

## Rare solid pseudopapillary neoplasm in a Caucasian man

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### ABSTRACT

*Pancreatic pseudopapillary neoplasms are rare pancreatic tumors and have a female predominance. Clinical manifestations include abdominal pain and/or an abdominal mass. Some patients are asymptomatic, and the tumors are found incidentally on imaging. Endoscopic ultrasound with fine needle biopsy with pathological tissue remains the gold standard for diagnosis. The malignant potential is low, and surgery remains the standard treatment.*

**Keywords:** Solid pseudopapillary neoplasm, pancreatic tumor, pancreatic malignancy, endoscopic ultrasound

### INTRODUCTION

Solid pseudopapillary neoplasms (SPN) are rare tumors of the pancreas. They occur more frequently in women and in patients of Asian or African American ethnic backgrounds. We report a rare case of a solid pseudopapillary neoplasm in a Caucasian man who initially presented with nonspecific acute abdominal pain.

### CASE

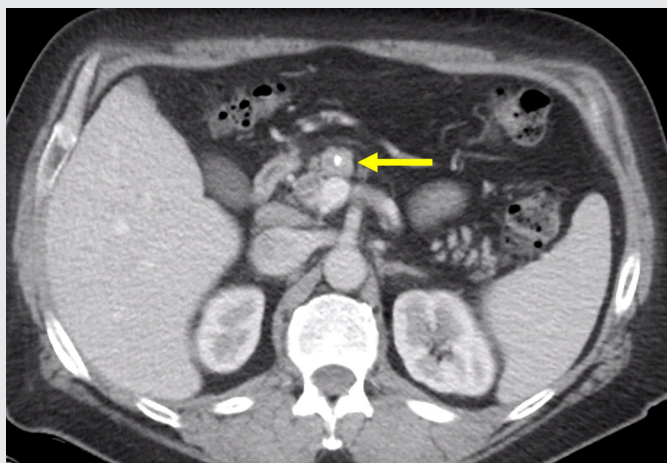
A 52-year-old Caucasian man with a past history of poorly controlled diabetes mellitus and hypertension presented to the emergency department with diffuse abdominal pain and nausea for one day. He described the pain as dull aching, with a pain score 2 out of 10, and generalized. He had lost 120 lbs. unintentionally in one year. The patient denied a history of alcohol, tobacco, or illicit drug use. His vital signs showed blood pressure 188/117 mm Hg, heart rate 96 beats per minute, respiratory rate 20 breaths/minute, temperature 97.4°F, and oxygen saturation 92% on room air. His body

mass index was 27.3 kg/m<sup>2</sup>. On examination, he was alert and in no acute distress. He had clear breath sounds on auscultation. His abdomen was soft and non-distended, with tenderness at the epigastrium and with normal bowel sounds.

Laboratory workup showed a white blood cell count of 12.31 k/ $\mu$ L (ref 4.23–9.07), hemoglobin 13.6 g/dL (13.7–17.5 g/dL), and platelet count 219 k/ $\mu$ L (165–400). Serum glucose was 247 mg/dL, hemoglobin A1c was 13.2% (4.0–6.0%), alkaline phosphate was 115 IU/L (35–129 IU/L), aspartate transaminase was 71 IU/L (5–37 IU/L), and alanine transaminase was 166 IU/L (5–41 IU/L). Computed tomography with a pancreatic protocol showed a 1.7 cm  $\times$  1.5 cm solid appearing mass in the body of the pancreas (Figure 1). Magnetic resonance imaging of the abdomen without contrast confirmed a 1.5 cm solid lesion in the body of the pancreas and a 5 mm central calcification in the mass. A few mildly prominent lymph nodes were seen in the porta hepatis.

An endoscopic ultrasound showed an 18.2 mm  $\times$  15.4 mm hypoechoic, homogeneous, round mass with central calcification and acoustic shadowing near the genu/proximal body of the pancreas (Figure 2), and fine needle biopsy was obtained. Pathology showed an admixture of solid and pseudopapillary areas forming fibrovascular stalks and rosette-like structures;

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**Figure 1.** Computed tomography with a pancreatic protocol showed a mass at the pancreatic body measuring 1.7 cm × 1.5 cm with a central calcification.

