An Inferior Mesenteric-Caval Shunt via the Internal Iliac Vein with Portosystemic Encephalopathy

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Abstract

We report here a case of an unusual extrahepatic portosystemic venous shunt in a 37-year-old woman without liver cirrhosis or portal hypertension, who developed portal systemic encephalopathy. Angiography demonstrated an inferior mesenteric-caval shunt characterized by the presence of direct communication of the inferior mesenteric vein with the left internal iliac vein. After the treatment with percutaneous transcatheter embolization of the shunt via a femoral vein approach using coils, she had no episode of portal systemic encephalopathy.

Key words: inferior mesenteric vein, inferior vena cava, hepatic encephalopathy, hyperammonemia, interventional radiology

Introduction

Extrahepatic portal systemic venous shunts develop preferentially in portal hypertension (1). Most of extrahepatic portal systemic collaterals are gastroesophageal, gastrorenal, splenorenal, paraumbilical, inferior mesenteric-hemorroidal, and superior mesenteric-caval shunts (2). In this report, we describe a case of an unusual extrahepatic portosystemic venous shunt characterized by the presence of an inferior mesenteric-caval shunt with direct communication between the inferior mesenteric vein and the left internal iliac vein in spite of the absence of liver cirrhosis and portal hypertension.

Case Report

A 37-year-old Japanese woman was admitted to a local hospital in August 1997 because of coma. On admission, her blood ammonia level was 317 µg/dl, and electroencephalogram showed δ waves with a frequency of 3 per second. A diagnosis of hepatic encephalopathy was suggested. Her consciousness improved with branched-chain amino acid supplement, and her blood ammonia level decreased. In April 1998, she had a similar episode again and was admitted to the same local hospital. At that time, her clinical and radiological findings showed no evidence of liver cirrhosis. Superior mesenteric arterial portography did not show any intra- or extra-hepatic portal systemic shunts. She was referred to Hamamatsu University Hospital for further evaluation of her encephalopathy. Her past medical history included two caesarean sections and spina bifida occulta. She did not drink alcohol and was not taking any medication. Her family history was unremarkable.

On physical examination, she had no evidence of anemia, icterus or cutaneous stigmata of chronic liver disease. Neither the liver nor spleen tip was palpable. There was no evidence of ascites or peripheral edema. Neurological examination did not reveal any abnormality. Laboratory studies disclosed the following values (the normal reference range is given in parentheses): erythrocyte count 381×10⁴/mm³; hemoglobin 9.0 g/dl; hematocrit 29.3%; leukocyte count 3,600/ mm³; platelet count 30.9×10⁴/mm³; total protein 6.9 g/dl (6.5–8.0 g/dl); albumin 3.7 g/dl (4.1–5.1 g/dl); total bilirubin 0.3 mg/dl (0.3–1.4 mg/dl); serum lactate dehydrogenase 168 IU/l (101–193 IU/l); serum aspartate aminotransferase 13 IU/l (11–30 IU/l); serum alanine aminotransferase 10 IU/l (5–42 IU/l); serum alkaline phosphatase 194 IU/l (117–356 IU/l); serum γ-glutamyl transpeptidase 12 IU/l (12–75 IU/l); fasting blood glucose 83 mg/dl; prothrombin time 12.2 s (control: 12.2, 100%). Neither hepatitis B surface antigen nor antibodies to Hepatitis C virus (HCV) were detected in serum by enzyme immunoassay. Indocyanine green excretion test showed 3% retention at 15 minutes (<10%). Ammonia concentration in peripheral blood was 50.9 µg/dl (30–80 µg/dl). Serum amino acid profiles were normal, and allopurinol load test (3) did not show any increase in urinary orotic acid excretion.

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Upper abdominal ultrasonography and computed tomography did not disclose any abnormal findings. No esophageal or gastric varices were found by endoscopy. Thallium-201 per rectal scintigraphy showed that the ratio of heart to liver radioactivities was 1.0, suggesting the presence of severe portal systemic venous shunts. Superior mesenteric arterial portography and splenic arterial venography did not detect any hepatofugal blood flow (Fig. 1A and B), while inferior mesenteric arterial venography demonstrated that the blood streams of the left colic, sigmoid and superior rectal veins merged into an abnormal vessel communicating with the left internal iliac vein. There was no hepatopedal blood flow through the inferior mesenteric vein, but considerable hepatofugal blood flow through the communicating vessel (Fig. 1C). Selective venous sampling of

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Figure 1. (A) Superior mesenteric arterial portography did not show any sign of hepatofugal blood flow. (B) Splenic arterial venography did not show any hepatofugal blood flow. (C) Inferior mesenteric arterial venography demonstrated that the blood flows of the left colic, sigmoid and superior rectal veins merged into an abnormal vessel (arrow) communicating with the left internal iliac vein. (D) After embolization, hepatopedal blood flow through the middle colic vein and inferior mesenteric vein was demonstrated without any evidence of shunt flow. IVC: inferior vena cava, IMV: inferior mesenteric vein, PV: portal vein.
blood ammonia showed a higher value of 192.0 µg/dl in the shunt vessel in contrast with 49.8 µg/dl in the inferior vena cava and 26.1 µg/dl in the left external iliac vein.

