

Portal-systemic Shunt Causing Hepatic Encephalopathy – A Case Report

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Abstract

Portal-systemic shunt, spontaneous connection between systemic veins and portal vein system, is a rare cause among the patients with hepatic encephalopathy. We reported a case of 57-year-old male with extrahepatic portal-systemic shunt who presented with repeated episodes of disturbance of consciousness and characteristic images of abdominal sonography, contrast-enhanced abdominal computed tomography and 3D reconstruction images. There are a few reports in the literature about symptomatic portal-systemic shunt presented in adulthood and it may be misdiagnosed as psychological diseases. So the diagnosis of this rare disease may rely on abdominal image studies only and associated symptoms may be resolved by avoidance of precipitating factors, surgical occlusion or radiological obliteration. (J Intern Med Taiwan 2018; 29: 401-407)

Key Words: Portal-systemic shunt, Hepatic encephalopathy, Disturbance of consciousness, Non-cirrhotic

Introduction

Hepatic encephalopathy is one of the common diseases with the prevalence of 39.9%¹ in cirrhotic patients due to insufficient detoxification from ammonia into urea². However, there are some patients without liver diseases but are diagnosed as hepatic encephalopathy. The intrahepatic and extrahepatic portal-systemic shunts may be the causes of hepatic encephalopathy³, especially in patients without liver cirrhosis. There are a few reports in the literature⁴ about symptomatic portal-systemic shunt

presented in adulthood and it may be misdiagnosed as psychological diseases. Spontaneous intrahepatic or extrahepatic shunts, especially between superior mesenteric vein and inferior vena cava, were previously reported^{1,4,5}. In Taiwan, however, only a case of intrahepatic shunt was reported⁵. We herein reported a non-cirrhotic patient whose extrahepatic shunt was located between the portal vein and the left renal vein.

Case report

A 57-year-old male, who denied liver disease,

abdominal surgery or consumption of alcohol, had history of unknown cause of consciousness disturbance and gastrointestinal bleeding about few years ago. He presented with conscious drowsy, coffee-ground vomiting and tarry stool in this admission. At admission to intensive care unit, his Glasgow Coma Scale (GCS) was E3M4V1 with body temperature of 36.4°C, heart rate of 72 beats per minute, respiratory rate of 19 times per minute with oxygen saturation of 95% under ambient air and blood pressure of 118/82 mmHg. On physical examination, pale conjunctiva and asterixis were noted. There were no signs of liver cirrhosis such as abdominal distension, ascites, hepatomegaly, spider angioma, leg edema, or palmar erythema. The laboratory data showed mild anemia (hemoglobin 11.6 milligram per deciliter (mg/dl)), leukocytosis (white blood count 13360 per microliter) but no thrombocytopenia, hyperbilirubinemia (total bilirubin 2.09 mg/dl), pre-renal azotemia (blood urea nitrogen 51mg/dl, creatinine 0.99 mg/dl), and strong positive for stool

occult blood (4+). Initially, the patient did not have hyperammonemia but the levels of ammonia rose on next day (74.4 mg/dl elevated to 135.7 mg/dl). The culture of blood and urine were sterile. The serologic tests for hepatitis B surface antigen (HBsAg) and anti-hepatitis-C-virus antibody (anti-HCV) were non-reactive. The levels of glutamate oxaloacetate transaminase (GOT), glutamate pyruvate transaminase (GPT), gamma-glutamyl transpeptidase (γ -GT) and alkaline phosphatase (ALP) were within normal range. The upper gastrointestinal endoscopy revealed an active ulcer (A2) over gastric angularis, endoscopic hemostasis was performed and no evidences of esophageal and gastric varices were found. The portable bed-side abdominal sonography demonstrated hepatic steatosis and torturous tube-like structures near hepatic hilum and pancreas (Fig. 1). There was no evidence of dilatation of biliary tract, hepatomegaly, cholelithiasis, portal vein thrombus, intrahepatic shunt, cirrhosis of liver or splenomegaly. The abdominal computed

