【 CASE REPORT 】

Intractable Hepatic Encephalopathy with a Large Portosystemic Shunt Successfully Treated Using Shunt-preserving Disconnection of the Portal and Systemic Circulation

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Abstract:
Hepatic encephalopathy (HE) is a significant symptom of decompensated liver cirrhosis. Occlusion of portosystemic shunts is used to treat refractory HE. Nevertheless, these treatments often cause adverse events, such as ascites and esophageal varices. We treated a 57-year-old man with refractory HE using shunt-preserving disconnection of the portal and systemic circulation (SPDPS). After SPDPS, there were no obvious complications, and the patient's ammonia level significantly decreased. To date, the patient has not experienced recurrent HE. SPDPS appears to be a safe and effective treatment method for portosystemic encephalopathy.

Key words: shunt-preserving disconnection of the portal and systemic circulation, hepatic encephalopathy, percutaneous transsplenic access

(Intern Med 59: 1047-1051, 2020)
(DOI: 10.2169/internalmedicine.3955-19)

Introduction

Hepatic encephalopathy (HE) is a significant symptom of decompensated liver cirrhosis. Hepatic encephalopathy due to a portosystemic shunt (PSS) is called portosystemic encephalopathy (1). In cases in which there is a poor response to treatments such as lactulose or rifaximin, interventional therapy for shunts is required. Treatments for large PSSs, such as balloon-occluded retrograde transvenous obliteration (B-RTO), are useful for patients with refractory HE. Nevertheless, these treatments often lead to ascites and/or the formation of esophageal varices (2-7). Kashida et al. reported that shunt-preserving disconnection of the portal and systemic circulation (SPDPS) was effective in patients with portosystemic encephalopathy without the development of these complications (8). Generally, SPDPS is performed using the transcutaneous transhepatic approach, while the transcutaneous transsplenic approach is rarely used. We report a case in which refractory HE was successfully treated using selective embolization of the splenic vein for SPDPS via the transcutaneous transsplenic approach.

Case Report

The patient was a 57-year-old man with hepatitis-B-related cirrhosis who had been hospitalized several times for multiple episodes of HE (West Haven Criteria: grades 3-4) between May-July 2018. There was no trigger factor for most of these episodes. His relevant medical history included: hepatocellular carcinoma (HCC), which was treated with central bisegmentectomy of the liver at 47 years of age; recurrent HCC, which was treated with right posterior segmentectomy and repeated hepatic arterial infusion che-

motherapy at 50 years of age; and a refractory liver abscess, which was treated with transgastric drainage tube at 54 years of age (this drainage tube remained in place).

The patient's laboratory data during the third admission (Table) were as follows: platelet count, 7.6×10⁴/μL; prothrombin time-international normalized ratio (PT-INR),

Table. Laboratory Data on Admission.

Variables	Value	Unit
WBC	3,600	/μL
RBC	2.84	×10^6/μL
Hb	9.0	g/dL
PLT	7.6	×10^4/μL
PT-INR	1.9	mEq/L
TP	6.7	g/dL
Albumin	2.5	g/dL
T-bil	1.2	mg/dL
AST	44	IU/L
ALT	28	IU/L
ALP	638	IU/L
γGTP	70	IU/L
LDH	252	IU/L
BUN	13	mg/dL
Creatinine	1.21	mg/dL
Na	144	mEq/L
K	4.1	mEq/L
Cl	115	mEq/L
Ammonia	356	μg/dL
	Child-Pugh score; 10 points	
	MELD score; 16 points	

WBC: white blood cell, RBC: red blood cell, Hb: hemoglobin, PLT: platelet, PT-INR: prothrombin time-international normalized ratio, TP: total protein, T-bil: total-bilirubin, AST: aspartate aminotransferase, ALT: alanine aminotransferase, ALP: alkaline phosphatase, γGTP: γ-glutamyl transpeptidase, LDH: lactate dehydrogenase, BUN: blood urea nitrogen, MELD: Model for End-Stage Liver Disease

1.9; total bilirubin, 1.2 mg/dL; total protein, 6.7 g/dL; and serum albumin, 2.5 g/dL. He was positive for Hepatitis B virus antigens. His Model of End-Stage Liver Disease (MELD) score was 16, his Child-Pugh Grade was C (score 10), and his blood ammonia level was 356 μg/dL (normal range, 30-80 μg/dL). Three-phase contrast-enhanced computed tomography (CT) revealed the presence of a large splenorenal shunt and splenomegaly (Fig. 1). Esophagogastroduodenoscopy revealed thin esophageal and mild gastric varices.

