Marchiafava–Bignami disease

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Hong Kong Med J 2024;30:417.e1–2
https://doi.org/10.12809/hkmj2310892

This article was published on 15 Oct 2024 at www.hkmj.org.

A 67-year-old female was admitted to the neurology department in October 2020 with abnormal behaviour and cognitive impairment. Her memory and numeracy had declined, and symptoms progressed over the preceding week. She had a 30-year history of chronic alcohol abuse with an average daily intake of 500-mL liquor.

Routine biochemistry including electrolytes, liver function, and vitamin B12 were within normal limits. Magnetic resonance imaging of the brain showed areas of hyperintensity of the corpus callosum (splenium, body, and genu), bilateral middle cerebellar peduncles, periventricular white matter, and subcortical white matter of the frontal lobe on T2-weighted and fluid-attenuated inversion recovery images. Diffusion-weighted imaging revealed prominent high-intensity signal lesions involving the splenium, and these corresponding lesions were hypointense on the apparent diffusion coefficient map (Fig). Based on her history, physical examination and magnetic resonance imaging features, the patient was diagnosed with Marchiafava–Bignami disease (MBD). Despite a normal level of serum vitamins, the patient was prescribed vitamin B and neurotrophic treatment. Symptoms improved and she made a good recovery over the next 30 days.

Marchiafava–Bignami disease is a rare neurological syndrome characterised by primary degeneration and necrosis of the corpus callosum associated with chronic alcoholism and malnutrition. The clinical manifestations of MBD are severe and nonspecific and include an altered

