AMSER Rad Path Case of the Month:

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

Aditi Chaurasia, Medical Student
All India Institute of Medical Sciences
Kara Lynn Gawelek, MD
Department of Pathology, Brigham and Women's Hospital
Shanna Matalon, MD
Department of Radiology, Brigham and Women's Hospital

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

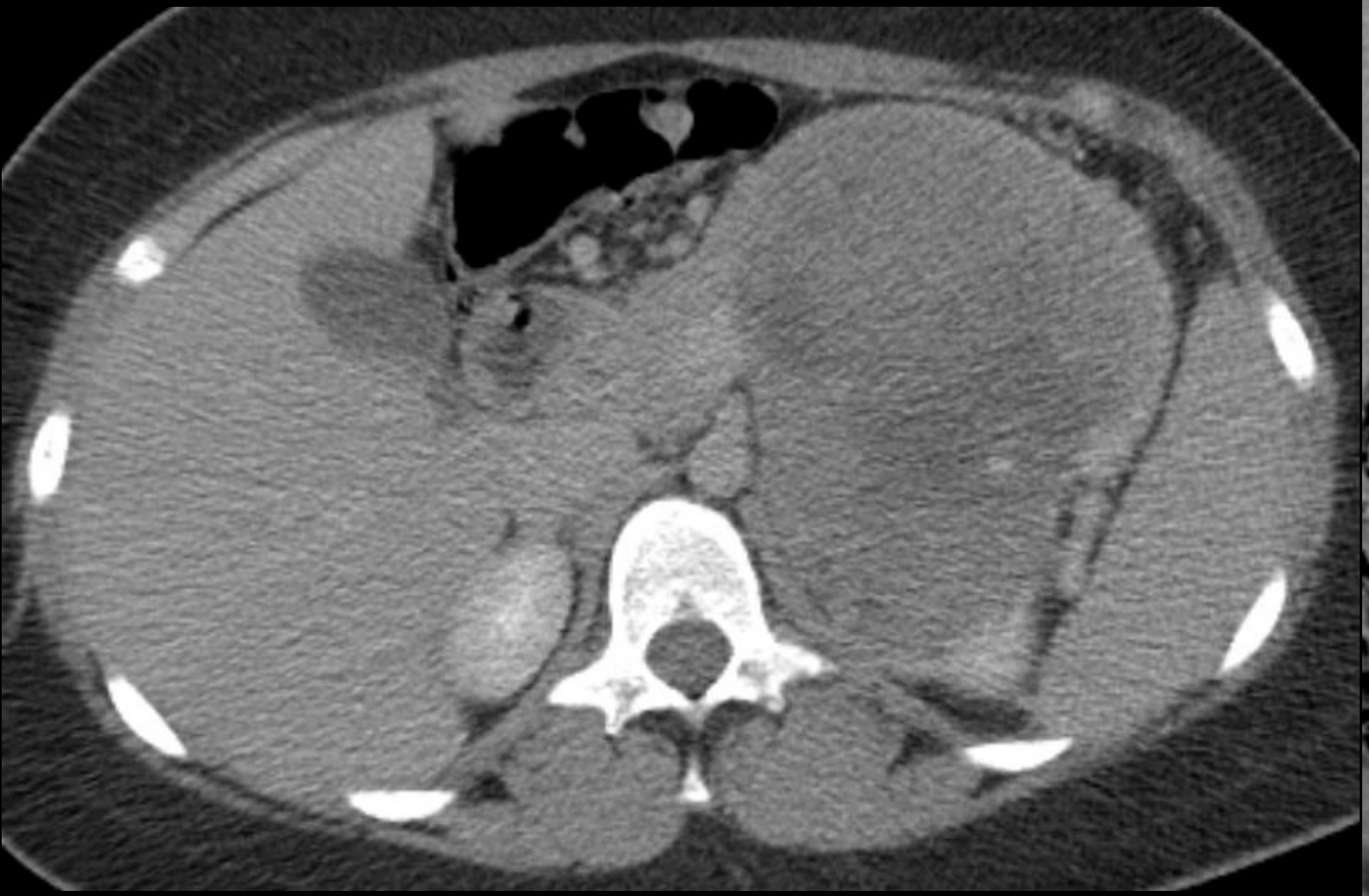
- Hemodynamically stable
- Abdomen: Soft, nontender. Vague mass palpated in LUQ.

