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Case Report

Transjugular intrahepatic portosystemic shunt for pyrrolidine-alkaloid-induced hepatic sinusoidal obstruction syndrome in a patient with situs inversus totalis and portal vein thrombus

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A B S T R A C T

We here report a rare case of pyrrolidine-alkaloid-induced hepatic sinusoidal obstruction syndrome (PA-HSOS) complicated with situs inversus totalis and portal vein thrombus (PVT). The patient had abdominal distention and ascites, which were not responsive to conservative medical treatment or peritoneal drainage. Transjugular intrahepatic portosystemic shunt (TIPS) was successfully performed in the patient via the left internal jugular vein approach. After the procedure, abdominal distension and ascites gradually resolved. The TIPS procedure could be considered as a useful treatment for PA-HSOS patients with PVT.

Keywords: Case reports; Hepatic veno-occlusive disease; Portal vein; Portasystemic shunt, transjugular intrahepatic; Situs inversus

Introduction

Hepatic sinusoidal obstruction syndrome (HSOS) is a hepatic vascular disease often caused by oral intake of herbal medicines containing pyrrolizidine alkaloids in China, such as Tusanqi (Gynura segetum), and it is classified as pyrrolidine-alkaloid-induced HSOS (PA-HSOS).[1–5] The use of transjugular intrahepatic portosystemic shunt (TIPS) in the treatment of PA-HSOS has been reported in the literature, and it is considered an effective treatment for patients whose portal hypertension and related complications do not respond to medical treatment.[2,4–6] However, there is no report of any PA-HSOS case with both situs inversus totalis (SIT) and portal vein thrombus (PVT) at the same time. SIT is a rare congenital anatomic variant with an incidence of 0.005%–0.02%.[7] Here, we describe a case of a successful TIPS creation in a SIT and PVT patient diagnosed with PA-HSOS caused by Tusanqi.

Case Report

A 67-year-old woman with a one-month history of ingesting Tusanqi had experienced abdominal distention and ascites for five months. These symptoms did not respond to conservative medical treatment or peritoneal drainage performed at the local hospital. The patient was transferred to our hospital for further treatment to relieve symptoms and improve liver function. Imaging examination revealed SIT, portal vein thrombosis, hepatomegaly, liver congestion, and thrombus in the superior mesenteric vein and splenic vein (Fig. 1). The laboratory tests were as follows: total serum bilirubin 57.3 μmol/L, alanine aminotransferase 64.2 U/L, aspartate aminotransferase 83.8 U/L, gamma-glutamyl transferase 77 U/L, serum albumin 28.0 g/L, white cell count 10.0 × 10^9/L, platelet count 82 × 10^9/L, INR 1.52, D-dimer 2.71 mg/L, prothrombin time 17.10 seconds, activated partial thromboplastin time 36.80 seconds, and Child-Pugh score 10, MELD score 23. Considering the history and imaging and laboratory examination,

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we diagnosed her with PA-HSOS caused by Tusanqi intake, portal vein thrombosis, and SIT.

After ruling out relevant contraindications, we created the TIPS via the left internal jugular vein. The procedure was carried out as a mirror image of the routine operation. The middle hepatic vein was initially catheterized as a marker. Indirect portal venography via the superior mesenteric artery showed the defect of portal vein, and intrahepatic portal vein branches and collateral compensation (Fig. 2A). The special puncture steel needle sheath tube system (RUPS-100; COOK, Bloomington, IN, USA) was used to puncture the confluence of the left and right branches of the portal vein under fluoroscopy via the middle hepatic vein. Direct portal venography by a pigtail catheter (Cordis, Milpitas, CA, USA) showed extensive thrombosis of the superior mesenteric vein and the left and right branches of portal vein. And the portal vein pressure measurement was 28 mmHg. We used an 8-Fr guiding catheter (Cordis) and a 50 mL syringe to aspirate thrombus several times. Then a 7 mm × 8 cm balloon catheter (Armada; Abbott Vascular Inc., Santa Clara, CA, USA) was used to dilate the puncture channel. A double stent was implanted for shunting (8 mm × 5 cm covered stent [Viabahn, W.L. Gore & Associates, Flagstaff, AZ, USA] combined with 8 mm × 8 cm bare stent [Cordis]). The final portal venography showed a little blood flow into the right and left branches of the portal vein and a reduced filling defect. And the portal vein pressure measurement was 17 mmHg. Finally, the pigtail catheter (Cordis) was retained for continuous thrombolysis (Urokinase, 400,000 U/24 hours, 48 hours, 2 days) (Fig. 2B). Three days later, repeat portal venography showed that there were still some filling defects from the trunk of the portal vein to the proximal part of superior mesenteric vein. We repeatedly aspirated a small amount of thrombus and implanted an 8 mm × 8 cm bare stent (Cordis) to the proximal part of the superior mesenteric vein. Final portal venography showed the stent position was good and the TIPS shunt was patent (Fig. 2C).

