Congenital intrahepatic portosystemic shunts: Imaging findings and endovascular management

Rajsekar Chandrasekharan, Sreekumar K Pullara, Tixon Thomas, Nazar Puthukudiyil Kader, Srikanth Moorthy

Department of Radiology, Amrita Institute of Medical Sciences and Research Center, Kochi, Kerala, India

Correspondence: Dr. Rajsekar Chandrasekharan, Department of Radiology, Amrita Institute of Medical Sciences and Research Center, Amrita Lane, P.O. Elamakkara, Kochi, Kerala, India. E-mail: raj462750@gmail.com

Abstract

We present two cases of congenital intrahepatic portosystemic shunts in which the right portal vein directly communicated with the inferior venacava (IVC) in one patient and with the hepatic vein in the other. Multiple hepatic nodules consistent with focal nodular hyperplasia (FNH) were seen in the first patient. The second patient presented with recurrent history of hepatic encephalopathy. Percutaneous transhepatic embolization was performed using coils and Amplatz device following which she completely recovered.

Key words: Amplatzer device; amplatz vascular occluder device; congenital portosystemic shunts; hepatic encephalopathy; hypertrophied hepatic artery; intrahepatic portosystemic shunts; portal vein hypoplasias

Introduction

Congenital portosystemic shunts (CPSS) are rare. They are classified into extrahepatic and intrahepatic shunts. Intrahepatic CPSS less frequently show association with focal nodular hyperplasia (FNH), regenerative nodules, and encephalopathy unlike extrahepatic CPSS. Clinical complications like hepatic tumors and hepatic encephalopathy have been frequently reported in association with extrahepatic CPSS. We report two cases of intrahepatic CPSS -one associated with FNH in one patient and the other with recurrent encephalopathy in which endovascular management was attempted.

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Case Reports

Case 1

An 8-year-old female child presented with right hypochondrial pain. Biochemical evaluation revealed increased serum glutamic pyruvic transaminase (SGPT)/serum glutamic oxaloacetic transaminase (SGOT) and alkaline phosphatase levels. Sonography showed right lobe atrophy and multiple heteroechoic focal lesions in the liver. Multidetector computed tomography (MDCT) images showed multiple lobulated and well-defined isodense and hypodense lesions scattered throughout the liver, predominantly in the right lobe. Arterial phase showed mild and homogenous enhancement of all focal lesions except one lesion which showed a heterogeneous enhancing pattern [Figure 1A and B]. Unenhanced CT images showed multiple small hyperdense foci within the segment VI/VII indicating hemorrhagic foci. The hepatic artery was hypertrophied and tortuous [Figure 1C]. Portal phase of the multiphase CT showed a hypoplastic left portal vein. Right intrahepatic portal vein was seen to drain directly into the hepatic segment of inferior venacava (IVC) [Figure 1B]. Biopsy of a liver focal lesion and its histopathological analysis showed

benign hepatocellular hyperplasia which may represent nodular hyperplasia or nodular regenerative hyperplasia.

Case 2

A 36-year-old female presented with features of encephalopathy and bleeding per rectum. Biochemical evaluation revealed elevated level of blood ammonia and prolonged prothrombin time. SGPT/SGOT was also mildly increased and the albumin to globulin ratio was reversed. Sonography showed features like altered echo texture and right lobe atrophy. MDCT revealed diffuse fatty changes. Anterior and posterior branches of the right portal vein appeared tortuous and dilated. Two intrahepatic portal vein to right hepatic vein shunts were seen. The larger shunt involved drainage of the entire anterior branch of the right portal vein into the right hepatic vein [Figure 2A]. The smaller one was between a branch of posterior division of the right portal vein and a tributary of the right hepatic vein. Middle and left hepatic veins were normal. Mild splenomegaly was present. No porto systemic collaterals were present. Liver biopsy showed mild steatosis with inflammation and portal fibrosis. In view of recurrent episodes of encephalopathy, it was decided to embolize the larger fistula. Under general anesthesia, ultrasound-guided transhepatic right portal vein puncture was performed using Chiba needle followed by insertion of 6F sheath. Portogram showed a large venous sac connecting the right anterior division of portal vein with right hepatic vein [Figure 2B]. Amplatz closer device was deployed to prevent distal migration of coils, following which 8 × 8 mm and 6 × 6 mm (0.35″) stainless steel coils (Cook, Bloomington, IN) were placed to embolize the shunt [Figure 2C-E]. The smaller shunt was not treated. The patient made an uneventful recovery.

She has had no episodes of encephalopathy in a follow-up period of 2 years.

Discussion

Broadly, portosystemic shunts have been classified as intrahepatic and extrahepatic.[1] Intrahepatic shunts are commonly seen in Budd-Chiari syndrome and sometimes following blunt trauma. Extrahepatic communications through collaterals are usually present in patients with cirrhosis and portal hypertension and in patients with thrombosis of splenic or portal vein. However, congenital CPSS are rare.[2] Morgan and Superina proposed a classification for congenital extrahepatic shunts, also known as "Abernethy malformations" based on the portosystemic veins that are involved.[3] Park *et al*. later reported a few cases and proposed a classification for congenital intrahepatic shunts into subtypes based on the location of the shunt, its number, and shunt characteristics.[1]

