Percutaneous Transhepatic Angioplasty for Portal Vein Cavernous Transformation after Choledochal Cyst Surgery

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Background: Cavernous transformation of the portal vein rarely occurs after a choledochal cyst surgery. Case characteristics: A 7-year-old boy with a history of a choledochal cyst surgery was admitted with recurrent oral and nasal bleeding over next two years. After excluding coagulopathies and hematopathies, we treated him with percutaneous transhepatic angioplasty. Outcome: The flow of the portal vein recovered immediately after balloon dilation. The patient’s symptoms were relieved, and no recurrence or complications occurred. Message: Stenosis and cavernous transformation of portal vein can be successfully managed by percutaneous transhepatic angioplasty.

Keywords: Complications, Hematomasis, Portal Hypertension.

Cavernous transformation of the portal vein (CTPV), known as portal cavernoma, is caused by stenosis or obstruction of the portal vein. This condition leads to spontaneous bypass across the stenosis, which sustains blood flow and liver function. Most cases of CTPV in children are caused by neonatal septicemia, and umbilical and intra-abdominal infections. However, CTPV has been rarely reported after choledochal cyst surgery. The traditional treatment for CTPV is surgery. We report a case of successful treatment of a CTPV by percutaneous transhepatic angioplasty.

CASE REPORT

A 7-year-old boy was admitted to our center with recurring oral and nasal bleeding. Physical examination revealed splenomegaly and an abdominal scar, and ultrasound examination showed extrahepatic portal vein stenosis (Fig. 1). The boy had a history of a choledochal cyst treated by Roux-en-Y hepatoenterostomy 6 years previously. The patient had no signs of fever or jaundice during the early follow-up period, and ultrasound examination findings were normal for 6 months after the operation. Over a period of next 4 years, his spleen size started increasing, platelet counts decreased, and he developed gastro-intestinal bleeding. Gastroscopy revealed moderately severe varicose veins in the gastric fundus and distal esophagus; barium meal examination showed signs of venous beading at the bottom of the esophagus, indicating a spontaneous portosystemic shunt (Fig. 1). Enhanced computed tomography (CT) indicated splenomegaly and the dilatation of the portal and splenic vein (Fig. 1). Color doppler ultrasound showed main portal vein diameter from 4.4 to 7.8 mm and a flow velocity of 11.0 cm/s. The diameter of the right portal, left portal, and splenic vein were 7.2, 5.0, and 9.3 mm, respectively. Three-dimensional CT reconstruction provided direct visualization of the severe stenosis of the main portal vein.

We treated the patient with percutaneous transhepatic angioplasty. The right portal vein was successfully accessed under ultrasound monitoring, and a 5F sheath was subsequently inserted. A 4F elbowed catheter was

FIG. 1 Enhanced computed tomography showing severe stenosis of the main portal vein (white arrow) and dilation of the splenic vein and coronary vein (a); and Barium swallow showing venous beading at the bottom of the esophagus indicating a dilated venous shunt (b).
advanced upstream into the superior mesenteric vein with the help of a guide wire. Digital subtraction angiography confirmed the presence of portal vein stenosis and pericholecystic collaterals coursing towards the liver. After the main portal vein was dilated three times with a Cordis 10-40-mm balloon at 14 atm, it was recanalized and the collateral vessels disappeared as blood flowed into the liver again (Web Fig. 1).

The blood flow of the portal vein was immediately restored. The size of the spleen below the costal margin decreased from 5 to 2 cm within 5 days after the therapy, and the platelet count normalized. One month after the therapy, CT and ultrasound examinations showed that the stenosis had been dilated to 4.8 mm with a rising flow velocity of 62.5 cm/s in the patent portal vein. The stenosis continued to expand to 5.1 mm within 3 months after the therapy. After 6 months of warfarin administration, the portal vein remained patent over a follow-up period of 2 years.

**DISCUSSION**

Postoperative complications of choledochal cysts usually include cholangitis, biliary stone formation, anastomotic stricture formation, and malignancy. Portal vein stenosis and CTPV are rare. CTPV is characterized by the presence of collaterals in the vicinity of the occluded blood vessels. Conditions such as inflammation, tumor metastasis, regional compression parasite infestation, and chronic liver disease can lead to occlusion of the portal vein, which then leads to CTPV [2,3]. However, CTPV may also be found in association with extrahepatic bile duct stenosis by choledochal varices. Jaundice and fever are common clinical manifestations. The common bile duct is extraluminally compressed and laminated in such cases [1]. Pre-operative ultrasound examination and operative exploration excluded this condition in the present case (Fig. 1).

Choosing the optimal surgical approach in CTPV is challenging. Simple disconnection may not effectively reduce the pressure, with chances of regeneration of collateral circulation and rebleeding [4]. Creation of a distal splenorenal shunt is the most commonly performed selective portal decompression procedure in children, but exposure of the shunt during the portacaval procedure is difficult. The minimum diameter required for splenic vein anastomosis is 6 mm. A superior mesenteric vein to intrahepatic left portal vein (Rex) shunt was recently used to bridge the thrombosis after liver transplantation, but the vascular condition in present case was highly complex [5]. Transjugular intrahepatic portosystemic shunt (TIPS) reduces the portal vein pressure gradient to clinically insignificant levels in the short term, but frequent shunt dysfunction and encephalopathy have precluded it from being the first-line treatment. Real-time ultrasound-guided percutaneous transhepatic inter-venital therapy has the advantages of minimal invasion, high efficiency, and high reproducibility [6]. It is widely applied in treatment of portal hypertension, tumor compression, tumor embolus, and tumor thrombus.

Interventional therapy for post-operative portal vein occlusion has been used after liver transplantation [7]. Interventional therapy has now become the first-line treatment for vascular complications after liver transplantation. Thrombolysis in the infrahepatic portal vein and angioplasty in the extrahaepatic portal vein through a splenic vein route are often successfully performed [8].

In conclusion, interventional therapy for portal vein stenosis and CTPV following intra-abdominal surgeries may be an effective therapeutic option.

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**REFERENCES**

WEB FIG. 1 Digital subtraction angiography confirming portal vein stenosis and pericholecystic collaterals coursing towards the liver (a); Venography after recanalization of the portal vein (b).