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Solid Pseudo-papillary Neoplasia: A Rare Malignancy of the Pancreas

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Abstract

Solid pseudopapillary neoplasms (SPNs) are exceedingly rare type of exocrine pancreatic malignancy, representing only 0.9%–2.7% of all exocrine pancreatic malignancies. They predominantly affect young women and unlike other pancreatic malignancies, they have excellent prognoses with 5-year survival following surgical resection approaching 97%. Given the rarity of the disease, little is known about their histopathogenesis as they do not harbor similar genetic mutational abnormalities like other pancreatic tumors. We describe a rare case of SPN in a young female who was found to have the rare diagnosis during the work up for deranged liver function tests.

Keywords: Solid pseudopapillary neoplasm, Pancreas, Exocrine malignancy, Low-grade tumors, Rare tumors

1. Introduction

Solid pseudopapillary neoplasms (SPNs) are exceedingly rare type of exocrine pancreatic malignancy,¹ representing only 0.9%–2.7% of all exocrine pancreatic malignancies.² They are classified as “low-grade,” malignant pancreatic tumors by the World Health Organization (WHO)³ and predominantly afflict young women with a female to male ratio of 9.8:1⁴ and a mean age of presentation of 28.5 years.⁵ In contrast to typical pancreatic malignancies, SPNs do not harbor similar genetic signatures and their histopathogenesis remains relatively unknown.⁶ Unlike other pancreatic malignancies, they also have a better prognosis with overall 5-year survival approaching 97% following surgical resection.⁶

We describe a case of a young woman who was diagnosed with SPN following evaluation of derangements in liver function tests.

2. Case report

A 22-year-old female presented to the emergency department (ED) for lightheadedness and a near

syncopal episode on the morning of the presentation. The episode of lightheadedness was gradual in onset, progressive, aggravated by standing or walking, and alleviated by lying down. She reported usual health until four days before presentation at which time she developed a sore throat that progressed to a dry cough. She also endorsed fatigue and myalgias but denied fevers, chills, sick contacts, new exposures, recent travel, or change in medications. She denied any recent change in weight or a history of eating disorders. Medical history was notable for history of polycystic ovarian syndrome, menorrhagia complicated by iron deficiency anemia, and World Health Organization (WHO) class III obesity. She was unvaccinated against COVID-19. Her only home medication included oral contraceptive pills (OCP).

On presentation to the ED, she was afebrile but slightly hypotensive (97/57 mmHg) with a preserved heart rate (81 beats per minute) and normal oxygen saturation on room air. Her weight was 90 kg; unchanged from prior. Physical examination was unremarkable. Laboratory diagnostics revealed an unremarkable complete blood count with a normal differential. Her coagulation profile was

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unremarkable. Metabolic panel, however, was remarkable for elevations of aspartate aminotransferase ((AST) 244 units/L (reference range: 14–36)) and alanine aminotransferase ((ALT) 211 units/L (reference range: 0–34)) with a normal total bilirubin (0.5 mg/dL (reference range: 0.2–1.3)) and alkaline phosphatase ((ALP) 92 units/L (reference range: 38–126)). Urine pregnancy test was negative. The patient's COVID-19 polymerase chain reaction (PCR) came back positive, and she was administered fluids and monoclonal antibodies. Her lightheadedness resolved; it was believed to be in the setting of orthostasis.

The patient had had a similar presentation a year prior when she had presented to the ED of another hospital with similar complaints and had been found to be hypotensive (78/36 mmHg) with a preserved heart rate (81 beats per minute). Laboratory diagnostics had revealed a normocytic anemia (hemoglobin (Hb) 9.8 g/dL (reference range: 12.0–16.0); mean corpuscular volume (MCV) 80.3 FL (reference range: 80.0–99.0)) with an elevated red blood cell distribution width ((RDW) 14.7% (reference range: 11.5–14.5)) and a low hematocrit ((Hct) 32.6% (reference range: 37.0–47.5%)). During that presentation, the metabolic panel was remarkable for mild transaminitis (AST: 62 IU/L; ALT: 56 IU/L) with a normal total bilirubin (0.3 mg/dL) and ALP (88 units/L). COVID-19 testing was negative. On that presentation, orthostatic vitals were positive, and she received IV fluids with resolution of her symptoms. She was counseled to get an ultrasound of the liver, but was lost to follow up.

