Cholangiocarcinoma: Current update and role of imaging

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Goals and Objectives

• Review the clinical presentation and distribution of cholangiocarcinoma (CC).
• Review the classification system, the characteristic imaging features, and typical growth patterns.
• Discuss important features which aid in determining potential resectability and staging of the tumor, which may affect prognosis.
• Illustrate post-resection appearance and complications, as well as typical pattern of metastatic spread.
Background and Etiology

• Cholangiocarcinoma (CC) is the second most common primary hepatic malignancy, after hepatocellular carcinoma.
• It is rare however, accounting for only 2% of all cancers.
• The tumor is associated with a poor prognosis, which may be improved if resection is deemed an option.
• CC arises from bile duct epithelium, anywhere along the intrahepatic or extrahepatic bile ducts.
• CC is more prevalent in men, between the 6th and 7th decades.
• The risk of developing CC increases with chronic inflammation of the biliary system. Risk factors include: primary sclerosing cholangitis, choledochal cysts, Caroli disease, hepatolithiasis, bile duct adenoma, Clonorchis infection, +HCV, among others.
Clinical Symptoms

- In perihilar tumors, clinical symptoms usually relate to bile duct obstruction, including: right upper quadrant pain, jaundice, pruritus, fatigue, anorexia/weight loss, dark urine, and clay-colored stools.
- In advanced disease, there can be a palpable mass, weight loss, and ascites.
- Typically, laboratory results will show an obstructive liver profile.
- Often, CC is clinically silent, leading to advanced stage at time of diagnosis.
Role of Imaging

- The role of imaging in CC is to provide noninvasive diagnosis and characterization of the tumor, determine stage and possible resectability, and screen high-risk patients for early detection.
- A range of modalities can be utilized to assist in diagnosis, staging, and assessment for potential resectability, including: CT, MR, MRCP, MRA, PET, ultrasound angiography, and cholangiography. (This exhibit will focus on CT, MR, and MRCP)
- MR and MRCP are deemed superior to CT for assessment of intraductal lesions, owing to better contrast resolution.
Classification, Staging Systems, and Intrahepatic Growth Patterns

- There are three broad categories of CC, separated anatomically into intrahepatic, perihilar, and distal extrahepatic.

- Perihilar CC are further divided according to the Bismuth-Corlette schematic (detailed on next slide).

- Staging of CC is according to the Tumor, Node, Metastasis (TNM) classification of the American Commission on Cancer.

- Three intrahepatic growth patterns have been described by the Liver Cancer Study Group of Japan: mass forming, periductal infiltrating, and intraductal.
The Bismuth-Corlette classification is a classification system for perihilar cholangiocarcinoma. It is based on the degree of ductal infiltration.

- **Type I**: Limited to CHD, below level of confluence of RHD and LHD.
- **Type II**: Involving the confluence of the RHD and LHD.
- **Type IIIa**: Type II + extends to bifurcation of RHD
- **Type IIIb**: Type II + extends to bifurcation of LHD
- **Type IV**: Extends to bifurcation of both RHD and LHD OR multifocal involvement.
- **Type V**: Stricture at the junction of CBD and CD.

CHD = Common hepatic duct  
RHD = Right hepatic duct  
LHD = Left hepatic duct  
CD = Cystic duct
Classic Imaging Features:
Mass-forming Cholangiocarcinomas

- Typically hypo-attenuating on unenhanced CT, with heterogeneous mild peripheral enhancement and delayed central enhancement.
- Capsular retraction may be present.
- Distal biliary ductal dilation.
- Portal vein (less commonly hepatic veins) may be narrowed by the mass.
- Rarely forms a tumor thrombus.
- With vascular invasion there may be associated lobar or segmental atrophy.
Classic Imaging Features: Periductal Infiltrating

- Intra-hepatic tumors identified as regions of periductal parenchymal thickening with narrowing or dilatation of the involved duct.
- Most common at the hepatic hilum.
- Distal dilatation of the biliary tree usually present.
Classic Imaging Features: Intraductal Tumors

