CASE REPORTS

ACQUIRED HEPATOCEREBRAL DEGENERATION: A CASE PRESENTATION WITH CRANIAL MAGNETIC RESONANCE IMAGING FINDINGS

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Gazi Medical Journal 1998; 9 : 136-138

SUMMARY : *We present a 54-year-old male patient who had previously been followed with the diagnosis of postnecrotic cirrhosis for twelve years. During the last 4 years, he had developed progressive dysarthria, tremor, and ataxic gait. Neurological examination revealed a mild right hemiparesis with bilateral cerebellar and pyramidal signs. Cranial magnetic resonance imaging study showed cerebral and cerebellar atrophy, as well as bilateral high intensity signals in dentate nuclei and periventricular white matter in T2 weighted images, reminiscent of Wilson's disease. These white matter lesions can also be erroneously diagnosed as multiple sclerosis plaques.*

Key Words: *Brain, Magnetic Resonance Imaging, Liver Diseases.*

INTRODUCTION

Acquired (non-wilsonian) hepatocerebral degeneration, characterised with permanent and progressive dysfunction of extrapyramidal and cerebellar systems, may occur in the advanced stages of chronic liver disease. Although clinical and laboratory examination findings are helpful in the differentiation from Wilson's disease (1), the underlying pathophysiology has not been clearly elucidated. We report cranial magnetic resonance imaging (MRI) findings of a case with postnecrotic cirrhosis, who presented with dysarthria, bradykinesia and ataxia, that had progressed for 4 years before the final diagnosis of acquired hepatocerebral degeneration was established. MRI findings in this disease have rarely been reported previously.

CASE REPORT

A 54-year-old male patient who had previously been followed with the diagnosis of postnecrotic cirrhosis for twelve years presented with difficulty walking. He reported repeated episodes of hepatic encephalopathy for the last 3 years. Speech was slurred and he progressively lost his ability to perform tasks, requiring skills with his hands in the last 4 years. He developed right-sided weakness a year before admission. Physical examination showed periumbilical venous enlargement suggesting collateral vessel formation, minimal ascites detected by percussion, hepato and splenomegaly. There were no Kayser-Fleischer corneal rings. Neurologic examination revealed intact mental status except for a slight impairment of recent memory. There was a minimal right hemiparesis. Cerebellar tests were abnormal on both sides, with marked intentional tremor and rebound phenomenon in the upper extremities. Deep tendon reflexes were hyperactive. Snout, glabella and palmomental reflexes were present.

Plantar responses were flexor bilaterally. Sensory examination was normal.

Laboratory examination revealed some hematological abnormalities. The platelets were reduced in number: 70,000/mm3 (N: 150,000-400,000). Fibrinogen level was slightly decreased at 153mg/dl (N: 160-415). Fibrin degradation products were increased, ranging from 10 to 40µg/ml (N: <10). These findings were compatible with a diagnosis of disseminated intravascular coagulation within the setting of a chronic liver disease. Total bilirubin and ammonia levels were elevated at 3.5mg/dl(N: 0.3-1) and 180µg/dl(N: 80-110) respectively. The following tests were normal: serum biochemistry profile, bleeding time, partial thromboplastin time, prothrombin time, blood and urinary copper levels and serum ceruloplasmin. An electroencephalogram (EEG) showed slight slowing of the background activity. Cranial computerized tomography revealed cerebral and cerebellar atrophy. Cerebrospinal fluid (CSF) examination, including the absolute IgG level and the IgG index was normal. There were no oligoclonal bands. Visual, brainstem auditory and posterior tibial somatosensory evoked potantials were normal. Cranial MRI showed cerebral and cerebellar atrophy, as well as bilateral high signal intensity in dentate nuclei (Fig. 1). There were periventricular hyperintense lesions of various sizes in T2 weighted images (Fig. 2).