A right upper quadrant ultrasound was then performed on an outpatient basis, ordered by her primary care physician, and demonstrated a hypoechoic mass within the pancreatic head measuring 2.1 × 2.1 × 2.7 cm. Magnetic resonance imaging (MRI) of the abdomen with and without contrast corroborated the ultrasound findings, demonstrating a T2 hyperintense mass with central scarring at the head of the pancreas with enhancement following contrast administration (Fig. 1). There was no pancreatic ductal dilatation. Endoscopic ultrasound with core biopsy was performed and confirmed the presence of a rounded hypoechoic mass within the genu. Histopathology demonstrated friable red-brown soft tissue that stained positive for β -catenin and progesterone receptor (PR) with patchy synaptophysin positivity (Fig. 2). The tumor cells were negative for chromogranin. The patient was diagnosed with solid pseudopapillary neoplasm of the pancreas and underwent pancreaticoduodenectomy.

3. Discussion

In this report, we describe a case of a rare type of exocrine pancreatic tumor - solitary papillary neoplasm - in a young female who had initially presented to the emergency department due to symptoms pertaining to orthostasis and found to have deranged LFTs. Indeed, SPNs have a predilection for young women, although cases have been described in patients ranging in age from 2 to 85 years.⁴ There is no known ethnic or racial predisposition for the tumor. The etiology of SPNs remains unknown and there have been no known risk-factors for the tumor,¹ though there appears to be a cluster of cases associated with familial adenomatous polyposis.⁷ They are not associated with any functional endocrine syndromes.³

Medical literature surrounding SPNs is based on scattered case series with sparse systematic reviews. The most common presenting symptoms were upper abdominal pain, bloating, and mass sensation.⁶ About one third of the patients were asymptomatic and diagnosed incidentally.⁵ Additional symptoms include weight loss, fever, jaundice, nausea, vomiting, or fatigue. In the case of our patient, the tumor size was too small to have caused the symptoms through obstruction of blood flow as the ALP was not elevated. Her symptoms were most likely in the setting of her menorrhagia compounded further by the COVID-19 infection. There are rare case reports of SPNs presenting with traumatic⁸ or spontaneous⁹ rupture leading to hemoperitoneum, with rupture more common in the pediatric population.⁶ Rarely, they may present with metastases; the most common site being the liver.¹⁰

The initial modality of diagnosis is imaging—the most common gross finding of SPN is an encapsulated mass with clear demarcation from the pancreas, containing solid and cystic components.¹¹ Contrast enhanced computed tomography (CECT) provides better delineation of solid and cystic components and aids in capsule definition.¹² An incomplete capsule may be predictive of malignant potential.¹¹ There may be complete or incomplete encapsulation of the mass with presence of both solid and cystic areas. Calcifications, representing solid areas, are usually peripheral while hemorrhagic degeneration, representing cystic areas, are central. There is weak arterial enhancement but strong portal-venous enhancement of the solid component on CECT.¹³ On MRI, T1-and T2-weighted images have heterogenous signal intensity because of the presence of hemorrhagic degeneration. There is also post-contrast enhancement on T2-

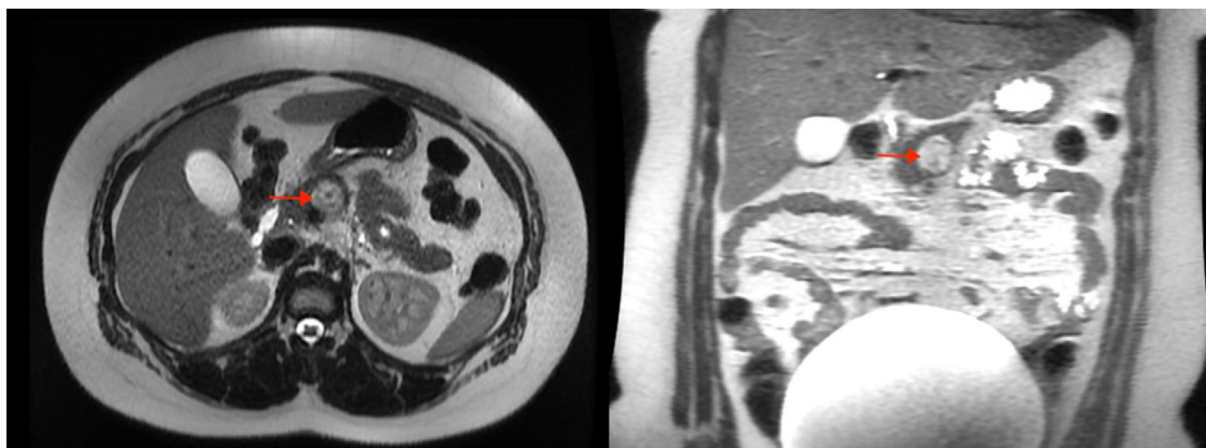


Fig. 1. MRI with and without contrast showing T2 hyperintense mass with enhancement following contrast administration (red arrows).

