Caroli's disease

Pratibha Issar • Sanjeev Kumar Issar

Published online: 11 September 2013
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Caroli's disease, also known as congenital communicating cavernous ectasia of the biliary tree, is a rare autosomal recessive developmental abnormality characterized by saccular dilatation of the intrahepatic bile ducts. The magnetic resonance (MR) images shown here are from a 21-year-old man who presented with jaundice, abdominal pain, and fever for 5 days. On clinical examination, he had tenderness in the right hypochondrium. Laboratory parameters showed elevated direct serum bilirubin, alkaline phosphatase, AST, ALT, and leukocyte count. Axial T2-weighted MR revealed hyperintense dilated cystic structures of varying sizes that communicate with the biliary tree. Hypointense multiple intraluminal filling defects due to intraductal lithiasis were seen (Fig. 1). MR cholangiography demonstrated the pathognomonic feature of saccular dilated and nonobstructed intrahepatic bile ducts that communicate with

Fig. 1 Saccular dilation of the biliary tree with intraductal lithiasis

Fig. 2 Magnetic resonance cholangiography shows a connection between the saccular dilated segmental bile duct and the centre biliary tree

the biliary tree (Fig. 2). The condition may be associated with cystic renal disease and hepatic fibrosis and the abnormality may be segmental or diffuse. Complications are recurrent bouts of cholangitis, intrahepatic calculi, abscess, and development of cholangiocarcinoma [1, 2].

References

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P. Issar (✉) • S. K. Issar
Department of Radiodiagnosis, J L N Hospital and Research Centre,
Bhilai, Chhattisgarh 490 006, India
e-mail: mareesh_23@yahoo.co.in