Marchiafava-Bignami Disease in a Patient with Anorexia Nervosa

Hiroaki Nishioka ⓘ , Hiroki Kamei ⓘ

Department of General Internal Medicine, Kobe City Medical Center General Hospital, Hyogo, Japan

A 15-year-old girl was transferred to our emergency room with a three-day history of altered mental status and impaired speech function. She also had appetite loss for seven days and received a drip containing thiamine 20 mg at the previous clinic. She had been diagnosed with anorexia nervosa (AN) two years ago, repeated anorexia, and overeating. She had no history of drug abuse, previous head injuries, seizures, or other neurological problems. Her vital signs were stable. She had a body mass index of 15.9 kg/m^2 and a Glasgow coma scale score of E4V4M6. She was seen as restless and agitated. No neurological focal signs were observed. Laboratory examination revealed albumin of 2.3 g/dl, alanine transaminase of 44 IU/l, aspartate transferase of 59 IU/l, and vitamin B1 of 5.5 µg/dl. Head computed tomography did not reveal any abnormal findings; however, diffusion-weighted magnetic resonance imaging (MRI) of the brain detected a hyperintense signal change in the splenium of the corpus callosum (Figure 1a). She was diagnosed with Marchiafava-Bignami disease (MBD) and given vitamin B complex. Seven days later, her mental status returned to normal, and the abnormal MRI signals disappeared (Figure 1b).

MBD is characterized by progressive demyelination of the corpus callosum, which can result in necrosis.[1] It commonly affects chronic alcoholics; however, it is sometimes associated with severe malnutrition.[1,2] To our knowledge, only a few cases have been reported in patients with AN.[3] Patients with MBD present with various clinical signs and symptoms including an altered mental state, impaired speech, altered personality, seizures, confusion, impaired memory, disorientation, unconsciousness, and signs of interhemispheric disconnection.[1-5] The differential diagnoses of MBD include Wernicke encephalopathy, hypoglycemic encephalopathy, infarction, pontine and extrapontine osmotic demyelination, and delirium tremens. Although computed tomography cannot detect MBD in the early stages, MRI enables early detection of lesions suggestive of MBD and is useful for the definitive diagnosis as in our case.[1] MRI typically demonstrates symmetric involvement of the corpus callosum. The splenium is the most commonly affected site, followed by the genu. The impaired regions initially show hyperintense signals on T2-weighted, fluid-attenuated inversion recovery, or diffusion-weighted imaging. After the acute stage of MBD, abnormal signals return to normal.[1] However, if the disease progresses to permanent demyelination and necrosis, the affected region shows atrophy and cystic transformation.[1] Cortical lesions were also reported in a few cases, which showed poor prognosis.[5] A vitamin B1 deficiency is considered to cause this condition; however, serum vitamin B1 levels do not directly correlate with those in the tissues; therefore, vitamin B complex therapy should be initiated in suspected patients regardless of serum vitamin B1 levels.[2,4] Early diagnosis and treatment are important to prevent progression resulting in irreversible corpus callosum damage.[4] Although no specific treatment regimen is approved for MBD, the initiation of vitamin B complex supplementation within two weeks after symptom onset was reported to have improved outcomes.[1] Clinicians should consider MBD in the differential diagnosis of patients with AN who present with altered mental status, regardless of serum vitamin B1 levels.
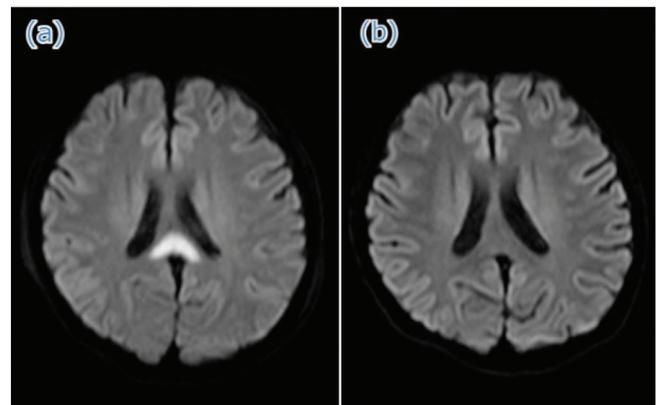

FIG. 1. Diffusion-weighted magnetic resonance imaging of the brain reveals a hyperintense signal in the splenium of the corpus callosum, without brain atrophy on admission (a), and the hyperintense lesion disappeared in seven days (b).

Informed Consent: Informed consent was obtained from the patient's family.

Authorship Contributions: Concept: H.N., H.K.; Design: H.N., H.K.; Data Collection or Processing: H.N., H.K.; Analysis or Interpretation: H.N., H.K.; Literature Search: H.N., H.K.; Writing: H.N., H.K.

Conflict of Interest: No conflict of interest was declared by the authors.

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. **[CrossRef]**

2. Fernandes LMP, Bezerra FR, Monteiro MC, et al. Thiamine deficiency, oxidative metabolic pathways and ethanol-induced neurotoxicity: how poor nutrition contributes to the alcoholic syndrome, as Marchiafava-Bignami disease. *Eur J Clin Nutr*. 2017;71:580-586. **[CrossRef]**

3. Tao H, Kitagawa N, Kako Y, et al. A case of anorexia nervosa with Marchiafava-Bignami Disease that responded to high-dose intravenous corticosteroid administration. *Psychiatry Res*. 2007;156:181-184. **[CrossRef]**

4. Parmanand HT. Marchiafava-Bignami disease in chronic alcoholic patient. *Radiol Case Rep*. 2016;11:234-237. **[CrossRef]**

5. Namekawa M, Nakamura Y, Nakano I. Cortical involvement in Marchiafava-Bignami disease can be a predictor of a poor prognosis: a case report and review of the literature. *Intern Med*. 2013;52:811-813. **[CrossRef]**