Congenital Arterioportal Fistula of the Liver: Report of a Case

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ABSTRACT

Congenital intrahepatic shunts are uncommon anomalies consisting of abnormal connections between the hepatic arteries, portal veins, hepatic veins or systemic venous system. It can be found incidentally or in symptomatic infants.

Intrahepatic arterioportal fistula is caused by an abnormal connection with hepatic artery and portal vein. It is a rare entity and an uncommon cause of portal hypertension in children.

We reported a 10 month-old child with Down syndrome who presented with growth failure, hepatosplenomegaly and ascites. Imaging findings and treatment of the patient are discussed.

Keywords: Portal hypertension, arterio-portal fistula, hepato-portal fistula, congenital hepatic shunts

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CASE REPORT

A 10 month-old child with Down syndrome, PDA, ASD and congenital hypothyroidism presented with growth failure and abdominal distention. He was admitted 3 months ago with RSV pneumonia. His mother noticed that the abdomen was gradually increased in size over the past 1 month. He was a term baby boy GA 38 weeks with birth weight of 2,600 gm.

Present body weight was 7.4 kg and height was 67 cm. Physical examination revealed anicteric sclera, normal heart sound, no murmur. Lungs were clear. Abdomen was distended with dilated superficial veins. Liver was palpated 3 fingers below right costal margin and spleen was palpated 3 fingers below the left costal margin. Basic laboratory results were within normal limits.

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The patient did not have symptoms of heart failure and clinical euthyroid. Due to abdominal distention, the patient was referred to our department for contrast enema. Barium enema result was unremarkable.

Abdominal sonography was performed due to hepatosplemegaly. Sonography revealed enlargement of the liver. The hepatic celiac trunk and common hepatic artery were dilated. There was a tortuous artery at the hepatic hilar region connecting to the left portal vein. The left portal vein was dilated and had mixed arterial and venous waveform within it. Fistulous tract between the hepatic artery and portal vein was suggested. (Fig 1)

Splenomegaly and dilated periumbilical veins were demonstrated. There were generalized bowel wall thickening and ascites.

CT scan showed hepatomegaly with no focal mass lesion. Dilated left portal vein was seen. There was a connection between his left hepatic artery and his left portal vein. Splenomegaly with multiple collateral vessels including

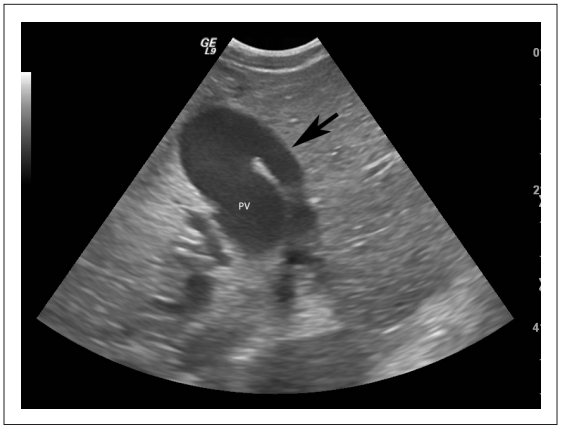

Fig 1A. Gray scale sonography of the liver showed dilated tubular structure connecting to the portal vein (PV)

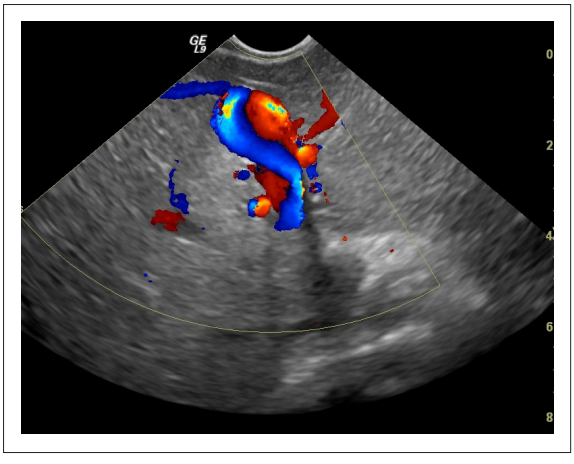

Fig 1B. Doppler Sonography of the liver demonstrated flow within the dilated tubular structure consistent with blood vessel. The flow within it has two opposite directions showing in red (toward the transducer) and blue (away from the transducer).

