DOI: http://doi.org/10.4038/jmj.v33i2.141

Case Report

Marchiafava- Bignami disease (MBD): A rare case report

Sharmika. S[1], Nalayini. J[1], Peranantharajah. T[1], Ajantha. K[1]

[1]Teaching hospital, Jaffna

Abstract

Marchiafava-Bignami disease (MBD) is a rare neuro degenerative disease characterized by demyelination of corpus callosum. Clinical diagnosis of MBD is challenging due to its nonspecific neurological manifestations. It's promptly diagnosed by brain Magnetic Resonance Imaging (MRI). Prompt treatment with high dose thiamine could be lifesaving. Here we report a 68 year old male with chronic alcoholism who was diagnosed to have MBD and successfully treated with high doses of intravenous thiamine administration.

Key words

Marchiafava-Bignami disease (MBD), Corpus Callosum, Chronic alcoholism, Thiamine

Introduction

Marchiafava-Bignami disease (MBD) is a rare neuro degenerative disorder characterized by demyelination of corpus callosum. [1] It is most commonly seen in ill-fed chronic alcoholics, but also observed in poorly nourished nondrinkers. [2] The clinical manifestations are nonspecific with a wide variation in neurological deterioration. It is diagnosed by degeneration of corpus callosum in brain Magnetic Resonance Imaging (MRI). The prognosis of the disease is variable with complete recovery, partial recovery or death. Administration of high doses of thiamine and vitamin B complexes result in remarkable clinical recovery and resolution of imaging abnormalities. [3]

Case report

A 68 year old male who presented to the Accident and Emergency unit with gradual worsening of altered mental status over two weeks. He did not have fever, headache or symptoms of meningitis. There was no history of head trauma or injury. There was no significant past medical illnesses; however he is a chronic alcohol abuser. He consumes approximately 40 units of alcohol weekly over 45 years. Examination revealed thin body built male with Glasgow Coma Scale of 10/15. (Eye opening 3, Verbal 3, Motor 4). His neurological examination was normal including his ophthalmic fundoscopy. There were no signs of dehydration or meningeal irritation on examination. He was haemodynamically stable with no other system abnormalities.

Investigations revealed normal including hematological and biochemical except Hyponatremia of 118mmol/L with normal other serum electrolytes. Low Sodium levels were attributed to SIADH (Syndrome of Inappropriate Anti Diuretic Hormone secretion) due to alcoholism with low serum osmolality, high urine osmolality and increased urinary Sodium excretion with normal TSH and cortisol levels. His initial imaging

Corresponding Author: Sharmika S. Email: sharmiga.a@gmail.com, https://orcid.org/0000-0001-7142-1628

Submitted June 2021 Accepted December 2021

of NCCT Brain revealed only atrophy. Despite adequate correction of Sodium, he did not show improvement but deteriorated further. He was subjected to further imaging with MRI brain that revealed hyper intensity signals in T2W1 and significant atrophy of corpus callosum.

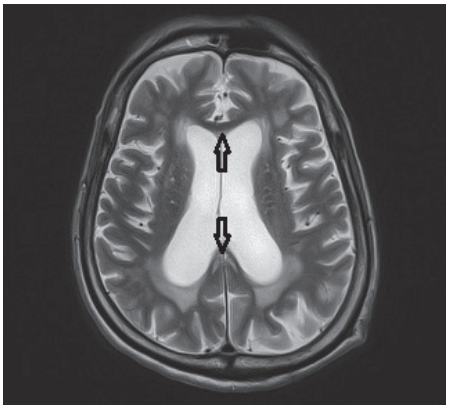

Figure1 Axial T2 weighted image shows significant atrophy of genu (upward arrow) and hyper intensity of splenium (downward arrow) of corpus callosum.

His thiamine dose was increased from 300 to 1500mg daily and continued for a week in the ward. Patient clinical condition markedly improved with that and he was able to walk and talk normally.

Discussion

MBD is a rare disease typically affects corpus callosum which can range from demyelination to frank necrosis. It is usually associated with chronic alcoholism leading to nutritional deficiencies which is also observed in malnourished nondrinkers too. The disease can be acute, sub-acute or chronic. The clinical manifestations are nonspecific with a wide variation including confusion, cognitive deficits, dementia, ataxia, apraxia, walking inability, dysarthria, seizures, coma or even death. [4], [6] The prognosis of the disease is variable with complete recovery, partial recovery or death.

Different conditions in alcoholics such as Wernicke's encephalopathy, delirium tremens and osmotic demyelination may have similar presentation. So brain MRI plays crucial role in prompt diagnosis and early intervention. The typical pathognomonic features on MRI are symmetrical lesions on corpus callosum. The central layers of corpus callosum are affected with sparing of dorsal and ventral layers producing sandwich sign. The disease typically affects body of the callosum followed by genu and splenium. Lesions also found on cerebral peduncle, middle cerebellar peduncle, hemispheric white matter and internal capsule. During acute phase, affected regions become oedematous with or without demyelination and appear as low signal density on T1 weighted images, high signal intensity on T2 weighted images and FLAIR (Fluid attenuated inversion recovery). As the acute phase passes, the oedema resolves. However, there will be permanent demyelination and necrosis, therefore MRI will show thinning and atrophy of corpus callosum and cystic transformation in untreated patients. If diagnosed and treated early, MRI demonstrate complete resolution of lesions of corpus callosum. [5]

Most of the case reports of MBD show favourable response to administration of high dose of parenteral thiamine, folate and vitamin B complexes.

Conclusion

MBD is often misdiagnosed and mistreated due to its nonspecific neurological manifestations. But it is a completely treatable condition with early initiation of high dose of parenteral thiamine and vitamin B complex. So treatment should be started in suspected patients before the radiological diagnosis as this is not harmful.

References

1. Marchiafava E, Bignami A. Sopra un alterazione del corpo calloso osservata in soggetti alcoolisti. Riv Patol Nerv 1903; 8: 544-9

2. Rusche-Skolarus LE, Lucey BP, Vo KD, Snider BJ. Transient encephalopathy in a postoperative nonalcoholic female with Marchiafava-Bignami disease. Clin Neurol Neurosurg 2007; 109(8): 713-5.

3. Nemlekar SS, Mehta RY, Dave KR, Shah ND. Marchiafava: Bignami Disease Treated with Parenteral Thiamine. Indian J Psychol Med. 2016;38(2):147-149. doi:10.4103/0253-7176.178810

4. S Raina, DM Mahesh, J Mahajan, SS Kaushal, D Gupta, DS Dhiman. Marchiafava Bignami Disease. Journal Assoc Physicians India 2008; 56: 633-5.

5. Kumar K, Sunil et al. "Marchiafava - bignami disease: a case report." Journal of clinical and diagnostic research : JCDR vol. 8,8 (2014): RD01-2. doi:10.7860/JCDR/2014/8377.4668

6. Hillbom M, Saloheimo P, Fujioka S, Wszolek ZK, Juvela S, Leone MA. Diagnosis and management of Marchiafava-Bignami disease: a review of CT/MRI confirmed cases. J Neurol Neurosurg Psychiatry. 2014;85(2):168-173. doi:10.1136/jnnp-2013-305979