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Case Report

Solid pseudopapillary tumor of the pancreas: A difficult presentation of an uncommon tumor

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ABSTRACT

We present a case of a solid pseudopapillary tumor of the pancreas incidentally discovered in a child who presented following sports-related abdominal trauma. This case is a unique presentation of an uncommon pancreatic tumor that posed a diagnostic dilemma to radiologists as it appeared as a hematoma by imaging and initial image-guided aspiration. Ultimately, MRI and core biopsy were needed to make the final diagnosis. This entity posed a problem during the differential diagnosis given the appearance and extenuating circumstances.

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Introduction

Solid pseudopapillary tumor is a mixed solid and cystic neoplasm of the pancreas [6]. It is an uncommon pancreatic neoplasm occurring most commonly in females in the second or third decade of life [2]. It is typically benign and treated with surgical resection [1,2]. They typically appear as well encapsulated pancreatic masses with cystic areas, internal hemorrhage, calcification and necrosis [1,3]. The rarity of this tumor makes arrival at the correct diagnosis sometimes

difficult. Knowledge of imaging features and awareness of this neoplasm can help arrive at the correct diagnosis.

Case report

A 14-year-old previously healthy female presented with abdominal pain approximately 2 weeks following blunt abdominal sports related trauma when she was hit in the abdomen during a basketball game by an opponent's elbow. She

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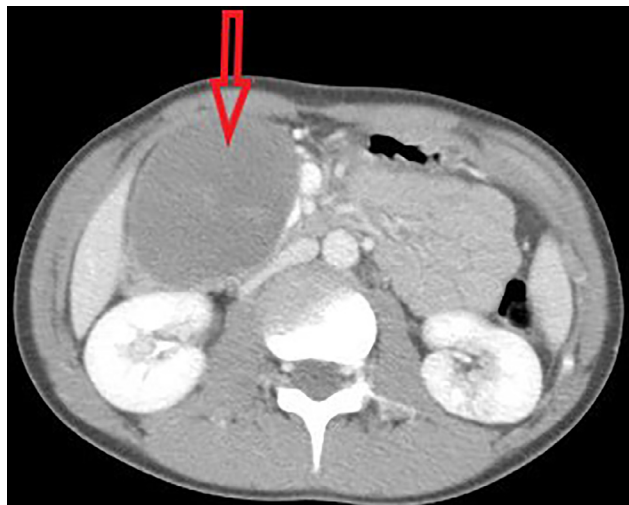


Fig. 1 – Axial contrast enhanced CT image obtained at an outside hospital shows a well-circumscribed low density mass (red arrow) in the right upper quadrant. (Color version of figure is available online.)

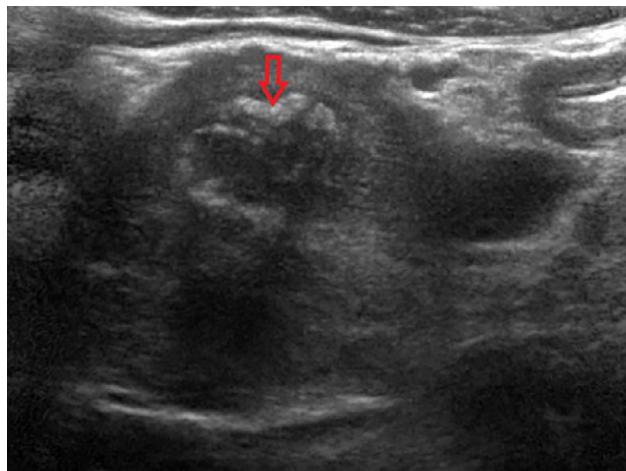


Fig. 2 – Initial US image shows a complex echogenic lesion in the right upper quadrant with areas suggesting calcification (red arrow). (Color version of figure is available online.)