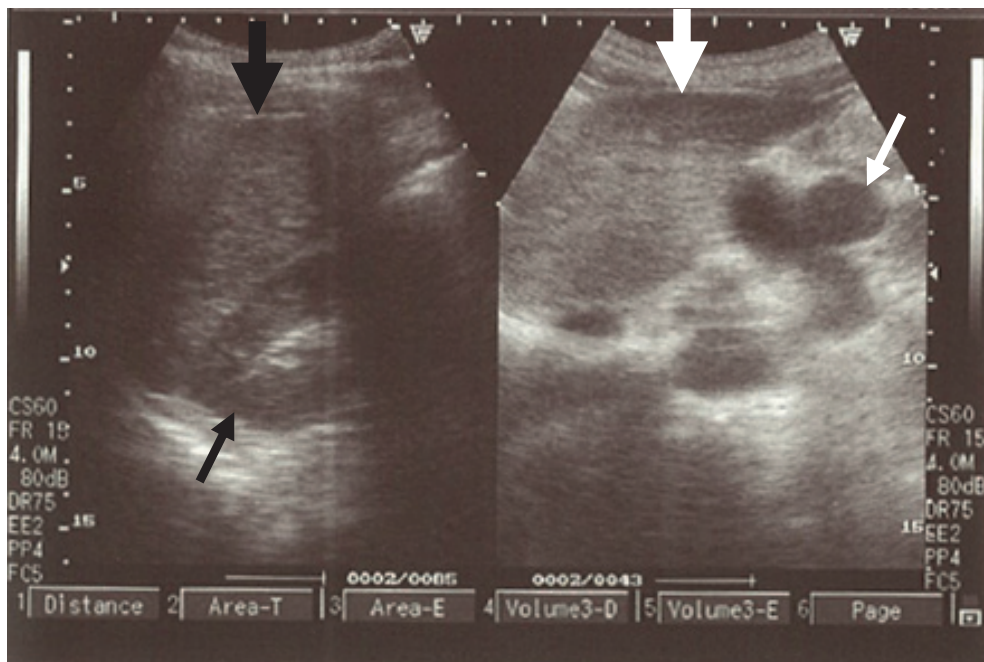


Figure 1. Abdominal sonography, right panel: right axillary line sagittal view shows right lobe of liver (black thick arrow) and right kidney (black thin arrow), left panel: mid-line sagittal view shows left lobe of liver (white thick arrow) and tortuous tube like structures near liver (white thin arrow).

tomography (CT) with contrast enhancement was arranged to study the nature of torturous tube-like structures near hepatic hilum and it revealed the portal-systemic shunt from confluence of superior mesenteric vein and splenic vein to left renal vein (Fig. 2) and no fibrotic changes or signs of recanalisation of paraumbilical vein, segmental hypertrophy, segmental atrophy, splenomegaly, nodular surface, calcification within portal system or ascites was found. Two extrahepatic portal veins were seen, one derived from confluence of superior mesentery vein and splenic vein; another portal vein derived from shunt (Fig. 3). The 3D images from reconstructing CT coronal view slices by MIIL 3.0 software [6] demonstrated the tortuous portal-systemic shunt more clearly (Fig. 4). Due to no clinic or radio-

logic evidence of liver cirrhosis, we thought the hepatic encephalopathy of this patient resulted from the large portal-systemic shunt. After administration of rectal lactulose for enema, intravenous high dose proton pump inhibitor, endoscopic hemoclips, blood transfusion, fluid resuscitation and empiric parenteral antibiotics, the symptoms of this patient subsided gradually. His consciousness became significantly clear. He was then transferred to general ward and the oral lactulose was given continuously. He was discharged on the 13th day of admission without complications and then he visited our out-patient clinic once but lost of follow up later.

