

A large spontaneous intrahepatic portosystemic shunt in a cirrhotic patient

Xingshun Qi^{1,*}, Chun Ye², Yue Hou¹, Xiaozhong Guo^{1,*}

¹Liver Cirrhosis Study Group, Department of Gastroenterology, General Hospital of Shenyang Military Area, Shenyang, China;

²Department of General Surgery, General Hospital of Shenyang Military Area, Shenyang, China.

Summary A spontaneous portosystemic shunt is a rare malformation of the vessels supplying the liver. This condition often leads to the development of hepatic encephalopathy due to excessive shunting of blood from the portal vein to the inferior vena cava. Some studies have suggested that the presence of spontaneous portosystemic shunts is inversely associated with the appearance of large esophageal varices. Spontaneous intrahepatic portosystemic shunts (SIPSS) are far less frequently observed than extrahepatic portosystemic shunts, which include spleno-gastric-renal shunts, mesenteric-caval shunts, and a large patent umbilical vein. Reported here is a case of decompensated liver cirrhosis with a large SIPSS without any incidence of overt hepatic encephalopathy.

Keywords: Spontaneous intrahepatic portosystemic shunt; liver cirrhosis; hepatic encephalopathy; embolization

1. Introduction

A spontaneous portosystemic shunt is a rare malformation of the vessels supplying the liver that can lead to the development of hepatic encephalopathy (HE) due to excessive shunting of blood from the portal vein to the inferior vena cava (1-2). That said, a study has indicated that patients with spontaneous portosystemic shunts had significantly fewer large esophageal varices than those without such shunts (3). Color Doppler ultrasound and computed tomography are the first-line imaging modalities that are used to diagnose a spontaneous portosystemic shunt. Angiography should be the gold standard for diagnosis of those shunts. Depending on the location of the shunt, spontaneous portosystemic shunts primarily include extrahepatic and intrahepatic shunts. Spontaneous intrahepatic portosystemic shunts (SIPSS) are far less frequently observed than extrahepatic portosystemic shunts, which include spleno-gastric-renal shunts, mesenteric-caval shunts, and a large patent umbilical vein. Reported here

is a case of decompensated liver cirrhosis with a large SIPSS without any incidence of overt HE.

2. Case Report

On October 10, a 58-year-old male was admitted to this Department for intermittent pain in the left upper abdomen that had persisted for about 10 years. The man was diagnosed with hepatitis B virus (HBV) infection-related liver cirrhosis 10 years prior. The man also had a 20-year history of alcohol abuse (100 g daily) but had abstained from alcohol consumption for 10 years. The man denied any previous history of trauma.

On physical examination, there was mild tenderness in the left upper abdomen with no rebound, shifting dullness, and edema in both lower extremities. Laboratory results revealed a white blood cell count of $3.9 \times 10^9/L$ (reference range: $3.5-9.5 \times 10^9/L$), a red blood cell count of $4.30 \times 10^{12}/L$ (reference range: $4.3-5.8 \times 10^{12}/L$), a hemoglobin level of 136 g/L (reference range: 130-175 g/L), and a platelet count of $45 \times 10^9/L$ (reference range: $125-350 \times 10^9/L$). Blood was positive for HbsAg and HBcAb-IgG, HBV DNA was 9.2×10^5 copies/mL (reference range: $< 1.0 \times 10^3$ copies/mL), total bilirubin was 48.9 $\mu\text{mol}/L$ (reference range: 5.1-22.2 $\mu\text{mol}/L$), alanine aminotransferase was 66.04 U/L (reference range: 9-50 U/L), aspartate aminotransferase was 93.11 U/L (reference range: 15-40 U/L), albumin

*Address correspondence to:

Drs. Xiaozhong Guo and Xingshun Qi, Department of Gastroenterology, General Hospital of Shenyang Military Area, No. 83 Wenhua Road, Shenyang 110840, China.

E-mail: guo_xiao_zhong@126.com (XG), xingshunqi@126.com (XQ)

