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**MR imaging of acquired hepatocerebral degeneration.**

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## MR Imaging of Acquired Hepatocerebral Degeneration

Acquired hepatocerebral degeneration is a well-known irreversible neurologic syndrome that occurs occasionally in patients who have chronic liver disease and portocaval shunting [1, 2]. Affected patients have some combination of coarse postural and action tremor, dyskinesias of cranial predominance, dysarthria, gait and limb ataxia, and variable impairment of intellectual function [1, 2]. Few neuroimaging studies of this condition, which affects the cortex, basal ganglia, and cerebellum, have been performed. We present a patient whose symptomatology correlates with MR abnormalities of the basal ganglia, a finding not previously described.

### Case Report

A 66-year-old woman had a 3-year history of progressive incapacitating tremor of the limbs, speech difficulties, and involuntary orofacial movements. Chronic hepatitis (positive test for hepatitis B surface antigen) had been diagnosed when she was 47 years old. When she was 58, she had her first attack of hepatic encephalopathy. Ten more episodes occurred in the following years before the onset of fixed neurologic symptoms. On repeated examinations, she had a normal mental status, choreic movements of the face and neck, a coarse dysarthria, mild hypokinesia, a large-amplitude postural and action tremor of the limbs, and truncal ataxia. Muscular tone was normal; reflexes were brisk, but plantar responses were flexor. No Kayser-Fleischer rings were present. Liver function tests were altered, but other blood studies, including levels of copper and ceruloplasmin, were normal. CT of the head (Fig. 1A) showed mild cortical atrophy. MR showed abnormal increased signal on T1-weighted images (Fig. 1B), 575/20/2 (TR/TE/excitations), in the corpus striatum, mainly in the globus pallidus, and subthalamic region. On T2-weighted images, 2000/40/90, the same area again showed abnormal signal hyperintensity relative to the brain on the early-echo sequence (Fig. 1C) that faded in the later echo (Fig. 1D). No signal abnormalities were found in the cerebellum.

### Discussion

Acquired hepatocerebral degeneration can occur with all varieties of chronic liver disease; it is associated most frequently with alcoholic cirrhosis or subacute or chronic hepatitis. Portosystemic shunts are always present [1, 2]. Macroscopic examination of the brain shows laminar or pseudolaminar necrosis with microcavitation at the junction of the cortex and white matter, at the striatum, and in the cerebellar white matter. Microscopic findings consist of a diffuse increase in the size and number of protoplasmic astrocytes; intranuclear inclusions (mainly glycogen) in astrocytes; and diffuse but patchy degeneration of neurons and myelin, especially in the deeper cortex and subcortex, basal ganglia, and cerebellum [2, 3]. Pathogenetic mechanisms are speculative. A diffuse astrocytic hyperplasia also occurs after episodic stupor or coma [4].

The history and clinical findings of our patient are typical of acquired hepatocerebral degeneration. To our knowledge, only one previous report [5] of MR imaging of this condition has been published. It described increased signal intensity in the dentate nuclei bilaterally on T2-weighted images without other abnormalities. In our patient, T2-weighted images did not show such an abnormality in the dentate region; however, correlation between the MR findings in the basal ganglia and the clinical state of the patient was good.

We do not know of other normal or pathologic states that result in a similar bilateral T1 hypersignal in the basal ganglia. We suggest that this appearance on MR may be similar to the shortening of T1 caused by intracellular phospholipids in the posterior pituitary gland related to the normal metabolism of the neurohypophysis [6]. On the other hand, a rim of short T1 and T2 frequently is seen in the capsule of brain abscesses. Pathologic studies in these cases have not identified hemorrhage. It has been suggested that the abnormal signal may reflect paramagnetic proton-electron dipole-dipole interaction and the presence of heterogeneously distributed free radicals that are the product of the increased metabolic activity produced by actively phagocytosing macrophages in the capsular wall [7]. Because acquired hepatocerebral degeneration is associated with an

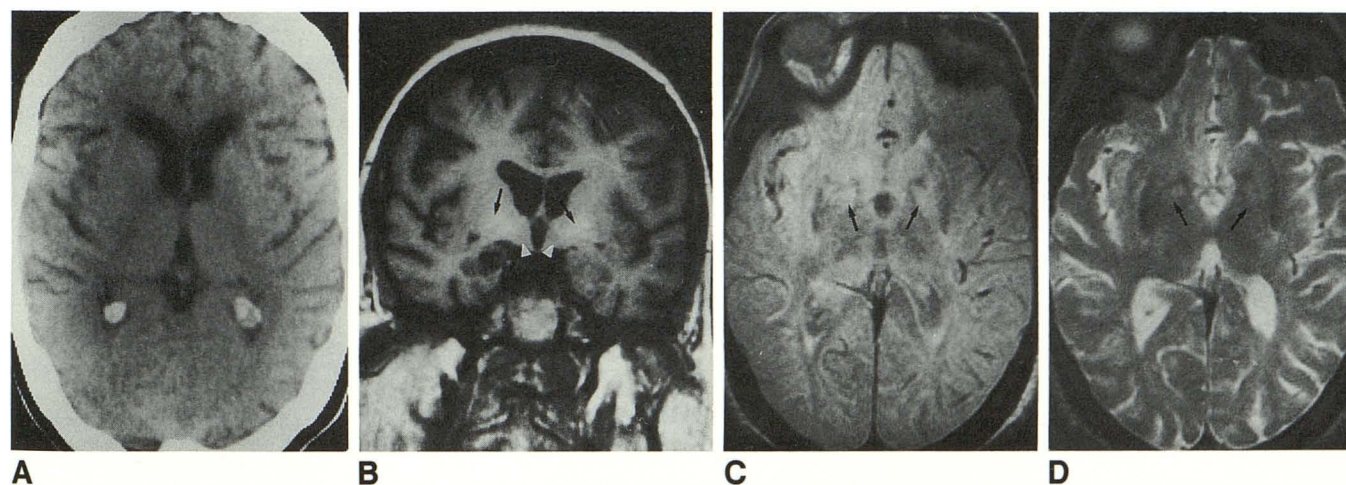


Fig. 1.—Acquired hepatocerebral degeneration.

A, CT scan shows no abnormalities at level of basal ganglia. Note mild ventricular enlargement.

B, Coronal T2-weighted MR image (575/20 at 1.5 T) at level of basal ganglia shows hypersignal in globus pallidus (arrows) and subthalamus (arrowheads).

C and D, On dual-echo MR images (2000/40/90), expected low signal in basal ganglia related to age is replaced by a hypersignal (arrows) in early-echo sequence (C), which persists but diminishes in later-echo sequence (D, arrows).

active process of phagocytosis with proliferation of protoplasmic astrocytes and the presence of large numbers of fatty macrophages [2-4], it seems reasonable to speculate that a similar paramagnetic effect could account for the T1 shortening observed in our case. The exact nature of these images awaits pathologic confirmation.

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