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Marchiafava Bignami Disease – A Corpus Callosum Anathema

KEYWORDS: chronic alcoholism, corpus callosum, seizures.

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ABSTRACT *Chronic alcohol abuse may lead to myriad spectrum of neuropsychiatric conditions out of which Marchiafava-Bignami Disease (MBD) is a rare one. It is characterised by symmetrical demyelination of the middle portion of corpus callosum. We present case history of 47 year old male with history of chronic alcoholism presenting with seizures and finally diagnosed as having Marchiafava-Bignami Disease (MBD).*

INTRODUCTION

Marchiafava-Bignami disease (MBD), a complication of chronic alcoholism though rare, cannot be ignored. It is characterised by primary demyelination of corpus callosum. It is defined by radiological and pathological aspects rather than its clinical aspects. There are also other conditions associated with heavy drinking having neuropsychiatric manifestations like peripheral neuropathy, delirium tremens, alcoholic dementia, rum fits, cerebellar degeneration, alcoholic amblyopia, beriberi and Wernicke-Korsakoff syndrome.

CASE REPORT

47 year old male presented to emergency department in an unconscious state. There was history of alcohol binge 1 day prior, followed by two episodes of generalised tonic clonic movements with uprolling of eye balls and tongue bite. There was no history of fever, headache, vomiting, head injury or bowel / bladder incontinence. Patient was a chronic alcoholic with history of alcohol abuse since 20 years, consuming 1 to 1.5 litres of liquor every day. Patient had been admitted 5 years back for alcoholic hepatitis. Patient was not on any drugs. Neurological examination showed a Glasgow Coma Scale of E2M4V4. No focal deficit on motor examination. Meningeal signs were absent. All cranial nerves were normal. Fundus examination was normal. Systemic examination of other systems was normal. A working diagnosis of rum fits v/s seizure disorder was made and patient was admitted in ICU. CT scan was done on emergency basis and was normal. After admission was in post ictal phase for around 6 hours after which his neurological status improved to a GCS of 15/15. On day 2 he started complaining of vertigo and imbalance while walking. On day 4 MRI was done which showed multiple areas of decreased signal intensity involving the genu and the anterior aspect of the corpus callosum on T1- weighted images as shown in figure I. Hyperintensities seen involving the genu and the anterior aspect of corpus callosum on T2 - weighted images as shown in figure II.

Biochemistry results revealed mild increase in AST/ALT and low serum albumin. Hemogram was suggestive of macrocytic anemia with haemoglobin of 9.8 g% and MCV of 102.Serum B12 levels were 89 pg/mL (Normal Range 150-900 pg/mL). Serum Thiamine levels were 61nmol/L (Normal Range 74-222 nmol/L). Chest X Ray, EEG and CSF studies were normal. On the basis of history, clinical features and imaging studies the diagnosis of acute form of Marchiafava-Bignami Disease (MBD) was made. Patient was treated with anti epileptic drugs, thiamine and other vitamin B complex and protein supplements with nutritious diet during hospitalization. At discharge he showed 75-80% improvement in his symptoms of vertigo and ataxia.

Fig I: MRI T1 Weighted Image showing decreased signal intensity involving the genu and anterior aspect of corpus callosum

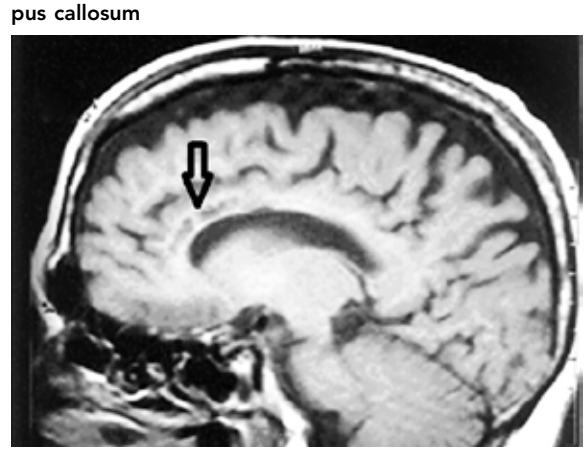

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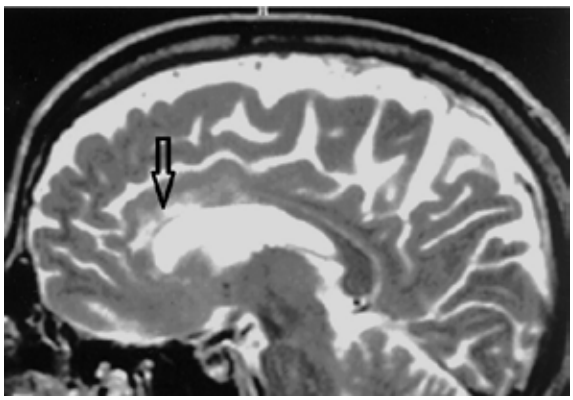
Fig II : MRI T2 Weighted Image showing hyperintensities seen involving the genu and the anterior aspect of corpus callosum.



DISCUSSION

The Italian pathologists Marchiafava and Bignami first described this disease following post-mortem brain analysis of three alcoholics (red wine drinkers) who presented with status epilepticus and coma in 1903.

The underlying mechanism of the disease is still poorly understood. It is probably caused by the combination of alcohol abuse and malnutrition, leading to metabolic, toxic and vascular disturbances[1].There are no characteristic clinical presentations of Marchiafava-Bignami disease. Clinical clues for the disease are reduced consciousness, psychotic and emotional symptoms, depression and apathy, aggression, seizures, hemiparesis, ataxia, apraxia and frequently leading to coma and death[2]. However these are non specific and are seen in many neurological disorders.

The disease typically affects the body of the corpus callosum, followed by the genu, and finally the splenium[3]. The entire corpus callosum may be also involved. Other white matter tracts such as the anterior and posterior commisures and the cortico-spinal tracts may be involved. Lesions may be also found in the hemispheric white matter and in the middle cerebellar peduncles. The subcortical U-fibers tend to be spared[4,5,6]. The corpus callosum degenerates and splits into three layers ("layered necrosis"). The middle layer is the most affected with relative sparing of upper and lower layers responsible for the so called radiological 'sandwich sign'. Only the body or all of the corpus callosum may be affected. The necrosis leads to cystic cavities. During the acute phase of the disease, there are infiltrating macrophages but only a mild inflammatory reaction. The initial loss of myelin eventually gives rise to axonal loss. The reason for preferential involvement of the corpus callosum is not known. Occasionally, the lateral putamina and the cortex (particularly layers 3 and 5) may also be involved[3,4,5].

The common differential diagnosis based on clinical features are rum fits, seizure disorder,wernicke's encephalopathy and stroke.

The diagnosis of MBD rests mainly on evidence of the typical callosal lesions on MRI. The corpus callosum may also be affected in other diseases such as ischemic stroke, contusion, multiple sclerosis, and lymphoma. MBD, however, is distinguished from these disorders by the symmetry of the callosal lesions with relative sparing of thin upper and lower edges[6].

Treatment for Marchiafava-Bignami disease (MBD) is not specific or proven. Common treatment options include supplementation with thiamine, vitamin B12 and folic acid. Patients should also receive nutrional counselling along with rehabilitation from alcoholism.

CONCLUSIONS

Marchiafava-Bignami disease (MBD) is a rare, alcohol induced neurological disorder requiring high index of suspicion and MRI brain for diagnosis. There is a layered necrosis of corpus callosum. Complete abstinence from alcohol is essential for recovery. Vitamin B12, folate and corticosteroids can be tried with variable results.

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