Approach to the Imaging of the Dilated Bile Duct

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Learning Objectives

1. Present a practical and imaging facilitated approach to biliary duct dilatation.
2. Review the pathophysiology and clinical manifestations of the spectrum of dilated bile duct pathologies including obstructive causes such as choledocolithiasis, cholangiocarcinoma, and IPMN-B, and non-obstructive causes such as Caroli disease, choledochal cyst, and primary sclerosing cholangitis.
3. Discuss the utility and limitations of the various imaging modalities used for the diagnosis of bile duct dilatation.
4. Describe and demonstrate the spectrum of imaging findings of bile duct dilatation on ultrasound, CT, MRI, and MRCP.
5. Review examples of important imaging findings for distinction between various dilated bile duct pathologies.
6. Review treatment options for bile duct pathologies.
Background

- Bile duct dilatation can be due to a myriad of benign and malignant etiologies, including obstructive causes such as choledocolithiasis, IPMN-B and cholangiocarcinoma, and non-obstructive causes such as Caroli disease, choledochal cyst, and primary sclerosing cholangitis.

- Thus, a systematic and thorough approach is critical for identifying the most accurate diagnosis in order to provide the appropriate management.

- Key features include:
  1. presence or absence of obstruction and/or stones
  2. location of dilatation
  3. communication with biliary tree
  4. type of pathology (e.g. neoplastic, inflammatory)
Outline

Obstructive biliary disorders
- Stone (choledocolithiasis)
- Stricture
  - Cholangiocarcinoma
  - IPMN-B

Non-obstructive biliary disorders
- Caroli disease
- Choledochal cyst
- Recurrent pyogenic cholangitis
- Primary sclerosing cholangitis
Obstructive Biliary Disorders
Choledocholithiasis

WHAT?
- Presence of gallstones within bile ducts (typically common hepatic duct and common bile duct).

CLINICAL PRESENTATION
- Uncomplicated: asymptomatic, or cholestatic picture.
- Complicated: acute pancreatitis, cholangitis

MANAGEMENT
- Remove stone (endoscopically or surgically) and treat complications.
  - Treatment of choice: ERCP with sphincterotomy, however it is associated with a complication rate of 6-24% (10 year follow-up).
Choledocholithiasis

KEY IMAGING FINDINGS

- **U/S**
  - Visualisation of stones – echogenic foci & posterior shadowing.
  - Dilated bile duct (>6mm + 1mm per decade >60)

- **CT** – sensitivity 65-88%
  - Target sign (central rounded density with surrounding low attenuating bile or mucosa)
  - Biliary dilatation

- **CT cholangiography** – sensitivity 93%, specificity 100%
  - However, 1. high contrast complication rate, 2. decreased excretion due to obstruction

- **MRCP** (gold standard) – sensitivity and specificity approaching 100%, replacing ERCP as Dx tool.
  - T2: filling defects
Cholangiocarcinoma

WHAT?

- Extrahepatic or perihilar cholangiocarcinoma (accounting for 90% of cholangiocarcinomas) are the subtypes that typically cause occlusion of bile ducts (see image).

CLINICAL PRESENTATION

- Usually age >65 with pruritus, jaundice.
- Increased risk in patients with PSC and choledochal cysts due to chronic biliary inflammation.

MANAGEMENT

- Resection is the only cure, where adjuvant or neoadjuvant chemotherapy may also be necessary. Recurrence is common.

Imaging is key in diagnosis and management, due to multidisciplinary approach.

Cholangiocarcinoma

Important considerations in imaging workup for proper evaluation and resectability, if cholangiocarcinoma suspected.

- Initial U/S or CT should be followed by MRI or MR cholangiography.
- Image prior to procedures (e.g. drainage, stenting).
- Early and portal venous phases important for relationship of tumor and hepatic artery.

