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.  
- SPN is a benign lesion with low grade malignant potential, often curative with surgery.  
- Hormones may play into the origin/development, with a strong correlation with females and progesterone receptor (PR).  
- SPN mimics pancreatic neoplasms, pseuodoblasts and pancreatic endocrine neoplasms on imaging and cytological features (Table 2, Image 1).

**Case Presentation**

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

**CASE 1:**

- A 29-week pregnant female with a solid pancreatic lesion in theuncinate 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 increased size in liver lesions, but PET scan was negative.

**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 centrocarcinoma cells that are hypersensitive to female sex hormone stimulation leading to proliferation.  
  - PR are located on 75% α-cells and 5-20% β-cells. Progesterone increases proliferation of acinar cells most in vivo and promotes proliferation of differentiated cells, not neogenesis.  
- SPN requires complete surgical resection despite low malignancy risk with 5yr survival ~97%.  
  - Metastatic disease include liver, regional lymph nodes, mesentery, omentum, peritoneum with local invasion to duodenum, stomach, spleen and major vessels.  
  - Post-op complications: pancreatic fistula, pancreatitis, prolonged gastric emptying, bleeding, infection, diabetes.  
- Possible IHC pattern for accurate SPN diagnosis is: CD10, β-catenin with negative membranous E-cadherin.  
  - Not utilized universally leading to misdiagnoses or inappropriate therapy.

**References:**


**Table 1: Common Features of SPN**

<table>
<thead>
<tr>
<th>Epidemiology</th>
<th>Clinical Presentation</th>
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<tbody>
<tr>
<td>2nd-3rd decade of life</td>
<td>Hypertrophic</td>
</tr>
<tr>
<td>Female gender (10:1)</td>
<td>Abdominal Pain/Mass</td>
</tr>
<tr>
<td>Rare pancreatic exocrine tumor</td>
<td>Asymptomatic</td>
</tr>
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<td>15% malignant</td>
<td>Nausea/Vomiting</td>
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**Table 2: Microscopic Features of SPN**

<table>
<thead>
<tr>
<th>Cytology</th>
<th>Distribution</th>
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<tbody>
<tr>
<td>Round/oval eccentric nuclei</td>
<td>Membranous E-cadherin</td>
</tr>
<tr>
<td>Sheets of uniform cells with papillary formation</td>
<td>E-cadherin membranous</td>
</tr>
</tbody>
</table>

**Table 3: Pathology and IHC Case Series Comparison**

<table>
<thead>
<tr>
<th>Case 1</th>
<th>Case 2</th>
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<tbody>
<tr>
<td>FNA</td>
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<tr>
<td>EUS-FNA</td>
<td></td>
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<tr>
<td>Membranous E-cadherin</td>
<td>Membranous E-cadherin</td>
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<td></td>
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<tr>
<td>E-cadherin, PR, AE1/AE3, CD10, S100, Vimentin, PR</td>
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<tr>
<td>Membranous E-cadherin, PR</td>
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<tr>
<td>Membranous E-cadherin, PR, S100, Vimentin, PR</td>
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<td>Membranous E-cadherin, PR, S100, Vimentin, PR</td>
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**Image 1:** Characteristic hyaline globules in clusters (arrow). Solid sheets of uniform cells with papillary formation from lack of adhesion (B).

**Image 2:** Case 1: CT of abdomen/pelvis that showed a solid pancreatic mass (arrow) located in the tail of the pancreas.

**Image 3:** Case 2: CT of abdomen/pelvis that showed a solid pancreatic mass (arrow) located in the body of the pancreas.