Cystic Pancreatic Tumors - Radiologic-Pathologic Correlation

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Objectives

- Review clinical features of the most common cystic pancreatic tumors
- Discuss pathologic features that contribute to their imaging appearance

### IPMN – Classification

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<th>WHO</th>
<th>ARIP</th>
<th>5 yr survival</th>
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<td>Adenoma</td>
<td>Noninvasive</td>
<td>Low-grade dysplasia</td>
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<tr>
<td>borderline</td>
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<td>Moderate dysplasia</td>
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<tr>
<td>In situ carcinoma</td>
<td></td>
<td>High-grade dysplasia</td>
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<tr>
<td>Invasive carcinoma</td>
<td>Invasive</td>
<td>IPMN with invasive component</td>
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### IPMN – Clinical

- Mean age 65, 60% male
- Symptoms - abdominal or back pain, weight loss, recurrent pancreatitis
- Associations
  - Extrapancreatic malignancies 30%
  - Ductal adenocarcinoma
  - Peutz-Jeghers

### IPMN – Pathology

- Papillary projections
- Thick mucin ducts
- Columnar epithelium – intestinal, pancreateobiliary, or gastric differentiation
- Main duct, branch duct, or combined types

Overview

- Cystic neoplasms
  - Intraductal papillary mucinous neoplasm (IPMN)
  - Mucinous cystic neoplasm
  - Microlithic serous cystadenoma
- Solid pseudopapillary tumor
- Differential considerations
  - Acinar cell carcinoma
  - Pseudocyst
  - Lymphoepithelial cyst

IPMN

- Mucin-producing neoplasm within the main pancreatic duct or its branches
- 20% of cystic pancreatic lesions
- All malignant potential
**IPMN – Main & Combined Types**

- Diffuse or segmental main duct dilatation
- Mural nodules

**IPMN – Main & Combined Types Diff Dx**

- Diffuse main pancreatic duct dilatation
  - Obstruction
  - Pancreatic or ampullary mass
  - Cholelithiasis
  - IPMN
  - Chronic pancreatitis

**IPMN – Main & Combined Types**

**IPMN – Branch Duct Type**

- Unilocular cyst or clustered small cysts with lobulated margins
- Mural nodularity
- Communication with ducts

**IPMN – Main & Combined Types**

**IPMN – Branch Duct Type**

**IPMN – Main & Combined Types**

- Other imaging features
  - Distal ulcer of major papilla into duodenum
  - Pancreatic atrophy
  - Rim enhancement of duct walls

**IPMN – Branch Duct Type Diff Dx**

- Pancreatic cyst
  - Cystic neoplasm
    - Branch duct IPMN
    - Mucinous cystic neoplasm
    - Oligocystic serous cystadenoma
    - Solid pseudopapillary tumor
  - Pseudocyst
  - Nonneoplastic cysts – retention cyst, lymphoepithelial cyst, epidermoid/dermoid, duplication cyst, echinococcus
IPMN – Invasive Carcinoma

- Findings associated with malignancy or invasion
  - Main duct or combined types, diffuse main duct involvement
  - Branch duct type >3 cm, larger caliber communicating tract
  - Mural nodule >5.10 mm
  - Common bile duct dilatation
  - Solid mass
  - Lymph node, liver, or peritoneal metastasis

IPMN – Consensus Guidelines

- Main duct & combined IPMN
  - Malignancy 60-95%, mean 70%
  - Invasive carcinoma 23-57%, mean 43%
  - Most if not all benign main duct IPMNs progress into invasive cancer
  - Recommendation: resect all main duct and combined IPMNs in good surgical candidates with reasonable life expectancy


IPMN – Invasive Carcinoma

- Branch duct IPMN
  - Malignancy 6-40%, mean 25%
  - Invasive carcinoma 0-31%, mean 15%
  - Recommendation: resect if symptomatic, consider observation if asymptomatic and <3 cm without mural nodularity and without main duct dilatation >6 mm


