



A RARE CASE OF MARCHIAFAVA BIGNAMI SYNDROME IN A CHRONIC ETHANOLIC PATIENT

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ABSTRACT

Marchiafava-Bignami disease (MBD) is a rare condition characterized by demyelination and degeneration of the corpus callosum seen in patients with chronic alcohol intake. This may also rarely occur in non alcoholics who are chronically malnourished. The etiopathogenesis of this condition is based on a complex deficiency of group B vitamins especially in alcoholics. Many patients improve with supplementation of these vitamins. However, a good response is not always observed. The definitive diagnosis of Marchiafava-Bignami disease can be problematic and is based on features of neuroimaging studies, especially magnetic resonance imaging. The present article is a case report of a Marchiafava-Bignami disease in an alcohol-dependent male patient who improved after the administration of parenteral B vitamins. This article also gives a review of literature of Marchiafava Bignami disease.

KEYWORDS : Marchiafava Bignami disease, corpus callosum

INTRODUCTION :

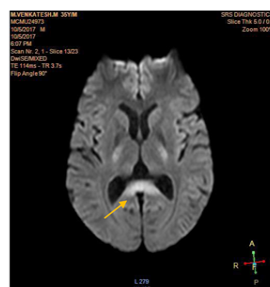
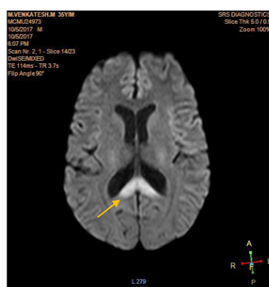
Marchiafava-Bignami disease (MBD) was first described in 1903 by the Italian pathologists Ettore Marchiafava and Amico Bignami. It is a disorder associated with chronic alcohol consumption and characterized by demyelination and necrosis of the corpus callosum [1]. In many cases, magnetic resonance imaging (MRI) also reveals hemispheric white matter lesions [2].

In 2001, Helenius et al reported that about 250 new cases have been reported since the original description of the disease, over 200 of them had died, 30 severely demented or bedridden, and only 20 favorably recovered [3]. Nevertheless, there have been some reported cases with clinical and imaging improvement [3-6]. In 2004, Heinrich et al described two clinical subtypes of MBD: type A predominantly characterized by stupor and coma, pyramidal symptoms and radiological involvement of the entire corpus callosum; type B, with slighter impairment of consciousness level and partial or focal lesions of the corpus callosum [1].

CASE REPORT :

A 35 year old male patient who is a chronic alcoholic was brought to the emergency room in a state of unconsciousness lasting for about 6 hours. There is no history of seizures or weakness of limbs. On examination the patient is not conscious and the vital parameters are within normal limits. Central nervous system examination showed no focal deficits, pupils were small and sluggish reacting to light. All the laboratory parameters like blood sugar, electrolytes, renal and liver function tests were in normal limits (except raised MCV- 105fl). MRI brain was ordered which revealed an isolated ill-defined significant restriction on diffusion showing hypo signal on ADC involving splenium of corpus callosum suggestive of MBD (figure 1). CSF analysis showed normal picture.

On this background patient was initially admitted in ICU and was started on parenteral supplementation of B complex vitamins and other supportive care was given. In a span of 2 days patient gradually regained consciousness and was able to communicate with gestures with a GC score of 10. Treatment was continued and the patient recovered completely in 11 days.



DISCUSSION :

MBD is seen most often in individuals with chronic alcoholism; however, some cases have been reported in malnourished patients with no history of alcoholism. Although chronic alcoholism and nutritional deficits are commonly associated with the disease, etiology is still controversial [7]. There is no prototypical clinical presentation of MBD. It may present as a varied presentation with subtle clinical signs such as reduced consciousness, emotional and psychotic symptoms, depression and apathy, aggression, seizures, hemiparesis, ataxia, and apraxia.

The disease can be acute, subacute, or chronic and may lead to death within weeks or months. The acute form of MBD includes seizures, decreased consciousness and rapid death. The subacute form includes varying degrees of confusion, dysarthria, behavioral abnormalities, memory deficits, signs of interhemispheric disconnection, apraxia, and gait disorders. The chronic form, which is less common, is characterized by mild dementia.

Until recently, a definitive diagnosis of MBD could only be made at autopsy. However, in the era of modern neuroimaging, it is possible to confirm the diagnosis based on a typical clinical profile with a past history of alcoholism and a CT scan or MRI demonstrating specific pathological lesions in the corpus callosum.

The corpus callosum appears hypodense on CT scans, except for cases characterized by subacute hemorrhage, in which the lesions may be isodense or hyperdense. However, when the lesion is small, the CT tends to be normal. In these cases, MRI has greater sensitivity. Callosal injuries are typically hypointense on T1 and hyperintense on T2, sometimes extending to the genu and adjacent white matter. The lesions do not present with a mass effect and, in the acute phase, may present with contrast enhancement [1]. Chronic lesions may progress to "cavitation" with well defined margins. This is called a "positive sandwich sign". The hyperintensities can be found in other regions, such as the centrum semiovale.

Treatment with thiamine, folic acid and other B complex vitamins has been described in several patients with favorable outcome. A review of CT/MRI confirmed MBD cases revealed a linear trend for better outcomes among patients treated with thiamine.

MBD is a rare complication associated with alcohol consumption and should not be forgotten. Despite the poor prognosis, an early diagnosis using MRI and prompt supportive therapy may enable a better outcome. Further studies are needed to clarify the etiopathogenesis of the disease and help define an effective treatment.

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