

Marchiafava-Bignami disease with asymmetric extracallosal lesions

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Marchiafava-Bignami disease (MBD) is a rare disease in patients with chronic alcoholism and is characterized by symmetric demyelination and necrosis of the corpus callosum. Clinical features include neuropsychiatric disorders, dysarthria, tetraparesis, astasia-abasia, impaired consciousness and symptoms of interhemispheric disconnection. The lesions in MBD are not restricted to the corpus callosum, but also occur in extracallosal regions. However, no case of MBD with extracallosal lesions confined to one side of the brain has been reported. We present a patient with MBD showing asymmetric extracallosal lesions and complete resolution of magnetic resonance imaging (MRI) abnormalities.

A 38-year-old, right-handed man admitted to our emergency department due to general weakness and slurred speech. There was no fever, headache, dysphagia, vertigo, tinnitus, or hemisensory change. He had a history of chronic alcohol abuse for 20 years. His brother reported that he had consumed a daily average of 250 ml of spirit (Chinese liquor). Neurological evaluation revealed the patient in an altered mental state and tetraparesis. He received cranial computed tomography (CT) scan 13 h after onset in the emergency department of our hospital. Cranial CT was normal. Another brain CT performed 2 days after onset was also normal. Then the patient was transferred to the neurology intensive care unit (NICU).

Laboratory tests found that serum folate and vitamin B12 levels were lower. Syphilis and HIV serology was negative. Cardiac evaluation including electrocardiogram and heart ultrasound was normal. Magnetic resonance imaging on day 3 demonstrated hyperintensity at the corpus callosum (splenium), right frontal lobe and right hemispheric white matter on diffusion weighted imaging (DWI). Apparent diffusion coefficient (ADC) mapping showed relative hypointensity in the aforementioned lesions. T1 and T2-weighted MR imaging did not clearly show the lesions (Figures 1–6).

On the basis of clinical and imaging features, MBD was diagnosed. The patient received thiamine every day. On day 13, the patient's conscious state improved. Neurological evaluation revealed that he was apathetic, dysarthric, tetraparetic. On day 22, follow-up MR imaging demonstrated disappearance of signal-intensity abnormalities on DWI (Figures 7–10). The patient was still apathetic, dysarthric and tetraparetic on day 39, and he was transferred to the community hospital for rehabilitation treatment.

Marchiafava-Bignami disease is a rare disease mainly associated with alcoholism, which results in symmetrical demyelination and necrosis of

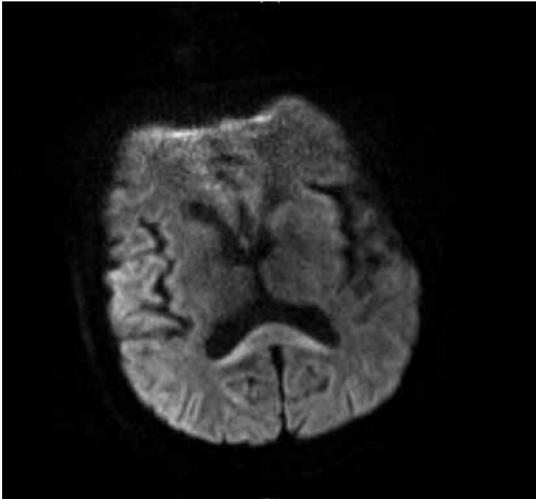


Figure 1. Hyperintensity in corpus callosum (DWI)

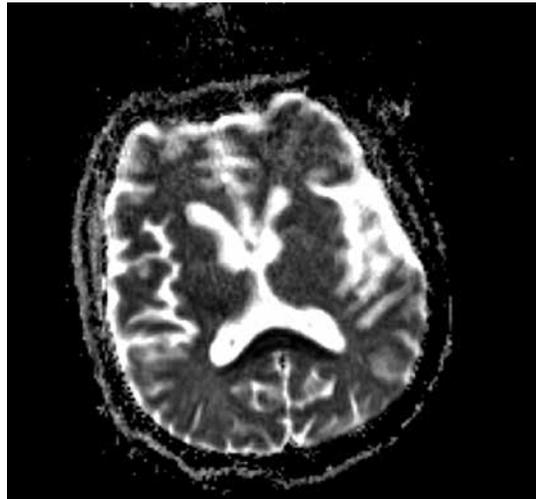


Figure 2. Hypointensity in corpus callosum (ADC)

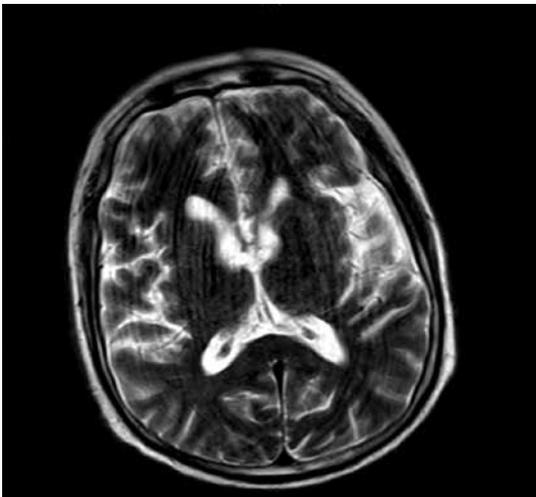


Figure 3. Isointensity in corpus callosum (T2)