Congenital intrahepatic portosystemic venous shunt is an uncommon condition that probably results from abnormal embryonic development by the fourth week of fetal life.[4,5] The vitelline and umbilical systems begin to break into intrahepatic sinusoids that give rise to the intrahepatic portal and hepatic veins, respectively. Congenital portosystemic shunts are thought to represent persistence of communications between the portal and vitelline venous systems.[4,5] Also, reduction in blood flow to the liver may result in fatty degeneration, hepatic dysfunction, and atrophy of the liver.[6] The usual sonographic findings include abnormal cystic or tubular, anechoic, serpiginous vascular structures which seems to communicate the portal with the systemic circulation.[1] Doppler study can confirm the vascular nature of the structures and calculate the shunt ratio (total blood flow volume in the shunt divided by the
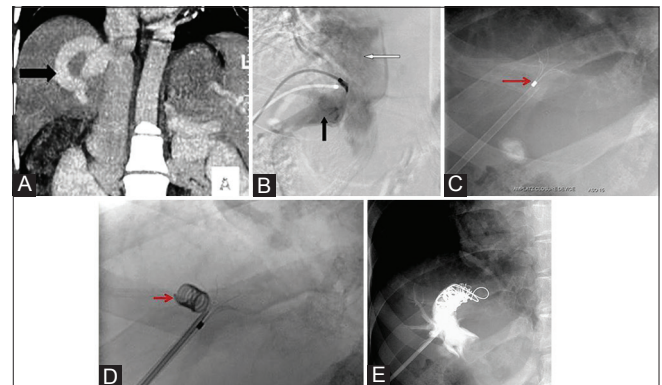

Figure 1 (A-C): An 8-year-old female child with multiple focal nodular hyperplasia-like lesions. Axial CT image in portal venous phase (A) shows multiple minimally enhancing lesions in both lobes of liver (long white arrow) (B) Abnormal portal venous channel connecting the right portal vein and IVC (curved arrow), consistent with portocaval shunt (C) Heterogeneous lesion at segment VI (black arrow) and hypertrophied hepatic artery (short white arrow)

Figure 2 (A-E): A 36-year-old female presented with encephalopathy. Axial CT image in portal phase (A) shows the anterior branch of the right portal vein joining with the right hepatic vein (black arrow). Portogram (B) showing venous sac connecting the portal vein (white arrow) with the right hepatic vein (black arrow) (C) Introduction of the Amplatz vascular device (arrow) through 6F vascular sheath (D) Deployment of metallic coils (arrow) proximal to the Amplatz occluder device. Post embolization angiogram (E) showing occlusion of the portosystemic venous shunt

blood flow in the portal vein). It has been recommended that a shunt ratio greater than 60% should be corrected to prevent complications.[1]

FNH is thought to be a hyperplasic hepatocellular response to increased arterial perfusion from an underlying congenital vascular malformation (like CPSS).[7] Hepatic ischemia and reduction in portal vein flow with subsequent inadequate delivery of growth factors and hormones have also been postulated as etiological factors.[8] FNH-like lesions have been reported in association with only extrahepatic CPSS and very rarely in patients with intrahepatic CPSS.[3,9] However, the number of cases in these reported series is very small. Moreover, the essential milieu for developing nodules like portal vein flow diversion and arterializations is present in both intrahepatic and extrahepatic shunts.[7-11] Both typical FNH and nodular regenerative nodules show homogenous and strong enhancement in the arterial phase. However, atypical FNH (FNH-like) may be multifocal, and may show heterogeneous enhancement due to necrosis and hemorrhage, as noted in our case.[7,8,11,12] FNH and atypical FNH have histopathologically similar appearance.[12] In case of congenital porto systemic shunt with liver tumors, interval follow-up with ultrasound and liver function tests is warranted.[2]

In an early cirrhotic or non-cirrhotic patient presenting with recurrent hepatic encephalopathy, porto systemic venous fistula should be a differential diagnosis. They often respond poorly to conventional medical therapy. Previous reports suggested surgical correction of the portosystemic shunt. But interventional radiological techniques are preferred now. Endovasvcular management techniques commonly performed use microcoils for the shunt embolisation. However few previous studies report use of balloon and sclerosing agents. Gupta *et al.* have reported successful treatment of congenital intrahepatic porto systemic shunt with embolization using *n*-butyl cynoacrylate in a 14-month-old child.[13] Lee *et al.* demonstrated successful embolization using the Amplatz vascular plug II.[14] In our patient, embolization was performed with a combination of the Amplatz device and coils, which has been rarely reported. The Amplatz device prevents distal migration of the coils while functioning as a scaffolding to form a dense coil mesh. Children are more resistant to hepatic encephalopathy than adults. In these patients, meticulous clinical and ultrasound follow-up must be performed. Mild metabolic abnormalities associated with a portosystemic shunt can be managed with medical therapy and dietary modifications such as a reduced protein diet.

Conclusion

Congenital CPSS are rare and represent persistent embryologic connection between portal system and the systemic veins. The detection of a portal vein to IVC shunt in a patient with no background cirrhosis or findings of Budd-Chiari syndrome should raise the possibility of CPSS.

CIHPSS rarely show association with benign hepatic tumors and hepatic encephalopathy. In our series, one patient presented with hepatic encephalopathy and the other presented with multiple FNH-like lesions. Embolization of the shunt with microcoils, Amplatz device, and both is now considered the treatment of choice for intrahepatic portosystemic shunt. In our patient, embolization of shunt was done with both coils and Amplatz device. Follow-up of the patient showed significant improvement.

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