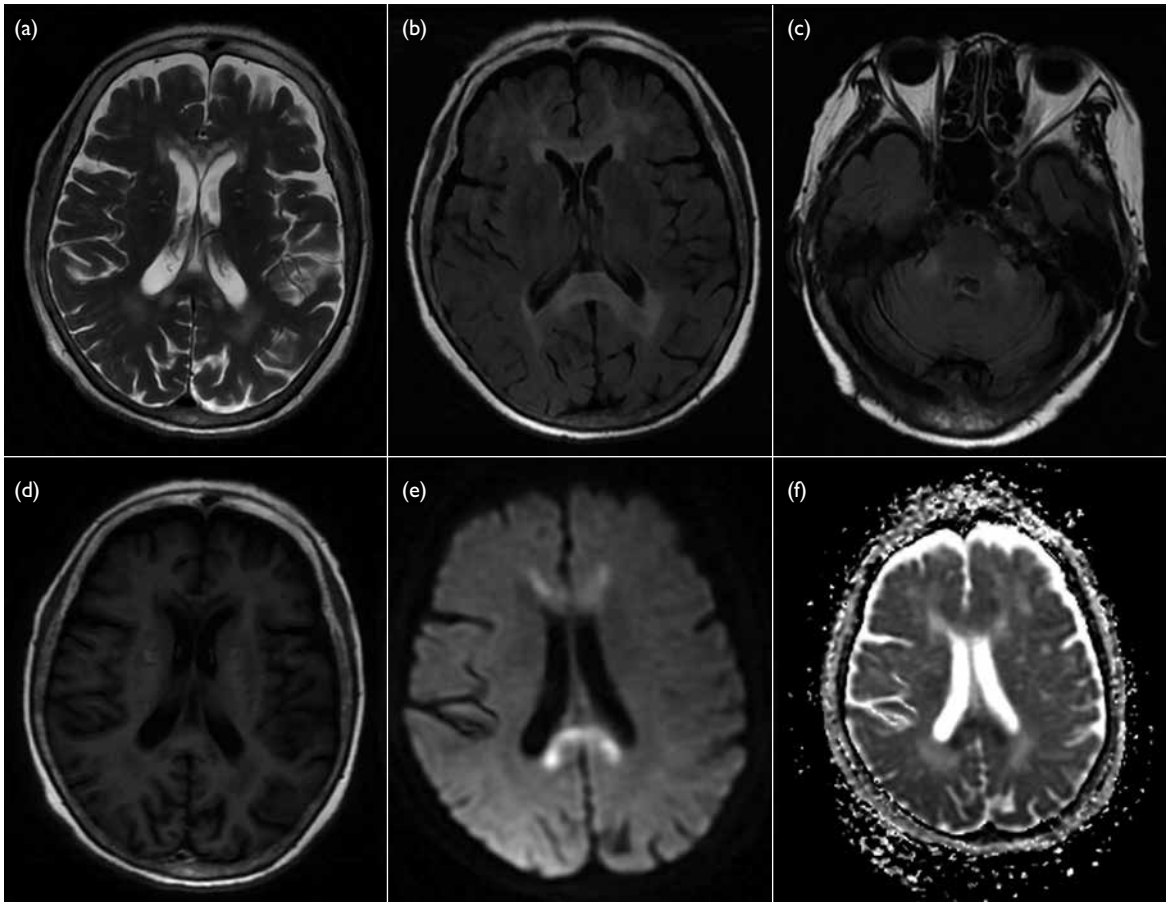

FIG. Magnetic resonance imaging of the brain showing (a) the entire corpus callosum with hyperintensity on T2-weighted imaging, (b) the entire corpus callosum with hyperintensity on fluid-attenuated inversion recovery imaging, (c) bilateral middle cerebellar peduncles with hyperintensity on fluid-attenuated inversion recovery imaging, and (d) the entire corpus callosum with hypointensity on T1-weighted imaging. The splenium was strongly hyperintense on diffusion-weighted imaging (e) and hypointense on apparent diffusion coefficient map (f). The genu was moderately hyperintense on diffusion-weighted imaging (e) and hypointense on apparent diffusion coefficient map (f)

mental state, impaired walking, deficient memory, and dysarthria. Symptoms and imaging findings may improve following thiamine treatment.[1,2] The role of magnetic resonance imaging is essential to confirm the diagnosis. Chronic alcohol abuse plays an important role in its development although MBD has been occasionally diagnosed in patients with no history of alcohol abuse, in particular individuals with poorly controlled diabetes mellitus or following surgery.[3,4] The aetiology and pathophysiology of MBD remain unclear. Possible mechanisms include cytotoxic oedema, blood-brain barrier breakdown, demyelination, and necrosis. Early pathological manifestations are mainly intramyelinic or cytotoxic oedema. In the later stages, demyelination and necrosis of the corpus callosum (especially in the genu and the body) may follow[5] with necrosis leading to atrophy and cavitation in chronic stages, and a decreased number of oligodendrocytes. Occasionally, similar lesions can involve extracallosal regions, such as the anterior and posterior commissures, subcortical white matter, middle cerebellar peduncle, optic chiasm, putamen, internal capsules, hippocampus, and frontal cortex.[2]

The recent advances in neuroradiology techniques help understand the pathophysiological processes of MBD. Studies with diffusion-weighted imaging have shown a low apparent diffusion coefficient, which has been interpreted as irreversible cytotoxic oedema, and may precede the development of demyelination and necrosis and predict a poor or partial recovery.[2,5] Nonetheless the high apparent diffusion coefficient showing reversible signal changes favoured an underlying vasogenic oedema-related process. In magnetic resonance spectroscopy studies, an increased choline/creatine ratio indicates demyelination during the acute phase of MBD, while a reduced N-acetylaspartate/creatine ratio suggests secondary axonal injury. In addition, decreased cerebral blood flow and cerebral blood volume in magnetic resonance perfusion suggest hypoperfusion. Recognition of the neuroradiological features is crucial to establish a diagnosis.

Author contributions

Concept or design: Both authors.
Acquisition of data: Q Wang.
Analysis or interpretation of data: F Ren.
Drafting of the manuscript: F Ren.
Critical revision of the manuscript for important intellectual content: Q Wang.

Both authors had full access to the data, contributed to the study, approved the final version for publication, and take responsibility for its accuracy and integrity.

Conflicts of interest

Both authors have disclosed no conflicts of interest.

Funding/support

This study received no specific grant from any funding agency in the public, commercial, or not-for-profit sectors.

Ethics approval

This study was approved by the Hospital of Chengdu University of Traditional Chinese Medicine Research Ethics Committee, China. Informed consent for all treatments and procedures, and consent for publication were obtained from the patient.

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