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Case Report

Marchiafava-Bignami disease: an interesting case report!

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ABSTRACT

Marchiafava-Bignami Disease (MBD) is a rare neurodegenerative disease usually associated with chronic alcoholism characterized by demyelination and necrosis of the corpus callosum. Here, we report a 57-year-old patient who presented to our hospital with acute aphasia, disorientation and altered mental state. Diffusion weighted imaging (DWI) magnetic resonance imaging (MRI) revealed areas of restricted diffusion in the corpus callosum, bilateral periventricular white matter and right cerebellar peduncle. Given that the patient was a chronic alcoholic, a diagnosis of Marchiafava-Bignami disease was made. Supportive management with intravenous Thiamine and multivitamins was offered. The patient improved clinically after one week of treatment and a repeat DWI MRI revealed complete resolution of the abnormalities. It is imperative to diagnose MBD promptly and distinguish this from other neurological manifestations related to chronic alcoholism. An early diagnosis can aid in a better outcome and faster recovery.

Keywords: Chronic alcoholism, Corpus callosum, Demyelination, Intravenous thiamine

INTRODUCTION

Marchiafava Bignami is a rare affliction of chronic alcoholism primarily involving the corpus callosum resulting in demyelination and necrosis.[1,2] The main pathophysiology revolves around chronic alcoholism with resultant hypovitaminosis, alteration in the neurotransmitter activity and oxidative injury.

The clinical presentation of the disease encompasses a wide range of symptoms ranging from reduced consciousness, emotional and psychotic symptoms, depression and apathy, aggression, seizures, hemiparesis, ataxia, and apraxia.[1] The disease can present in 3 stages - acute, subacute and chronic form.[3,4]

However, if left untreated, these patients may land in coma as well. The gold standard investigation is magnetic resonance imaging (MRI) brain which may show typical features of restricted diffusion in the corpus callosum and helpful in appropriate clinical setting to guide in early diagnosis.[4,5] A relatively accepted treatment for this condition is intravenous thiamine (vitamin B1) infusion. Some authors suggest usage of high dose steroids, folic acid, and amantadine in the treatment protocol. Our patient is a 57-year-old alcoholic who presented with acute confusion and showed DWI abnormalities in corpus callosum and periventricular white matter.

CASE REPORT

A 57-year-old chronic alcoholic presented to our hospital acutely in a state of confusion, with disorientation and aphasia. Patient had been consuming alcohol for almost 25 years.

We obtained the history from the patient's relative. There was no prior history of sensory or motor deficit, memory loss or previous similar episodes. On examination, patient was hemodynamically stable. CNS examination revealed

GCS of 11, with spontaneous eye opening, incomprehensible speech and purposeful movement to painful stimulus. The patient was stabilized. Investigations revealed normal values of serum electrolytes, serum glucose, transaminases, total counts, C reactive protein and erythrocyte sedimentation rate. Toxicology screen was negative. MRI brain was performed to evaluate for intracranial pathology. It revealed multiple areas of restricted diffusion in the corpus callosum, posterior limb of bilateral internal capsule, parietal white matter and right cerebellar peduncle (Figure 1).

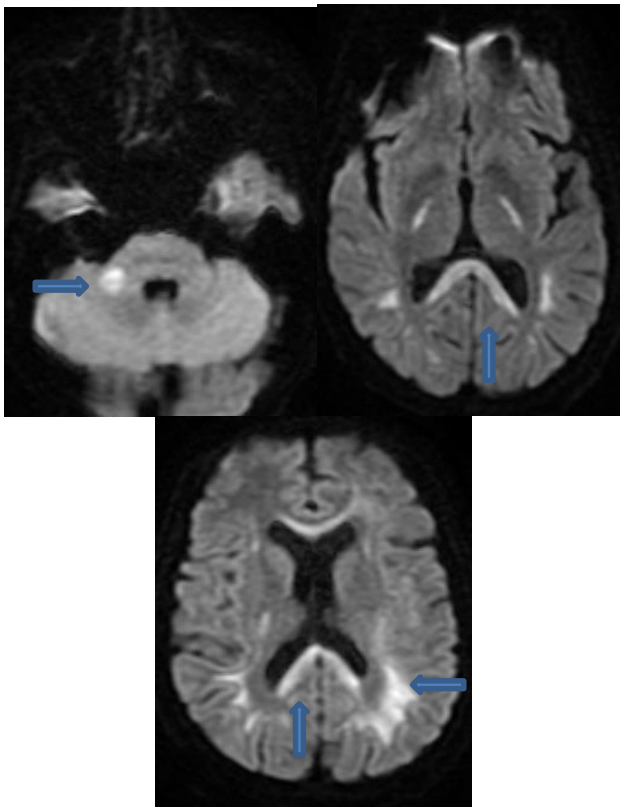

Figure 1: Pre-treatment diffusion weighted images show restricted diffusion areas (arrows) involving corpus callosum, bilateral parietal white matter and right middle cerebellar peduncle.

The diffusion restricted areas showed low intensity on apparent diffusion coefficient (ADC) map and appear hyperintense on FLAIR sequences (not shown).

With these imaging findings, in correlation with the history of alcoholism and clinical presentation, a diagnosis of Marchiafava-Bignami disease was made.

The patient was conservatively managed with intravenous thiamine infusion, intravenous fluids and multivitamins.

He made a significant clinical recovery in one week and a follow up DWI MRI was performed which revealed resolution of the previous restricted diffusion changes (Figure 2).