Follow-up computed tomography (CT) and ultrasonography performed three days after the TIPS procedure confirmed that the stent was patent and the liver congestion was improved (Fig. 3A, 3B). The laboratory tests were as follows: total serum bilirubin 73.6 μmol/L, alanine aminotransferase 27.0 U/L, aspartate aminotransferase 37.5 U/L, Child-Pugh score 9, MELD score 14. The symptoms of abdominal distension and ascites gradually resolved. The patient was discharged on postoperative day 6. The one- and three-month follow-up ultrasonography confirmed the patency of blood flow (Fig. 3C, 3D). The three-month follow-up laboratory tests were as follows: total serum bilirubin 60.4 μmol/L, alanine aminotransferase 20.3 U/L, aspartate aminotransferase 40.4 U/L, Child-Pugh score 7, MELD score 13. The 6-, 12-, and 18-month telephone follow-up results showed that the patient was generally in good condition.

This study was approved by the Ethics Committee of our

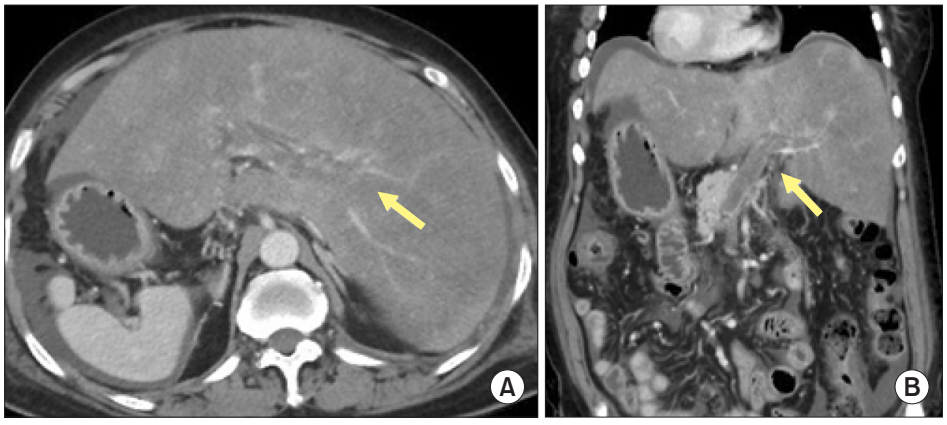

Fig. 1. Computed tomography showing situs inversus totalis, diffuse hepatomegaly, liver congestion, and thrombus in the branches of the portal vein (arrow) (A) and its main trunk (arrow) (B).

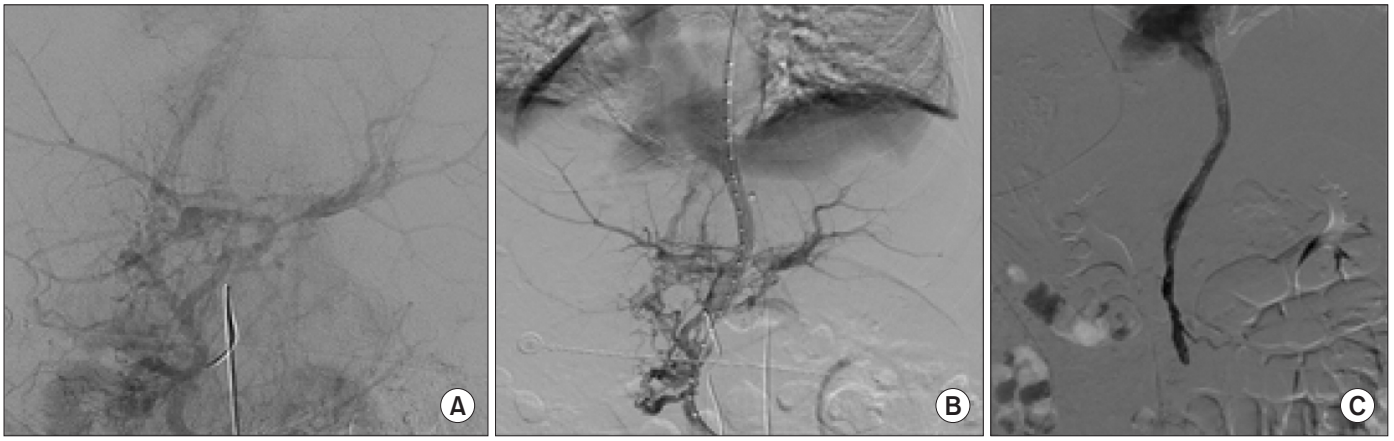

Fig. 2. (A) Indirect portal venography showing the significant filling defect of portal vein branches, and the main trunk of the portal vein is not shown. (B) Portal venography showing the recanalization of the main trunk of the portal vein after TIPS. (C) Final portal venography showing the recanalization of the superior mesenteric vein and the loss of the collateral vessels.

