

CASE REPORT

Metastatic Solid Pseudopapillary Tumor of the Pancreas: Diagnostic Challenges in the Setting of Abdominal Trauma

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ABSTRACT

Solid pseudopapillary tumor of the pancreas is a rare neoplasm of the exocrine pancreas that predominantly affects young females. Patients may be asymptomatic or present with non-specific symptoms. Asymptomatic patients are diagnosed with an incidental finding on imaging obtained for other reasons. This entity comprises only 0.3%–2.7% of pancreatic cancers; therefore, diagnosis requires a high level of suspicion. Solid pseudopapillary tumor of the pancreas consists of a well-circumscribed, heterogeneous mass that contains solid and cystic components with some foci of calcification, and is usually located in the distal pancreas. Diagnosis may be confirmed with preoperative biopsy. Complete surgical resection is the standard of care and carries an excellent prognosis, even in the setting of large tumors or advanced disease.

INTRODUCTION

Solid pseudopapillary tumor (SPT) of the pancreas, also known as Gruber-Frantz tumor, is a rare neoplasm of the exocrine pancreas that accounts for approximately 0.3%–2.7% of all pancreatic tumors [1, 2]. This entity predominantly affects young females, with a male to female ratio of 1:8–1:10. The median age of diagnosis is 20–30 years old [2, 3, 4].

Patients with SPT typically have non-specific symptoms, which can include anorexia, weight loss, abdominal pain, and/or a palpable abdominal mass [1, 5]. Patients may be asymptomatic, in which case, tumors are incidentally found when imaging studies are motivated by another process, such as abdominal trauma [6].

This neoplasm is often localized to the distal pancreas and consists of a large mass with cystic, solid, and necrotic components surrounded by a fibrinous pseudocapsule [7]. Diagnostic imaging consists of abdominal ultrasound, computed tomography (CT), and magnetic resonance imaging (MRI). On CT, SPTs appear as a well-defined heterogeneous masses with central cystic and peripheral solid areas [7]. Calcifications within the tumor have also been described.

SPTs are low-grade malignant tumors with a favorable prognosis following complete surgical resection. Malignant behavior can occur and is generally associated with neurovascular invasion, peripancreatic infiltration and nodal or distant metastases [2, 3, 8].

We present the case of a teenage girl who was initially diagnosed with a peripancreatic hematoma after sustaining a blunt abdominal trauma. She was later diagnosed with SPT, when complaints of an abdominal mass prompted repeat imaging and subsequent biopsy.

CASE REPORT

The patient was a 15 year-old female with history of blunt abdominal trauma, who presented to her primary care physician for evaluation of a new complaint of a non-tender abdominal mass. She had suffered a blunt abdominal trauma almost 2 years prior, when she fell while ice-skating and hit her abdomen with the upper bar of a skating trainer. A CT scan obtained at that time revealed hemoperitoneum and large left upper quadrant hematoma with active extravasation, possibly arising from the pancreas (**Figure 1**). Interventional radiology performed a visceral angiography that showed frank extravasation from the left inferior phrenic artery, which was subsequently embolized. Magnetic resonance cholangiopancreatography was obtained to evaluate for ductal involvement, and showed a large hematoma with effacement and inferior displacement of the pancreatic body and tail, associated with mild ductal dilatation. The patient did well after embolization and was discharged home on hospital day seven.

The patient noticed a non-tender mass located in the left lower abdomen 2 years later. An abdominal ultrasound showed at least five left-sided abdominal masses and a

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right ovarian mass. An abdominal CT scan demonstrated interval decrease in the size of a heterogeneous lesion within the pancreatic tail, with peripheral solid components and calcifications. Additional lesions were noted bilaterally in the adnexa, the caudate lobe of the liver, and mesentery. Presentation raised concern for SPT with intraperitoneal seeding (**Figure 2**). Percutaneous ultrasound-guided fine-needle aspiration (FNA) biopsy of an omental nodule confirmed the diagnosis. The neoplasm was positive for CD56, β -catenin, cyclin D1, neuron-specific enolase, and progesterone, with low Ki67 expression.

The patient underwent an exploratory laparotomy, distal pancreatectomy and splenectomy, omentectomy,

resection of multiple intra-abdominal tumors, and right ovarian cystectomy (**Figure 3**). The large pelvic mass that was felt to be an ovarian lesion was, in fact, involving the omentum. The right ovarian mass was an ovarian cyst; therefore, a cystectomy was performed for ovarian preservation. Pathology confirmed SPT measuring 9.8 cm in diameter. There was lymphovascular invasion, 4 out of 22 positive lymph nodes, and metastases to the peritoneum and omentum. Staging was pT3, N2, M1. The patient had an uneventful post-operative course and was discharged on post-operative day 7. She is being followed closely with repeat CT scans every 3 months for the first 1-2 years, then at increased intervals. Currently the patient has no evidence of recurrent disease.