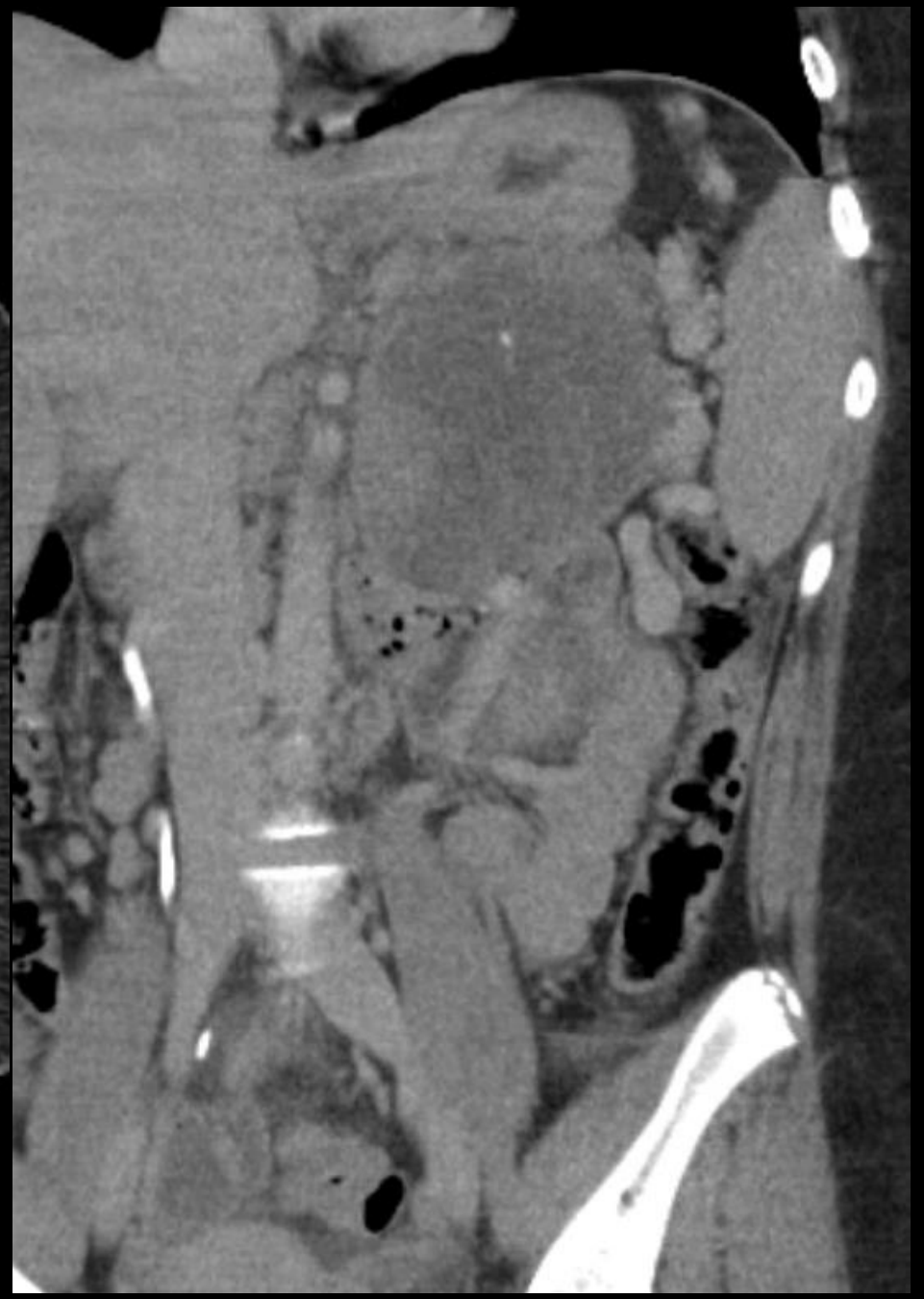
Pertinent Labs

- 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

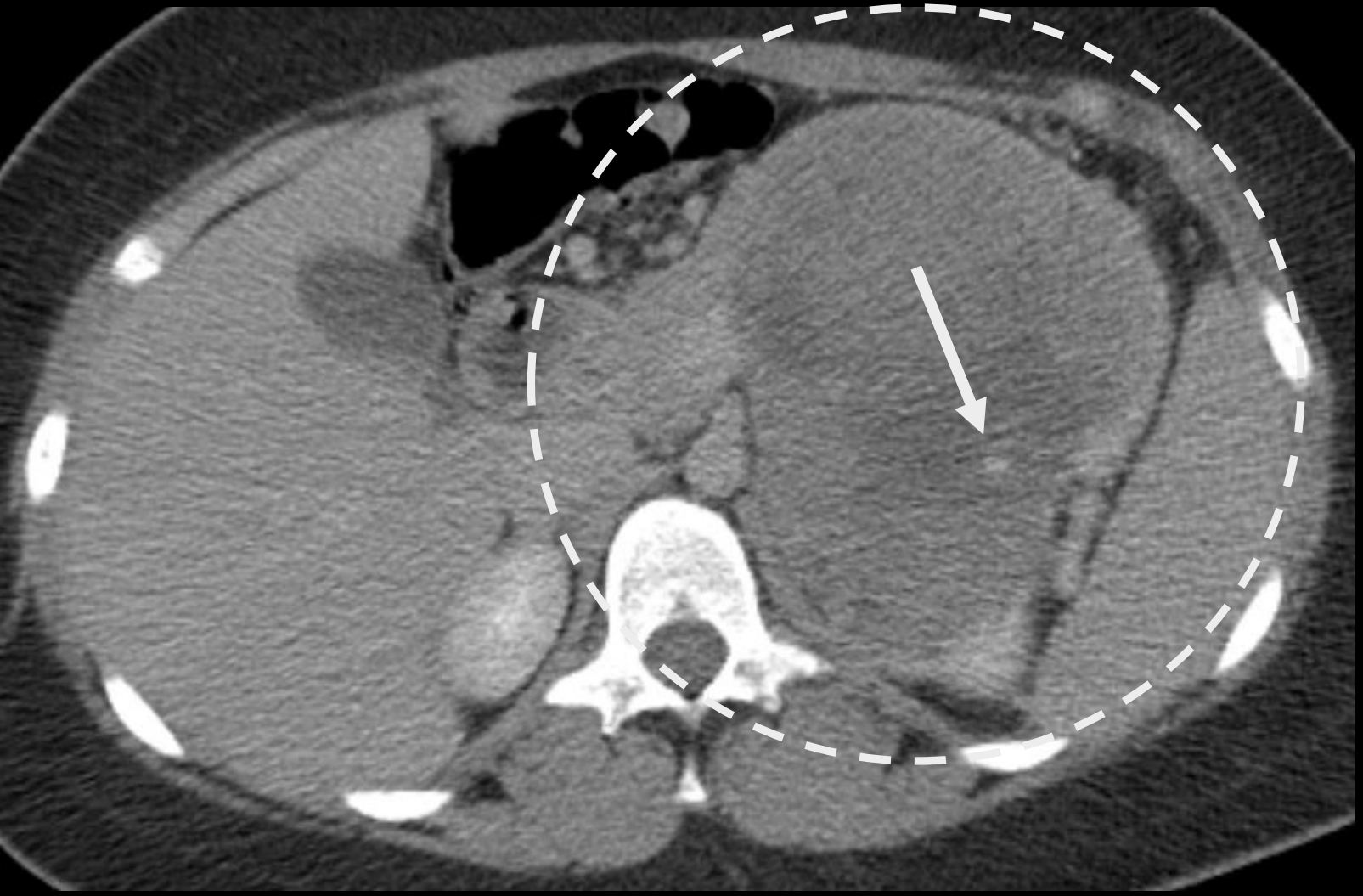
Radiology Images