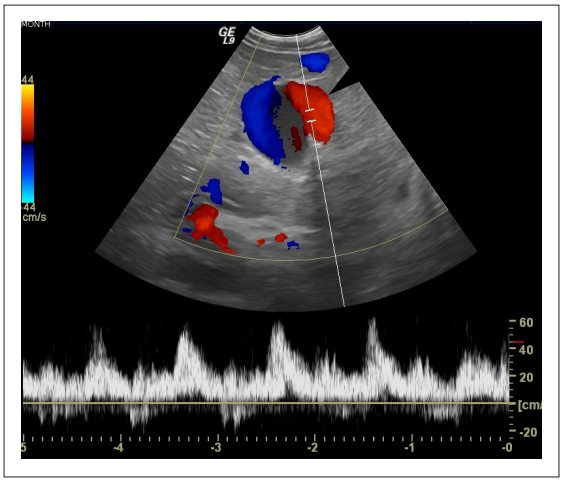

Fig 1C. Doppler sonography of the liver. The spectral waveform of the vascular structure on the right (arrow) is consistent with arterial waveform.

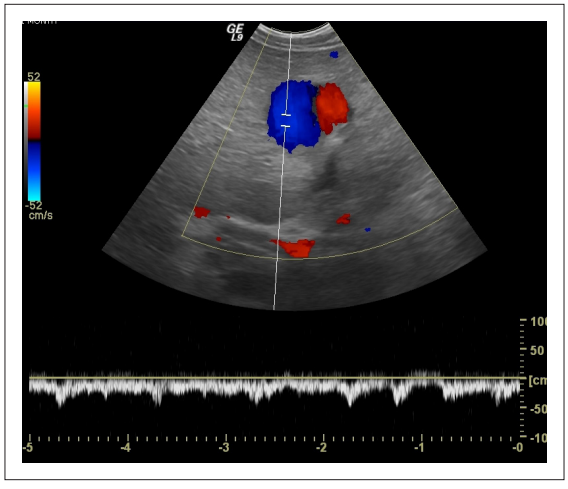

Fig 1D. Doppler sonography of the liver. The spectral waveform of the portal vein showed mixed arterial and venous waveform, suggesting fistula between portal vein and hepatic artery.

esophageal, paraesophageal and gastric varices were seen, consistent with portal hypertension. There were large ascites and diffuse thickening of bowel loops. (Fig 2)

Management

Hepatic angiography was performed showing a large hepatic-portal fistula, fed by multiple small vessels from hepatic arterial proper, right and left hepatic arteries to medial and lateral branch of the left portal vein. Due to small size and multiple arterial feeders, selection from arterial side was not successful. Direct puncture into the dilated portal vein was performed and selection of the fistula was performed under fluoroscopic and ultrasound guidance. The patient was treated with coil embolization (Micronester coils). Post embolization hepatic angiography showed 90% obliteration of the hepato-portal fistula. (Fig 3)

DISCUSSION

Abnormal connection between hepatic artery and portal vein can occur intrahepatic or extrahepatic. It can be congenital or acquired. The acquired anomalies are more common and can be caused by cirrhosis, hepatic neoplasms, trauma or iatrogenic cause such as biliary surgery.

Congenital arterioportal fistula is an intrahepatic communication between hepatic artery and portal venous system, without any systemic venous connection. The fistula may consist of a

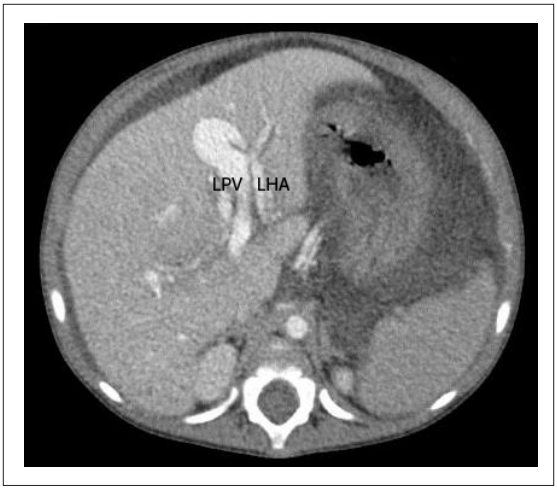

Fig 2A. CT scan through the liver, arterial phase demonstrated abnormal connection between the left hepatic artery (LHA) and left branch of the portal vein (LPV). The left portal vein is mildly dilated.

