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Hepatic encephalopathy caused by congenital extrahepatic portosystemic venous shunt

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Abstract Congenital portosystemic venous shunt is a relatively rare disease. Recently, a 60-year-old woman was admitted to our hospital for hepatic encephalopathy caused by congenital extrahepatic portocaval shunt. She had been in good health until the onset of this event, with no liver damage and no experience of abdominal surgery or history of abdominal trauma. In May 1993, hepatic encephalopathy manifested suddenly, with the chief complaint of orthostatic disturbance. Although conservative treatment was administered during the subsequent 5 years, on admission, liver damage and slight splenomegaly were shown, for which complete resection of the shunt vessel and splenectomy were performed. Postoperatively, the patient's symptoms have been alleviated. Hepatic encephalopathy caused by congenital portosystemic venous shunt requires long-term conservative treatment, and the patient's quality of life is reduced. For this reason, surgical intervention or embolization with interventional radiology should be considered, and the maintenance of hepatic blood flow should also be considered.

Key words Hepatic encephalopathy · Congenital portosystemic venous shunt · Extrahepatic portocaval shunt · Liver dysfunction · Splenomegaly

Introduction

Portosystemic encephalopathy is cerebral intoxication in which portal venous blood bypasses the detoxification site in the liver and drains directly into the systemic circulation.[1] A portosystemic shunt often occurs as a symptom secondary to portal obstruction, liver cirrhosis, or idiopathic portal hypertension (IPH). However, there have been reports of a sudden episode of hepatic encephalopathy with no marked histological changes in the liver, the so-called congenital portosystemic venous shunt. Congenital portosystemic venous shunt was first reported by Raskin et al.[2] in 1964. We report here a patient with congenital extrahepatic portocaval shunt with secondary liver damage caused by ischemic changes, and slight splenomegaly that developed gradually during conservative treatment.

Case report

A 60-year-old woman was admitted to our hospital in January 1998 because of recurrent episodes of consciousness disturbance that had continued for 5 years. Orthostatic disturbance while she was walking began to appear around May 1993. Her clinical course was observed by a general practitioner. In April 1995, she was admitted to another hospital because of severe consciousness disturbance. At that time, hepatic encephalopathy caused by extrahepatic portocaval shunt was diagnosed. In January 1998, astasia appeared, and the patient was referred to our hospital in response to the family's wishes. There was no history of alcohol or drug abuse. She denied prior abdominal surgery or history of abdominal trauma. Laboratory findings on admission are summarized in Table 1. On hematological studies, slight anemia and coagulopathy were observed. After admission, the platelet count varied, being around $14 \times 10^4/\mu l$, with the lowest value, $7 \times 10^4/\mu l$. On blood chemistry tests, elevated serum aspartate aminotransferase (AST) and serum alanine aminotransferase (ALT) levels, with decreased serum cholinesterase level, were observed. Serum total protein and albumin were also decreased. Her serum ammonia level was markedly high, at 240 μg/dl. Hepatitis B surface antigen and antibody, as well as hepatitis C antibody, were all negative. The indocyanine green test showed 40% retention at 15 min. The molar ratio of branched-chain amino acids was low, at 0.92.

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Table 1. Laboratory data on admission

Hematology	
WBC	3000/mm^3
RBC	394 × 10^4/mm^3
Hb	11.0 g/dl
Ht	32.1%
Plt	7.3 × 10^4/mm^3
Coagulation	
PT	57%
APTT	49.6 s
HPT	49%
Fibrinogen	234 mg/dl
Serological examination	
HbsAg	(−)
HbsAb	(−)
HCV	(−)
Blood chemistry	
TP	5.9 g/dl
Alb	3.2 g/dl
T-Bil	1.3 mg/dl
D-Bil	0.4 mg/dl
AST	59 IU/l
ALT	68 IU/l
LDH	241 IU/l
ALP	426 IU/l
ChE	2.6 IU/l
γ-GTP	19 IU/l
LAP	73 IU/l
T-Cho	112 mg/dl (120–220 mg/dl)
TG	49 mg/dl
BUN	7.0 mg/dl
Cr	0.6 mg/dl
CRP	0.1 mg/dl
ICGR15	40%
NH_3	240 μg/dl (<75 μg/dl)
Fisher ratio	0.92 (2.43–4.40)
AFP	3.7 ng/ml (<8.7 ng/ml)
CEA	2.1 ng/ml (<7.6 ng/ml)

