Puzzles of the Pediatric Pancreas: A Pictorial Review of Pediatric Pancreatic Pathologies

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Background

- Pancreatic masses are rare in the pediatric population
- Pancreatic tumors only account for 0.2% of malignant pediatric deaths
  - Clinical courses are distinct from those in the adult counterpart
    - Pediatric pancreatic tumors have a better prognosis and different histological spectrum
      - Pancreatic neoplasms can be divided into:
        - Epithelial origin
        - Non-epithelial origin
  - Non-neoplastic pathology can present as mass-like lesions and should be differentiated from neoplasm
Learning Objectives

In this presentation, we will:

- Demonstrate the imaging findings of pancreatic masses in pediatric patients using a multi-modality approach with ultrasound (US), computed tomography (CT) and magnetic resonance imaging (MRI) through a series of “puzzles” from our institution.

- Discuss the differential diagnosis and key imaging features that allow differentiation of these pathologies.

- Describe the pathologic basis of these features and management of these pancreatic lesions.
## Pancreatic Lesion Differential

<table>
<thead>
<tr>
<th>Neoplastic</th>
<th>Non-Neoplastic</th>
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<tbody>
<tr>
<td><strong>Epithelial Origin</strong></td>
<td><strong>Non-Epithelial Origin</strong></td>
</tr>
<tr>
<td>Pancreatoblastoma</td>
<td>Lymphoma</td>
</tr>
<tr>
<td>Solid Pseudo-papillary Tumor (SPEN)</td>
<td>Leukemia</td>
</tr>
<tr>
<td>Endocrine Tumors</td>
<td>Lymphangioma/ Venolymphatic malformation Hemangioma</td>
</tr>
<tr>
<td>Insulinoma Gastrinoma ACThoma VIPoma Nesidioblastosis</td>
<td>Dermoid Cyst</td>
</tr>
<tr>
<td>Ductal Adenocarcinoma Acinar Cell</td>
<td>Mesenchymal Tumors</td>
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</tbody>
</table>
Puzzle #1:
10 year-old female presenting after an episode of projectile vomiting with elevated lipase

- **Pieces of the Puzzle:** Imaging from initial US obtained - midline abdomen

- How would you describe this lesion?
- What is your differential?
- Will you suggest follow up imaging? What modality?

Go through the case and see the “clues” for imaging descriptions and the discussion at the end for the answers to the questions you encounter.
Puzzle #1:
Pieces of the Puzzle: Additional imaging obtained

• How would you describe this lesion on MRI?
• Does it have mass effect?

Now that you have all the pieces of the puzzle...

Can you solve the case?

• What effect does it have on the pancreatic duct?
• Does this change your differential?
Puzzle #1:
A Few Clues: Imaging descriptions

**Figure A:** Ultrasound image demonstrating heterogeneously hypoechoic mass in the head of the pancreas (blue arrow). There is pancreatic ductal dilatation (red arrow).

**Figure B:** T2 axial image demonstrating heterogeneously hypointense mass at the head of the pancreas (blue arrow).

**Figure C:** T1 axial image demonstrating intrinsic T1 hyperintensity suggestive of blood or proteinaceous products (blue arrow).

**Figure D:** T2 coronal imaging demonstrating the degree of pancreatic ductal dilatation (blue arrow).
Puzzle #1:
Solid Pseudo-papillary Tumor (SPEN)

- Accounts for 0.2-2.7% of all non-endocrine tumors of the pancreas
  - 22-52.6% of patients are children
  - *Most common pancreatic tumor of Asian children*
- **Clinical Presentation:** Abdominal discomfort, palpable mass
  - Jaundice is rare
  - Often asymptomatic
- **Pathology:** *Solitary slow growing tumor*
  - Well circumscribed, large on presentation
    - Fibrous capsule
  - Marked degenerative and hemorrhagic change
    - Soft, fleshy and friable with solid and cystic component
    - Small tend to be more solid
  - Can occur throughout the pancreas
# Solid Pseudo-papillary Tumor (SPEN): Imaging Features

