Neonatal Cholestasis as Initial Presentation of Portosystemic Shunt: A Case Report

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Congenital intrahepatic portosystemic shunts are rare in children. Portosystemic venous malformations are characterized by extreme clinical variability. We report a full-term 33-day-old male infant presenting with neonatal jaundice. On physical examination, he had generalized icterus and the liver was palpable 3.5 cm below the right costal margin. He had no other symptoms. Laboratory tests showed AST 632 U/L, ALT 198 U/L, total bilirubin 12.1 mg/dL, conjugated bilirubin 10.2 mg/dL, alkaline phosphatase 753 U/L, GGT 47 U/L and glucose 67 U/L. Colour Doppler ultrasonography showed the left portal vein was more dilated than the right portal branch and communication with dilated left hepatic vein. There was no evidence of portal hypertension, heart failure, hepatopulmonary syndrome and encephalopathy during his hospital stay, so he was discharged from the pediatric department and his parents advised to attend monthly follow-up. Congenital portosystemic shunts are rarely observed in the childhood period.[1] Depending on anatomic characteristics they may be intrahepatic or extrahepatic.[2] Intrahepatic portosystemic shunts (PSS) are observed between the portal vein and hepatic vein or vena cava inferior.[3,4] Small shunts may close themselves before the age of 2 years.[5] With the increase in use of imaging methods, diagnosing PSS has become easier, with an increase in the number of cases reported.[6] Neonatal cholestatis is a frequent complication of PSS.[1] We present a case presenting with neonatal cholestasis diagnosed with congenital intrahepatic PSS. (J Clin Exp Hepatol 2016;6:331–334)

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CASE

A 33-day old male applied to the pediatric gastroenterology clinic due to complaint of jaundice. It was learned that jaundice began at 4 days old, with occasional white (acholic) stools. The case was born at term by cesarean section weighing 3500 g and no previous known disease history was found for him or his family. Physical examination found weight 4600 g (50–75 percentile), length 53 cm (25–50 percentile), skin and sclera icterus, the liver was palpable 3.5 cm below the right costal margin, and spleen could not be palpated. Neuromotor development was normal. Laboratory studies showed a serum aspartate transaminase 632 U/L, alanine transaminase 198 U/L, total bilirubin 12.1 mg/dL, conjugated bilirubin 10.2 mg/dL, alkaline phosphatase 753 U/L, LDH: 515 U/L, glucose 67 mg mg/dL, PT: 14.2 s, INR: 1.26, stool steatocrit +3, TSH: 3.8 uU/mL and FT4: 1.3 ng/ml. Stool samples had acholic appearance. Considering metabolic disease, there was no hypoglycemia or septic appearance. Eye examination found no cataracts. Examination for infectious etiologies was negative (toxoplasma, cytomegalovirus, Epstein–Barr virus, hepatitis A, B, C). Alpha 1-antitrypsin (PiMM) and cystic fibrosis mutations were not detected. Eye examination was normal in terms of metabolic diseases. Abdominal sonography showed hepatomegaly (80 mm) with normal contours and echo pattern of the liver. The gall bladder and intrahepatic biliary system were normal. The main and right portal veins were normal on Doppler ultrasound examination (DUSI). The left portal vein was more dilated than the right portal branch. It was linked to the dilated left hepatic vein through a 0.8 mm aberrant venous connection (Figure 1). The left portal vein showed a continuous waveform spectrum on DUSI, whereas the left hepatic vein showed a mild

Keywords: cholestasis, portosystemic shunt, infant
Received: 7.03.2016; *Accepted:* 12.08.2016; *Available online:* 31 August 2016
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Abbreviations: AST: aspartate transaminase; ALT: alanine transaminase; DSUG: Doppler ultrasound; GGT: gamma glutamyl transferase; LDH: lactate dehydrogenase; PSS: portosystemic shunts
http://dx.doi.org/10.1016/j.jceh.2016.08.007