Figures in parentheses are normal ranges

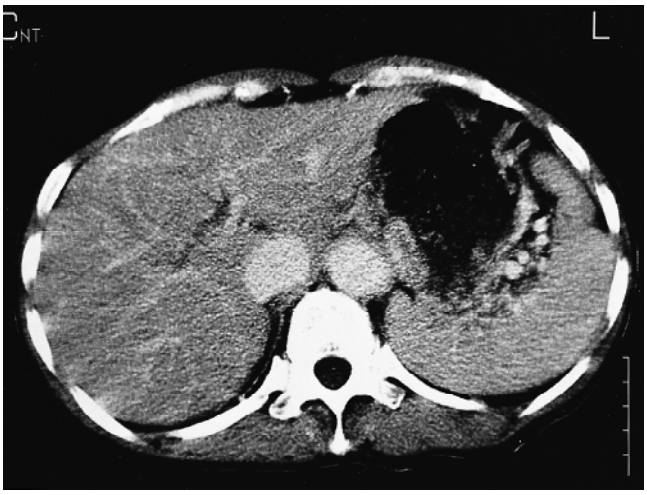


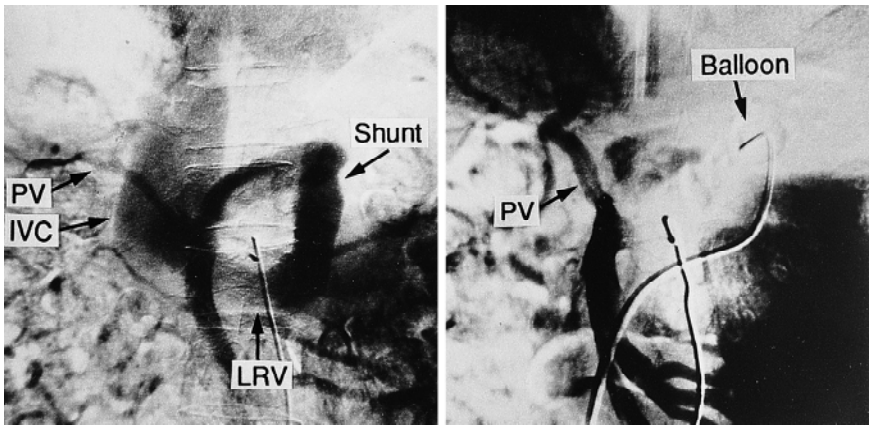
Fig. 1. Abdominal computed tomography shows fatty infiltration of the liver and slight splenomegaly

An endoscopic examination revealed no esophageal or gastric varices. Abdominal computed tomography (CT) showed severe fatty infiltration of the liver and slight splenomegaly (Fig. 1). The venous phase of a superior mesenteric arteriogram revealed a dilated shunt vessel between the superior mesenteric vein, with a slender portal vein, and the inferior vena cava via the left renal vein (Fig. 2a). When the shunt route was occluded by a balloon, the portal blood flow increased, and diameter of the portal vein was enlarged about three fold (Fig. 2b). The wedge pressure in the hepatic vein, measured simultaneously, was 17 cmH_2O. Conservative treatment (lactulose syrup; restricted protein intake; branched-chain amino acids; enema) improved the serum ammonia level to 70–80 μg/dl. To control the serum ammonia level and to diminish the patient's symptoms, surgical intervention was decided on. At surgery, the shunt vessel was noted at a site corresponding to the communication of the superior mesenteric vein and splenic vein. After separation of the left gastric vein, it was confirmed that the shunt vessel connected with the left renal vein. Further, the splenic vein was also observed to be dilated and tortuous (Fig. 3). Resection of the shunt vessel and splenectomy (180 g) were then performed. The portal pressure measured during operation increased from 15 to 21 cmH_2O. Histologic examination of surgically obtained wedged biopsy specimen of the liver showed considerable fatty change. Mild fibrous changes in the central vein and portal area were also shown. On the other hand, no noticeable inflammatory cells were present (Fig. 4).