## Ultrasound
- Large, well-circumscribed lesion
- Variable echogenicity depending on composition
- Echogenic rim of the fibrous capsule

## CT
- Hypoattenuating capsule
- Compresses adjacent structures without invasion
- Cystic contents are higher in density than gallbladder fluid density
  - Due to presence of hemorrhage and debris
- Less than 1/3 have internal septation
- Calcification can be seen at the periphery
- Solid portion enhances

## MRI
- T1 and T2 hypointense fibrous capsule
- Solid portions of tumor are iso to hypointense – but enhance with contrast
- Hemorrhage – a distinctive feature is best shown by MRI
  - Variable due to degradation of hemoglobin

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**Key Imaging Features:**
- Fibrous capsule and internal hemorrhage
Solid Pseudo-papillary Tumor (SPEN): Treatment and Prognosis

- Slow-growing tumor usually with benign clinical course
  - Due to potential for aggressive behavior → surgical resection
- 95% with local disease are cured by complete resection
- Metastatic disease – 7-16%
  - Liver metastasis - resected
Puzzle #2:
3 month-old male with scleral icterus and intermittently dark urine with additional findings of elevated leukocytosis and abundant blasts

- **Pieces of the Puzzle:** Imaging from initial US obtained – gallbladder and common bile duct

- Can you identify the gallbladder? Is there biliary ductal dilatation? What size do you expect the common bile duct to be in 6 month old?
- Where is the lesion?
- Will you suggest follow up imaging? What modality?

Go through the case and see the “clues” for imaging descriptions and the discussion at the end for the answers to the questions you encounter.
Puzzle #2: Pieces of the Puzzle: Additional imaging obtained

- How would you describe this lesion's effect on adjacent structures?
- What effect does it have on the biliary system?
- Does this change your differential?

Now that you have all the pieces of the puzzle...

Can you solve the case?
Puzzle #2: A Few Clues: Imaging descriptions

**Figure A:** Ultrasound imaging did not show mass; however, there was significant dilatation of the common bile duct (blue arrows). Gallbladder is full of sludge (red arrow).

**Figure B:** Coronal T2 demonstrating significant intra- and extra-hepatic biliary ductal dilatation (blue arrows).

**Figures C and D:** T2 Axial images demonstration homogenous enlarged pancreas consistent with leukemic infiltration (blue arrows).
Puzzle #2: Pancreatic Involvement of Acute Lymphocytic Leukemia (ALL)

- Most common non-epithelial tumor of the pancreas
  - Primary pancreatic lymphoma is much less common than secondary involvement
    - Secondary involvement = diffusely disseminated disease
    - Usually Large – cell and Burkitt’s involve the pancreas
  - Infant leukemia – seen before the age of one
    - Higher risk in female infants than male (reverses after the age of one)

Clinical Presentation: Non-specific, palpable abdominal mass, weight-loss, obstructive jaundice

- Infant leukemia is more aggressive with hepatosplenomegaly, CNS involvement and skin infiltration
Puzzle #2:
Pancreatic Involvement of Acute Lymphocytic Leukemia (ALL)

- **Pathology**: Two recognized morphologic patterns
  - Focal form occurs in the pancreatic head 80%
  - Diffuse form is infiltrative leading to glandular enlargement and poor definition
    - Can mimic acute pancreatitis
  - Infant leukemia – characterized cytogenetically with translocation of the MLL gene and associated with poorer prognosis

- **Treatment & Prognosis** – chemotherapy, treat the underlying leukemia
  - **ALL of the Infant has a poor prognosis**
    - Mortality associated with toxicity from aggressive chemotherapy regimen
**Pancreatic Involvement of Lymphoma: Imaging Features**