During the periods of hospitalization, the patient received lactulose, branched-chain amino acids, and protein restriction. Nevertheless, these medications were not sufficient to prevent new episodes of HE and hepatic coma. Thus, treatment for repeated portosystemic encephalopathy was considered necessary, and we consulted with radiologists regarding the indications for an interventional radiological splenorenal shunt. We were concerned about the exacerbation of refractory ascites and esophageal varices that could occur following a B-RTO because the patient had a history of hepatectomy and a low hepatic reserve. Thus, SPDPS was performed (Fig. 2). First, we selected percutaneous transsplenic puncture because percutaneous transhepatic puncture of the patient's residual liver was difficult to perform. A long sheath and catheter were prophylactically placed in the splenic artery from the femoral artery to manage hemorrhage due to splenic puncture. Next, we inserted a catheter from the left intercostal space to the splenic vein under ultrasound guidance. We placed the catheter between the junction of the inferior mesenteric vein and the splenorenal shunt; then, an indwelling Amplatzer® vascular plug II (AV II, diameter 16 mm, St. Jude Medical, St Paul, USA) was placed in the same region. Additionally, n-butyl-2-cyanoacrylate (NBCA) (33%) was slowly injected into the same region using a microcatheter. Although slight flow to the portal vein was noted when contrast medium was injected from within the plug, the microcatheter was unable to pass beyond the plug. The splenorenal shunt was not visual-
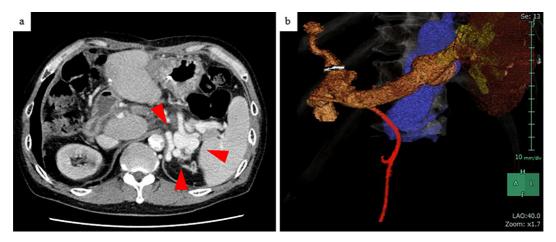

Figure 1. (a) Three-phase contrast-enhanced computed tomography (CT) in the late phase showing huge portosystemic shunt (arrowhead). (b) 3D-CT showing huge portosystemic shunt (blue: portosystemic shunt; brown: portal vein; red: superior mesenteric vein).
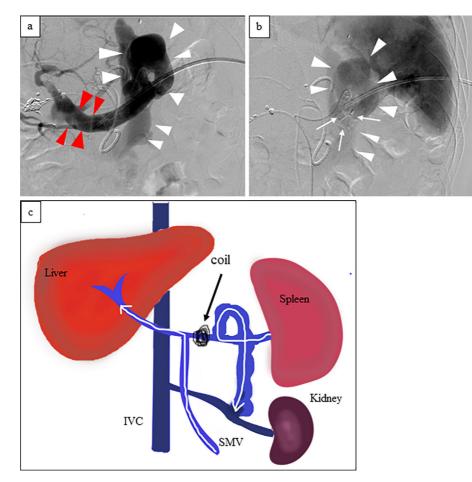
Figure 2. **(a) Before embolization, splenic venography reveals the portal vein (red arrowhead) and huge portosystemic shunt (white arrowhead). (b) After coil embolization (arrow), splenic venography reveals only a large portosystemic shunt (white arrowhead), without the portal vein. (c) A schematic illustration of the hemodynamics after selective embolization of the splenic vein for shunt-preserving disconnection of the portal and systemic circulation. Arrows show the direction of blood flow in the splenic vein and superior mesenteric vein.**

ized during superior mesenteric arterial portography; thus, we concluded that SPDPS was achieved. Finally, the splenic puncture point was filled with AV II and NBCA (33%) to complete the treatment.

