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"Marchiafava-Bignami Disease In A Nonalcoholic Younge Male: A Rare Disease"

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Abstract*: Marchiafava-Bignami disease (MBD) is a rare neurological disorder mostly seen in chronic alcoholic and malnourished patient with pathological hallmark of corpus callosum demyelination and necrosis. We report a case of marchiafava bignami disease in a chronic kidney disease patient, without alcoholism or malnutrition. A 20-year-old man presented with sudden onset of altered sensorium with multiple falls in three days. Neurologic examination showed dysarthria and ataxia but, otherwise, normal cranial nerves, motor and sensory functions, and tendon reflexes. Brain MRI showed symmetric abnormalities in the corpus callosum. In addition, demyelination was also observed in subcortical white matter in bilateral lateral frontal lobes. His symptoms significantly improved after stabilization and normalization of his serum creatinine level after a dialysis. The underlying pathophysiology of the development of MBD in our case is likely to be osmotic stress from chronic kidney disease causing structural and functional disturbance of oligodendrocytes.*

Keywords: *Nonalcoholic, Corpus callosum, Sandwich sign*

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Case Report:

A 20-year-old male patient presented to our institution with complaints of altered sensorium and multiple falls since three days followed by loss of consciousness since one day. He was nonalcholoic.. There was no history of fever, headache, vomiting, seizure, diabetes or hypertension. On clinical examination patient was not malnourished and his Glasgow coma scale was 9 (E2V3M4). Signs of meningeal irritation were absent. Pupils were of normal size and normal reacting. Laboratory results revealed normal haematological profile. Liver function test showed increased serum gamma glutamyl transpeptidase, bilirubin, alkaline phosphatase, and alanine transaminase. Renal function test showed elevated creatine level- 5.5. mg/dl.

Imaging Findings:

MR imaging showed abnormal T2W/ FLAIR image hyperintensity in genu and body of corpus callosum with involvement of its central part with sparing of its superior and inferior margin which give characteristic Sandwich sign [Figure 1]. Corpus callosum, posterior limb of internal capsule, cerebral peduncle showed multiple foci of blooming on Fast Field Echo images (FFE), suggestive of hemorrhagic necrosis. [Figure 2] . Splenium of corpus callosum showed diffusion restriction on diffusion weighted image (DWI) [Figure 3]. Few linear T2WI and Fluid attenuated inversion recovary (FLAIR) hyperintense foci were seen in subcortical white matter in bilateral lateral frontal lobes suggestive of Morel Laminar Sclerosis. [Figure 4]. Other imaging findings included periventricular demyelination . On the basis of history, clinical features and imaging findings the diagnosis of Marchiafava bignami disease was made. However, other lesions involving corpus callosum like multiple sclerosis, hemorrhagic leukoencephalopathy were included in the differential diagnosis.

I. Discussion:

Marchiafava-Bignami disease (MBD) is a rare, fatal neurodegenerative disease. Pathological hallmark is corpus callosum demyelination [1]. It was first reported in Italian wine drinker about 110 years ago.[2]. It was first described by two Italian pathologists, Ettore Machiafava and Amico Bignami [3]. Pathological changes include symmetrical demyelination and necrosis of the central part of the corpus callosum, with relative sparing of dorsal and ventral layers. Other structures of the CNS like optic chiasm and tracts, putamen, anterior commissure, cerebellar peduncles and, cortical gray matter and U fibers may be involved [3,4] Cortical damage in the lateral frontal and the temporal lobes, mainly in the third (although sometimes also in the fourth) cortical

layer with damage to the white matter tracts of the corpus callosum occure. In these area of cortical damage the neurons degenerated and were replaced by glial cells. Morel described this as cortical laminar sclerosis (now known as Morel cortical laminar sclerosis). [5] Morel cortical laminar sclerosis is secondary to MBD.

Chronic alcoholism and/ or malnutrition is closely associated with chronic MBD. [3,4] MBD can also occur in patients who have no history of alcohol abuse [6,7]. The main hypothesis for MBD pathogenesis is that the condition may be associated with malnutrition or vitamin B deficiency [6]. Other etiologies are cyanide, CO poisoning and sepsis, as well as sickle cell disease and Plasmodium infection. After a literature review of 100 studies in 2017, Fernandes et al. [8] suggested a synergism between ethanol-induced neurotoxic effects and hypovitaminosis B, particularly B1. MBD have been reported in patients with dramatic fluctuations in serum glucose in the setting of poorly controlled diabetes. [9,10,11]. Myelinolysis of the corpus callosum occur due to abrupt changes in serum osmolality. In addition to alcoholism, malnutrition, and wide fluctuations in serum glucose, Jorge et al. describe a case of MBD in a young trauma patient in 2015. [12]

Imaging findings:

On CT, the corpus callosum appears hypoattenuated. The areas with subacute bleeding may be iso or hyperattenuated. The corpus callosum appears hyperintense on T2-WI, hypointense on T1-weighted images and proton density-weighted MR images during the acute phase. During the subacute phase, cystic lesions and small foci of T2 hypointensity can develop, most likely because of hemosiderin deposition. In chronic stage signal intensity alterations become less evident but residual atrophy of the involved structure is seen[14].

Clinical and neuroradiological classification of MBD

- Type A: acute to subacute onset of consciousness impairment, pyramidal tract signs, limb hypertonia, seizures, hyperintense swelling of the corpus callosum on T2-weighted MR sequences and is associated with poor prognosis.
- Type B: normal or slightly impaired level of consciousness, dysarthria, gait disturbance, signs of interhemispheric disconnection and hyperintense lesions on T2-weighted MR sequences partially involving the corpus callosum. Type B has favourable prognosis and lesions may reverse suggesting an underlying oedema rather than demyelination[15].

On FLAIR images, central hypointense signal and peripheral hyperintense rim producing "sandwich sign" is seen, which is the characteristic diagnostic sign of MBD. Central hypointense signal represents necrosis & hyperintense signal represents gliosis. [16]. MR spectroscopy (MRS) shows increase in choline, increased choline/creatine (Cho/ Cr) ratio in acute phase. Lactate peak is usually seen in the acute/subacute phase of demyelination. SPECT studies shows bilateral reduction in cerebral blood flow [17].

Differential diagnosis includes infarction of recurrent artery of Heubner, neoplastic disease such as astrocytoma or lymphoma, demyelinating disease such as multiple sclerosis (MS), progressive multifocal leukoencephalopathy, or acute disseminated encephalomyelitis. MS is by far the most common and needs to be ruled out. Compared to MS, MBD has symmetric and oedematous spots restricted in corpus callosum on brain CT scans or MRI [18].

Treatment of MBD is usually empirical and consists of multivitamins, corticosteroids, stabilization of plasma glucose and other deranged metabolites and supportive care. Early diagnosis and prompt appropriate management are critical in reversing the underlying pathophysiology in the early stage.

II. Conclusion

Marchiafava-bignami disease is a rare neurodegenerative disease with chronic alcoholism is a most common etiology. But it can be seen in patients with acute osmotic imbalance. It can be diagnosed by its characteristic findings with MRI .