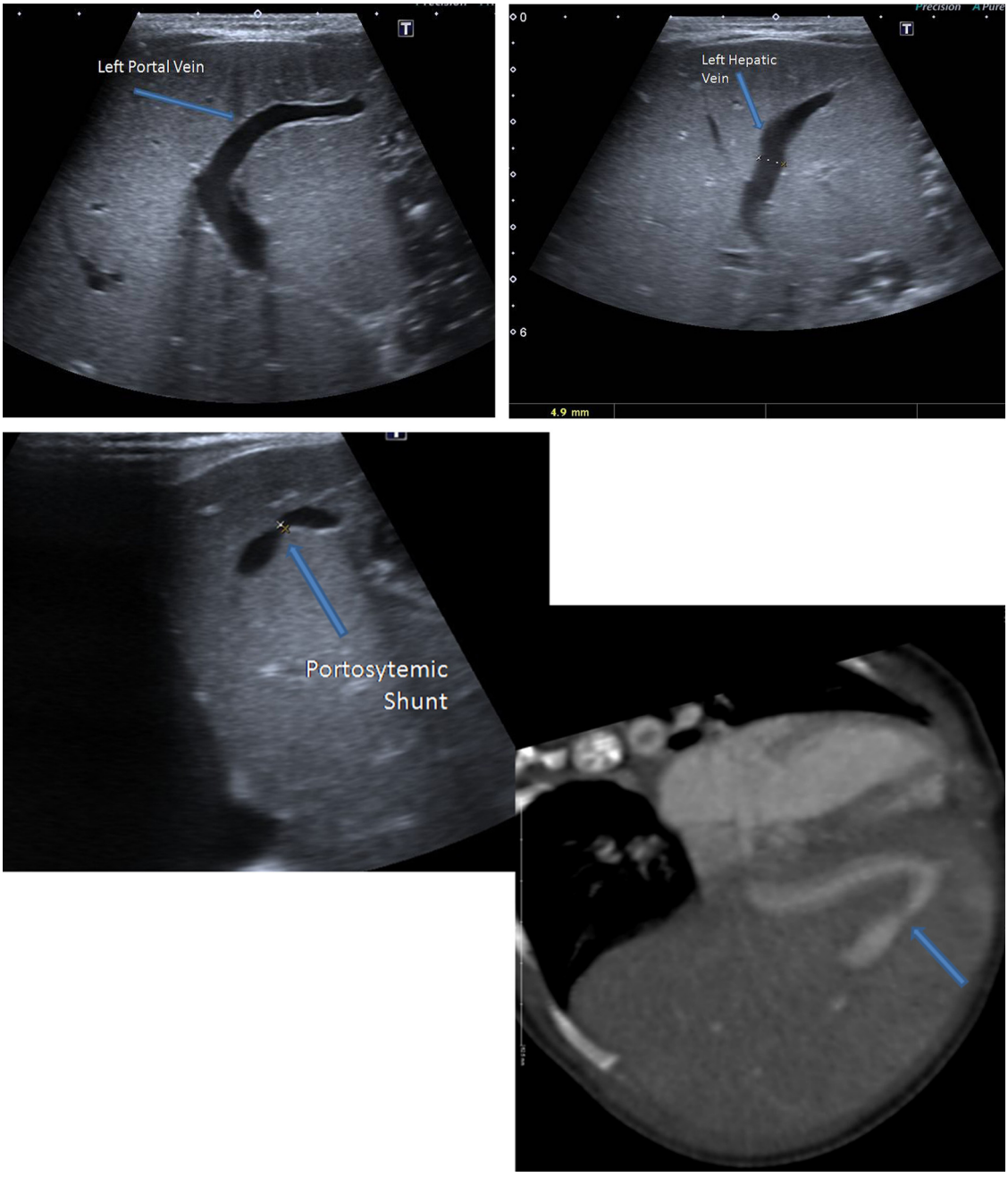

Figure 1 Ultrasonography and reformatted contrasted CT imaging show connection between dilated vena porta and left hepatic vein (white arrow).

increased turbulent flow. Other hepatic veins had normal phasic flow pattern and diameter. As cardiac pathologies may occur in shunt patients, echocardiography was performed for screening purposes. It was interpreted as normal. Echocardiographic evaluation was normal. Neurological examination found no evidence of encephalopathy. (Evaluating encephalopathy in this patient was not as easy as with older children. However the patient was active, mobile, with lively appearance, had no difficulty feeding, and had normal sleep habits for his age, so encephalopathy was not primarily considered.) Due to cholestasis, the patient was begun on ursodeoxycholic acid treatment. During hospital stay, no complications such as portal hypertension, heart failure and hepatopulmonary syndrome developed. The family was informed of complications that might develop. The patient was discharged after an 8-day hospital stay, with good activity, nutrition and weight gain and called for regular follow-up. The patient was monitored for 5 months. On follow-up the patient's hemogram, biochemistry and coagulation values

and stool color returned to normal. As a result the patient was not directed to another center for tests that our hospital does not perform. Control USI when the patient was 6 months observed the shunt had closed.

