Para-duodenal Pancreatitis An Imitator of Pancreatic Adenocarcinoma: A Diagnostic Challenge, MRI Characteristics, Mimics And Histopathological Correlation

Pardeep Mittal, MD; Peter A Harri, MD; Lauren F Alexander, MD; Courtney Coursey-Moreno

Emory University School of Medicine
Atlanta, Georgia USA
Disclosures

Neither the authors nor their immediate family members have a financial relationship with a commercial organization that may have a direct or indirect interest in the content.
Learning Objectives

• To demonstrate MR Imaging characteristics of paraduodenal pancreatitis differentiating from pancreatic adenocarcinoma

• To educate participants regarding variety of lesions which mimic paraduodenal pancreatitis such as chronic pancreatitis, pancreatic adenocarcinoma, ampullary carcinoma, giant cell reaction, Bruner gland hyperplasia etc.
Pathophysiology

- Paraduodenal pancreatitis may result from obstruction or anatomic variation of the accessory duct (Adsay and Zamboni, 2004)
- Risk factors are the same as for other forms of chronic pancreatitis, with alcohol abuse the most common identified etiology
  - Other factors, including peptic ulcer disease, gastric resection, preexisting cystic or solid masses in the duodenum
    - Heterotopic pancreas in duodenal wall
    - Brunner gland hyperplasia
  - Altered flow pattern in the main pancreatic duct, may also play a role (Shanbhogue et al., 2009)
- Disease process is centered within the submucosa of the second portion of the duodenum, GI tract obstructive symptoms—upper abdominal pain, nausea, and vomiting—are prominent

In the segmental form of paraduodenal pancreatitis, the pancreatic head, which is drained by the accessory duct, will also be inflamed
Clinical Presentation

• Similar to both chronic pancreatitis and pancreatic cancer
  ➢ Usually middle-aged men with history of alcohol abuse
  ➢ Acute setting: Postprandial abdominal pain, vomiting, or acute gastric outlet obstruction
  ➢ Chronic setting: Chronic weight loss, jaundice
  ➢ Prospective diagnosis is very uncommon; difficult to exclude malignancy with imaging or biopsy
  ➢ Tumor markers are usually normal
  ➢ Surgery (Whipple procedure) may be required to rule out malignancy or due to intractable symptoms

Diagnostic clue: Curvilinear soft tissue between pancreas and duodenum
Pancreatic Heterotopia

- Pancreatic heterotopia is presence of pancreatic tissue at aberrant location (lacks anatomical and vascular continuity with proper pancreas)
- Cystic transformation with heterotopia most often occur in second part of duodenum
Brunner Gland Hyperplasia

No neoplastic hyperplasia of duodenal submucosal glands
- Diffuse type: Multiple, small, submucosal nodules < 5 mm
- Solitary type: Solitary, sessile or pedunculated lesion > 5 mm

Diagnostic Clue:
- Polypoid, multicystic mass in duodenum
- No significant enhancement
- Tapering duct without cutoff

MRCP
T1 post
T2

Diagnostic Clue: "cobblestone" appearance in proximal duodenum
Paraduodenal Pancreatitis

Pancreatic groove is a theoretical space defined by pancreatic head (medially), 2nd portion of duodenum (laterally), 3rd portion of duodenum and IVC (posteriorly), and duodenal bulb (superiorly) also contains distal common bile duct (CBD), main/accessory pancreatic ducts, and major/minor papilla

- Chronic pancreatitis affecting pancreaticoduodenal groove
  - Pure form (sheet like): Affects only pancreaticoduodenal groove
  - Segmental form (mass like): Affects pancreaticoduodenal groove and extends medially into pancreatic head
MR Imaging Features

• Sheet-like thickening of the pancreatic groove (mildly T1 hypointense)
• Variable T2 signal (depending on acuity):
  • T2 hyperintense in acute phase
  • T2 hypointense chronic
  • T2 hyperintense cysts in medial duodenal wall or groove
• Pancreatic head may be enlarged with low T1 signal in segmental form (due to fibrosis/atrophy)
• Duodenal wall thickening leading to stenosis
• T1WI C+: Delayed enhancement of affected areas
• MRCP: Smooth narrowing of distal CBD and pancreatic duct with widened space between ampulla and duodenal lumen
Paraduodenal Pancreatitis

- Sheet-like mass
- Cystic changes in duodenum
- Delayed enhancement
- Regular tapering of CBD
Paraduodenal Pancreatitis

Extensive cystic changes in duodenum and pancreatic head and delayed enhancement of mass in groove

Sheet-like mass delayed enhancement regular tapering of CBD
Differential Diagnosis

- Adenocarcinoma of the pancreatic head
- Duodenal or ampullary carcinoma
- Chronic pancreatitis
- Cystic lesions of pancreatic head or duodenum
- Multiple entities can coexist!
  - There are case reports of pancreatic adenocarcinoma developing within the pancreatic elements present in the duodenal wall in patients with a history of paraduodenal pancreatitis
    - In part because of this, some authors recommend pancreaticoduodenectomy (Whipple procedure) as the definitive treatment of choice for paraduodenal pancreatitis

Pancreatic Adenocarcinoma

Malignancy arising from ductal epithelium of exocrine pancreas

- Most common pancreatic neoplasm (85-90% of all pancreatic tumors)
- High mortality, overall 5-year survival of 4% (resectable 25%, unresectable 1%)
- Although predominantly solid, cyst-like features (cystic degeneration/necrosis, retention cysts, side-branch ductal obstruction or attached pseudocysts) are associated with 8%

Differentiating features from paraduodenal pancreatitis
- Mass confined to pancreatic head
- Abrupt cutoff of CBD and pancreatic duct with “double duct” sign
- Atrophy of pancreas
- Signs of vascular invasion or metastases
- Lack of cystic change (usually)

MRI:
- T1WI Tumor is conspicuous appearing low signal and juxtaposed against high signal pancreatic parenchyma
- T2WI less useful, as tumors isointense to pancreas
- T1WI C+ tumors demonstrating progressive delayed enhancement

Cystic changes are more commonly seen in large poorly differentiated adenocarcinomas
Pancreatic Adenocarcinoma

- No cystic change in duodenum or pancreatic head
- Abrupt cutoff of CBD and pancreatic duct
- T1 hypointense lesion in pancreatic head/neck
- Hypoenhancing

Thickening of duodenal wall or cystic change is uncommon
Cystic and Solid Pancreatic Adenocarcinoma

- Cystic and solid mass in the pancreatic head
- Delayed enhancement
- Atrophy of distal pancreas
Pancreatic Adenocarcinoma

• Cystic change in pancreatic head
• T2 heterogeneous lesion in pancreatic head/neck
• Hypoenhancing lesion
• Abrupt cutoff of CBD and pancreatic duct which are dilated

Diagnostic Clue: Poorly marginated, hypoenhancing mass with abrupt obstruction of pancreatic duct ± common bile duct
Acinar Cell Cystadenocarcinoma

Rare 1-2% of exocrine neoplasms
   Pancreatic enzyme production
      (trypsin and lipase)
Most common site
   head of the pancreas 56%
   Tail of the pancreas 36%
   Body of the pancreas 8%
Lipase hypersecretion syndrome in 10-15%
   Elevated lipase levels causing fat necrosis, subcutaneous nodules and polyarthritis

- Large (usually >10cm)
- Hypoenhancing
- Varying degrees of necrosis with cystic change
- Well-marginated with surrounding capsule
- Can have intratumoral calcification and hemorrhage

Acinar cell carcinoma may not cause jaundice even when the tumor is located in the head of pancreas
Heterogeneous group of malignant epithelial neoplasms (adenocarcinoma) arising from ampulla of Vater

- **Enhancing (not delayed) mass** in the region of the ampulla
- **Abrupt cutoff** of markedly dilated ducts
- **No cystic changes**

Diagnostic clue:
Soft tissue mass centered in ampulla
Double duct sign with obstruction of both common bile duct (CBD) and pancreatic duct (PD)
Duodenal Carcinoma

- Intraluminal mass at or distal to ampulla of Vater
- Irregular thickening of duodenal wall
- Concentric narrowing of duodenal lumen
- Polypoid intraluminal mass
- Local lymphadenopathy and local infiltration
- Biliary ± pancreatic duct dilatation
  - periampullary tumors
- 15% in 1st portion of duodenum
- 40% in 2nd portion of duodenum
- 45% in distal duodenum

Mass in duodenum involving ampulla Causing double duct sign
Diffuse delayed enhancement

Mass centered in duodenum (not in pancreatic groove) without cystic change
Duodenal Adenoma

- Adenomas account for one-half of all benign duodenal neoplasms
- Villous adenomas are subtype of adenomatous polyps
- Mostly found in the second portion of the duodenum, with malignant transformation rate of 30%–60% arise in:
  - ampulla
  - periampullary region
- Patient may present with no symptoms other than vague dyspepsia,
- May present with obstructive jaundice, depending on the location of the tumor.

