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Case Report

Ectopic duodenal variceal bleed successfully treated with TIPS and 2 years follow-up: A Case Report ☆

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ABSTRACT

Duodenal varices are an uncommon presentation of portal hypertension and can result in significant gastrointestinal bleeding with a high mortality. Diagnosis can be difficult and therapeutic options limited. We present a case of upper gastrointestinal bleeding in a woman aged 54 years with primary biliary cholangitis who was ultimately diagnosed with ectopic duodenal variceal bleed, which was successfully treated with transjugular intrahepatic portosystemic shunt. Transjugular intrahepatic portosystemic shunt provide an effective treatment for ectopic duodenal variceal bleed caused by liver cirrhosis, though interventional radiologists need to remain aware of and vigilant for the complications and risks of this treatment option.

Introduction

Portal hypertension is the progressive complication of liver cirrhosis, and gives rise to the development of portosystemic collaterals commonly at the oesophagogastric junction, the abdominal wall and the rectum [1]. Ectopic varicose veins refer to varicose veins with portal hypertension different with the esophageal and gastric varices, which may exist alone or with varices in other parts [2]. About 17% of ectopic varicose veins occur in the duodenum, and it can occur in any part of the duodenum [2]. The most common site of duodenal varices is the duodenal bulb, followed by the descending part of duodenum [3]. Hemorrhage caused by ectopic varicose vein accounts for about 5% of portal hypertension hemorrhage, but the mortality rate can reach 40% [2,4]. The current clinical treatment methods for bleeding from duodenal varicose veins include: medical drug treatment, surgical treatment, endoscopic interventional treatment (endoscopic ligation or sclerotherapy), and interventional embolization [5–6]. However, there are no

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standard guidelines for treatment of ectopic duodenal variceal bleed. We report a case of ectopic duodenal variceal bleed treated successfully with transjugular intrahepatic portosystemic shunt (TIPS) combined with embolization of varicose veins.

Case Report

A 54-year-old woman presented with a 2-week history of melaena, abdominal distension, fatigue, and without abdominal pain, diarrhea. She had no history of hepatitis, drinking, nonsteroidal medications and gastrointestinal bleeding. There was no significant family history. She received fluid rehydration treatment in Fugu county hospital 2 weeks ago, and ultrasound scan indicated a cirrhotic liver.Physical examination revealed chronic disease face and liver palms. The liver could not be palpated and the spleen was enlarged with its lower edge 3cm below the left costal margin. Shifting dullness was negative. There was slight edema over both lower extremities.Blood routine examination diaplayed hemoglobin of 88 g/L, leukocytes 2.0×10^9/L and platelet 81×10^9/L. Serum biochemistry revealed creatinine 65 umol/L, blood ureanitrogen 3.49 umol/L, albumin 32.9 g/L, total protein 59.9 g/L, aspartate aminotransferase 18 U/L, alanine aminotransferase 24 U/L, alkaline phosphatase 167 U/L, γ-glutamine transaminase 163 U/L, total bilirubin 18.4umol/L and international normalized ratio 1.35. Anti-hepatitis B virus antibody and anti-hepatitis C virus antibody were all seronegative. Autoimmunity liver antibodies included antinuclear antibody, antimitochondrial antibody (AMA), AMA-M2 and AMA-M2-3E (BPO) were all seropositive. Esophagastroduodenoscopy (EGD) showed: 1. Three nodular varicose veins, about 0.4 to 0.6 cm in diameter, can be seen at a level of 28 cm from the incisors; 2. One varicose vein can be seen at the fundus of stomach; 3. Multiple varicose veins can be seen in the descending part of duodenum (Fig. 1A). Computed tomography vein (CTV) indicated that the tributaries of superior mesenteric vein circle around the duodenum and merge into the right renal vein (Fig. 2A). In conclusion, she was diagnosed with duodenal variceal bleed, primary biliary cirrhosis and Child-Pugh grade B (7 points).

