AMSER Rad Path Case of the Month:

13-year-old woman with incidental left upper quadrant abdominal mass

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Patient Presentation

Clinical history

• 13-year-old female with a past history of appendectomy and at that time a mass was incidentally noted in the left upper quadrant. She reported some early satiety. No other clinical symptoms.

Pertinent social history

• None

Pertinent physical exam findings

• Hemodynamically stable
• Abdomen: Soft, nontender. Vague mass palpated in LUQ.
Pertinent Labs

- WBC 12.70
- HGB 7.5 (L)
- HCT 22.1 (L)
- PLT 168
- BMP WNL
- Ca 8.6 (L)
- Glucose 136 (H)
- Amylase 173/101
Radiology Images
Heterogeneous mass arising from the pancreatic tail (circle) possibly with internal calcification (arrow).
Heterogeneous mixed cystic (T2 hyperintense components – long arrows) and solid (enhancing components – short arrows) mass with capsule (dotted arrows) arising from the pancreatic tail (circle). The calcification seen on prior CT is not well seen by MRI technique.
DDX (based on imaging)

- Pancreatic solid pseudopapillary tumor
- Pancreaticoblastoma
- Nonfunctioning islet cell (neuroendocrine) tumor
• OPERATION: Exploratory laparotomy, distal pancreatectomy, splenectomy.

• INDICATIONS FOR PROCEDURE:
• 13-year-old female with mass in the tail of her pancreas; it was felt that resection was indicated. The tumor was large enough that it was compressing the splenic vein with significant left-sided portal hypertension. Because of this concern and the possibility of malignancy, laparotomy and splenectomy considered appropriate.
Well-circumscribed mass with a heterogeneous cystic and solid cut surface. There is an area of necrosis/degeneration with a papillary appearance (white arrow).
Solid growth on top of photo, pseudo-papillary growth on bottom

Nested cells forming perivascular cuffs = pseudo-papillary architecture (black arrows) with stromal degeneration, cells with irregular nuclei, hyaline globules, perinuclear vacuoles (grey arrows).
Beta-catenin positive
Final Dx:

Pancreatic solid pseudopapillary tumor
Solid Pseudopapillary Tumors (SPTs)  
General Information

- Solid pseudopapillary tumor is a rare primary neoplasm of the pancreas, representing only ~1% of all pancreatic tumors.
- Typically affects young women, as in this case (often referred to as the "daughter tumor").
- Frequently asymptomatic or present with a gradually enlarging abdominal mass. Jaundice is a rare presentation.
- Usually characterized by an encapsulated heterogeneous mass, most of which are located in the pancreatic body and tail.
- Has malignant potential and so is usually surgically resected.
Solid Pseudopapillary Tumors (SPTs) Imaging Appearance

**CT findings:**
- Encapsulated heterogeneous mass with varying amounts of solid and cystic components
- Following contrast material administration, enhancing solid areas are typically noted peripherally, whereas cystic spaces are usually more centrally located

**MRI findings:**
- Encapsulated mass with heterogeneous signal intensity on T1- and T2-weighted images, reflecting the complex nature of the mass
- Areas of high signal intensity on T1-weighted images and/or high signal intensity on T2-weighted images can help identify blood products and/or cystic components, respectively
- In differentiating SPTs from islet cell/neuroendocrine tumors, SPTs typically do not demonstrate the hypervascularity typically seen in islet cell tumors
Solid Pseudopapillary Tumors (SPTs)  
Pathologic Appearance

**Gross examination:**
- Large mass (mean maximum dimension, 9.3 cm) and well encapsulated
- Contains varying amounts of necrosis, hemorrhage, and cystic change.

**Microscopic analysis:**
- Two distinct types of cellular arrangements: solid and papillary.
- The hallmark histologic pattern occurs when the tumor cells form papillary configurations composed of a fibrovascular stalk surrounded by several layers of epithelial cells.
- Solid areas containing necrosis, foamy macrophages, cholesterol granulomas, and calcifications may also be seen.

**Immunohistochemistry (IHC):**
- Characteristically positive for: beta-catenin (as in this case), α1-antitrypsin, CD56, CD10, and vimentin.
References:


• Coleman et al. Solid-Pseudopapillary Tumor of the Pancreas. RadioGraphics 2003; 23:1644 – 1648 *Published online* 10.1148/rg.236035006

• Peyman Dinarvand, Jinping Lai. Solid Pseudopapillary Neoplasm of the Pancreas A Rare Entity With Unique Features. Arch Pathol Lab Med—Vol 141, July 2017