**KEY IMAGING FEATURES**

- Dilated segmental bile ducts with lack of communication between right and left hepatic ducts.
- Crowding of bile ducts
- Ductal wall thickening
- Lobar atrophy

Dilated common hepatic and left hepatic duct with significant intra-hepatic biliary dilatation on CT.
**IPMN-B**

- Intraductal papillary mucinous neoplasms of the bile duct (IPMN-B) has recently been re-classified as a distinct type of bile duct neoplasm.
  - Unlike biliary mucinous cystic neoplasm (MCN), IPMN-B lacks ovarian-like stroma and communicates with the bile ducts.
  - Unlike intrahepatic cholangiocarcinoma (ICC), IMPN-B can be resected and demonstrates favorable prognosis.

- Therefore, correct diagnosis of IPMN-B is crucial.
IPMN-B

KEY IMAGING FEATURES

- **U/S:** viscous mucin as echogenic structure, though usually less viscous and anechoic.

- **CT & MRI:** large mural nodule in duct on contrast-enhanced studies is considered a sign of malignancy, but a small or flat tumor is difficult to detect, as mucin has similar density as bile.

- **MRCP & ERCP:** In addition to detecting large mural nodules, ERCP depicts mucin as defect in bile duct filling which is additional information to CT and MRI findings.

- **Endoscopic U/S & Intraductal U/S:** Can provide precise information regarding continuation between bile duct and cystic IPMN-B.
IPMN-B

MRCP
- arrows: hilar cystic tumor
- arrowheads: gallbladder
- diffusely dilated intrahepatic bile ducts

ERCP
- no definite hilar cystic tumor and gallbladder
- arrows: hypo-enhancement of the proximal portion of the right hepatic duct

Mismatch between ERCP and MRCP indicates a biliary obstruction by mucin.

1. CECT PV, 2. MR T2WI
3. MR T1W1, 4. Enhanced MR T1W1 showing irregular-shaped mural nodules in dilated upper CBD. Nodules show enhancement.

Takanami 2011
Non-Obstructive Biliary Disorders
Caroli Disease

WHAT?
- Congenital cystic dilation (large, saccular) of intrahepatic biliary ducts with a 1/1,000,000 incidence.

PATHOPHYSIOLOGY
- Malformation of ductal plate (precursor to intrahepatic bile ducts), causing bile ducts to be too numerous and ectatic.

CLINICAL PRESENTATION
- Recurrent cholangitis, portal hypertension and derivatives (hematemesis from varices, splenomegaly, cirrhosis) in younger population.
- Often associated with polycystic kidney disease and liver disease.

MANAGEMENT
- Localized: segmentectomy or lobectomy
- Diffuse: conservative; liver transplant

PROGNOSIS
- Poor, due to many complications: intraductal stones, cholangitis and abscess, cirrhosis, increased risk of cholangiocarcinoma

Levy 2009
Caroli Disease

KEY IMAGING FEATURES

- Distribution:
  - Segmental
  - Lobar
  - Diffuse

- Morphology:
  - Saccular (75%) – can help in DDx
  - Fusiform (25%)
  - Dilated intrahepatic bile ducts (IHBD)
  - Central dot sign: portal vein surrounded by dilated bile ducts

Levy 2009
Choledochal Cyst

WHAT?
- Congenital dilatation of extrahepatic bile duct

PATHOPHYSIOLOGY
- Dilatation of an anomalous pancreatico-biliary junction
- With contraction of sphincter of Oddi, enzymes back flow into cystic duct

CLINICAL PRESENTATION
- Usually age <10 with palpable mass and jaundice.
- Associated with cholangiocarcinoma, Klatskin tumor, distal cholangiocarcinoma.

DIAGNOSIS: Imaging is key.

MANAGEMENT & PROGNOSIS
- Resection, especially for most common types (I & IV) due to increased risk of cholangiocarcinoma (with risk reduced if cyst resected).

Levy 2009
Choledochal Cyst

KEY IMAGING FEATURES
- Dilated cystic structure that communicates with biliary system and is separate from gallbladder.
  - U/S & CT can be used to make the diagnosis, where ERCP, MRI, EUS might be utilized for better visualization of communication between cyst and biliary tree.