In order to prevent rebleeding, TIPS and venous embolization were performed with the consent of the patient and her families. After the complement of relevant preopreative examinations, Digital Substraction Angiography (DSA, Philips H5000, Netherlands)-guided TIPS was performed using a 0.035-inch guide wire (TERUMO, Japan). RUPS-100 (COOK Medical, USA) was sent to the liver along the guide wire and punctured to the left branch of the portal vein through hepatic parenchyma. According to the radiography (Fig. 3A & B), a large tortuous ectopic varicose vein, which was issued from the superior mesenteric vein and merged into the right renal vein was embolized with a releasable spring coil (Interlock, Boston Scientific Corporation, USA) (Fig. 3C); a tortuously expanded gastric coronary vein, was embolized with tissue gel (FAL, China) (Fig. 3D). Then, a shunt was established with a cover stent (8mm, BARD, USA) combined with a bare stent (8mm, BARD, USA), and the duodenal varices and gastric coronary veins were disappeared (Fig. 3D). Hepatic vein pressure gradient (HVPG) was decreased from 14 to 7 mmHg.

The patient was followed at 1, 3, and 6 months and every 6 months thereafter. Medical history, physical examination, biochemistry, haematological tests, and abdominal ultrasound were recorded. During the follow-up period, no adverse events occurred including bleeding, hepatic encephalopathy and ascites. Meanwhile, ultrasound showed blood flow smoothly in the TIPS stent. EGD at 1- and 6-month follow-up (Fig. 1B and 1C) and CTV of the portal vein at 24- month follow-up (Fig. 2B) showed that the duodenal varix had completely resolved.

Discussion

Duodenal varicose veins are mainly caused by portal hypertension [7]. Causes of portal hypertension can be prehepatic (such as portal vein thrombosis), hepatic (commonly liver cirrhosis and hepatic fibrosis) or posthepatic (such as hepatic vein obstruction) [8]. Two-thirds of duodenal varices are a result of portal vein hypertension due to hepatic cirrhosis [9]. Duodenal varices can result in massive gastrointestinal bleeding, reported at around 40% mortality; diagnosis and treatment are often difficult and controversial as experience is limited [2,4]. The therapies have been reported to be efficacious for duodenal variceal bleed, including endoscopic variceal ligation (EVL), endoscopic injection sclerotherapy (EIS) and TIPS [5,10–12].

EIS and EVL are the major endoscopic interventions for duodenal variceal bleed [13]. EIS is the treatment of acute variceal bleeding and prevention of rebleeding by endoscopic injection of sclerosing agent [14]. However, the veins passing through the duodenum are tributaries or common tributaries of the portal vein and the retroperitoneal vena cava, some of which can flow into the inferior vena cava or right renal vein [15]. The injection of sclerosing agent to the inferior vena cava or right renal vein may cause ectopic embolization [16]. EVL is the treatment of variceal hemorrhage by endoscopic loop ligature [17]. However, EVL could hardly acquire enough operative vision and accessibility for the treatment of duodenal varices bleed [17]. Simultaneously, endoscopic treatment including EIS and EVL cannot completely eliminate the varices, and it is easy to cause ulcer, perforation or rebleeding, as ectopic duodenal varices being located mainly in the serosal layer of weak intestinal wall [18]. Therefore, the potential risks and complications of EIS and EVL treatment for duodenal varices bleed should be fully considered.

TIPS is considered the first-line treatment for refractory variceal bleeding, with many studies reporting favorable outcomes in patients with duodenal varices bleed [6,19–20]. Tyler House et al. reported a case of massive duodenal variceal hemorrhage and ultimately required TIPS with coil embolization for control of bleeding. Postoperative review Model for end-stage liver disease scores improved significantly without any encephalopathy [19]. Rohit Anand et al. reported that a 43-year-old man diagnosed with duodenal varices bleed. Angiography suggested a large and competing nonphysiological shunt was found between the left mesenteric vein and the left renal vein. TIPS stent was performed to embolize completely

