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# An Unlikely Cause of Possible Hormone-Dependent Abdominal Pain in Young Fertile Females: A Case Series

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## Background

- Solid pseudopapillary neoplasm (SPN) is a rare pancreatic exocrine tumor (Table 1) with unknown etiology, first described by Dr. Frantz in 1959.<sup>1-3</sup>
- SPN is a benign lesion with low grade malignant potential, often curative with surgery.<sup>1</sup>
- Hormones may play into the origin/development, with a strong correlation with females and progesterone receptor (PR).
- SPN mimics pancreatic neoplasms, pseudocysts and pancreatic endocrine neoplasms on imaging and cytomorphological features<sup>2</sup> (Table 2, Image 1).

Table 1: Common Features of SPN <sup>1-3</sup>	
Epidemiology	Clinical Presentation
2nd-3rd decade of life	Asymptomatic
Non-Caucasian	Abdominal Pain/Mas
Female gender (10:1)	Nausea/Vomiting
Rare (1-2% exocrine tumors)	Weight loss
Benign (15% malignant)	Jaundice

Table 2. Microscopic Features of SPN <sup>1-3</sup>	
Size	Distribution
Cytology	Round/oval eccentric nuclei Uniform cells with fine granular chromatin No mitotic figures Extracellular hyaline globules
Histology	Pseudopapillary, solid, cystic Thin walled vessels Bands of fibrous tissue
Immunohistochemistry (IHC)	+ CD56, CD10, PR, $\beta$ -catenin (nuclear) - E-cadherin membranous - Chromogranin/Synaptophysin (G/S)*

## Case Presentation

We present two cases of young females with **epigastric pain, nausea/vomiting**, without pruritus, jaundice or steatorrhea.

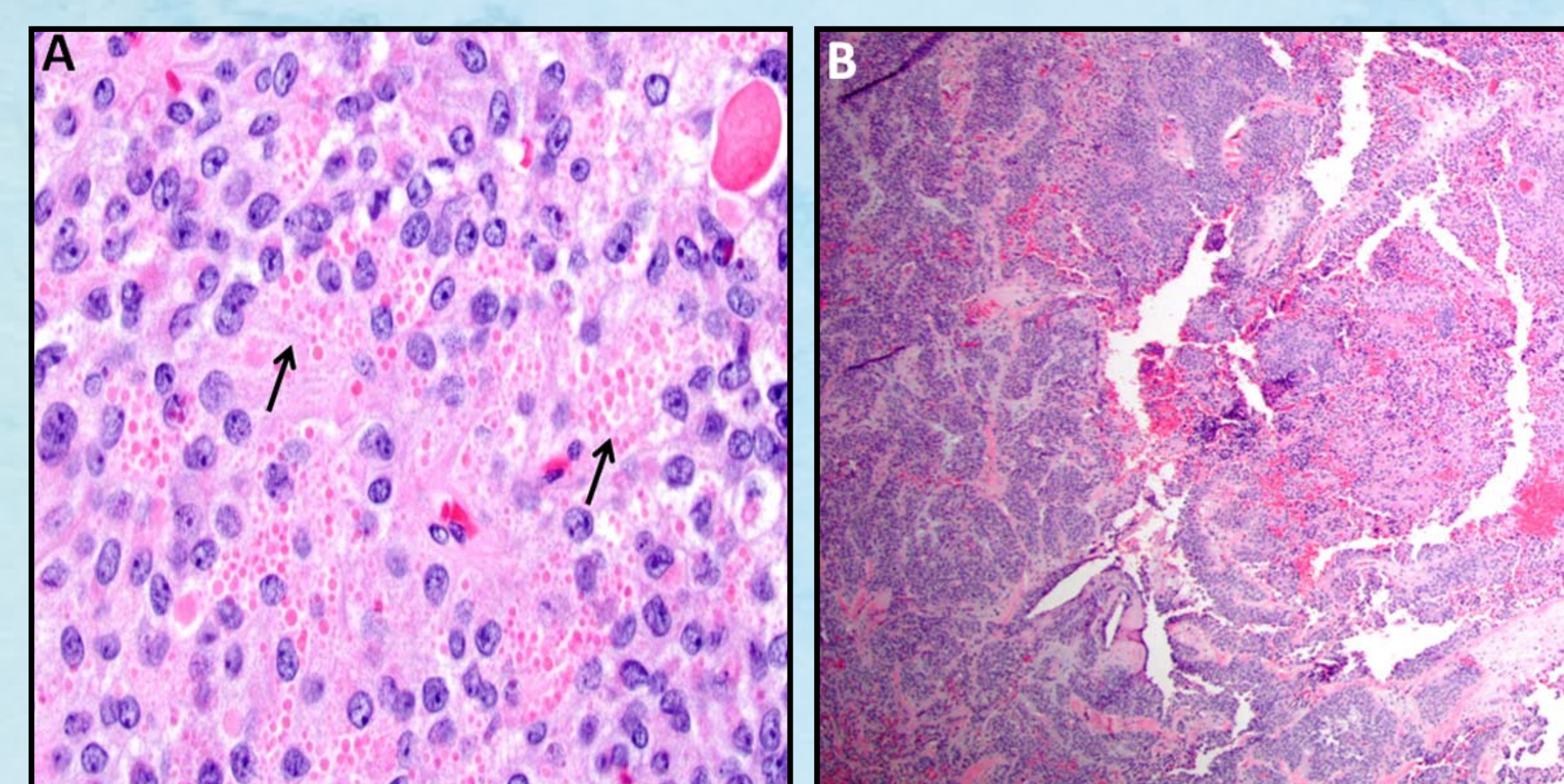
### CASE 1:

- A **20-week pregnant** female with a **solid pancreatic lesion** in the **uncinate process** (Image 2) as incidental finding on imaging. **Mirena** was inserted post-partum and repeat imaging showed an increase in tumor size after 1 year.
- Endoscopic ultrasound-fine needle aspiration (EUS-FNA) was performed (Table 3), followed by a **Whipple procedure**.
- Post-op complications included intra-abdominal abscess.

### CASE 2:

- A **solid mass** in the **pancreatic tail** was found on CT imaging (Image 3) with follow-up MRI indicating **spontaneous lesion regression**. Two years later and starting **oral contraceptive pills**, repeat imaging showed **increase tumor size** and **enhancing liver lesions**.
- EUS-FNA was performed (Table 3) and CT guided liver biopsy consistent with focal nodular hyperplasia. **Distal pancreatectomy** and splenectomy were performed.
- Repeat imaging showed increase size in liver lesions, but PET scan was negative.

Table 3. Pathology and IHC Case Series Comparison		
	Case 1	Case 2
FNA	+ C56, Vimentin, CD10, AE1/AE3 - C/S Oval nuclei No necrosis/mitotic figures Salt/pepper chromatin	+ CD10, Vimentin, PR, AE1/AE3, $\beta$ -catenin - C/S Monomorphic uniform cells Round/oval nuclei with grooves Finely dispersed chromatin
Surgical Pathology	+ CD56, Vimentin, CD10, $\beta$ -catenin (nuclear) + weak C/S Ki67 <2% 22 benign lymph nodes	+ CD56, CD10, $\beta$ -catenin (nuclear) + C/S Ki67 25% 25 benign lymph nodes



**Image 1.** Characteristic hyaline globules in clusters (arrows) (A). Solid sheets of uniform cells with papillary formation from lack of adhesion (B).



**Image 2.** Case 1- CT of abdomen/pelvis that visualizes the solid pancreatic mass (arrow) located in the uncinate process.



**Image 3.** Case 2- CT of abdomen/pelvis that shows solid pancreatic mass (arrow) located in the tail of the pancreas.

## Discussion:

- Both cases demonstrate a classic presentation of SPN; a young, fertile female with abdominal pain and pancreatic lesion on imaging.
- Lack of known etiology/pathophysiology, specific IHC markers/patterns to distinguish SPN from other pancreatic masses is a challenge.
  - Etiology: ectopic ovarian stroma from female genital bud or primitive centroacinar cells that are hypersensitivity to female sex hormone stimulation leading to proliferation.<sup>4-5</sup>
  - PR are located on 75%  $\alpha$ -cells and 5-20%  $\beta$ -cells. Progesterone increases proliferation of acinar cells most in vivo and promotes proliferation of differentiated cells, not neogenesis.<sup>6-7</sup>
- SPN requires complete surgical resection despite low malignancy risk with 5 yr survival ~97%.<sup>1,4</sup>
  - Metastatic disease include liver, regional lymph nodes, mesentery, omentum, peritoneum with local invasion to duodenum, stomach, spleen and major vessels.<sup>8</sup>
  - Post-op complications: pancreatic fistula, pancreatitis, prolonged gastric emptying, bleeding, infection, diabetes.<sup>8</sup>
- Possible IHC pattern for accurate SPN diagnosis is +CD10, +nuclear  $\beta$ -catenin with negative membranous E-cadherin.
  - Not utilized universally leading to misdiagnoses or inappropriate therapy.

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