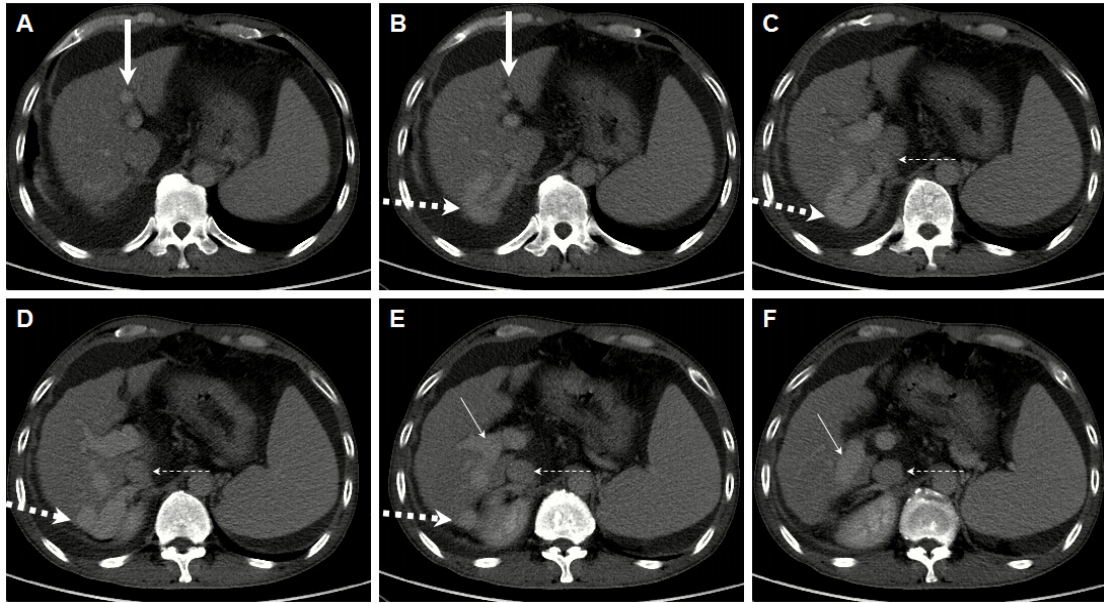


Figure 1. Abdominal axial contrast-enhanced computed tomography scans in the venous phase showing a spontaneous intrahepatic portosystemic shunt. Panels A-F are CT scans from top to bottom. The right portal branch (*fine solid arrow*) communicates with the inferior vena cava (*fine dashed arrow*) through a large portosystemic collateral vessel (*thick dashed arrow*). Notes: A thick solid arrow represents the left portal vein branch, a fine solid arrow represents the right portal vein branch, a thick dashed arrow represents the collateral vessel between the right portal vein branch and inferior vena cava, and a fine dashed arrow represents the inferior vena cava.

was 31.0 g/L (reference range: 40-55 g/L), prothrombin time was 19.3 seconds (reference range: 11.5-14.5 seconds), the international normalized ratio was 1.69, blood ammonia was 35 $\mu\text{mol/L}$ (reference range: 9-54 $\mu\text{mol/L}$), and alpha fetoprotein was 3.3 ng/mL (reference range: 0-6.7 ng/mL).

Contrast-enhanced computed tomography scans of the abdomen in the venous phase showed moderate ascites, splenomegaly, and right pleural effusion. Notably, there was a large collateral vessel between the right portal vein branch and the inferior vena cava (Figure 1). Thus, the patient was diagnosed with SIPSS, but no overt HE was observed. The presence of esophageal varices was confirmed by upper gastrointestinal endoscopy. Symptomatic treatment was given, including diuretics and hepatoprotective drugs. Abdominal pain gradually disappeared. On October 19, laboratory tests were performed again. Total bilirubin was 27.0 $\mu\text{mol/L}$, alanine aminotransferase was 44.97 U/L, aspartate aminotransferase was 68.02 U/L, albumin was 23.6 g/L, serum sodium was 137.2 mmol/L, and blood ammonia was 69 $\mu\text{mol/L}$. The patient was discharged and followed-up on an outpatient basis.

3. Discussion

Portal hypertension may be a major predisposing factor for SIPSS. Theoretically, the shunting of blood from the portal vein to the inferior vena cava may help to decrease portal pressure (2). In an experimental study, Geraghty *et al.* found that portal pressure and extent of portosystemic shunting were correlated a day after the

portal vein was partially ligated (4), but the correlation was absent in cirrhotic portal hypertension. However, SIPSS reduces the liver's ability to detoxify blood, thereby increasing the entry of toxic substances into the brain (2). In an early experimental study, Vogels *et al.* found that only rats with a portacaval shunt, rather than rats with a sham portacaval shunt, developed encephalopathy after hyperammonemia was induced with infusion of ammonium acetate (5). In contrast, Hsin *et al.* suggested that significant liver damage, rather than portosystemic shunts, would be required to induce the development of HE in cirrhotic rats (6). Despite the controversial findings from experimental studies, the clinical evidence appears to be unequivocal. A case-control study indicated that the prevalence of a large portosystemic shunt was significantly higher in cirrhotic patients with recurrent or persistent HE than in those without previous or present signs of overt HE (1). Notably, patients and the control group in that study were well matched in terms of age and the severity of liver dysfunction.

To the extent known, few studies have reported the presence of SIPSS in cirrhotic patients. More importantly, the patient in the current case was asymptomatic and did not receive embolization. Similarly, Tsauo *et al.* reported the presence of SIPSS in a patient with autoimmune hepatitis-related liver cirrhosis (7). Ding *et al.* also reported the presence of SIPSS in a patient with Budd-Chiari syndrome (8). In the two previous cases, no SIPSS-induced HE was observed. Thus, surgical or interventional vascular treatment of SIPSS was unnecessary.

