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CASE STUDY Open Journal of Psychiatry & Allied Sciences

Marchiafava-Bignami disease: a rare neuropsychiatric sequelae of alcohol dependence syndrome in a tertiary care hospital in South India

Abstract

Marchiafava-Bignami disease (MBD) was an entity that was discovered only upon autopsies of afflicted individuals. Magnetic resonance imaging (MRI) is now the most sensitive tool to diagnose MBD in vivo. Most common aetiology remains alcohol dependence with malnutrition, and a type A and B have been delineated. We present a case of MBD in chronic alcohol dependence, treated with parenteral thiamine wherein early clinical suspicion proved fruitful and there was significant recovery in the patient.

Keywords: Malnutrition, demyelination, thiamine, magnetic resonance imaging

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INTRODUCTION

The rare, progressive demyelinating, degenerative affliction, Marchiafava-Bignami disease (MBD) has been credited to two pathologists of Italian origin, who explained the acute demyelination of the corpus callosum (CC) in three chronic alcohol dependent patients, discovered at autopsy. CC functions as the crucial white matter bundle in the brain, with axons forming the connections between the cortices of the two cerebral hemispheres. Callosal lesions are best delineated by sagittal magnetic resonance imaging (MRI) wherein entire CC can be assessed; hence, remains the primary investigation in sifting out MBD from other diseases. The hallmark in MBD is lesions that affect the central layers of CC and in a symmetrical fashion.[1]

Prior to the era of brain imaging, MBD was categorised into acute, subacute, and chronic. There are two clinical subtypes of MBD: type A primarily manifested by stupor and coma, pyramidal symptoms and imaging revealing involvement of the entire CC; type B, a less severe variety with lesser degrees of impairment of consciousness and imaging showing partial or focal lesions of CC.[2,3]

We present a case of chronic alcohol dependence in complicated withdrawal with MBD of type B variety.

CASE REPORT

Mr. X, 37-year-old illiterate, unemployed, and married man was brought by his wife with history of single episode of unconsciousness with involuntary movements of upper and lower limbs with irrelevant talk. Initial admission to Intensive Care Unit (ICU) followed by consultation-liaison by psychiatry revealed Mr. X to have a history of long-standing alcohol consumption and was exhibiting confusion. Consuming alcohol (brandy) for the last 20 years with quantity of about 180 ml/day and increased consumption of 750-900 ml/day from the last five years was discovered. Prior attempts to quit alcohol or attempts to seek treatment had never occurred.

Further examination revealed Mr. X to be poorly built and nourished with a body mass index (BMI) of 17.2 kg/m^2. Mental status examination showed dishevelled appearance, poor hygiene, confusion, slowness in activity, and irrelevant speech. Disorientation to time, place, and person was confirmed. Neurological examination revealed hypotonia and decreased power (three/five) in all four limbs and exaggerated tendon reflexes while plantar responses were normal. An ataxic gait was observed. Cranial nerves' examination was normal. There were no sensory deficits. Meningeal signs were absent. Mini-mental state examination (MMSE)[4] score was 14 at the time of admission and the Clinical Institute Withdrawal Assessment of Alcohol Scale, Revised (CIWA-Ar)[5] score was 18. Provisional diagnosis of F10.41 (alcohol withdrawal state with delirium with convulsions) according to the tenth revision of the International Statistical Classification of Diseases and Related Health Problems (ICD-10)[6] was made.

Laboratory investigations revealed hypokalaemia (2.5 mEq/L) which was corrected. His serum bilirubin was

3.1 mg/dL which has later normalised to 1.0 mg/dL. Renal and thyroid functions were normal. There was no evidence of anaemia and the total leucocyte count was normal. He was treated for detoxification with tapering doses of chlordiazepoxide started at 240 mg in four divided doses and was also given oral thiamine 300 mg. His disorientation and ataxia persisted beyond 21 days although overall CIWA-Ar scores fell to six. Degenerative disorders were considered; hence, MRI of the brain was done.

MRI brain revealed findings of hyper-intense signal on T2-weighted and hypointense signal on T1-weighted images. Central hypo-intense with peripheral hyper-intense signal was seen along the splenium of CC on fluid-attenuated inversion recovery (FLAIR) images. Multiple small, discrete FLAIR hyper-intense foci were noted in bilateral fronto-parietal subcortical region (Figures 1 and 2).