DISCUSSION

Congenital intrahepatic shunts are rarely observed venous anomalies forming between the portal vein and hepatic vein or portal vein and inferior vena cava.[7] It is thought that congenital intrahepatic portosystemic venous shunts develop due to anastomosis between the subcardinal venous system and vitelline venos system in the early period of embryological development.[4,6,8,9] Another theory for intrahepatic PSS is rupture or portal vein aneurysm into the hepatic vein.[4,8] Percutaneous liver biopsy is a frequent cause of intrahepatic PSS.[10] Patients are questioned about liver biopsy, trauma and operation history before diagnosis of congenital PSS. Intrahepatic PSS cause partial or full changes in direction of portal blood flow to systemic circulation.[1,11] Park et al. divided intrahepatic PSS into 4 separate types depending on morphology.[1] In the most common type I a wide diameter single vein linking the right portal vein into the vena cava inferior is observed. Type II is observed with single or multiple linkages between peripheral branches of portal and hepatic veins localized in the single hepatic segment. In Type III peripheral portal and hepatic vein branches are linked due to an aneurysm. In Type IV there are multiple linkages between peripheral portal and hepatic vein branches in both lobes of the liver.[10] According to this classification, our case was in accordance with Type II. There are reports of cases diagnosed with PSS due to abnormalities during galactosemia scans or liver enzyme tests, increased serum bile acid levels and increased blood ammonia levels.[11,12] In the literature there are rare cases reported with acute liver failure, heart failure, rectal hemorrhage, glomerulopathy and hyperandrogenism.[11]

There are cases of neonatal cholestasis diagnosed with congenital shunt. In a study assessing 265 children with congenital PSS,[1] 24 cases were reported with neonatal cholestasis. Of these cases, 10 were diagnosed while researching jaundice, while 14 were diagnosed during galactosemia screening. Our patient had jaundice and acholic stool complaints, with hepatomegaly identified and disrupted biochemical parameters and coagulation, so when full abdominal USI was requested to determine the cause of cholestasis the shunt was identified. Considering that cholestasis may not be linked to the shunt, we researched all other causes of cholestasis in our patient according to our resources. During hospital stay and clinical follow-up, jaundice regressed, weight gain was in accordance with age and activity appeared good. With cholestasis resolved, UDC treatment was stopped for the patient. Our case applied with jaundice, and considering the role of congenital shunts in neonatal cholestasis, imaging methods were used and diagnosis made. In a study of 10 cases,[7] two cases were observed to have neonatal cholestasis. A different study evaluated 22 children with PSS and found 5 cases had cholestatic jaundice since birth, while 3 of these 5 cases were diagnosed in the prenatal period.[11] Researching PSS should be a part of history, examination and testing of children with neonatal cholestasis. However, the presence of a shunt should not prevent research into other causes of neonatal cholestasis etiology. Our case was tested for other causes of cholestasis etiology for this reason and results were found to be normal.

Diagnosis of congenital portosystemic shunt has become more common with the increase in use of methods such as DUSI, computed tomography and magnetic resonance imaging.[1] With careful obstetric examination and prenatal USI, diagnosis may even be made in the prenatal period.[7,11] Neonatal cholestasis, hypoglycemia, hepatopulmonary syndrome, pulmonary hypertension, portosystemic encephalopathy, liver tumors and pancreatitis are significant complications observed with PSS.[3,11] Small intrahepatic shunts may spontaneously close in patients under the age of 2 and monitoring of patients under the age of 2 is recommended in the literature.[1,11] In accordance with the literature we began monitoring our patient; in the 6th month of follow-up the patient's shunt was observed to have closed. To date development is in accordance with age and there are no other complaints. As a result we did not request other cholestasis tests that are not performed at our hospital. For cases above the age of 2, to prevent development of complications, it is necessary to close them with endovascular methods or surgery. As our case was diagnosed at an early age and no complications developed, they were monitored at regular follow-ups.

In conclusion, in children with neonatal cholestasis possible PSS should be kept in mind and appropriate investigations performed.

CONFLICTS OF INTEREST

The authors have none to declare.

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