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Marchiafava-Bignami disease with hyperintensity on late diffusion-weighted imaging

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Abstract

A 69-year-old man with a decades-long history of chronic alcohol consumption was admitted with gait disturbance (short steps and spasticity), deterioration of activity, and stuporous consciousness. Head magnetic resonance imaging (MRI) revealed hyperintensity on fluid-attenuated inversion recovery imaging in the corpus callosum and frontal white matter. The lesion later became more apparent on diffusion-weighted imaging. The clinical diagnosis was Marchiafava-Bignami disease (MBD). As temporary treatment, refraining from alcohol consumption and administration of vitamins were prescribed. The condition of the patient gradually improved. The purposes of this study were to demonstrate the clinical and radiological variety of MBD and to identify practical methods of treatment of this pathology.

Keyword

Marchiafava-Bignami disease (MBD); late diffusion-weighted imaging; white-matter lesions

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Introduction

Marchiafava-Bignami disease (MBD) is a rare neurological complication of chronic alcoholism, pathologically characterized by demyelination and necrosis in the corpus callosum. With wide usage of diagnostic criteria and progress in imaging techniques, early diagnosis is now possible. Many reports containing findings from magnetic resonance imaging (MRI), including diffusion-weighted imaging (DWI), have been published since 2000 (1), and most of these have reported hyperintensity on the first DWI during hospitalization (2–4). We report herein a rare case in which hyperintensity in the corpus callosum was noted on follow-up DWI during treatment.

Case report

A 69-year-old man had a decades-long history of chronic alcohol consumption, consuming approximately 500–550 mL of Japanese sake daily (standard alcohol content, 15% v/v; about 60–66 g ethanol/day). Up to about 1 year before admission, although he often experienced falls, he was independent in activities of daily living. Four days prior to admission, he had difficulty walking and stopped drinking. Three days prior to admission, the level of activity deteriorated, and he ceased speaking at home. On the day preceding admission, he was unable to walk or stand.

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On admission, the patient showed normal vital signs. In terms of the level of consciousness, he showed stupor, confabulation, delusive talking, agraphia, and ideomotor apraxia. Hyperreflexia of deep tendons, bilateral positive Babinski reflex, and muscle hypertonia (spasticity) were observed on neurological examination with brachybasia and instability by walking. He scored 15/30 on the Mini Mental State Examination (MMSE). Laboratory tests, including blood count, biochemical tests, blood sugar, ammonia, thyroid function tests, and levels of vitamins B1, B2, and B12, folic acid, and soluble interleukin-2 receptor, were all within normal limits. Cerebrospinal fluid examination also showed normal results.

Head MRI performed on admission revealed hyperintensity on fluid-attenuated inversion recovery (FLAIR) imaging in the corpus callosum and frontal white matter (Fig. 1a), but DWI did not reveal any abnormal signals (Fig. 1b). Based on the characteristic MRI findings and clinical history, MBD was diagnosed and treatment was started.

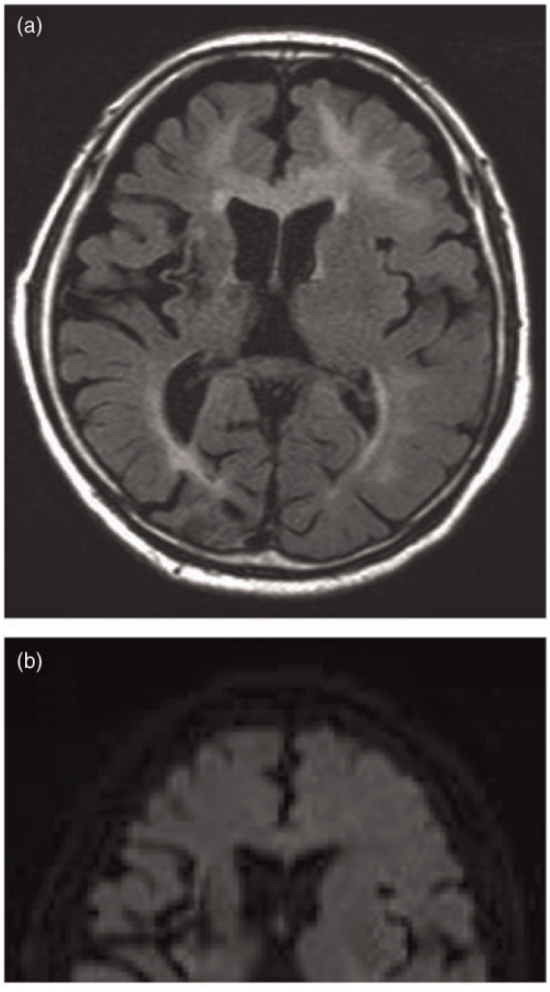

Fig. 1. Images taken at the primary admission. (a) Fluid-attenuated inversion recovery (FLAIR) imaging of the corpus callosum and frontal white matter. Hyperintensity is prominent and wide. (b) Diffusion-weighted imaging (DWI). No hyperintensity is apparent in the corpus callosum.

