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Marchiafava-Bignami Disease: A Case Study

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Abstract **Case Report**

Marchiafava-Bignami disease is a rare neurological condition characterised by necrosis and demyelination of the corpus callosum, usually associated with chronic alcoholism and/or malnutrition. The clinical manifestations of Marchiafava-Bignami disease are diverse and often non-specific. The diagnosis of Marchiafava-Bignami disease is based on magnetic resonance imaging results, which reveal significant and symmetrical damage to the corpus callosum. We report the case of a 63-year-old man with chronic alcoholism who had been experiencing symptoms of confusion for several days. Brain CT imaging revealed diffuse and complete damage to the corpus callosum, characteristic of a severe form of Marchiafava- Bignami disease.

Keywords: Marchiafava-Bignami disease, case report.

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INTRODUCTION

Marchiafava-Bignami disease (MBD) is a rare neurological complication characterised by necrosis and demyelination of the corpus callosum (CC) [1]. It is mainly linked to chronic alcoholism and/or malnutrition, often associated with a deficiency in B vitamins, particularly thiamine [2].

The clinical manifestations of MBD are diverse and often non-specific, including neuropsychiatric disorders, dysarthria, tetraparesis, astasia-abasia, convulsions, altered consciousness and symptoms of interhemispheric disconnection [1, 3].

The diagnosis of MBD is generally based on both medical history and imaging, which reveals significant and symmetrical damage to the corpus callosum, particularly its middle body, genu and splenium, with or without extracallosal lesions [3].

Objective: The aim of this study is to report the case of a patient and to support the role of imaging in the diagnosis of this entity.

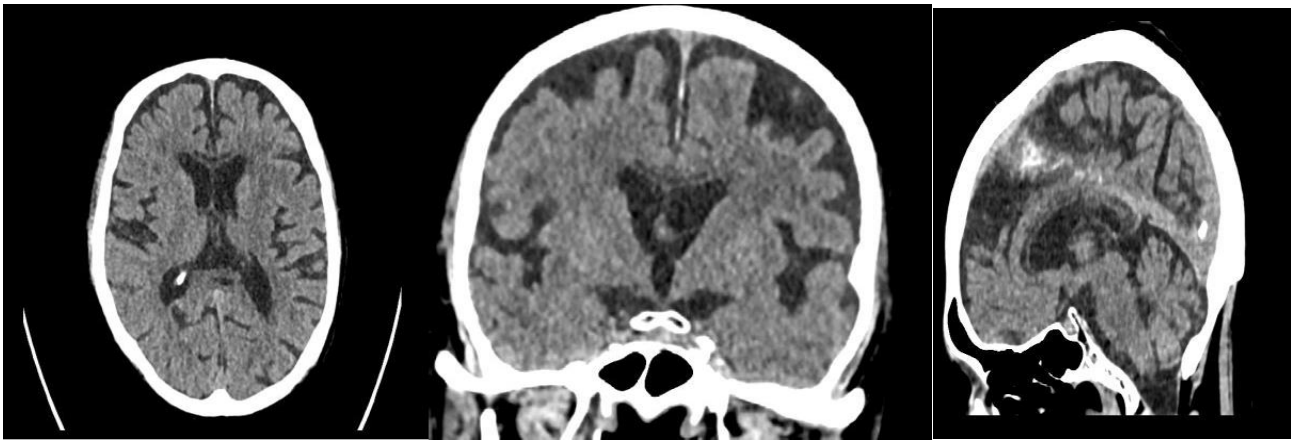

Figure 1: Computed tomography showing linear hypodensity of fluid-like appearance in the knee and splenium of the corpus callosum

PATIENT AND METHOD

Patient information: We report the case of a 63-year-old male patient with a history of chronic alcoholism.

Clinical findings: confusion and the rest of the examination was unremarkable.

Diagnostic approach: Suspicion of subdural haematoma, the patient was referred for a CT scan, which showed a linear hypodensity resembling fluid in the knee and splenium of the corpus callosum, appearing to have developed since the MRI scan in November 2022: chronic phase of Marchiafava-Bignami?

DISCUSSION

The pathophysiological mechanism of MBD remains unclear; however, published case reports consistently show that thiamine deficiency contributes to the development of the disease [3]. Cytotoxic oedema appears to play a key role in the early stages, followed by demyelination and necrosis in later stages [4, 5]. In addition, laminar sclerosis of the cerebral cortex, known as Morel's laminar sclerosis, is observed [6].

The symptoms of MBD can manifest acutely, subacutely, or chronically. The acute form is characterised by severe disturbances of consciousness, convulsions, and hypertonia of the limbs. The subacute form manifests as confusion, dysarthria, behavioural changes, drowsiness and visual disturbances. If diagnosis and treatment are not administered promptly, MBD can progress to coma and even death [2, 7]. The chronic form, which is less common, usually manifests as persistent dementia [8].

Given the variability and non-specific nature of the clinical manifestations of MBD, early diagnosis and differentiation from other conditions can be difficult. Nevertheless, MRI plays a key role in early diagnosis, as the identification of pathognomonic signs is crucial for prompt management. The acute form of MBD is generally characterised by hyperintensity on T2/FLAIR and DWI sequences, hypointensity on ADC and T1 sequences, and swelling of the

CC. The lesions observed are symmetrical and may show peripheral contrast-en . The lesions observed are symmetrical and may show peripheral contrast enhancement.

High-dose parenteral supplementation with thiamine and vitamin B complex remains the primary treatment for disorders related to alcoholism and malnutrition. In addition to thiamine, steroids are also commonly used to treat MBD, as they can stabilise the blood-brain barrier and reduce inflammatory oedema; however, their efficacy remains hypothetical [12, 13].

The progression and prognosis of MBD can vary. Favourable outcomes with regression of lesions visible on brain MRI are possible [14]. Severe disturbances of consciousness, low CC ADC values, total CC involvement, and extracallosal lesions, as in our case, are factors that may be associated with a poor prognosis [1, 10].

CONCLUSION

MBD is a rare condition that neurologists and neuroradiologists should consider when observing partial or diffuse CC lesions, detected by MRI or CT, in a patient with chronic alcoholism and malnutrition. Early diagnosis, combined with prompt treatment with parenteral thiamine, may offer the patient a chance of survival and recovery.

Conflicts of interest: The authors declare no conflicts of interest.

Author contributions: All authors contributed to the diagnostic and therapeutic management of patients and to the writing of this paper. They also declare that they have read and approved the final version of the manuscript.

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