The patient's liver function and coagulation capacity were improved after the operation, and serum ammonia levels were also normalized. Her neurological symptoms also showed amelioration after the operation.

Discussion

A portosystemic venous shunt is usually induced subsequent to a disease that exhibits portal hypertension, such as liver cirrhosis or IPH; however, some patients with such shunts without portal hypertension have been reported in recent years. This may occur in a disease state such as so-called congenital portosystemic venous shunt.[1–8] Congenital portosystemic venous shunt can be classified into intrahepatic or extrahepatic, according to the site of the shunt. Encephalopathy caused by congenital shunt has been reported very rarely. In 1964, the first case was reported by Raskin et al.[2] Since then, more than 40 cases of intrahepatic portosystemic venous



a b

Fig. 2. a Portogram through the superior mesenteric artery showing slender portal vein. A dilated shunt vessel flows into the left renal vein (*LRV*), forming a shunt with the finely depicted inferior vena cava (*IVC*). **b** When the shunt was occluded by a balloon, the blood flow from the portal vein (*PV*) to the liver increased

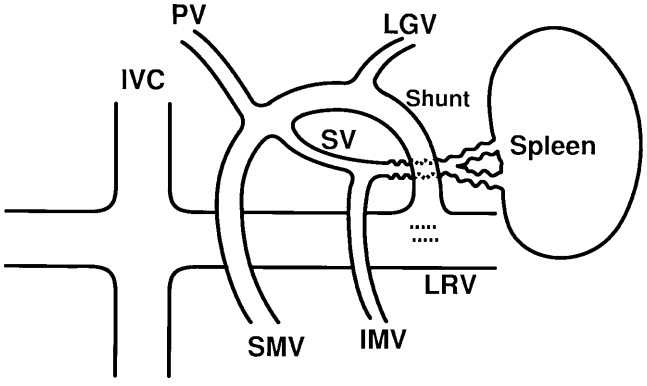


Fig. 3. Shunt vessel during operation. The shunt vessel branched from the superior mesenteric vein (*SMV*) and communicated with the left gastric vein (*LGV*) and the tortuous splenic vein (*SV*), subsequently flowing into the left renal vein (*LRV*). *IMV*, Inferior mesenteric vein

Fig. 4. Microscopic findings of the excised liver specimen indicate considerable fatty changes, consisting of large and small fatty drops. The portal area was slightly infiltrated by lymphocytes; however, cirrhosis or hepatitis was not apparent. ×20

shunt have been reported in the literature. Compared with congenital intrahepatic portosystemic venous shunt, congenital extrahepatic portosystemic venous shunt seems to have been reported less, frequently, with 18 cases, including the present case, reported, as far as we know.

According to Ohwada et al.,[3] the diagnostic criteria for congenital portosystemic shunt can be summarized as follows: (1) There are no remarkable microscopic changes in liver specimens, such as those of hepatitis, cirrhosis, or IPH. (2) The patient has no hypersplenism and no portal hypertension. (3) Portograms shows a hypoplastic portal vein with no arterioportal fistula. (4) There is no previous history of abdominal surgery or of inflammation such as pancreatitis.