<table>
<thead>
<tr>
<th>Ultrasound</th>
<th>CT</th>
<th>MRI</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Focal hypoechoic lesion in the head of pancreas (focal form)</td>
<td>• <strong>Focal</strong>&lt;br&gt;  • Hypoattenuating focus usually in the head of the pancreas&lt;br&gt;  • Retroperitoneal adenopathy</td>
<td>• <strong>Focal</strong>&lt;br&gt;  • T1 low intensity and T2 intermediate intensity (higher than pancreatic parenchyma but lower than fluid)&lt;br&gt;  • Faint contrast enhancement&lt;br&gt;  • Head of pancreas</td>
</tr>
<tr>
<td>• Hypoechoic enlargement of the pancreas (diffuse form)</td>
<td>• <strong>Diffuse</strong>&lt;br&gt;  • Hypoattenuating pancreas&lt;br&gt;  • Adenopathy&lt;br&gt;  • Multiple non-contiguous masses</td>
<td>• <strong>Diffuse</strong>&lt;br&gt;  • T1 and T2 low signal intensity&lt;br&gt;  • Homogenous contrast enhancement with small foci of reduced or absent enhancement</td>
</tr>
<tr>
<td>• Secondary effect on biliary system with compression of adjacent structures causing biliary ductal dilatation</td>
<td>• <strong>Diffuse</strong>&lt;br&gt;  • Secondary effect of compression of the biliary system</td>
<td>• Secondary effects of compression of the biliary system</td>
</tr>
</tbody>
</table>

**Key Distinguishing Imaging Features:**
- Bulky localized tumor without main pancreatic ductal dilatation
- Enlarged lymph nodes
- Lack of vascular invasion
Puzzle #3:
15 year-old female presenting after ATV bike accident

► **Pieces of the Puzzle:** Imaging from CT Body Trauma Scan

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► How would you describe this lesion?
► What should you look for?
► Who needs follow up imaging?

*Go through the case and see the “clues” for imaging descriptions and the discussion at the end for the answers to the questions you encounter*
Puzzle #3:

Pieces of the Puzzle: Additional imaging obtained after patient presented with nausea 4 weeks after accident

• Where is this lesion?
• What does it contain?

• Is this lesion vascular?
• Why did this arise?

Now that you have all the pieces of the puzzle...

Can you solve the case?
Puzzle #3:
A Few Clues: Imaging descriptions

**Figure A and B:**
Arterial and Venous phase axial CECT images demonstrating pancreatic laceration through the mid-body of the pancreas (blue arrows).

**Figure C and D:**
Subsequent US and Doppler imaging shows a hypoechoic focus in the pancreas with laying internal debris without vascular flow (blue arrows).
Puzzle #3: Pseudocyst Formation after Traumatic Laceration

Pancreatic injury is rare in the pediatric population

- However, it is the fourth most common injury, after that to the liver, spleen and kidneys
- 2/3 of injuries occur in the pancreatic body
  - Usually a result of a direct blow to the epigastrium and compression against the rigid spinal column

Direct signs are often difficult to detect and secondary signs of injury should be sought

- Peripancreatic fluid – fluid in the anterior pararenal space or lesser sac
- Findings of pancreatitis – diffuse gland enlargement or peripancreatic fat stranding, thickening of the anterior renal fascia and free fluid

Direct trauma to the pancreas is graded (next slide)
Puzzle #3: Pseudocyst Formation after Traumatic Laceration

Organ Injury Scaling of the American Association for Surgery in Trauma

- **Grade 1:** Hematoma with minor contusion/laceration but without duct injury
- **Grade 2:** Major contusion/laceration but without duct injury
- **Grade 3:** Distal laceration but without duct injury
- **Grade 4:** Proximal (to the right of the superior mesenteric vein) laceration or parenchymal injury WITH injury to the bile duct/ampulla
- **Grade 5:** Massive disruption of the pancreatic head
Puzzle #3: Pseudocyst Formation after Traumatic Laceration

