A rare neurological manifestation of a malnourished alcohol-dependent acute pancreatitis patient with Marchiafava–Bignami disease

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Introduction
Marchiafava–Bignami disease (MBD) is a rare demyelinating disorder, usually associated with heavy chronic alcohol consumption. It may occasionally occur in patients who are not alcoholics but are chronically malnourished and patients with severe nutritional deficiencies [1]. White-matter degeneration affects the corpus callosum and results in neurocognitive impairment and dysfunction of the corticobulbar and pyramidal tracts [2]. Neuroimaging, diffusion-weighted magnetic resonance imaging (MRI) specifically plays a pivotal role in confirming the diagnosis of MBD and in determining prognosis [3]. The treatment remains controversial and shows variable results, with complete recovery being a rare outcome [4]. We present a case of an alcohol-dependent female patient with subacute MBD and concurrent acute pancreatitis.

Clinical case report
A severely malnourished 49-year-old woman (body mass index, 16.5) with a long history of alcohol dependency was admitted to the Emergency Department with speech impairment and gait abnormality that had developed gradually over the preceding 2 weeks. Neurological examination revealed cognitive impairment, dysarthria, and tetraparesis (more pronounced in the lower limbs), which indicated cortical, bilateral corticobulbar and pyramidal tracts impairment. Physical examination showed a distended abdomen with rebound tenderness. Blood tests revealed moderately elevated serum aspartate transaminase (90 U/L), alanine transaminase (72 U/L), gamma-glutamyltransferase (96.00 U/L), significantly elevated pancreatic lipase (324 U/L), and high C-reactive protein (143 mg/L). Additional laboratory tests detected a decreased level of total albumin (44 g/L), albumin (23.8 g/L), iron (0.31 mg/L), and calcium (0.078 g/L), but not vitamins B1, B6, B12, or other electrolytes. The lipid profile was within normal limits. Abdominal computed tomography (CT) showed a lack of pancreatic parenchymal enhancement with multiple pancreatic cysts suggestive of acute pancreatitis and significant liver fatty change. Diffuse hypodensity of corpus callosum was seen on cranial CT imaging, confirmed by hyperintensity of the corpus callosum on FLAIR and T2 images in MRI (Figure 1A and B). Similar hypodense changes were found in the cortical and subcortical regions of the frontal lobes. No corresponding hypointensity was observed on ADC images. MBD was diagnosed based on clinical presentation, history of chronic alcohol abuse, and characteristic MRI findings. The patient commenced pharmacological treatment consisting of calcium, iron, vitamin B complex, and a 5-day course of intravenous steroids with 500 mg methylprednisolone per day and dietary supplementation via percutaneous endoscopic gastrostomy.

After 4 weeks of treatment, the acute pancreatitis subsided, with pancreatitis-function biochemical markers normalizing (lipase 41.00 U/L, gamma-glutamyltransferase 56.00 U/L).
frequently found and are associated with a poor prognosis. Extracallosal lesions are localized in the entire corpus callosum. Extracallosal lesions are associated with cognitive deterioration. The brain lesions depicted on MRI are caused by poor nutritional supply to brain tissue. The permeability of intracranial small vessels, which results in a gastrointestinal malabsorption, is detrimental and can impair chronic malnutrition, whether caused by inadequate intake or specific nutritional deficiencies in our patient. Nonetheless, a specific factor has not been identified, although in alcoholics, necrosis or demyelination of the corpus callosum is specific for MBD, a pathophysiological mechanism attributed to the toxic effects of alcohol, but this is unlikely in view of the prevalence of alcoholism and the rarity of corpus-callosum degeneration. A nutritional etiology has been considered in the central pons [8]. Conditions not related to alcohol abuse such as multiple sclerosis, acute disseminated encephalomyelitis, infection, viral and bacterial infections, and HIV infection should also be considered in the differential diagnosis.

Effective treatment is not available due to the uncertainty of the etiology of MBD. Thiamine and other forms of vitamin B supplementation are routinely used. Haas et al. [9] reported a favorable response to corticosteroids, although other authors do not support these findings [10]. Regardless of treatment, partial or complete recovery is rare and most patients remain with severe neurological deficits.

Conclusions

Our case report documents the diagnostic process of rapidly developing MBD in an alcohol-dependent and severely malnourished patient with concurrent acute pancreatitis. Despite treatment, only partial symptom resolution was observed, probably due to deep and irreversible cellular changes in the brain tissue and the gastrointestinal tract. However, prompt diagnosis and supportive treatment possibly prevented a dramatic or even fatal outcome. This case should raise awareness of the coexistence of different multiorgan complications and the need for a multidisciplinary approach. Alcohol-dependent and malnourished patients presenting with acute neurological deficits should be thoroughly investigated to exclude potentially fatal conditions such as MBD. The mutual interplay between the gastrointestinal tract and nervous systems has been a matter of great interest in recent years.

Authors’ contributions

M.S., J.B., K.P., and A.S.S. conceived and designed the project. M.S., A.S.S., and J.B. collected the data. M.S., J.B., A.S.S., K.P., and K.R. analysed and interpreted the data. M.S. and J.B. drafted the manuscript. All authors read and approved the final manuscript.

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Conflicts of interest
None declared.

References
1. Kakkar C, Prakashini K, Polnaya A. Acute Marchiafava-Bignami disease: clinical and serial MRI correlation. BMJ Case Rep 2014;2014:bcrr2013203442.
2. Yao-Yao S, Chen-Guang Z, Ning H et al. Clinical and neuroradiological features of 15 patients diagnosed with Marchiafava-Bignami disease. Chin Med J 2019;132:1887–9.
3. Hlaihel C, Gonnaud PM, Champin S et al. Diffusion-weighted magnetic resonance imaging in Marchiafava-Bignami disease: follow-up studies. Neuroradiology 2005;47:520–4.
4. Carrilho PE, Santos MB, Piasecki L et al. Marchiafava-Bignami disease: a rare entity with a poor outcome. Rev Bras Ter Intensiva 2013;25:68–72.
5. Bachar M, Fatakhov E, Banerjee C et al. Rapidly resolving non-alcoholic Marchiafava-Bignami disease in the setting of malnourishment after gastric bypass surgery. J Investig Med High Impact Case Rep 2018;6:232470961878431.
6. Adams RD, Victor M, Ropper AH. Principles of Neurology, 6th edn. New York, USA: The McGraw-Hill Companies, 1997: 998–9.
7. Sera S, Ichiba T. Marchiafava-Bignami disease in a patient with no alcohol abuse. Neurol India 2019;67:1169.
8. Muccio CF, De Lipsis L, Belmonte R et al. Reversible MR findings in Marchiafava-Bignami Disease. Case Rep Neurol Med 2019;2019:1–3.
9. Haas L, Tjan D, Van Die J et al. Coma in an alcoholic: Marchiafava-Bignami disease. NZ Med J 2006;119:1244.
10. Hillbom M, Saloheimo P, Fujoka S et al. Diagnosis and management of Marchiafava-Bignami disease: a review of CT/MRI confirmed cases. J Neurol Neurosurg Psychiatry 2014;85:68–173.