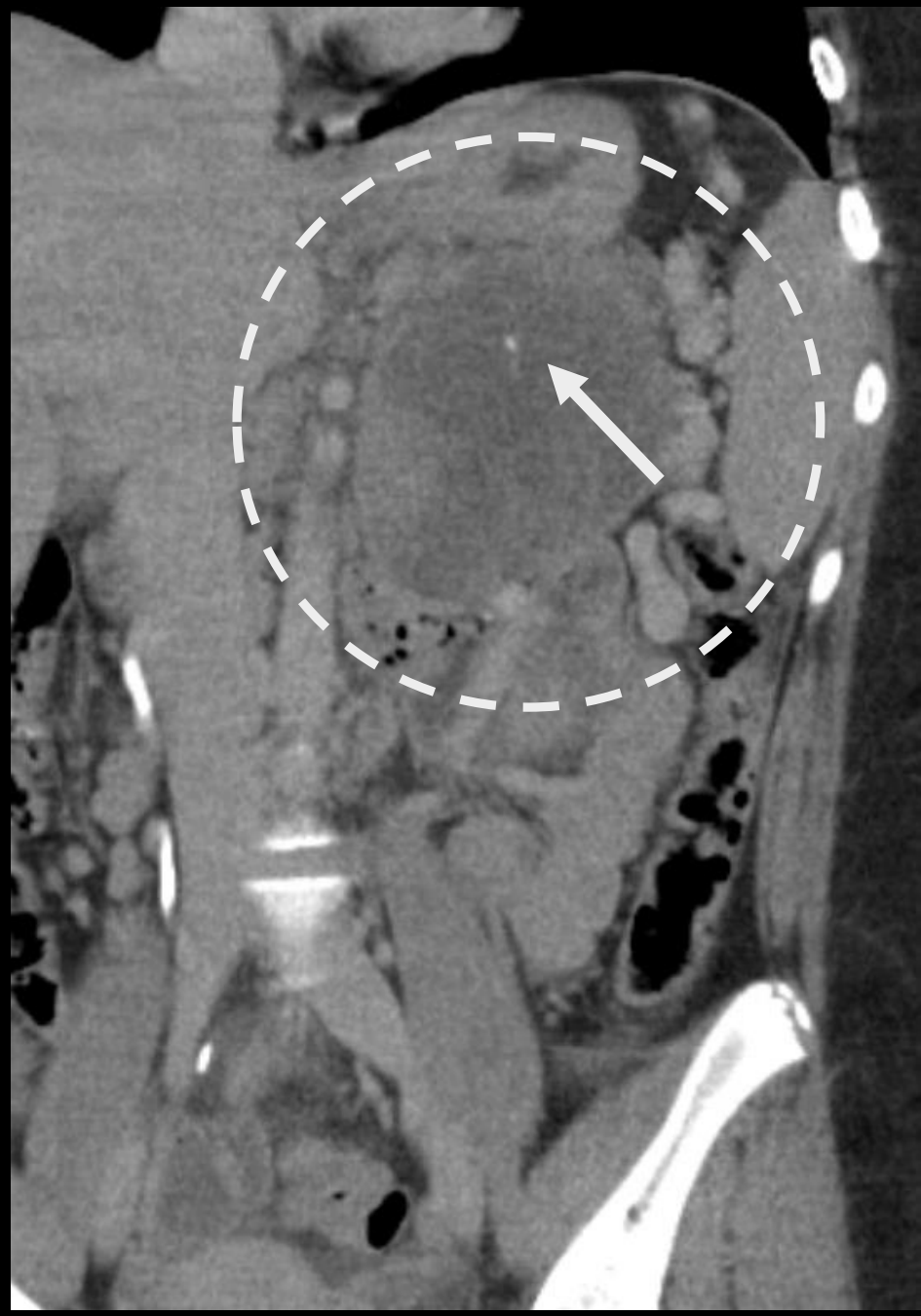
Noncontrast CT



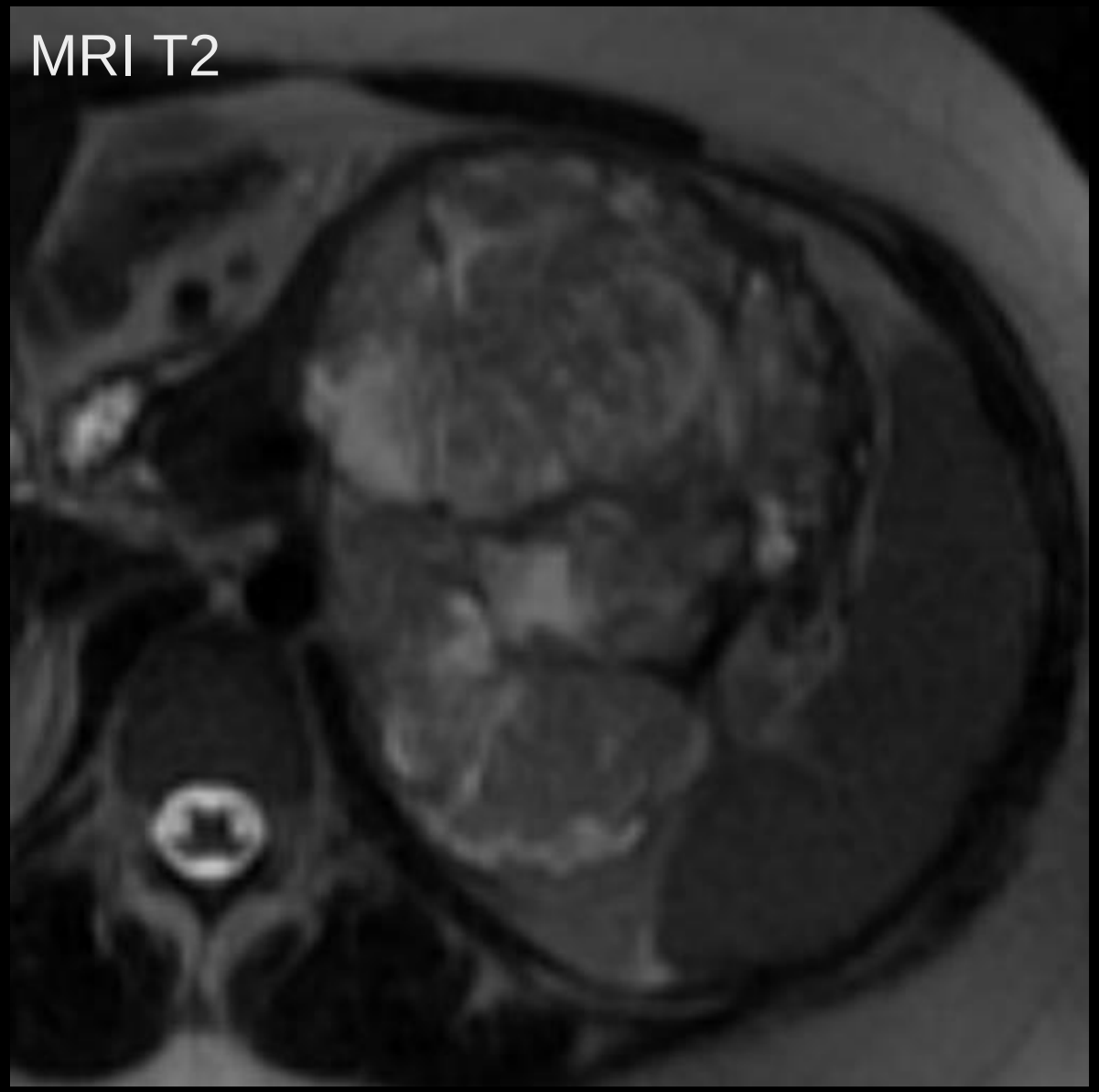


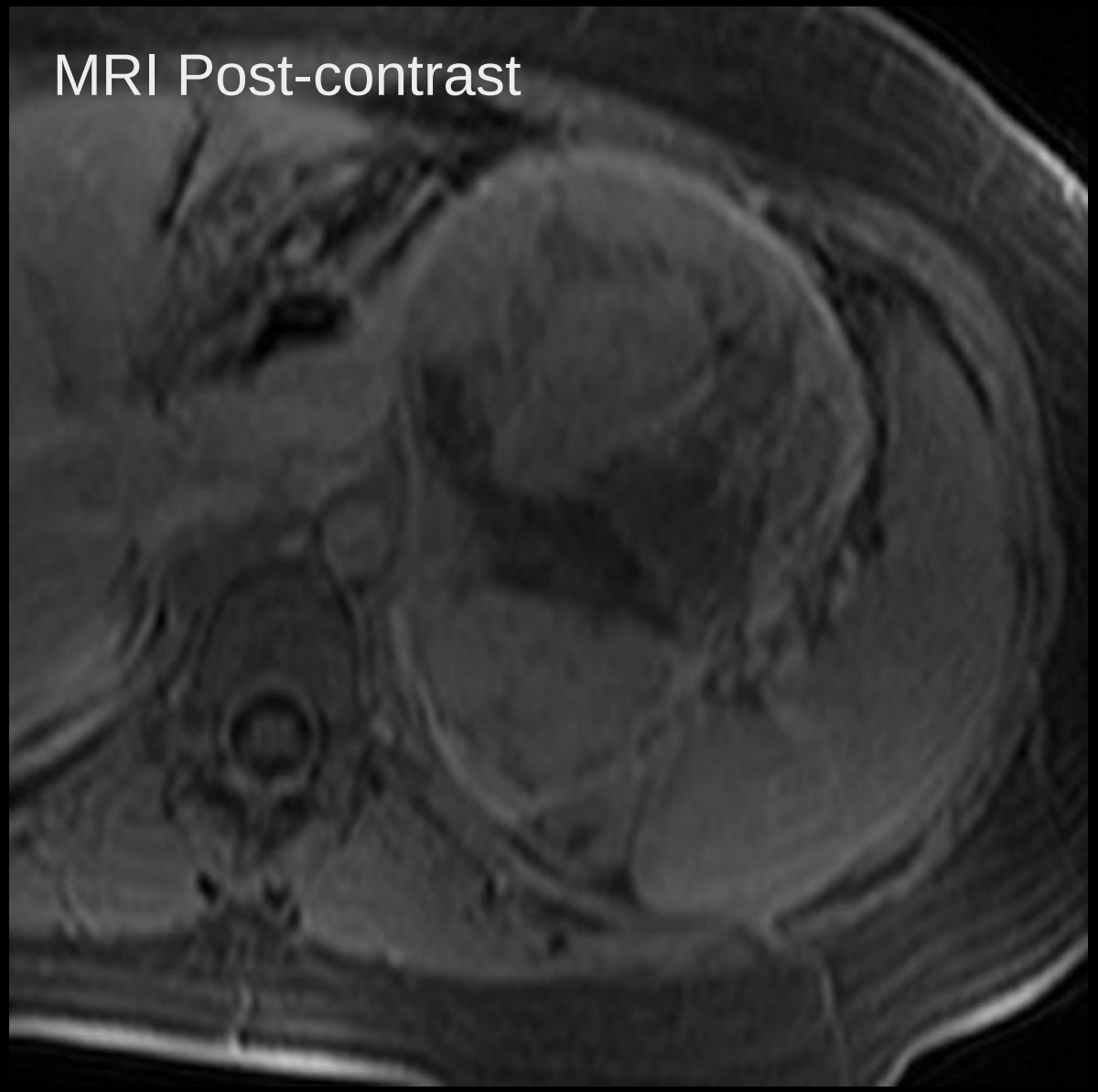
Noncontrast CT

