

Portosystemic Encephalopathy without Liver Disease Masquerading as Dementia

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ABSTRACT

An 84-year-old woman was hospitalized due to consciousness disorder as hyperammonemia. She had no etiology of liver disease. Twelve months before the current admission, she had been diagnosed with dementia based on her low level of daily perception and physical activity. Abdominal computed tomography revealed a large portosystemic shunt between the medial branch of the portal vein and middle hepatic vein. After the improvement of her consciousness disturbance by medical treatment, percutaneous shunt embolization was electively performed. The patient showed a remarkable clinical improvement. Consciousness disturbance caused by hyper-ammonemia might be underlying in dementia patients. Increase of hepatopetal portal blood flow might have contributed to the improvement of her consciousness disturbance. Embolization of the portosystemic shunt might be more effective for patients without liver disease as in the present case.

Key words: *Portosystemic Encephalopathy, Portosystemic shunt, Dementia*

Portosystemic shunts primarily occur in cases of portal hypertension with liver disease and subsequently can lead to portosystemic encephalopathy (PSE), a reversible brain dysfunction. Although PSE has been reported to develop rarely in patients with portosystemic shunts without liver disease, these patients have often been diagnosed as having dementia or other psychiatric diseases²⁾. We describe a case of PSE without liver cirrhosis which was diagnosed as dementia and showed a remarkable clinical improvement after shunt obliteration.

CASE REPORT

Case:

The patient was 84 years old woman. She had no etiology of liver disease. She was often referred to as dull. Twelve months before the current admission, she was diagnosed as having dementia based on her Mini-Mental State Examination (MMSE) score 22. Her family found her after losing consciousness in their home and she was rushed to our hospital while unconscious. Then, her degree of awakening was 10 (eye opening 3,

verbal response 3, motor response 4) on the Glasgow Coma Scale (GCS) and biochemical examination indicated hyperammonemia. We diagnosed unconsciousness caused by hyperammonemia and instilled a branched-chain amino acid preparation. After that, she recovered consciousness. Abdominal computed tomography (CT) revealed a large portosystemic shunt between the medial branch of the portal vein and middle hepatic vein (Fig. 1).

Physical findings:

Height 148.5 cm, Weight 55.5 kg, GCS-10, flapping tremor positive, anemia and jaundice not seen in palpebral conjunctiva or bulbar conjunctiva, flat and soft abdomen, back pain negative, liver and spleen not palpated, leg edema negative.

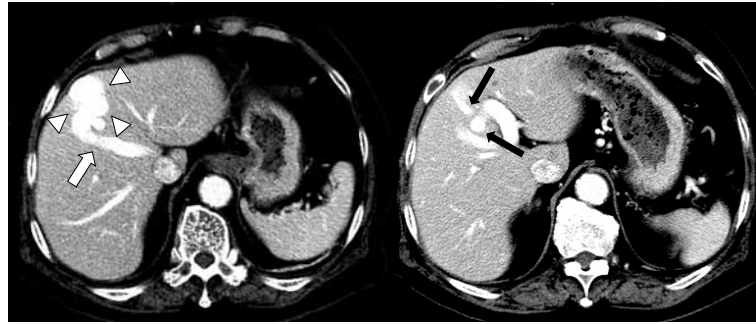
Blood and urine examinations at the hospitalization are shown in Table.

Increase of serum ammonia, blood sugar, blood urea nitrogen, and serum creatinine were noted. Although the levels of serum albumin and platelets were decreased, there were no markers of underlying liver disease.

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A. Pre Embolization



B. Post Embolization (4 months)

Fig. 1. Portal phase of enhanced CTs before and after embolization

(A) Pre-embolization.

There was a large shunt (arrowheads) between two medial branches of the portal vein (large and small black arrows) and middle hepatic vein (large white arrows).

(B) Four months after embolization.

The portosystemic shunt was completely embolized. Arrows show coils at the medial branches and the shunt.

