



Images in Neuroscience: Answer

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

Álvaro Machado^{a,*}, João Soares-Fernandes^b, Manuel Ribeiro^b, Margarida Rodrigues^a, João Cerqueira^a, Carla Ferreira^a

^aNeurology Department, Hospital de São Marcos, Largo Carlos Amarante, Apartado 2242, 4701-965, Braga, Portugal

^bNeuroradiology Department, Hospital de São Marcos, Braga, Portugal

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.⁵ 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 myoinositol 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.

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DOI of question: [10.1016/j.jocn.2008.08.040](https://doi.org/10.1016/j.jocn.2008.08.040)

* Corresponding author. Tel.: +351 253209089, +351 964124582; fax: +351 253613334.

E-mail address: alvmac@gmail.com (Á. Machado).