Final diagnosis was extrahepatic portal systemic venous shunt without portal hypertension. The previous episodes of the disturbed consciousness of this patient were considered to have been extrahepatic portal systemic shunt encephalopathy. During hospitalization, she had no similar events of encephalopathy. The precipitating factors remained unclear. However, considering the risk of recurrence of coma due to portosystemic encephalopathy in the near future, we decided to occlude the shunt using an angiographic technique. A catheter was inserted into the shunt through the left internal iliac vein via a percutaneous right femoral vein approach. Accidental venous spasm of the shunt vessel due to catheter insertion provided the simulation of shunt occlusion. At that time, inferior mesenteric arterial venography confirmed the appearance of hepatopedal blood flow through the inferior mesenteric vein. Transcatheter embolization of the shunt vessel was performed via a percutaneous femoral vein approach using four platinum coils (Tornade embolization coil; Cook Inc., Bloomington, IN, USA). After the embolization, inferior mesenteric arterial venography demonstrated no evidence of shunt flow. Hepatopedal blood flow through the middle colic vein and inferior mesenteric vein was detected (Fig. 1D). The patient has been very well without recurrence of any symptoms.

Discussion

Extrahepatic portosystemic venous shunts frequently develop in patients with portal hypertension (1); such shunts without portal hypertension are rare. In the present case, there was little clinical evidence of the presence of portal hypertension, although portal pressure and liver histology were not evaluated. She did not demonstrate any of the typical, physical signs of portal hypertension such as splenomegaly, varices or ascites. There were no abnormal collateral vessels except for a single portosystemic venous shunt detected by computed tomography or angiography. It is, therefore, difficult to consider that portal hypertension is present without any clinical signs, and that only one portosystemic venous shunt had developed without any other collateral pathways under portal hypertension.

There are various types of extrahepatic portal systemic collateral, most of which are gastroesophageal, gastrorenal, splenorenal, paraumbilical, inferior mesenteric-hemorrhoidal, and superior mesenteric-caval shunts (2). An inferior mesenteric-caval shunt is rarely encountered. The portal systemic venous shunt observed in the present case can be regarded as an inferior mesenteric-caval shunt with direct communication of the inferior mesenteric vein with internal iliac vein. Five cases of inferior mesenteric-caval shunts have been reported (4–8) (Table 1). All of the cases showed that the majority of the superior mesenteric venous flow drained into the dilated inferior mesenteric vein, and the inferior mesenteric venous flow was retrograde, while in the present case, the superior mesenteric venous flow entered the portal vein normally, but the shunt vessel, not the inferior mesenteric vein, drained the descending and sigmoid portions of the colon. Interestingly, patients younger than our patient did not experience portal systemic encephalopathy despite the presence of larger shunt vessels.

The cause of the inferior mesenteric-caval shunt observed in the present case remains unclear. It is considered likely to have been a congenital portosystemic venous shunt, since the

<table>
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<th>Case No.</th>
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<th>Sex</th>
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<th>Gastroesophageal varices</th>
<th>Histology of the liver</th>
<th>History of abdominal surgery</th>
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<td>caesarean sections</td>
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Table 1. Five Reported Cases of Inferior Mesenteric-Caval Shunts without Liver Cirrhosis

Case No. 4 showed congenital absence of the portal venous system. IVR: Interventional radiology. ND: not done.
shunt vessel was single and occurred in a young patient without liver cirrhosis and portal hypertension. Patency of the embryonic channel, demonstrated by Edward, might have occurred (9). However, this raises the question why our patient grew up without developing encephalopathy until the age of 37. The brain sensitivity to ammonia or other toxic metabolites may increase with aging as proposed previously (10). Another speculation is that homeostatic control of the production of intestinal flora-derived ammonia may gradually become disordered with increasing age, resulting in the predisposition to hyperammonemia-induced encephalopathy at high age as proposed by Nishimoto et al (8). Such mechanisms may contribute to the delayed presentation with hyperammonemia-related encephalopathy in the present case.

Another possible cause of the shunt is vascular formation involved in postoperative adhesion. Moncure et al proposed adhesion-related mesenteric-caval shunt as a cause of portosystemic shunt without portal hypertension (11). Although our patient had a prior history of caesarean section, such laparotomy cannot easily become a cause of adhesion-related inferior mesenteric-caval shunt.

Chronic or recurrent disabling portosystemic encephalopathy that is refractory to conventional treatment is generally considered an indication of shunt occlusion. However, the management of such patients must be judged according to their characteristic risk/benefit ratio. In portosystemic shunt associated with portal hypertension, the therapeutic blockade leads to overload of the portal venous system, increasing the risk for ascites formation and variceal bleeding (12); portal pressure measurement before and after temporary balloon occlusion of the shunt is recommended to test tolerance to subsequent shunt occlusion (6, 13, 14). In the present case, such risks were judged to be much lower because there was no evidence of liver cirrhosis or portal hypertension.

Primary treatment choices for extrahepatic portosystemic venous shunts are surgical ligation (10, 15) and interventional embolization (16, 17). Recent advances in interventional radiological techniques have increased the option of transcatheter embolization for extrahepatic portosystemic venous shunts. As we were concerned about postoperative development of adhesion-related portosystemic collaterals, we chose interventional embolization, not surgical ligation in the present case. Coils were used as embolic materials, because they may occlude the shunt progressively, avoiding acute overload of the portal venous system. The posttreatment evaluation after two years confirmed that there is no evidence of recanalization of the shunt vessel. However, long-term observation will be necessary to evaluate the outcome of interventional embolization.

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References