Table 1. Blood examination at the hospitalization

CBC	AST	13 IU/ liter	Virus Markers
WBC 15800 / μ l	ALT	17 IU/ liter	HBs antigen 0.00 IU/ml
RBC 4.28×10^6 / μ l	LDH	219 IU/ liter	HCV antibody 0.1 C.O.I
MCV 89.7 fl	ALP	282 IU/ liter	Autoimmunity Markers
MCH 31.3 pg	γ -GTP	25 IU/ liter	ANA < $\times 40$ times
MCHC 34.9 %	Na	147 mEq/ liter	AMA2 7.0 U/ml
Hb 13.4 g/dl	K	4.7 mEq/ liter	Tumor marker
Ht 38.4 %	Cl	114 mEq/ liter	AFP 8.5 ng/ml
Plt 14.5×10^3 / μ l	TP	7.1 g/dl	DCP 17 mAU/ml
	Alb	3.5 g/dl	
Blood coagulation test	BUN	59.1 mg/dl	
PT 77.1 %	Cr	0.99 mg/dl	
PTT 28.4 S	CRP	2.26 mg/dl	
APTT 28.4 S	TC	198 mg/dl	
fib 260 mg/dl	TG	115 mg/dl	
	FBS	231 mg/dl	
Blood Chemistry	HbA1c	6.4 %	
T-Bil 1.1 mg/dl	NH3	243 μ g/ml	
D-Bil 0.3 mg/dl	CPK	702 U/ liter	
	AMY	28 U/ liter	

AFP, a-fetoprotein; DCP, des- γ -carboxy prothrombin; Alb, albumin; ALP, alkaline phosphatase; ALT, alanine aminotransferase; AST, aspartate aminotransferase; Cr, creatinine; CRP, c-reactive protein; FBS, fasting blood sugar level; Hb, hemoglobin; LDH, lactate dehydrogenase; Plt, platelets; PT, prothrombin time; RBC, red blood cells; T-Bil, total bilirubin; D-Bil, direct bilirubin; TC, total cholesterol; WBC, white blood cells; ANA, Anti-nuclear antibody; AMA2, anti-mitochondrial M2 antibody.

CLINICAL COURSE

After admission to our hospital, her consciousness became clear. The diagnosis of PSE was confirmed by elevated serum ammonia levels of 243 µg/dl (normal, < 66) and a portosystemic shunt between the medial branch of portal vein and middle hepatic vein on abdominal enhanced CT. We considered that a portosystemic shunt might influence her competency for affiliation because she had no etiology of liver disease. We electively performed shunt obliteration.

The shunt embolization procedure was as follows. Before embolization, portal blood pressure

was directly measured as 9 mmHg. Sandwich embolization technique was used for obliteration of the large portosystemic shunt. First, for stopping outflow from the shunt, an Amplatzer vascular plug II of 20 mm diameter was deployed at the middle hepatic vein through a right jugular vein approach. Secondly, for stopping the inflow into the shunt, nine detachable microcoils and two pushable microcoils from 7 mm to 14 mm diameter were placed at the medial branches of portal vein through a transhepatic direct approach of a right portal branch. Shunt obliteration was completely achieved without complications (Fig. 2).

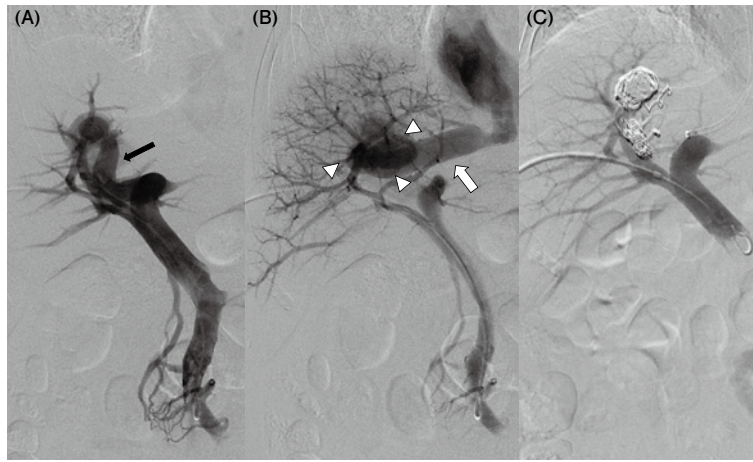


Fig. 2. Portographies before and after embolization

(A) Early and (B) delayed phase of pre-embolizations.

There was an aneurysmal change of shunt (arrowheads) between the dilated medial branches of portal vein (black arrow) and middle hepatic vein (white arrow).

(B) After embolization.

The portosystemic shunt was completely obliterated.