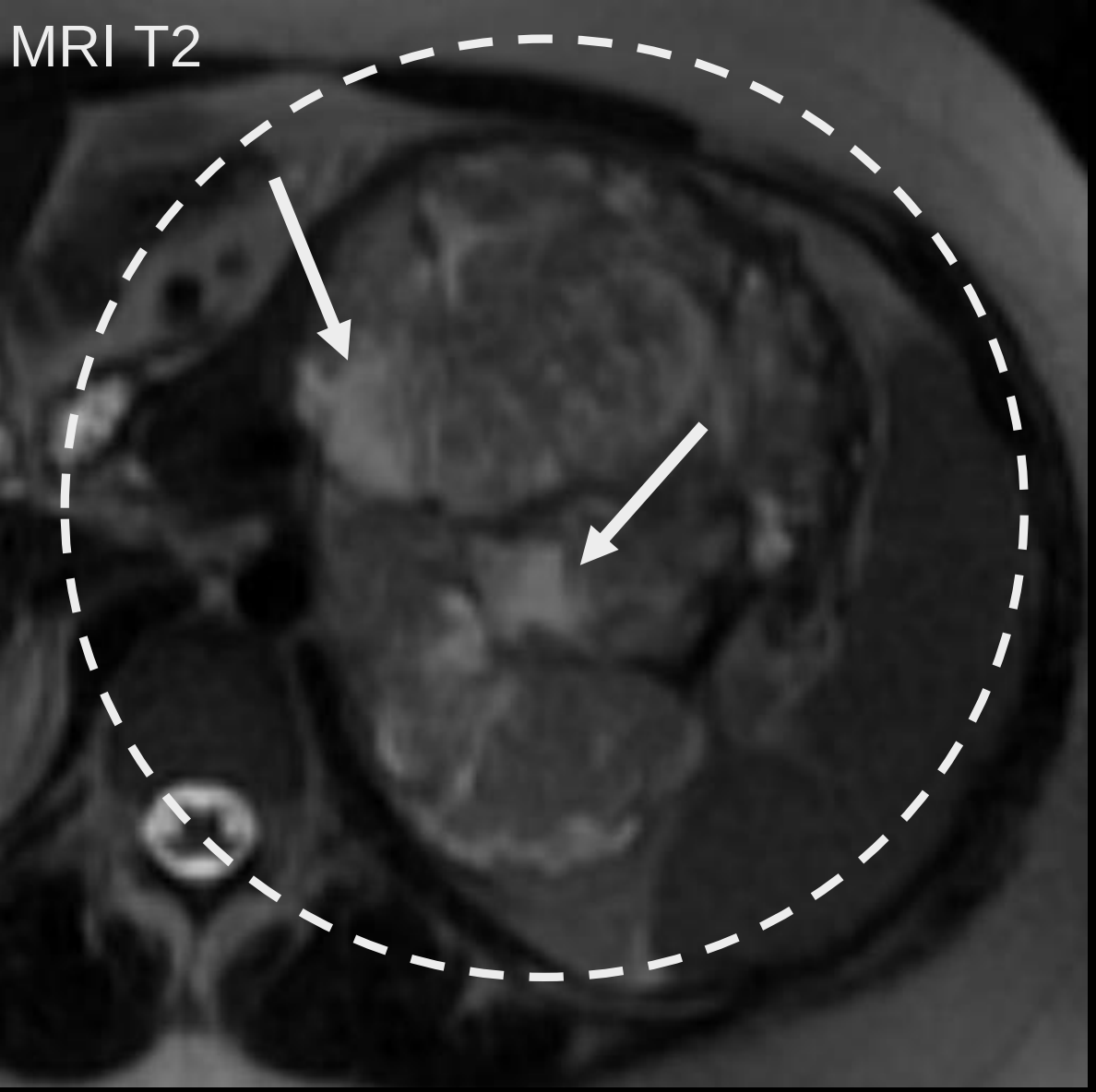


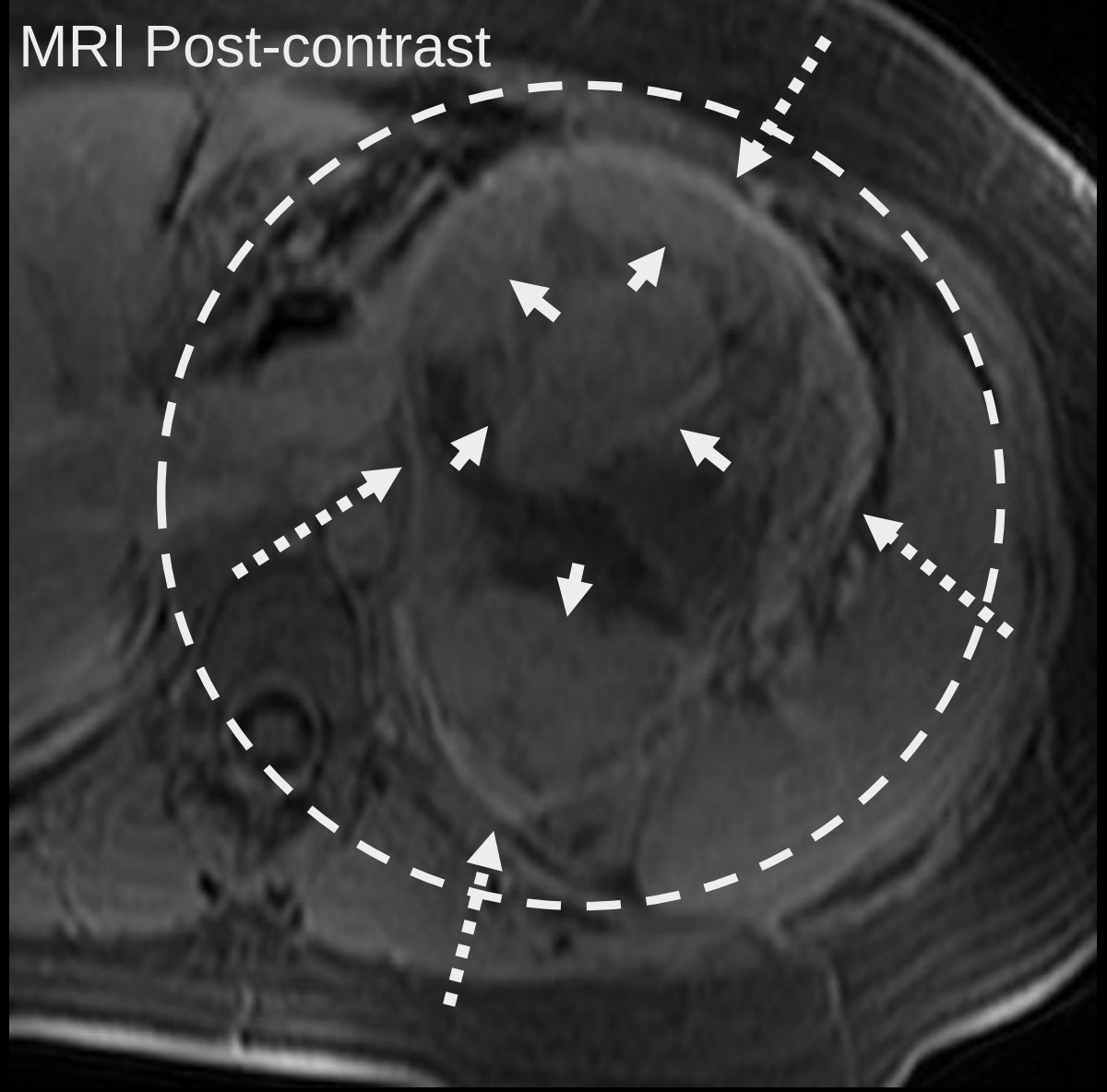


Heterogeneous mass arising from the pancreatic tail (circle) possibly with internal calcification (arrow)


