Peribiliary cysts - a morphologic mimicker

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Special Focus: Cysts Case Type: Clinical Cases
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Patient: 74 years, male

Clinical History:

A 74-year-old man was referred to the hospital for further evaluation of a suspected biliary duct dilatation. The patient had a significant history of chronic alcohol consumption (>40g/ day) for about 20 years. There were no significant findings on physical examination and laboratory studies revealed no signs of cholestasis.

Imaging Findings:

Ultrasonography showed an irregular hepatic contour, an enlarged caudate and left lobe, relative atrophy of the right lobe and widening of the hepatic fissures. Additionally, clustered cysts around the hepatic hilum and large portal tracts in the left and right liver lobes were identified. There were no cysts in the periphery of the liver. It is possible to identify thin septa between the clustered cysts. (Fig.2)

On CT (Fig.1) multiple round or tubular hypodense lesions were seen around the large portal tracts, mimicking bile duct dilatation (Fig.1)

Axial and coronal reformatted MR (Fig.3) examination showed multiple cysts distributed along the hepatic hilum and large portal tracts. Cysts are identified on both sites of the portal vein. Magnetic resonance cholangiopancreatography (CPRM) showed no communication between the cysts and the bile duct lumen (Fig.3). These features were suggestive of peribiliary cysts associated with severe liver disease.

Discussion:

Peribiliary cysts are benign and usually an incidental finding with imaging studies in the context of advanced liver disease. Its pathogenesis is related to obstruction and cystic dilatation of the extramural glands in the periductal connective tissue [1, 2, 3, 4]. Inflammatory changes and disturbances in periportal circulation were proposed to explain the gland obstruction [1, 2, 3, 4]. Genetic factors, such as autosomal polycystic kidney disease and other congenital cystic diseases, may also be associated with the pathogenesis of peribiliary cysts [1, 2, 4, 5].

Usually, it is possible to find a variable number of clustered cysts coursing exclusively along the hepatic hilum and larger portal tracts with variable size, the larger reaching up to 3cm in diameter [1, 5, 7].

Although peribiliary cysts are generally asymptomatic and incidentally found, in rare cases, may cause obstruction of the biliary tree [2].

A recent study by Nakanuma et al. reports that peribiliary cysts are more likely to occur in chronic alcoholic liver disease than in other causes of cirrhosis and that their occurrence was related to the degree of alcohol-related hepatic fibrosis and pancreatic fibrosis [5, 1].

The key elements for diagnosis of this entity are the presence of an underlying cirrhotic liver and the recognition of
cystic lesions on both sides of the portal veins. In addition, the presence of small fluid-filled cavities independent of the biliary tree on MRCP provides support to the diagnosis [4, 6].

The differential diagnosis to consider:

Biliary duct dilatation: the identification of cysts distributed on both sides of the portal vein is important to differentiate peribiliary cysts from ductal dilatation [4, 6, 8].

Primary sclerosing cholangitis (PSC): a linear cluster of cysts may mimic a “string of beads” appearance, corresponding to alternating strictures and dilation of biliary ducts associated with PSC [6, 8, 9].

Carolí’s disease: it is characterised by multifocal cystic or fusiform dilatation of intrahepatic bile ducts. Unlike peribiliary cysts, in Carolí’s disease cystic dilatations exhibit continuity with the bile ducts [4, 7].

Bile duct hamartomas (BDH): BDH are typically multiple round or irregular focal lesions of nearly uniform size(?15mm) scattered throughout the liver [4, 7].

Periportal oedema: it is seen as an ill-defined hypodensity on CT or as a high signal intensity on T2-weighted images with a perportal distribution. It is a nonspecific finding that can occur with acute hepatitis, hepatic veno-occlusive disease, after liver/bone marrow transplantation, rapid intravenous hydration, trauma and congestive heart failure [4, 6].

Peribiliary cysts are a potential source of misdiagnosis and the recognition of its predisposing factors and classic signs allows a diagnosis.

**Differential Diagnosis List:** Peribiliary cysts, Peribiliary cysts, Biliary ductal dilatation, Primary sclerosing cholangitis, Carolí’s disease, Bile duct hamartomas, Periportal oedema

**Final Diagnosis:** Peribiliary cysts

**References:**


Figure 1

Description: Origin:
Figure 2

Description: Subcostal US examination showed multiple round or tubular anechoic lesions around the large portal tracts mimicking bile duct dilatation. Origin: Hospital de Braga
Description: (a, b) MRI reveals multiple T1 hypointense, T2 hyperintense round structures coursing along the portal tracts, with a diameter ranging from 0.2 to 2 cm, consistent with peribiliary cysts. Origin: Hospital de Braga
Description: (a, b) MRI reveals multiple T1 hypointense, T2 hyperintense round structures coursing along the portal tracts, with a diameter ranging from 0.2 to 2 cm, consistent with peribiliary cysts. **Origin:** Hospital de Braga.

Description: Coronal maximum intensity projection (MIP) MRCP reformat shows a (c) shows no communication between the cyst and the bile duct lumen. **Origin:** Hospital de Braga.