Extrahepatic Portosystemic Venous Shunt without Portal Hypertension

Yasuhiro Nishimoto, Hiroshi Hoshino, Shinji Sato, Akihiko Oguri, Masahiko Yamada, Daisaku Nishimura, Naoyuki Katada, Hiroshi Sano and Katsumoto Kato

A 72-year-old woman was admitted for recurrent episodes of encephalopathy. Laboratory data showed mild liver dysfunction and hyperammonemi a, while she had neither anemia nor splenomegaly. The dilated inferior mesenteric vein (IMV) was opacified retrogradely from the superior mesenteric vein by superior mesenteric arteriography, and IMV was found to connect with the inferior vena cava (IVC) through a torturous shunt. No obstruction of the extrahepatic portal vein or hepatic vein was observed by arteriography. Histological evaluation of the liver biopsy indicated remarkable fatty change without cirrhosis. Finally, we diagnosed this case as extrahepatic portosystemic venous shunt without portal hypertension.

Key words: inferior mesenteric vein (IMV), inferior vena cava (IVC), hepatic encephalopathy

Introduction

Intrahepatic microscopic portosystemic venous shunt is sometimes observed in liver cirrhosis (1–3), however, intrahepatic macroscopic shunt has been rarely reported since the first description by Raskin et al (4). Excluding traumatic causes, the extensive intrahepatic portosystemic shunt is thought to originate either from congenital vascular malformation or as a result of massive necrosis postnatally induced by factors such as viral infection or hepatotoxic substances (2, 5). On the other hand, extrahepatic collateral veins are frequently observed in portal hypertension such as in liver cirrhosis or idiopathic portal hypertension. In cases without portal hypertension, extrahepatic portosystemic venous shunt is extremely rare. To the best of our knowledge, only 8 cases have been described in the world literature (6–13). Here, we report a rare case of extrahepatic portosystemic venous shunt without portal hypertension.

Case Report

A 72-year-old woman had been in good health except for spondylosis until November 1992, when she experienced an episode of abnormal behavior and disorientation persisting for 1 or 2 days. Similar episodes occurred at half year intervals and the patient’s family consulted our hospital, Kamo Hospital, and the patient was admitted on October 21, 1993. Although the patient underwent an operation for intraocular lens implantation for a cataract at age 70, she had no past history of abdominal trauma, diseases or operations. Nobody in her family had either hepatic, neurological or metabolic disease. The patient never smoked and seldom drank alcoholic beverages. She had no history of drug abuse.

The patient’s height was 141.5 cm and her weight was 58 kg. Body temperature on admission was 37.1°C and blood pressure was 134/94 mmHg. She was well nourished, overweight and did not appear to be suffering from any chronic disease. Although the patient’s consciousness was alert, she responded slowly to questions and commands. On physical examination, her skin was free of eruption, anomalous pigmentation or spider angiomas, but palmar erythema was observed. The bulbar conjunctiva showed no icterus and the palpebral conjunctiva showed no anemia. The chest was clear to percussion and auscultation. The liver and spleen were not palpable. There was no tenderness over the abdomen. The superficial lymph nodes were not palpable, and no edema was observed. Hemorrhoid was not noticed in the anus. Flapping tremor of the wrist and digits was elicited.

Hematological studies disclosed the following values; white blood cell count 10,800/mm³ with 52% neutrophils, 43% lymphocytes, 2% monocytes, 2% eosinophils and 1% basophils, red blood cell count 407 x 10⁴/mm³, platelet count 26.1 x 10⁴/mm³, hemoglobin 14.1 g/dl, hematocrit 39.4%, prothrombin time 100.0%, activated partial thromboplastin time 100.4%, and heparplastin test 120.1%. Blood chemical studies showed
mild liver dysfunction as follows; total bilirubin 0.5 mg/dl (normal 0.2–1.0 mg/dl), aspartate aminotransferase 60 IU/l (normal 8–38 IU/l), alanine aminotransferase 80 IU/l (normal 5–33 IU/l), and lactic acid dehydrogenase 418 IU/l (normal 200–420 IU/l). Serum total protein was 7.0 g/dl (normal 6.1–8.4 g/dl), and the serum albumin was 3.6 g/dl (normal 3.6–5.1 g/dl). Serum ammonia level rose to 211 μg/dl (normal 35–85 μg/dl). Hepatitis B surface antigen and antibody were both negative, as was a test for the hepatitis C virus antibody. Tumor markers such as carcinoembryonic antigen or alpha-fetoprotein were normal. Tests for antimitochondrial antibody, antismooth muscle antibody and antinuclear antibody were all negative. The fasting aminogram showed mild elevation of taurine, aspartic acid, glutamic acid, proline, citrulline, α-aminobutyric acid, cystine, methionine, tyrosine and phenylalanine. Specific patterns of aminogram as in citrullinemia and argininosuccinic aciduria, usually accompanied by hyperammonemia, were not disclosed. The molar ratio of branched-chain amino acids (leucine, isoleucine and valine) to aromatic amino acids (tyrosine and phenylalanine) decreased to 1.83 (normal 2.43–4.40). An indocyanine green test showed 25% retention at 15 minutes (normal <10%).