MRI T2


MRI Post-contrast





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.

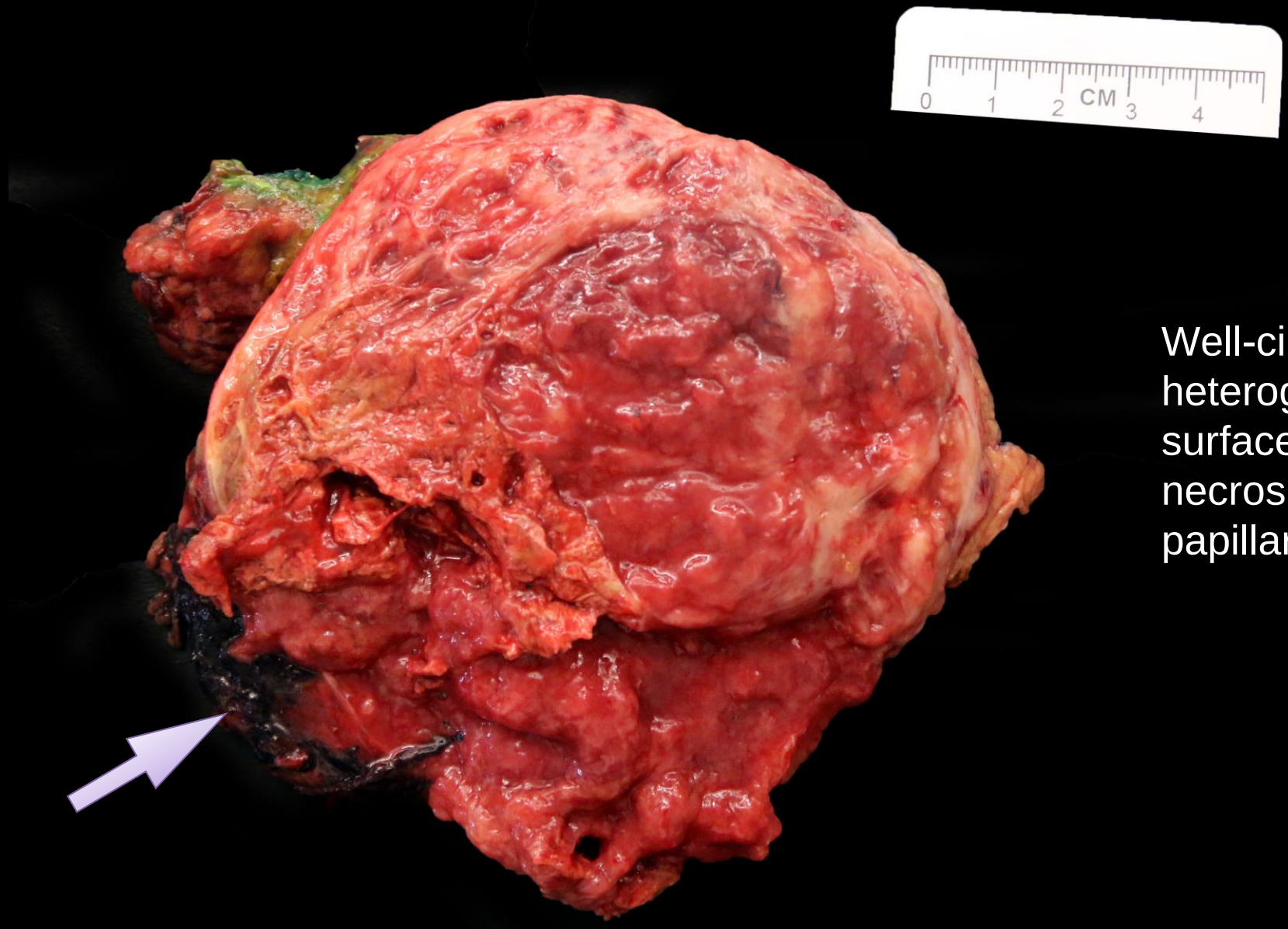
DDX (based on imaging)

- Pancreatic solid pseudopapillary tumor
- Pancreaticoblastoma
- Nonfunctioning islet cell (neuroendocrine) tumor

- OPERATION: Exploratory laparotomy, distal pancreatectomy, splenectomy.

