Alcohol abuse and acute behavioural disturbances in a 24-year-old patient

Álvaro Machado, João Soares-Fernandes, Manuel Ribeiro, Margarida Rodrigues, João Cerqueira, Carla Ferreira

1. Answer

C. Marchiafava-Bignami disease (MBD).

2. Discussion

MBD was named after two Italian pathologists who described acute demyelination of the corpus callosum at necropsy in 3 South-Italian male red-wine drinkers.1 Etiology is unknown. The main pathological features range from demyelination with preserved axon structure, to extensive necrosis with cystic formation and microbleedings.2

Clinical features are highly variable and include reduced consciousness, unsteady gait, behavioural disturbances, motor defects, seizures and, rarely, interhemispheric disconnection syndromes.2 Most of these can be seen in much more frequent alcohol-related disorders like Wernicke’s encephalopathy or central pontine myelinolysis, which may not have distinct ocular findings.3 Recent brain-imaging methods, particularly MRI, disclosed highly specific lesion patterns which, combined with the clinical features, were used to divide MBD in 2 subtypes: type A, characterized by consciousness impairment, extensive T2-weighted hyperintense swelling of the corpus callosum, and bad prognosis; and type B, characterized by behavioural and gait disturbances, restricted “sandwich-like” T2-weighted hyperintense lesions in the corpus callosum genu or splenium, and a better outcome.4 MRI also assumes a pivotal role in distinguishing MBD from other diseases, as the lesions affect the central layers of the corpus callosum and are remarkably symmetric.3 Diffusion weighted imaging (DWI) and fluid-attenuated inversion recovery are most sensitive, depicting striking hyperintense lesions.3 This was seen in our patient (Fig. 1C, D).

Spectroscopy, showing increased myo-inositol and choline peaks, without a decrease in the NAA/Cr ratio, suggested demyelination with absent or minor axonal damage. Perfusion-weighted imaging, has not, to our knowledge, been published in relation to MBD. In our patient no perfusion abnormality was seen, arguing against an acute disruptive lesion. Fiber-tracking showed callosal interhemispheric fiber disruption (Fig. 1B).

The combination of preserved NAA/Cr ratio and preserved perfusion may be a better outcome marker, as both argue against a necrotic lesion, which could be expected considering conventional imaging and DWI findings.

References