The patient was able to eat and drink, so he consumed regular meals, stopped drinking alcohol, and began medical rehabilitation. He was given thiamine 75 mg/day beginning the day after admission and nicotinamide 1.5 g/day beginning 5 days after admission.

Follow-up head MRI was performed 12 days after admission. The hyperintensity in the total area of corpus callosum remained the same on FLAIR imaging (Fig. 2b), but the white-matter lesion in the left frontal area was decreased in size (Fig. 2a). The callosal lesion showed hyperintensity on DWI (Fig. 2c). Part of this lesion appeared to have a reduced apparent diffusion coefficient (ADC) (Fig. 2d).

Another follow-up MRI was performed 39 days after admission, showing cavitary changes in the corpus callosum (Fig. 3a and b). After treatment, motor functions gradually recovered, but mild agraphia and apraxia remained. He was independent for activities of daily living and scored 29/30 on MMSE 67 days after admission.

Discussion

This patient met the criteria for subacute MBD according to the classification of Brion (5). We believe that the hyperintensity on FLAIR imaging on admission reflected damage to myelin and vasogenic edema of the corpus callosum and extracallosal projections (6).

Subsequent improvements in vasogenic edema and myelin damage probably accounted for the reduced hyperintensity and smaller size of the lesion in the corpus callosum on FLAIR imaging on day 12. The hyperintensity appearing on follow-up DWI may have reflected cytotoxic edema. Many previous reports of MBD have observed hyperintensity in the corpus callosum on early DWI (2–4). Several articles have indicated that abnormalities on MRI disappeared with improvements in symptoms (2,4). Hyperintensity on early DWI may be caused by reversible myelin vacuolation or intramyelinic edema, which has been described in patients with epilepsy and mild encephalitis (2).

On the other hand, we report here a rare case of MBD showing hyperintensity on delayed DWI. To the best of our knowledge, only one similar case exists in the literature (7). The authors suggested that the hyperintense lesions on initial FLAIR and late DWI represented vasogenic and cytotoxic edema, respectively. These characteristic findings indicate a pathological variety of MBD.

MBD has been regarded as a disease with poor prognosis. However, various cases have been reported with