complained of sharp and intense abdominal pain as well as an episode of emesis. An abdominal CT was performed at an outside hospital showing a well-circumscribed cystic mass in the right upper quadrant (Fig. 1). This was thought to be a pancreatic pseudocyst versus evolving duodenal hematoma. She underwent ultrasound-guided aspiration of the lesion, revealing only a small amount of remote blood products. At this point the lesion was felt to be a hematoma. She was discharged but

continued having abdominal pain and subsequently returned to the emergency room the following year. An abdominal US at that time revealed a complex echogenic lesion in the right upper quadrant (Fig. 2). It was uncertain whether it was arising from the pancreatic head or duodenum. The etiology was uncertain and was thought to represent a duodenal hematoma, gastrointestinal duplication cyst or cystic pancreatic lesion. Two additional ultrasound examinations found the lesion to be slightly decreased in size from the initial exam. Etiologies

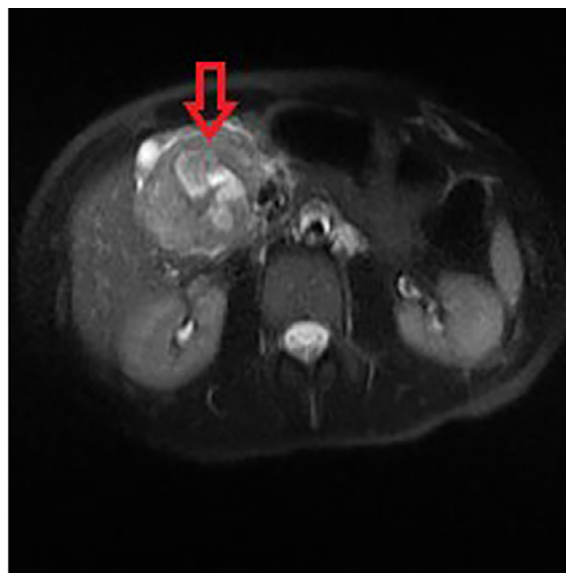
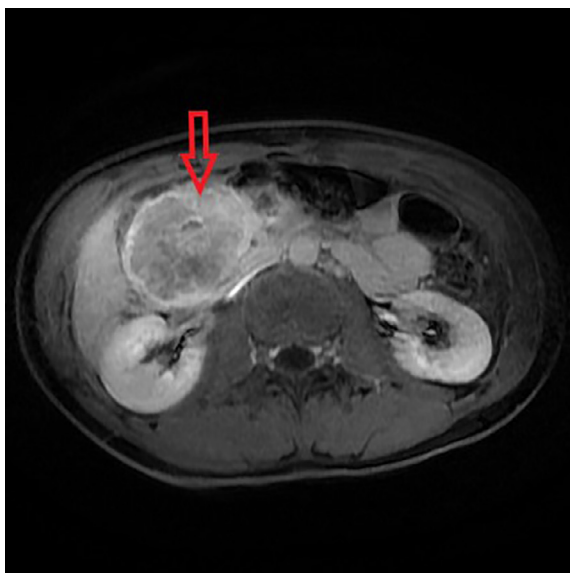


Fig. 3 – Axial contrast enhanced LAVA (left) and Axial T2 SSFSE fat saturated (right) MRI abdomen images (red arrows) show a heterogeneous enhancing mass with areas suggesting cystic change and hemorrhage in the right upper quadrant. (Color version of figure is available online.)

