Acquired hepatocerebral degeneration

25 year-old male patient with a history of hepatic cirrhosis, he had attacks characterized by disturbed consciousness (mostly confusion) associated with movement disorders, dysarthric speech, tremor.

Imaging Findings:

A 25 year-old male patient had a history of hepatic cirrhosis developing secondary to hepatitis B and/or C infection for 11 months. He was referred for MRI study in a stuporous state which was evident for the last two days. He had milder attacks in his medical history, characterized by disturbed consciousness (mostly confusion) associated with movement disorders, dysarthric speech, tremor, and somewhat resolving over several weeks. His serum ammonium level was considerably high. MRI study of the cranium was performed with a 1.5 T MR scanner, T1 and T2 weighted sequences on three planes were obtained.

Discussion:

Acquired hepatocerebral degeneration (AHD) is a rare nonhereditary irreversible neurologic syndrome that occurs in patients with chronic liver disease associated with multiple metabolic insults. Most of the disorders of cerebral function develop rapidly, and result mainly in a disturbance of consciousness ranging from confusion through stupor to coma and death. Progressive movement disorders, dysarthria, tremor and ataxia may also be observed. The patient's neurological status correlates best with serum ammonium levels. The pathophysiology and the locations of the cerebral injuries are incompletely understood. Enlargement and hyperplasia of protoplasmic astrocytes in the cerebral and cerebellar cortex, basal ganglia, and diencephalic nuclei is seen in acute forms of ACD. Gray and white matter necrosis with cavitation, gliosis, and myelin degeneration may occur in chronic forms. A variety of different MRI findings have been reported in HCD patients. Hyperintense signal changes on T1 weighted, and hypointense or no signal changes on T2 weighted sequences have been reported in the basal ganglia, subthalamic area, quadrigeminal plate, cerebral peduncles, internal capsules, anterior pituitary gland. Especially bilateral symmetrical hyperintensity of the globus pallidus is reported in T1-weighted cerebral magnetic resonance images in 52-100% of patients with chronic liver diseases. This hyperintense signal change on T1 weighted images is reported to be due to the deposition of manganese most probably reflecting the presence of an adaptive process designed to improve the efficacy of ammonia detoxification by astrocytes.

Differential Diagnosis List: Acquired hepatocerebral degeneration
**Final Diagnosis:** Acquired hepatocerebral degeneration

**References:**


Figure 1

Description: Sagittal T1 weighted MR image shows diffuse hyperintense signal changes in the cerebral peduncles, quadrigeminal plate, anterior pituitary gland, cerebellar white matter. Origin:
Description: Axial T1 weighted MR image reveals symmetrical diffuse hyperintense signal changes of the putamina, globus pallidi, kaput of kaudat nuclei. Origin:
Description: Axial T2 weighted MR image shows globus pallidi to be diffusely and symmetrically hypointense. Origin:
Description: Axial T1 weighted MR image shows clearly delineated hyperintense signal changes of the quadrigeminal plate. Origin: