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Case Report

CASE REPORT

Marchiafava-Bignami Disease: A Rare Disease with MRI Findings and Literature Review

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ABSTRACT

Marchiafava bignami disease (MBD) is an unusual complication of chronic alcoholism that typically results in demylination of corpus callosum, but sometimes it can involve the adjacent white matter and even the subcortical and cortical regions. The pathophysiology of MBD is still unknown but thought to be due to vitamin B complex deficiency. Magnetic Resonance Imaging (MRI) is crucial for its diagnosis that shows the characteristic lesions in corpus callosum which may also extend into deep white matter, subcortical and cortical regions. We present a case of Marchiafava bignami disease in 55-year chronic alcoholic referred to us for MRI brain by Medical unit. This case is presented here for its rarity along with its clinical and radiological findings as well as literature review to generate awareness regarding this uncommon toxic complication of chronic alcoholism.

Key Words: Alcoholism, Corpus callosum, Marchiafava bignami disease, MRI

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Introduction

Marchiafava-Bignami disease (MBD) is a rare neurological disease related to chronic alcohol Consumption. It is characterized by primary degeneration of the corpus callosum seen in their middle or late ages.[1] This disease may occasionally occur in patients who are not alcoholic, hence, it is thought that alcohol is not a single reason for MBD.[2] The exact pathophysiology of MBD is still not elucidated but thought to be due to vitamin B complex deficiency.[1,3] Patients usually present with variety of neurological features such as seizures, deterioration of consciousness, irritability, dysarthria, dementia, behavior disorder and limb weakness which are difficult to differentiate from other alcohol related disorder and even sometimes from motor neuron disease. So diagnosis is generally the result of neuroimaging, particularly by MRI, that shows hyperintense lesions without significant mass effect within the corpus callosum, that may extend to adjacent white matter on T2WI, FLAIR and DWI.[2]

We are reporting this case of MBD in a chronic alcoholic who presented with altered level of consciousness, irritability, parasthesia of right upper and lower limb, dysarthria, gait disturbances and dementia, and evaluated with MRI brain. It highlights the importance of MRI as good imaging tool to diagnose MBD in the relevant clinical scenario.

Case Report

A 55-year old chronic alcoholic with daily consumption of alcohol about one pint for last 35 years referred to

radiology department, Civil Hospital Karachi for MRI brain. He had complaints of progressive dysarthria, gait disturbances and dementia over a year but now presented with altered level of consciousness, irritability, and paresthesia of right upper and lower limb. There was no history of fever. On neurological examination, he responded to eye opening on pain with incomprehensible sounds and flexion withdrawal of limbs to painful stimulus, with total Glasgow coma scale of 8/15. Reflexes were diminished. There were no signs of meningeal irritation and cranial nerve palsies. There was nothing special in the patient's medical and personal history except the alcoholism. Family history was also unremarkable. His routine laboratory investigations did not show any pathology. AFB (Acid Fast Bacilli) smear and culture was negative. His liver and renal functions were almost normal. Initial diagnosis of motor neuron disease was made on the basis of clinical picture but his electrophysiological analysis was normal. Cerebrospinal fluid examination wasn't performed. So he was evaluated with MRI which shows hypointense signals on T1WI and hyperintense signals on T2WI and FLAIR in genu and splenium of corpus callosum without significant mass effect, extending into adjacent deep and subcortical white matter (figure 1).

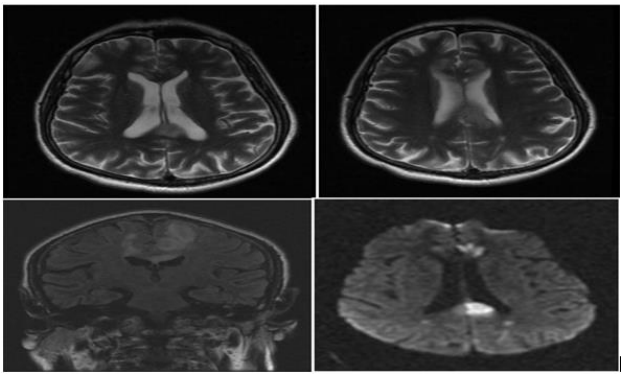

Figure 1: MRI axial T2WI, coronal FLAIR and axial DWI demonstrate hyperintense signals in genu and splenium of corpus callosum extending into subcortical regions (best seen on FLAIR image) showing diffusion restriction.

These lesions showed diffusion restriction on DWI. On the basis of clinical picture, neurological examination findings and typical corpus collasum involvement on MRI, workup for vitamin B level was done that showed low serum level so the diagnosis of Marchiafava bignami disease was made. Then patient was treated with vitamin B complex and showed clinical improvement on 1-month follow up that led to definite diagnosis of MBD.

Discussion

Marchiafava-Bignami disease is a rare toxic neurological disease seen mostly in chronic alcoholics but may rarely occur in non-alcoholics. It is characterized by progressive demyelination and necrosis of the corpus callosum which may extend into the adjacent white matter and occasionally as far as the subcortical and cortical regions.[1,2] The pathological mechanism of MBD is still not completely understand but thought to be due to vitamin B complex deficiency.[1,3] Majority of patients are male between 40 and 60 years of age, and have a history of chronic alcoholism and malnutrition.[1,4] This disease was described by Italian pathologists, Amico Bignami and Ettore Marchiafava, in the postmortem study of a chronic Italian alcoholic in 1903 and approximately 250 cases are reported in the literature.[3,4] The clinical spectrum of this disease is very diverse, including impaired consciousness, seizure, dysarthria, behavior disturbance, limb weakness, dementia, interhemispheric disconnection syndromes and even coma and death.[4,5] The clinical diagnosis of Marchiafava-Bignami disease (MBD) has considerably changed during recent decades with improvement in modern neuroimaging especially brain MRI providing the opportunity of a reliable in-vivo diagnosis.[6] The lesions typically involved the corpus callosum with affection most commonly towards the body of the corpus callosum followed by the genu and finally the splenium.[1] The other sites can also be involved and include the cerebral peduncles, middle cerebellar peduncles, hemispheric white matter as well as internal capsules.[3,7] Cortical involvement in very rare but is a poor prognostic indicator.[8] The pathognomonic MRI findings for the diagnosis of MBD are hyperintense signal lesions without significant mass effect within the corpus callosum, which may extend to adjacent white matter on T2WI, FLAIR and DWI sequences[2], as seen in our patient's MRI. Diffusion restricted lesions on DWI are associated with poor outcome, however these do not predict irreversibility of the lesion.[9] There is no specific therapy for MBD but after prompt diagnosis, early initiation of treatment by Alcohol abstinence along with vitamin B complex and folic acid can improve the outcome.[7]

Conclusion

Marchiafava bignami disease is a rare condition and MRI is very helpful to make early diagnosis especially if the clinical signs are not specific and severe, as prompt therapy may result in survival of these patients. MRI has also role in follow up of patients with MBD. This case was reported not only due to its rarity but also to emphasize the role of MRI in diagnosing MBD and differentiating it from other alcohol related disorders, and to assist the clinicians by reviewing literature and summarizing important features of the MBD.

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