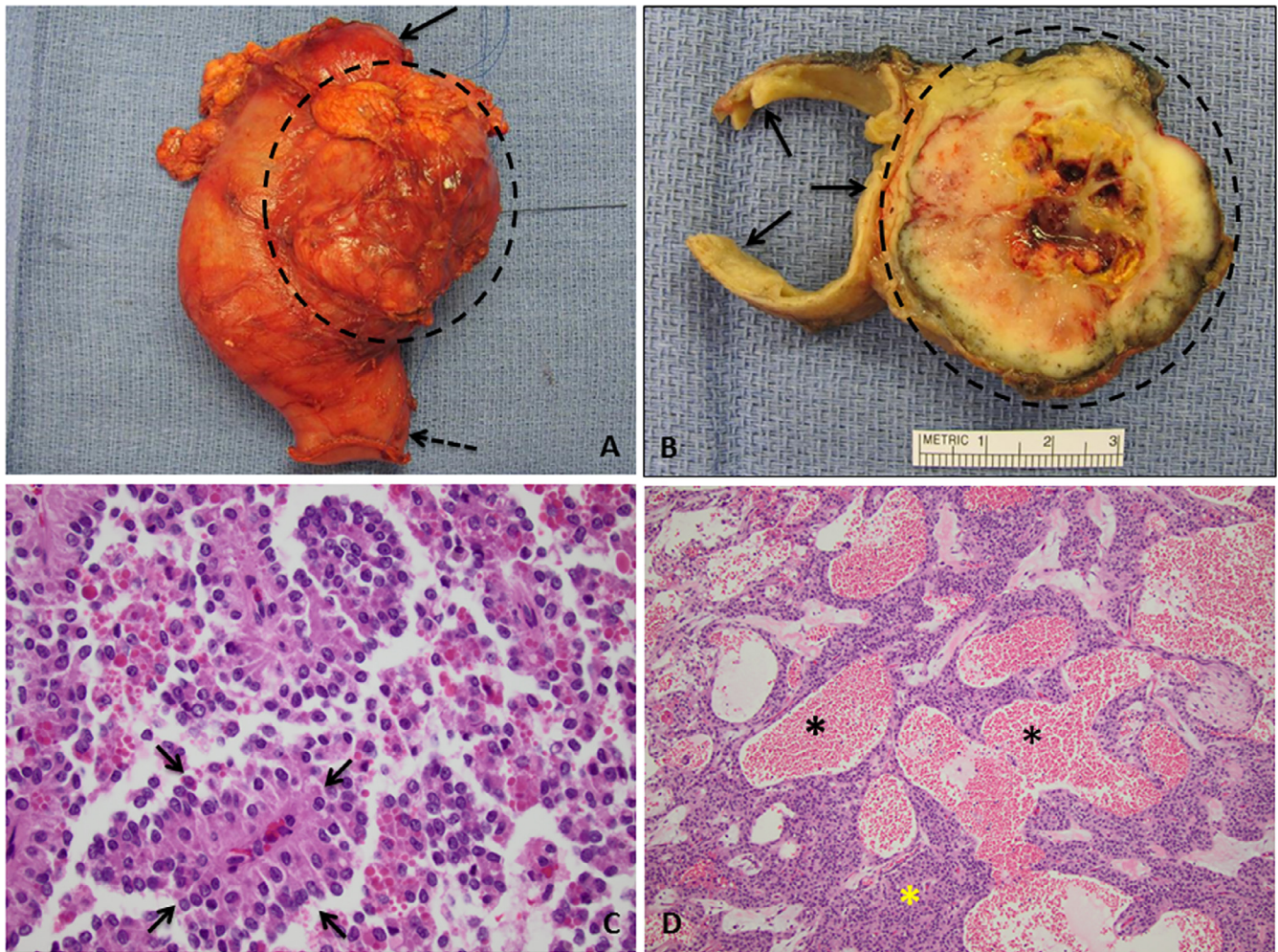


Fig. 4 – (A) Gross picture of the specimen including a segment of duodenum (on the left side of the picture; solid arrow indicating proximal resection end and dashed arrow indicating distal duodenum), and pancreatic head containing the tumor (on the right side of the picture; the tumor is highlighted by a dashed circle). **(B)** Gross picture of the cut section of tumor. The tumor has a solid and cystic appearance with hemorrhage. The tumor measures 6.5 × 4.5 × 4.3 cm and it is highlighted by the dashed circle. The duodenum is located on the left side of the specimen and its luminal surface is highlighted by arrows. **(C)** Microscopic picture of tumor (H&E stain): the tumor shows pseudopapillae with hyalinized fibrovascular cores lined by several layers of bland epithelial cells with eosinophilic cytoplasm and clusters of hyaline globules. The arrows highlight one of the pseudopapillae. **(D)** Microscopic picture of tumor (H&E stain): the tumor contains multiple cystic structures filled by blood content (representative cysts are highlighted by black stars). The yellow star highlights the solid portion of the tumor. (Color version of figure is available online.)

such as organizing hematoma and benign cystic mass were suggested.

