Severe Portal Hypertension due to Congenital Hepatoportal Arteriovenous Fistula Associated with Intrahepatic Portal Vein Aneurysm

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ABSTRACT: A 13-year-old girl was referred for assessment of severe gastrointestinal tract bleeding. Her liver function tests were normal, and she had no evidence of chronic liver disease or history of significant trauma. Clinical and sonographic findings suggested the presence of a portal vein aneurysm associated with a hepatoportal arteriovenous fistula. Abdominal angiography confirmed the diagnosis. The arteriovenous fistula was congenital, and the associated portal vein aneurysm was either congenital or secondary to hemodynamic changes in the portal venous system. © 1998 John Wiley & Sons, Inc. *J Clin Ultrasound* **26**:357–360, 1998.

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Intrahepatic or extrahepatic arterioportal fistula associated with portal vein aneurysm is an uncommon cause of portal hypertension. The predominant clinical manifestations are complications of portal hypertension, most commonly bleeding from the gastrointestinal tract.[1–3] We report the gray-scale and color Doppler sonographic findings in a child with severe variceal bleeding due to portal hypertension caused by a large intrahepatic arteriovenous fistula and portal vein aneurysm.

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CASE REPORT

A 13-year-old girl was referred to the Pediatric Gastroenterology Clinic at Dr. Sami Ulus Children's Hospital because of severe gastrointestinal tract bleeding. Her medical history was unremarkable, with no significant abdominal trauma, jaundice, or infections. On physical examination, she was noted to be normally developed and well nourished. The liver was not palpable, and the spleen was palpable 4 cm below the left costal margin. No dilated veins were noted on the abdominal wall. A clear continuous murmur was heard over the right upper quadrant of the abdomen, in association with a thrill. The patient's hemoglobin was 5 g/dl, and her white blood cell count was 7,400/ml with a normal differential. All other laboratory findings, including prothrombin time and serum levels of total protein, albumin, globulin, aspartate transaminase, alanine transaminase, and total bilirubin, were within normal limits. Hepatitis B surface antigens were not detected. Endoscopy of the esophagus showed grade 4 varices.

Real-time sonography of the liver demonstrated an anechoic oval lesion that was in continuity with the left branch of the portal vein (LPV) (Figure 1) and was assumed to be an aneurysmal dilatation of the LPV. Color Doppler examination confirmed the diagnosis. The distal portion of the LPV showed focal enlargement, with a diameter of 20 mm, and turbulent flow (Figure 2). Reversed flow was detected in the LPV. The left branch of

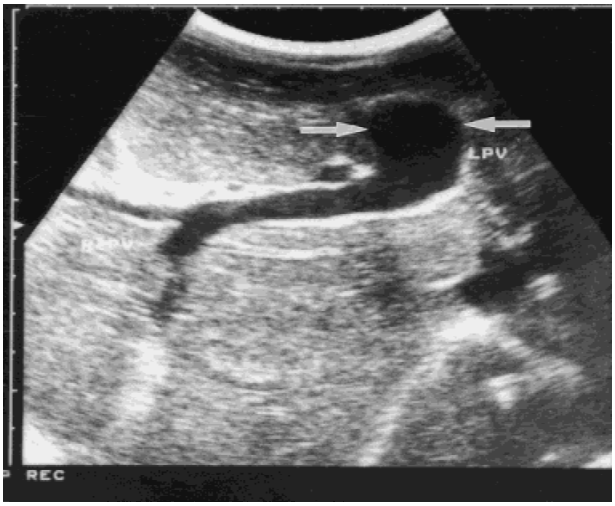

FIGURE 1. Gray-scale sonogram of the liver demonstrating an aneurysmal dilatation of the distal segment of the left branch of the portal vein (arrows).

the hepatic artery was also enlarged, with a diameter of 8 mm. In the hepatic tissues adjacent to the aneurysmal dilatation, color speckling (mosaic pattern), which is considered to be a typical sign for arteriovenous fistula, was present (Figure 3).

