Journal of the Neurological Sciences 346 (2014) 303–306

Contents lists available at ScienceDirect

Journal of the Neurological Sciences

journal homepage: www.elsevier.com/locate/jns

Short communication

Long-term clinical and radiological improvement of chronic acquired hepatocerebral degeneration after obliteration of portosystemic shunt: Report of a case

Shin Hisahara [a,*,1], Takashi Matsushita [a,1], Mizuki Kitamura [a,b,1], Shinichi Mezawa [c], Michio Nonaka [a,d], Tomihiro Imai [a,e], Shun Shimohama [a]

[a] Department of Neurology, School of Medicine, Sapporo Medical University, Sapporo 060-8556, Japan
[b] Department of Neurology, Yokohama Rosai Hospital, Yokohama 222-0036, Japan
[c] Department of Gastroenterology, Sapporo Kyoritsu Hospital, Sapporo 007-0848, Japan
[d] Department of Neurology, Hokuyukai Neurological Hospital, Sapporo, 063-0802, Japan
[e] Department of Occupational Therapy, School of Health Science, Sapporo Medical University, Sapporo 060-8556, Japan

ARTICLE INFO

Article history:
Received 22 January 2014
Received in revised form 2 July 2014
Accepted 29 July 2014
Available online 15 August 2014

Keywords:
Chronic acquired hepatocerebral degeneration
Cerebellar ataxia
Portosystemic shunt
Liver disease
MRI
Neuroradiology

ABSTRACT

Neurological manifestations are common in patients with decompensated cirrhosis. The majority of these patients show hepatic encephalopathy or chronic acquired (non-Wilsonian) hepatocerebral degeneration (CAHD). They characteristically present with dysarthria, ataxia, involuntary movements, and altered mental status. Neuroradiological examination in patients with hepatic encephalopathy often shows abnormal signals in multiple regions of the brain, such as the pallidum, putamen, caudate nucleus, hemispheric white matter, and ventral midbrain. The pathogenesis of hepatic encephalopathy and CAHD is poorly understood and the response to conventional therapies is often poor.

We report a male patient with cirrhosis of unknown cause, who developed slowly progressive cerebellar truncal and limb ataxia and slurred speech. Magnetic resonance imaging (MRI) showed focal T2 hyperintensity in bilateral dentate nuclei and middle cerebellar peduncles (MCPs). After treatment by obliteration of the portosystemic shunt, clinical manifestations and MRI abnormalities were dramatically improved. He was followed for six years until he died of uncontrollable bleeding due to hepatocellular carcinoma. At the last examination 9 months before death, he showed no apparent aggravation of neurological symptoms, and no abnormal signal intensities in the MCPs and supratentorial compartment. The clinical course and changes of brain MRI findings of this case are extremely rare, suggesting that obliteration of the portosystemic shunt may be effective for CAHD over long term.

1. Introduction

Chronic acquired (non-Wilsonian) hepatocerebral degeneration (CAHD) is an uncommon but clinically significant complication of liver cirrhosis [1–3]. Diverse clinical manifestations of CAHD have been described, such as involuntary movements, cognitive dysfunction, parkinsonism, and cerebellar ataxia. The existence of portosystemic shunts is likely associated with deterioration of clinical condition. Neuroradiological studies have indicated hyperintensity in the globus pallidus on T1-weighted MRI images [4]. Some previous reports have shown CAHD patients presenting ataxia with bilateral middle peduncle lesions, but the correlation between ataxia and brain images has not been entirely elucidated [5–8]. Manganese deposition in the brain seems to play a pivotal role for the pathogenesis of CAHD [4]. While liver transplantation is considered to be the only effective therapy for CAHD [9], some patients did not respond to transplantation or showed deterioration of neurological deficits after transplantation [10,11].

In this report, we describe a patient who had cirrhosis and manifested slowly progressive cerebellar ataxia. After treatment by obliteration of the portosystemic shunt, ataxia and MRI abnormalities in the middle cerebellar peduncle were dramatically improved. The patient showed no apparent aggravation of neurological symptoms and neuroradiological examinations for six years.

2. Case report

A 67-year-old Japanese man was admitted with a three-year history of gradually progressive gait instability and speech difficulty.

* Corresponding author at: Department of Neurology, School of Medicine, Sapporo Medical University, South 1, West 17, Chuo-ku, Sapporo 060-8556, Japan. Tel.: +81 11 611 2111x3821; fax: +81 11 622 7668.
E-mail address: hisahara@sapmed.ac.jp (S. Hisahara).
[1] These three authors contribute equally to this work.

http://dx.doi.org/10.1016/j.jns.2014.07.068
0022-510X/

Approximately two years before admission, he was diagnosed with chronic subdural hematoma based on a tendency to fall and underwent surgical treatment. Four months before admission, the patient needed a wheelchair. At age 59, he was found to have abnormal liver function and diagnosed with cirrhosis of unknown etiology. There was no history of alcoholism and hepatitis B or C viral infection. No familial hepatic or neurological diseases were reported. He had no special preference for protein- and fat-rich foods such as beans and peanuts. At ages 61 and 64, he underwent endoscopic injection sclerotherapy for esophageal varices.

Physical examination revealed no Kayser–Fleischer rings. Neurological examination showed no disturbance of consciousness but mild cognitive dysfunction. No abnormal posture and no spontaneous movements including asterixis were observed. He showed severe ataxic dysarthria characterized by scanning and explosive speech. He was unable to stand and walk alone due to truncal and limb ataxia. Deep tendon reflexes were symmetric and hyperactive. Babinski's sign was present bilaterally. There was no evidence of extrapyramidal signs and sensory disturbance.

