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A Case of Cortical Involvement in Marchiafava-Bignami Disease Accompanying Wernicke's Encephalopathy

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Dear Editor,

Marchiafava-Bignami disease (MBD) is a rare condition that is mainly associated with chronic alcoholism and malnourishment,[1] and characterized by symmetrical demyelination and necrosis of the corpus callosum. Clinically diagnosing MBD can be difficult due to the diversity of its symptoms, which range from cognitive impairment, reduced consciousness, and apathy, to seizures.[2,3] Its pathogenesis may involve a complex deficiency of group B vitamins, similar to Wernicke's encephalopathy (WE), which characteristically involves the mammillary bodies, periaqueductal regions, medial thalami, third ventricular walls, pons, medulla, basal ganglia, and cortical gray matter on magnetic resonance imaging (MRI).[4] Although MBD cases with cortical involvement have been reported, WE with cortical involvement accompanied by MRI findings of MBD has rarely been reported. We report a case of cortical involvement in MBD accompanying WE.

A 56-year-old male presented with altered mental status and a 20-year history of daily alcohol consumption. The initial neurological examination revealed drowsiness, impaired awareness, and impaired vertical and horizontal vestibulo-ocular reflexes, with no definite lateralizing sign, where the patient could lift both extremities against gravity with some effort. Ataxia and gait could not be evaluated, and nystagmus was not present. Blood tests revealed elevated creatinine (1.82 mg/dL), phosphorus (7.2 mg/dL), and ketone bodies (916 μmol/L). T2-weighted and diffusion-weighted MRI images demonstrated hyperintense lesions and multifocal diffusion-restricted lesions involving both superior frontal lobes, corpus callosum, medial thalami, periaqueductal gray matter, and dorsal medulla (Fig. 1).

The intravenous administration of thiamine (500 mg/day), high-dose vitamin B complex, and corticosteroid resulted in mild improvement in awareness, with the patient becoming alert and able to obey two-step commands. Left tactile anomia and apraxia were observed, but other disconnection syndromes were indeterminate due to poor cooperation. Electroencephalography showed diffuse slowing in all leads. Further evaluations including of the cerebrospinal fluid and MRI in sagittal views could not be performed because of noncooperation and withdrawal of approval from both the patient and his family. The patient was admitted for 12 days, and then transferred to a nursing hospital.

Both WE and MBD are strongly associated with chronic alcoholism and malnutrition, and often occur concurrently.[3] Although early thiamine treatment can reduce the neurological complications of MBD, the outcome is usually unfavorable.[5] Corticosteroids stabilize the blood–brain barrier by reducing the increased vasogenic permeability and decreasing the inflammatory edema.[2] Cortical involvement in MBD is secondary to callosal lesions and may be associated with a change in osmolality, whose mechanism is similar to that of central pontine and extrapontine myelinolysis (CPM and EPM, respectively).[3] The rare reports of WE with splenial involvement[6] have indicated that focal diffusion restriction is characteristic, which differs from MBD. Cortical involvement in WE has also been reported,[7] where

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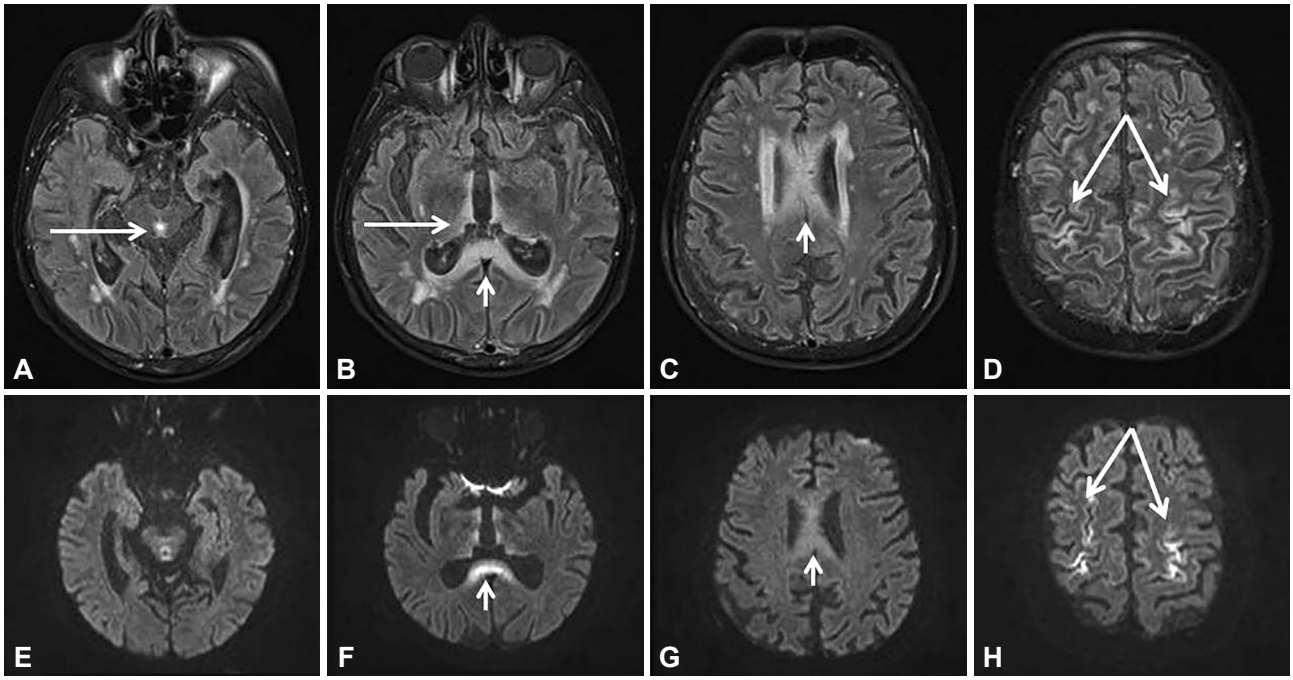

Fig. 1. Brain magnetic resonance imaging (MRI) images with fluid-attenuated inversion recovery (A–D) and diffusion-weighted MRI images (E–H) of our patient. High signal intensities in the periaqueductal gray and bilateral medial thalami are evident in panels A and B (long arrows). Widespread high signal intensities involving the corpus callosum are evident in panels B, C, F, and G (short arrows). Linear high signal intensities are evident in the bilateral frontal cortices in panels D and H (double head arrows).

tetraplegia and spasticity were observed, and cortical laminar necrosis with hemorrhage was revealed on susceptibility-weighted MRI images; however, these findings were not observed in our patient. The cortical lesions in our patient may have been caused by a mechanism involving types of myelinolysis similar to CPM and EPM. Although cases of MBD accompanying WE have been reported, to the best of our knowledge this is the first reported case of cortical involvement in MBD accompanying WE.

Author Contributions

Conceptualization: Michelle Youn, Jung-Ju Lee. Data curation: Jung-Ju Lee, Byung-Kun Kim, Ohyun Kwon, Woong-Woo Lee. Formal analysis: Jung-Ju Lee, Jong-Moo Park, Kyusik Kang. Methodology: Jung-Ju Lee. Validation: Jong-Moo Park, Kyusik Kang. Writing—original draft: Michelle Youn, Jung-Ju Lee. Writing—review & editing: Michelle Youn, Jung-Ju Lee.

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Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

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