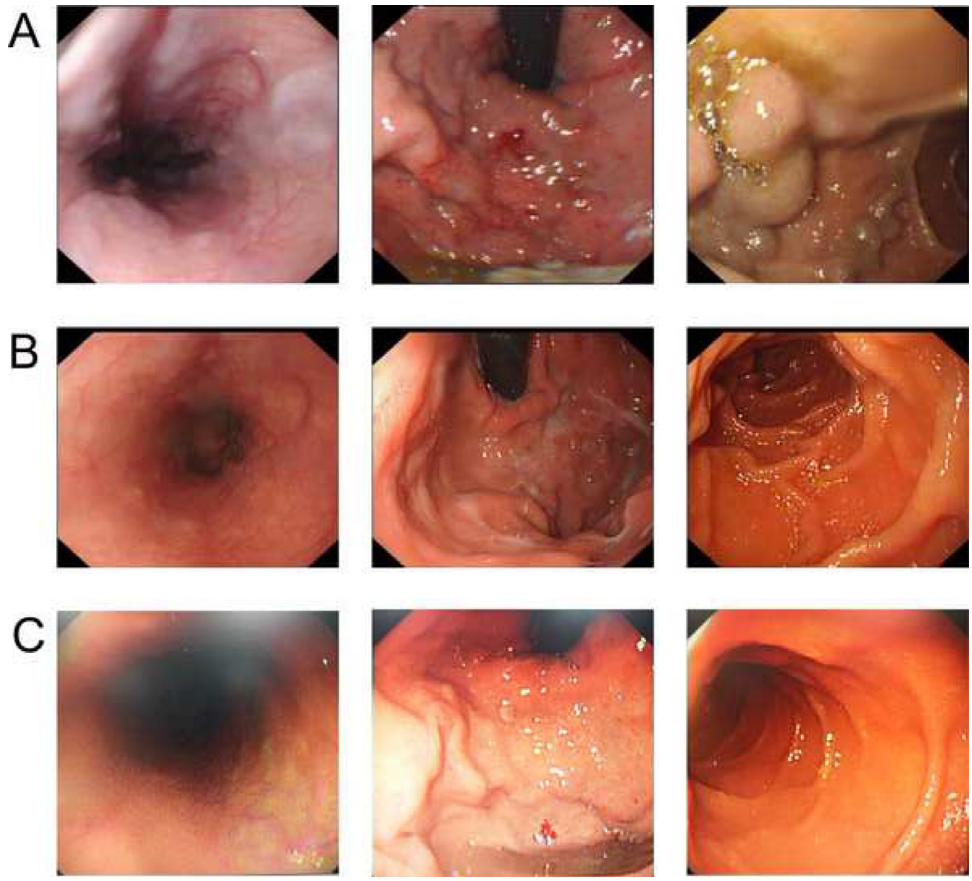

Fig. 1 – The images of EGD in esophagus, fundus of stomach and duodenum. A: **Preoperative EGD showed esophageal varices, gastric varices (severe) and descending duodenal varices (severe).** B: **EGD at 1-month follow-up showed that mild esophageal varices existed, gastric varices and descending duodenal varices disappeared.** C: **EGD at 6-month follow-up showed that mild gastric varices appeared.**

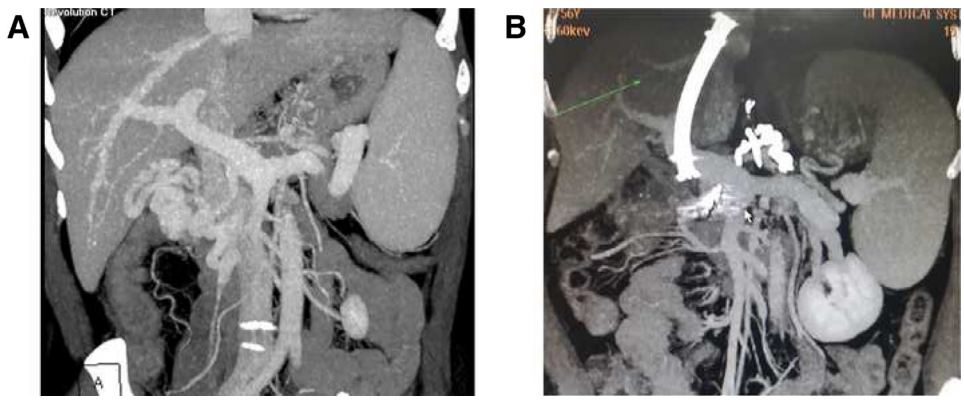

Fig. 2 – The images of CTV. A: **Preoperative CTV indicated: (1) cirrhosis and portal hypertension; (2) esophageal and gastric varices; (3) the left gastric vein flow into the splenic vein and the tributaries of superior mesenteric veins flow into the right renal vein; (4) splenomegaly.** B: **CTV at 24-month follow-up indicated: (1) cirrhosis and portal hypertension; (2) the embolization of esophageal and gastric varices; (3) the smooth blood flow in the stent and the metal image of spring coil; (4) splenomegaly.**

