Congenital Cystic Lesions of Bile Ducts: imaging-based differential diagnosis

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Learning objectives

Congenital cystic lesions of the biliary tract (CCLBT) are rare congenital biliary pathologies due to an abnormality of the development of ductal plaque during embryogenesis or they may be secondary to abnormalities of the pancreaticobiliary junction. Depending on the time of the stoppage of this development, various conditions may occur, gradually affecting the smaller or larger bile ducts.

The objectives of this essay are:

• to illustrate congenital cystic lesions of the biliary tract on the basis of knowledge of embryologic development;
• to illustrate the imaging findings of congenital cystic lesions of the intrahepatic biliary tract and choledochal cysts and provide differential diagnosis on imaging.
Background

**Congenital cystic lesions of the biliary tract** (CCLBT) are rare congenital biliary pathologies due to an abnormality of the development of ductal plaque during embryogenesis (Congenital Cystic Lesions of the Intrahepatic Bile Ducts, CCLIBD) or they may be secondary to abnormalities of the pancreaticobiliary junction (Congenital Cystic Lesions of the Extrahepatic Bile Ducts, CCLEBD) [1].

As regards the CCLIBD, the caliber of malformed ducts determines the type of ductal plate malformation. If small interlobular bile ducts are affected, **Von Meyenburg complex** or **congenital hepatic fibrosis** results. If medium-sized intrahepatic ducts are affected, **autosomal dominant polycystic liver disease** results. **Caroli disease** is the result of ductal plate malformation of large intra- hepatic bile ducts [2].

Although the cause is unclear, it is widely accepted that the prototype of CCLED, **choledochal cysts**, arises from an anomalous pancreaticobiliary ductal junction, particularly a long common channel, which is seen in 50-80% of patients [3, 4, 5].

Clinically, most of these conditions, such as biliary hamartomas, may be asymptomatic and they may be found incidentally on abdominal imaging performed for other indications. On the other hand, in a small group of patients, CCLBT may present in childhood or young adulthood with pain in the right upper quadrant and recurrent attacks of cholangitis with fever and jaundice (in the Caroli disease) or pancreatitis (in choledochal cysts), which are the most frequent complications [6]. Another known complication is malignancy [6, 7]. The overall risk of cancer in choledochal cysts is 10-15%, commonly affecting the extrahepatic bile ducts and gallbladder [7].

The different types of fibropolycystic liver disease demonstrate characteristic findings at computed tomography (CT) and magnetic resonance (MR) imaging. Patients with congenital hepatic fibrosis typically have imaging evidence of liver morphologic abnormalities, varices, splenomegaly, renal lesions, and other associated ductal plate abnormalities. Biliary hamartomas usually manifest as multiple cysts that are nearly uniform in size and measure up to 15 mm in diameter. Autosomal dominant polycystic disease typically manifests as an enlarged and diffusely cystic liver. In Caroli disease, cystic or fusiform dilatation of the intrahepatic ducts is seen, as well as the "central dot sign," which corresponds to a portal vein branch protruding into the lumen of a dilated bile duct. Choledochal cyst manifests as a fusiform or cystic dilatation of the extrahepatic bile duct [2].

MRI modalities for the analysis of the biliary tree are mainly represented by T2-weighted sequence, also known as MR cholangiography (MRCP), and T1-weighted gadolinium-enhanced sequences [8].
In this educational exhibit, we describe the most common imaging findings of these conditions and provide differential diagnosis on imaging.
Findings and procedure details

In Von Meyenburg complex (biliary hamartomas), lesions are usually scattered, multiple and can be solid, cystic, or mixed; "the starry sky" appearance is pathognomonic in Magnetic Resonance Cholangiopancreatography (Fig. 1, Fig. 2). Hepatobiliary contrast agents will demonstrate no communication with the biliary tree. At Ultrasound, small hamartomas are usually echogenic. Often tiny individual hamartomas cannot be resolved and are instead interpreted as diffuse heterogenous liver echotexture. Larger hamartomas (>10 mm) may appear hypoechoic or anechoic and "comet tail artefact" may be seen [9]. Differential diagnosis from multiple liver metastases may be not simple by using Ultrasound, so it could be necessary to use another technique (MR, CT or even CEUS) to provide differential diagnosis of these conditions (metastases present more variable size and prominent enhancement), as well as with the clinical history of the patient.

Congenital hepatic fibrosis is usually associated with autosomal recessive polycystic kidney disease [10]. It is characterized by enlargement of portal spaces due to the presence of fibrosis and numerous, more or less ectatic, abnormal bile ducts communicating with the biliary tree [11]. The main feature of the disease is portal hypertension without liver insufficiency, but progression of fibrosis can evolve to true cirrhosis [12]. Morphologic changes in the liver with hypertrophy of the lateral segments of the left lobe and normal volume or hypertrophy of segments IVa and IVb (Fig. 3), associated ductal plate malformations, signs of portal hypertension, such as varices, splenomegaly, and renal abnormalities, are frequently found in combination in patients with congenital hepatic fibrosis [12].