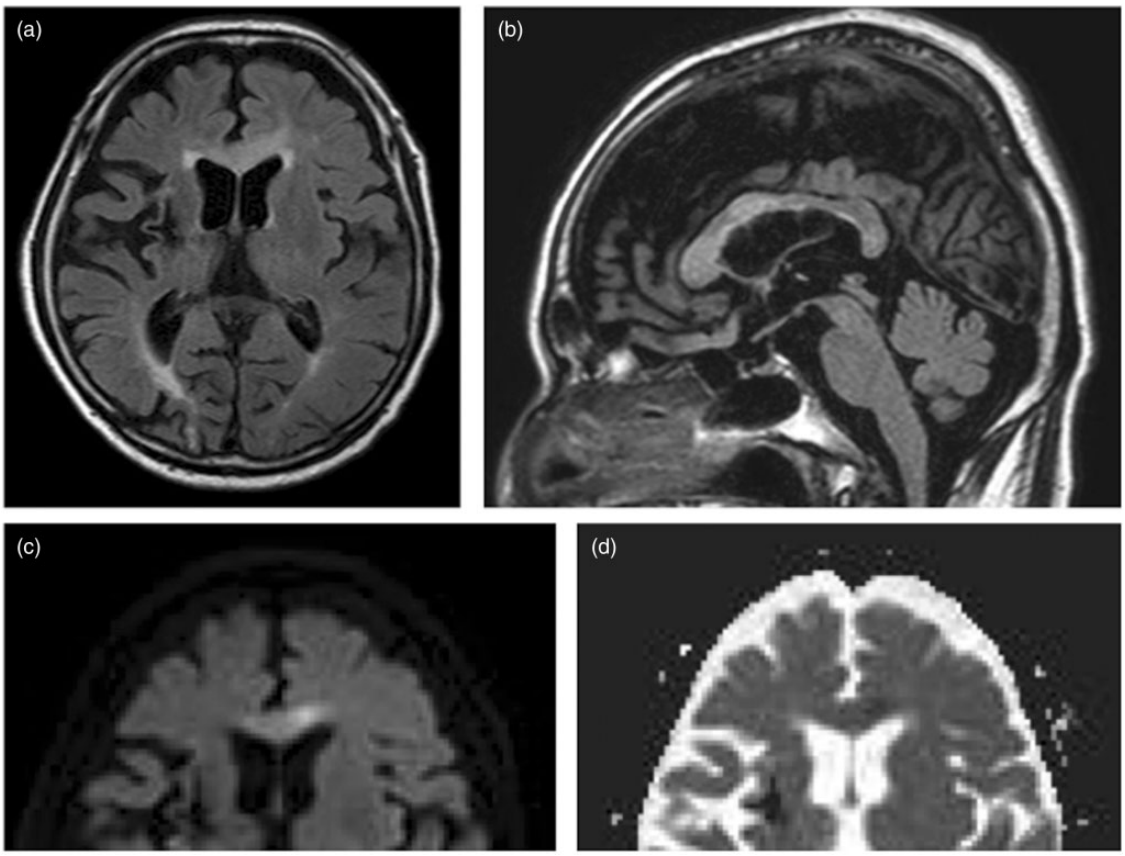

Fig. 2. Images taken on the 12th hospital day after admission. (a, b) FLAIR imaging of axial and sagittal section. Hyperintensity in the frontal white matter is reduced. Hyperintensity of the corpus callosum is still apparent. Cavity formation at this site is not apparent at this time. (c) DWI hyperintensity of the corpus callosum is apparent. (d) ADC mapping. Hypo- to isointensity in the corpus callosum is found.

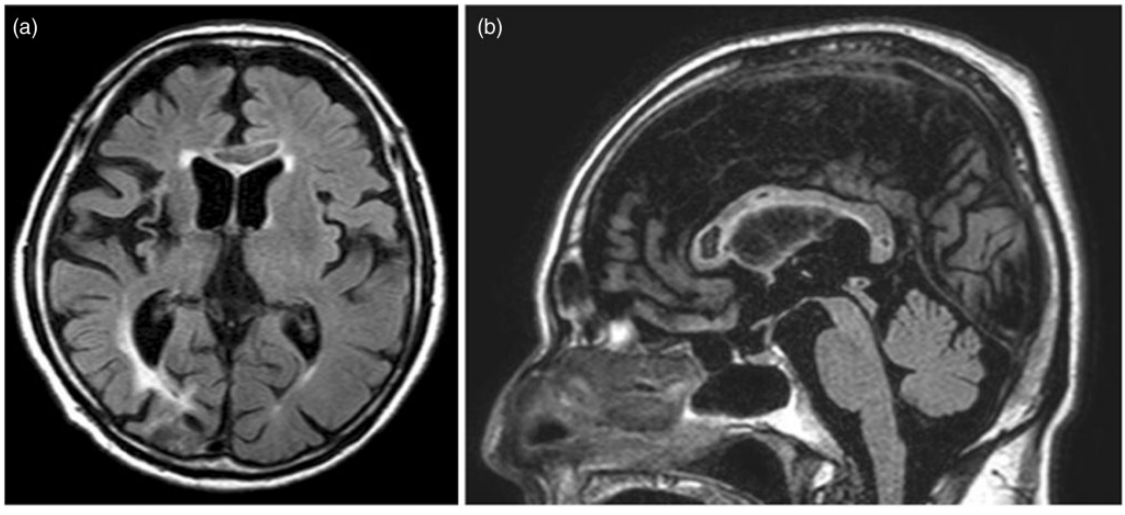

Fig. 3. Representative images from the final follow-up (on 39th hospital day) (a, b) FLAIR imaging of axial and sagittal section. Further reduction of the hyperintensity and cavitation of the corpus callosum are noted.

the technical progress in MRI. Some reports have discussed and presented prognostic indices for MBD, such as level of consciousness, degree of lesion in the corpus callosum (8) and cortical involvement (9). Quick and exact diagnosis and initiation of appropriate treatment for MBD as soon as possible is thus important.

In conclusion, we should be aware of the clinical and radiological variety of MBD. Radiological changes on MRI at each stage of the clinical course should be evaluated. Further studies are needed to confirm the underlying pathology of the hyperintense lesions in the corpus callosum.

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