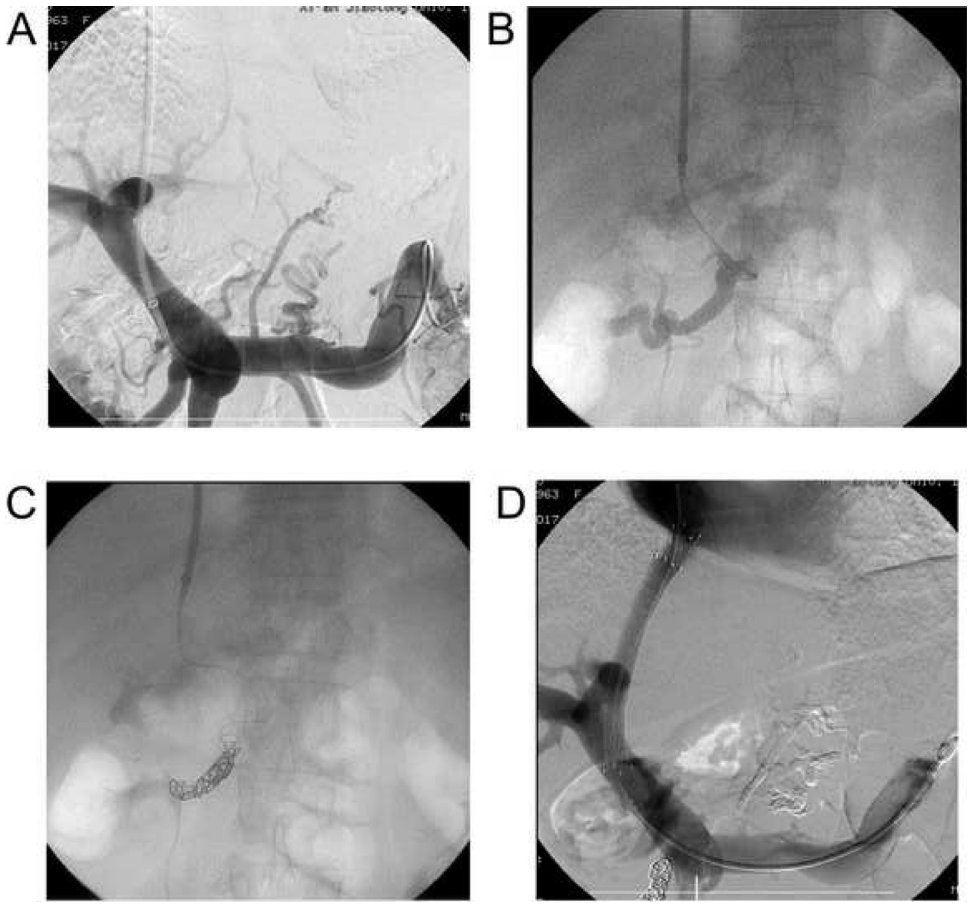

Fig. 3 – The images of angiography. A & B: **(1) the intrahepatic portosystemic shunt puncture was successful; (2) a large tortuous ectopic varicose vein was issued from the superior mesenteric vein and merged into the right renal vein; (3) a tortuously expanded gastric coronary vein was issued from the spleen vein.** C: **the duodenal variceal was embolized with a releasable spring coil.** D: **the blood flow in stent is smooth and the varicose vein was disappeared.**

the nonphysiological shunt, and no upper gastrointestinal bleeding occurred [20]. Our case presented a woman diagnosed with duodenal variceal bleed and CTV revealed that duodenal veins were merged into the right renal vein. EIS and EVL had been tried in multiple studies for bleeding ectopic varices with some degree of success. However, in our case, EIS or EVL may fail to control the bleeding and avoid the severe complications. Therefore, we considered TIPS combined with varices embolization as the therapies of the patient. Angiography suggested that there is a nonphysiological shunt and the releasable spring coil was used to avoid the ectopic embolization. The stents were used to establish a shunt and reduce the HVPG, which results in portal venous decompression and variceal bleeding control. After TIPS, the HVPG of the patient was decreased to 7 mmHg and no serious complications including rebleeding, hepatic encephalopathy and ascites occurred during 2 years follow-up. At present, we continue to follow up the patient regularly.

In conclusion, the difficult diagnosis and nonstandard treatment of duodenal varices bleed result in a low detection rate and high mortality. Therefore, we recommend improving gastroscopy and adding the portal venous angiography to assess the severity degree of varices and the presence of ectopic varices, for the patients with cirrhosis and upper gastrointestinal bleeding. In terms of treatment, we also recommend TIPS combined with varicose vein embolization as the first choice, for embolizing nonphysiological shunts, reducing portal pressure gradients, and improving liver perfusion. However, as high operating difficulty and expensive cost of TIPS, we should comprehensively assess the conditions of patients to choose the therapies.

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