Selective arteriography of the hepatic artery was performed by digital subtraction angiography, and a large left hepatic artery–left portal vein fistula was visualized (Figure 4). A β-blocker (propranolol) was prescribed to decrease portal venous pressure. The hepatic artery was surgically ligated, and the postoperative period was uneventful.

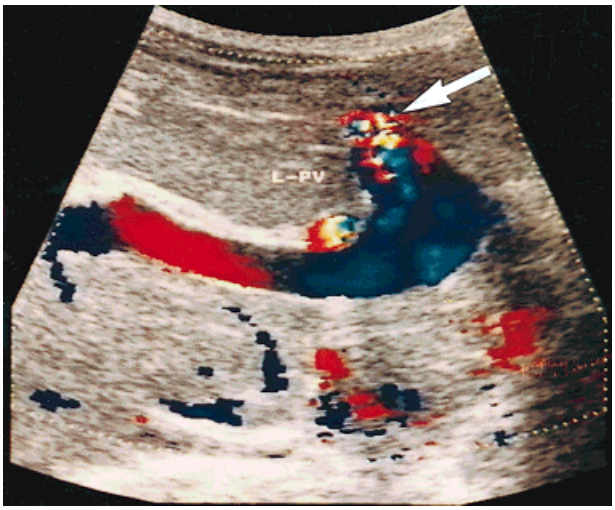

FIGURE 2. Color Doppler sonogram of the portal vein aneurysm. The flow in the left portal vein (L-PV) is reversed, and there is turbulence (arrow) at the distal portion of the aneurysmal sac.

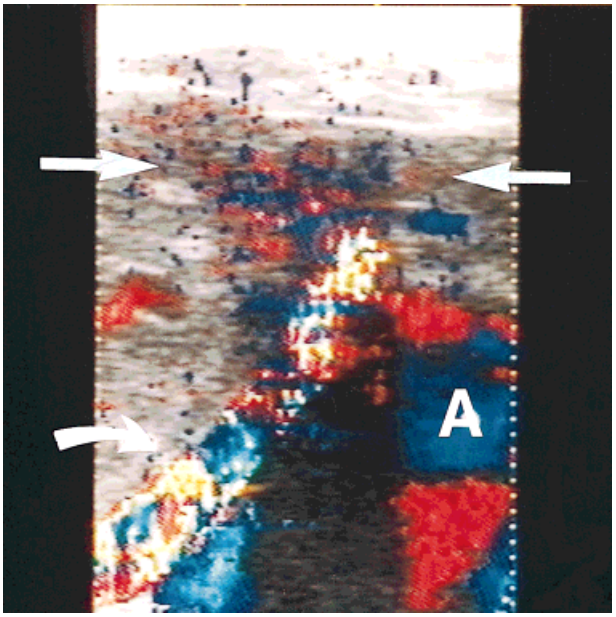

FIGURE 3. Color Doppler sonogram showing speckling of perivascular liver parenchyma (large arrows) adjacent to the arteriovenous communication. This characteristic finding is due to vibrations transmitted from the fistula to surrounding tissues. The hepatic artery (curved arrow) contains high-velocity turbulent flow. A, aneurysm.

DISCUSSION

Portal vein aneurysm is rare, with fewer than 30 cases reported[4,5] since Barzilai and Klekner's first description in 1956.[6] The etiology is believed to be congenital, secondary to portal hypertension, or associated with abnormal weakness of the vein wall.[4] Portal vein aneurysm often presents in conjunction with major gastrointestinal tract bleeding.[1] Variceal bleeding in our patient led us to suspect this etiology. Initially, the portal vein aneurysm was demonstrated by gray-scale sonography. The arteriovenous fistula was subsequently identified as the etiologic factor by color Doppler imaging.