weighted images. Ultrasonography shows hypoechoic lesions. Endoscopic ultrasound-guided biopsy is usually performed in most cases to establish a diagnosis,¹⁴ as in the case presented. In a systematic review, the most common site of tumor location was the body/tail of pancreas (59.3%) followed by the head of the pancreas (36.0%); 1.1% cases of SPN were extra-pancreatic.⁵ 15% of the tumors may metastasize, are usually confined to the liver, and are synchronous.¹⁵

Histology with immunohistochemistry (IHC) is necessary to confirm the diagnosis of SPNs. Histologic examination shows mixed solid and pseudo-papillary components. The solid component is comprised of poorly cohesive cells whereas the pseudo-papillary component corresponds to monomorphic, discohesive neoplastic cells after detachment from fibrovascular stalks.¹⁶ Although not always present, single cells with cytoplasmic tails - cercariform cells - are highly suggestive of SPNs.¹⁷

On IHC, β -catenin (nuclear or cytoplasmic) is characteristic, similar to the case presented. They may also be positive for CD56, PR, CD10, vimentin, α -1-antitrypsin, claudin5, and cyclin D1.¹

Management of SPNs is focused on primary surgical resection depending on the location of the tumor. Pancreaticoduodenectomy is indicated for SPNs affecting the head of the pancreas, whereas distal pancreatectomy with or without splenectomy (preferably through a laparoscopic approach) is indicated for tumors in the body or tail of the pancreas.^{1,6} When feasible, surgical removal is indicated even in the setting of metastatic disease.¹⁵ The overall prognosis of SPN is excellent with overall 5-year survival rates approaching 97%.⁶ There is a dearth of evidence in the utility of tumors markers to predict prognosis.⁶ The recurrence rate was 4.4% with mortality due to SPN in 1.5%.⁵ There is insufficient evidence regarding the use of chemotherapy in patients who were not surgical candidates.⁶

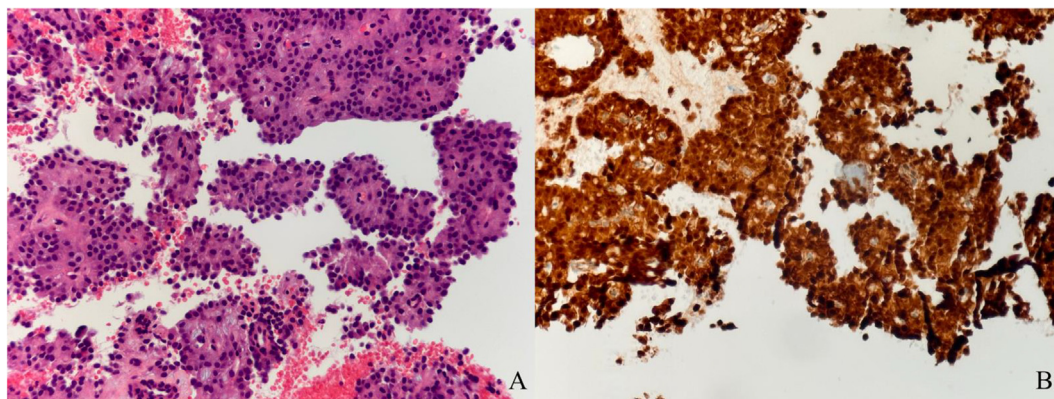


Fig. 2. Panel A. Hematoxylin and Eosin stain—40X magnification. Solid sheets of tumor cells are characteristically discohesive, causing them to fall apart at the periphery of their fibrovascular cores and imparting a pseudopapillary (or finger-like) appearance. Panel B. β -catenin immunohistochemical stain—40X magnification. Nuclear beta-catenin staining is highly characteristic of SPN.

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Conflict of interest

The authors have no conflicts of interest to disclose.

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