Available online at www.sciencedirect.com

ScienceDirect

journal homepage: www.elsevier.com/locate/mjafi

Case Report

Overreligious beliefs with neuropsychiatric manifestations in a young lady—Rule out Marchiafava–Bignami disease

Vijoy Kumar Jha ^{a,*}, Dev Prakash Sharma ^b, Vivek Sharda ^c,
Abhilasha Kumari ^d

^a Classified Specialist (Medicine) and Nephrologist, INHS Kalyani, Vishakapatnam, India

^b Classified Specialist (Radiology), INHS Kalyani, Vishakapatnam, India

^c Classified Specialist (Medicine) and Neurologist, INHS Kalyani, Vishakapatnam, India

^d Medical Officer, INS Circars, Vizag, India

ARTICLE INFO

Article history:

Received 25 September 2018

Accepted 25 February 2019

Available online 15 April 2019

Keywords:

Marchiafava–Bignami disease

Neuroimaging

Demyelination

Thiamine

Psychiatric counseling

ABSTRACT

Marchiafava–Bignami disease is a rare condition characterized by demyelination of corpus callosum due to alcohol or malnutrition. Here we report a young lady who, due to her religious beliefs had stopped taking food and presented with neuropsychiatric manifestations. Neuroimaging was suggestive of Marchiafava Bignami disease and with adequate nutritional and thiamine therapy she had dramatic improvement clinically and had near complete resolution of lesions in neuroimaging.

© 2019, Armed Forces Medical Services (AFMS). All rights reserved.

Introduction

Marchiafava–Bignami disease (MBD) is a rare condition characterized by demyelination of the corpus callosum. The etiology of this disease is either toxic or nutritional as it is seen predominantly in alcoholics with poor nutrition, suggesting synergism between alcohol-induced neurotoxic effects and hypovitaminosis B.

We report a young female patient with overreligious beliefs who presented with neuropsychiatric manifestation and was

detected to have demyelination of the corpus callosum on neuroimaging. With nutritional supplementation and parenteral thiamine over 1 month, she improved remarkably and had complete resolution of brain lesion.

Case report

A 36-year-old female patient was brought to our hospital with complaints of the generalized weakness of 3- to 4-month duration which got worsened over 2–3 days. She also

* Corresponding author.

E-mail addresses: vkjhamd@gmail.com, vkjhamd@rediffmail.com (V.K. Jha).

<https://doi.org/10.1016/j.mjafi.2019.02.011>

0377-1237/© 2019, Armed Forces Medical Services (AFMS). All rights reserved.

complained of blurred vision, diplopia, vertiginous sensation, and gait imbalance during walking and standing. She was not taking food for the last 3–4 days and was on a fluid diet due to some religious reasons. She claimed that she had been challenging herself in the past and also wanted to prove that one can survive without food and water if someone has a spiritual inclination. Further history revealed that she had lost about 20 kg of body weight over the last 6 months and has reduced from 56 kg to 36 kg at present due to frequent fasting and surviving on liquid diets on non-fasting days. She denies any significant past history of medical/surgical illness. She denies any history of alcohol abuse/any alternative medicines consumption. She had two children through normal delivery, the youngest being 8 years old.

Clinically on admission, she was sick. Her pulse rate was 102/min, regular, BP was 130/84 mm Hg, SPO₂ was 98% without oxygen, weight was 36 kg, no pallor, icterus, edema, and

lymphadenopathy. Random blood sugar was 112 mg/dl. Central nervous system examination revealed bilateral horizontal gaze palsy with upbeat nystagmus on the vertical gaze. Romberg's sign was positive and plantars were normal. Other systemic examination was normal.

Investigations revealed the following: erythrocyte sedimentation rate (ESR) was 14 mm fall; Hb was 10.2 gm/dl; mean corpuscular volume (MCV) was 88 fl; total leucocyte count (TLC) was 4500/mm³; platelet count was 80,000/mm³; HIV, HBsAg, and hepatitis C antibody (anti-HCV) tests were negative; kidney function test was normal; liver function test was normal; serum albumin level was 3.2 gm/dl; serum sodium/potassium ratio was 136/4.8 mEq/L; serum calcium level was 8 mg/dl; and HbA1C was 6.2%. Ultrasonography (USG) abdomen was normal, serum vit B-12 and folic acid levels were normal, non-contrast computerized tomography (NCCT) head/chest/abdomen was normal, and urine routine examination/

DWI

ADC

Fig. 1 – Magnetic resonance imaging brain shows white/bright splenium of the corpus callosum on diffusion weighted imaging. It is dark on apparent diffusion coefficient, suggesting diffusion weighted imaging restriction. ADC, apparent diffusion coefficient; DWI, diffusion weighted imaging.

DWI

ADC

Fig. 2 – Magnetic resonance imaging brain shows the right corona radiata hyperintensity lesions correspondingly dark/black on apparent diffusion coefficient, suggesting diffusion weighted imaging restriction. ADC, apparent diffusion coefficient; DWI, diffusion weighted imaging.

microscopic examination (RE/ME) was normal. The thyroid profile was normal, and electroencephalography (EEG) and cerebrospinal fluid (CSF) study was normal. In view of severe neurological signs, magnetic resonance imaging (MRI) of the brain was performed which revealed focal areas of true restriction of diffusion in the splenium of the corpus callosum and bilateral corona radiata. These lesions were not apparent on T1WI, T2WI, or FLAIR sequences. No magnetic susceptibility effects were noted on gradient-echo images [Figs. 1 and 2](#).

