A Rare Congenital Disorder of Intrahepatic Bile Ducts

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A 34-year-old male presented with chief complaints of jaundice for last 4 weeks and abdominal distension and swelling of the legs for 3 weeks. There was no associated abdominal pain, pale stools, or itching. He is a known diabetic on insulin therapy, and occasionally consumes alcohol. There was no family history of any liver-related diseases. His initial workup showed hemoglobin of 8.8 g%, total counts of 9800 cells/cu mm, and platelets counts of 58000 cells/cu mm. Liver function tests showed total bilirubin of 13.5, direct bilirubin of 10.4,

alanine transaminase of 1560 IU/L (normal 8 < 40), aspartate transaminase of 633 IU/L (normal <50), and alkaline phosphatase of 1385 IU/L (normal 25–140). Viral markers (HBsAg, anti HCV) were negative. An ultrasound abdomen showed hepatosplenomegaly with dilated common bile duct, common hepatic duct, and intrahepatic biliary radicals, with presence of intrahepatic biliary calculi. MRI images are shown in Figures 1–3.

What is the diagnosis?

Figure 1 T2 hASTE axial showing gross multifocal cystic dilatation of intrahepatic biliary ducts.

Figure 2 T2 hASTE coronal view.

Keywords: intrahepatic biliary dilatation, hepatomegaly, aminotransferases, cholestasis, choledocal cyst

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Abbreviations: AST: aspartate transaminases; ALT: alanine transaminases; USG: ultrasound; IHBR: intrahepatic biliary dilatation; CBD: common bile duct; CHD: common hepatic duct; MRI: magnetic resonance imaging; PCKD: polycystic kidney disease; CT: computed tomography

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Figure 3 Post-contrast T1 axial scan.

Images in Hepatology—Answer

T2 HASTE axial and coronal images show gross multifocal cystic dilatation of bilobar peripheral as well as central intrahepatic bile ducts with associated splenomegaly. The post-contrast T1 axial images show enhancing portal radicles within dilated intrahepatic bile ducts, giving rise to the “Central Dot Sign” suggestive of Caroli disease (Figure 3A).

Figure 3A Post-contrast T1 axial ‘Central Dot Sign’-enhancing portal radicles within dilated intrahepatic biliary radicles.

Caroli disease is a rare, autosomal recessive congenital disorder comprising of multifocal segmental cystic non-obstructive dilatation of intrahepatic bile ducts secondary to in-utero malformation of the ductal plate. It is also classified as a type V choledochal cyst, according to the Todani classification. Its simple or pure form only involves intrahepatic bile ducts, whereas the complex form is associated with congenital hepatic fibrosis and polycystic kidney disease (Caroli syndrome).¹ The “central dot” sign^{2,3} corresponds to enhancing dots within the dilated intrahepatic bile ducts, representing portal radicles in contrast-enhanced CT, and is highly suggestive of Caroli disease. If the disease is localized, segmentectomy or lobectomy may be considered. However, for diffuse disease liver transplantation may be required.

CONFLICTS OF INTEREST

The authors have none to declare.

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