In our patient, orthostatic disturbance appeared suddenly in 1993, and hepatic encephalopathy with hyperammonemia was considered to be the cause shortly thereafter. Further, neither liver dysfunction nor splenomegaly was observed at that time. Although conservative treatment had been continued for about 5 years until she was admitted to our hospital, her serum AST and ALT levels were in the range of 40–100 IU/l, and splenomegaly, coagulopathy, and thrombocytopenia (platelet count, 7×10^4–$14 \times 10^4/\mu l$) were observed after admission. Our patient's features differed from those cited in the criteria of Ohwada et al.,[3] mentioned above, in that she had liver damage with splenomegaly. In the reported cases of congenital extrahepatic portosystemic venous shunt, liver damage was reported only by Norimoto et al.,[4] and slight splenomegaly was reported by Ohkubo et al.[5]

What contributed to the liver damage with slight splenomegaly in our patient? First, no abnormality had been shown in the patient's liver function until the time

that hepatic encephalopathy appeared, suggesting that effective blood flow volume in the hepatic circulation could be preserved. On admission, slight splenomegaly appeared on CT images, although no esophageal or gastric varices were detected by upper gastrointestinal endoscopic examination. After shunt occlusion, using a balloon technique during abdominal angiography, the portal blood flow increased considerably. In addition, considerable fatty change in the liver was observed in the excised liver specimen. Considering these results, her clinical course, and the histological findings of the liver, it seems unlikely that a basic disease that may have caused portal hypertension existed before the onset of the hepatic encephalopathy. It was therefore presumed that an effective circulatory flow capable of maintaining liver function had been preserved for some time, after which the blood flow volume of the shunt began to increase, resulting in secondary liver damage caused by ischemic changes. From the variations in her serum AST and ALT levels, the possibility of an increase/decrease in the blood flow volume of the shunt was also considered. In addition, the low-protein diet she received for preventing hepatic encephalophathy may have played a role in the induction of fatty change in the liver. Concerning the slight splenomegaly we observed, an influence of liver dysfunction was suggested from the observed tortuous splenic vein; however, the possibility of splenic congestion caused by altered blood flow through a complicated shunt network, without portal hypertension, was strongly considered. In fact, changes in the platelet count were observed with histologically confirmed splenic congestion. On the other hand, there was no finding suggestive of thrombosis.

The additional diagnostic possibilities included IPH. Compression of portal branches and non-cirrhotic fibrosis are characteristically shown in liver specimens of IPH, but IPH is often observed with various histological findings. In this patient, however, the diagnostic criteria for IPH did not seem to be fulfilled. Based on the evidence mentioned above, it seemed reasonable to consider a congenital event.

However, the question remains: why does encephalopathy caused by congenital portosystemic venous shunt frequently occur in and after middle age, as seen in most of the reported patients. Although it is possible that the blood flow of the shunt increases gradually with advanced age, as mentioned previously, Kerlan et al.[8] suggested decreased toleration of toxic substances, (such as NH3) in the aging brain. According to Alagille et al.[9] and Eckhauser et al.,[10] the incidence of encephalopathy was reported to be rather low in children with portosystemic shunt for noncirrhotic portal hypertension; this finding supports the idea that symptoms occur late in life.

For this type of encephalopathy, surgical ligation has been, thus far, the predominant treatment. In our patient, however, shunt vessel dissection and splenectomy were performed because of the slight splenomegaly associated with thrombocytopenia. When we consider the altered or increased shunt flow caused by branched collateral vessels, it must be kept in mind that postoperative control may be insufficient by the ligation approach alone. On the other hand, transcatheter embolization has been performed with advanced interventional radiology (IVR), which can be performed in massive collateral vessels as well.[11,12]

Conservative treatment for hepatic encephalopathy caused by congenital extrahepatic portocaval shunt is necessary to diminish the symptoms over a long period of time; however, it is difficult to control serum ammonia levels while maintaining the patient's quality of life. In order to avoid the nutritional disturbance caused by a low-protein diet, and possible ischemic changes caused by continuously decreased hepatic blood flow volume, it should be noted that early surgical intervention and IVR are recommended in some patients after the onset of this disease.

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