VOLUME - 13, ISSUE - 08, AUGUST - 2024 • PRINT ISSN No. 2277 - 8160 • DOI : 10.36106/gjra

Original Research Paper

General Medicine

A CASE REPORT OF MARCHIAFAVA-BIGNAMI DISEASE: A RARE CLINICAL ENTITY IN CHRONIC ALCOHOLISM

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ABSTRACT Marchiafava-Bignami disease (MBD) is a rare neuro degenerative disease characterized by demyelination of corpus callosum. The disease clinically presents with various manifestations resulting in MBD type A and type B on the basis of clinical condition, extent of callosal involvement and extracallosal involvement at brain magnetic resonance imaging (MRI), and prognosis. We report a patient affected by MBD, who presented with genu and body of corpus callosum lesion in brain MRI and achieved a favorable recovery.

KEYWORDS : Corpus callosum, Centrum semiovale, Thiamine, Alcohol use disorder

INTRODUCTION

Marchiafava-Bignami disease (MBD) is a rare neurological disease characterized by primary degeneration of the corpus callosum associated with chronic consumption of ethanol. MBD occasionally occurs in chronically malnourished, despite nonalcoholics. Clinical manifestation of MBD is nonspecific with a wide variation, like difficulty in walking, paraparesis or quadriparesis, altered mental status, seizure, and even coma or death. A deficiency of group B vitamins is the main etiopathogenic hypothesis, and many patients improve after the administration of the multivitamins . Magnetic resonance imaging (MRI) brain is generally used to support the diagnosis.

Case Report

We present a case of a 35 year old male brought to our tertiary health care centre by his relatives in altered sensorium with complaints of headache since 15 days , tremors, bilateral upper limb and lower limb weakness since 8 days, slurring of speech since 4 days and multiple episodes of vomiting 2 days ago before presenting to the hospital. He had no history of loss of consciousness, fever, head injury, convulsion, any known cardiac illness. He was not a known case of diabetes mellitus, hypertension.

Patient was known case of alcohol use disorder since last 10 years , average 30 standard units of alcohol/week for the past 7 years, country liquor with last drink taken 15 days back.

On presentation to the casualty, his Glasgow coma scale (GCS) score was 13 (E4 V4 M5). He showed no signs of meningeal irritation, no fever, and had normal vital signs.

His pupils were normal and reactive to light. His examination revealed upper motor neuron signs in the bilateral upper and lower limbs. Fundoscopy was normal ,the cerebrospinal fluid (CSF) showing no evidence of pleocytosis or elevated protein levels.

Vitally the patient had a pulse rate of 102/min, regularly regular, good volume, no radio radial or radio femoral delay, no pulse apex deficit with all peripheral pulses palpable. The blood pressure of the patient was 100/70 mm Hg and the SpO2 was 97% on room air. The patient had no pallor, icterus, clubbing, cyanosis, oedema or lymphadenopathy. Other systems normal.

Neurological Examination –

The patient was lying in a supine position, conscious, not oriented to time, place and person not obeying given commands, afebrile with a lean built. Cranial nerve examination was normal; The patient had no bowel and bladder incontinence; slurring of speech present ;Power in bilateral upper limb was 4/5 and bilateral lower limb was 4/5 ;There was spasticity in all four limbs. The deep tendon reflexes of left upper limb and lower limb were brisk and that of right upper limb and lower limb were normal. The bilateral plantar reflex was extensor. The patient could experience pain sensations in the bilateral upper limbs and lower limbs. Pupils were bilaterally symmetrical and reactive to light. No signs of meningeal irritation ;No cerebellar signs noted.

Investigations

On ECG the heart rate was 110/min with normal axis and no significant ST-T segment changes. Chest Xray of the patient revealed no significant findings. Fundus examination of the patient was normal.Hemoglobin – 10.3 g/dL; TLC – 8,600/cumm; RBC – 3.51 x 10 ^ 6/cumm; Platelet count – 2.16 x 10 ^ 6/cumm; hematocrit – 32.2%; mean corpuscular volume – 108.6 fL; urea – 44 mg/dL.His laboratory tests for Vitamin B12, folate was normal. HIV status was negative; serum creatinine – 1.2 mg/dL;Na – 139 mEq/L; K – 3.9 mEq/L; total bilirubin – 0.9 mg/dL of which direct – 0.3 and indirect 0.6; total protein – 6.4 g/dL of which serum albumin – 3.1 and serum globulin – 3.3. CSF cytology – No Nucleated cells; CSF protein – 36mg/dL and sugar –68mg/dL.

Mri Brain (plain And Contrast)

Genu and body of corpus callosum appears bulky with T2/FLAIR hyperintensities in genu and body of corpus callosum and adjacent bilateral subcortical white matter with involvement of subcortical U fibers of frontal, parietal lobe. Few areas of blooming noted within adjacent to white matter likely microhemorrhage. It show no diffusion restriction on DWI corresponding drop on ADC with no enhancement on post contrast study. The lesions spared the thalamus, cerebellum, pons, and subcortical region. From above Imaging findings possibility of Marchiafava-Bignami disease more likely.