Discussion

Clinically, the diagnose of portal-systemic

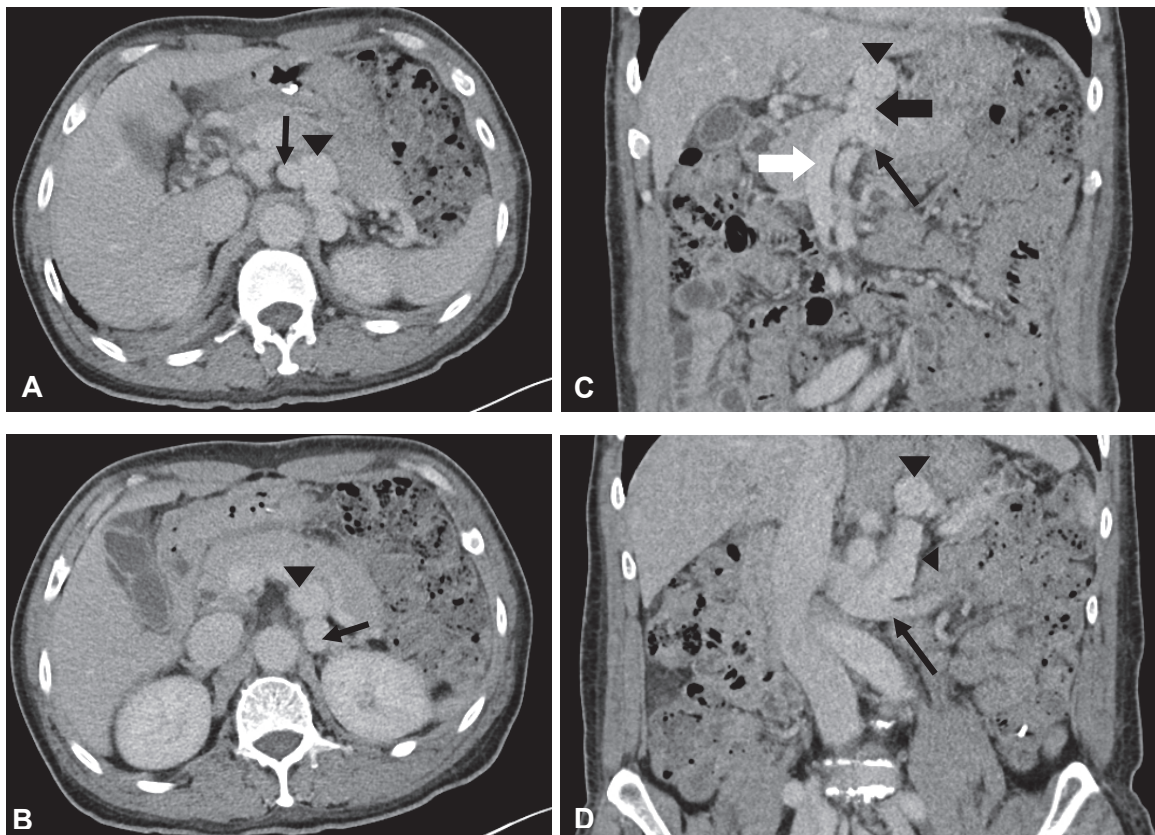


Figure 2. Portal-systemic shunt: (A) axial view, shunt (black arrow head) starts from confluence of superior mesenteric vein and splenic vein (black arrow); (B) axial view, shunt (black arrow head) ends at left renal vein (black arrow); (C) coronal view, shunt (black arrow head) starts from confluence (black thick arrow) of superior mesenteric vein (white thick arrow) and splenic vein (black thin arrow); (D) coronal view, shunt (black arrow heads) ends at left renal vein (black arrow).