The electroencephalography demonstrated sporadic delta waves and slow waves. Computed tomographic (CT) evaluation of the brain disclosed mild atrophy of the cerebral cortex and calcifications in the basal ganglia. No varices were observed in either esophagus or stomach by upper digestive tract endoscopy. CT scans of the abdomen showed extreme fatty infiltration in the liver with hepatomegaly (Fig. 1). The density of parenchyma of the liver was indicated much lower than that of blood vessels. Splenomegaly and ascites were not detected, and the remainder was not remarkable.

Conservative treatment such as oral administration of lactulose (45 ml per day) and infusion of branched-chain amino acid enriched solution (500 ml per day) diminished the ammonia level to 124 μg/dl, whereas the patient was free from mental disturbance. She was discharged on November 11, 1993. The patient’s ammonia level has never decreased to less than 100 μg/dl in spite of anti-hyperammonemia therapy such as oral administration of lactulose (45 ml per day) and kanamycin sulfate (1.5 g per day) since then. However, she sometimes experiences mental disturbances.

The patient was admitted on October 18, 1995 for the second time, because of recurrent symptoms such as anomalous behavior, drowsiness and disorientation. Her physical findings and laboratory data were also the same as on the first admission. Arteriography of the abdomen was performed and revealed the following findings. Celiac arteriography showed no abnormal finding, and the venous phase of splenic arteriography also showed normal splenic and portal veins. The venous phase of a superior mesenteric arteriography revealed that a dilated, 10 mm in diameter, inferior mesenteric vein (IMV) was opacified retrogradely from the superior mesenteric vein, and that the IMV connected with the inferior vena cava (IVC) through a torturous shunt (Figs. 2 A, B). Pressure of portal vein was not examined.

Ultrasound-guided fine needle biopsy of the liver was performed and the specimens indicated considerable fatty change of large and small droplet type in the lobule. Mild lymphocytic infiltration were shown at the portal area, however, there was no evidence of liver cirrhosis. Histologically, the present case was diagnosed as fatty liver. Idiopathic portal hypertension was excluded as follows; neither splenomegaly nor anemia was observed clinically, or no histological finding such as fibrosis with narrowing of portal veins was recognized in the patient. Finally, we diagnosed the case as extrahepatic portosystemic venous (IMV-IVC) shunt without portal hypertension such as in liver cirrhosis or idiopathic portal hypertension.

In order to manage serum ammonia and to diminish the symptoms, we recommended that the patient undergo an operation for shunt ligation, however, she rejected the surgical treatment. She was discharged on November 10, 1995. The patient remains alive and is treated with 100 g of high branched-chain amino acid diet, oral administration of 45 ml of lactulose and 1.5 g of kanamycin sulfate per day. She was well for six months after discharge, but hyperammonemia sometimes still occurs.

**Discussion**

In an early stage of embryological development, the right subcardinal vein becomes a part of the hepatic segment of the inferior vena cava, and the vitelline vein devies to the vitelline sinusoids, which become the intrahepatic portal vein branches and the hepatic veins (14). Intrahepatic portosystemic shunt was first reported by Raskin et al in 1964 (4), and more than 30 cases have been reported since then. Raskin et al (4) proposed...
Figure 2.  A) The venous phase of a superior mesenteric arteriography. B) The schema of the venous phase of a superior mesenteric arteriography. A dilated IMV is opacified retrogradely from the superior mesenteric vein, and that IMV connects with the IVC through a torturous shunt. Direction of blood flow is shown by arrows.


Figure 3. Specimens obtained by ultrasonically guided needle biopsy from the liver (HE stain, x40) indicate remarkable fatty change of large and small droplet type in the lobule. The portal area was mildly infiltrated by lymphocytes, however liver cirrhosis was not apparent.

intrahepatic shunt as congenital origin based on persistence of portions of the omphalomesenteric venous system. On the other hand, Kozuka et al considered the abnormal shunt to be acquired because microscopic examination showed both the muscular layer and the elastic lamellae to disappear abruptly from the wall of the shunt (5). It is still obscure whether intrahepatic portosystemic venous shunt is congenital or acquired.

Extrahepatic portosystemic venous shunt without portal hypertension or abdominal trauma is extremely rare, only 9 cases (including the present one) have been reported since Kerlan et al (6) first described it in 1982. Including the cases with liver cirrhosis or idiopathic portal hypertension more cases were added, but it is difficult to distinguish extrahepatic portosystemic shunt from the collateral pathway caused by portal hypertension. Moreover, the shunt caused by trauma is apparently different from the other, non-portal hypertensive non-traumatic, portosystemic venous shunt from the viewpoint of etiology. For our review, we excluded reports with cirrhosis, idiopathic portal hypertension or a past history of trauma here. The male to female ratio in the patients of extrahepatic portosystemic shunt without portal hypertension was 1:8, fe-
It is still obscure whether extrahepatic portosystemic venous shunt is congenital or not, thus further investigation is necessary.

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References