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LETTER TO THE EDITOR

Bilateral middle cerebellar peduncles involvement a malnourished man with Marchiafava-Bignami disease

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Dear Editor,

Marchiafava-Bignami disease (MBD), as a rare condition mainly associated with alcoholism and malnutrition, is characterized by demyelination and necrosis of the corpus callosum (CC) [1]. The clinical presentation of MBD can vary from altered mental status, cognitive impairment, gait disturbance, seizure, depression, and even death [2]. The characteristic neuroimaging findings are symmetric lesions of the CC. Extracallosal lesions such as cortex or subcortical white matter have also been well documented in the literature. Nevertheless, MBD involving bilateral middle cerebellar peduncles (MCPs) is relatively rare. Here, we describe a case of MBD with widespread lesions including bilateral MCPs.

An 85-year-old man with a long-term history of chronic obstructive pulmonary disease (COPD) was admitted to our department due to altered mental status, difficulty in walking and speaking for 7 days. Three weeks prior to admission, he was sent to local hospital for expectoration, dyspnea, and loss of appetite on March 16, 2018. Several days later, he was unable to feed. Chest computed tomography (CT) showed massive pleural effusion and he was treated with thoracal closed drainage as well as empirical antibiotics. As complicated with neurological presentation, the patient was referred to our hospital. He was skeletization with a body mass index (BMI) of 12.9 kg/m^2 and did not have a habit of drinking alcohol. On admission, his vital signs were a temperature of 36.5 °C, a heart rate of 78 beats per minute, blood pressure of 145/72 mmHg, and body weight of 35 kg. Breathing sounds were reduced in both lower lungs. The findings of neurological examinations included lethargy, severe cognition impairment, tetraparesis, and generalized muscular hypertonia. Laboratory studies yielded macrocytic anemia (hemoglobin 10.3 g/dL, normal range 12.9–17.0 g/dL; corpuscular volume 99 fL, normal 80.0–95.0 fL), hypoproteinemia (albumin 2.8 g/dL, normal range 3.5–5.2 g/dL), hypokalemia (potassium 3.38 mmol/L, normal range 3.5–5.5 mmol/L), and low level of vitamin B (vitamin B12 100 pg/mL, normal range 191–663 pg/mL; vitamin B1 11 ng/mL, normal range 20–50 ng/mL). Tests of blood glucose, liver and renal function, endocrine hormones, and tumor markers were all within normal limits. The CSF analysis showed slightly increased protein (76 mg/dL, normal range 15–45 mg/dL) without pleocytosis. Work-up for specific viral and atypical infectious pathogens were all negative. On day 2 after admission, brain magnetic resonance imaging (MRI) showed abnormal signal intensities in the splenium of corpus callosum (SCC), which were hyperintense on T2-weighted and diffuse-weighted images with hypointense on T1-weighted image (Fig. 1a–c). In addition, hyperintense lesions in bilateral middle cerebellar peduncles (MCPs) and subcortical white matter could also been visualized on fluid-attenuated inversion recovery (FLAIR) sequence (Fig. 1d). The patient was diagnosed with MBD and administrated with high-dose multivitamins, methylprednisolone, and nutritional support. On day 14 after admission, a repeated MRI disclosed partial resolution in the SCC and MCPs (Fig. 1e, f). However, he was still dementia, global aphasia, and tetraplegia in flexion without any meaningful response. He was discharged 1 week later.

Discussion

MBD, a rare neurological syndrome frequently associated with a history of alcoholism and malnutrition, is first reported in Italian alcoholics. It can also occur in the setting of other conditions including carbon monoxide poisoning, sepsis,

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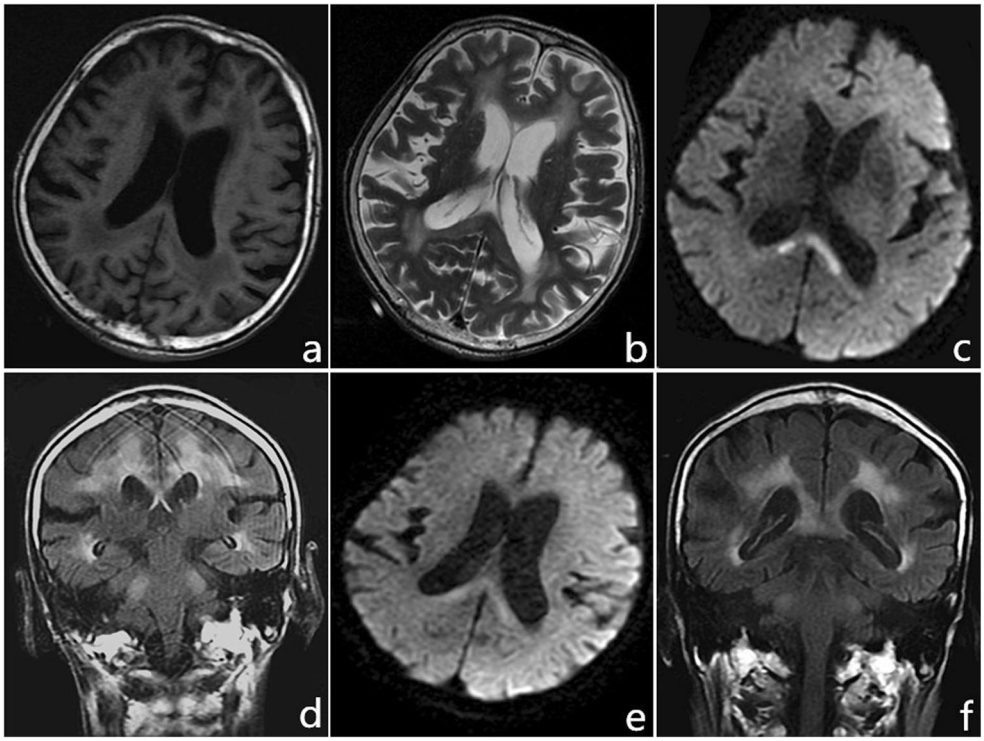

Fig. 1 On day 2 after admission, brain MRI showed hyperintensity of the splenium of corpus callosum (SCC) on T2-weighted image (**b**) and diffuse-weighted image (DWI) (**c**) with hypointensity on T1-weighted image (**a**). Fluid-attenuated inversion recovery (FLAIR) sequence (**d**) showed hyperintensity in bilateral middle cerebellar peduncles (MCPs) and subcortical white matter. Two weeks later, a repeated MRI demonstrated partial resolution in the SCC and MCPs on DWI (**e**) and FLAIR (**f**)

sickle cell disease, *Plasmodium falciparum* infection, cardiac carcinoma surgery, and diabetes [3]. MBD is characterized by degeneration, necrosis, and atrophy of the corpus callosum (CC) that facilitates cognitive, sensory, and motor interhemispheric exchange of information.

The physiopathologic mechanism that underlies MBD remains unknown, but current evidence from human and animal studies suggests a synergism between ethanol-induced neurotoxic effects and hypovitaminosis B. Ethanol and its metabolite directly damage the central nervous system (CNS) by means of inducing reactive oxygen species (ROS) production, which contributes to oxidative stress and neurodegeneration. In the term of thiamine deficiency, it decreases thiamine pyrophosphate-dependent enzyme activity, resulting in diminishing adenosine triphosphate (ATP) production, the development of oxidative damage, and influencing neurotransmitter pathways [3]. Two subtypes of MBD have been summarized according to their clinicoradiological features. Type A is characterized by major impairment of consciousness, the entire CC involvement, and poor outcome. In addition, type B manifests as slight impairment of consciousness, partial callosal lesions, and a favorable outcome [4]. Advanced neuroimaging techniques are helpful for understanding the pathophysiologic processes of MBD. In Magnetic resonance spectroscopy (MRS) studies, the increased choline/creatine (Cho/Cr) ratio and reduced *N*-acetyl aspartate/creatine (NAA/Cr) ratio suggest myelin destruction and secondary neuronal loss, respectively [5]. In addition, there is decreased cerebral blood volume in the CC in MR perfusion indicative of hypoperfusion. Positron emission tomography (PET) scan shows reduced glucose metabolism in CC, cerebral hemispheres, and hemispheric white matter, which may indicate that in MBD, perfusion and metabolism defects may affect structures beyond the CC [6].

In our case, the acute onset of neurological presentation together with symmetric lesions in the SCC, eliminating the possibilities of infectious and vascular factors, a diagnosis of type A-MBD may be established. We speculate that low levels of albumin and vitamin B, suggested a malnourished status, play an important role in the development of MBD. MR findings of MBD involving extracallosal areas have been abundantly described in the literature, especially for cortex or subcortical white matter. Previously published cases indicate that cortical lesions may be a marker of poor outcomes. In this case, widespread subcortical white matter hyperintense may similarly suggest a poor prognosis. Bilateral MCPs involvement in patient with MBD is infrequent [7, 8]. Hence, MBD should be taken into consideration when MR findings of bilateral MCPs insults are present, especially in patient with alcoholism or malnutrition.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflicts of interest.

References

1. Ironside R, Bosanquet FD, McMenemey WH (1961) Central demyelination of the corpus callosum (Marchiafava-Bignami disease) with report of a second case in Great Britain. Brain 84:212–230
2. Hillbom M, Saloheimo P, Fujioka S, Wszolek ZK, Juvela S, Leone MA (2014) Diagnosis and management of Marchiafava-Bignami disease: a review of CT/MRI confirmed cases. J Neurol Neurosurg Psychiatry 85(2):168–173
3. Fernandes LMP, Bezerra FR, Monteiro MC, Silva ML, de Oliveira FR, Lima RR et al (2017) Thiamine deficiency, oxidative metabolic pathways and ethanol-induced neurotoxicity: how poor nutrition contributes to the alcoholic syndrome, as Marchiafava-Bignami disease. Eur J Clin Nutr 71(5):580–586
4. Heinrich A, Runge U, Khaw AV (2004) Clinicoradiologic subtypes of Marchiafava-Bignami disease. J Neurol 251(9):1050–1059
5. Tuntiyatorn L, Laothamatas J (2008) Acute Marchiafava-Bignami disease with callosal, cortical, and white matter involvement. Emerg Radiol 15(2):137–140
6. Nalini A, Kovoor JM, Dawn R, Kallur KG (2009) Marchiafava-Bignami disease: two cases with magnetic resonance imaging and positron emission tomography scan findings (2009). Neurol India 57(5):644–648
7. Xuehan L, Zhiyu LV, Wang P, Chen X (2017) A rare case of Marchiafava-Bignami disease: extracallosal lesions involving bilateral medipeduncle. Neurol India 65(3):642–643
8. Bellido S, Navas I, Aranda MA, Ginestal R, Venegas B (2012) Unusual MRI findings in a case of Marchiafava Bignami disease. Neurology 78(19):1537