- Mass like thickening of the duodenum
- Early enhancement
- And abrupt cutoff of CBD and pancreatic duct is more consistent with mass
Main Duct IPMN with Adenocarcinoma

**Main duct IPMN**

- Markedly dilated, tortuous MPD $\geq 5$ mm (segmental or diffuse) without evidence of distal obstructing mass and often with "bulging" ampulla filled with fluid (mucin) at duodenal sweep
- Presence of polypoid enhancing nodularity within MPD lumen is very suspicious for malignancy
- Pancreas often atrophic overlying dilated duct

**Complex cystic lesion** in the head of pancreas and groove with soft tissue components

**Enhancing soft tissue components**

**Dilatation of pancreatic duct and bile duct**

IPMN: Metaplasia of mucin producing columnar epithelium within pancreatic ducts, commonly with papillary projections

Endoscopically characterized gaping papilla extruding mucin "fish mouth papilla" 

Seen in 25-50% main duct IPMNs
Acute Pancreatitis

Acute pancreatitis:
- Caused by acinar cell injury and premature activation of trypsinogen to trypsin
- Focal or diffuse enlargement of the pancreas
- Not primarily centered in pancreaticoduodenal groove, usually involves entire pancreas
- Associated with peripancreatic inflammation, including retroperitoneal fluid and fat stranding
- Should resolve on follow-up examinations
- Pseudocyst is the most common complication of acute pancreatitis

- Inflammatory mass in groove and pancreatic head with delayed enhancement
- Surrounding increased T2 signal on fat-saturated images consistent with acute inflammation
- Prominent pancreatic duct
Acute on Chronic Pancreatitis with Giant Cell Reaction

Fibroinflammatory mass related to chronic pancreatitis may be very difficult to distinguish from pancreatic adenocarcinoma

- Inflammatory mass in groove with delayed enhancement
- Surrounding increased T2 signal on fat-saturated images consistent with acute inflammation
- Atrophy of tail from chronic pancreatitis
Chronic Pancreatitis

- Progressive, irreversible inflammatory damage to pancreas resulting in parenchymal fibrosis, morphologic changes, and loss of endocrine/exocrine function
- Atrophic pancreatic parenchyma with a dilated, beaded main pancreatic duct (MPD) and intraductal calculi

MRI:
- **T1WI** loss of normal high signal of Pancreatic parenchyma
- **Post contrast:** Decreased enhancement on arterial phase, delayed progressive enhancement
- Pancreatic duct: Dilated (> 3 mm), irregular with strictures and dilated side branches
- Stones within pancreatic duct appear as signal voids

- **Diffuse decreased T1 signal** of the pancreas
- **Diffuse atrophy** of the gland
- **Pseudocyst** in the pancreatic head but no solid mass
- **Adjacent duodenum** is normal

Adjacent duodenum is normal
Lymphoepithelial Cysts

Rare (<1% of all pancreatic cysts)
Benign
Usually middle aged men (mean age 55 years, M>F 4:1)
Cysts are lined by squamous epithelium, filled with keratinaceous debris and surrounded by a band of dense lymphoid tissue
MR Imaging:
- findings are not well described in the literature, variable MR appearance either unilocular or multilocular

- **Heterogeneous multilocular cystic mass** in the uncinate process of pancreas
- **Hyperintense** T1 signal due to hemorrhagic /proteinaceous material
- **No contrast enhancement**
Squamoid Cyst Of Pancreatic Duct

Unilocular
Median size 1.5 cm
May be large and have high CEA levels
Resected with clinical impression being IPMN

- Unilocular cyst in pancreatic head
- Obstructing the biliary and pancreatic ducts
- No abnormal enhancement or internal complexity
Treatment

Principal Indications for Surgery
- Pain
- Jaundice
- Gastrointestinal symptoms
  - Marked duodenal stenosis (or extensive fibrosis)
  - Pancreatic insufficiency
  - Weight loss
  - Steatorrhea
  - Diabetes

Acute phase:
- Conservative treatment
  - Bed rest
  - Fasting
  - Analgesics
  - Parenteral nutrition
- Reassess after 4-6 weeks (symptoms improve in most patients but relapses can occur)

Resistant to medical treatment:
- Groove pancreatitis occasionally resistant to medical treatment so surgical treatment is required
  - Pancreaticoduodenectomy (Whipple procedure)
  - Pylorus sparing pancreaticoduodenectomy
Paraduodenal pancreatitis is a focal form of pancreatitis centered in pancreatic elements normally present in the submucosa of the second portion of the duodenum in the region of the minor papilla.

Clinical and imaging features of paraduodenal pancreatitis can mimic multiple disease processes, with the most important differential consideration being pancreatic adenocarcinoma.

The most specific imaging features of paraduodenal pancreatitis, including duodenal wall thickening, enhancement, and cystic change, reflect the underlying pathophysiology and anatomy.
References


THANK YOU

pmittal@emory.edu