- Alterations in duct caliber.
- Duct ectasia, with or without a discernible mass.
- If a polypoid mass is identified, it is usually hypo-attenuating on pre-contrast imaging and demonstrates enhancement.
72 year-old male.
Contrast-enhanced CT image demonstrates a large, heterogeneously enhancing lesion that encases the intrahepatic inferior vena cava and the confluence of the hepatic veins, extending into both lobes (thin solid arrow). Fat saturation post contrast T1 weighted images show the mass with initial continuous rim enhancement and progressive central patchy enhancement on delayed phase images (block arrow), consistent with cholangiocarcinoma. The mass is seen abutting the right hepatic vein and intrahepatic portal venous branches (dashed arrow).
51 year-old female with history of HCV and abdominal pain. Axial T2-weighted image shows a large, lobulated, well-circumscribed mass in the right hepatic lobe, with associated capsular retraction and peritoneal ascites (solid arrows).

Subsequent post-contrast delayed axial T1-weighted images demonstrate delayed/persistent enhancement (dashed arrows).

Mass was path-proven CC.
80 year-old male presents with painless jaundice. Enhanced, fat saturation T1 weighted images demonstrate thickened, enhancing wall of the proximal common bile duct (solid arrows), as well as intrahepatic peri-ductal enhancement and signal abnormality involving the right hepatic lobe, consistent with peri-biliary spread of cholangiocarcinoma (dashed arrows). Intrahepatic biliary dilatation is also noted.
60 year-old male with history of HCV presents abnormal liver profile. Pre- and post-contrast T1 weighted fat saturation images demonstrate a circumferential mass encasing the common hepatic duct at the bifurcation which progressively enhances following contrast administration, compatible with Bismuth II CC.

The mass also resulted in moderate narrowing of the CHD and distal LHD, as well as high-grade stricture of the distal RHD.

Thick slab MRCP image reveals marked intrahepatic biliary ductal dilatation and non visualization of the strictured segment of CBD.
46 year-old male presents with jaundice and abnormal liver profile. T1 weighted images from a high resolution isotropic volume excitation sequence demonstrate moderate intrahepatic biliary ductal dilatation secondary to a polypoid intraductal mass, centered at Klatskin point, which extends to the level of the pancreatic head.

On three minute delayed imaging, the lesion demonstrates delayed enhancement.

No evidence of intrahepatic biliary ductal extension of this lesion.
Distal Extrahepatic

78 year old female presenting with painless jaundice. MRCP image reveals moderate to severe intrahepatic biliary dilatation. There is abrupt stricturing of the common bile duct approximately 2-cm distal to the confluence, with areas of subtle nodularity.

T₁ post-contrast images from liver acquisition with volume acceleration (LAVA) sequence with 15 minute delay confirm thickening of the common bile duct at the level of the stricture, with delayed enhancement, compatible with cholangiocarcinoma.

T₁ post-contrast LAVA sequence image reveals an enlarged portocaval lymph node anterior to the head of the pancreas, medial to the second portion of the duodenum, which was proven to be metastasis after resection and regional dissection.
Associated Features

- Lobar or segmental atrophy
- High signal on pre-enhancement T1 in the periphery of segments or lobes where tumor is present without portal obstruction around areas of duct dilatation distal to the tumor
- Biliary dilatation and duct wall enhancement
- Lymph node enlargement
- Vascular involvement approaches 50% in hilar CC
66 year-old female.
Single shot fast spin echo T2 weighted image demonstrates an ill-defined hilar mass centered at, and splaying, the portal vein bifurcation.

Post contrast fat saturation T1 weighted images reveal that the mass invades the right portal vein and a focal area of the portal vein bifurcation (solid arrow). The left portal vein is patent.

