Cholangiocarcinoma: the ugly one

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Learning objectives

The objectives in this review are:

- To Outline the risk, the epidemiology factors, and the morphological classification of cholangiocarcinoma.

- To describe of the radiological presentation of the colangiocarcinoma, presenting different cases collected in our hospital.

- To show the radiological manifestations of cholangiocarcinoma according to their anatomical location.
Background

Cholangiocarcinoma is the second most common primary liver tumor after hepatocarcinoma. It is an adenocarcinoma that arises from the bile duct epithelium. Cholangiocarcinoma may be well, moderately, or poorly differentiated, and it contain abundant fibrous stroma.

According to the reports, it represents the 10-20% of liver cancers and less than 3% of gastrointestinal tumors.

The prevalence of cholangiocarcinoma varies markedly from one geographic region to another, with the highest prevalence in Southeast Asia.

The typical age of presentation is between 50 and 70 years, the risk factors have the chronic inflammation of the bile duct in common, some of which are:

- Hepatolithiasis
- Parasitic infections of the biliary tract (trematodes), endemic in Asia
- Primary sclerosing cholangitis (10% of patients, 1.5% per year)
- Congenital abnormalities of the biliary tree (Caroli disease, cysts of bile duct, congenital hepatic fibrosis)
- Exposure to toxic (Thorotrast, alcohol)
- Viral infection (HCV, HBV, HIV, EBV)
- Familial polyposis
- Diabetes Mellitus

Most patients have an advanced disease stage at the time of their diagnosis, and the most common clinical presentation is constitutional symptoms and jaundice.
Cholangiocarcinomas display a wide spectrum of imaging findings. Most cholangiocarcinomas (with the exception of peripheral, intrahepatic type) typically cause biliary ductal obstruction. The initial imaging technique for a patient with obstructive jaundice is ultrasound, however in most cases the ultrasound detects dilatation of bile duct without observing the cause.

The CT has high reliability to determine the level of obstruction, and is useful in evaluating metastatic disease in the same study. At our institution, precontrast and triphasic CT, including late arterial phase, hepatic venous phase, and equilibrium phase scanning, is usually performed in a patient who has not undergone prior CT.

MR cholangiopancreatogram has a high diagnostic accuracy in determining the level and cause of obstruction. These two modalities (CT and MRI) are equally effective in the detection and correct diagnosis of the tumor.

Cholangiography, has a higher spatial resolution to show the bile ducts, although it is an invasive technique.

From an anatomical point of view we can classify the different types of cholangiocarcinoma as intrahepatic, hilar extrahepatic. Fig. 1 on page 7

**INTRAHEPATIC**

They can be classified according to their growth pattern in three types: 1) mass-forming, 2) periductal, 3) intraductal. Fig. 2 on page 7

1. Mass-forming type: it is morphologically presented as an homogeneous mass with an irregular but well-defined margin and it is frequently associated with the dilatation of the biliary tree in the periphery of the tumor. Fig. 3 on page 8 Vascular encasement by the tumor is also common, but grossly visible intravascular tumoral thrombosis is rare. Fig. 4 on page 9

At ultrasonography, mass-forming cholangiocarcinoma manifests as an homogeneous hypoechoic mass with an irregular but well-defined margin. Fig. 5 on page 9
greater than 3 cm in size are usually hyperechoic, but tumors less than 3 cm are hypo-
or isoechoic.

The typical CT features of a mass-forming cholangiocarcinoma include homogeneous hipodense attenuation lesion, irregular peripheral enhancement in the early phase (meaning actively growing areas), with gradual centripetal enhancement in the equilibrium phase (the degree of the enhancement in the late phase is related to the volume of fibrous stroma, which is usually located in the center of the tumor). Fig. 6 on page 10

The MR imaging features of the mass-forming cholangiocarcinoma are similar to its CT features. The mass shows an irregular margin with high signal intensity at T2-weighted images and with low signal intensity at T1-weighted images. Fig. 7 on page 10 Both the peripheral and the centripetal enhancement may be more prominent at MR imaging than on CT scans.

2. Periductal Infiltrating Type: it is characterized by its growth along a dilated or narrowed bile duct without mass formation.

In the different imaging techniques it shows a diffuse periductal thickening, with an irregular narrowing that often causes proximal biliary dilatation. It presents a greater contrast enhancement when compared to liver parenchyma Fig. 8 on page 11.

3. Intraductal Type: five patterns of growth have been described.

a. diffuse and marked ductectasia with a grossly visible papillary mass.

b. diffuse and marked ductectasia without a visible mass.

c. intraductal polypoid mass within localized ductal dilatation. Fig. 9 on page 11

d. intraductal castlike lesions within a mildly dilated duct.

e. focal stricture-like lesion with mild proximal ductal dilatation.

HILAR

Hilar cholangiocarcinomas account for more than 50% of all large bile duct malignancies. They are usually small lesions near the bifurcation of the hepatic duct, causing intrahepatic biliary dilatation with normal caliber of the extrahepatic biliary ducts. Fig. 10 on page 12 Presents delayed contrast enhancement.
Depending on their growth pattern differentiate various subtypes can be differentiated: 1) infiltrating, 2) exophytic and 3) polypoid.