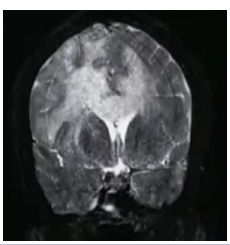

Fig.No. 1(t2 Coronal Mri)

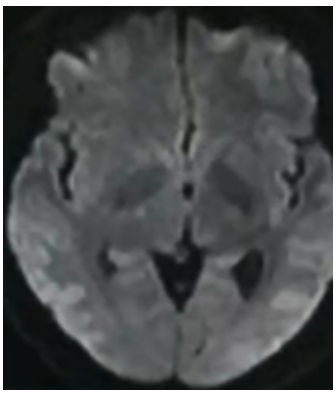

Fig. No.2 (Diffusion weighted image (DWI))

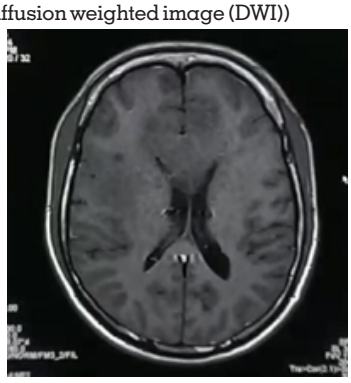

Fig.No.3 (T1 contrast MRI)

Showing no contrast enhancement

The patient was administrated with intravenous thiamine (300 mg/ d for 5 d), vitamin B complex, vitamin B12 (2ml/d for 5 d), folate (5 mg/d) from day of admission. His sensorium improved gradually and he got oriented to time, place and person after 2 days of initiating treatment. Power got improved in all four limbs ,diplopia improved, however hypertonia ,hyperrefexia and slurring of speech was present at the time of discharge .On discharge, the patient was advised strict alcohol abstinence with rehabilitation and proper nutrition.
On follow-up after 6 weeks, patient was independent with daily activities, obeys commands, power, tone and reflexes , speech was also normal. On follow up MRI after 6 weeks, there was definite resolution in the radiological lesions.

DISCUSSION

Although first described by Carducci in 1898 in Italian red wine drinkers, it was in 1903, that the Italian pathologists Marchiafava and Bignami described a unique alteration of the corpus callosum in three alcoholic patients who died after having seizures and coma. The disease affects persons in middle and late adult life. With a few exceptions, the patients have been males and severe chronic alcoholics.

MBD is a rare disorder of unknown etiology characterized by demyelination of the corpus callosum with various clinical manifestations which is often mismanaged and mistreated. Chronic alcohol abuse plays an important role in its development, even though MBD has been occasionally diagnosed in nonalcoholic patients as well.

The underlying mechanism of the disease is still not understood. It is probably caused by the combination of alcohol abuse and malnutrition, leading to metabolic, toxic and vascular disturbances. Possible mechanisms include cytotoxic edema, blood-brain barrier breakdown, demyelination, and necrosis. Pathologically, MBD is characterized by symmetrical demyelination and necrosis of the central part of the corpus callosum, with relative sparing of thin upper and lower edges. Subsequently, necrosis leads to cavitation and atrophy of the corpus callosum in chronic stages .

The diagnosis of MBD rests mainly on evidence of the callosal lesions. The corpus callosum may also be affected in other diseases such as ischemic stroke, contusion, multiple sclerosis, and lymphoma, epilepsy, antiepileptic drug withdrawal, acute disseminated encephalomyelitis, infarction, viral and bacterial infections, especially influenza, HIV, and hypoglycemia, other demyelinating diseases. MBD, however, is distinguished from these disorders by the symmetry of the callosal lesions with relative sparing of thin upper and lower edges.

Other neuropsychiatric conditions associated with chronic heavy drinking are: Wernicke's encephalopathy, Korsakoffs psychosis, alcoholic dementia, cerebellar degeneration, central pontine myelinolysis, and peripheral neuropathy. These conditions can be differentiated on the basis of clinical and laboratory analysis, such as occurred in the patient reported herein. Wernicke's encephalopathy was ruled out on the basis of clinical presentation and site of brain MRI lesion.

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. The course of the disease may be acute, subacute or chronic and may lead to death within weeks to months. Marchiafava-Bignami disease may present in various clinical forms.

Acute Marchiafava-Bignami disease includes seizures, impairment of consciousness, and rapid death.

Subacute Marchiafava-Bignami disease includes variable degrees of mental confusion, dysarthria, behavioral abnormalities, memory deficits, signs of interhmispheric disconnection, and impairment of gait.

Chronic Marchiafava-Bignami disease, which is less common, is characterized by mild dementia that is progressive over years.

There are two types of Marchaifava Bignami Disease(MBD):

Type A MBD is a subacute presentation with hypertonia, pyramidal signs, impaired consciousness, T2 callosal hyperintensity and an overall dismal prognosis.

Type B MBD presents with gait disturbance, interhemispheric disconnection syndrome, normal to slightly impaired mentation, T2 callosal hyperintensity and an overall good prognosis

CONCLUSIONS

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

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