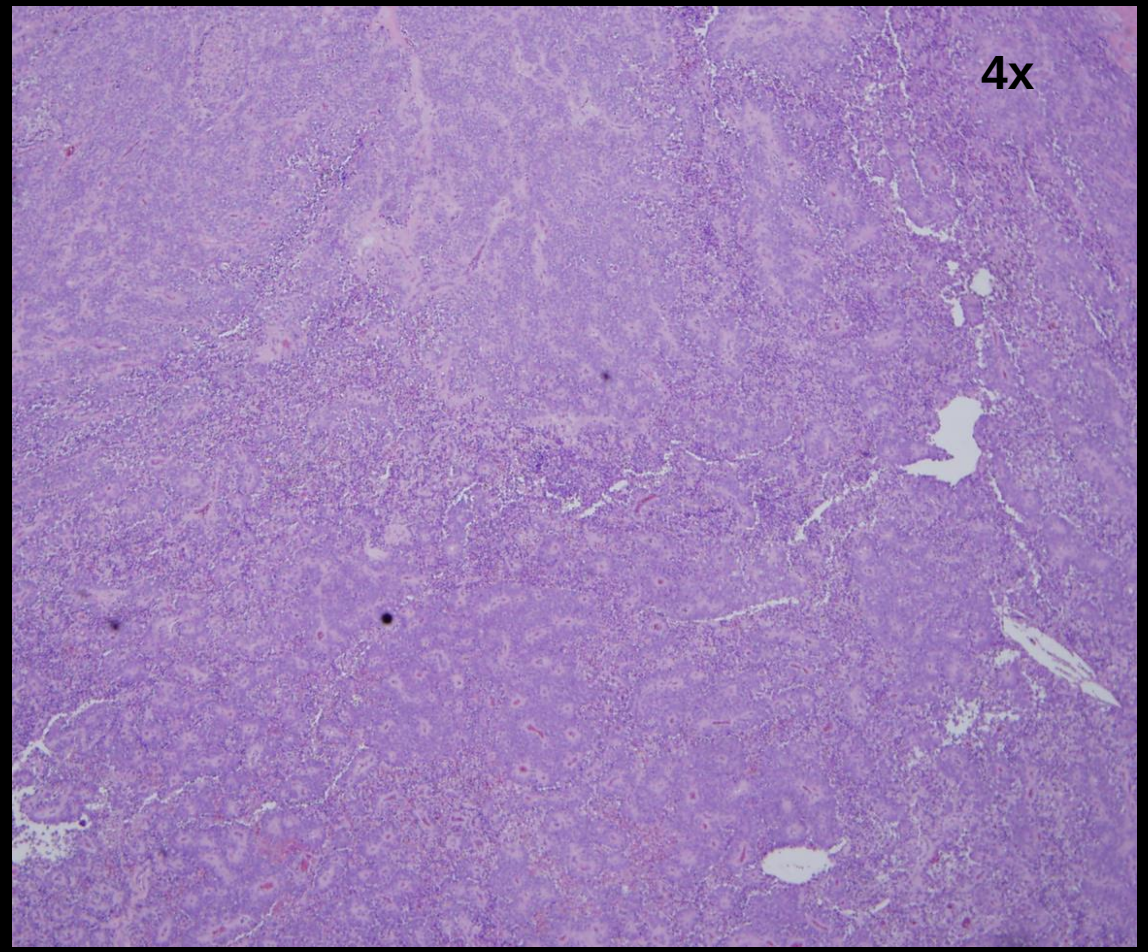
- INDICATIONS FOR PROCEDURE:
- 13-year-old female with mass in the tail of her pancreas; it was felt that resection was indicated. The tumor was large enough that it was compressing the splenic vein with significant left-sided portal hypertension. Because of this concern and the possibility of malignancy, laparotomy and splenectomy considered appropriate.