Final diagnosis of G31.2 (degeneration of nervous system due to alcohol [MBD]) with F.10.2: alcohol dependence syndrome, according to ICD-10 was made. He was given intravenous thiamine 500 mg/day with injection five per cent dextrose for seven days followed by oral thiamine 600 mg/day for 14 days. Improvement quantified with improved MMSE score (24/30), ataxia resolving, and absence of confusion after 42 days of admission. He had come for follow-up after one month and has been maintaining abstinence (Figure 3).

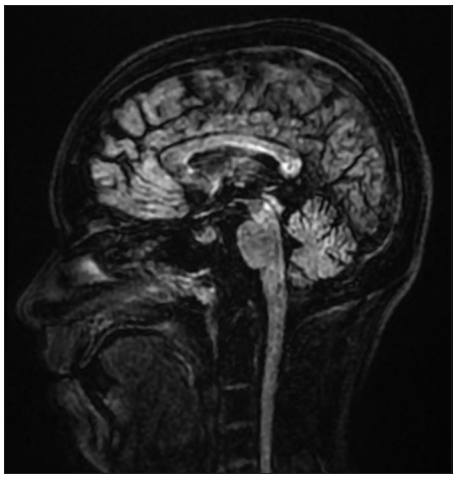

Figure 1: Central hypo-intense with peripheral hyper-intense signal was seen along the splenium of corpus callosum on fluid-attenuated inversion recovery (FLAIR) images in Magnetic resonance imaging (MRI) of brain

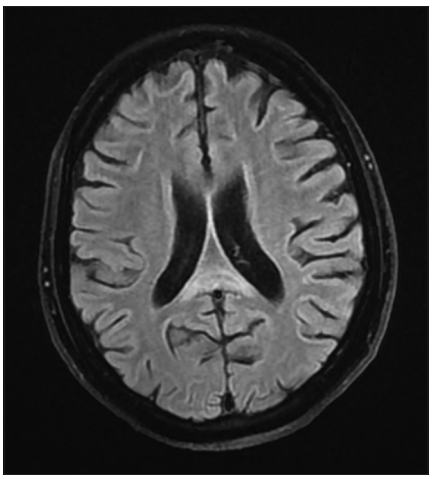

Figure 2: Hyper-intensity of splenium on T2W images in magnetic resonance imaging (MRI) of brain

DISCUSSION

Since 1903, MBD bears the name of two Italian pathologists; but, was first historically described by Carducci in 1898 among red wine drinkers of Italian origin.[7] The aetiology of MBD is most often attributed to long standing alcohol consumption, in association with malnutrition and at times, reports of occurrence with diabetes mellitus, poisoning with cyanide, carbon monoxide poisoning, and sepsis; additionally, sickle cell disease and plasmodium infection are present.[8]

Predominantly though the disease is a manifestation of alcohol abuse with severe malnutrition leading to metabolic, toxic, and vascular pathologies, MBD has varied presentations. Red flags include impaired consciousness, emotional and behavioural abnormalities, low mood, apathy, psychosis or aggression, seizures, paresis, ataxia, apraxia, dysarthria, and coma.[7] The course of the illness may be acute, subacute, and chronic, and eventually death. Acute subtype manifests as a rapid progression from seizures, impairment of consciousness to death. Subacute subtype presents as an admixture of confusion, dysarthria, behavioural disturbances, memory impairments, signs of interhemispheric disconnection with gait abnormalities. Chronic MBD, a rare entity, is marked by mild dementia which is of a progressive deteorating nature.

