



A large spontaneous extrahepatic portosystemic shunt in a cirrhotic patient: from main portal vein to right renal vein

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Abstract: In this paper, we illustrated a very rare type of spontaneous portosystemic shunt originating from main portal vein to right renal vein in a cirrhotic patient. The diagnosis of such a large spontaneous portosystemic shunt was established according to the axial contrast-enhanced computed tomography scans. The cause and outcome were also discussed.

Keywords: Spontaneous portosystemic shunt; liver cirrhosis; hepatic encephalopathy (HE)

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Introduction

Spontaneous portosystemic collateral vessels are frequently observed in liver cirrhosis with portal hypertension. Common types of spontaneous portosystemic collateral vessels in these patients include gastro-oesophageal varices, para-oesophageal veins, gastro-renal or splenorenal shunts, and para-umbilical veins (1). In this case report, we illustrated a very rare type of spontaneous portosystemic shunt originating from the main portal vein to right renal vein.

Case presentation

On January 20, 2014, a 59-year-old male was admitted to our department due to recurrent abdominal distension and weakness for about 5 years and intermittent fever for about one month. He had a 5-year history of alcoholic liver cirrhosis and a 20-year history of diabetes. He drunk white wine with a dose of 50 g per day for more than 30 years. At his admission, the temperature was 38.5 °C, heart rate was 118 b.p.m., and blood pressure was 143/58 mmHg. On physical examinations, there was mild edema of both lower limbs. On laboratory tests, hepatitis B virus surface antigen and hepatitis C virus antibody were negative, alpha

fetoprotein (AFP) level was 2.66 IU/mL (0–6.7 IU/mL), white blood cell (WBC) was $5.6 \times 10^9/L$ (4×10^9 – $10 \times 10^9/L$), percentage of neutrophil was 85.8% (50–70%), hemoglobin concentration was 86 g/L (110–170 g/L), platelet count (PLT) was $162 \times 10^9/L$ (100×10^9 – $300 \times 10^9/L$), total bilirubin (TBIL) was 17.5 $\mu\text{mol/L}$ (0–20.5 $\mu\text{mol/L}$), alanine aminotransferase (ALT) was 19 U/L (9–72 U/L), aspartate aminotransferase (AST) was 32 U/L (8–50 U/L), alkaline phosphatase (ALP) was 95 U/L (38–126 U/L), albumin was 25.2 g/L (37–53 g/L), serum creatinine was 59.6 $\mu\text{mol/L}$ (42–133 $\mu\text{mol/L}$), serum sodium was 125.5 mmol/L (130–150 mmol/L), prothrombin time (PT) was 15.5 seconds (11.5–14.5 seconds), and international normalized ratio (INR) was 1.23. On axial contrast-enhanced computed tomography scans, liver surface was irregular, spleen was remarkably enlarged, and a large tortuous collateral vessel was communicated between main portal vein and right renal vein (*Figure 1*). After a written informed consent from this patient and his relatives was obtained, an upper gastrointestinal endoscopy was performed, which demonstrated mild esophageal varices and portal hypertensive gastropathy. After intravenous infusion of ceftriaxone and hepatoprotective drugs were given for



Figure 1 A large spontaneous extrahepatic portosystemic shunt shown at contrast-enhanced computed tomography scans. Solid white arrows refer to a large collateral vessel. Dashed white arrows refer to main portal vein. Solid black arrows refer to inferior vena cava. Dashed black arrows refer to right renal vein. White arrowhead refers to a point of junction between main portal vein and collateral vessel. Black arrowhead refers to a point of junction between right renal vein and collateral vessel.

5 days, he was discharged.

On May 14, 2014, he was re-admitted to our department due to recurrent fever and unconsciousness for 2 days. At this admission, the temperature was 38.8 °C, heart rate was 80 b.p.m., and blood pressure was 179/80 mmHg. On laboratory tests, blood ammonia was 73 $\mu\text{mol/L}$ (9–54 $\mu\text{mol/L}$), WBC was $7.5 \times 10^9/\text{L}$, percentage of neutrophil was 89.2%, hemoglobin concentration was 90 g/L, PLT was $99 \times 10^9/\text{L}$, TBIL was 27.8 $\mu\text{mol/L}$, ALT was 16 U/L, AST was 22 U/L, ALP was 63 U/L, albumin was 28.5 g/L, serum creatinine was 91 $\mu\text{mol/L}$, serum sodium was 123.6 mmol/L, PT

was 16.3 seconds, INR was 1.31, and AFP was 3.37 IU/mL. MELD score was 11.27 points. Hepatic encephalopathy (HE) and infection were considered. After intravenous infusion of ceftriaxone, L-ornithine-L-aspartate, and hepatoprotective drugs were given for 3 days, his consciousness and temperature became normal with a blood ammonia level of 54 $\mu\text{mol/L}$. Then, he was discharged.

On October 7, 2014, he was re-admitted to our department due to massive haematemesis and unconsciousness for one day. On laboratory tests, hemoglobin concentration was 51 g/L, WBC was $5.3 \times 10^9/\text{L}$, percentage of neutrophil

was 78.9%, PLT was $114 \times 10^9/L$, blood ammonia was $34 \mu\text{mol/L}$, TBIL was $32.7 \mu\text{mol/L}$, ALT was 99 U/L , AST was 178 U/L , ALP was 42 U/L , albumin was 21.7 g/L , serum creatinine was $129.8 \mu\text{mol/L}$, serum sodium was 138.2 mmol/L , PT was 15.6 seconds, INR was 1.28, and AFP was 1.88 IU/mL . MELD score was 15.02 points. After a written informed consent from this patient and his relatives was obtained, an upper gastrointestinal endoscopy was performed again, which demonstrated mild esophageal varices and massive duodenal ulcer. After transfusion of red blood cells and pharmacological treatments, including esomeprazole, L-ornithin-L-aspartate, and hepatoprotective drugs, etc., repeated haematemesis and hemafecia remained. Considering the peri-operative risk, his relatives refused surgery. He died on October 12, 2014.

Discussion

The causes for the development of this large collateral vessel were uncertain in our case. Because we did not have any prior CT scans for this patient, we were not sure about when this collateral vessel developed. We had two major considerations, as follows. The first consideration was that this collateral vessel was acquired. If so, embryonic channels were reopened in the setting of portal hypertension. Before the occurrence of liver cirrhosis and portal hypertension, this collateral vessel was not existent. With a gradual increase in the portal pressure, portal blood was spontaneously diverted into renal vein through this collateral vessel.

The second consideration was that this collateral vessel was inherent or congenital. According to the review by Sokollik *et al.* (2), congenital portosystemic shunt is divided into five types: (I) extrahepatic congenital portosystemic shunt type I with absence or severe hypoplasia of intrahepatic portal venous system; (II) extrahepatic congenital portosystemic shunt type II with maintained intrahepatic portal venous system; (III) intrahepatic congenital portosystemic shunt within the left lobe; (IV) intrahepatic congenital portosystemic shunt within the right lobe; and (V) patent ductus venosus (2). According to the review by Gupta *et al.* (3), extrahepatic congenital portosystemic shunt type I is further classified as two subtypes: (Ia) splenic vein and superior mesenteric vein drain separately into the inferior vena cava; and (Ib) splenic vein and superior mesenteric vein form a common channel before draining into the inferior vena cava. Extrahepatic congenital portosystemic shunt type II is

further classified as three subtypes: (IIa) the portosystemic shunt arises from intrahepatic portal vein branches; (IIb) the portosystemic shunt arises from main portal vein; and (IIc) the portosystemic shunt arises from gastric, mesenteric, or splenic veins. If this case was attributed to the congenital extrahepatic portosystemic shunt, it should be extrahepatic congenital portosystemic shunt type IIb. But it should be noted that the extrahepatic congenital portosystemic shunt should be observed on CT scans obtained at the childhood or at the time when he was not cirrhotic. If the evidence was lacking, congenital portosystemic shunt should be often accompanied with cardiac anomalies.