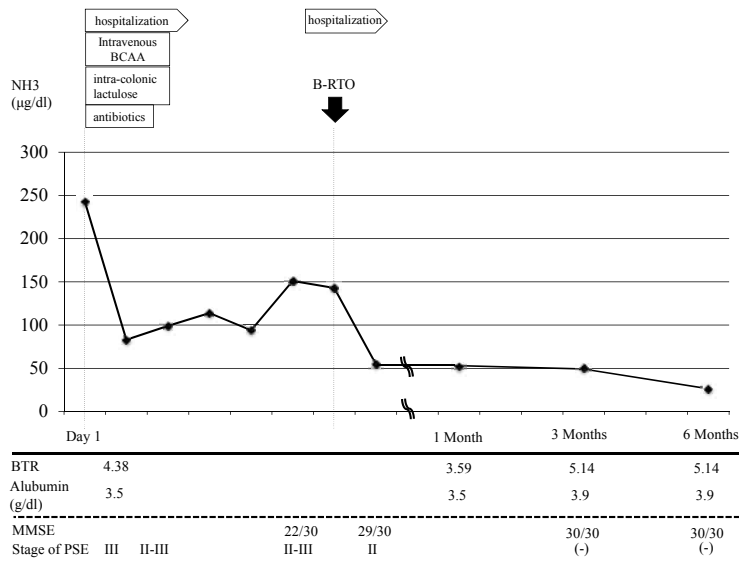


Fig. 3. Clinical course

Although the level of consciousness was improved with intravenous BCAA, intra-colonic lactulose, and antibiotics after the first admission, the low competency for affiliation and hyperammonemia remained. However, after shunt obliteration, the hyperammonemia disappeared, and both the score of Mini-Mental State Examination (MMSE) and the stage of PSE were dramatically improved. BTR (branched chain amino acids / tyrosine molar ratio) and Serum albumin were also raised.

Figure 3 shows the clinical course. After obliteration of the shunt, the hyperammonemia disappeared. Both the MMSE score and PSE stage were dramatically improved. Six months after shunt obliteration, (Cancel: there was no recurrence of portosystemic shunt nor appearance of ectopic shunt) the patient is living a completely independent life.

DISCUSSION

The present case of PSE occurred in a patient without liver disease, who had been continuously treated for dementia. Other cases complicated by both PSE and dementia have been reported, as in our case^{1,5)}. However, it is difficult to diagnose their pathophysiology correctly, because they show poor findings of chronic liver disease and tend to focus on dementia or some kind of psychiatric disease. Watanabe et al⁷⁾ reported characteristics suspiciously like those of PSE in patients without liver disease, as below: (a) High serum ammonia levels with no or slightly abnormal liver function. (b) The repeated development of psychiatric-like symptoms. (c) Abnormally large vessels connected with portal flow detected on enhanced abdominal CT. In the present case, we found all of these characteristics and they provided useful clues for the diagnosis. The apparent reasons for late diagnosis were that the patient had no history of liver disease, serum ammonia was not routinely measured, and PSE was not suspected.

It is reported that the etiology of PSE without liver cirrhosis includes congenital abnormalities in the intrahepatic vascular system, degeneration of hepatic parenchyma and anastomosing vascularization after abdominal surgery, liver biopsy or trauma²⁾. In the present case, the patient received abdominal surgery for colon cancer 13 years ago, and showed abnormally large vessels connected by the intrahepatic portal vein. When we consider this, the etiology of the present case might be related to congenital abnormalities in the intrahepatic vascular system. If so, we should consider why PSE never occurred before this episode. Ammonia is mainly metabolized in the urea cycle in the liver. In addition to the liver, for the ammonia flux, skeletal myocytes provide ammonia metabolism by incorporating ammonia into glutamine via glutamine synthetase³⁾. The reason for hyperammonemia in this case might be the decrease of skeletal myocytes due to aging or lower activity in daily life.

The utility of an intrahepatic portosystemic shunt for embolization has been reported^{4,6,8)}. Six months after embolization, significant improvement was observed in the patient's clinical status, and no signs of recurrent hepatic encephalopathy or com-

plaints due to portal hypertension were registered. Furthermore, BTR (branched chain amino acids / tyrosine molar ratio) and serum albumin were raised. We presume it reflects an increase of the portal flow volume into the liver parenchyma.

CONCLUSION

In a case of unstable dementia accompanied by symptoms similar to PSE, the possibility of a portosystemic shunt and hyperammonemia should be considered in spite of the presence or absence of underlying liver disease.

The embolization of a portosystemic shunt might be more effective, especially for patients without liver disease.

All authors declare that they have no conflict of interest (COI).

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