Case Report

Marchiafava: Bignami Disease Treated with Parenteral Thiamine

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ABSTRACT

Marchiafava - Bignami disease is rare sequelae of chronic alcohol use. We present a case with transient ischemic attack like presentation and its management with parenteral thiamine. A 53 year old male with history of country liquor use since 32 years was brought to hospital with acute onset of delirium & mild weakness involving motor functions of left side of the body, non-reactive planters and exaggerated tendon reflexes on left side. The MRI showed bilateral hyper intense signal on T2W and FLAIR images & Hypo intense lesion on T1W images involving body, genu and splenium of corpus callosum. The features are suggestive of Marchiafava - Bignami Disease. There have been few guidelines for management of MBD and literature supports use of parenteral thiamine 500mg leading to remission of symptoms and symptomatic improvement. It is advisable to use parenteral thiamine in all cases as it overlaps management of other co-morbidities of chronic alcoholism.

Key words: *Alcohol, marchiafava bignami disease, parenteral thiamine*

INTRODUCTION

Patients of Alcohol use disorder have a conglomeration of clinical features. Marchiafava-Bignami (MBD) a disease seen mostly in chronic alcoholics that results in progressive demyelination and necrosis of the corpus callosum.[1] Magnetic Resonance Imaging (MRI) of brain showing Corpus Callosum (CC) involvement forms the hallmark of the disease. The clinical presentation however includes a large variety of presentations, with no specific or pathognomonic clinical features. Following is a case report of a patient who presented with clinical picture resembling a transient ischaemic attack.

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CASE REPORT

A 53-year-old illiterate man, working as a peon, was brought to hospital with acute onset of unresponsiveness and a fall at home. Thereafter he was unable to walk by himself and not able to speak clearly. Patient had been consuming alcohol (Both Indian Manufactured foreign liquor and country Liquor) for last 32 years. Daily use was up to 1200-1500 ml/day of country liquor. Patient had developed heavy use for last 12 years; there was a past history of hepatic de-compensation with history of jaundice.

Patient had poor built and nourishment with darkened areas over the sun exposed parts indicative of pellagra lesions. The patient was not oriented to time and place and had difficulty in speaking clearly. Patient was able to identify his relatives. The neurological examination revealed mild weakness involving left side of the body, non-reactive planters and exaggerated tendon reflexes on left side. Patient also had impaired joint position sense and fine touch involving lower extremities with swelling of both knee joints. Mini Mental Status Examination (MMSE) was 11 at the time of admission.

Blood investigations showed reduced platelets (92000/cu.mm) and raised ALT (112 IU/l). Patient's computed

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tomography (CT) scan showed features suggestive of changes of global ischemia. Due to suspected cerebrovascular event and inconclusive CT findings MRI was done.

The MRI revealed findings of bilateral hyper intense signal on T2W and FLAIR images. There was Hypo intense lesion on T1W images involving body, genu and splenium of corpus callosum (CC). Subtle restriction diffusion noted. The features are suggestive of Marchiafava-Bignami Disease (MBD).

Hyper intense signal on T2W and FLAIR images involving bilateral fronto-parietal subcortical region [Figures 1 and 2].

Patient was given intravenous (IV) Thiamine 500 mg/day with Injection 5% Dextrose and Normal Saline (NS). For alcohol withdrawal Tab. Lorazepam 2 mg HS was started and tapered slowly. During the course of admission gradually there was return of power and gait improved. Increment in MMSE was till 21 but deficits in memory, attention and language remained [Figure 3].

DISCUSSION

MBD was named after two Italian pathologists who described acute demyelination of the CC at necropsy in three South-Italian male red-wine *drinkers*.[2] The CC comprises axons connecting the cortices of the two cerebral hemispheres and is the principal white matter fiber bundle in the brain.[3] The best method to assess Callosal lesions is sagittal MRI for visualizing the entire CC, which also assumes a pivotal role in distinguishing MBD from other diseases, as the lesions affect the central layers of the CC and are remarkably symmetric.[4]

Heinrich *et al*.,[5] have discussed it as two principal subtypes of MBD may be differentiated: Coma or stupor is the predominant clinical feature in one — referred to as type A–, versus a normal or at most slightly impaired level of consciousness in the other, type B respectively. Since neuroimaging has proved to be a suitable tool for in-vivo diagnosis of MBD.[5]

Raina *et al*.,[13] have described on basis of the onset as acute MBD disease includes seizures, Impairment of consciousness, and rapid death. Sub-acute MBD includes variable degrees of mental confusion, dysarthria, behavioural abnormalities, memory deficits, signs of interhemispheric disconnection, and impairment of gait. Chronic MBD, which is less common, is characterised by mild dementia that is progressive over years. Table 1 shows few case reports in literature with respect to clinical presentation, treatment and outcome.