Our case did not develop any episodes of variceal bleeding or presented with large esophageal varices, but developed HE. This might be associated with the presence of such a large spontaneous portosystemic shunt, which decreased the portal pressure and the liver detoxification. However, the clinical significance of spontaneous portosystemic shunt in cirrhotic patients remained controversial. The first issue was whether or not spontaneous portosystemic shunt decreased the incidence of esophageal varices or variceal bleeding. The second issue was whether or not spontaneous portosystemic shunt increased the incidence of HE. Some researchers suggested no association between spontaneous portosystemic shunt and esophageal varices. In an early study, Rousselot *et al.* explored the effects of natural portal-systemic shunting in 203 patients, including 135 cirrhotic patients, 15 patients with extrahepatic obstruction of portal system, and 53 patients without any evidence of pathology in the portal system (4). They suggested that the presence of large portosystemic shunts was not associated with the reduction in the portal pressure and incidence or severity of variceal bleeding. Lam *et al.* compared the incidence of variceal hemorrhage among cases of chronic liver diseases with and without large spontaneous shunts (5). They found a similar proportion of variceal hemorrhage between them. In a retrospective study of 20 patients with large self-established portosystemic shunts, Aseni *et al.* also concluded that the risk of bleeding was not correlated with presence of massive spontaneous portosystemic shunt (6). By contrast, other researchers suggested that spontaneous portosystemic shunt was a protective factor for the occurrence of esophageal varices and that spontaneous portosystemic shunt could not influence the development of HE. In an early study, Wexler *et al.* described that a single large spontaneous portosystemic shunt prevented from the development of esophageal varices in six patients with biopsy-proven

liver cirrhosis (7). However, one of them presented with severe HE. Iannello *et al.* also reported that a patient with liver cirrhosis had a large spontaneous splenorenal shunt without any varices at endoscopy or HE (8). Tarantino *et al.* also confirmed that only one of 15 cirrhotic patients with splenorenal shunts had large esophageal varices (9), but the risk of HE was similar between patients with and without splenorenal shunts. Culafic *et al.* reported that a case did not have any episodes of HE or variceal bleeding despite the presence of large spontaneous portosystemic shunt (10). Collectively, the heterogeneous findings among studies might be explained by the discrepancy in the sample size and diameter and type of portosystemic shunt. Indeed, Riggio *et al.* have confirmed the role of large spontaneous portosystemic shunt in the development of persistent HE (11). However, they also acknowledged that two patients with splenic- or mesenteric-renal shunt did not have any episodes of HE and four patients with persistent HE did not have large portosystemic shunts. Thus, other factors should be important in determining the occurrence of HE.

Our case did not receive any interventions for this large collateral, because portosystemic shunt related encephalopathy was readily resolved. Similarly, as recently reported by our team, a case with a large spontaneous intrahepatic portosystemic shunt had no episodes of overt HE and did not receive any therapeutic interventions for portosystemic shunts (12). If there were recurrent of persistent episodes of HE in patients with large portosystemic shunts, angiographic embolization should be considered. In a multicenter European study by Laleman *et al.*, 37 patients with refractory HE were treated with embolization of large spontaneous portosystemic shunts (13). In a single-center study from Mayo Clinic, Rochester, 23 patients with severe persistent HE were also treated with embolization of large spontaneous portosystemic shunts (14). In a single-center case-control study from South Korea, 17 patients with recurrent HE underwent embolization of spontaneous portosystemic shunt (15). Taken together, these studies confirmed the efficacy and safety of embolization of spontaneous portosystemic shunt in patients with recurrent or persistent HE.

In conclusion, this case report demonstrated a very rare case with a large spontaneous extrahepatic portosystemic shunt from main portal vein to right renal vein in a cirrhotic patient.

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Footnote

Conflicts of Interest: The authors have completed the ICMJE uniform disclosure form (available at <http://dx.doi.org/10.21037/amj.2017.02.04>). Dr. Qi serves as an Editor-in-Chief of AME Medical Journal. The other authors have no conflicts of interest to declare.

Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient for publication of this manuscript and any accompanying images.

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