asymptomatic.⁷

On ultrasound, the tumor could be predominantly solid, predominantly cystic, or mixed solid and cystic mass. Fine-needle aspiration is usually diagnostic.^{8,9}

On cross-sectional imaging, SPT is encapsulated homogeneous and enhancing when small and heterogeneous if large, due to hemorrhage and cystic degeneration. The fluid level has been reported in 18% of the cases. Peripheral calcification is seen in 30% of the cases.¹⁰ Central dystrophic calcifications may occur in areas of hemorrhage. Lamellar calcifications may also be present.¹¹ If predominantly cystic, calcified and enhancing soft tissue components are seen in the periphery of the lesion. A study by Cantisani et al¹² showed the most common enhancement pattern of SPT as early, peripheral, and heterogeneous enhancement during the arterial phase

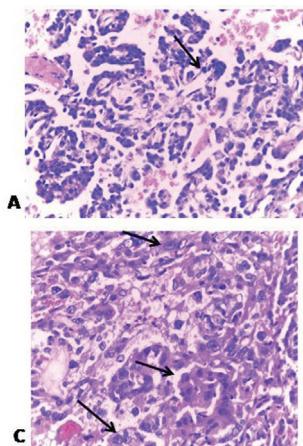


Fig. 6 (A–C) H&E stain (magnification x400). Pseudopapillae (arrow in A) with vascular stalks surrounded by loosely cohesive fairly monomorphic tumor cells. Arrows in B and C show a tumor with high-grade areas showing increased nuclear atypia and mitosis. H&E, hematoxylin and eosin.

with a progressive but heterogeneous filling of the lesion during the portal venous and equilibrium phases. The tumor capsule usually shows early and more intense enhancement.

Magnetic resonance imaging (MRI) has an added advantage over CT in the diagnosis of SPT as most of the tumors show evidence of hemorrhage with a variable signal on MRI.¹³ A hypointense rim suggestive of capsule can be seen on both T1 and T2 weighted images. CT has limitations in demonstrating hemorrhage, cystic degeneration, and capsule.¹⁴

Malignant transformation is rare and is suspected radiologically when the lesion is large with eccentric lobulated margin rather than smooth lobulation and when there is focal nodular or amorphous scattered calcification rather than rim calcification or if there is invasion into the surrounding tissue, or metastasis.^{15,16} The most common site of metastasis is the liver. Metastasis can also occur in the peritoneum, omentum, or lymph node. Solitary liver metastasis is not a contraindication to surgery.

The histogenesis of SPN is still enigmatic. This tumor may have its origin from the pluripotent stem cells of the genital ridges. This theory is strongly supported by features such as expression of progesterone and androgen receptors in the tumor, lack of expression of pancreatic epithelial markers, and regression of some tumors after menopause. The tumors are hormone dependent and may increase in size during pregnancy. Pathologically, although SPN was considered a “benign” or “borderline” tumor for many years, recent molecular evidence demonstrates alterations in cancer-associated genes and the ability to metastasize, and hence, this entity is now considered to be a low-grade malignant pancreatic tumor.¹⁷ The tumor shows a slow doubling time (765 days).¹³

SPN with foci of high-grade malignant transformation is an aggressive histologic subtype, the presence of nuclear atypia and high-mitotic rate may suggest an aggressive subtype.¹⁸

There is a lack of consensus regarding the utility of the Ki-67 index as a prognostic marker for SPT; however, Yang et al reported that a Ki-67 index >4% might indicate a poor

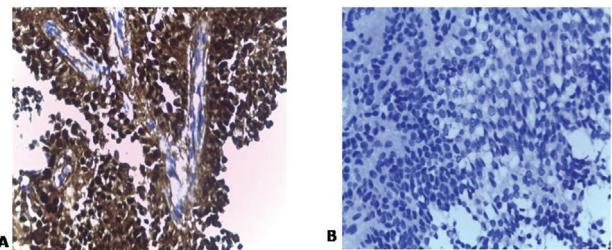


Fig. 7 (A, B) Immunohistochemistry showing positivity for β -catenin (both nuclei and cytoplasm) in A and negative for chromogranin in B (magnification x400).

prognosis in patients with SPT.^{19,20} Our patient had a Ki-67 index of 3% but showed malignant transformation.

The literature search showed only a few cases of SPT with minimal intravascular extension. The key finding in our case is extensive intravascular tumor extension, not described so far. In spite of intravascular extension, the patient was taken for surgery, being the only treatment option available even in aggressive lesions. The role of chemotherapy and radiotherapy is not well established in invasive/malignant SPT.

Complete surgical resection is the treatment of choice in SPT.¹⁸ About 10 to 15% of patients have metastases at the time of diagnosis or develop metastases at some point in the future. The histopathology does not always completely and precisely predict the biological behavior of tumor, recurrence, or malignant potential.²¹ Hence, the importance of imaging and regular follow-up in all cases of SPT after surgical resection to diagnose early recurrence or distant metastasis should be emphasized.

Conclusion

A pancreatic mass in a young adult female with a heterogeneous appearance should raise suspicion for SPT. Malignant transformation should be suspected radiologically when there is angioinvasion or infiltration of adjacent structures as the role of the pathology of predicting malignant behavior is still uncertain. This is a unique case with extensive intravascular extension of thrombus not described in the literature so far.

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Conflict of Interest

None declared.

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