Journal of Pediatric Surgery (2012) 47, E27–E31

ELSEVIER

Journal of Pediatric Surgery

www.elsevier.com/locate/jpedsurg

Congenital intrahepatic portohepatic shunt managed by interventional radiologic occlusion: a case report and literature review ☆

Chiara Grimaldi [a], Lidia Monti [b], Piergiorgio Falappa [c], Giuseppe d'Ambrosio [a], Antonio Manca [d], Jean de Ville de Goyet [a,*]

[a] *Liver surgery and Transplantation Unit, Children's Hospital Bambino Gesù, Rome, Italy*
[b] *Hepatobiliopancreatic Radiology Unit, Children's Hospital Bambino Gesù, Rome, Italy*
[c] *Interventional Radiology Unit, Children's Hospital Bambino Gesù, Rome, Italy*
[d] *"B. Trambusti" Unit, " Policlinico" Hospital, Bari*

Received 4 June 2011; revised 27 August 2011; accepted 23 October 2011

Key words:
Liver anatomy;
Congenital malformation;
Portosystemic shunt;
Hepatopulmonary syndrome;
Interventional radiology

Abstract Congenital intrahepatic portosystemic shunts are rare hepatic vascular anomalies that often lead to severe secondary conditions. A 6-year-old boy was referred for assessment of severe hypoxia, and a large liver mass was diagnosed with such a malformation and was managed by direct closure of the venous fistula by interventional radiology. Follow-up assessment shows normalization of the respiratory condition and a progressive reduction of the vascular liver lesion. Review of literature suggests that radiologic interventions are currently the criterion standard for managing these conditions, with surgery reserved for patients who are not eligible for radiologic procedure and those requiring liver transplantation.
© 2012 Elsevier Inc. All rights reserved.

Congenital portosystemic shunts (CPSS) are rare vascular malformations (1/30,000) classified into 2 types: type I (extrahepatic) and type II (intrahepatic). Although the condition can be symptom free and discovered incidentally, patients can present with severe related complications including hepatopulmonary syndrome, pulmonary arterial hypertension, liver tumor, encephalopathy, heart failure, or neonatal cholestatic jaundice [1,2].

Because of the anatomical proximity of the vascular structures involved (portal vein and hepatic vein or vena cava), the anatomy of the shunt itself is characterized by a short fistulous communication between the 2 venous systems; therefore, percutaneous shunt closure can be challenging. When the shunt is large in diameter, clinical symptoms can develop early in childhood, making the technical challenge of closing a large shunt in a small child even greater. Large congenital shunts are usually associated with severe hypoplasia of a part or the entire intrahepatic portal venous system. This results in diversion of a large part of the splanchnic flow directly into the caval system and deprives the latter veins from flow and causes development failure. These factors make it difficult or dangerous to close the shunt as a single

☆ Note: Amplatzer: see http://international.amplatzer.com.

* Corresponding author. Department Surgery and Transplantation Center, Ospedale Pediatrico Bambino Gesù, Piazza Sant'Onofrio 4, 00165 Roma, Italy.
E-mail address: deville@opbg.net (J. de Ville de Goyet).

0022-3468/$ – see front matter © 2012 Elsevier Inc. All rights reserved.
doi:10.1016/j.jpedsurg.2011.10.079

step intervention, and may indicate closing the shunt by a staged approach. Several cases have been reported in literature that mention successful surgical banding or transplantation. More recently, interventional radiologic occlusion of the shunt has been proposed as a cure.

We describe a child with a huge intrahepatic CPSS, presenting with hypoxia (hepatopulmonary syndrome) and a vascular liver mass, both considered as complications associated and secondary to portosystemic shunting who was managed successfully by radiologic intervention.

1. Case report

A 6-year-old white boy with a history of recurrent mild bronchopulmonary infections was seen at a local hospital for progressive dyspnea and fatigue. Physical examination showed digital clubbing and cyanosis with an oxygen saturation (SaO_2) of 72% on room air. Thoracic auscultation, lung function tests, bronchoscopy, chest radiograph, and computed tomographic scan of thorax and lungs were unremarkable, with only mention of a slightly enlarged heart. A provisional diagnosis of hypoxia caused by pulmonary fibrosis resulting from viral infection was proposed: the child was receiving nasal O_2 at the rate of 8 L/min in resting condition to maintain a 100% level of SpO_2.

