

Case Report

Marchiafava bignami syndrome: A rare case report

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Abstract

We have presented here a case report of Marchiafava Bignami Syndrome in a 36 years old male. Marchiafava Bignami Syndrome is a rare entity with few reports in literature across the world. This syndrome is associated with chronic alcoholism in old and aged alcoholics. The exact etiology is unclear and the clinical course can be mild to a very terrible fatal illness. The only treatment is alcohol abstinence and proper nutrition.

Key words

Marchiafava Bignami Syndrome, Chronic alcoholism, Alcohol abstinence.

Introduction

Marchiafava Bignami Syndrome is a rare entity with few reports in literature across the world. This syndrome is associated with chronic alcoholism in old and aged alcoholics. The exact etiology is unclear and the clinical course can be mild to a very terrible fatal illness. The only treatment is alcohol abstinence and proper nutrition.

Case report

36 years old male had sudden onset giddiness associated with blackouts which started 5-6 years back. He took symptomatic treatment for the

same. Recently, six weeks back he noted that he could not walk normally. There was swaying bilaterally but more on the left side. The relatives at home noted that his gait was altered. He came to our OPD clinic for swaying, impaired gait, dysarthria. On enquiry it was found that he had a long history of irritability, depression, sleep disturbances, mood fluctuations and intermittent giddiness since last one and a half to two years. He was a chronic alcoholic since ten years with poor appetite and irregular food habits. He was not a known hypertensive, diabetic or any past medical illness. His parents were both hypertensive with adequate control on anti hypertensives. He had had a road traffic accident

six months back for which his CT brain was done and was reported to be normal. His routine investigations at that time were found to be normal.

On examination, he was conscious, with depressive look, except for mild pallor his general examination was insignificant. He had a pulse of 118 beats per minute, regular and blood pressure of 210/120 mm of Hg in right upper arm supine position. He had loud A2, rest everything was normal. He had mild dysarthria with slightly impaired coordination bilaterally. Romberg's test was negative. Rest of the CNS examination was normal. His per abdomen examination revealed soft to firm non tender mild hepatomegaly. Rest of the abdominal examination was non revealing. Lungs were clear.

He was admitted and investigated for blood pressure and his neuropsychiatric symptoms. His CXR, KFT, LFT, serum proteins, Vit B12, folate, thiamine levels were normal. His blood glucose levels were also found to be normal. 2D ECHO was reported as hypertensive heart disease. However his MRI of the Brain revealed diffuse white matter hypodensities in frontal, parietal lobe, cerebellum including sub cortical involvement. (Figure – 1, 2)

Figure - 1: Sagittal T2-weighted image showed hyper intense signal in the central layer of corpus callosum with the sparing of the dorsal and ventral layers producing the 'sandwich sign'.

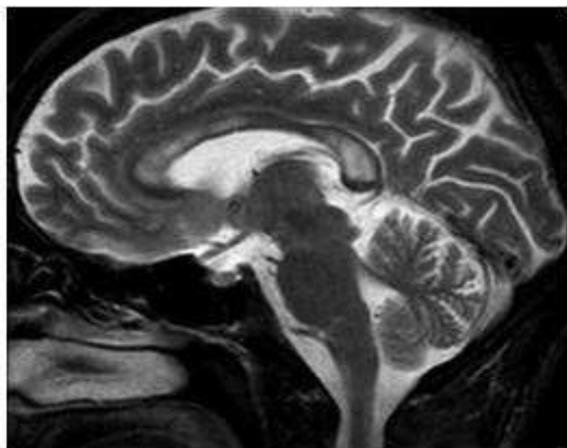
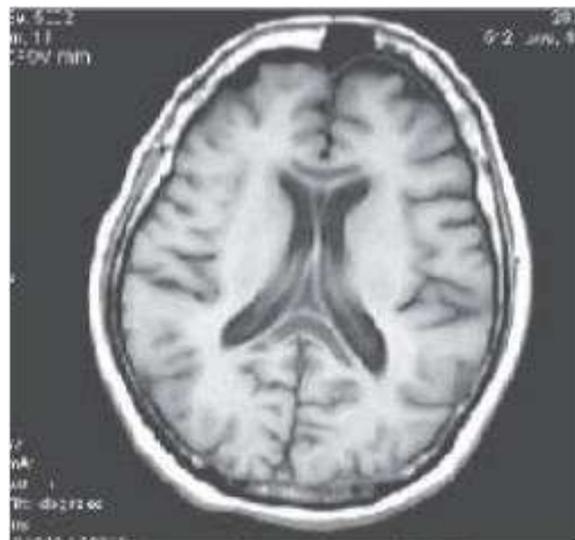


Figure - 2: Axial T1 weighted M R image shows hypointense signal in the corpus callosum.



