Acquired hepatocerebral degeneration: A case report

Wei-Xing Chen, Ping Wang, Sen-Xiang Yan, You-Ming Li, Chao-Hui Yu, Ling-Ling Jiang

Acquired hepatocerebral degeneration (AHD) is an exceptional type of hepatic encephalopathies (HE). It is characterized by neuropsychiatric and extrapyramidal symptomatology similar to that seen in hepatolenticular degeneration (Wilson’s disease). In this paper, we report a case of AHD with unusual presenting features.

METHODS: A 28-year-old man with AHD was described and the literature was reviewed.

RESULTS: The man had a history of HBV-related liver cirrhosis. He was admitted to our hospital with apathy, dysarthria, mild consciousness impairment and extrapyramidal symptoms after hematemesis. By review of the literature, cases with AHD often did not present consciousness impairment. So our case was once diagnosed incorrectly as Wilson’s disease.

CONCLUSION: AHD is a rare syndrome and its variable clinical manifestations make it difficult to be diagnosed. But we believe that extensive examination and thorough understanding of the disease are beneficial to a correct diagnosis. Moreover, biocoen is effective in treating the case.

© 2005 The WJG Press and Elsevier Inc. All rights reserved.

Key words: Acquired hepatocerebral degeneration

INTRODUCTION

Acquired hepatocerebral degeneration (AHD) is a rare syndrome characterized by neuropsychiatric and extrapyramidal symptomatology in patients with portocaval shunt. The pathophysiology of this disorder is still unknown, but hyperammonemia and/or brain manganese overload may play a role. Medical treatment is often disappointing. We reported a case of AHD whose clinical features led us to a misdiagnosis at one time.

CASE REPORT

The patient was a 28-year-old man. He had a history of HBV-related liver cirrhosis. No familial hepatic or neurological diseases were reported. Two weeks ago, he had an episode of tarry stool and was cured. But he developed apathy, dysarthria, mild consciousness impairment and involuntary abnormal movements after some days. Physical examination revealed splenomegaly and shifting dullness. Abdominal ultrasonic examination suggested portal hypertension and a great deal of ascites. Cranial magnetic resonance imaging (MRI) revealed increased signal intensity in basal ganglia bilaterally on T2-weighted images similar to hepatolenticular degeneration (Wilson’s disease) (Figure 1). Electroencephalogram (EEG) disclosed diffuse slow wave activity (Figure 2A). Slit lamp examination found no Kayser-Fleischer (K-F) ring of the cornea associated with Wilson’s disease. Copper balance and cerebrospinal fluid examinations gave normal results. Liver failure was grade B-9 of the Child-Pugh classification. The level of serum ammonia was measured several times, and was above the reference range (132 µmol/L) one time. He was treated with hepatic protector tablets, ammonia lowering agents, bowel relaxants and branched chain amino acid infusion. But therapeutic effect was not so good. Finally, after administration of biocoen (coenzyme complex injection), his neuropsychiatric abnormalities and the presentation of EEG were added, but the symptoms did not relieve obviously. Finally, after administration of biocoen (coenzyme complex injection), his neuropsychiatric abnormalities and the presentation of EEG markedly improved (Figure 2B). By 3 mo follow-up, we found that he recovered well and could take care of himself after leaving hospital.

Figure 1 Cranial MRI in the patient with similar hepatolenticular degeneration (Wilson’s disease). A: increased signal intensity in basal ganglia bilaterally on T2WI; B: increased signal intensity in basal ganglia bilaterally on T1WI.

DISCUSSION

AHD is an exceptional type of hepatic encephalopathies (HE). It has been described in patients with severe liver disease of many causes, and notably in patients with surgically or spontaneously induced porto-systemic shunts. But many questions remain unanswered. Its frequency and pathogenesis remain largely uncertain. The clinical, neuroradiological, and biological characteristics of AHD have not yet been fully determined. The response to treatment is also incompletely understood. Our review of the literature (including MEDLINE, from 1981 to April 2003) yielded 36 similar cases[13], among them, 8 (22.2%) were Chinese and 28 (77.8%) were from Western countries.

Studies have indicated that the disease is associated with multiple metabolic insults, such as ammonia, manganese, etc. Especially, the toxic effects of manganese might be the major determinant[13,14]. It has been proved that manganese is cleared by the hepatobiliary system and whole blood and cerebrospinal...
fluid manganese concentrations in some patients with AHD are several fold above the reference range. Thus deposition of manganese in the brain is postulated in patients with AHD, which may induce diffuse degeneration in parenchymal brain. Microscopically, neuronal loss, Alzheimer type II astrocytes and cytoplasmic glycogen granules in basal ganglia are characteristic. The spectrum of clinical presentations can be neuropsychiatric (apathy, lethargy, excessive somnolence, secondary dementia, etc.), extrapyramidal symptoms (focal dystonia, postural tremor, myoclonus, rigidity, dysarthria, choreoathetosis, etc.), or both[1,2,10,14]. But it was also reported that some patients showed increased signal intensity in the dentate nuclei bilaterally on T2-weighted images reported that some patients showed increased signal intensity in vivo which may induce diffuse degeneration in parenchymal brain.