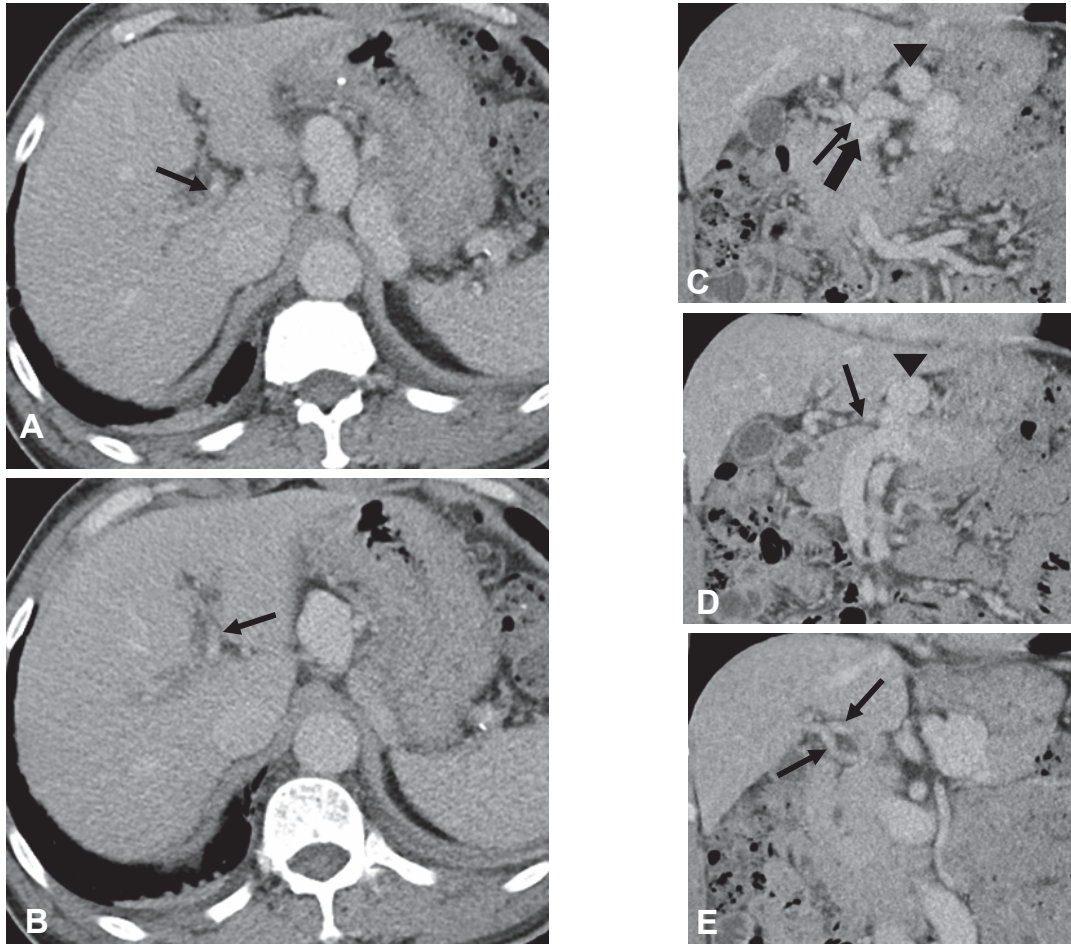


Figure 3: Portal veins: (A) axial view, small caliber of right branch of portal vein (black arrow); (B) axial view, small caliber of left branch of portal vein (black arrow); (C) coronal view, one portal vein (black arrow) starts from confluence (black thick arrow) of superior mesenteric vein and splenic vein, shunt (black arrow head); (D) coronal view, another portal vein (black arrow) starts from shunt (black arrow head); (E) coronal view, two portal veins (black arrows) near hilum of liver.

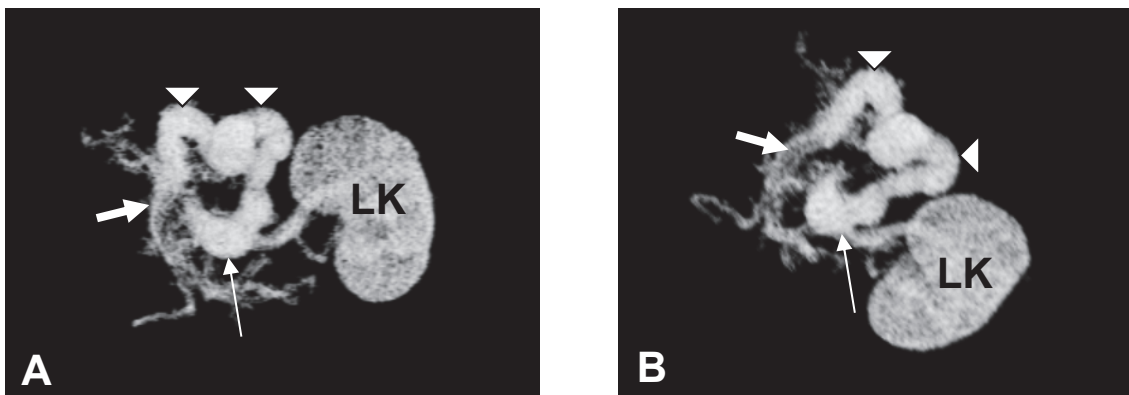


Figure 4. 3D picture: (A) 'Coronal' view, tortuous portal-systemic shunt (white arrow head) starts from confluence of superior mesenteric vein (white thick arrow) and splenic vein and ends at left renal vein (white thin arrow), LK: left kidney; (B) 'rotated coronal' view, tortuous portal-systemic shunt (white arrow head) can be more clearly seen. White thick arrow: superior mesenteric vein, white thin arrow: left renal vein, LK: left kidney.