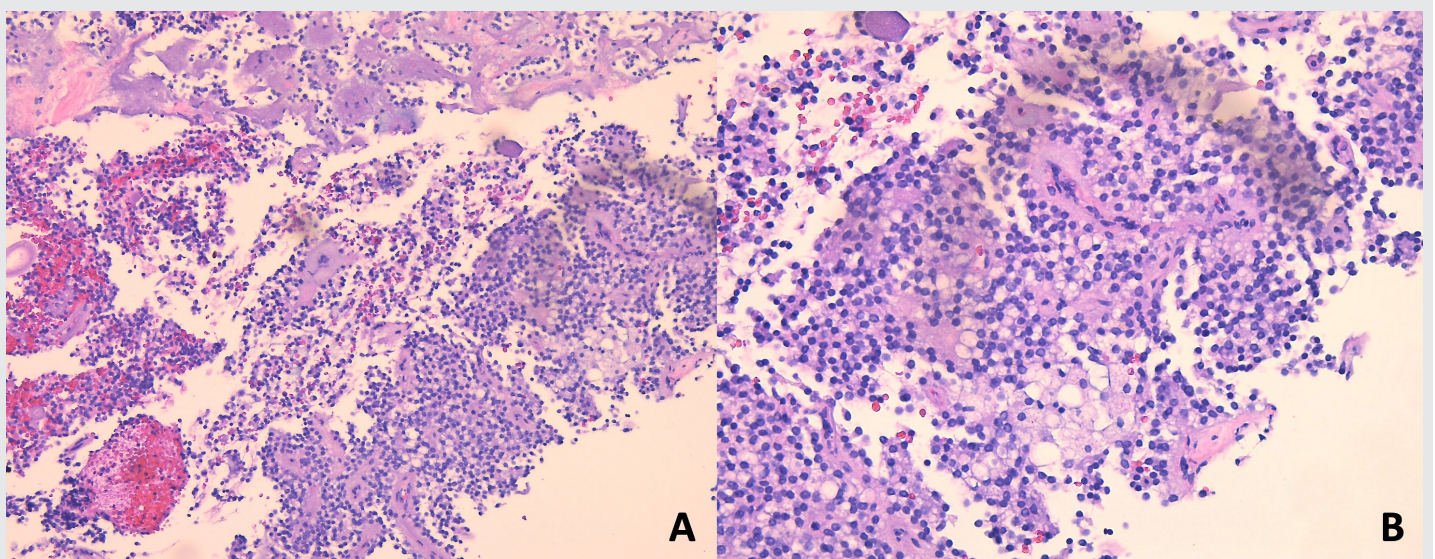


**Figure 2.** Endoscopic ultrasound showed an 18.2 mm × 15.4 mm hypoechoic, homogeneous, round mass with central calcification and acoustic shadowing near the genu/proximal body of the pancreas.

stroma showed various degrees of hyalinization and evidence of degeneration, foamy macrophages, and calcification (Figure 3A and 3B). Tumor cells stained positive for beta-catenin, androgen receptor, CD56, CD10, focally positive for pancytokeratin and synaptophysin and stained negative for E-cadherin and CD45 (Figures 4A and 4B). The morphology and

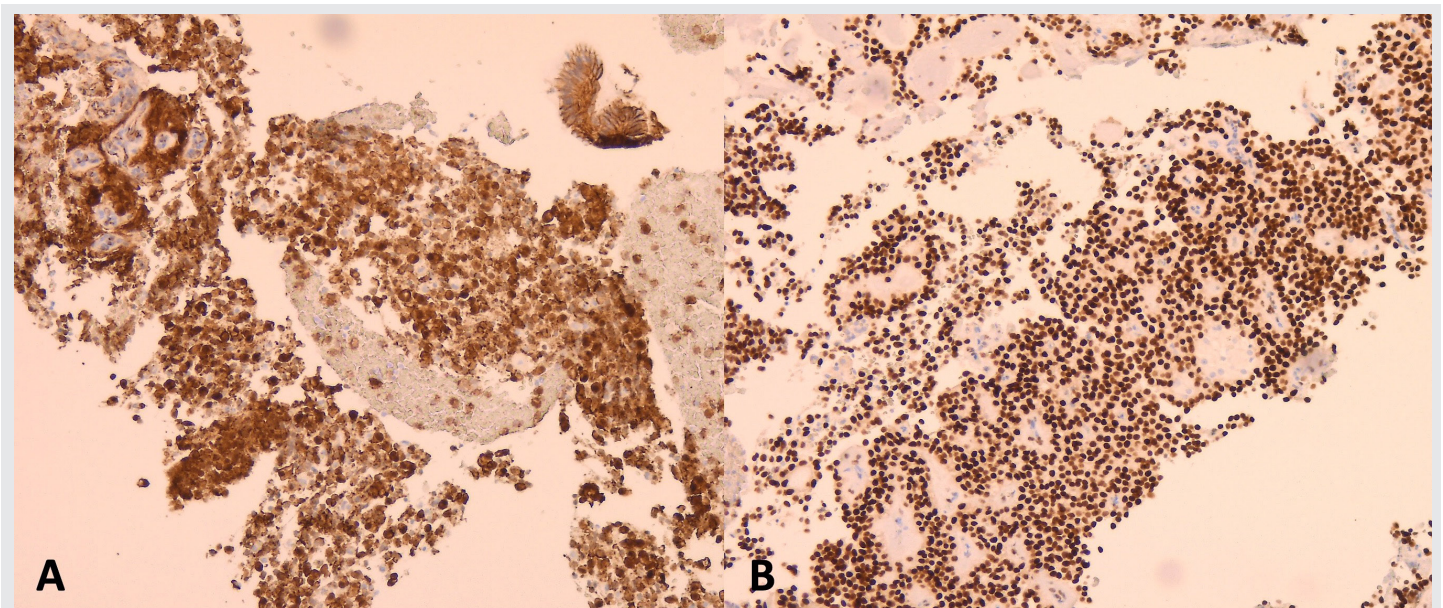
immunophenotype on pathology were most consistent with solid pseudopapillary neoplasm of the pancreas.

The patient was referred to hepato-pancreato-biliary surgery for surgical evaluation. With uncontrolled hypertension and diabetes, he was considered high risk for surgery at this time. He is currently managed conservatively with annual imaging surveillance.



**Figure 3.** (A) Hematoxylin and eosin stain 10× and (B) Hematoxylin and eosin stain 20× show admixture of solid and pseudopapillary areas (tumor cells getting detached from blood vessels forming fibrovascular stalks/rosette-like structures). (A) Stroma shows various degrees of hyalinization, evidence of degeneration, and hemorrhage.





**Figure 4.** (A) shows the immunohistochemical stain beta catenin with aberrant nuclear expression (B) shows the immunohistochemical stain androgen receptor with strong nuclear positivity.

## DISCUSSION

Pancreatic solid pseudopapillary neoplasms (SPN) are rare exocrine pancreatic tumors that account for 1–3% of all pancreatic tumors and were first described by Frantz in 1959.<sup>1,2</sup> These neoplasms have a female predominance with a female to male ratio of 10:1. In addition, they primarily occur in Asian and African American women who present at a younger age than men, with the mean ages at presentation being 25 and 35, respectively.<sup>3,4</sup> Typical clinical manifestations include vague abdominal pain (40%) and a palpable abdominal mass (33%). Some patients may have poor appetite and nausea, which may be secondary to compressive effects of the tumor on the stomach and adjacent organs.<sup>5,6</sup> However, 20% of females and up to 40% of males are asymptomatic.<sup>5</sup> An SPN is a rare occurrence in men. The common location of the mass in male patients is in the body-tail region of the pancreas, which accounts for up to 65% of the total cases; the location in females usually involves the pancreatic head.<sup>6</sup>

A pancreatic SPN is detected by imaging incidentally in asymptomatic patients. Multiple imaging

modalities for SPN can be used. Abdominal ultrasound shows a hypoechoic, clear-bordered cystic or cystic-solid mass. Computed tomography and magnetic resonance imaging of the abdomen describe a large well-circumscribed lesion with heterogeneous density and solid and cystic components.<sup>5,7</sup> Endoscopic ultrasound-guided fine needle aspiration with biopsy (FNA/B) is diagnostic.<sup>8</sup>

The tumor markers for pancreatic cancer are usually negative in patients with pancreatic SPN. These markers include the carcinoembryonic antigen (CEA), serum cancer antigen (CA) 19-9, and serum CA 72 and are elevated in less than 10% of patients with SPN. Therefore, no tumor markers or specific laboratory tests have been established for diagnosing pancreatic SPN to date. The definite diagnosis is based mainly on imaging studies and pathological tissue analysis from FNA/FNB specimens.

The risk of malignant potential is estimated to be 15%. The tumor can be locally aggressive and extend into adjacent blood vessels and organs. There is potential for local recurrence and distant metastasis.<sup>8</sup> Distant metastases were reported in about 4% cases

at the time of diagnosis. According to a multicenter study by Matos, surgical resection is the treatment of choice for pancreatic SPN.<sup>9</sup> Patients have an excellent prognosis with a 5-year survival rate as high as 97% in patients undergoing surgical resection and a 10-year survival rate at 96%.<sup>6,10,11</sup>

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