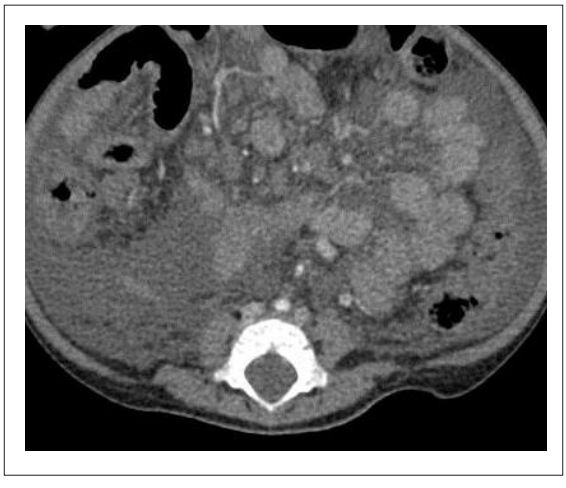

Fig 2B. CT scan at mid abdomen showed moderate ascites. There are mild thickening small bowel loops.

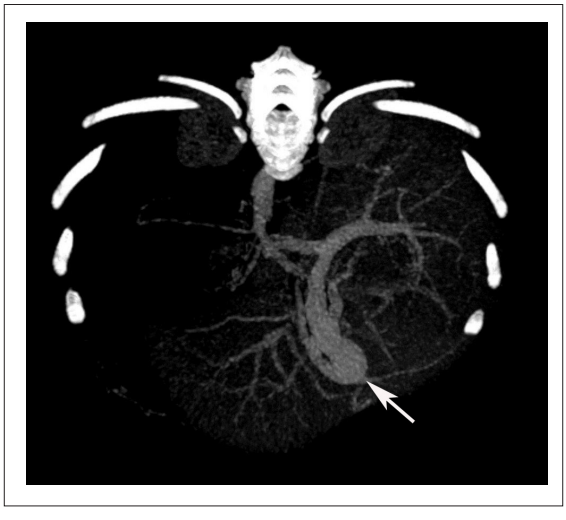

Fig 2C. CT scan with maximal intensity projection (MIP) demonstrated abnormal connection between left hepatic artery and left branch of the portal vein (arrow).

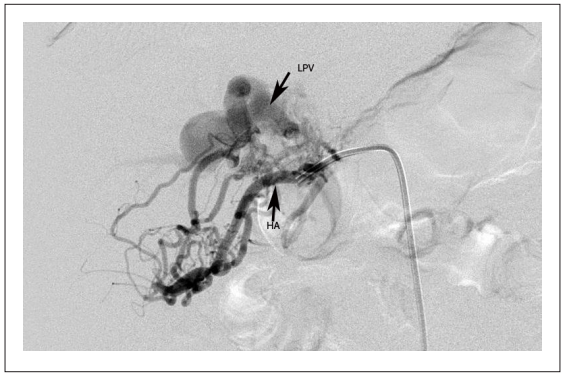

Fig 3A. Hepatic angiogram demonstrated hepatic-portal fistula from small branches of hepatic artery (HA) to left portal vein (LPV).

