OBSERVATION

Prominent Cerebellar Symptoms With Unusual Magnetic Resonance Imaging Findings in Acquired Hepatocerebral Degeneration

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Background: Cerebellar lesions revealed by abnormal signals on magnetic resonance images are extremely rare in acquired hepatocerebral degeneration (AHCD).

Objective: To report a case of AHCD with prominent cerebellar findings both clinically and radiologically.

Design and Setting: Case report and tertiary-care hospital.

Patient: A 46-year-old man complained of progressive speech difficulties of 5 months' duration. Two years earlier, he had been diagnosed as having cirrhosis of the liver caused by alcoholism and hepatitis B virus infection.

Results: The patient had progressive ataxic dysarthria and limb and gait ataxia as manifestations of AHCD. Magnetic resonance imaging of the brain revealed distinctive symmetrical T2 high-signal intensities in the bilateral cerebellar hemispheres and brachium pontis, which were consistent with his neurologic deficits. Simultaneously, high T1 signals in the bilateral pallidum and ventral midbrain were noted, which are typical manifestations of AHCD. Follow-up magnetic resonance imaging 3 months later showed the same cerebellar signs and abnormal signals.

Conclusions: The cerebellar cortex and middle cerebellar peduncle are considered highly vulnerable structures to metabolic insults in liver disease. Findings from our patient suggest that dominant cerebellar deficits with compatible T2 high-signal lesions are another type of clinical manifestation in AHCD.

Arch Neurol. 2004;61:1458-1460

ACQUIRED HEPATOCEREBRAL degeneration (AHCD) is a chronic and largely irreversible neurologic syndrome associated with acquired liver disease. It is often combined with ataxia, and postmortem studies have shown frequent involvement of the cerebellum. However, direct neuroimaging evidence of cerebellar damage in patients has rarely been reported. In this article, we describe a patient who had AHCD and progressive ataxia in whom abnormal magnetic resonance imaging (MRI) signals were noted in the bilateral cerebellar cortex and brachium pontis.

REPORT OF A CASE

A 46-year-old man complained of progressive speech difficulties of 5 months' duration. He had been well until 2 years earlier, when he was diagnosed as having cirrhosis of the liver caused by alcoholism and hepatitis B virus infection. Since then, he had developed recurrent ascites twice but had never had hepatic encephalopathy. His medical history included diabetes diagnosed 4 months earlier. Physical examination results showed hepatosplenomegaly and spider nevi on the upper chest but no Kayser-Fleischer rings. At neurologic examination he was alert and oriented, and there was no evidence of abnormal spontaneous movements or posture. His facial expressions were limited, and he showed minimal clumsiness during finger tapping tests; however, he had impaired bilateral coordination of the arms and legs as well as gait ataxia on tandem gait tests. This patient's most prominent impairment was ataxic dysarthria characterized by scanning, slurred, staccato, explosive, and hesitant speech. The volume...
and tone of his voice fluctuated, and frequent unnatural breathing interrupted his speech. Neither abnormal muscle tone nor tremor was present. Deep tendon reflexes were symmetric and normoactive, and plantar responses were flexor. Intellectual functions were not impaired; his attentiveness was intact, he had normal verbal and visual memory, and his executive functions were preserved.

Laboratory tests showed normal serum electrolyte, blood urea nitrogen, and creatinine levels. His fasting blood glucose level was as high as 338 mg/dL. His complete blood cell count was normal except for slight thrombocytopenia. Hepatic dysfunction was noted, including an aspartate aminotransferase level of 65 U/L (normal, 13-34 U/L), an alanine aminotransferase level of 58 U/L (normal, 5-46 U/L), a prothrombin time of 16.6 seconds (normal, 10.7-14 seconds), a total bilirubin concentration of 1.5 mg/dL (normal, 0.2-1.2 mg/dL), a serum ammonia concentration of 170 µg/dL (normal, 10-100 µg/dL), and a γ-glutamyltransferase level of 83 U/L (normal, 12-54 U/L). A hepatitis B viral marker assay showed current infection, but his serum folate and vitamin B₁₂ levels were not decreased.

Brain MRI showed increased T1 signals within the bilateral pallidum and ventral midbrain, along with increased T2 signals in the bilateral brachium pontis and cortex of the cerebellum (Figure). Conservative treatments in addition to diabetes control were continued thereafter. A follow-up neurologic evaluation 3 months later using MRI revealed persistence of the previously mentioned cerebellar signs and lesions.

According to postmortem pathologic studies, the involved areas in AHCD are widespread and include the cerebral cortex, subjacent white matter, basal ganglia, and cerebellum. These findings are consistent with the neurologic symptoms commonly present in AHCD, which include combinations of dementia, dysarthria, choreoathetosis, tremor, and ataxia. In most patients, however, abnormal MRI signals are observed only in more restricted areas, including the pallidum, putamen, and ventral midbrain. A high T1 signal in the pallidum is consistently found in patients with AHCD and is considered a neuroimaging marker for advanced liver disease. The extent of abnormal signals has been correlated with the impairment of motor performance. The accumulation of paramagnetic materials, especially manganese, is thought to be a likely mechanism for high T1 signals.

Cerebellar lesions have rarely been observed on MRI studies as abnormal signals, although ataxia frequently appears in AHCD. Although our patient had prominent cerebellar symptoms including progressive ataxic dysarthria and gait and limb ataxia, he had minor extrapyramidal motor symptoms. This type of clinical manifestation is unusual in AHCD. In addition, our patient had no prior episodes of clinical encephalopathy, which usually precede the development of AHCD. The presence of a cerebellar lesion was consistent with his symptoms. We also observed neuroimaging results consistent with AHCD in the pallidum and ventral midbrain, which were minimally relevant to this patient’s clinical symptoms. It is interesting that our patient’s T2 high-signal cerebellar abnormality was more efficient than his T1 abnormality at producing clinical symptoms. To our knowledge, this case is the first report of MRI findings showing focal abnormal cerebellar lesions in AHCD and offering results of simultaneously performed neurologic examination and follow-up. Our results indicate that one possible type of clinical manifestation in AHCD consists of dominant cerebellar deficits with compatible T2 high-signal lesions.

Focal abnormalities in the cerebellum are thought to be due to spongiform degeneration, increased water content, or myelinolysis. Toxic metabolites and osmotic changes caused by liver disease may also play a major role. We cannot exclude the possibility, however, that diabetes and previous alcoholism may have contributed to our patient’s condition. A follow-up examination 3 months later showed that the cerebellar

Figure. A and B, Axial T2- and T1-weighted images show increased T2 and decreased T1 signals in the bilateral cerebellar cortex. C and D, Axial T2- and T1-weighted images demonstrate symmetrically increased T2 and decreased T1 signals in the bilateral cerebellar cortex and middle cerebellar peduncles. E and F, T1-weighted image shows increased signal intensity in the ventral midbrain and globus pallidus.
lesion was still present, suggesting that it is not transient and may be irreversible.

Accepted for Publication: January 7, 2004.
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Author Contributions: Study concept and design: Park and Heo. Acquisition of data: Park and Heo. Analysis and interpretation of data: Park and Heo. Drafting of the manuscript: Park and Heo. Critical revision of the manuscript for important intellectual content: Park. Administrative, technical, and material support: Park and Heo. Study supervision: Park.

REFERENCES


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