Hepatoportal arteriovenous fistula is also uncommon and usually results from either a ruptured aneurysm of the hepatic artery or penetrating trauma.[7] Intrahepatic arteriovenous fistulas have been described secondary to needle puncture of the liver, blunt abdominal trauma, familial hereditary telangiectasia, and arteriovenous malformation.[7] A few cases of congenital fistulas from the hepatic artery to the portal vein have been reported,[1,2,7,8] but this abnormality is not a common cause of portal hypertension. Increased blood flow in the portal system is considered to be the cause of hyperkinetic portal hypertension in patients with hepatoportal arteriovenous fistulas. The predominant clinical manifestation of portal

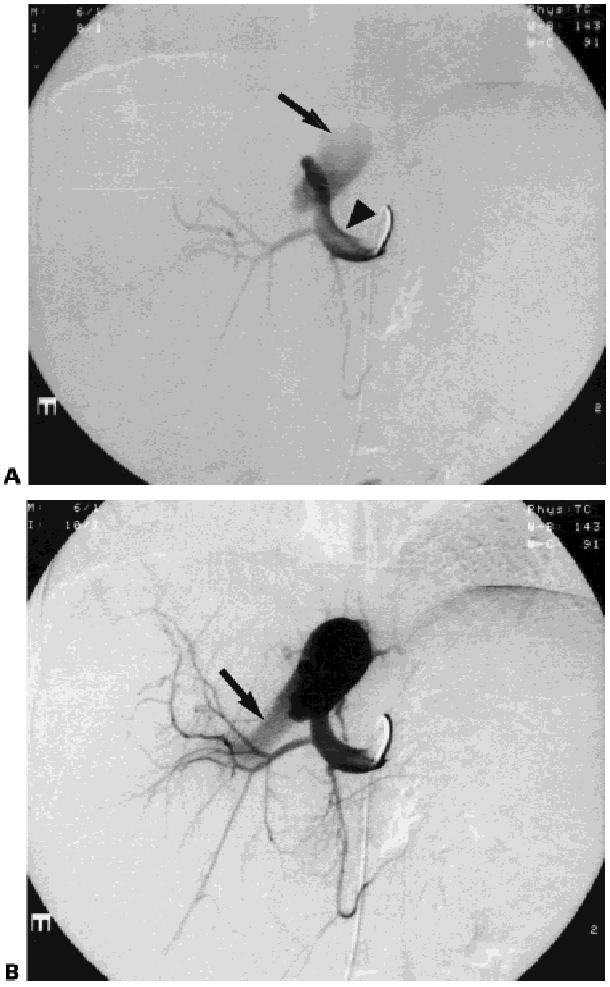

FIGURE 4. (A) Digital subtraction angiogram showing opacification of the aneurysmal segment of the portal vein (arrow) via the left hepatic artery (arrowhead). **(B)** Subsequent visualization of the more proximal segment of the left branch of the portal vein (arrow).

hypertension in our patient was variceal bleeding. It is of interest that the continuous thrill and murmur over the right upper quadrant of the abdomen were helpful in the diagnosis. In our case, the portal vein aneurysm associated with the arteriovenous fistula might have been congenital or secondary to hemodynamic changes in the portal venous system.

Both gray-scale sonography and color Doppler sonography are fast and noninvasive initial imaging modalities for establishing a diagnosis of portal vein aneurysm and arteriovenous fistula.[9,10] In our patient, the dilated portal vein branch, enlarged left hepatic artery, and arteriovenous fistula were noted by color Doppler sonography. The turbulence and disturbed flow in the arteriovenous fistula caused the adjacent soft tissues to vibrate. The vibrations induced by the fistula caused the color speckling of the perivascular liver parenchyma.[11,12] This must be differentiated from color artifacts resulting from inappropriately high color gain. Speckling is best seen in systole. Although clinical and sonographic findings suggested the presence of an arteriovenous shunt, hepatic angiography was the only method to confirm the suspected anatomic vascular malformation.

Many therapeutic options for hepatoportal arteriovenous fistula and aneurysm of the portal vein have been reported. In general, surgery is the procedure of choice for extrahepatic fistulas, whereas embolization is optimal for intrahepatic fistulas.[7] In recent years, transcatheter arterial embolization has been increasingly used to treat intrahepatic arteriovenous fistulas.[3,13–15] In our patient, propanolol was administered to decrease portal venous pressure and to control variceal bleeding, and then the hepatic artery was surgically ligated.

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