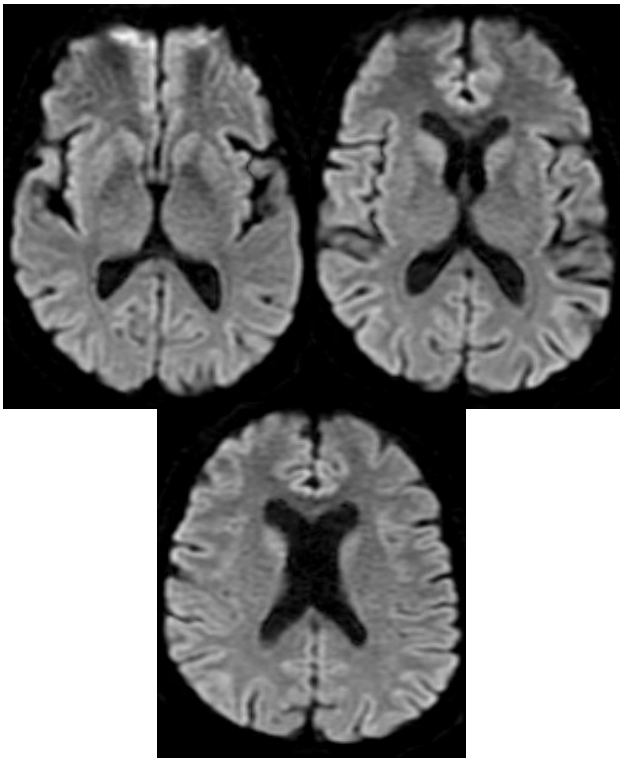

Figure 2: Post treatment DWI show complete resolution of the abnormal areas of restricted diffusion in corpus callosum and parietal white matter.

DISCUSSION

Marchiafava-Bignami disease (MBD) is a rare disorder of demyelination and necrosis of the corpus callosum and the subcortical white matter.[5] The disease also can involve cerebellar peduncles and putamen. The vast majority of patients are male between 40 and 60 years of age with history of chronic alcoholism and malnutrition.[6-8] Prognosis can be worse with background chronic alcoholism, which is the usual clinical scenario.

MBD pathophysiology is unclear. One of the hypotheses is alcohol-associated hypovitaminosis, mainly B1 and oxidative injury to the corpus callosum producing an initial phase of cytotoxic edema, and a later phase of focal demyelination followed by necrosis.[1,9]

Although the clinical features may be quite variable and nonspecific, MBD should be suspected in patients with chronic alcohol abuse and/or malnutrition who present with certain common neurological symptoms.[10] Acute presentation is characterized by a sudden onset of loss of consciousness and seizures. Also, other features can be apathy, aggressiveness, confusion, seizures, and psychosis. Acute MBD needs to be distinguished from Wernicke's encephalopathy, another alcohol-related disease that presents with ataxia, ophthalmoplegia, nystagmus, and confusion.[4] Subacute features can be depression, ataxia, apraxia and agraphia. Chronic forms

can present as progressive severe global dementia, visual hallucinations, and behavioral abnormalities, needs to be differentiated from Alzheimer disease.

Evaluation relies heavily on imaging findings and correlation with a thorough history and physical exam. Laboratory exam including serum electrolytes, serum transaminases, serum glucose, complete blood count, toxicology screening and serum/spinal fluid infectious serology panel form a part of evaluation.

MRI is the gold standard imaging study of choice although computed tomography (CT) may reveal hypodense lesions in the corpus callosum, especially the central portion.[11,12] The typical pathognomonic features on MRI are lesions on the corpus callosum, which shows restricted diffusion in the acute stage, due to cytotoxic edema with or without demyelination.[6,13] Later, this can progress to necrosis and result in atrophy of corpus callosum in the chronic form. Lesions in other brain regions such as anterior/ posterior commissures, optic chiasm, and middle cerebral peduncles are also described and associated with a higher incidence of severe cognitive impairment.[14]

There are no management guidelines or specific proven treatment to date. Literature search on MBD have shown a favorable response to the intravenous thiamine, folate, and vitamin B complexes as well as high-dose corticosteroids.[1,5] Management also includes alcohol cessation, guidance and rehabilitation therapy, and undergo nutritional counselling.[4]

The following differentials may be considered in all patients with acute delirium or acute ataxia, or structural diseases that overlap in the neuroimaging findings which would include Wernicke's encephalopathy, vitamin B12/folate deficiencies and encephalopathy due to drugs/toxins/ infections/metabolic impairments.

Disease severity is variable.[1] Outcomes are poor if there are lesions outside corpus callosum, history of heavy alcohol consumption and severe disturbance of consciousness. Early diagnosis and effective treatment are therefore important to the patient's recovery.

CONCLUSION

It is generally accepted that the Marchiafava Bignami disease is related to alcohol abuse and hypovitamosis (of B complex vitamins). Majority of the patients respond to the treatment with intravenous thiamine and other vitamin B complex. Since the prevalence of alcohol consumption and the incidence of the neurodegenerative sequelae are significantly morbid, it is imperative that the clinician can promptly identify and treat this condition. Also, clinicians should be able to differentiate it from other neurodegenerative conditions with the combination of clinical and MR imaging findings. In appropriate clinical background, the diagnosis of Marchiafava-Bignami disease can be made with reasonable accuracy using DWI MRI and prompt therapy may revert the condition almost completely.

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