

Meige syndrome: full recovery after liver transplantation



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Background

Meige syndrome is a focal orofacial dystonia involving symmetrical blepharospasm and oromandibular dystonia. It is also known as Brueghel syndrome and was first described more than 100 years ago. The typical symptom of Meige syndrome is difficulty keeping the eyelids open. The etiology of this disorder is unknown, it is considered by many investigators to be a form of idiopathic torsion dystonia. In the majority of the patients, Meige syndrome is primary or idiopathic, where the cause of spasm is not known, however secondary cases can occur following prolonged use of neuroleptics or secondary to underlying brain disorders and also in case of hyperthyroidism. No cases of Meige syndrome due to acquired hepatocerebral degeneration (AHD) are known.

Objective

To describe a case of Meige syndrome due to AHD and its reversibility with liver transplantation.

Materials and Methods

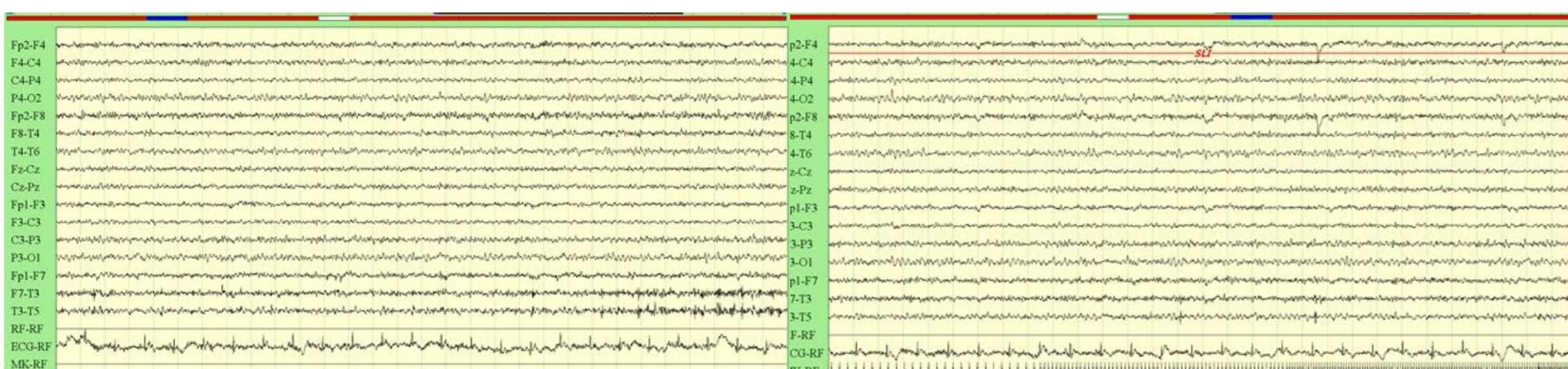
A 63-year-old man with an HCV-related liver cirrhosis and a recent onset of hepatic encephalopathy received an extensive neurologic assessment, in order to undergo a liver transplantation (LT), and he was seen again after surgery