Gross Path

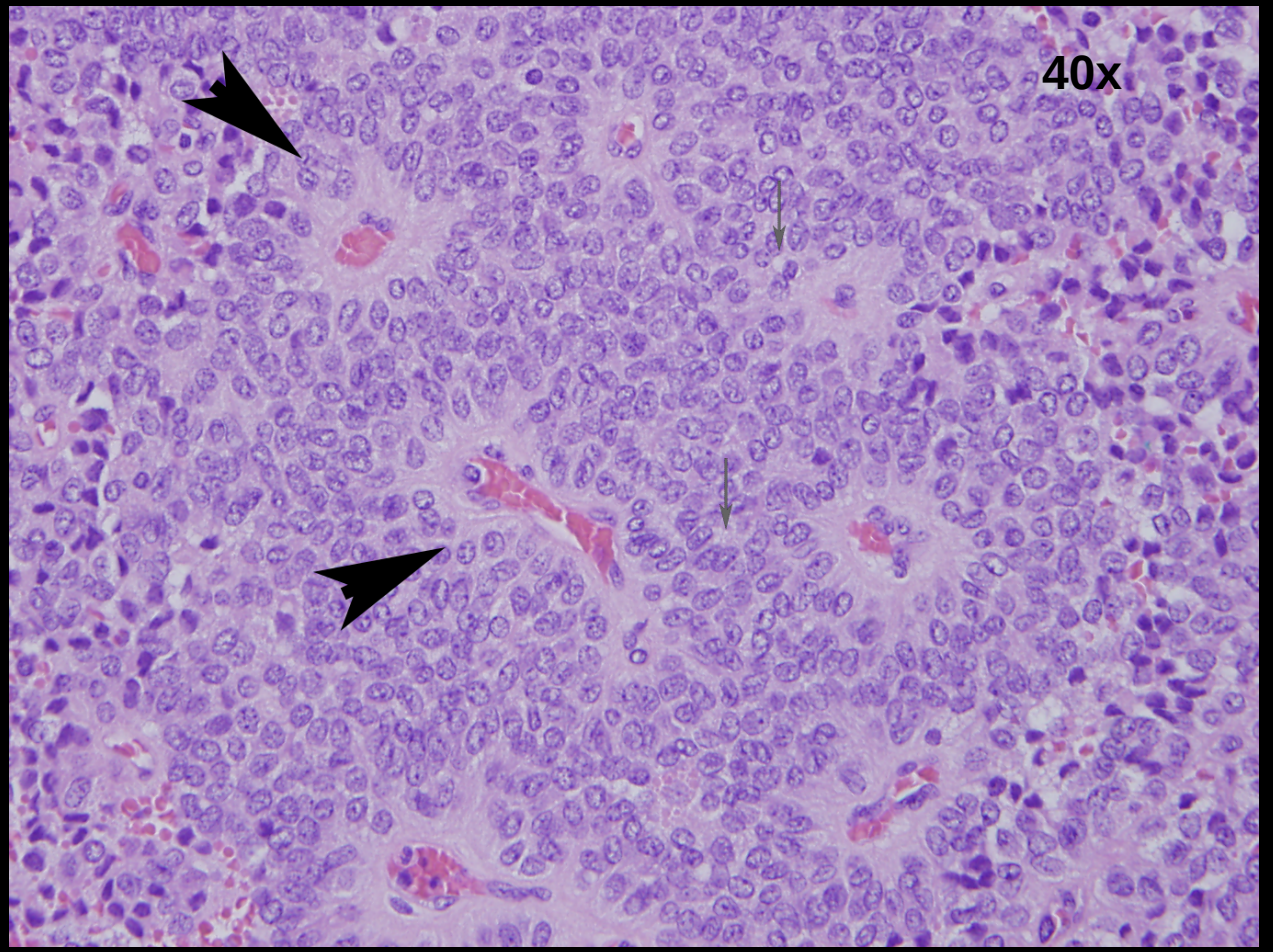


Well-circumscribed mass with a heterogeneous cystic and solid cut surface. There is an area of necrosis/degeneration with a papillary appearance (white arrow).

Micro Path

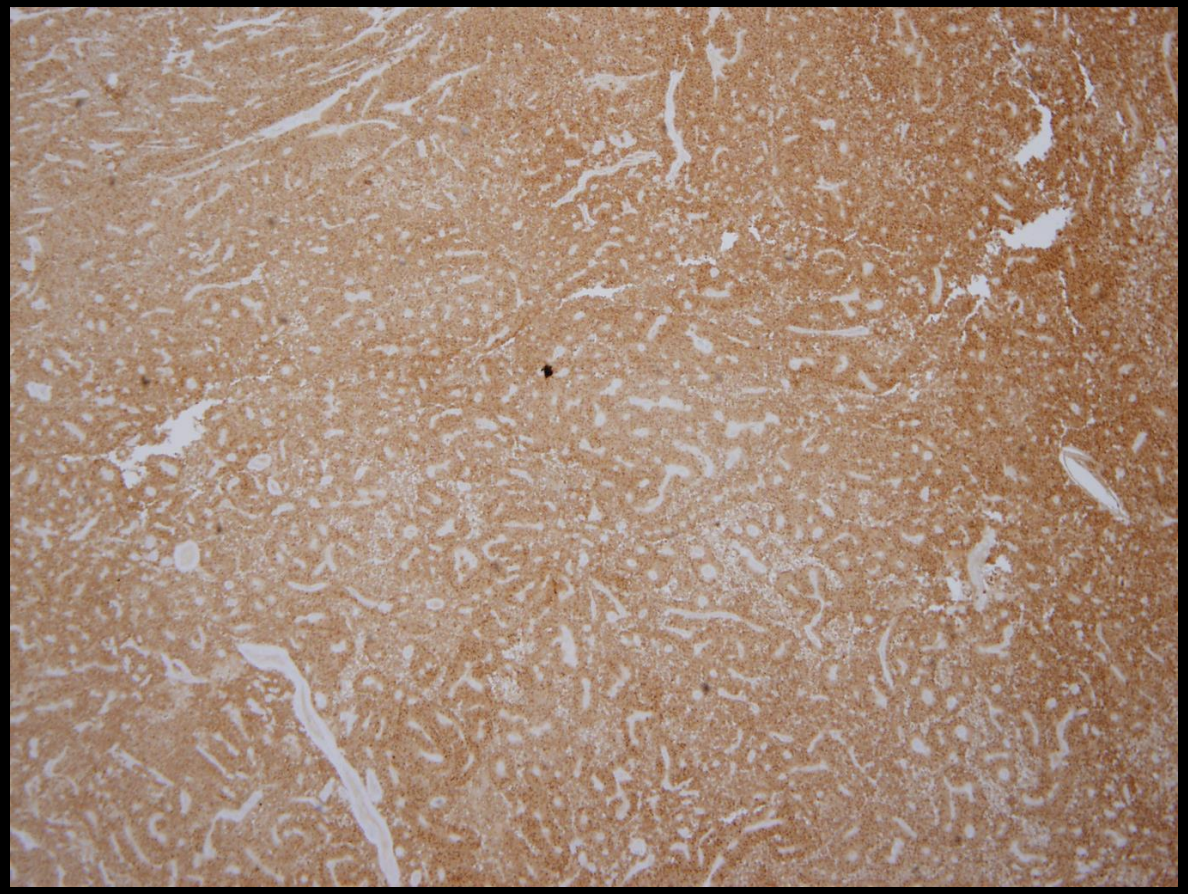


Solid growth on top of photo, pseudo-papillary growth on bottom



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

IHC



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)**, α*1*-antitrypsin, CD56, CD10, and vimentin.

References:

- Asim Shuja and Khalid A. Alkimawi. *Solid pseudopapillary tumor: a rare neoplasm of the pancreas*. Gastroenterology Report 2 (2014) 145–149, doi:10.1093/gastro/gou006
- 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*