Laboratory findings showed hepatic dysfunction, including an ammonium level of 122 (normal range: 20–70) μg/dL, total bilirubin level 1.6 (0.2–1.2) mg/dL, and type IV collagen level 155 (0–140) ng/mL. Fischer ratio was decreased to 1.08 (2.6–4.5). Although serum ceruloplasmin was slightly decreased (17.5 mg/dL: normal range 21–37), serum copper and cerebrospinal fluid protein were normal. Serum cholesterol and triglyceride were also normal. An abdominal computerized tomography scan showed splenomegaly, cirrhosis, splenorenal shunt, paraumbilical vein, and dilatation of portal and esophageal veins (Fig. 1A). An electroencephalogram revealed poorly organized background activity with posteriorly dominant α rhythm and intermittent irregular delta waves. T2-weighted MRI showed mild cortical atrophy and hyperintense lesions in deep white matter, indicating chronic ischemia. Additionally, MRI study revealed symmetric hyperintense lesions in bilateral dentate nuclei and middle cerebellar peduncles (MCPs) (Fig. 2A). Although slightly hyperintense lesions on T1-weighted images were found in bilateral putamen, there were no obvious lesions in the globus pallidus, cortex and brain stem (Fig. 2D). Single photon emission computed tomography (SPECT) depicted no significant decreases of cerebral and cerebellar blood flow. Therefore, CAHD was diagnosed. Treatment with hepatic protector together with intravenous and oral branched-chain amino acids (BCAA) failed to relief the symptoms. He then underwent partial splenic artery embolization and percutaneous transhepatic obliteration (PTO) to occlude the splenorenal shunt. Three months after the procedure, dysarthria and ataxia improved dramatically. He was able to walk with a mildly wide-based gait. Fischer ratio improved to 1.44. The area of hyperintense signal in MCPs decreased markedly (Fig. 2B). Putaminal abnormal signal showed little change (Fig. 2E). The patient was prescribed oral BCAA. There were no elevations of transaminases and ammonia. Two years later, he was able to walk with a wide-based gait, but did not need any assistance in his house. Three years later, an abdominal CT showed no obvious progression of cirrhosis and no re-exacerbation of varices and portosystemic shunt (Fig. 1B). At 71 years of age, he was diagnosed with hepatocellular carcinoma and treated with transcatheter arterial chemoembolization. Although blood ammonia level fluctuated

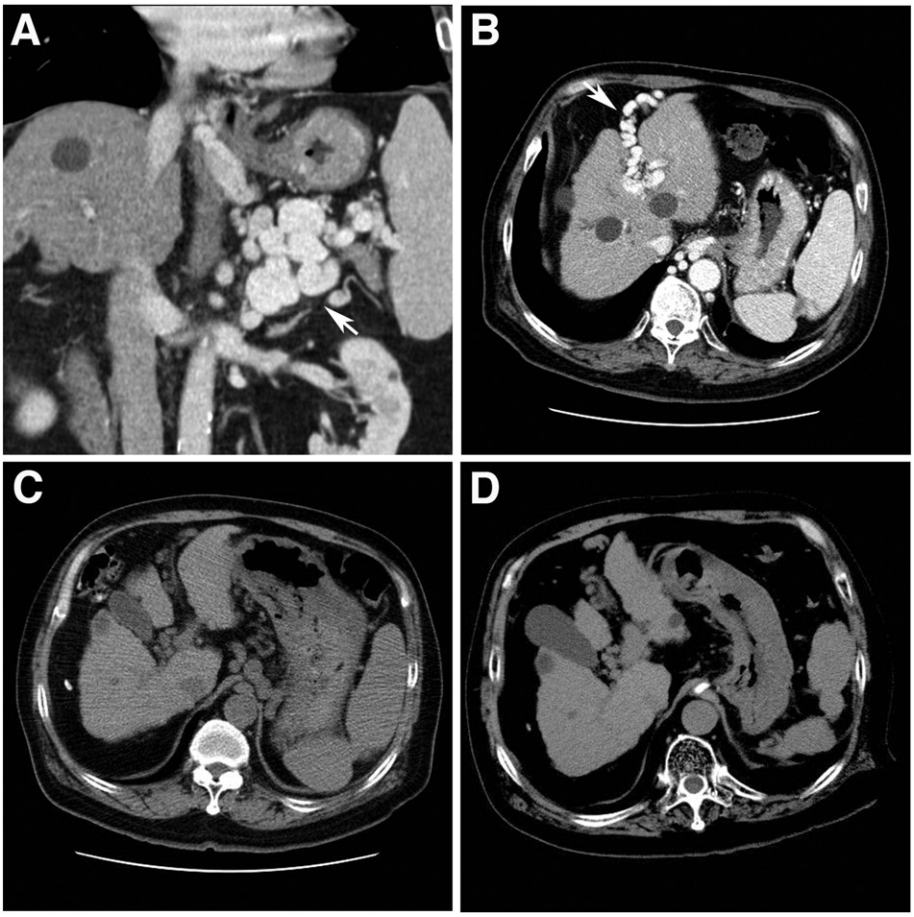

Fig. 1. Representative enhanced abdominal CT images on admission (A and B). These images show significant liver atrophy with multiple cysts, splenomegaly, a splenorenal shunt (arrow in A), and dilated paraumbilical vein (arrow in B) indicating cirrhosis. Plain CT images on admission (C) and three years after portosystemic shunt obliteration (D). Compared with the image on admission, no obvious progression of liver atrophy and splenomegaly is observed three years later.