IPMN – Colloid Carcinoma

- Mucinous Cystic Neoplasm
  - Mucin-producing neoplasm without ductal involvement
  - 10% of cystic pancreatic lesions
  - All malignant potential

Mucinous Cystic Neoplasm – Clinical

- Mean age 40-50, 95% women
- Nonspecific symptoms when large, 20-30% incidental
- Cyst fluid – elevated carcinoembryonic antigen (CEA), normal amylase
  - IPMN elevated amylase and CEA
Mucinous Cystic Neoplasm - Pathology
- Micro
  - Columnar epithelium with surrounding ovarian-type stroma
  - Spectrum of low, moderate, or high-grade dysplasia and associated invasive carcinoma
- Gross
  - 17% solid & solid
  - Multilocular cystic mass with fibrous capsule
  - Cysts contain smooth lining, papillary projections, or mural nodules

Mucinous Cystic Neoplasm - CT
- Multilocular cystic mass with enhancing wall and septa
- Wall-circumscribed, smooth round margin
- Calcifications in wall or septa 10%

Mucinous Cystic Neoplasm - MR
- Cyst contents
  - High T2, low T1 signal intensity most common
  - May vary with high proteinaceous content or hemorrhage
- Delayed enhancement of fibrous capsule

Mucinous Cystic Neoplasm - Invasive
- Findings associated with malignancy or invasive component
  - Size >4 cm
  - Mural nodules

Mucinous Cystic Neoplasm - Invasive
- Noninvasive – complete resection curative
- Associated invasive carcinoma
  - Resectable 5-10% 5 year survival
  - Unresectable 10-20% 2 year survival
- Recommendation: resect all mucinous cystic neoplasms unless contraindications since life expectancy will allow development of invasive carcinoma

Microcystic Serous Cystadenoma
- Benign serous fluid-producing neoplasm
- 20% of cystic pancreatic lesions

Microcystic Serous Cystadenoma – Clinical
- Mean age 62, 75% women
- Nonspecific symptoms when large, 40% asymptomatic
- Cyst fluid – elevated pancreatic and salivary isoenzymes, negative tumor markers
Microcystic Serous Cystadenoma – Pathology
- Clear cuboidal cells with centroacinar cell differentiation
- Sponge-like appearance of numerous small cysts
- Fibrous septa +/− central stellate scar
- Thin incomplete pseudocapsule

Microcystic Serous Cystadenoma – Solid Serous Variant
- No cysts
- Similar appearance to endocrine tumors and hypervascular metastases with marked arterial enhancement

Microcystic Serous Cystadenoma – Imaging
- Numerous small cysts or honeycomb appearance with lobulated margins
- Central scar – 30%, may calcify
- Only hypervascular cystic pancreatic neoplasm

Microcystic Serous Cystadenoma – von Hippel-Lindau
- Autosomal dominant, chromosome 3p25
- Prevalence 1/31,000 – 1/53,000
- Multiple clear cell neoplasms
  - CNS and retinal hemangioblastomas
  - Renal cysts and renal cell carcinoma
  - Pheochromocytoma
  - Pancreatic involvement – 50–80%, pancreatic cysts, serous cystadenoma, endocrine tumors, adenocarcinoma

Microcystic Serous Cystadenoma – Imaging
- Younger age – average 42
- Often multifocal & macrocystic but identical pathology to microcystic serous cystadenoma
- Diffuse cystic replacement may occur

Microcystic Serous Cystadenoma – von Hippel-Lindau
- Younger age – average 42
- Often multifocal & macrocystic but identical pathology to microcystic serous cystadenoma
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Solid Pseudopapillary Tumor
- Unclear cell of origin
- <5% of cystic pancreatic lesions
- All malignant potential
Solid Pseudopapillary Tumor – Clinical

- Mean age 22, 96% female
- Rare rupture can cause hemoperitoneum
- Good prognosis
  - 15-20% local invasion or distant metastasis
  - 95% 5 year survival