Now he was diagnosed as a case of accelerated systemic hypertension with alcoholic leukoencephalopathy (mild Marchiafava Bignami syndrome). He was given anti-hypertensives and his blood pressure was adequately controlled. He was also given vitamin supplements, micronutrients and mild sedatives, antidepressants and tranquilizers. His neuropsychiatric symptoms abated and he improved with the given treatment and alcohol abstinence. He was discharged with an advice to take antihypertensives, vitamins and other medicines. The patient is under follow up. These medicines are Telmisartan, amlodipine, metoprolol, venlafaxine, stalopam and clonazepam in minimal doses.

Discussion

Alcohol has often mild and at times severe and devastating general and systemic manifestations [1]. Marchiafava-Bignami disease (MBD) is a rare neurological disease related to chronic and heavy alcohol consumption and malnutrition, and is characterized by primary demyelination and necrosis of the central part of the corpus callosum [2]. MBD is most frequently seen in middle-aged or elderly chronic alcoholic males [3].

MBD was first reported in 1903 by Marchiafava and Bignami, who originally described the symptoms in Italian men with increased consumption of inexpensively manufactured Chianti red wine [4]. Currently, however, MBD is known to occur in patients with chronic consumption of other sorts of alcohol including whisky and French liqueur. MBD has also been found in severely malnourished people without a history of alcoholism [5].

The underlying mechanism of the disease is still not understood [6]. It is probably caused by the combination of alcohol abuse and malnutrition, leading to metabolic, toxic, and vascular disturbances [7]. The main pathologic changes seen in MBD include symmetrical demyelination and necrosis of the central part of the corpus callosum, with relative sparing of dorsal and ventral layers. Rarely, 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 [8].

Marchiafava-Bignami disease may present in various clinical forms [9]. Acute Marchiafava - Bignami disease includes seizures, impairment of consciousness, and rapid death. Subacute Marchiafava - Bignami disease includes variable degrees of mental confusion, dysarthria, behavioural abnormalities, memory deficits, signs of interhemispheric disconnection, and impairment of gait. Chronic Marchiafava - Bignami disease, which is less common, is characterized by mild dementia that is progressive over years [10].

CNS manifestations chiefly associated with chronic alcoholism are alcoholic neuropathy, brief amnesic episodes, blackout intervals, Rostral vermis syndrome, generalized cognitive decline, Korsakoff's psychosis and Wernicke's encephalopathy apart from cerebrovascular diseases [11]. Marchiafava Bignami Syndrome is an illness which may have mild to very severe manifestations [12]. It is associated with alcohol intake for years together and may have

unnoticeable manifestations like irritability, mood fluctuations, sleep disturbances, mild cognitive decline, giddiness, swaying, mild visual disturbances [13].

It may also present as a severe neuropsychiatric illness, altered sensorium, to an illness relentlessly progressive to a comatose state and death [14]. There is history of chronic alcoholism and MRI Brain is characteristic showing white matter densities in the cortex, periventricular areas, cerebellum, corpus callosum and subcortex [15]. The burden of white matter hypodensities may not correlate with the clinical history, symptoms and neurological deficits [16].

Diagnosis is made on the basis of clinical findings in combination with radiological imaging features. MRI is currently the most sensitive diagnostic tool [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 [18]. The clinical course of the illness may range from unnoticeable symptoms to downhill fatal illness [19].

No standardized treatment protocols have been established in MBD [20]. Because the aetiology of the disease is uncertain, a specific therapy is not available. Cessation of alcohol intake is mandatory [21]. However, most often patients are treated with thiamine, vitamin B-complex and folate, with good clinical recovery in many patients [22].

Treatment consists of alcohol abstinence and proper care of nutrition. Very few cases of Marchiafava Bignami Syndrome are reported in literature and these cases are of old and aged alcoholics. Our case stands unique as far as age is concerned that is 36 years old.

Conclusion

This condition is relatively unknown among physicians, neurologists and psychiatrists. Most of the cases reported in literature are those with fatal outcome. We have reported this case with an intention to highlight the existence of such a syndrome related to alcohol abuse and alcoholism and that timely diagnosis and intervention may go along with in avoid in a mortality. Proper timely diagnosis of Marchiafava-Bignami syndrome may bring down the high reported mortality rate.

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