Caroli disease is a rare autosomal recessive disease characterized by segmental or diffuse communicating, non-obstructive, saccular or fusiform dilatation of large intrahepatic bile ducts (up to 5 cm). The "central dot sign" is pathognomonic (Fig. 5) and represents cystic enveloping of portal tract vessels [2]. Liver specific hepatobiliary MRI contrast agents can be useful to prove communication with the biliary tree and provide differential diagnosis from hamartomas.

We refer to "Caroli syndrome" when Caroli disease and congenital hepatic fibrosis coexist: this condition is even more common than Caroli disease alone [13]. Perhaps, the two conditions may represent different stages of the same disease [13].

Choledochal cysts are predisposing factors for cholangitis, pancreatitis, and malignancy. The use of hepatobiliary contrast agents will show a communication with the biliary tree. They are classified according to the Todani modification of the Alonso-Lej classification (Fig. 8).
Although the cause is unclear, it is widely accepted that choledochal cysts arise from an anomalous pancreaticobiliary ductal junction, particularly a long common channel, which is seen in 50-80% of patients [3, 4, 5]. Anomalous pancreaticobiliary ductal junction may allow mixture of pancreatic and biliary juices, which activates pancreatic enzymes leading to consequent inflammation and weakening of the duct wall [3, 5]. In 22% of cases, anomalous pancreaticobiliary ductal junction is seen without dilatation, but patients are still at high risk of carcinogenesis, particularly of gallbladder cancer [14].
Fig. 1: Axial MR image (T2-W fat sat) of a 52-year-old woman with biliary hamartomas shows multiple millimetric cystic lesions in almost all liver segments, frankly hyperintense on T2-weighted sequences and hypointense on T1-W sequences.

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Fig. 2: Magnetic Resonance Cholangiopancreatography of the same patient of Fig. 1 shows the "starry sky" appearance, pathognomonic for the Von Meyenburg complex.

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Fig. 3: Axial CT scan on portal venous phase of a 68-year-old man, with no medical history of hepatitis or cirrhosis, shows hepatomegaly with marked hypertrophy of segment IV, left lobe and caudate lobe. Features are consistent with congenital hepatic fibrosis.

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**Fig. 4:** Coronal MR image T2-W sequence of a 47-year-old woman with Caroli disease shows in S7, below the liver dome, dilatation of the intraepathic biliary ducts, up to the third order branches, which appear hyperintense (arrow).

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**Fig. 5:** Axial T1-W image on portal venous phase of the same patient of Fig. 4 shows the "central dot sign" (arrow), due to the enhancement of the central portal radicles within a dilated intrahepatic biliary duct: this sign is pathognomonic for Caroli disease.

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Fig. 6: Coronal reformatted CT image on portal venous phase of a patient with choledocal cyst shows a dilated cystic lesion (maximum diameter of 1.8 cm) of the proximal common bile duct (arrow).

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Fig. 7: Magnetic Resonance Cholangiopancreatography of the same patient of Fig. 6 shows fusiform dilatation of the common hepatic and common bile ducts up to the junction with main pancreatic duct. Features are consistent with choledocal cyst type Ic according to Todani classification.
Fig. 8: Todani modification of Alonso-Lej classification and relative percentage of occurrence of each type of choledochal cyst. Type IA is marked cystic dilatation of entire extrahepatic bile duct; IB, focal segmental dilatation of extrahepatic bile duct, usually distal to cystic duct insertion; IC, smooth fusiform dilatation of entire extrahepatic bile duct. Type II: discrete diverticulum of extrahepatic bile duct. Type III: dilatation of distal common bile duct confined to wall of duodenum, often bulging into duodenal lumen (choledochocele). Type IVA: multiple sites of dilatation of both extrahepatic and intrahepatic biliary tree. Type IVB: multiple sites of dilatation of extrahepatic bile duct only
(string-of-beads appearance). Type V: multiple sites of saccular or cystic dilatation of only intrahepatic biliary tree (Carolii disease or communicating cavernous ectasia) [15].

Conclusion

CCLBTs are rare congenital biliary pathologies. The clinical significance of these conditions is varied, ranging from none in biliary hamartomas to high risk of cholangiocarcinoma in Caroli disease and predisposition for cholangitis, pancreatitis and malignancy in choledochal cysts.

CCLBTs can be recognized with characteristic radiological features, using almost all the diagnostic techniques. Familiarity with imaging findings is crucial for their appropriate detection and for differential diagnosis with other similar hepatic cystic lesions (from simple hepatic cysts to metastatic lesions).
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