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Marchiafava-Bignami Disease

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Marchiafava-Bignami Disease (MBD) is considered to be a rare and severe complication of chronic alcohol abusers; atrophy and necrosis of the corpus callosum is the hallmark of the disease.

We report a 32-year-old female who presented with headache and diplopia. A brain MRI showed high signal intensity in the splenium of the corpus callosum. This case supports the occurrence of MBD in non-alcoholic patients.

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MBD is a demyelination of corpus callosum without inflammation but other areas of the CNS may be involved. Symptoms of MBD range from non-specific symptoms to coma and status epilepticus[1-4]. The rarity of MBD is a challenge in diagnosis and therapy.

The disease could present in two forms, an acute form characterized by sudden onset, coma, and epileptic seizures, which might end up with stupor and quick death; a chronic form, which could be featured by dementia and dysarthria; this form could last for several years[4].

The aim of this report is to discuss the variety of symptoms that patients with MBD present with and to rule out other differential diagnosis.

THE CASE

A 32-year-old single Saudi female with no known medical illness presented to the ER with one week history of frontal headache, compressing in nature and moderate in intensity which was partially relieved with simple analgesia. Patient gave history of fatigue, dizziness and double vision for 4 days for far objects before presentation. She experienced vertical diplopia in all directions, no history of weakness, numbness or decrease in vision, no history of menstrual charges or alcohol consumption.

On examination, the patient was conscious, alert, oriented, MMSE 30/30 and well-nourished.

Cranial nerves were normal apart from abnormal saccades in both vertical and horizontal gaze with normal pursuits. The tone was normal, power 5/5, DTR normal with downgoing plantars bilaterally. Sensory, coordination and gait were normal. Laboratory work including blood sugar, electrolytes, thyroid function test, auto immune profile and hormonal profile were within normal limits; Bl2 level was 200. Cranial CT was normal. Cranial MRI showed swollen splenium of the corpus callosum with high T_2 and FLAIR signal intensity in splenium. Figures 1 and 2 show high signal on DWI and low signal on ADC keeping with restriction. Visual evoked potential was normal.

During admission, the patient's headache has improved on simple analgesia. The patient was discharged with supplementation of Cobolamin and Thiamin.

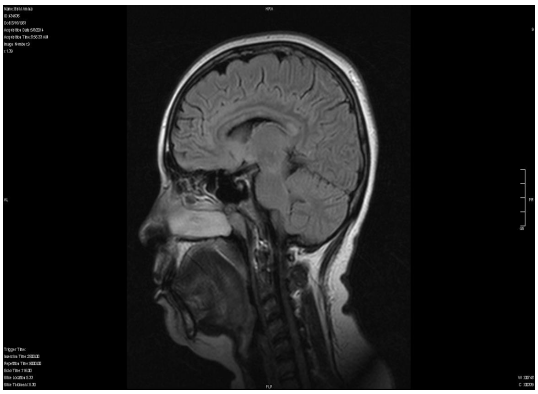

Figure 1a: Sagittal MRI FLAIR of Brain

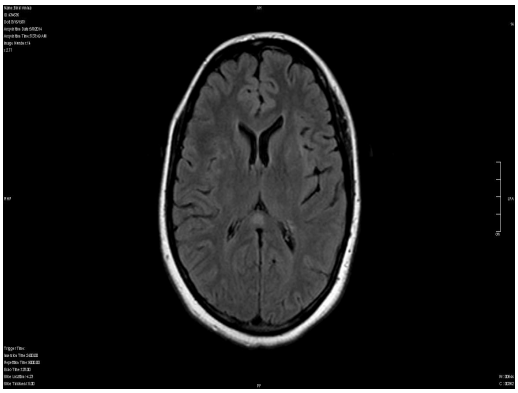

Figure 1b: Axial Showing Hyperintensity in Splenium of Corpus Callosum

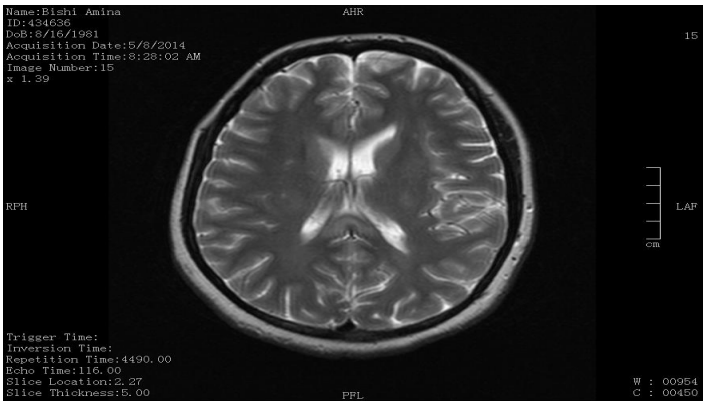

Figure 2: Axial T_2 of Brain Showing High Signal Intensity of Splenium of Corpus Callosum

DISCUSSION

MBD is characterized by primary degeneration of the corpus callosum leading to altered mental status, seizure and multifocal CNS signs as dysarthria, ataxia, aphasia, hemiparesis and dementia and these findings are not fully explained by the characteristic corpus callosum lesion alone. The cause of this condition is unknown but it is more noted in red wine consumers; however, it is not confined to red wine consumers as in our patient[5-9]. Nutritional deficiencies have also been implicated. Toxic factors have been suggested, but no agent has been implicated. Genetic susceptibility has been suspected because of the frequency of report in Italian men in old literature.

MBD diagnosis is rarely made before death because the symptoms and findings are non-specific. Onset is usually in middle age or later[6-9,11]. Well-defined therapy is not available but treatment with vitamin-B supplements has been used with variable degree of improvement.

The literature suggests that MBD is a slowly progressive disease resulting in death in few years; rarely spontaneous remission has been reported; however, any patient with suspected MBD should be evaluated carefully and other treatable conditions should be considered before the diagnosis of MBD arrived at[10,12].

Few previous reports of MBD were documented, but the prognosis in each was different. Lack of follow-up in each case report minimizes the experience about the prognosis of this condition.

CONCLUSION

MBD in non-alcoholics is very rare. MRI could lead to a suspicious diagnosis of MBD; biopsy is the only confirmatory procedure.

We presented a rare case in a non-alcoholic patient presented with headache and diplopia. MRI showed high signal intensity in the splenium of the corpus callosum.

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