Hemodialysis-related Portal-Systemic Encephalopathy

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Abstract

Hemodialysis-related portal-systemic encephalopathy (HRPSE) is characterized by the presence of porto-systemic encephalopathy without liver dysfunction, usually caused by changes in the systemic venous flow related to hemodialysis. We herein describe the case of a 75-year-old woman who developed hepatic encephalopathy five years after the initiation of hemodialysis. Abdominal contrast-enhanced computed tomography (CT) and three-dimensional CT angiography revealed a portosystemic venous shunt, and the patient was diagnosed with portosystemic encephalopathy. Occlusion therapy ameliorated her disturbance of consciousness. HRPSE should be recognized as a treatable neuropsychiatric disorder.

Key words: portal-systemic shunt, encephalopathy, hemodialysis, Parkinsonism, occlusion therapy


Introduction

There are various causes of disturbance of consciousness in patients undergoing hemodialysis, including uremic encephalopathy, drug-induced encephalopathy and cerebral stroke (1-3). Hemodialysis-related portal-systemic encephalopathy (HRPSE) can also result in impaired consciousness in hemodialysis patients (4). HRPSE is characterized by the presence of portosystemic encephalopathy without liver dysfunction, usually caused by changes in the systemic venous flow related to hemodialysis. However, this disease may be overlooked since its features are not well known. We encountered a case of HRPSE in a 75-year-old woman who developed repeated disturbances of consciousness, asterixis and Parkinsonism following the initiation of hemodialysis therapy. The aim of this case report is to discuss the features of HRPSE and summarize the findings of earlier cases.

Case Report

A 75-year-old woman was admitted to our department with a disturbance of consciousness. On the day of admission, her consciousness level had fluctuated following the administration of hemodialysis. This was her first episode of disturbance of consciousness. She had been previously diagnosed with hypertension, diabetes mellitus and diabetic nephropathy with end-stage renal dysfunction, and hemodialysis had been initiated five years before the current admission. On the initial examination, her body weight, blood pressure and heart rate were 59.6 kg, 145/82 mm Hg and 68 beats/min, respectively. She was afebrile, and a physical assessment revealed no significant findings. However, she displayed an impaired consciousness, and a neurological examination showed symmetrical rigidity of the neck and bilateral extremities, with asterixis of both feet. The tendon reflexes, muscle tone and sensory examination findings were normal. A blood analysis performed under hemodialysis revealed no abnormalities, except for high serum ammonia (179 μg/dL; normal =0-80) and creatinine (3.5 mg/dL; normal =0.4-1) levels. The white blood cell count was 4,700/μL, red blood cell count was 2.38×10⁶/μL and hemoglobin concentration was 8.3 g/dL. The platelet count was normal. The aspartate aminotransferase level was 26 IU/L and the alanine aminotransferase level was 9 IU/L. The electrolyte levels were as follows: sodium, 137 mEq/L; potassium, 3.7 mEq/L; and chloride, 105 mEq/L.

The patient’s serum was negative for anti-hepatitis C vi...
rus antibodies (according to an enzyme immunoassay) and hepatitis B surface antigens. Abdominal computed tomography (CT) revealed no significant finding in the liver, and her urine was negative for glucose and positive (1+) for proteins. Head CT and radiography of the chest yielded normal findings. However, T1-weighted magnetic resonance imaging with 1.5 Tesla showed findings typical of hepatic encephalopathy, including symmetrical hyperintense areas in the globus pallidus (Fig. 1). An electroencephalogram also demonstrated triphasic waves characteristic of hepatic encephalopathy. In order to identify the cause of the consciousness disturbance, we performed CT of the abdomen, with and without contrast enhancement, which disclosed the presence of a large portosystemic venous shunt (Fig. 2). Furthermore, three-dimensional CT angiography showed a large shunt between the left splenic and left renal veins (Fig. 3).

The administration of balanced-chain amino acids partially mitigated the patient’s disturbance of consciousness, and the serum ammonia level decreased to nearly the normal range (Fig. 4). Her clinical history, symptoms, physical examination findings, head CT features and blood test data excluded the possibility of uremic encephalopathy, drug-induced encephalopathy, cerebral stroke, electrolyte imbalances, hypovolemia and blood pressure fluctuations. During hospitalization, her level of consciousness fluctuated, and the serum ammonia concentration increased to 320 μg/dL after five days of constipation (Fig. 4). We therefore diagnosed her with HRPSE and applied catheter therapy to occlude the abnormal shunt vessel (Fig. 5). The impaired consciousness and Parkinsonism were ameliorated, and she has since experienced no episodes of recurrence of encephalopathy.

**Discussion**

We reviewed 13 cases of HRPSE (Table) (4-15). Of these patients, nine were women and the average patient age was 62.2 years (range, 32-80 years). The interval between the initiation of hemodialysis and onset of HRPSE ranged from one month to 15 years. The most common cause of kidney dysfunction was diabetic nephropathy, and the majority of the abnormal shunt vessels were linked with the left renal vein via extrahepatic veins, such as the left gastric, splenic
and superior mesenteric veins. A history of abdominal surgery was present in only three cases. Surprisingly, all cases involved Asian individuals, with 92% of the patients being Japanese. We suspect that this finding may be attributed to the large number of patients undergoing hemodialysis therapy (approximately 300,000 in 2011) in Japan.

In Japan, the onset of encephalopathy due to portosystemic shunt formation after hemodialysis has recently attracted considerable attention (4). These shunt vessels may be congenital or develop spontaneously or following external abdominal injury or surgery (16). In addition, the patient usually remains asymptomatic throughout life. However, in the presence of additional factors, such as hemodialysis-induced hemodynamic changes, the shunt may cause symptoms. In anuric patients undergoing hemodialysis, a large volume of fluid may be rapidly removed within 3-4 hours, inducing a transient decrease in the total body volume, including the venous blood volume. Due to the effects of a decreased intravenous pressure after hemodialysis, the portal venous blood readily flows into the inferior vena cava via large extrahepatic shunts. The mingling of ammonia-rich portal venous blood with the systemic circulation potentially causes central nervous system toxicity, resulting in hepatic encephalopathy. Furthermore, hyperammonemia tends to occur when large amounts of fluid are removed during hemodialysis. Peritoneal dialysis does not typically induce these changes as it involves the very gradual removal of body fluids. Nevertheless, one case of a French patient who devel-

Figure 4. Time course of the patient’s clinical features, treatment and serum ammonium level. The serum ammonium level changed after the administration of balanced-chain amino acid therapy. Hemodialysis may partially decrease the serum ammonium level. Finally, abnormal vessel occlusion therapy ameliorated the patient’s disturbance of consciousness.

Figure 5. (A) Angiography of the superior mesenteric vein showing an abnormal shunt vessel between the left splenic and left renal veins. (B) After occlusion therapy, no abnormal flow was observed.
The authors state that they have no Conflict of Interest (COI).

References