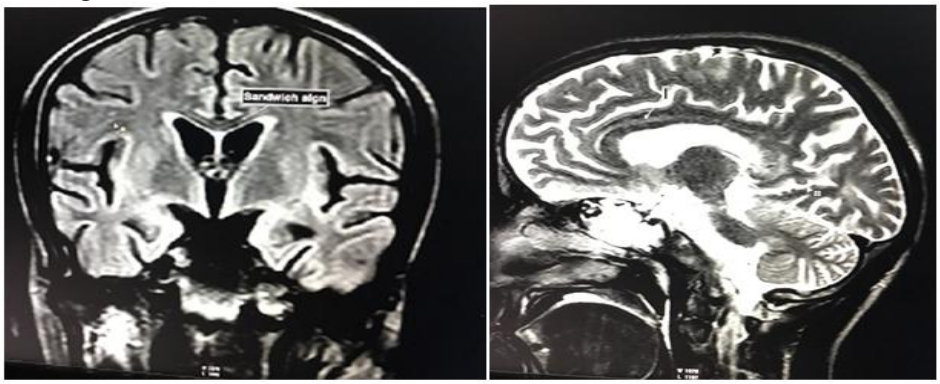
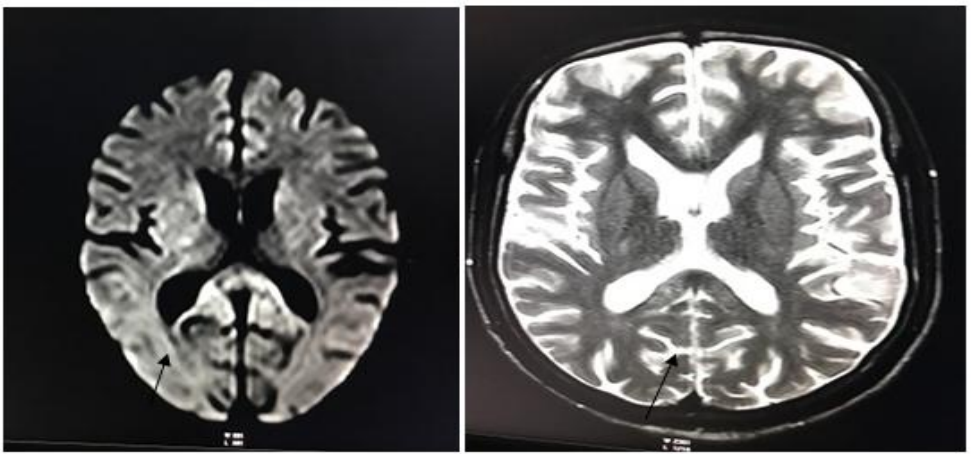
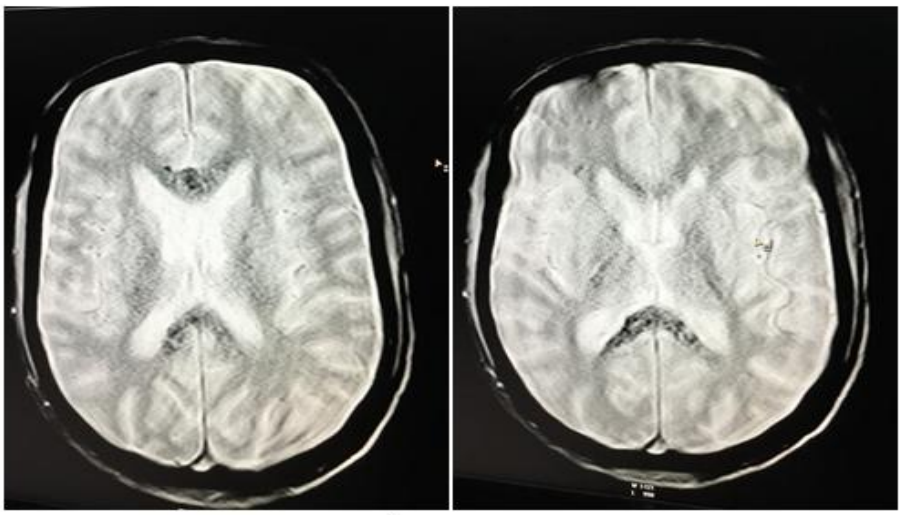


Figure 1: (a)FLAIR Coronal image (b)T2W saggital image- Sandwich sign (arrow).



a b

Figure 2: (a)Diffusion weighted image (b= 1000) showing diffusion restriction in splenium of corpus callosum (arrow) (b) T2W Axial image showing hyperintensity in splenium of corpus callosum more on right side (arrow).



a b

Figure 3: (a and b) FFE-(Fast Field Echo) showing areas of blooming and genu and splenium of corpus callosum suggestive of hemorrhagic necrosis (arrow)

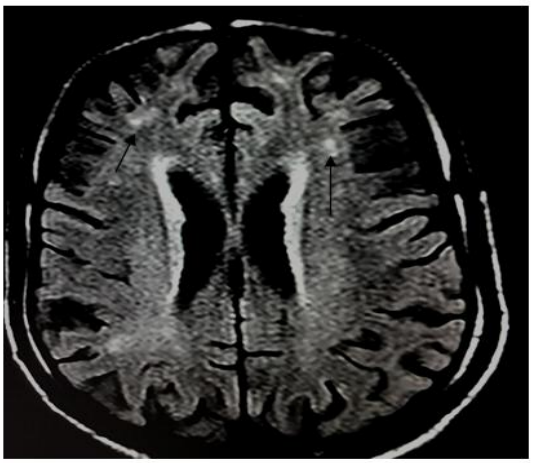


Figure 4: FLAIR Axial image showing patchy hyperintensity in subcortical and deep white matter in bilateral frontolateral cortex (arrow).

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