Typical Imaging of Acquired Hepatocerebral Degeneration in a Paucisymptomatic Patient

Alterações Imagiológicas Típicas de Degenerescência Hepatocerebral Adquirida num Doente Paucisintomático

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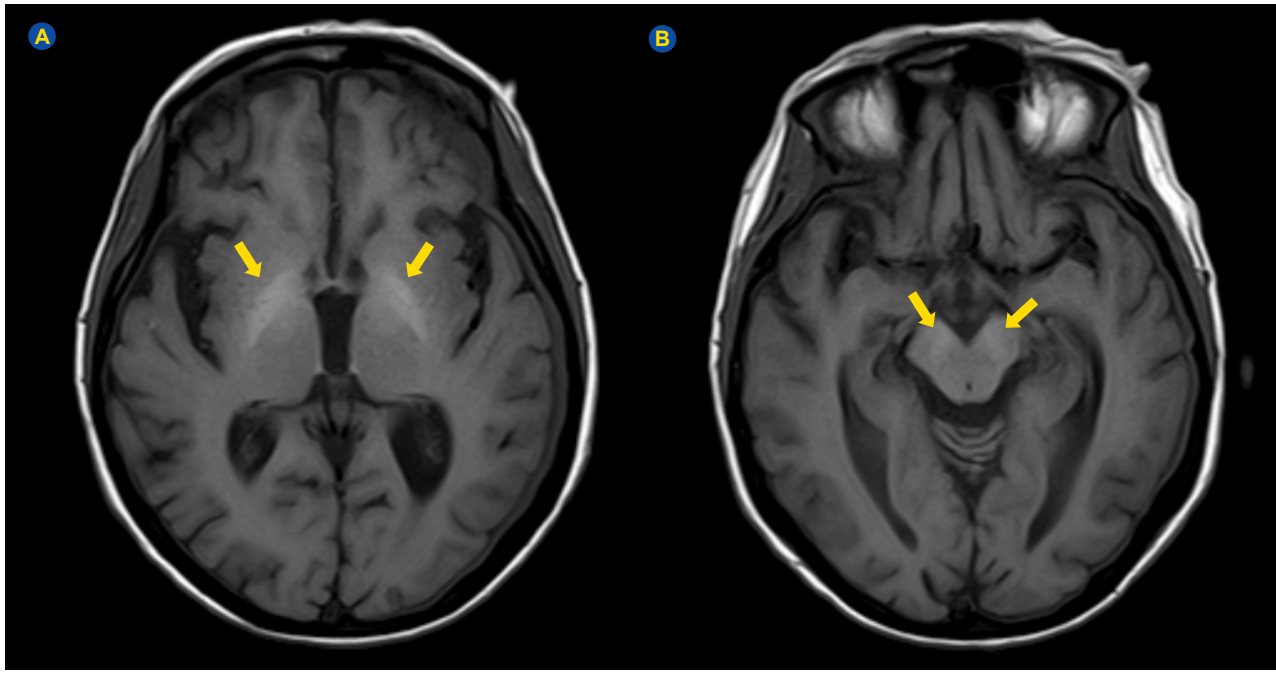

Figure 1 – Brain MRI (axial plane) depicted symmetric T1 hyperintensity in *globus pallidus* (A; yellow arrows), *substantia nigra* and mid-brain (B; yellow arrows). These changes are typical of acquired hepatocerebral degeneration, and are believed to be due to manganese deposition in the CNS of patients with chronic hepatic failure.

A 63-year-old woman with a portosystemic shunt in the context of hepatic cirrhosis but without history of encephalopathic episodes, had been complaining of arm tremor for six months. Her examination showed a mild extrapyramidal symmetrical parkinsonism with bilateral discrete bradykinesia, symmetric rest and action tremor and slight bilateral arm rigidity (particularly on the right side). She underwent brain magnetic resonance imaging (MRI) which depicted typical findings of acquired hepatocerebral degeneration (Fig. 1).

This is a rare, underdiagnosed and frequently irreversible neurologic syndrome that occurs in the context of chronic hepatic failure, particularly with portosystemic shunts.[1,2] It is characterized by extrapyramidal, neuropsychiatric and cerebellar symptoms,[1] and is believed to be caused by manganese deposition in the central nervous system - particularly in the basal ganglia. Brain MRI typically shows bilateral T1- hyperintensity in the basal ganglia (particularly in the pallidum), sometimes extending to the thalamus.[2]

PROTECTION OF HUMANS AND ANIMALS: The authors declare that the procedures were followed according to the regulations established by the Clinical Research and Ethics Committee and to the Helsinki Declaration of the World Medical Association.
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