Course in hospital— She was treated with adequate nutritional support. Psychiatry counseling was provided, and she was administered anxiolytic drugs along with vitamin supplements, parenteral thiamine 500 mg 8 hourly, and other supportive measures. It was planned to administer methylprednisolone to her but deferred as she improved with nutritional support within 3 days. Repeated MRI brain after 1 month revealed a focal area of Diffusion Weighted Imaging (DWI) restriction involving splenium showing near complete

resolution. Mild T2WI hyperintensity along with few punctate foci of DWI restriction was seen in previously noted lesion. There was no focal T1WI hypointensity, and no magnetic susceptibility effects were noted on gradient-echo images [Figs. 3 and 4](#).

Discussion

Marchiafava-Bignami disease (MBD) is one of the rarest conditions characterized by demyelination of the corpus callosum and seen most often in alcoholic patients. Italian pathologists Marchiafava and Bignami, in 1903, described three alcoholic men who were found dead after coma, and postmortem examination revealed that the middle two-thirds of the corpus callosum were severely necrotic. This condition was also reported in non-alcoholics (7.2% of all the MBD cases).¹

DWI

ADC

Fig. 3 – Follow-up magnetic resonance imaging brain shows splenium of the corpus callosum with punctate areas-white/bright on diffusion weighted imaging. Correspondingly dark/black on apparent diffusion coefficient suggestive of mild diffusion weighted imaging restriction. ADC, apparent diffusion coefficient; DWI, diffusion weighted imaging.

DWI

ADC

Fig. 4 – Follow-up magnetic resonance imaging brain shows normal bilateral corona radiata, and there is decrease in the extent and degree of diffusion restriction. ADC, apparent diffusion coefficient; DWI, diffusion weighted imaging.

Based on a review of 50 radiologic cases, two clinical subtypes of MBD have been described—type A and type B. Type A has features of coma and stupor and noted to have a high prevalence of pyramidal tract symptoms and involvement of the entire corpus callosum. Type B is characterized by normal or mildly impaired mental status and radiologically partial or focal callosal lesions.² Our case had features suggestive of type 2 MBD. Few case reports of MBD have also been reported in patients with dramatic fluctuations in the serum glucose level in the setting of poorly controlled diabetes suggesting abrupt changes in serum osmolality leading to demyelination of the corpus callosum.^{3,4} Chronology of events and the range of clinical symptoms may vary. Psychiatric disturbances, dysarthria, apraxia, and signs of interhemispheric disconnection may also be noticed. The condition is distinguished by other callosal lesions that the lesions are usually symmetrical and located in the middle of the corpus callosum. MRI brain may show increased T2/FLAIR signal, involving the mamillary bodies, dorsomedial thalami, tectal plate, and periaqueductal area around the third ventricle. Serum electrolyte and glucose levels, a complete blood count, and toxicology screening should be done. The “sandwich sign” is pathognomonic for MBD in MRI and is seen as hypointensity in the central layers of the corpus callosum on T1-weighted images with sparing of the dorsal and ventral layers. In MBD, unlike a stroke, areas of restricted diffusion resolve completely without apparent permanent damage like in this case.⁵ Early administration of parenteral thiamine is associated with better outcomes, particularly if administered within 2 weeks of symptom onset.¹ In this case, the patient had been administered parenteral thiamine in the doses prescribed for Wernicke's encephalopathy, and improvement in her neuropsychiatric conditions was noticed within few days. Corticosteroids are also being used in MBD to reduce brain edema, suppress demyelination, reduce inflammation, and stabilize the blood–brain barrier. A case report has also described the treatment of MBD with amantadine along with thiamine, vitamin B-12, and folate.⁶ Individual who survives the disease requires rehabilitation and alcohol and nutritional counseling.

Conclusion

In a patient with deliberate poor oral intake due to overfasting related to religious sentiments or other reasons (to deliberately lose weight) presenting with neuropsychiatric manifestations, it is imperative to suspect MBD, which shows dramatic improvement with thiamine therapy. Sharp clinical acumen and appropriate urgent neuroimaging can help in early diagnosis and early initiation of treatment, thereby improving the prognosis even in cases with severe diffusion restriction of the complete corpus callosum.

Conflicts of interest

The authors have none to declare.

REFERENCES

1. 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;85:168–173.
2. Heinrich A, Runge U, Khaw AV. Clinicoradiologic subtypes of Marchiafava-Bignami disease. *J Neurol*. 2004;9:1050–1059.
3. Pérez Álvarez AI, Ramón Carbajo C, Morís de la Tassa G, Pascual Gómez J. Marchiafava-Bignami disease triggered by poorly controlled diabetes mellitus. *Neurologia*. 2016;31:498–500.
4. Kilinc O, Ozbek D, Ozkan E, Midi I. Neurological and psychiatric findings of Marchiafava-Bignami disease in a nonalcoholic diabetic patient with high blood glucose levels. *J Neuropsychiatry Clin Neurosci*. 2015;272:e149–e150.
5. Ihn YK, Hwang SS, Park YH. Acute Marchiafava-Bignami disease: diffusion-weighted MRI in cortical and callosal involvement. *Yonsei Med J*. 2007;48:321–324.
6. Staszewski J, Macek K, Stepień A. Reversible demyelination of corpus callosum in the course of Marchiafava-Bignami disease. *Neurol Neurochir Pol*. 2006;40:156–161.