Original Research Paper

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General Medicine

A RARE CASE OF ACUTE MARCHIAFAVA BIGNAMI IN CHRONIC ALCOHOLIC MALE

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ABSTRACT Marchiafava Bignami is a rare disorder occurring in chronic alcoholics and malnourished individuals. Clinical presentations include seizures, apraxia, demetia, and comatose state. It is diagnosed by specific MRI abnormalities. Treatment is usually with supplementation of micronutrients and supportive care. It is associated with poor prognosis.

KEYWORDS :

INTRODUCTION

Marchiafava Bignami disease (MBD) is a very rare disorder seen in chronic alcoholics characterised by demyelination and necrosis of corpus callosum. It was discovered in 1903 by Italian pathologists Ettore Marchiafava and Amico Bignami. The etiology of Marchiafava Bignami disease(MBD) is attributed to combination of alcohol induced neurotoxicity and deficiency of B – complex vitamins. Other causes may include callosal myelinolysis due to sudden fluctuation of serum osmolality, occurring as complication of ketoacidosis in diabetic patients or alcoholics. It has also been related to other non-alcoholic conditions like carbon monoxide poisoning, sepsis, cerebral malaria and sickle cell disease.

Histologically, macroscopic features in body and genu of corpus callosum consist of necrotising or cystic lesions. Microscopic features consist of white matter necrosis, abundant macrophages, foamy histiocyte infiltration, small perivascular lymphocytes, gliosis and prominent demyelination that extends symmetrically into centrum semiovale. Oligodendrocytes reduced in number.

The symptoms of MBD can present in acute, subacute and chronic form:
(a) Acute – seizures, sudden loss of consciousness, aggressiveness, confusion, psychosis
(b) Subacute – depression, ataxia, apraxia, agraphia, dysarthria, anomia.
(c) Chronic – progressive severe dementia, visual hallucinations.
On neurological examination, patient may be lethargic, comatose or unconscious. Apraxia may be present due to interhemispheric disconnection in view of damage to fibres of corpus callosum. Tremors, spasticity, delirium tremens and gait abnormalities may be seen.

Routine investigations like hemogram, blood glucose level, renal and liver function tests, serum electrolytes, cerebrospinal fluid routine and electron microscopy are uneventful. Hemogram may show macrocytic anaemia due to vitamin B12 deficiency.

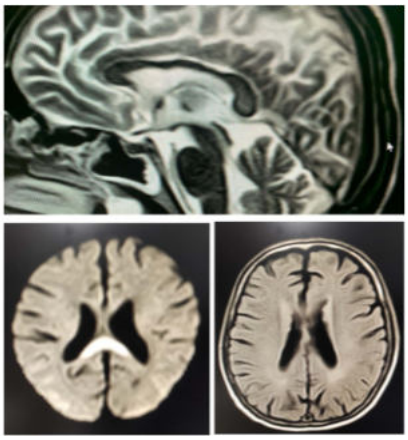
Radiographic features:
(a) CT scan may show hypoattenuating regions in the corpus callosum.

(b) MRI – Sagittal MRI shows **Sandwich Sign** due to involvement of central layers with relative sparing of dorsal and ventral extremes.

T1**- hypointense** foci in corpus callosum in acute phase
T2- **hyperintensity** in corpus callosum in acute phase.Hypointense focal lesions in subacute phase. **Ear of the lynx sign** seen

MR Spectroscopy – NAA/Cr ratio shows progressive decrease to minimum level after the first few months followed by partial recovery after around 11 months.

MRI Brain of MBD Patient
Sources: Radiology department of B.J.M.C, Ahmedabad



CASE REPORT

A 47 year male patient, who is chronic alcoholic since 20 years, was brought to casualty in medicine department with history of alcohol binge that morning followed by single episode of convulsion followed by loss of consciousness. Patient did not have any comorbidities and no significant family history present. On examination, patient was unconscious, non responsive to deep pain stimulation. Deep tendon reflexes were absent and bilateral plantar reflexes were flexor. Pupils were mid dilated, non reactive to light. Pulse rate was 110/min, blood pressure was 120/80 mmHg, RBS was 130 mg/dl and oxygen saturation was 85% at room air. Patient was intubated and kept on mechanical mode of ventilation. On further evaluation, routine investigations like hemogram, renal and liver function tests, electrolytes, urine routine and microscopy, thyroid function tests, vit B12 levels, arterial blood gas analysis, cerebrospinal fluid routine and microscopy were unremarkable. HIV,HBsAg, HCV were negative. ECG, chest x-ray, fundus, ultrasonography of abdomen, NCCT Brain did not show abnormality. MRI BRAIN showed (i) altered signal intensity in splenium and body of corpus callosum with sparing of peripheral dorsal and ventral layers – SANDWICH SIGN. (ii) hypointense lesion on T1WI, hyperintense on T2 & FLAIR images and shows true diffusion restriction. (iii) Abnormal T2W/FLAIR hyperintensity is noted at tip of frontal horn of bilateral lateral ventricles – EAR OF THE LYNX SIGN (iv) few linear abnormal T2/FLAIR hyperintensities noted in bilateral parietal and left anterior temporal region in subcortical location which shows true restriction on DWI s/o MOREL LAMINAR SCLEROSIS (v) Early changes of

cerebral atrophy. From above findings, patient was diagnosed as having ACUTE MARCHIAFAVA BIGNAMI DISEASE. Patient was treated with thiamine, vitamin B1, B6,B12 and supportive critical care in ICU.

CONCLUSION

Chronic alcoholics presenting in casualty with seizures and comatose condition should be evaluated for MBD once all other possible parameters for altered neurological function are ruled out. Extracallosal lesions, cerebral lobe impairment, severe disturbance of consciousness and heavy alcohol consumption are associated with poor prognosis in MBD.

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CONFLICT OF INTERESTS

There are no conflicts of interests.

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