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Original Research Paper

General Medicine

MARCHIAFAVA - BIGNAMI DISEASE

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KEYWORDS :

INTRODUCTION

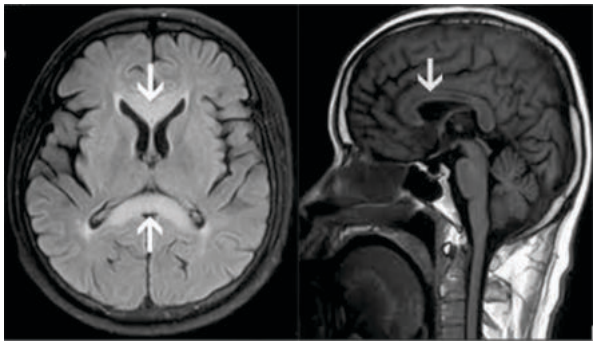
Marchiafava-Bignami disease (MBD) is a very rare disorder of demyelination/necrosis of the corpus callosum and the near subcortical white matter that is especially predominant in ill-fed alcoholics. The disease can be acute, subacute, or chronic. The clinical picture is marked by dementia, dysarthria, spasticity, and walking inability. It is a very rare condition. In the United States, one study found 250 published cases were reported before 2001, suggesting it is likely many cases have gone undiagnosed. International data is similar, always reminding that the prevalence is underestimated because of the non-autopsied patients. Published in approximately 300 case reports.

CASE REPORT

A 43 year old male patient was admitted to GMCH Aurangabad with complaints of acute onset altered sensorium and generalized weakness since 1 day. He was a chronic alcoholic who consumed approximately 90 ml country liquor daily since 15 years. His last drink was 2 days back. He had no other comorbidities.

There was no history of headache or seizures.
On examination he was drowsy, disoriented.
PR-90 bpm, BP-180/90 mmHg, Spo2-99%
No significant findings on general examination
CNS- Patient was drowsy, disoriented
Following simple commands
Not responding verbally.
No cranial nerve FND
Moving all 4 limbs
Power 5/5 all joints
Planters bilateral flexors

IMAGES



MRI BRAIN showing altered signal intensity in selenium of carpus callous appearing hyper intense on T2 ,and showing restricted diffusion on DWI suggestive of MARCHIAFAVA BIGNAMI DISEASE.

CLINICAL COURSE

On admission,NCCT Brain was done to rule out SDH which does not reveal any abnormality Blood investigation
CBC -> hb 13.9 ,TLC -6100 ,platelets 1.26 lac
Urea 30 ,creat 0.8 ,TB 1.0 ,SGOT 33
Electrolyte- Na 135 ,K 4.8 ,BSL 122 mg%

Considering alcohol withdrawal,he was started on Inj Thiamine,Chlordiazepoxide and T. Lorazepam . Repeat NCCT Brain was done on day 2 of admission to rule out infarct which might have been missed on first NCCT.

MRI Brain was done on day 3 of admission in view of no improvement in sensorium. It was reported as altered signal intensity in splenium of corpus callosum appeaing hyperintense on T2 and FLAIR, and showing restricted diffusion on DW with corresponding low ADC values and no blooming on GRE with surrounding cytotoxic edema suggestive of MARCHIAFAVA BIGNAMI DISEASE.

Patient was started on Inj Thiamine TDS and other supportive care .

Clinical improvement was observed over 5 days in which he was responding to verbal commands and was able to ambulate by himself He was discharged on day 7 of illness.

DISCUSSION

MBD does not show any specific geographical distrubution. Cases from all over the world are reported.

No definite etiology has yet been described. The main hypothesis for its pathogenesis is that the disease is a result of B vitamin deficiency. There is no prototypical clinical presentation of MBD. It may present as a varied presentation with subtle clinical signs such as reduced consciousness, emotional and psychotic symptoms, depression and apathy, aggression, seizures, hemiparesis, ataxia, and apraxia. (3)

CT can be normal in this disease. MRI is usually required for the diagnosis. The radiological lesions involve the central portion of the corpus callosum, with sparing of the dorsal and ventral layer, producing the well-known "sandwich sign", the characteristic diagnostic sign of MBD. The antimortem diagnosis of MBD mainly depends on the neuroimaging characteristics rather than the clinical features,which are often quite varied.

CONCLUSION

Marchiafava-Bignami disease is a rare disorder. Limited case reports have been published from India. Three case reports from Maharashtra were found. 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. (2)

This patient showed a good response to Inj. thiamine and could be discharged from the hospital. Further studies are needed to clarify the etiopathogenesis of the disease and help define an effective treatment.

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Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

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