1. Infiltrating: it is the most common, as we described previously it shows a thickening of the ductal wall obliterating that obliterates the lumen, what causes proximal biliary dilatation.

2. Exophytic: it manifests as a low-attenuation mass with peripheral enhancement. It could be difficult to distinguish from intrahepatic cholangiocarcinoma.

3. Polypoid: it represents an intraductal soft tissue density mass, showed as hypodense/hypointense when compared to liver parenchyma.

We can also classify them according to their location and the involvement of the hepatic ducts, known as Bismuth-Corlette classification. Fig. 12 on page 13

EXTRAHEPATIC

These are those cholangiocarcinomas that affect the extrahepatic bile duct. According to their growth pattern they can be infiltrative or polypoid.

1. Infiltrative: hyperenhanced mural thickening causing focal stenosis or duct caliber change in the extrahepatic duct with proximal ductal dilatation. Fig. 13 on page 14 Fig. 14 on page 14

2. Polypoid: low density/intensity intraluminal mass that provoke proximal ductal dilatation. Fig. 15 on page 15
Fig. 1: Anatomical classification of cholangiocarcinoma: intrahepatic (yellow), hilar (blue), extrahepatic (orange).

**Fig. 2:** Classification of cholangiocarcinoma by its growth pattern.


**Fig. 3:** Polilobulated hypodense mass in the study without contrast(a), which progressively enhances in the venous phase contrast(b). The late phase(c) shows retention of contrast in the areas of fibrous stroma.

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Fig. 4: Abdominal CT with contrast in portal phase: Coronal reconstruction shows a hypodense lesion compatible with cholangiocarcinoma, which encases the left portal branch.

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**Fig. 5:** Poorly defined hypoechoic lesion located at LHI, which causes dilation of distal intrahepatic biliary radicals.

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**Fig. 6:** Hypodense lesion located at LHI with fingerlike extensions that have peripheral enhancement in the arterial phase and define hypodense central region in the venous phase(b) that enhances in the late phase(c).

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Fig. 7: MRI of the same patient as in figure 4: lesion in LHI with dilatation of peripheral bile ducts; the lesion appears as hypointense on T1-weighted(a), with peripheral enhancement in the arterial phase(b) and central enhancement in late phase(c).

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Fig. 8: Periductal infiltrating cholangiocarcinoma.(a) Axial T2-weighted MR image shows a dilated peripheral intrahepatic duct with a slightly hyperintense lesion surrounding it (arrow).(b) Contrast-enhanced equilibrium phase MR image shows periductal enhancement around the dilated intrahepatic duct (arrowheads).

**Fig. 9:** Abdominal enhanced CT: axial images of arterial (a) and venous (b) phase and a coronal reconstruction (c); we can see dilatation of intrahepatic bile canaliculi (yellow arrows) and a soft tissue mass, which enhances inside the proximal common bile duct.

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**Fig. 10:** Contrast MRI, MR cholangiopancreatogram and Cholangiography. There is a severe intrahepatic biliary dilation due to a filiform stenosis with tumoral involvement of both liver ducts and the common hepatic duct (Klatskin tumor).

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Fig. 11: Infiltrating hilar cholangiocarcinoma: axial MR images in T2 sequence(a) contrast-enhanced T1-weighted MR(b). We can see a marked dilatation of intrahepatic bile ducts. No proximal bile duct is identified, the suprapancreatic and intrapancreatic portions show normal caliber. Expanding lesions or pathological hepatic hilum enhancements are not seen. MR cholangiopancreatogram images(c) and Cholangiography(d) confirmed the intrahepatic bile duct dilatation with stenosis at the hilar confluence of several ducts.

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**Fig. 12:** Type I: tumor distal to the bifurcation of the common hepatic duct. Type II: tumor affecting the bifurcation but sparing the principal left and right hepatic ducts. Type III: tumor affecting the principal right or left hepatic ducts. Type IV: tumor that affects the secondary or tertiary biliary ducts or multicenter tumor.


**Fig. 13:** T2-weighted axial (a, b) and coronal(c) MR images show a marked dilatation of the intrahepatic and extrahepatic bile ducts. The distal common bile duct shows an abrupt stop, without mass formation.

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**Fig. 14:** Axial postcontrast MR images (a,b) MR cholangiopancreatogram (c). These images show the same findings as described in the previous figure. We see a subtle thickening of the walls of the bile duct in the distal portion, with postcontrast enhancement in the late sequences. It suggests a distal bile duct cholangiocarcinoma with infiltrative, periductal growth.

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**Fig. 15:** Axial enhanced CT images in the venous phase (a,b) and a coronal reconstruction (c). We observe dilated bile ducts (black arrow), and distal common bile duct occupation by a soft tissue mass (yellow arrows) with enhancement after the introduction of contrast (higher in late phase).

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Conclusion

Early diagnosis of cholangiocarcinoma is a challenge due to their different morphological appearance and image. The morphological classification is useful in the radiographic interpretation.

Ultrasound plays a role of diagnostic suspicion, usually is the first imaging technique done. The CT and MRI are the imaging modalities of choice for characterization with a high degree of reliability for the diagnosis of cholangiocarcinoma.
References