The challenging diagnosis of cholangiocarcinoma

Poster No.: C-0108
Congress: ECR 2012
Type: Scientific Exhibit
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Keywords: Abdomen, Liver, Biliary Tract / Gallbladder, CT, MR, Ultrasound, Contrast agent-intravenous, Neoplasia
DOI: 10.1594/ecr2012/C-0108

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Purpose

To show the different imaging features of cholangiocarcinoma found in US, CT and MR, according to the different growth pattern and morphologic classification.

Methods and Materials

At our institution, precontrast and dynamic CT including arterial phase, hepatic venous phase and delayed phase scanning is usually performed.

Precontrast CT is useful for the detection and differentiation of an intraductal stone. Arterial phase is useful for understanding arterial anatomy and planning the surgical method. Venous and delayed phases show the best characterization of the lesion. Cholangiocarcinoma has a higher enhancement than does the adjacent liver parenchyma or normal bile duct, at venous and delayed phase due to the abundant fibrous stroma.

According to the morphologic and growth pattern classification, cholangiocarcinoma is classified into mass-forming, periductal infiltrating and intraductal growth types.

Results

Cholangiocarcinomas are malignant tumors arising from the biliary tract, originating from the epithelial cell lining the intra and extrahepatic ducts. Most cholangiocarcinomas are well, moderately, or poorly differentiated adenocarcinomas with abundant fibrous stroma.

Intrahepatic cholangiocarcinoma is the second most common primary malignancy of the liver, after hepatocellular carcinoma. The prevalence of cholangiocarcinoma varies markedly from one geographic region to another, with the highest prevalence in Southeast Asia.

There are a number of recognized risk factors for cholangiocarcinoma that all share the common feature of chronic biliary inflammation. Among these risk factors, infection with liver flukes (eg, Clonorchis sinensis and Opisthorchis viverrini) and hepatolithiasis are common causes in endemic areas. In Western countries primary sclerosing cholangitis and liver cirrhosis are the most common risk factors. There are other main risk factors such as hepatic viral infections (chronic hepatitis B and C infection, Epstein-Bar virus,
HIV), various bile duct diseases (Caroli disease) and environmental or occupational toxins (thorotrast, dioxin…).

The typical CT features of a **mass-forming cholangiocarcinoma** include homogeneous attenuation, irregular peripheral enhancement with gradual centripetal enhancement (Fig 1, 2), capsular retraction, the presence of satellite nodules and dilatation of the biliary tree in the tumor periphery (Fig 3, 4). Vascular encasement by the tumor is also common, but visible intravascular tumor thrombosis is rare (Fig 5).

At histological analysis, the viable tumor cells are usually located at the periphery of the tumor. By contrast, the central portion of the tumor is composed of variable degree of fibrosis. The degree of enhancement of a tumor in the delayed phase image is directly related to the amount of interstitial space in the fibrous stroma.

At ultrasonography, mass-forming cholangiocarcinoma manifests as a homogeneous mass with an irregular but well defined margin, with hypoechoic peripheral rim. However, tumors greater than 3 cm in size are usually hyperechoic (Fig 6).

The MR features are very similar to those found in CT. The mass shows high signal intensity at T2-weighted imaging with low signal intensity at T1-weighted imaging. Both the peripheral and centripetal enhancement may be more prominent at MR imaging than in CT (Fig 7).

**Periductal infiltrating cholangiocarcinoma** is characterized by growth along dilated or narrowed bile duct without mass formation and manifests as a diffuse periductal thickening and increased enhancement due to tumor infiltration. This type is rare in intrahepatic cholangiocarcinoma, but most hilar cholangiocarcinoma are of this type. At US, infiltrating type appears as a small, masslike lesion or diffuse bile duct thickening with or without obliteration of the bile duct lumen depending on tumor extent (Fig 8, 9).

**Intraductal type** is an intriguing type of cholangiocarcinoma because it has a variety of imaging features. Imaging patterns include diffuse and marked ductectasia with a gross visible papillary mass, diffuse and marked ductectasia without a visible mass, an intraductal polypoid mass within localized ductal dilatation, intraductal castlike lesions within a mildly dilated duct, and a focal stricture-like lesion with mild proximal ductal dilatation.

Several tumors or tumor-like conditions should be included in the differential diagnosis for a mass-forming type, like hepatocellular carcinoma with cirrhotic-stroma, sclerosing HCC, and combined HCC-cholangiocarcinoma, especially in patients with chronic liver
disease. In addition, metastasis from other sites and melanoma may manifest with similar findings. Not only are primary sclerosing cholangitis and recurrent pyogenic cholangitis risk factors, but they can also mimic a cholangiocarcinoma especially in the periductal and intraductal types.

Images for this section:

![Fig. 1](image1)

**Fig. 1:** Mass-forming cholangiocarcinoma. CT features in arterial phase CT (A), venous phase CT (B), and ten-minute delayed phase (C). CT shows gradual centripetal enhancement of the tumor and significant capsular retraction.

![Fig. 2](image2)

**Fig. 2:** Mass-forming cholangiocarcinoma. CT shows a rim enhancement at the periphery in the arterial phase (A), that becomes gradual, with important delayed enhancement (C). There is important parenchymal atrophy and distal bile duct dilatation. It’s encasing the left portal branch.
**Fig. 3:** Typical imaging findings in cholangiocarcinoma. A: satellite nodule posterior to the tumor in the right hepatic lobe. B: important capsular retraction. C: distal bile duct dilatation.

**Fig. 4:** Mass-forming cholangiocarcinoma located in the hilar region infiltrating the gallbladder infundibulum, with secondary important intrahepatic bile duct dilatation.
Fig. 5: Mass-forming type cholangiocarcinoma in the left hepatic lobe, with vascular encasement without thrombosis.
Fig. 6: Ultrasonography image of case in figure 4 shows a mass bulging the hepatic contour.

Fig. 7: CT shows a hypodense mass next to the gallbladder with a high signal intensity at T2-WI.
Fig. 8: CT shows an irregular and poorly defined lesion in the confluence of the hepatic bile ducts, with secondary intrahepatic bile duct dilatation.
Fig. 9: Cholangiogram depicts a narrowed segment in the distal right and left bile duct and common bile duct with secondary intrahepatic bile dilatation.
Conclusion

Diagnosis of cholangiocarcinoma is challenging according to its different appearances. A wide spectrum of neoplastic and non-neoplastic conditions of the biliary tract may masquerade as cholangiocarcinoma, adding more complexity. In most cases, a definitive diagnosis can be established only with histopathologic examination of a biopsy specimen.

References


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