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Central dot sign in Caroli disease

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Abstract

The central dot sign is a non-pathognomonic radiological sign, but suggestive of caroli disease, where the central dot corresponds to fibrovascular bundles surrounded by cystic dilatations of the intrahepatic bile ducts. We present the case of a 24-year-old girl with Caroli disease diagnosed by the central dot sign observed on MRI.

Keywords: Caroli disease; Central dot sign; Biliary dilatation; MRI.

Description

A 24-year-old girl with caroli disease presenting to emergency for biliary colic. Clinical examination revealed an icteric patient. Biological tests revealed cholestasis with elevated bilirubin (BT: 54 mg/L) and moderate hepatic cytolysis. Viral hepatitis serology was negative.

Abdominal ultrasound showed dilatation of the intrahepatic bile ducts without any obstacle. CP MRI demanded to identify the obstacle revealed a diffuse cystic dilatation of the intrahepatic bile ducts centred by a portal branch, giving the characteristic "central dot sign" (Figure 1) without dilatation of the main bile duct associated to lithiasis of the main bile duct.

Discussion

Caroli disease or obliterative cavernous ectasia is a congenital ductal plate malformation that may involve different levels of the biliary tree without abnormality of the hepatic parenchyma [1]. It may be accompanied by bacterial cholangitis or biliary lithiasis causing recurrent episodes of hepatic colic and pancreatitis.

Diagnosis is based on imaging, which shows a characteristic radiological sign - the central dot sign - best seen in the axial plane on CT or MRI. This central dot represents the cross-section of a branch of the portal vein surrounded by an abnormally dilated bile duct which enhances after injection [2], while the main bile duct is of normal calibre.

The central dot sign differentiates caroli disease from other biliary dilatations. However, it is also found in peri-biliary cysts, periportal lymphoedema and severe obstructive biliary dilatation [3], although it is absent in primary sclerosing cholangitis or recurrent pyogenic cholangitis.

Treatment can be conservative, with antibiotic therapy and biliary drainage, or surgical, based on partial hepatectomy in the focal form or liver transplantation in the diffuse form. Prognosis depends on the severity of episodes of cholangitis, comorbidities and increased risk of biliary cancer.

Author's contributions: All the authors contributed to study concept, data analysis and writing the paper. All authors read and approved the final version of the manuscript.

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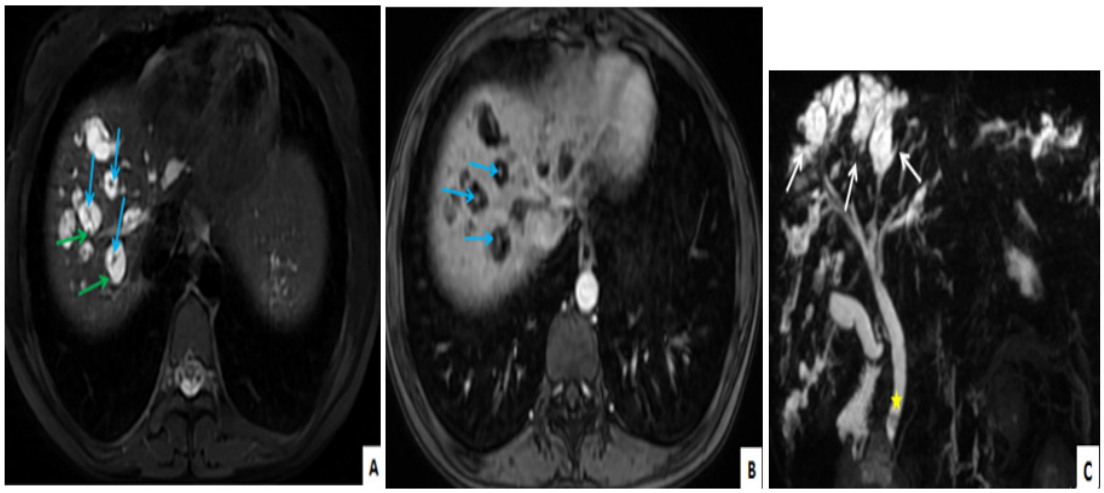

Figure 1: Liver MRI on T2 weighted images with fat saturation **(A)** showing large diffuse cystic ectasias of the intrahepatic bile ducts (green arrow) centred by branches of portal vein (blue arrow) enhanced on T1 sequence with portal injection **(B)** This is the central dot sign. MR cholangiography sequence **(C)** in the coronal plane showing biliary ectasia (white arrow) with gallstones of the main bile duct (star).