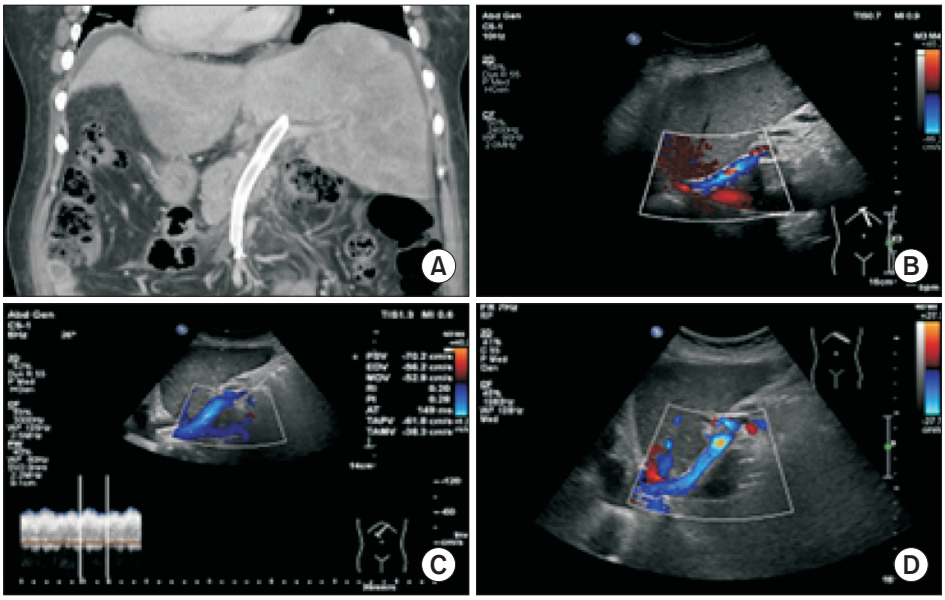

Fig. 3. (A) Computed tomography scan and ultrasonography showing the patency of blood flow in the stent and significant improvement in portal perfusion over with the image taken before the procedure. The flow rate in the stent observed three days (B), one month (C), and three months (D) after the procedure taken by ultrasonography were 171 cm/s, 70 cm/s, and 93 cm/s.

hospital and was in accordance with the ethical standards of the 1964 Helsinki declaration and its later amendments.

Discussion

HSOS is caused by injury to the sinusoidal endothelial cells and subendothelial edema. HSOS present with abdominal distension, pain in the hepatic region, ascites, jaundice, and hepatomegaly. Intake of pyrrolizidine alkaloids (PAs) is the major etiology of HSOS in China.[1–5] In 2017, the Hepatobiliary Diseases Committee of the Chinese Society of Gastroenterology developed the "Nanjing criteria" to diagnose and treat PA-induced HSOS.[5] Based on the Nanjing criteria, we diagnosed her as PA-HSOS using her medical history, clinical manifestations, laboratory tests, and imaging results. The expert consensus recommends anticoagulation-TIPS ladder treatment as the therapeutic strategy for PA-HSOS patients.[5] When the effect of symptomatic supportive treatment and anticoagulant therapy is poor, TIPS could improve the prognosis of PA-HSOS patients.[8,9] Given the failure of the conservative medical treatment administered at the local hospital and the detectable peritoneal drainage, we finally chose TIPS to reduce ascites and save the liver.

The special feature of this case is that the patient had both SIT and PVT, which was diagnosed by enhanced abdominal CT. SIT is defined as the complete mirror-image transposition of the thoracic and abdominal viscera.[10–12] TIPS treatment on SIT patients has only rarely been reported in the literature. Because of their distinctive anatomical structure, the right internal jugular vein should not be used as a standard access for the TIPS procedure. Thus, we chose the left jugular vein.

It is very uncommon for PVT is to place a large burden on patients with HSOS. As the patient had no history of liver cirrhosis, the thrombus formation was thought to be associated with the portal hypertension caused by the HSOS. While this is not a contraindication to TIPS, it poses a significant technical challenge to the necessary puncture and recanalization of the portal venous system.[13] Given this challenge, we placed the catheter in the hepatic artery as a marker to guide the puncture tool, and we performed portal vein thrombectomy, thrombolysis, and stenting to restore the patency of the portal venous system and reduce portal pressure.

In conclusion, PA-HSOS with SIT and PVT is very rare. TIPS was effective in the management of this difficult and unusual case.

Funding

None.

Conflicts of Interest

No potential conflict of interest relevant to this article was reported.

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