Patients with symptomatic SIPSS should benefit from treatment. Recently, An *et al.* have shown that angiographic embolization of a spontaneous portosystemic shunt can improve the survival and liver function of cirrhotic patients with recurrent HE (9). Endovascular embolization of large spontaneous portosystemic shunts should be considered first (10-11). Several studies have recently described the Amplatzer device for embolization of large shunts. Kessler *et al.* used the Amplatzer Vascular Plug to embolize a large retroperitoneal shunt during a transjugular intrahepatic portosystemic shunt procedure (12). Ramirez-Polo *et al.* reported the use of the Amplatzer device to occlude an extrahepatic portosystemic shunt (*i.e.*, a porto-femoral "umbilical" shunt) in a patient with persistent HE (13). Wu *et al.* also reported the use of the Amplatzer device for embolization of a spontaneous splenorenal shunt in a cirrhotic patient presenting with HE after placement of a transjugular intrahepatic portosystemic shunt (14). Since an endovascular procedure is technically difficult, laparoscopic closure of SIPSS could be a treatment alternative (15).

Based on this unique case report, hepatologists should be mindful of the possibility of SIPSS in patients with liver cirrhosis. The decision to embolize SIPSS may depend primarily upon the presence of HE.

References

1. Riggio O, Efrati C, Catalano C, Pediconi F, Mecarelli O, Accornero N, Nicolao F, Angeloni S, Masini A, Ridola L, Attili AF, Merli M. High prevalence of spontaneous portal-systemic shunts in persistent hepatic encephalopathy: A case-control study. *Hepatology*. 2005; 42:1158-1165.
2. Chan CC. Portal-systemic collaterals and hepatic encephalopathy. *J Chin Med Assoc*. 2012; 75:1-2.
3. Tarantino G, Citro V, Conca P, Riccio A, Tarantino M, Capone D, Cirillo M, Lobello R, Iaccarino V. What are the implications of the spontaneous spleno-renal shunts in liver cirrhosis? *BMC Gastroenterol*. 2009; 9:89.
4. Geraghty JG, Angerson WJ, Carter DC. Portal venous pressure and portosystemic shunting in experimental portal hypertension. *Am J Physiol*. 1989; 257:G52-57.
5. Vogels BA, van Steynen B, Maas MA, Jorning GG, Chamuleau RA. The effects of ammonia and portal-systemic shunting on brain metabolism, neurotransmission and intracranial hypertension in hyperammonaemia-induced encephalopathy. *J Hepatol*. 1997; 26:387-395.
6. Hsin IF, Wang SS, Huang HC, Lee FY, Chan CY, Chang CC, Hsu CY, Lin HC, Lee SD. Portosystemic collaterals are not prerequisites for the development of hepatic encephalopathy in cirrhotic rats. *J Chin Med Assoc*. 2012; 75:3-9.
7. Tsauo J, Liu L, Tang C, Li X. Education and Imaging. Hepatobiliary and pancreatic: Spontaneous intrahepatic portosystemic shunt associated with cirrhosis. *J Gastroenterol Hepatol*. 2013; 28:904.
8. Ding PX, Li Z, Han XW, Zhang WG, Zhou PL, Wang ZG. Spontaneous intrahepatic portosystemic shunt in Budd-Chiari syndrome. *Ann Vasc Surg*. 2014; 28:742 e1-4.
9. An J, Kim KW, Han S, Lee J, Lim YS. Improvement in survival associated with embolisation of spontaneous portosystemic shunt in patients with recurrent hepatic encephalopathy. *Aliment Pharmacol Ther*. 2014; 39:1418-1426.
10. Laleman W, Simon-Talero M, Maleux G, *et al.* Embolization of large spontaneous portosystemic shunts for refractory hepatic encephalopathy: A multicenter survey on safety and efficacy. *Hepatology*. 2013; 57:2448-57.
11. Gopalakrishna R, Hurkadli PS, Puthukudy NK, Nair HR. Embolization of portosystemic shunt for treatment of recurrent hepatic encephalopathy. *J Clin Exp Hepatol*. 2014; 4:60-62.
12. Kessler J, Trerotola SO. Use of the Amplatzer Vascular Plug for embolization of a large retroperitoneal shunt during transjugular intrahepatic portosystemic shunt creation for gastric variceal bleeding. *J Vasc Interv Radiol*. 2006; 17:135-140.
13. Ramirez-Polo A, Márquez-Guillén E, González-Aguirre AJ, Casanova-Sánchez IE, Chávez-Ruiz R, Carrillo-Maravilla E, López-Méndez E. Persistent hepatic encephalopathy secondary to portosystemic shunt occluded with Amplatzer device. *Ann Hepatol*. 2014; 13:456-460.
14. Wu W, He C, Han G. Embolization of spontaneous splenorenal shunt for after-TIPS hepatic encephalopathy in a patient with cirrhosis and variceal bleeding. *Hepatology*. 2015; 61:1761-1762.
15. Kwon JN, Jeon YS, Cho SG, Lee KY, Hong KC. Spontaneous intrahepatic portosystemic shunt managed by laparoscopic hepatic vein closure. *J Minim Access Surg*. 2014; 10:207-209.

(Received January 6, 2016; Revised January 25, 2016; Accepted January 29, 2016)