Results

Before LT

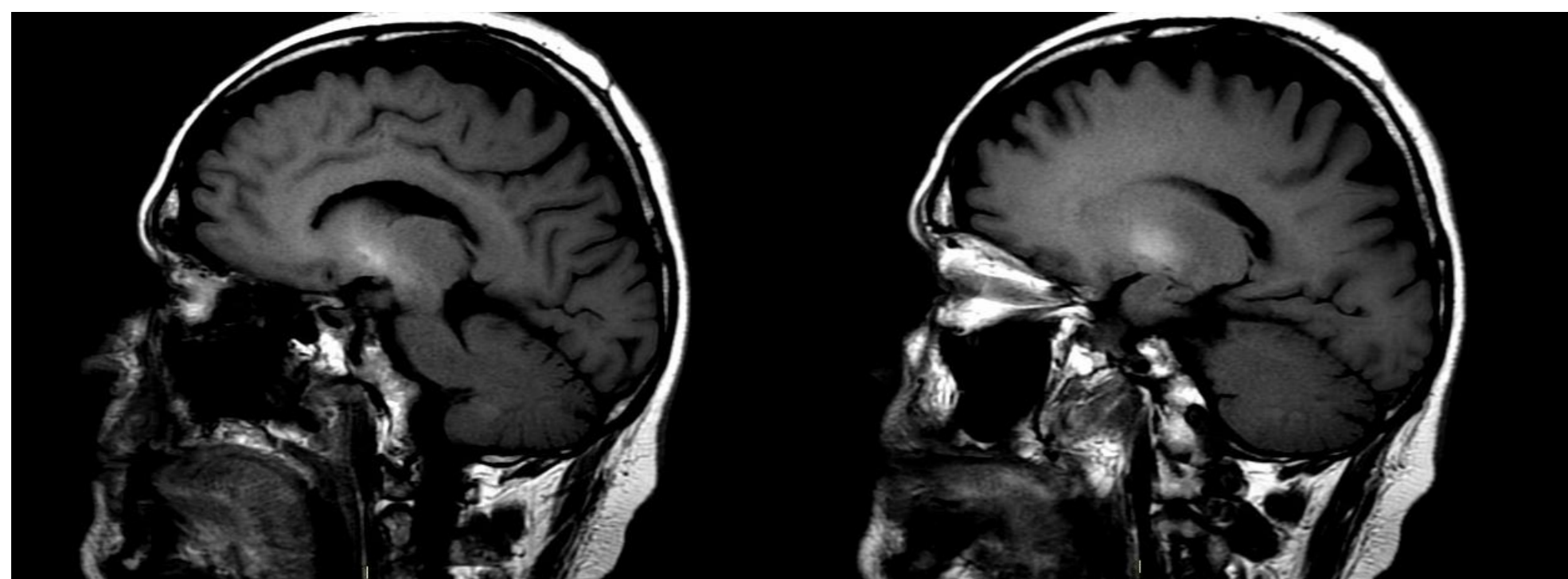
Clinically: somnolence, mild cognitive dysfunction, delirium and psychosis, associated with symmetrical **blepharospasm and oromandibular dystonia leading to the diagnosis of Meige syndrome**

EEG background activity was diffusely slow



Brain CT scan: normal

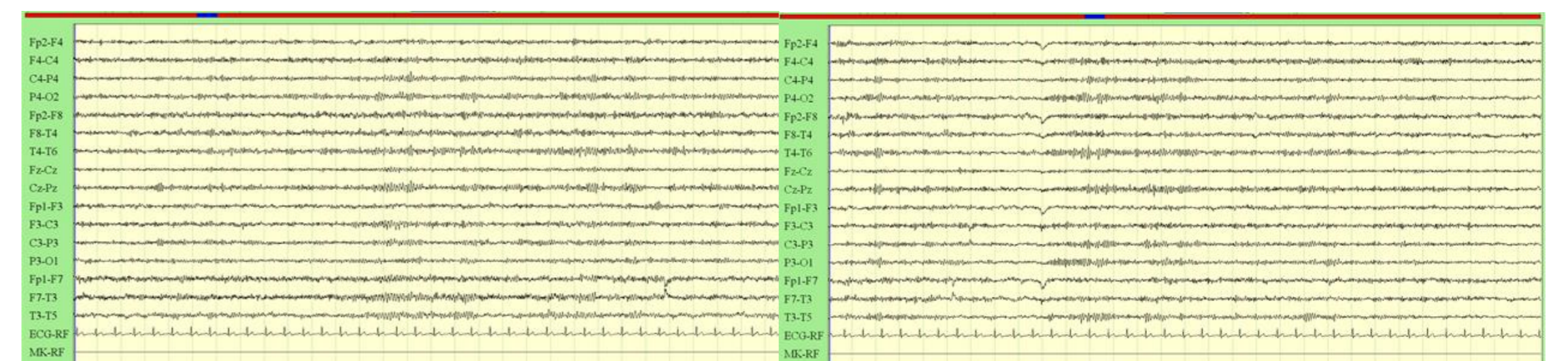
Brain MRI: bilateral pallidal hyperintensity on T1-weighted images



After LT

Clinically: the movement disorder disappeared, on behavioural examination only mild psychoaffective disturbances persisted for a few months. Neurological examination was normal

EEG: normal



Brain CT scan: normal

Brain MRI: complete resolution of bilateral pallidal hyperintensity on T1-weighted images

Follow-up: No further neurological problems occurred during 5 years of follow-up

Discussion and Conclusion

This is the first description of AHD-related Meige syndrome. Our observation confirms that AHD is a potentially reversible syndrome that may be healed by liver transplantation. Therefore, its presence should not be considered a contraindication for this operation.