- Management of injury is somewhat *controversial in the pediatric population*
  - Standard of care currently is that of NON-operative management
    - Presence of ductal injury is a predictor of failure of non-operative management
      - External drainage is proposed for treatment of contusions and lacerations
        - Prevention of complications due to presence of proteolytic enzymes
    - Spleen sparing distal pancreatectomy is favored in the setting of distal ductal injuries
  - Complications of injury include:
    - Formation of pancreatic fistulae
    - Pancreatitis
    - Development of pseudocysts
      - Formation of a pseudocyst is seen as a favorable complication - easily drained percutaneously or cystogastrostomy
Puzzle #4:
16 year-old male presenting with watery profuse diarrhea

- **Pieces of the Puzzle:** Imaging from CT in the arterial phase

- How would you describe this lesion?
- What is happening to the gastric wall?
- How can you confirm this lesion? What follow-up imaging can you pursue?

Go through the case and see the “clues” for imaging descriptions and the discussion at the end for the answers to the questions you encounter.
Puzzle #4:
**Pieces of the Puzzle:** Additional imaging obtained

- What type of exam is this?
- What radiotracer is used and why?
- What does this show?

Now that you have all the pieces of the puzzle...

Can you solve the case?
**Puzzle #4:**

**A Few Clues:** Imaging descriptions

**Figure A and B:**
Arterial phase axial and sagittal CECT images demonstrating an arterial enhancing mass emanating from the head of the pancreas (blue arrows).

**Figure C:**
Axial CECT demonstrating diffuse gastric wall thickening (blue arrows).
**Puzzle #4:**

**One More Clue: Imaging descriptions**

**Figure D:**
Selected SPECT images from an In-111 labeled Octreoscan. Images demonstrate focal radiotracer uptake in the region of the pancreatic head corresponding to the lesion seen on CT scan (blue arrows). No metastatic disease to the liver is shown.
Puzzle #4:
Endocrine Cell Derived Islet Cell Tumors: Gastrinoma

- Islet cell tumors are seen in **older children**
  - More often encountered in middle age adults
    - Age range of 7-83 years
- **May or may not secrete** hormonally active polypeptide capable of producing a clinical syndrome
  - Functioning or hyperfunctioning islet cell tumors
    - Most common: Insulinoma (47%)
    - Second: Gastrinoma (30%)
  - Non-functioning or silent islet cell tumors
### Puzzle #4:
Endocrine Cell Derived Islet Cell Tumors:

<table>
<thead>
<tr>
<th>Tumor Type</th>
<th>Clinical Features</th>
</tr>
</thead>
<tbody>
<tr>
<td>Insulinoma</td>
<td>Composed of B-cells → hyperinsulinemic hypoglycemia</td>
</tr>
<tr>
<td>Gastrinoma</td>
<td>G cells → Zollinger-Ellison Syndrome</td>
</tr>
<tr>
<td></td>
<td>• Multiple/recurrent peptic ulcers – duodenal/post bulbar</td>
</tr>
<tr>
<td></td>
<td>• Gastroesophageal reflux</td>
</tr>
<tr>
<td></td>
<td>• Gastrin hypersecretion → diarrhea</td>
</tr>
<tr>
<td>ACTHoma/</td>
<td>Cushing Syndrome</td>
</tr>
<tr>
<td>VIPoma</td>
<td>Massive watery diarrhea, hypokalemia, achlorhydria</td>
</tr>
<tr>
<td>Somatostatinoma</td>
<td>D-cells → diabetes, gallbladder disease steatorrhea</td>
</tr>
<tr>
<td>Glucagonoma (only two cases reported in children)</td>
<td>A cells → diabetes, stomatitis, necrolytic migratory erythema</td>
</tr>
</tbody>
</table>
Puzzle #4: Endocrine Cell Derived Islet Cell Tumors

- Neuroendocrine tumors range in size from 0.5mm to 20 cm
  - Non-functioning tumors often manifest later and larger in size as symptoms are due to mass effect
  - Insulinomas often present at very small sizes due to their striking clinical symptoms
    - Usually involve the body and tail of the pancreas