shunt in non-cirrhotic patients is considerably difficult because the symptoms of hepatic encephalopathy are nonspecific. The hepatic injury in these patients is very mild and the portal pressure does not rise even there was a large shunt. As a consequence, these patients might be misdiagnosed as dementia, depression or psychological diseases⁷. Signs and symptoms may include repeated episodes of disturbance of consciousness. The laboratory data might be normal or slight elevated in liver enzyme, bilirubin or ammonia. The methods to diagnose in the patients with portal-systemic shunt include clinical manifestation, radiological imaging such as abdominal sonography, computed tomography (CT), magnetic resonance imaging (MRI) and transcolonic portal scintigraphy⁴. The abdominal sonography was most frequently adapted in these patients with liver problem. It is important to find if any intrahepatic or extrahepatic torturous tube-like structure in sonography examination of patients with hepatic encephalopathy. If torturous tube-like structures exist such as this case, further abdominal CT or MRI studies can identify the portal-systemic shunt. Furthermore, 3D reconstruction images using CT or MRI slice images can demonstrate more comprehensive shunt pictures. The classification of portal-systemic encephalopathy by Watanabe et al. was based on the location of portal-systemic shunt (origin, end and involvement of the liver or not). Type I (intrahepatic type) is a type of shunt whose origin and end are both in the liver. This type is usually resulting from congenital vascular anomaly such as failed degeneration of patent venous duct or Rendu-Osler-Weber disease. Type II (intra- and extrahepatic type) has an extrahepatic origin from the portal vein and go through into the liver. Type III (extrahepatic type), most frequently seen, has extrahepatic origin such as the left gastric vein, superior mesenteric vein or splenic vein and end at inferior vena cava or left renal vein. Type IV, another extrahepatic type, is as same as type III anatomi-

cally but the dynamics of hepatic blood circulation are similar to those of idiopathic portal hypertension. Type V represents the other type of extrahepatic type without portal perfusion of liver⁴. In our case, the origin of shunt was the confluence of superior mesenteric vein and splenic vein and the end was left renal vein, without liver involvement, his portal-systemic encephalopathy type can be classified as type III. For this case, small calibers of portal vein branches and double extrahepatic portal veins were found (Fig. 3), the small portal veins might be associated with the formation of portal-systemic shunt. Watanabe reported that some patients (type V) with portal-systemic shunt did not have portal veins⁴. However, there were two portal veins in our case. The multiple branches of portal vein and shunt could be found in the patient with cavernous transformation of the portal vein (CTPV). The CTPV is the consequence of portal vein thrombosis [8]. The typical changes in CTPV are atrophy of the left lateral segment, hypertrophy of segment IV and caudate lobe⁹ and especially linear areas of calcification within the portal vein indicating chronic venous thrombosis¹⁰. In our case, both abdominal echogram and computed tomography could not find any evidence of portal vein thrombus. Moreover, in the patients with CVPT, the strictures or displacements of biliary tracts are often occurred¹¹⁻¹², which was not found in our case. The current treatments for portal-systemic shunt includes interventions, such as surgically or radiologically, and medical management. Surgical interventions are surgical occlusion of shunts¹³ and liver transplantation¹⁴. The radiologic intervention includes endovascular occlusion¹⁵ or embolization¹⁶. Medical managements are the avoidances of precipitant factors such as high-protein diet, gastrointestinal bleeding, constipation, hypokalemia, hypnotic medication, diuretics, hypovolemia or metabolic alkalosis. These therapies are similar for cirrhotic patients with hepatic encephalopathy⁴.

Conclusion

A non-cirrhotic patient with hepatic encephalopathy due to portal-systemic shunt was relatively rarely seen. It's important to obtain images of splanchnic vessels to identify spontaneous portal-systemic shunts in the patients who have repeated episodes of consciousness disturbance. This unusual presentation of hepatic encephalopathy may provide further clinical thinking in the future.

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一位男性因罹患因肝門靜脈系統靜脈分流造成的 肝性腦病變：病例報告

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摘 要

在臨床上，大部分的肝性腦病變是導因於肝硬化造成的高血氨症，然而，肝門靜脈系統靜脈分流造成的肝性腦病變相對罕見，因此，我們今天報告一例57歲男性因為上腸系膜靜脈與左腎靜脈之間的分流而導致肝性腦病變的病人，希望我們的診斷與治療經驗能提醒各位臨床醫師。