After SPDPS, there were no obvious complications (e.g., increased ascites) and the ammonia level decreased from 356 μg/dL to 77 μg/dL (Fig. 3a). The postoperative course was uneventful, and the patient was discharged. One week after SPDPS, the patient underwent contrast-enhanced CT. This revealed an embolic agent in the AV II and slight reduction of the splenorenal shunt. To date, the patient's ammonia level has remained within normal levels, and HE did not recur during 12 months of follow-up. Furthermore, the patient's albumin and platelet levels improved for approximately 2 months after SPDPS (Fig. 3b). The patient provided his consent for the publication of this case report and associated clinical images.

Discussion

We reported a case in which SPDPS was effective for re-

peated HE. HE in patients with liver cirrhosis is classified into two types: end-stage coma and chronic recurrent coma (9); this patient's case was classified as the latter type. A large PSS is the most frequent cause of recurrent or persistent HE in patients with cirrhosis (10). In patients who fail to respond to treatments, such as those with lactulose or rifaximin, shunt occlusion should be promptly considered as the next option (7). In Asia, B-RTO is often chosen as the treatment for large shunts, which cause HE (11, 12). Nevertheless, this treatment has drawbacks, including increased portal vein pressure and the resulting risk of esophageal varices aggravation and the formation of ascites. In this case, it was considered that B-RTO would likely worsen the portal hypertension and cause the deterioration of the patient's general condition because only the left lateral segment of the liver remained after surgery. We therefore determined that SPDPS would involve fewer complications than B-RTO.

SPDPS was devised by Kashida et al. (8). The splenic vein is occluded on the hepatic side from the inflow portion of the splenorenal shunt, resulting in blockade of the out-
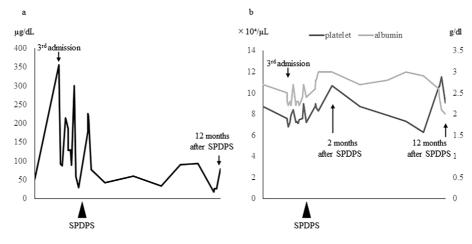
Figure 3. (a) The ammonia levels before and after shunt-preserving disconnection of the portal and systemic circulation (arrowhead). The ammonia levels significantly declined following the procedure. (b) The albumin and platelet levels after shunt-preserving disconnection of the portal and systemic circulation (arrowhead). The albumin and platelet levels improved for approximately 2 months.

flow of toxic substances (e.g., as ammonia) to the systemic circulation. Unlike B-RTO, the splenic vein drains from the splenorenal shunt into the systemic circulation, thereby preventing portal hypertension. When discussing the indications for SPDPS, shunt localization and the hepatic reserve should be considered. Shunts that flow from the splenic vein and which are more than a few centimeters distal to the superior and inferior mesenteric veins are adaptations of SPDPS, as they may allow embolization of the splenic vein. However, patients with end-stage cirrhosis might be ineligible for this treatment because therapeutic effects cannot be expected unless encephalopathy-inducing substances (e.g., ammonia) are metabolized in the hepatocytes.

In this patient, only the left lateral segment of the liver remained after surgery. It was difficult to perform portal vein puncture via the liver; thus, the transsplenic approach was selected. Regarding transsplenic approaches, Boulvin et al. (13) succeeded in performing percutaneous transsplenic portal angiography, and this has become a standard technique for portal angiography. Nevertheless, because the transsplenic approach involves a risk of splenic hemorrhage, it is difficult to consider it a standard technique. In the present case, we first placed a catheter in the splenic artery in preparation for bleeding. Further, the splenic puncture line was embolized with AV II and NBCA at the end of treatment. As a result, the treatment could be completed without splenic hemorrhage. There are no reports of SPDPS using a transsplenic approach; thus, this can be considered a rare case. The present study was associated with some limitations. We did not have data on the portal venous pressure, wedged hepatic venous pressure, or the hepatic venous pressure gradient before or after SPDPS. If we had shown the changes in blood pressure before and after SPDPS, we could have further suggested the usefulness of SPDPS.

In conclusion, our patient showed improvement in portosystemic encephalopathy without developing ascites or esophageal varices, and his blood ammonia level decreased during the follow-up period. SPDPS was an effective and safe method for treating portosystemic encephalopathy in our patient.

The authors state that they have no Conflict of Interest (COI).

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