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Clinical Image

Neurological Symptoms in Cirrhotic Patients – Not Always Encephalopathy

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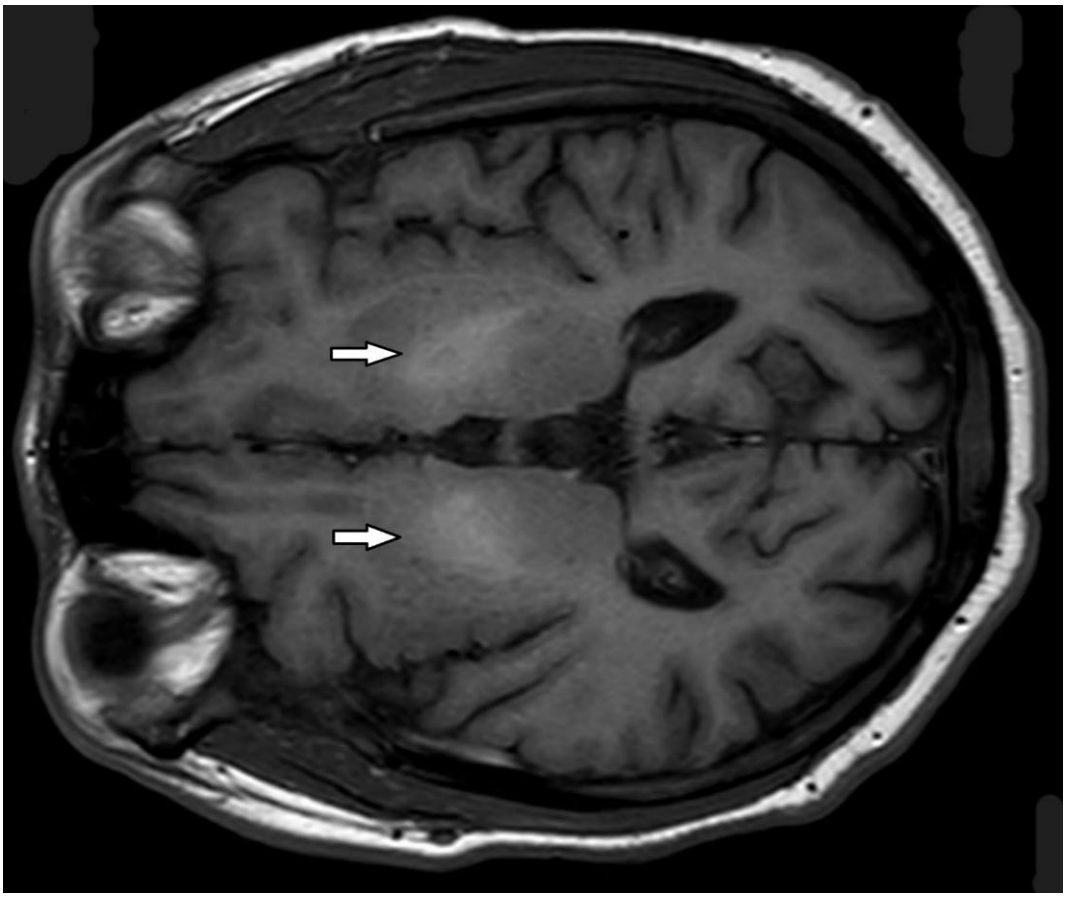

Figure 1: Brain MRI revealing cortical atrophy and hyperintensity in the globus pallidus bilaterally on T1 weighted sequences, suggesting acquired hepatocerebral degeneration in this clinical context.

Chronic liver disease often leads to neuropsychiatric disorders related to decreased protein synthesis and ammonemia [1]. While hepatic encephalopathy (EH) is more common and often reversible, acquired hepatocerebral degeneration (AHD) is rare, difficult to diagnose, and irreversible without liver transplant [2]. When both are present simultaneously, diagnosis is even more challenging [3]. A 39-year-old male with alcohol- and hepatitis B-related cirrhosis was admitted to the emergency department with a 1-week history of disorientation and psychomotor retardation, constipation and upper limb tremor. No bowel movements were reported in the previous 5 days. Physical examination was significant for jaundice, lentification, spatio-temporal disorientation, and mild dysarthria, without language compromise, facial paralysis or motor deficits. Examination also revealed dysmetria, ataxic gait and flapping. Blood tests revealed pancytopenia, hyperbilirubinemia (total bilirubin 7.97 mg/dL, direct bilirubin 1.99 mg/dL), elevated liver enzymes (AST/ALT 53/51 U/L), prolonged prothrombin time and activated thromboplastin time, and ammonia 81 μmol/L. Inflammatory parameters were unremarkable. CA head CT scan did not reveal any abnormalities. Abdominal CT scan documented a splenorenal shunt and portal cavernoma.

The patient was admitted with a diagnosis of hepatic encephalopathy (West-Haven grade II) in the context of constipation. Laxative therapy was optimized, with gradual improvement of neurologic symptoms. However, after 1 week of therapy, dysarthria, ataxia and upper limb tremors were still present and now obviously asymmetric. A brain magnetic resonance imaging (MRI) was performed, revealing cortical atrophy and hyperintensity in the globus pallidus bilaterally on T1 weighted sequences, highly suggestive of manganese deposits, which were compatible with liver dysfunction (Figure 1). Susceptibility-weighted imaging (SWI) and diffusion MRI did not show any relevant findings. Electroencephalograms were negative for signs of epileptic activity. Copper metabolism was normal, and there was no history of drugs or specific metabolic context. Tremor asymmetry and extrapyramidal symptoms, as well as its persistence despite improvement of other neurologic symptoms and normalization of ammonia, was the key to trigger further investigation [4]. MRI was compatible with acquired hepatocerebral degeneration, which occurs in the late stages of chronic liver disease and is associated with the development of portosystemic shunts [5]. In this case, concomitant hepatic encephalopathy hindered the diagnosis [3].

AHD is associated with globus pallidus hyperintensity on MRI T1-weighted images, which results from manganese deposition because of both portosystemic shunting and liver dysfunction. Follow-up has focused on best supportive care and clinical monitorization. Prognosis is gruesome in this case, since AHD developed at a particularly young age and the patient is still awaiting liver transplantation, which is the only possible treatment.

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Conflicts of Interest: None.

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