Co-occurrence of Marchiafava-Bignami disease and Wernicke's Encephalopathy in a Single Patient: A Rare Case Report

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Abstract

Background

Marchiafava-Bignami disease (MBD) and Wernicke's encephalopathy (WE) are rare alcohol-related neurological disorders caused by chronic malnutrition, particularly thiamine deficiency.

Case Presentation

We report a 56-year-old male with chronic alcoholism who presented with altered mental status, slurred speech, and limb weakness. MRI revealed T2/FLAIR hyperintensities in the genu of the corpus callosum and dorsomedial thalami, with features of both MBD and WE. Diffusion-weighted imaging showed restricted diffusion in the corpus callosum.

Conclusions

This case highlights the rare co-occurrence of MBD and WE in the same patient. Early recognition and vitamin supplementation are key, although prognosis remains guarded in alcohol-related encephalopathies.

Keywords: *Alcohol-Related Disorders, Case Report, Marchiafava-Bignami Disease, Magnetic Resonance Imaging, Thiamine Deficiency, Wernicke Encephalopathy.*

Introduction

Marchiafava-Bignami disease (MBD) and Wernicke encephalopathy (WE) are rare but serious complications of chronic alcoholism, primarily due to thiamine deficiency1. MBD is characterized by demyelination and necrosis of the corpus callosum, while WE typically affects regions around the third and fourth ventricles including the thalami, mammillary bodies, and periaqueductal gray matter2. Clinical features range from confusion and ataxia to coma. Accurate diagnosis requires high clinical suspicion and MRI, which reveals hallmark imaging patterns1,2. We report a unique case of a patient presenting with overlapping radiological features of both MBD and WE, emphasizing the role of MRI in timely diagnosis and treatment initiation.

Case Report

A 56-year-old male with a history of chronic alcohol use disorder arrived at the Emergency Department (ED) due to altered mental status, weakness of bilateral upper and lower limbs, and slurred speech, as reported by a family member. Upon examination in the ED, the patient exhibited hypertension (blood pressure: 170/100 mmHg) and tachycardia (123 beats per minute). Although the rest of his vital signs were otherwise normal, he appeared drowsy, non-verbal, and displayed gait instability. Laboratory results indicated elevated aspartate aminotransferase (205 U/L), while the remaining laboratory tests were within normal limits. Ultrasonography showed features of chronic liver disease with portal hypertension.

The patient is a chronic alcoholic, consuming alcohol daily for the past 30–40 years, typically drinking five glasses of homemade alcohol each day. He had previously been hospitalized for similar problems associated with altered mental status and ataxia in both legs; however, no documentation was available. For the past one and a half months, he experienced decreased appetite, weakness of limbs, reduced interaction, and altered sleep patterns. He visited the psychiatric outpatient department and was diagnosed with alcohol dependence syndrome (ADS)-induced psychosis and was prescribed Tab Fluoxetine, Tab Olanzapine, and Tab Lorazepam. His last intake of alcohol was recorded seven days prior to admission.

A computed tomography (CT) scan of the head showed no significant abnormalities. After receiving primary care, the patient was admitted for further workup. Contrast-enhanced MRI of the brain (Figure 1) revealed T1 hypointense and T2/FLAIR hyperintense lesions in the middle of the genu of the corpus callosum, sparing the dorsal and ventral aspects, showing mild diffusion restriction in DWI images with an ADC value of 0.5×10^{-3} mm^2/s. No areas of hypointense blooming were noted in SWI images. The lesion showed heterogeneous enhancement in the post-contrast study. T2/FLAIR hyperintensity was noted around the tip of the frontal horns of the bilateral lateral ventricles, dorsomedial thalami, basal ganglia, periaqueductal region, and around the fourth ventricle. These lesions showed no diffusion restriction in DWI images and no significant enhancement in the post-contrast study (Figure 1(d)). The combination of chronic alcoholism and specific imaging findings favored the diagnosis of Marchiafava-Bignami disease (MBD) and Wernicke's encephalopathy (WE).

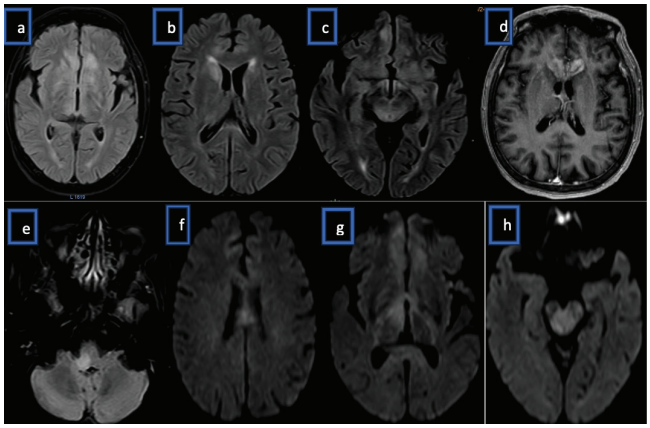

Figure 1. *Axial brain MRI images showing (a, b) FLAIR hyperintensities in the genu of the corpus callosum and dorsomedial thalami; (c) hyperintensities in the periaqueductal region; (d) post-contrast T1 image with heterogeneous enhancement in the genu of the corpus callosum. DWI images (e–h) show diffusion restriction in the genu of the corpus callosum, and (g) bilateral symmetrical hyperintensities in the frontal cortices.*

During hospital admission, neurology and psychiatry consultations were done. The patient was started on intravenous thiamine along with folic acid and multivitamin supplementation. The patient showed slight overall improvement; however, he remained drowsy (GCS E4V3M4). There was minimal progress in daily activities, particularly regarding sitting balance, and weakness in all limbs persisted. A week later, the patient was discharged at the request of family members, as they observed no significant changes in his condition, and a follow-up MRI could not be conducted due to missed appointments.