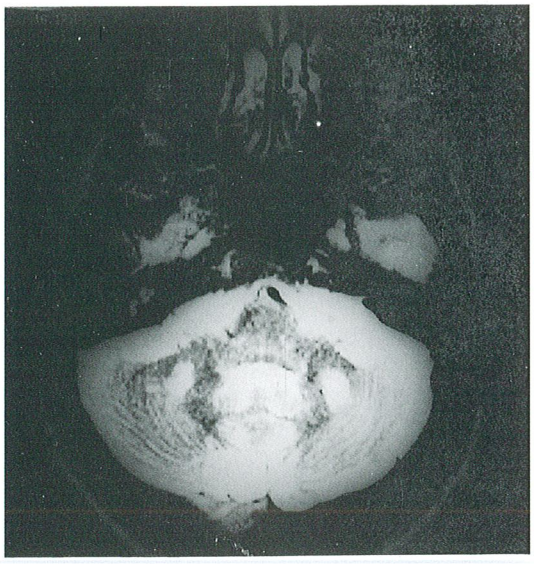

Fig - 1 : Bilateral high signal intensity in dentate nuclei on T2 weighted images.

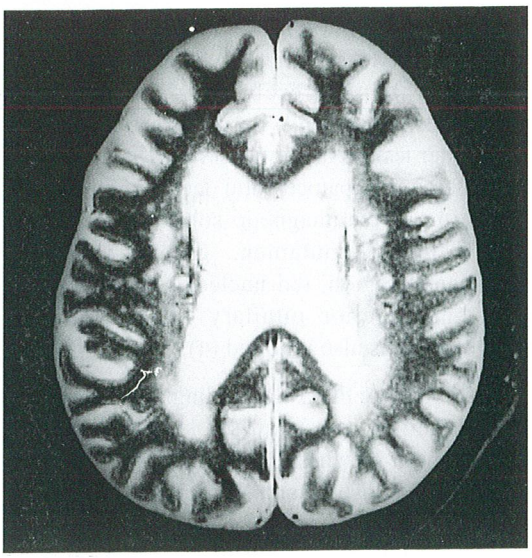

Fig - 2 : Periventricular hyperintense lesions can be mistaken for multiple sclerosis plaques.

DISCUSSION

Acquired hepatocerebral degeneration is characterized by progressive neurological signs; such as dysarthria, ataxia, tremor and dementia during the course of chronic liver disease, with recurrent attacks of hepatic encephalopathy (1-3). Pyramidal tract signs are usually present. EEG background activity is slowed (1). Our patient has all of the clinical and laboratory features typical of this disorder. The presence of chronic and irreversible symptoms rules out hepatic encephalopathy. Absence of a positive family history and Kayser-Fleischer rings, the late onset of symptoms, , normal serum copper, ceruloplasmin and 24 hour-urinary copper excretion levels make Wilson's disease highly unlikely. The clinical picture developing in the setting of chronic liver disease, absence of immunologic abnormalities in CSF examination and normal evoked potential studies rule out multiple sclerosis (MS).

Demyelination and microcavitation are the major histopathological findings of acquired hepatocerebral degeneration (4, 5). MRI findings reflect these lesions situated usually symmetrically within the white matter of the cerebral hemispheres (6). Hyperintense signal abnormalities in dentate nuclei suggest that grey matter is also involved, as previously reported (5, 7). In chronic acquired hepatic failure, T1-weighted images demonstrate increased signal intensity in the basal ganglia, although cerebellum seems to be spared (5).

However, signal abnormalities in the dentate nuclei bilaterally on T2-weighted images can occur in acquired hepatocerebral degeneration, indistinguishable from Wilson's disease (7). Such grey matter lesions are atypical for MS, but can be encountered in hepatocerebral degeneration due to deposition of paramagnetic substances such as copper in the putamen, globus pallidus, subthalamic region, red nucleus, quadrigeminal plate and anterior pituitary (4, 5). Pallidal hypointensity is also reported (4).

We conclude that MRI findings are helpful in the diagnosis of acquired hepatocerebral degeneration, but they can possibly be confused with Wilson's disease and MS plaques.

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