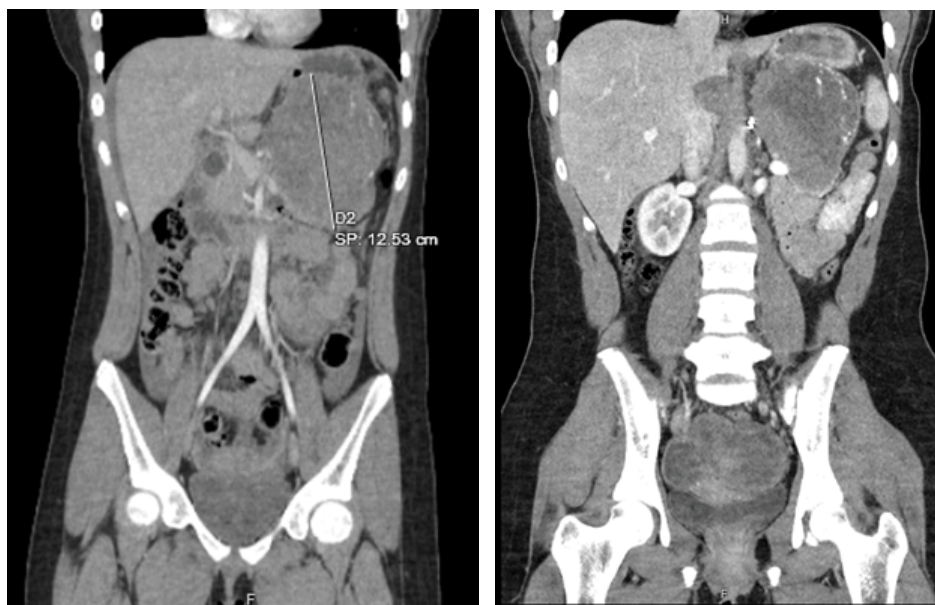


Figure 1. Left: CT scan obtained after abdominal trauma showing large hematoma with effacement of pancreatic tail (arrow). Right: CT scan two years after blunt abdominal trauma, showing a large heterogeneous mass with calcifications (bold arrow) and smaller perihepatic (arrow) and pelvic lesions (curved arrow).



Figure 2. Solid pseudopapillary tumor (circled) seen in surgical specimen of distal pancreatectomy and splenectomy with smaller omental lesions (arrows).

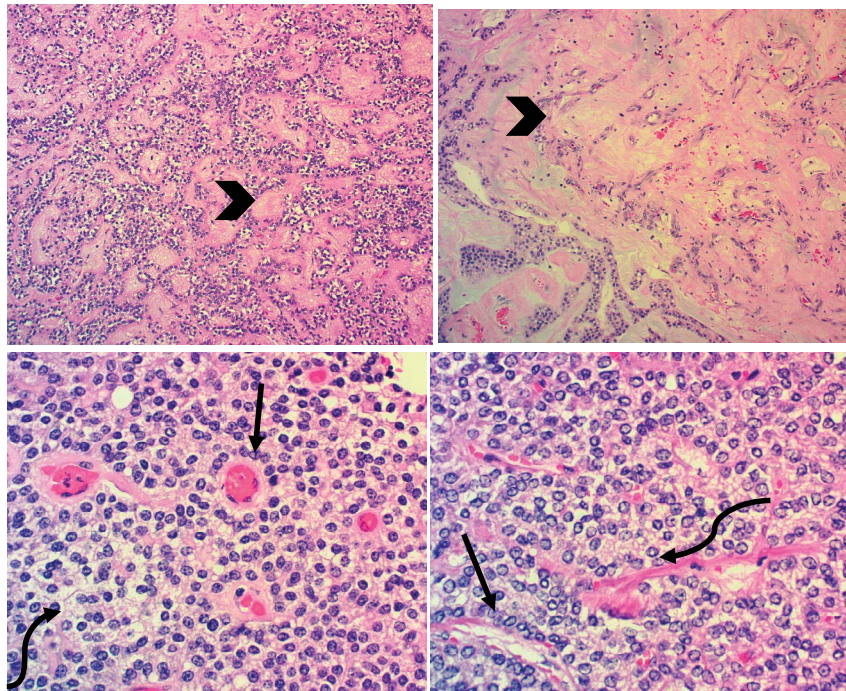


Figure 3. Histopathology of tumor specimens. Stroma with varying degrees of hyalinization (arrow heads) and characteristic pseudopapillary appearance. Solid nests of uniform round cells which surround blood vessels which creates the pseudopapillary architecture (straight arrow). Eosinophilic cytoplasm with perinuclear vacuoles (curved arrow).

DISCUSSION

Solid pseudopapillary neoplasm of the pancreas is a rare entity that characteristically presents in young women. This neoplasm was first described by Frantz in 1959, and has been depicted with different terms in the literature, until the World Health Organization renamed it as SPT in 1996. More recently, the WHO reclassified it from a borderline to a low-grade malignant epithelial neoplasm [3, 8].

