



Marchiafavae Bignami Disease in Chronic Alcoholic Patient

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Abstract

Marchiafava-Bignami disease (MBD) is a rare, alcohol-associated disorder characterized by demyelination and necrosis of the corpus callosum. It is mostly seen in male alcoholics. It is caused by the combination of alcohol abuse with malnutrition, leading to metabolic, toxic, and vascular disturbances. MBD carries poor prognosis. There are 2 clinical subtypes- TYPE A where patient mainly presents with coma and stupor has a mortality of 90%; TYPE B, which of lesser severity and presents with only mild mental impairment. Clinical symptoms include reduced consciousness, psychotic and emotional symptoms, depression and apathy, aggression. Diagnosis of the disease is made on the basis of clinical findings in combination with neuro imaging. Treatment options tried include thiamine, folate and VITAMIN B12, amantadine and high dose corticosteroids have been tried with limited success. We present a patient who presented with acute onset symptoms and typical Imaging findings findings.

Case Presentation

Clinical Presentation

35-year-old male alcoholic for 7 years presented with 3 episodes of GTCS followed by altered sensorium since 1 day. No history of fever, vomiting. Each episode of seizure lasted for around 10 minutes. First 2 episodes were in home and last one was at casualty while being evaluated by ER physician. He did not regain consciousness in between the seizures. Patient was not responding appropriately to oral commands. History of loss of bowel and bladder control was present. His last binge of drinking was 48 hours prior to admission. Family history was unremarkable. No other comorbidities, or history of similar complaints in the past.

On examination, patient was conscious, disoriented and follows simple commands.

Vitals stable. Moves all 4 limbs against gravity. lip and tongue bites were present. Patient was afebrile, BP - 130/70 mm of hg no neck stiffness. bilateral pupils were equal round and reactive. Examination of fundus was normal.

Investigations

Baseline investigations were done.

Hemoglobin was 14.4. TC was 8900. liver function tests were normal. renal parameters were also normal with creatinine of 0.82 and urea of 24.7

CSF analysis was done which also turned out to be normal with only 3 cell/cumm with predominantly lymphocytes.

Imaging studies were done with CT as the initial modality. Images and discussion of the imaging follows.

Course in the hospital and diagnosis

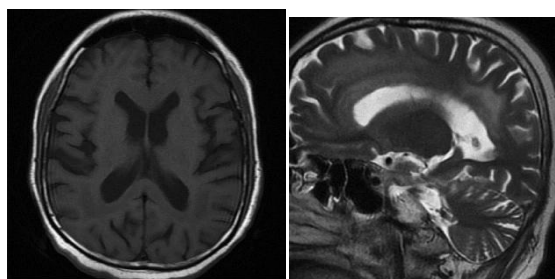
Provisional diagnosis of alcohol withdrawal was done and patient was treated accordingly. Patient's sensorium did not improve even after 72 hours with appropriate treatment.

Electroencephalogram showed diffuse slow waves of 5-8 Hz without epileptiform discharge. CSF study done to rule out infection was normal. CT brain was done as an initial imaging study revealed hypo attenuating lesions in the corpus callosum.

Subsequent MRI brain revealed hypo-intense corpus callosum and periventricular white matter in T1 weighted images with corresponding T2 images showed hyper intensities with true restricted diffusion in DWI image which confirmed the diagnosis of Marchiafava bignami syndrome. He was treated with IV thiamine,



CT coronal section showing hypo densities in the corpus callosum



MRI showed hypo intense corpus callosum and periventricular white matter in T1 weighed images with corresponding T2 showing hyperintensities

Discussion

Marchiafava-Bignami disease, which results from the demyelination of the corpus callosum, can be observed in people with chronic alcoholism. The

presentations of MBD are variable. In the acute stage, the patient often has non-specific neurologic changes, such as confusion, coma, seizure, dysarthria, and hemiparesis^{1,2}. Classic inter-hemispheric disconnection syndrome is often obscure patients with confusional state and it has often been misdiagnosed as Wernicke encephalopathy, alcohol withdrawal syndrome, or encephalitis. The early diagnosis depends on MRI. Conditions with acute onset, impairment of consciousness, wide spread lesions, and cortical involvement indicate an unfavourable prognosis^{1,2}.

The characteristic MR imaging findings are symmetric lesions of the corpus callosum. Lesions maybe also found in the hemispheric white matter, cortex, middle cerebellar peduncles, and internal capsules^{2,3}. During the acute phase, the affected areas demonstrate oedematous change with or without demyelination, appearing hypo intense on T1WI and hyper intense on T2WI. After the acute stage, oedematous change may subside and the corpus callosum gradually recovers its normal intensity. In the case of permanent myelin damage and necrosis, MR imaging typically reveals atrophy of the corpus callosum with cystic change, particularly in the central layer.

The diagnosis of MBD rests mainly on evidence of the callosal lesions. While the corpus callosum may also be affected in other diseases such as ischemic stroke, contusion, multiple sclerosis and lymphoma, in MBD however, it is distinguished from these disorders by the symmetry of the callosal lesions⁴.

Tumours in this location generally have a significant mass effect and contrast enhancement and are easily distinguished from MBD. Differentiating acute MBD from Wernicke encephalopathy is not difficult because, in the latter, MRI shows abnormal signal intensity and contrast enhancement in the mammillary bodies, periaqueductal region and the walls of the third ventricle⁵. Corpus callosal hyper intensity may also be seen in extrapontine myelinolysis (EPM), which may occur in isolation or in association

with central pontine myelinolysis. The lesions in EPM are usually bilateral and symmetrical and may involve any of the subcortical structures⁶.

No standardized treatment protocols have been established in MBD. However, most often patients are treated with thiamine, vitamin B-complex and folate, with good clinical recovery in many patients. Staszewski et al., treated a patient with thiamine, vitamin B-12 and folate and amantadine with improvement⁷. Clinical improvement has been documented using high dose of corticosteroids⁸. The available evidence suggests that an effective and aggressive early treatment is often associated with marked clinical improvement.

In summary, MBD is considered a radiological and medical emergency and early recognition is critical for good clinical outcome. The antimartum diagnosis of MBD mainly depends on the neuroimaging characteristics rather than the clinical features, which are often quite varied. Acute MBD may have a rapid course resulting in death. Early diagnosis and prompt institution of appropriate treatment are essential to improve the prognosis of acute MBD.

Conclusion

Marchiafava-Bignami disease though rare should be considered as a possible differential in alcoholics who otherwise present with an unexplained altered sensorium, aggressive behavior.

CT can be used as an initial screening tool, MRI is found to be more sensitive in the diagnosis. Early recognition and treatment of this syndrome is essential for a favorable clinical outcome

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