Discussion

MBD and WE are distinct neurological issues linked to chronic alcoholism. Italian pathologists first identified MBD through observations of corpus callosum degeneration in heavy red wine drinkers, leading to seizures and coma. This rare disorder primarily affects long-term alcohol users and is characterized by demyelination, necrosis, and sometimes hemorrhages, often due to thiamine and folate deficiencies[3]. While 92.8% of MBD cases occur in those with alcohol dependence, it can also affect non-drinkers with conditions like depression or post-bariatric surgery complications[4]. Alcohol disrupts protein expression in white matter, lipid metabolism, and blood vessel integrity, causing small vessel necrosis, blood-brain barrier disruption, and cytotoxic edema, resulting in symmetric demyelination and ischemic damage to the corpus callosum and other central nervous system structures[2,4].

Clinical manifestations vary significantly and are categorized into three types: acute (mental clarity, ataxia, delirium, dysarthria, coma, or death), subacute (confusion, memory issues, walking difficulties, behavioral changes, excessive sleepiness), and chronic (interhemispheric disconnection syndrome, progressive dementia, unusual behaviors)[3]. Heinrich et al. (2004) identified two main clinic-radiological subtypes: Type A MBD, which presents subacutely with hypertonia, pyramidal signs, altered consciousness, and a poor prognosis, and Type B MBD, characterized by gait issues, interhemispheric disconnection syndrome, and a generally positive prognosis with normal to mild cognitive impairment[5].

Historically, postmortem tissue examination was essential for diagnosing MBD. Now, diagnosis primarily relies on clinical assessment and radiological evaluations, with MRI as the "gold standard." CT scans typically show MBD as a hypodense corpus callosum, while hemorrhages may appear isodense or hyperdense. MRI reveals hypointense lesions in the corpus callosum on T1-weighted images and hyperintense lesions on T2/FLAIR, and DWI sequences, often affecting the cortex and white matter without mass effects. Early stages show reduced apparent diffusion coefficient (ADC) values

due to cytotoxic edema, while later stages may show increased ADC values, indicating demyelination without axonal damage[4]. Magnetic resonance spectroscopy (MRS) indicates elevated choline levels and a high choline/creatine (Cho/Cr) ratio during the acute phase, along with a lactate peak during acute or subacute demyelination. Single-photon emission computed tomography (SPECT) shows bilateral decreases in cerebral blood flow. In acute MBD, the "sandwich sign" appears as hyperintensity in the central corpus callosum on T2/FLAIR images, while chronic lesions may show well-defined cavitations[3,4].

In WE, the medial thalamic nuclei, tegmentum, periaqueductal gray matter, tectal plate, and mammillary bodies around the third ventricle are most affected due to their high oxidative metabolism and susceptibility to thiamine deficiency. MRI shows increased T2/FLAIR hyperintensity involving these regions along with contrast enhancement and restricted diffusion[6].

The enhancement in the corpus callosum likely indicated active inflammation or demyelination related to MBD, while the lack of enhancement in the mammillary bodies, periaqueductal region, and midbrain suggested a chronic condition of WE. Our patient's clinical features and the imaging (CT and MRI) findings suggested a patient presented with MBD rather than WE.

When assessing MBD and WE, differential diagnoses like Korsakoff syndrome, and combined Wernicke-Korsakoff syndrome, as well as demyelinating disorders (e.g., multiple sclerosis), recurrent artery of Heubner infarction, central pontine myelinolysis, and tumors such as astrocytoma and lymphoma should be considered[7].

There is no standardized treatment for MBD, but early thiamine and folic acid administration can aid recovery, while corticosteroids may reduce inflammation. The effectiveness of amantadine is unclear, as it has only been tested with vitamin B[3,8]. Significant clinical improvement was not seen following vitamin supplement treatment in our case. It has been suggested that the combination of MBD and WE influenced the prognosis. A limitation of this case report is the inability to obtain follow-up MRI images, as the family declined further treatment and follow-up imaging was unsuccessful.

MBD has a high mortality rate, particularly in its acute phase, and can lead to various outcomes, including recovery, persistent symptoms, vegetative state and death[9]. Over half of non-alcoholic MBD patients fully recover, compared to about 10% of those with alcoholic MBD. Survivors often experience significant neurological impairments, though some may recover partially or completely[8,9]. Survivors should focus on quitting alcohol, seeking rehabilitation, and obtaining nutritional support.

Conclusion

This case demonstrates the rare co-occurrence of Marchiafava-Bignami disease and Wernicke's encephalopathy in a patient with chronic alcoholism. MRI plays a pivotal role in diagnosis. While treatment with thiamine and nutritional support is essential, outcomes depend on early recognition and intervention. This report highlights the need for heightened clinical suspicion and the importance of addressing chronic alcohol use and malnutrition.

Consent

Written informed consent was obtained ensuring patient's anonymity.

Declaration of competing interest

There are no conflicts of interest.

Acknowledgement

N/A

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