Its incidence varies among different series, representing 0.3% to 2.7% of pancreatic cancers [1, 4, 9, 10]. The majority of cases have been reported in the last two decades, an observation that can be explained by the adoption of a defined nomenclature, the advancement of imaging studies and the increased awareness of this condition [1, 9]. It mainly affects females in the 3rd and 4th decades of life, although 25% of cases present in children [1, 4, 9, 10]. The incidence is higher in individuals of Asian and African-American descent [1, 4].

Symptomatic patients usually present with non-specific complaints, most commonly with abdominal pain or discomfort. Many of them are asymptomatic and diagnosed after an incidental finding on imaging. Other complaints are palpable abdominal mass, weight loss, vomiting, jaundice, and hematemesis [1, 4, 9]. Rarely, patients present after tumor rupture, usually after sustaining a blunt abdominal trauma [4]. The patient we present noted a mass in the lower abdomen, which prompted further investigation. What was originally identified as a large hematoma in the context of blunt abdominal trauma, was shown to be a mass with intratumoral hemorrhage. As spontaneous rupture is rare, this inciting event likely led to rupture and subsequent intraperitoneal seeding. In retrospect,

the peripancreatic hematoma was larger than would be expected for a low-impact abdominal trauma. A high index of suspicion may have prompted an earlier diagnosis.

MRI and CT are valuable tools in aiding the differential diagnosis, as well as surgical planning. SPTs appear as a well-circumscribed, heterogeneous masses that contain solid and cystic components with some foci of calcification [4, 9]. The differential diagnosis includes pseudocyst, mucinous cystic tumors, serous cystadenoma, microcystic adenoma, islet cell tumor, acinar cell carcinoma, cystadenocarcinoma, pancreaticoblastoma, and vascular tumors [1, 4]. In our patient, active intratumoral bleeding likely obscured the classic features normally seen on CT. The diagnosis can be confirmed with percutaneous FNA or core needle biopsy, as well as endoscopic ultrasound-guided FNA (EUS-FNA) [4, 9]. Virgilio *et al.* advocate for a preoperative diagnosis based solely on imaging due to concerns for tumor rupture and intraperitoneal seeding secondary to percutaneous and EUS-FNA biopsy [11].

Some authors postulate that SPTs originate from primitive pancreatic ductal cells, acinar cells or endocrine cells. Others hypothesize that female predominance is due to the proximity of pancreatic primordial cells to the ovarian primordial cells of the genital ridge [1, 4, 9, 12]. Histologically, SPTs are characterized by pseudopapillary structures made up of poorly cohesive, monomorphic cells and lined by neoplastic cells [4, 9]. After immunohistochemical staining, SPTs tend to have β -catenin activity in both the nuclei and the cytoplasm [7]. There is no clear relationship between pathologic features and clinical behavior.⁴The following have been proposed as indicators of malignant potential: high Ki-67 index, chromosomal abnormalities, nuclear pleomorphism and

nuclear atypia, extensive tumor necrosis, venous invasion, and infiltrative growth pattern [9].

Unlike other pancreatic tumors, SPTs are considered a low-grade entity. Only about 10%–15% of cases are reported to exhibit malignant behavior [9, 10]. Surgical resection is the standard of care in the management of SPTs, because of the excellent prognosis even in the setting of large tumors or advanced disease [1, 4, 10, 12]. According to Vassos [9], tumor enucleation and incomplete resections should be avoided due to the risk of recurrence. Spătaru *et al.* [1] advocate for preservation of pancreatic tissue as able in pediatric patients. Metastases can involve the liver, lymph nodes, peritoneum, and omentum. Extensive lymphadenectomies are not deemed necessary due to the low rate of lymph node metastasis. Chemotherapy, alcohol injection, and radiotherapy are valid options for the management of metastatic disease to the liver, although more recent studies have shown increased survival after metastasectomy [4]. SPT carries a good prognosis, with an overall 5-year survival of up to 97% after surgical resection [9, 10]. Long term surveillance is recommended, since late recurrences have been described [12].

Our patient had SPT of the pancreatic tail, with lymph node invasion and metastatic disease to omentum and peritoneum. She underwent complete surgical resection and at the present time she has no evidence of disease recurrence.

CONCLUSION

Solid pseudopapillary tumor of the pancreas is a rare, low-grade tumor that typically presents in adolescent females. It generally has a favorable prognosis after complete surgical excision. However, the paucity or lack of symptoms makes diagnosis very challenging in a trauma setting and demands a high level of suspicion. Our patient experienced a serious intra-abdominal bleed that did not correlate with the low-impact mechanism of trauma. As it turns out, she had a SPT that ruptured secondary to the blunt trauma, which led to diffuse intra-abdominal seeding. Bleeding from the tumor obscured the classical CT findings associated with SPT. Further work-up and interval imaging could be considered in these scenarios after the acute presentation has been resolved.

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Ethical approval: Not considered research by the Marshfield Health System Institutional Review Board, and therefore, exempt from review and approval.

Conflict of interest

None to report on the part of any author.

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