INTRODUCTION

Congenital intrahepatic portosystemic shunt is a rare anomaly. In humans, the incidence of the portosystemic shunt is estimated at 1/30,000 births. The intrahepatic portosystemic shunt in association with an aneurysm is not reported in neonates as most case reports mention such shunt in the adult age group.[1,2] Furthermore, the early occurrence of pulmonary hypertension in neonatal age is very unusual.

CASE REPORT

A late-preterm male newborn (35 weeks) delivered by cesarean section was referred to our center for further management as the baby developed respiratory distress. The initial chest radiograph showed cardiomegaly with bilateral diffuse lung opacities. In view of increasing oxygen requirement, the baby was intubated and the surfactant was administered. Echocardiography on day 2 of life showed dilated right and left ventricles with moderate tricuspid regurgitation and severe pulmonary arterial hypertension (PAH) (mean pulmonary artery pressure ~58 mm Hg) for which baby was started on intravenous sildenafil infusion. The baby maintained oxygen saturation; however, there was persistent tachypnea. Initial liver function tests on day 2 of life showed elevated bilirubin levels with normal liver enzymes. Bilirubin levels showed progressively decreasing values with time. Blood ammonia levels measured on day 8 of life were elevated (~163.6 μmol/lit; Laboratory reference value was 1–35 μmol/lit).

Ultrasound of the abdomen on day 2 of life showed a Park et al. type 3[3] congenital intrahepatic portosystemic shunt through an aneurysm with associated mild ascites and diffuse subcutaneous edema [Figures 1-5]. Initially, the shunt was managed conservatively using intravenous sildenafil and optimizing ventilator settings based on serial radiographs. However, tachypnea and severe PAH failed to respond to conservative management. Echocardiography done on day 7 of life showed persistent severe PAH (mean pulmonary artery pressure ~58 mm Hg). Persistent severe PAH...
was attributed to intrahepatic shunt and percutaneous shunt closure was planned. Contrast-enhanced computed tomography of the abdomen was acquired before shunt closure which confirmed a tubular vascular channel connecting the left branch of the portal vein and left hepatic vein through an aneurysm [Figure 6].

Percutaneous shunt closure was performed on day 14 of the life. In the catheterization laboratory, after securing the right internal jugular vein access, left hepatic vein injection reconfirmed the diagnosis of type 3 intrahepatic portosystemic shunt connected through aneurysm in accordance with the sonogram and CT scan [Figure 7]. Assessment of pulmonary hemodynamics revealed pulmonary hypertension before the shunt closure. Aneurysmal sac measured ~6 mm × 6 mm and shunt measured ~4 mm in width. The shunt was successfully closed by deploying V-Trak microvention coils (Terumo Corporation, Japan) into the aneurysmal sac. Closure of the shunt was confirmed in postembolisation hepatic venous contrast run. The child showed clinical improvement after the procedure and was discharged subsequently in a stable condition. Liver function tests including blood ammonia levels measured after the procedure showed normal values. Ultrasound of the abdomen showed no residual flow in the portosystemic shunt. During the 6 months postprocedure follow-up period, the child remained asymptomatic with normal liver function and complete resolution of pulmonary hypertension.

**DISCUSSION**

During prenatal life, congenital intrahepatic portosystemic shunt on ultrasound can cause fetal growth restriction in
the neonatal age group, patients usually present from complications of the shunt.\[4\]

Various complications such as hepatic encephalopathy, neonatal cholestasis, liver tumors, PAH, and hepatopulmonary syndrome have been described in the literature. Bernard et al. reported the incidence of pulmonary hypertension in 30 out of 180 children at ages ranging from the neonatal period to 15 years (mean: 5 years 4 months) with all anatomic types of the shunt.\[5\] Congenital portosystemic shunt commonly coexists with congenital malformations, such as cardiovascular anomalies, polysplenia, annular pancreas, and biliary atresia. Some small intrahepatic portosystemic shunts located between the portal branches and hepatic veins disappear spontaneously by the age of 1–2 years.\[4,6\]

Park et al.\[3\] categorized intrahepatic portosystemic shunts arbitrarily into four different morphologic types. The first and most common type is a single large tube of a constant diameter that connects the inferior vena cava to the right portal vein. The second type includes single or multiple communications between peripheral branches of hepatic and portal veins in one hepatic segment. The third type is the rarest, with aneurysmal communication between the peripheral portal vein and hepatic vein. The fourth type has a peripheral portal and hepatic veins showing multiple communications diffusely involving both lobes.