MRI remains the most sensitive tool for diagnosis of MBD in vivo. The classical radiological feature involves the central layers of CC being affected, with sparing of the dorsal and ventral layers (sandwich sign on MRI).[9] Radiological finding of callosal lesions forms the basis for diagnosis of MBD. Involvement of CC is seen in neurological diseases like ischaemic stroke, contusion, multiple sclerosis, and lymphoma. A diagnosis of MBD is arrived at by the symmetry of the callosal lesions with relative sparing of thin upper and lower edges on MRI.[9] Other differential diagnoses to be considered in chronic alcohol dependence are Wernicke's encephalopathy, Korsakoff's psychosis, alcoholic dementia, cerebellar degeneration, central pontine myelinolysis, alcoholic amblyopia, alcoholic pellagra encephalopathy, and peripheral neuropathy. MBD has not yet found its way into specific treatment guidelines.[10] Cessation of alcohol intake remains crucial. Treatment with thiamine, vitamin B-complex, and folate remains mainstay of therapy, with judicious control of agitation with benzodiazepines/low-dose antipsychotics.[11,12] Our case report highlights the role of early clinical suspicion of a degenerative sequelae of alcohol, with parental thiamine playing a significant role in recovery. Our case can best be categorised as type B MBD associated with lesser severity and radiologically lesser callosal lesions, and fairer prognosis. Table 1 shows case reports of MBD illustrating clinical presentation, therapy, and clinical outcome.

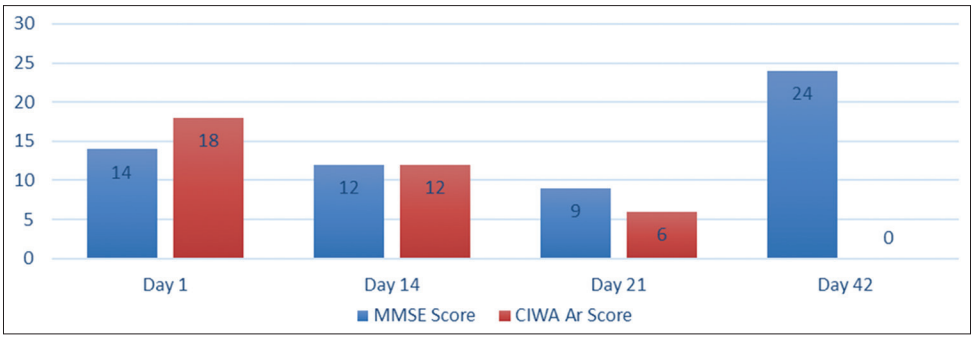

Figure 3: Progress of patient's condition during hospital stay representing cognition by Mini-mental state examination (MMSE) and alcohol withdrawal by the Clinical Institute Withdrawal Assessment of Alcohol Scale, Revised (CIWA-Ar)

Table 1: Case reports of Marchiafava-Bignami disease illustrating clinical presentation, therapy, and clinical outcome

Authors and year of publication	Clinical Presentation	Treatment	Outcome
Ellepola and Abayaweera [13]	Confusion, ataxia, dressing apraxia, and agitation for one week	High doses of parenteral thiamine and a flexible dose of oral chlordiazepoxide	Showed improvement after two weeks
Naaz *et al*. [14]	Suspiciousness, decreased sleep, and violent behaviour for 15 days, urinary incontinence with difficulty in speaking for ten days	Olanzapine 10 mg, oral thiamine 300 mg, folic acid 5 mg, and lorazepam 2 mg	Delusions and hallucinations subsided by four days, incontinence and dysarthria improved by 14 days
Malhotra *et al*. [15]	Acute confusional state and progressive inability to walk	High-dose corticosteroid and intravenous thiamine, intravenous immunoglobulin (IVIG) 0.4 g/kg/day for ten days	Death
Rawat *et al*. [16]	Tremors, sleep impairment, disorientation, faecal and urinary incontinence for three days, four episodes of seizures	Injection thiamine 100 mg and anti-withdrawal medications	Symptom-free by 20 days
Carrilho *et al*. [17]	Memory disturbances for one year, excessive drowsiness developing into torpor and coma	IV thiamine 500 mg/day, high doses of parenteral B vitamins	Death
Hoshino *et al*. [18]	Slurred speech for three weeks, dysphagia and disorientation for three days	IV thiamine 100 mg and complex B vitamin therapy	Symptom-free after 17 days of admission

Conclusion

The neuropsychiatric sequelae of chronic alcohol consumption remain varied and are most often fatal. Our case report goes to prove the necessity to maintain a high clinical vigilance for MBD in alcohol dependence disorders, and advocates for early radiological confirmation with long-term thiamine therapy as a means of reversing the minor type of MBD.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his/her consent for his/her images and other clinical information to be reported in the journal. The patient understands that his/her name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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