CLASSIFICATION: Todani
- Type I: true choledochal cyst with focal dilatation of extrahepatic duct
  - Most frequent type: 90-95%
  - (Type II: diverticulum of the extrahepatic duct)
  - (Type III: choledochocele – dilatation of distal part of bile duct)
- Type IV: dilatation of the entire extrahepatic duct with involvement of portions of intrahepatic ducts
  - (Type V: Caroli’s disease)
Type 1 Choledochal Cyst

CT: True choledochal cyst with focal dilatation of extrahepatic duct
Primary Sclerosing Cholangitis (PSC)

WHAT?
- Chronic progressive disorder of unknown cause of fibrosis causing inflammation, fibrosis and strictures of medium and large ducts and/or extrahepatic biliary tree.
- Associated with IBD.

CLINICAL PRESENTATION
- Fatigue, pruritis, jaundice (obstructive), IBD findings.

DIAGNOSIS
- Cholangiographic evidence of bile duct changes → imaging is key.

PROGNOSIS
- Complications: cholestasis, hepatic failure.
- 10-12 years without treatment.
Primary Sclerosing Cholangitis (PSC)

KEY IMAGING FEATURES

- Intrahepatic bile duct dilatation with
  - multifocal strictures (short segments: 3-5mm) and
  - mild dilatation.

U/S: Thickening of bile duct wall is one of earliest features

CT:

- Discontinuous dilatation
- Bile wall thickening at level of porta hepatis
- Lymphadenopathy

Cholangiography: used in initial diagnosis of disease

- Beading: alternating strictures and normal ducts
- Pruned-tree: distal bile ducts are narrowed
- Mural irregularity: irregular luminal margin
- Diverticula: specific for PSC
Recurrent Pyogenic Cholangitis

WHAT?

- Recurrent infections of biliary system with formation of intrahepatic pigmented stones and recurrent infection of unknown etiology, but many secondary to biliary parasites.

CLINICAL PRESENTATION

- Recurrent cholangitis, septic picture.

MANAGEMENT – Imaging key for Dx.

- 1. Acute complications: supportive management.
- 2. Prevention of long-term complications
  - Resection, clearance of stones, minimally invasive techniques

PROGNOSIS

- Long-standing infections lead to complications such as biliary cirrhosis and cholangiocarcinoma, many of which responsible of mortality.
Recurrent Pyogenic Cholangitis

KEY IMAGING FEATURES

- Initial evaluation with U/S: Ductal dilatation and stones in 85-90%.
- Additional evaluation: CT & Cholangiography (from ERCP, MRCP or PTC):
  - intra- and extrahepatic duct dilatation, enhancement of duct walls
  - straightened intrahepatic ducts with less acute or right-angled branching patterns (as a result of extensive periductal fibrosis)
  - decreased arborization and acute tapering of the peripheral ducts
    - classic "arrowhead"
    - "missing duct" sign: complete obstruction of a bile duct.
  - hepatic abscesses, bilomas, and stones

Intrahepatic lithiasis with focal dilatation, which can be indistinguishable from focal Caroli’s disease, thus clinical context is crucial.
Conclusion

Obstructive biliary disorders
- Stone (choledocolithiasis)
- Stricture
  - Cholangiocarcinoma
  - IPMN-B
  - Miscellaneous
    - Post-inflammatory (pancreatitis, post-radiation)
    - Inflammatory (AIDS cholangiopathy, biliary parasites)

Non-obstructive biliary disorders
- Caroli disease
- Choledochal cyst
- Recurrent pyogenic cholangitis
- Primary sclerosing cholangitis

A systematic approach, with consideration of:
1. presence or absence of obstruction and/or stones
2. location of dilatation
3. communication with biliary tree
4. type of pathology (e.g. neoplastic, inflammatory)

...is key to differentiate malignant vs benign disease.
References