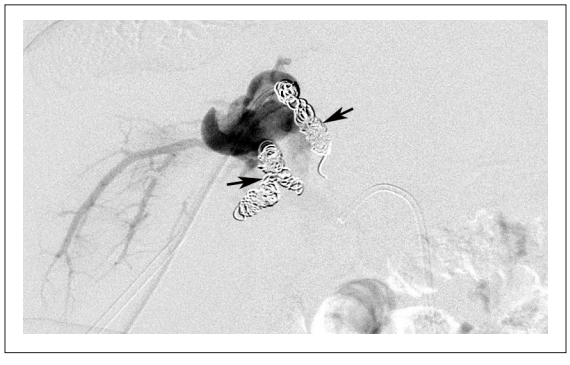

Fig 3B. Embolization was performed by portal vein approach from direct puncture. Micronester coils (arrows) was used to occlude two fistulae.

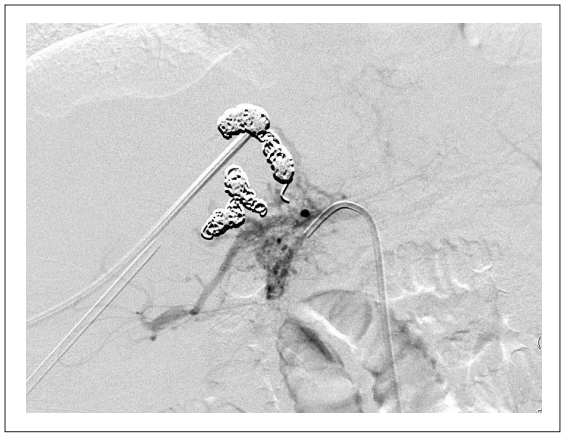

Fig 3C. Postembolization angiography demonstrated 90% obliteration of arterioportal portal fistula.

single arteriovenous connection or may be multiple connects with the same varix. It can be associated with hereditary hemorrhagic teleangiectasia, Ehlers-Danlos syndrome and biliary atresia.[1] Our reported case was considered to be congenital because there were no known secondary factors. Moreover, the patient was presented at a very young age.

The most common presentation of arterioportal fistula is the presence of portal hypertension with associated ascites due to increased blood flow in the portal system. Intestinal dysfunction such as malabsorption, diarrhea and steatorrhea can occur. Intestinal dysfunction is related to venous congestion and stasis secondary to reversal of flow in the portal and superior mesenteric veins.[2] Some patients may present with high output cardiac failure. Our patient had a complicated underlying conditions which included Down syndrome and congenital heart disease, and also presented with growth failure. Hepatosplenomegaly and multiple dilated superficial veins of the abdomen were detected on physical examination, suggesting portal hypertension.

Diagnosis can be performed by sonography, CT or MRI. Sonography is a fast, reliable, non-invasive low cost method to establish the diagnosis. Dilated portal vein, saccular dilatation of its branches and enlarged hepatic artery can be seen.[3] Doppler ultrasound shows pulsatile hepatofugal flow in the shunt portal vein.

Other congenital hepatic vascular malformations include arteriovenous malformation (AVM), portosystemic shunts, venous and lymphatic malformations. Sonographic findings of AVM include a nest of tortuous enlarged vessels in the liver with Doppler shifts in arteries and veins, a low arterial RI and increased pulsatility of veins.[4]

Portosystemic shunts, venous and lymphatic malformations are considered low-flow vascular malformations. Sonography shows no arterial flow and can be differentiated from arteriovenous shunts.

Definitive therapy is aimed to obliterate the shunt and restore normal portal and hepatic arterial hemodynamics. The options include transarterial embolization, surgical ligation of the implicated hepatic artery, partial hepatectomy or liver transplantation.[2,5]

Since the APF usually has a single feeding artery, the chance of successful occlusion by interventional radiological procedure is increased compare to arteriovenous malformation which has multiple feeding vessels.[6] The advantages of interventional procedures are shorter hospital stay, less pain and lower morbidity. The major complication of the large APF is portal vein thrombosis.[6]

Our patient was treated with Micronester coil embolization with satisfactory occlusion of the fistula. There were no immediate complications.

CONCLUSION

Intrahepatic shunt with the fistula between hepatic artery and portal vein can occur congenitally and should be in the differential diagnosis in children with clinically portal hypertension. Diagnosis can be easily established by color Doppler ultrasound by demonstrated anomalous connection of the vessels.

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