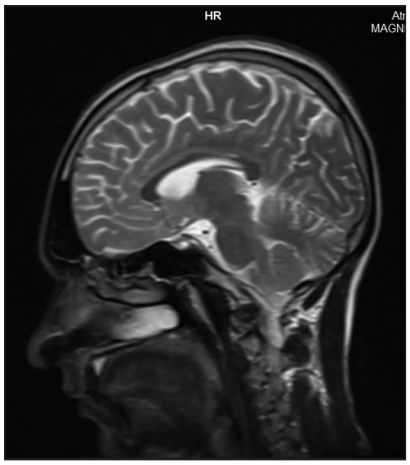

Figure 1: T2 weighted showing hyperintense lesion at Genu of Corpus callosum

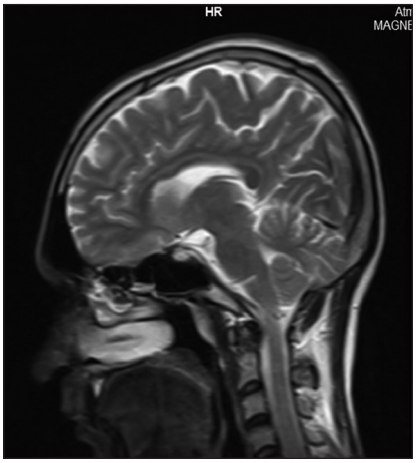

Figure 2: Image showing hyperintense lesion involving body of corpus callosum

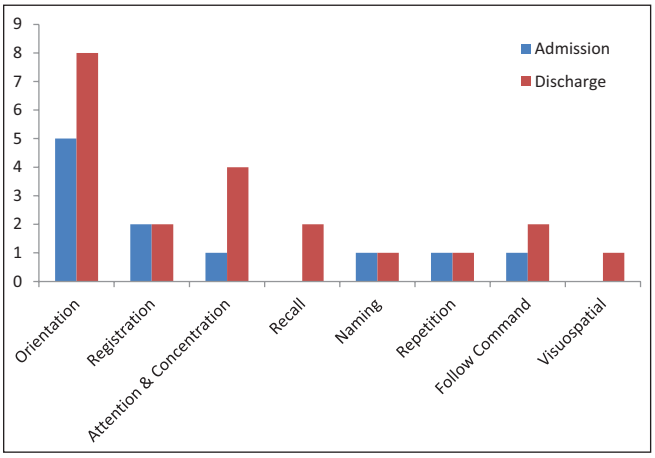

Figure 3: Showing the improvement of MMSE score on parenteral thiamine therapy

CONCLUSION

It is evident from above that it may be imperative to suspect MBD in a patient with delirium and neurological

Table 1: Case reports of marchiafava bignami disease

No.	Authors	Clinical Presentation	Treatment	Outcome
1.	C. S. Tung *et al*.[6]	Vertigo, slurred speech, and progressive gait disturbance	IV B complex 1000mg/day and IV Prednisolone for 3 days	Improvement by day 17, Improved attention and Lesion resolution
2.	SJ Wagh *et al*.[7]	Unconsciousness followed by Complex Partial Seizures	Thiamine, B12, IV Valproate and Phenytoin	-
3.	Yasunobu Hoshino *et al*.[8]	Slurring of Speech. Progressive dysarthria and dysphagia with upper and lower motor neuron signs, and the limb weakness with upper motor neuron involvement mimicking Motor Neuron Disease	IV B complex including Thiamine 100mg daily	Improved Clinically and Reversal of MRI findings in Splenium and Corpus Callosum.
4.	Pedro Sena *et al*.[9]	Acute Confusitional state	IV B-complex and Corticosteroids including Thiamine 100mg	Death
5.	Leeneke Hans *et al*.[11]	Altered Consciousness and Dysarthria. Coma	Thiamine 100mg IV over two days	Improvement of GCS from 3 to GCS 10 after 72 hours of treatment.
6.	Duk Lyul Na *et al*.[10]	Sub-acute onset of Emotional liability, dysarthria, and abnormal behaviour	Multivitamins therapy	-
		Sudden onset of stuporous status		Improvement
7.	Jagdeo P. Rawat *et al*.[12]	Forgetfulness.	Inj. Thiamine (100 mg) OD, Tab. Multivitamin BD and Tab. Lorazepam 10 mg in divided doses.	Improvement and Discharge
		Disorientation	Inj Thiamine 100 mg and anti-withdrawal line of management.	Improvement and Discharge

signs, which show improvement with Thiamine therapy. 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.[10] Hence sharp clinical acumen and urgent neuroimaging can help in early diagnosis. It is advisable to use parenteral thiamine in all cases as it overlaps management of other co-morbidities of nutritional deficiencies and Wernicke Korsakoff syndrome commonly seen in alcohol use disorders. There have been few guidelines for management of MBD and literature supports use of parenteral thiamine[14] (500 mg/day for 5 days atleast) leading to remission of symptoms and symptomatic improvement.

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