- Gastrinomas are variable in the production of clinical symptoms
  - Gastroesophageal reflux may be the ONLY manifestation
  - Gastrin hypersecretory diarrhea is occasionally the dominant symptom
    - Can cause clinical confusion and delay in diagnosis

- Gastrinomas tend to occur in the “gastrinoma triangle”
  - Bound by the porta hepatitis and the second/third portions of duodenum
Puzzle #4: Endocrine Cell Derived Islet Cell Tumors: Gastrinoma – Imaging Features

**Ultrasound**
- Difficult to identify due to smaller size
- May be hypoechoic with hyperechoic rim

**CT**
- Gastrinomas are usually larger than insulinomas
- Tend to have a heterogeneous appearance
- Small gastrinomas enhance avidly, greater than pancreas
- Avid on the arterial phase

**MRI**
- T1 and T2 hyperintense
- Ring-like enhancement while insulinomas enhance homogeneously
- Tend to be visible on T1 weighted imaging prior to contrast administration
- Easier to identify metastasis
  - Larger tumors are associated with liver metastasis

**Key Distinguishing Imaging Features:**
- Markedly avid arterial enhancement
- Enhancement greater than pancreas
- Arterial enhancing mets
**Puzzle #4:**
Endocrine Cell Derived Islet Cell Tumors: Gastrinoma – Imaging Features

- **Somatostatin-receptor imaging**
  - Somatostatin – naturally occurring neuropolypeptide released by endocrine or nerve cells
    - High density of somatostatin receptors in numerous endocrine tumors
  - Radiolabeled analog – **indium labelled penetreotide (Octreoscan)**
    - Less gastrointestinal activity – good for abdominal tumor sites
  - Imaging at **4, 24 and 48** hours with SPECT imaging
    - Tumors – foci of increased uptake
  - **Do not use for pancreatic carcinomas of exocrine origin**
    - False negative
Puzzle #4: Endocrine Cell Derived Islet Cell Tumors: Gastrinoma – Imaging Features

- **Treatment and Prognosis**
  - **Surgical resection**
    - Leads to complete cure without any recurrence in 20-25% of patients
    - Whipple (pancreaticoduodenectomy) = greatest probability of cure
      - Removal of the entire gastrinoma triangle
      - Increased morbidity and mortality from the surgery – primarily recommended only for larger tumors
  
- **Long-term**
  - Monitoring of gastrin levels as screening
  - Metastatic disease is managed with chemotherapy and long-term acid suppression
    - Chemo targeting somatostatin receptors - octreotide
Puzzles #5 and 6: Incidental pancreatic findings

- **Pieces of the Puzzle #5:** Imaging from CT in the arterial and venous phase
  - How would you describe this lesion?
  - Does it change on the venous phase?
  - How can you confirm this lesion? Does this resemble another organ?

- **Pieces of the Puzzle #6:** Imaging from MRI T1 out of phase and T1 in phase
  - How would you describe this lesion?
  - Does it change on the T1 out of phase?
  - What does this change suggest about the contents of this lesion?
Puzzles #5 and 6: Intra-pancreatic lesions mimicking neoplasms!

**Figure A:** Axial CT image in the arterial phase demonstrating an arterial enhancing lesion in the tail of the pancreas (blue arrow).

**Figure B:** Axial CT imaging in the venous phase demonstrating the same lesion (blue arrow) with venous enhancement following the enhancement pattern of the spleen (red arrow). This incidental lesion was felt to represent an intra-pancreatic spenule.

**Figure C:** Axial T1 out of phase image demonstrating “india-ink” artifact surrounding the intra-pancreatic lesion suggesting fluid fat interface (blue arrow).

**Figure D:** Axial T1 fat saturated image demonstrating signal drop out of this fat containing intra-pancreatic lesion (blue arrow). This incidental lesion was felt to represent an intra-pancreatic lipoma.
References:


