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CASE REPORT

Acute Marchiafava–Bignami disease with plexiform neurofibroma: a case report

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Abstract

Background: Marchiafava–Bignami disease (MBD) is a rare neurological disorder characterized by demyelination and necrosis of the corpus callosum. It is most commonly associated with chronic alcohol use and nutritional deficiency. Clinical manifestations are variable and may include seizures, altered sensorium, gait disturbance, and speech impairment. Early recognition is often difficult due to the nonspecific nature of presenting symptoms. Magnetic resonance imaging (MRI) plays a crucial role in establishing the diagnosis. Plexiform neurofibroma, a benign peripheral nerve sheath tumor typically associated with neurofibromatosis type 1, is rarely encountered alongside unrelated neurological conditions.

Case description: We report the case of a 37-year-old man with a long history of alcohol dependence who presented with acute confusion, seizures, slurred speech, and gait instability following heavy alcohol intake. MRI of the brain demonstrated symmetrical signal abnormalities involving the corpus callosum consistent with MBD. Physical examination also revealed a long-standing cervical plexiform neurofibroma.

Management and outcome: The patient was treated with high-dose intravenous thiamine along with supportive measures and anticonvulsant therapy. Gradual neurological improvement was observed during hospitalization.

Conclusion: This case highlights the importance of considering MBD in individuals with chronic alcohol use who present with acute neurological symptoms. Prompt neuroimaging and early thiamine administration are essential to improve clinical outcomes.

Keywords: Marchiafava–Bignami disease, corpus callosum, chronic alcohol use, thiamine deficiency, plexiform neurofibroma

Introduction

Marchiafava–Bignami (MBD) is an uncommon neurological disorder characterized by progressive demyelination and necrosis of the corpus callosum, the principal commissural structure connecting the cerebral hemispheres. The condition was first described in 1903 among individuals with chronic alcohol consumption and was historically associated with a poor prognosis due to delayed diagnosis and limited treatment options (1). More recent clinical and radiological studies have improved understanding of the disease and demonstrated that early recognition and treatment can

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significantly improve outcomes (2). The corpus callosum plays a vital role in interhemispheric integration of motor, sensory, and cognitive functions. Damage to this structure can therefore lead to a broad spectrum of neurological manifestations, including confusion, dysarthria, seizures, gait disturbance, cognitive impairment, and behavioral changes (2, 3). In severe cases, patients may develop altered levels of consciousness or interhemispheric disconnection syndromes (3). Chronic alcohol use and malnutrition remain the most commonly recognized risk factors for MBD. However, cases have also been reported in individuals with metabolic disorders, malignancies, and severe nutritional deficiency in the absence of alcohol use (3). Thiamine deficiency is considered a key pathogenic factor, as thiamine functions as an essential cofactor for enzymes involved in cerebral glucose metabolism. Deficiency results in impaired oxidative metabolism, accumulation of lactate, and subsequent neuronal injury (4). In individuals with chronic alcohol dependence, thiamine deficiency may result from poor nutritional intake, impaired intestinal absorption, reduced hepatic storage, and decreased utilization within neural tissue (4). Magnetic resonance imaging (MRI) has become the most important diagnostic tool for MBD. Typical findings include symmetrical lesions involving the corpus callosum, particularly the body and splenium (5). These lesions usually appear hypointense on T1-weighted images and hyperintense on T2-weighted and fluid-attenuated inversion recovery (FLAIR) sequences. Diffusion-weighted imaging (DWI) often demonstrates restricted diffusion in the acute phase, reflecting cytotoxic edema (6). In more severe cases, extracallosal involvement of cortical and subcortical white matter structures may also be present and is often associated with poorer clinical outcomes (7). Patients with chronic alcohol use frequently present with multiple comorbidities that may complicate diagnosis. The presence of neurocutaneous lesions such as plexiform neurofibromas may introduce additional diagnostic considerations. Plexiform neurofibromas are benign tumors arising from peripheral nerve sheaths and are typically associated with neurofibromatosis type 1.

In this report, we describe a patient with acute MBD presenting after heavy alcohol consumption, with coexisting clinical features suggestive of plexiform neurofibroma.

Case report

A 37-year-old married man with a history of chronic alcohol consumption and tobacco use for approximately 18 years was brought to the emergency department following a binge drinking episode. According to family members, he had consumed large quantities of alcohol over several consecutive days prior to presentation. He subsequently developed persistent vomiting, fever, abnormal involuntary movements involving all four limbs, progressive confusion, and reduced responsiveness. The family also reported urinary incontinence and difficulty speaking before hospital admission. On examination, the patient appeared drowsy and confused with a Glasgow Coma Scale score of 10. There were no signs of meningeal irritation. Neurological examination revealed marked dysarthria and impaired coordination suggestive of cerebellar dysfunction. The patient exhibited agitation, bilateral hand tremors, and intermittent involuntary limb movements. Speech was slurred, and he demonstrated difficulty naming objects, indicating possible anomia. Gait was markedly impaired, and he required assistance to maintain posture. General examination revealed pallor and low-grade fever. Multiple firm nodular swellings were noted over the facial region and posterior neck. These lesions were consistent with cutaneous neurofibromas, and the pattern raised suspicion of plexiform neurofibroma. The patient had a documented history of repeated alcohol binges, previous alcohol withdrawal seizures, and occasional jaundice related to alcohol-associated liver disease. Laboratory investigations revealed macrocytic anemia, leukocytosis, and electrolyte abnormalities. Liver function tests showed elevated transaminases consistent with chronic alcohol-related hepatic injury. Given the acute neurological presentation, differential diagnoses included meningoencephalitis, Wernicke encephalopathy, metabolic encephalopathy, and cerebrovascular events. A non-contrast computed tomography (CT) scan of the brain performed at admission did not reveal significant abnormalities. However, due to persistent neurological deficits, MRI of the brain was performed. DWI demonstrated restricted diffusion involving the central fibers of the corpus callosum, suggesting acute cytotoxic injury (**Figure 1**). FLAIR imaging showed hyperintense signal changes within the corpus callosum (**Figure 2**). Sagittal T2-weighted images revealed diffuse signal abnormalities involving the body and splenium of the corpus callosum without mass effect (**Figure 3**). These findings were consistent with acute MBD. Based on the clinical presentation and neuroimaging findings, treatment was initiated immediately with high-dose intravenous thiamine (500 mg twice daily). The patient also received anticonvulsant therapy with phenytoin and supportive management. Empirical antibiotics were started initially while infectious causes were being excluded. Dextrose-containing fluids were administered only after thiamine supplementation to avoid worsening thiamine deficiency. Additional management included vitamin B complex supplementation, correction of electrolyte abnormalities, and supportive care. Over the following 7 days, the patient demonstrated gradual neurological improvement. His level of consciousness improved, speech became clearer, and

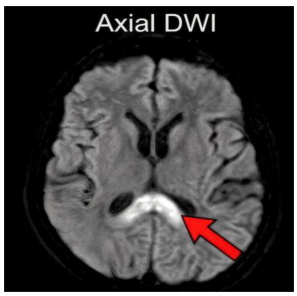

FIGURE 1 | Axial diffusion-weighted imaging (DWI) demonstrating diffusion restriction involving the central fibers of the corpus callosum, predominantly affecting the splenium and body (red arrow), consistent with acute cytotoxic edema seen in Marchiafava–Bignami disease (MBD).

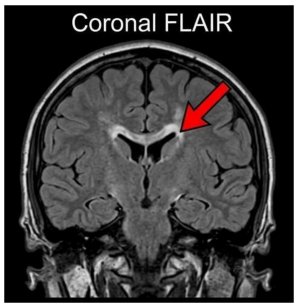

FIGURE 2 | Coronal fluid-attenuated inversion recovery (FLAIR) magnetic resonance imaging (MRI) sequence showing hyperintense signal changes within the body of the corpus callosum (red arrow), reflecting inflammatory and demyelinating involvement typical of the acute phase of MBD.

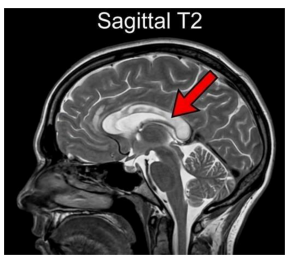

FIGURE 3 | Sagittal T2-weighted MRI showing diffuse hyperintensity involving the corpus callosum (red arrow), predominantly affecting the body and splenium, consistent with toxic-metabolic demyelination seen in acute MBD.

orientation progressively returned. By the time of discharge, he had regained significant cognitive and motor function.

Discussion

Marchiafava–Bignami disease is an uncommon neurological disorder most frequently associated with chronic alcohol use and severe nutritional deficiency. The pathological hallmark of the disease is demyelination and necrosis of the corpus callosum, leading to disruption of interhemispheric communication (1). The pathogenesis of MBD is thought to involve metabolic and nutritional mechanisms, particularly thiamine deficiency. Thiamine plays a critical role in cerebral energy metabolism as a cofactor for enzymes involved in mitochondrial oxidative pathways (4). Deficiency results in impaired energy production, oxidative stress, and neuronal injury. The corpus callosum appears particularly vulnerable to these metabolic disturbances due to its dense concentration of myelinated fibers and high metabolic demand (2). Clinical manifestations are variable and may include confusion, dysarthria, gait disturbance, seizures, behavioural changes, psychosis, or coma (3). Because of this variability, the condition may be mistaken for other alcohol-related neurological disorders such as Wernicke encephalopathy or metabolic encephalopathy (3). Neuroimaging plays a central role in establishing the diagnosis. MRI typically demonstrates symmetrical lesions involving the corpus callosum, particularly the body and splenium (5). DWI frequently reveals restricted diffusion during the acute phase, reflecting cytotoxic edema within affected white matter (6). In the present case, MRI showed diffusion-restricted lesions involving the splenium of the corpus callosum, a finding commonly reported in acute MBD (8). Additional involvement of cortical regions, including the precentral gyrus and bilateral frontoparietal areas, suggested extracallosal extension of the disease process, which has been associated with more severe clinical presentations (7). Another notable feature in this patient was the presence of nodular lesions suggestive of plexiform neurofibroma. Plexiform neurofibromas are benign tumors arising from peripheral nerve sheaths and are typically associated with neurofibromatosis type 1. Although there is no established pathogenic link between neurofibromatosis and MBD, the coexistence of neurocutaneous lesions may complicate clinical evaluation and warrants careful neurological assessment. Early treatment plays a crucial role in determining patient outcomes. High-dose parenteral thiamine remains the cornerstone of therapy and should be initiated promptly when the diagnosis is suspected. Thiamine supplementation helps restore impaired cerebral metabolism and may prevent further neuronal damage (4). Importantly, thiamine must be administered before glucose or dextrose-containing fluids because glucose administration in thiamine-deficient patients may precipitate or worsen Wernicke encephalopathy (9). Supportive management includes correction of electrolyte disturbances, treatment of alcohol withdrawal, seizure control, and nutritional support. Early diagnosis and timely treatment have been associated with favourable neurological recovery in several reported cases (10).

Conclusion

Marchiafava–Bignami disease is a rare but potentially reversible neurological disorder primarily associated with

chronic alcohol use and nutritional deficiency. The condition predominantly affects the corpus callosum and can present with a wide spectrum of neurological manifestations (1). MRI plays a critical role in early diagnosis by demonstrating characteristic callosal lesions, particularly involving the body and splenium of the corpus callosum (5). Early recognition is essential, as prompt treatment with high-dose parenteral thiamine can significantly improve clinical outcomes (4). This case emphasizes the importance of considering MBD in patients with chronic alcohol dependence who present with acute neurological deterioration. Timely neuroimaging and early thiamine therapy may prevent irreversible neurological damage and facilitate recovery (10).

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Conflict of interest

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