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IMAGE | LIVER

Congenital Extrahepatic Portosystemic Shunt in a Child With Caroli's Syndrome

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CASE REPORT

A 7-year-old boy presented with recurrent cholangitis, failure to thrive, and progressive splenomegaly. On examination, he had severe pallor, clubbing, and splenohepatomegaly. Lab results showed anemia (hemoglobin 7.2 g/dL) and normal liver functions. Abdominal ultrasound showed hepatosplenomegaly with coarsened liver echotexture and multifocal intrahepatic biliary radical (IHBR) dilatation. The main portal vein was markedly attenuated beyond the splenomesenteric confluence. Bilateral kidneys were enlarged with indistinct corticomedullary junction. Contrast-enhanced computed tomography demonstrated right liver lobe atrophy with compensatory left lobe hypertrophy. There were skip areas of bilobar saccular or tubular IHBR dilatation, with a central dot sign, typical of Caroli's disease (Figure 1). Coronal maximum intensity projections showed an atretic main portal vein and its intrahepatic branches with a prominent and tortuous portosystemic (lienorenal) shunt draining portal blood into systemic circulation (Figure 2), indicative of congenital extrahepatic portosystemic shunt (CEPS, Type 2). There was asymmetric enlargement of the kidney with renal tubular ectasia and multiple renal calculi. Upper gastrointestinal endoscopy showed large esophageal varices with red color signs. The patient required 5 sessions of variceal ligation for eradication. Liver

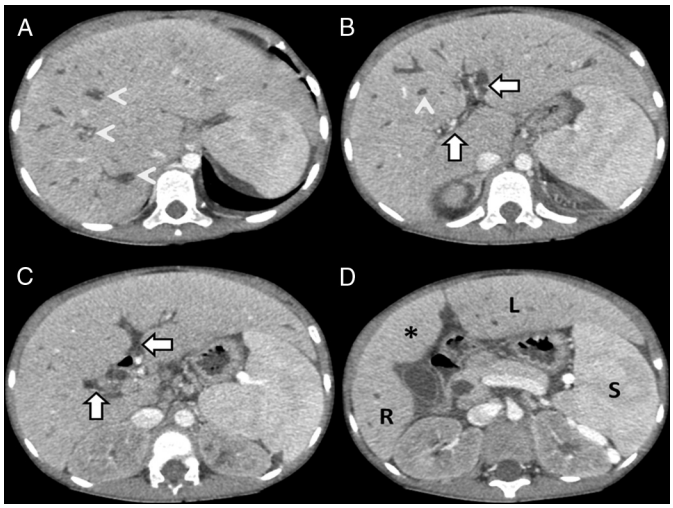

Figure 1. (A) Axial contrast-enhanced computed tomography depicting skip areas of bilobar biliary dilatation with central dot sign (arrows). (B and C) Note absent portal venous branches at the hepatic hilum (arrows). (D) The liver shows volume redistribution in the form of relative right lobe atrophy (R) and compensatory left lobe enlargement (L) with preserved segment IV volume (asterisk). Note the attendant splenic enlargement (S).