The patient then sustained another similar blow to the middle abdomen while playing in a soccer game and again presented with similar abdominal pain and an episode of emesis. A repeat abdominal US revealed a persistent right upper quadrant mass again concerning for an organizing hematoma or a neoplasm. During this hospital course, the patient underwent an MRI examination of the abdomen. This revealed a heterogeneous mass in the region of the pancreatic head and the proximal duodenum with areas suggesting cystic change, hemorrhage and calcification (Fig. 3). At this time, the diagnosis of solid pseudopapillary tumor of the pancreas was suggested with additional considerations of a com-

plex duodenal hematoma, complex pseudocyst or duodenal neoplasm. She then underwent ultrasound guided core biopsy and fine-needle aspiration of the lesion. Pathology revealed fibroconnective tissue with a neoplastic proliferation consisting of fibrovascular cores, myxoid stromal change and surrounding ovoid epithelioid cells. This was PAS-D positive with rare cytoplasmic inclusions. The patient was subsequently diagnosed with a solid pseudopapillary tumor of the pancreas.

The patient subsequently had a pancreaticoduodenectomy (Whipple resection) to remove the tumor including a segment of duodenum and pancreatic head (Fig. 4A). On gross examination and sectioning, there was a well-circumscribed, nonencapsulated, solid and cystic mass (6.5 × 4.5 × 4.3 cm) in the pancreatic head (Fig. 4B). The mass was confined within

the pancreas and did not invade the adjacent duodenum and common bile duct. Microscopic examination revealed pseudopapillae with hyalinized fibrovascular cores lined by several layers of bland epithelial cells with eosinophilic cytoplasm and clusters of hyaline globules (Fig. 4C). Areas of hemorrhage and blood-filled cysts were also present (Fig. 4D). Immunohistochemical stain showed strong nuclear staining. These findings were consistent with a solid pseudopapillary tumor of the pancreas.

Discussion

Solid pseudopapillary tumors of the pancreas are uncommon neoplasms with nonspecific imaging features. On ultrasound, they present as large heterogeneous cystic and solid masses with a thick capsule and peripheral calcifications [3,5]. On computed tomography, they are usually well-defined solid masses with intermixed cystic components and calcifications [3,4]. On T1 weighted MRI, they appear as hypointense masses with areas of increased signal corresponding to hemorrhage and necrosis. There can be a peripheral rim of decreased signal corresponding to a fibrous capsule [3,6]. On T2 sequences, they have heterogeneous signal with enhancement on post-contrast sequences [3,6]. The differential diagnosis of a large pancreatic lesion includes pancreatic pseudocyst, hematoma (especially with a history of trauma such as in our patient), pancreatoblastoma, or less likely pancreatic adenocarcinoma. The patient's clinical course of persistent abdominal pain over several years makes a hematoma less likely. Also, the relative stability over several years makes malignant tumors such as pancreatoblastoma and adenocarcinoma less likely. Additionally, the patient had no history of pancreatitis to suggest that the lesion was a pseudocyst.

Awareness of solid pseudopapillary tumors of the pancreas and their ability to mimic other entities such as a hematoma may help expedite the diagnosis. After initial detection on ultrasound, knowledge of this tumor may lead to prompt workup with MRI and possible tissue typing to make the diagnosis. Diagnosis of this entity is important as surgical resection usually can be curative with a greater than 95%

survival rate [6]. The tumors contain areas of hemorrhage and necrosis so on FNA they may appear as only blood products if no solid elements are obtained. Additionally, given the close proximity to the gastrointestinal tract, these tumors may mimic pathologies such as duodenal hematomas, primary duodenal neoplasms and complex gastrointestinal duplication cysts. While this patient's age and sex were typical of a SPIN tumor (female in the second decade of life), the radiologic appearance was atypical. Although SPIN tumors may have areas of hemorrhage, the tumor in this case presented with an atypically large area of hemorrhage, which obscured the underlying solid components. This appearance combined with the clinical history of abdominal trauma made the presentation atypical and the diagnosis initially elusive.

This case highlights how the imaging findings and clinical presentation of solid pseudopapillary tumors of the pancreas make it a difficult yet important diagnosis to reach. Knowledge that such tumors may mimic other pathologies can lead to a prompt workup and diagnosis.

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