Solid Pseudopapillary Tumor – MR

- Hemorrhage – high T1, heterogeneous T2 signal intensity
- Fibrous capsule – low T2, delayed enhancement
- Enhancement – early mid arterial, progressive portal venous and delayed phases

Solid Pseudopapillary Tumor – Pathology

- Micro
  - Solid areas of uniform polygonal cells
  - Degeneration results in pseudopapillae of stromal cells surrounding capillaries

Solid Pseudopapillary Tumor – Pathology

- Gross
  - 10% solid & cystic
  - Central cavities with hemorrhage and necrosis
  - Thick capsule that may calcify

Solid Pseudopapillary Tumor – Diff Dx

- Pancreatic neoplasms with cystic degeneration
  - Solid pseudopapillary tumor
  - Acinar cell carcinoma
  - Pancreatic endocrine tumor
  - Ductal adenocarcinoma

Solid Pseudopapillary Tumor – CT

- Peripheral capsule – calcification 30%
- Varying cystic and solid components
- Enhancement – early heterogeneous peripheral with progressive fill-in

Acinar Cell Carcinoma

- Malignant neoplasm with pancreatic enzyme production
- 1-2% of exocrine pancreatic neoplasms
- 15% of pediatric pancreatic neoplasms
Acinar Cell Carcinoma – Clinical
• Mean age 56, M:F 2:1
• Lipase hypersecretion syndrome – 10-15%
  – Lipase >10,000 U/dl
  – Subcutaneous nodules
  – Polyarthritis
• Poor prognosis
  – 8% 5 year survival

Nonneoplastic Cysts
• Pseudocyst
• Lymphoepithelial cyst
• Mucinous nonneoplastic cyst
• Epidermoid/dermoid
• Retention cyst
• Duplication cyst
• Echinococcal cyst

Acinar Cell Carcinoma – Pathology
• Hypercellular, acinar structure with glandular or trabecular appearance
• Large, circumscribed – average 10 cm
• Necrosis & cystic change common

Pseudocyst
• 30% of pancreatic cystic lesions
• Most history of pancreatitis
  – Incidental cystic lesion more likely to be neoplastic
• Pathology
  – No epithelial lining
  – Necrotic-hemorrhagic material surrounded by inflamed fibrous capsule

Acinar Cell Carcinoma – Imaging
• Cystic or necrotic areas when large
• Calcification
• Capsule
• Rare intratumoral hemorrhage

Pseudocyst – Imaging
• Pancreatic fluid collection
• Thin or thick uniform enhancing wall
• Debris – low 12 signal, nonenhancing

Acinar Cell Carcinoma – Imaging

Lymphoepithelial Cyst
• 0.5% of pancreatic cystic lesions
• Mean age 56, 80% male
• Pathology
  – Squamous epithelium lined cyst filled with keratinaceous debris
  – Surrounding band of lymphoid tissue
Lymphoepithelial Cyst – Imaging

- Unilocular or multilocular cyst
- Keratinized material – central hypodense, hyperechoic, low T2 areas
- May contain lipid

Incidental Cyst – ACR Guidelines

- <2 cm – follow-up in 1 year with MRI
  - No growth – no further follow-up
  - Growth – treat as 2-3 cm category
- 2-3 cm – characterize with MR/MRCP
  - Serous cystadenoma – follow-up every 2 years
  - Branch duct IPMN – follow-up every 6 months for 2 years, then yearly
  - Uncharacterized – follow-up yearly
- >3 cm
  - Serous cystadenoma – consider resection when >4 cm
  - Other cystic neoplasm or uncharacterized – cyst aspiration
  - Then resect depending on comorbidities and risk

Key Imaging Features

- IPMN
  - Main/combined – main duct dilatation, mural nodules
  - Branch – communication with main duct, mural nodules
- Mucinous cystic neoplasm
  - Pancreatic body or tail
- Encapsulated multilocular mass
- Microcystic serous cystadenoma
  - Honeycomb +/- central scar
  - Only hypervascular cystic neoplasm
- Solid pseudopapillary tumor
  - Encapsulated solid and cystic mass
  - Evidence of hemorrhage