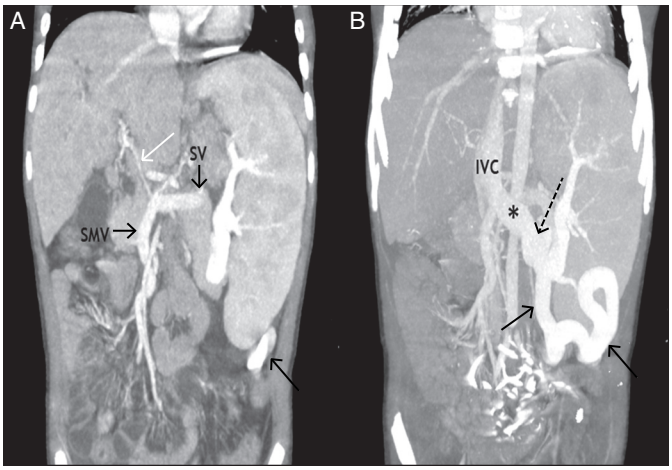

Figure 2. (A) Coronal maximum intensity projections depicting a thread-like atretic main portal vein (white arrow) beyond the confluence of superior mesenteric vein (SMV) and splenic vein (SV). (B) Attendant large tortuous portosystemic shunt vessel (black arrow) can be seen draining portomesenteric blood from the splenic vein into the systemic circulation (left renal vein [*]) via a large portosystemic shunt (dotted arrow).

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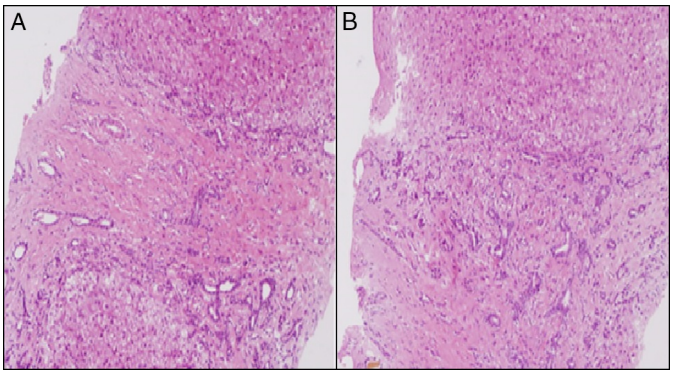

Figure 3. Liver biopsy showed nodules separated by broad fibrous septae with maintained acinar architecture. Note the absence of portal vein in portal tract and ductular proliferation.

biopsy showed nodules separated by large, broad, fibrous bands and maintained acinar architecture, suggestive of congenital hepatic fibrosis (Figure 3). Portal tracts showed absence of portal vein profiles, ductular proliferation, and conspicuous hepatic arteries, which confirmed the diagnosis of CEPS. On the basis of clinical presentation, imaging, and biopsy, a diagnosis of Caroli's syndrome (Caroli's disease + congenital hepatic fibrosis) with type 2 CEPS and renal tubular ectasia was made. The child was treated with intravenous antibiotics, and his cholangitis resolved. He was discharged on oral cyclical antibiotics (trimethoprim-sulphamethoxazole and cefixime, alternating every 4 weeks). At his 1-year follow-up, his varices had been eradicated with no recurrence. There has been no recurrence of cholangitis, and his anemia resolved, although growth centiles remain affected.

Association of CEPS with congenital hepatic fibrosis and Caroli's disease have been reported only rarely and have clinical, diagnostic, and therapeutic implications.[1-4] It has been postulated that portal venous blood supply plays a central role in the formation and remodeling of the ductal plate. Congenital anomalies in the development of the portal vein are likely to lead to ductal plate malformation, which can then lead to congenital hepatic fibrosis as in our case. This association also has important diagnostic and therapeutic implications. There is an increased risk of hepatic malignancies as decreased flow in the portal vein leads to a compensatory increase in hepatic artery flow, predisposing a patient to malignancy.[5] The portosystemic shunt needs to be differentiated from secondary shunts caused by portal hypertension related to congenital hepatic fibrosis. In our case, an atretic main portal vein on imaging and absent portal vein profiles in the portal tract on histology confirmed the congenital origin of the portosystemic shunt. Our patient presented with recurrent cholangitis with an incidental diagnosis of asymptomatic CEPS. Previously reported cases in the literature also had incidentally diagnosed CEPS in patients presenting with symptoms of cholangitis or growth failure.[2-4]

DISCLOSURES

Author contributions: B. Bihari Lal, A. Arora, and V. Sood wrote the manuscript. S. Alam, D. Rawat, and R. Khanna reviewed and revised the manuscript. A. Arora and B. Bihari Lal prepared the figures. B. Bihari Lal is the article guarantor.

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