The mass abuts the right hepatic artery, but the left hepatic artery is not in contact with the tumor (dashed arrows).
Associated Features: Biliary Dilatation and Lobar Atrophy

49 year old male with right upper quadrant pain and jaundice.

Post contrast coronal T1 fat saturation images demonstrate a lesion in segment IVa, with progressive, delayed enhancement, causing moderate left and mild right intrahepatic biliary dilatation and associated atrophy of the left hepatic lobe (circle and solid arrows).
68 year old female with known history of CC. Sequential axial MR images from an IN/OUT phase post BH sequence demonstrate intrahepatic periductal enhancement and signal abnormality involving the right hepatic lobe.

Associated Features:
Duct Wall Enhancement
Treatment & Prognosis

- Prognosis is extremely poor with a five-year survival rate of 1%; median survival of 6 months.
- Chemotherapy and radiation have failed to show significant benefit.
- Only potential cure is complete surgical resection or liver transplantation.
- Palliative surgery can be undertaken to improve obstructive symptoms.
- Similarly, percutaneous and endoscopic procedures, including arterial chemoembolization and radiofrequency ablation, can be attempted for palliation.
Assessment of Resectability

- Imaging is performed to screen for local and distant metastatic disease.

Criteria for unresectability:
- Bilobar infiltration beyond second order bile duct branches
- Invasion of major vessels [including the main portal vein or main hepatic artery, both right and left main branches of the portal vein, or involvement of the main portal vein in one lobe and the main hepatic artery in the contralateral lobe].
- Combination of vascular involvement in one lobe and extensive bile duct invasion in contralateral lobe.
- Lymph node metastasis beyond N1.
- Hepatic or distant metastasis.
Post-Operative Appearance

47 yo male with history of periductal CC. Axial and coronal contrast-enhanced CT images demonstrate expected pneumobilia secondary to Roux-en-Y with hepaticojejunostomy.
Complications

Prolonged biliary stasis from malignant obstruction may cause acute complications including ascending cholangitis, hepatic abscess, or portal vein thrombosis.

48 year old male with history of hilar CC. Axial contrast-enhanced CT revealed moderate intrahepatic ductal dilatation secondary to an obstruction at the level of the ductal confluence.

Axial and coronal enhanced CT images demonstrated a large, multi-lobulated right hepatic lobe abscess, with associated hypo-perfusion of the right hepatic lobe. The abscess extends to the capsular surface, with possible extension into the peritoneal cavity (solid arrows).
Typical Patterns of Metastases

- Lymph nodes- Up to 50% of patients have positive nodes at the time of diagnosis. Portocaval and porta hepatis being the most common sites.
- Peritoneal- Up to 20% of patients have peritoneal involvement at the time of diagnosis. This may appear as peritoneal thickening and/or enhancing nodules.
- Pulmonary and hepatic metastasis are less common, but can occur.
Metastases: Regional Lymph Nodes

64 year old male with unexplained weight loss and elevated alkaline phosphatase. Post contrast fat saturation T1 weighted images revealed a peripherally enhancing left hepatic lobe mass (solid arrows) with associated distal intrahepatic biliary dilatation (dashed arrow). As well as an enlarged/enhancing retroperitoneal lymph node (circle). A conglomeration of enlarged porta hepatis nodes are visualized on an axial fat saturation T2 weighted image (oval).
Metastases: Peritoneal Carcinomatosis

63 year old female with history of intrahepatic CC and subsequent biliary drain placement for decompression of biliary obstruction. Contrast-enhanced CT demonstrates diffuse peritoneal carcinomatosis (solid arrows) and new bilateral adnexal lesions consistent with ovarian metastasis (dashed arrows).
Conclusions

• Cholangiocarcinoma has a poor prognosis with high morbidity and mortality.

• Specific features on CT and MRI can allow a confident diagnosis and aid with determining resectability, which can improve prognosis and prevent unnecessary surgery.

• Imaging plays a major role in diagnosis and management.
References