Chevallier et al.\[7\] classified intrahepatic portosystemic venous shunts into four categories based on their clinical and anatomic features: Type I includes paraumbilical veins, such as those encountered in portal hypertension; Type II involves a connection between a portal branch and hepatic vein in adjacent liver segments; Type III comprises portal and hepatic vein connections between nonadjacent liver segments, and Type IV includes any connection between the right portal branch and inferior vena cava. Findings in our case were consistent with the Type III shunt per Park et al.\[3\] and Type II shunt per Chevallier et al.\[7\]

Grayscale sonography demonstrated a tubular anechoic channel connecting the portal vein branch with the hepatic vein. Color Doppler sonography demonstrated the presence of blood flow within the shunt including the direction of flow. De Gaetano et al.\[8\] have reported low velocity, bidirectional, or helical flow in the aneurysm on Doppler sonography. Multidetector computed tomography with contrast or magnetic resonance imaging of the abdomen with contrast can be used for delineation of shunt anatomy before intervention, excluding associated anomalies, especially if the ultrasound evaluation is suboptimal.

The vascular anomaly may regress spontaneously during infancy.\[9\] The definitive treatment for
portohepatic shunts can be considered in patients beyond the 1st year of life or those presenting with life-threatening complications related to the shunt. Most authors also agree that patients with symptomatic congenital portosystemic shunt should be treated.[10]

As there are no well-established guidelines, treatment protocols may vary between various centers.

Endovascular intervention is minimally invasive and usually the preferred treatment modality for shunt closure. Prompt reduction in the symptoms with correction of biochemical abnormalities can be noted following endovascular closure of shunt. Various endovascular embolization materials such as coils, balloons, and vascular plugs can be used depending on the shunt.[11] Surgery is preferred in cases with large shunts with a risk of inadvertent migration of embolic agents during embolization or after failed embolization with shunt recurrence or persistence.[11] Surgical options include ligation of the portal vein, resection, or even lobectomy. Treatment must be tailored according to the clinical presentation, type, and hemodynamics of the shunt, and availability of the treatment modalities.

CONCLUSION

The portosystemic shunt can present with varied clinical manifestations. It is crucial to suspect it in case of unexplained pulmonary hypertension, as in most of the cases, it is treatable either by surgical or catheter interventions.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Tanoue S, Kiyosue H, Komatsu E, Hori Y, Maeda T, Mori H. Symptomatic intrahepatic portosystemic venous shunt: Embolization with an alternative approach. AJR Am J Roentgenol 2003;181:71-8.
2. Chagnon SF, Vallee CA, Barge J, Chevalier LJ, Le Gal J, Blery MV. Aneurysmal portohepatic venous fistula: Report of two cases. Radiology 1986;159:693-5.
3. Park JH, Cha SH, Han JK, Han MC. Intrahepatic portosystemic venous shunt. AJR Am J Roentgenol 1990;155:527-8.
4. Uchino T, Matsuda I, Endo F. The long-term prognosis of congenital portosystemic venous shunt. J Pediatr 1999;135:254-6.
5. Bernard O, Franchi-Abella S, Branchereau S, Pariente D, Gauthier F, Jacquemin E. Congenital portosystemic shunts in children: Recognition, evaluation, and management. Semin Liver Dis 2012;32:273-87.
6. Franchi-Abella S, Branchereau S, Lambert V, Fabre M, Steinberg C, Losay J, et al. Complications of congenital portosystemic shunts in children: Therapeutic options and outcomes. J Pediatr Gastroenterol Nutr 2010;51:322-30.
7. Chevallier P, Oddo F, Souci J, Diaine B, Padovani B. Macroscopic intrahepatic portosystemic venous shunt: Review of the literature and reclassification. J Radiol 2000;81:597-604.
8. De Gaetano AM, Rinaldi P, Barbaro B, Mirk P, Di Stasi C, Gui S, et al. Intrahepatic portosystemic venous shunts: Color Doppler sonography. Abdom Imaging 2007;32:463-9.
9. Gitzelmann R, Forster I, Willi UV. Hypergalactosaemia in a newborn: Self-limiting intrahepatic portosystemic venous shunt. Eur J Pediatr 1997;156:719-22.
10. Franchi-Abella S, Gonzales E, Ackermann O, Branchereau S, Pariente D, Guerin F, International Registry of Congenital Portosystemic Shunt members. Congenital portosystemic shunts: Diagnosis and treatment. Abdom Radiol (NY) 2018;43:2023-36.
11. Papamichail M, Pizanias M, Heaton N. Congenital portosystemic venous shunt. Eur J Pediatr 2018;177:285-94.