As described above, the disease is characterized by neuropsychiatric and extrapyramidal symptoms developing from chronic hepatic encephalopathies. The patient was irresponsive to the routine therapy of anti-hepatic encephalopathies (such as branched chain amino acids or levodopa therapy). But it has been reported that some patients with AHD are responsive to branched chain amino acids or levodopa therapy. However, the disease develops gradually and the symptoms become progressively worse. Medical treatment is often disappointing. It has also been reported that endovascular occlusion of a porto-systemic shunt is temporarily effective. Moreover, liver transplantation in selected cases could be curative. is that AHD also distinct from the more acute and transient episodes of HE. The neurological symptoms of HE disappear when the disease relieves, and there is no organic damage in HE[11]. The disease develops gradually and the symptoms become progressively worse. Medical treatment is often disappointing. But it has been reported that some patients with AHD are responsive to branched chain amino acids or levodopa therapy. It has also been reported that endovascular occlusion of a porto-systemic shunt is temporarily effective. Moreover, liver transplantation in selected cases could be curative. is that AHD might be a reversible and treatable disorder partly[1,2,10,14].

REFERENCES

1. Burkhard PR, Delavelle J, Du Pasquier R, Spahr L. Chronic parkinsonism associated with cirrhosis: a distinct subset of acquired hepatocerebral degeneration. Arch Neurol 2003; 60: 521-528

2. Condat B, Dusoleil A, Bernardneau M, Roche A, Pelletier G, Buffet C. Chronic acquired hepatocerebral degeneration: the role of manganese and treatment by endovascular occlusion.
of a porto-systemic shunt. Gastroenterol Clin Biol 1999; 23: 268-270

3 Jog MS, Lang AE. Chronic acquired hepatocerebral degeneration: case reports and new insights. Mov Disord 1995; 10: 714-722

4 Levy VG, Cameron E, Ollat H, Opolon P, Darnis F, Contamin F. Chronic hepatic encephalopathies. Acquired cerebral degeneration not due to Wilson’s disease. Sem Hop 1983; 59: 1369-1373

5 Spencer DC, Forno LS. February 2000: Dementia with motor dysfunction in a patient with liver disease. Brain Pathol 2000; 10: 315-316, 319

6 Soffer D, Sherman Y, Tur-Kaspa R, Eid A. Acquired hepatocerebral degeneration in a liver transplant recipient. Acta Neuropathol 1995; 90: 107-111

7 Spitaleri DL, Vitolo S, Fasanaro AM, Valiani R. Choreaathetosis. Uncommon manifestation during chronic liver disease with portocaval shunt. Riv Neurol 1983; 53: 293-299

8 Thobois S, Giraud P, Debat P, Gouttard P, Maurizi A, Perret-Liaudet A, Kopp N, Broussolle E. Orofacial dyskinesias in a patient with primary biliary cirrhosis: a clinicopathological case report and review. Mov Disord 2002; 17: 415-419

9 Stracciarri A, Guarino M, Pazzaglia P, Marchesini G, Pisi P. Acquired hepatocerebral degeneration: full recovery after liver transplantation. J Neurol Neurosurg Psychiatry 2001; 70: 136-137

10 Ueki Y, Isozaki E, Miyazaki Y, Koide R, Shimizu T, Yagi K, Hirai S. Clinical and neuroradiological improvement in chronic acquired hepatocerebral degeneration after branched-chain amino acid therapy. Acta Neurol Scand 2002; 106: 113-116

11 Wang DY. Clinical analysis of hepatic cirrhosis with chronic hepatocerebral: Clinical analysis of 8 cases. Shanxi Yixue Zazhi 1997; 26: 406-407

12 de Santi MM, Lungarella G, Luzi C, Miracco C, Tosi P. Ultrastructural features in active chronic hepatitis with changes resembling Wilson’s disease. Am J Clin Pathol 1986; 85: 365-369

13 Lee J, Lacomis D, Comu S, Jacobsohn J, Kanal E. Acquired hepatocerebral degeneration: MR and pathologic findings. AJNR Am J Neuroradiol 1998; 19: 485-487

14 Layargues GP. Movement dysfunction and hepatic encephalopathy. Metab Brain Dis 2001; 16: 27-35

15 Finlayson MH, Superville B. Distribution of cerebral lesions in acquired hepatocerebral degeneration. Brain 1981; 104: 79-95

Edited by Kumar M and Wang XL Proofread by Zhu LH