Because of the incidental finding of a liver mass on magnetic resonance imaging (MRI), the child was referred to our institution for further evaluation. On detailed ultrasonographic (US) liver examination, B-mode images showed an abnormal communication between the right portal branch and the median hepatic vein, the former malformation appearing as a complex and dysmorphic vascular structure. The left portal venous system was not visible. Color Doppler examination confirmed the diagnosis of a veno-venous shunt, with flow characterized by a monomorphic spectrum and high velocity within the shunt and inferior vena cava (up to 100 cm/s). The hepatic arterial signal was slightly increased with a hyperkinetic spectrum. On review of the MRI, the presence of the large right hepatic mass was confirmed, characterized by a vascular rim, radial vascular architecture, and a central scar, at first suggesting focal nodular hyperplasia (normal serum α-fetoprotein levels). The angio-MRI study, however, confirmed the presence of a complex vascular malformation within the right liver, between the right branch of the portal vein and the median hepatic vein. Contrast echocardiography showed massive right to left shunting without an abnormal intracardiac communication, confirming the diagnosis of hepatopulmonary syndrome as the cause of the boy's hypoxia.

With a diagnosis of intrahepatic CPSS complicated by hepatopulmonary syndrome and vascular liver mass, a conventional angiographic study was obtained to evaluate the precise vascular anatomy of the malformation and possibly occlude the shunt. Transjugular hepatic catheterization and venography with subsequent retrograde portography showed a double veno-venous fistulous tract connecting the right portal vein and the median hepatic vein (Fig. 1). Venography confirmed that the left portal veins were present but hypoplastic. Temporary balloon occlusion of the dominant fistula completely interrupted the flow through the whole malformation (direct portography through the catheter showing no residual fistulous flow) and confirmed that the portal pressure remained within acceptable range during shunt closure (portocaval gradient measured 7 mm Hg during shunt occlusion). Interestingly, the portal flow immediately rerouted into the right and left hepatic sectors (including the left portal system).

These preliminary observations indicated that radiologic occlusion of the fistulous tract was appropriate. After measurement of the internal diameter of the fistula by balloon catheter (Amplatzer sizing balloon II), a 18-mm Amplatzer II device was chosen with the device diameter to be 150% of the diameter of the fistulous tract, as measured by the sizing balloon (as recommended by the manufacturer) and positioned inside the dominant fistula, with both ends of the device coming to the edge of portal vein and hepatic vein, respectively (Fig. 2). During the procedure and after positioning the device, angiography and US Doppler study confirmed absence of flow through the malformation, with the second fistulous tract closed by compression inside the malformation, so that no further treatment was necessary.

The child's postoperative (PO) course was characterized by:

a) transient worsening of hypoxia (SaO_2 reduced to 50% in absence of supplemental O_2) and increased O_2 requirements (up to 10 L/min at rest). A computed tomography of the chest ruled out pulmonary embolism secondary to the procedure. Progressive improvement was observed during the second week after intervention, with continuing lowering needs of O_2 and improving resting SaO_2 with time thereafter.
b) A transient increase in serum transaminases, γ-glutamyl transferase, and bilirubin levels was

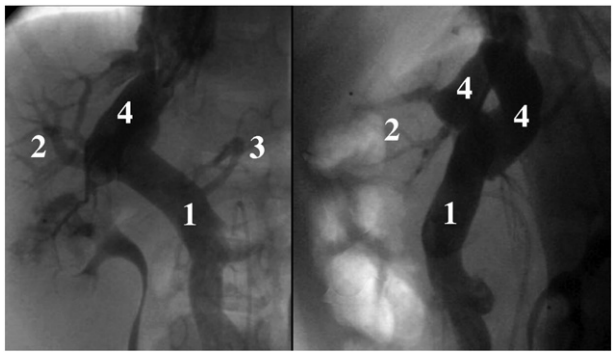

Fig. 1 Transjugular retrograde transfistula portography in a 6-year-old boy with a huge high-flow congenital intrahepatic portohepatic shunt: frontal and lateral views show a large portal vein trunk (1) flowing directly into the median hepatic vein through a large double portohepatic fistula (4). A hypoplastic right (2) and left (3) portal venous system is visible.

Download English Version:

https://daneshyari.com/en/article/4156162

Download Persian Version:

https://daneshyari.com/article/4156162

Daneshyari.com