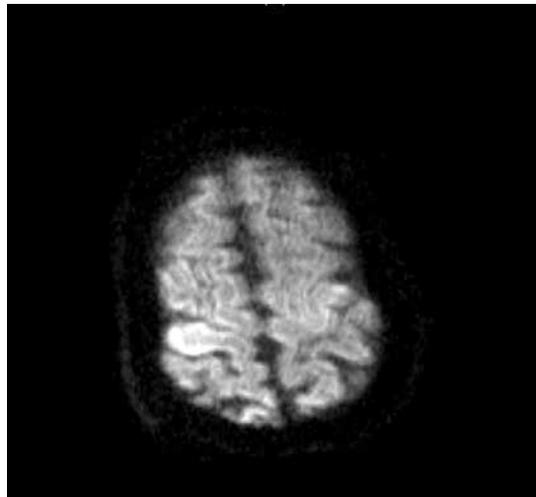


Figure 4. Hyperintensity in right frontal lobe (DWI)

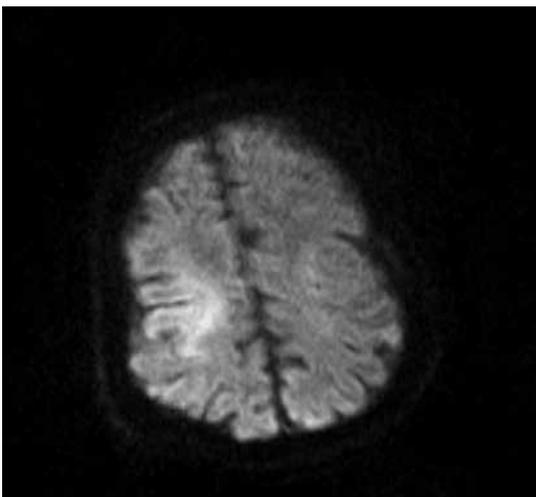


Figure 5. Hyperintensity in right hemisphere (DWI)

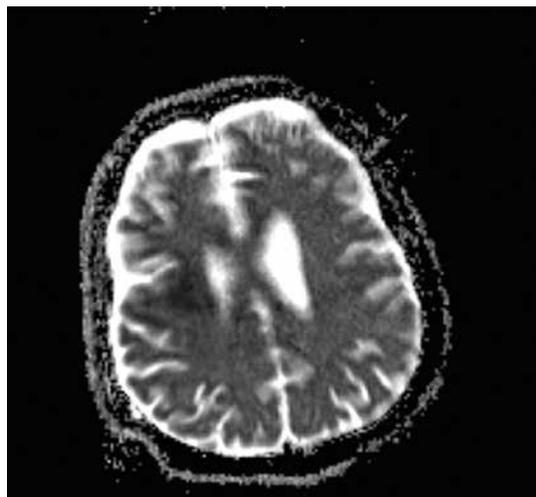


Figure 6. Hypointensity in right hemisphere (ADC)

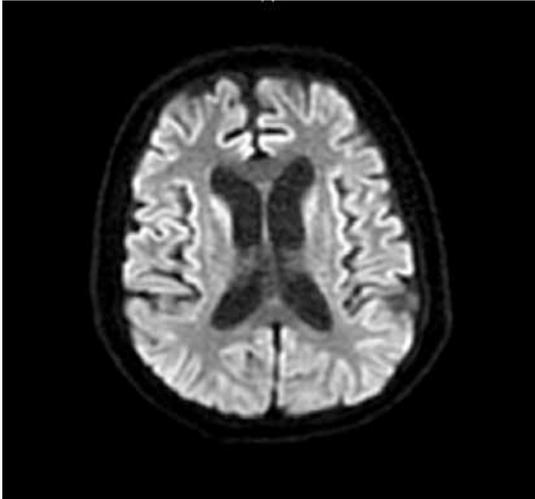


Figure 7. Isointensity in corpus callosum (DWI)

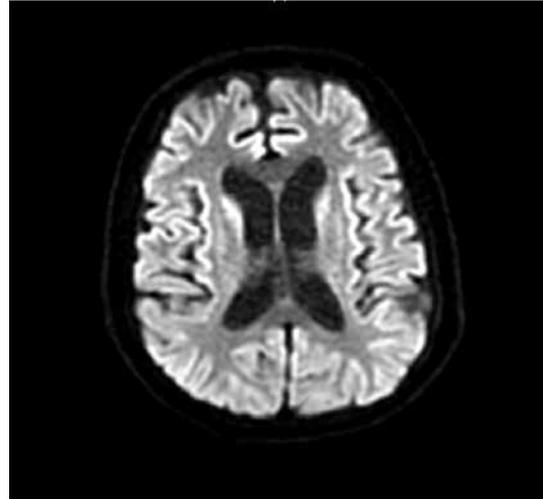


Figure 8. Isointensity in corpus callosum (DWI)

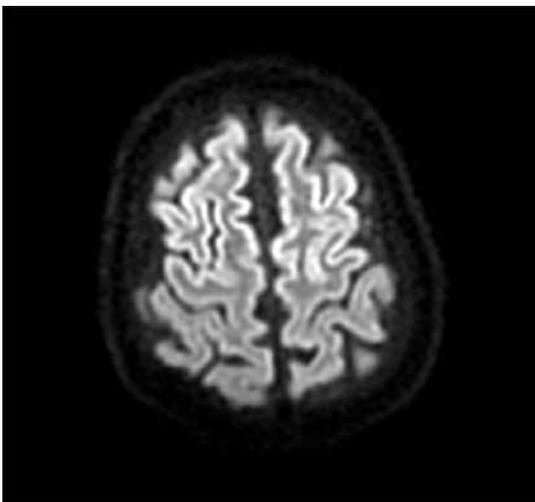


Figure 9. Isointensity in frontal lobe (DWI)

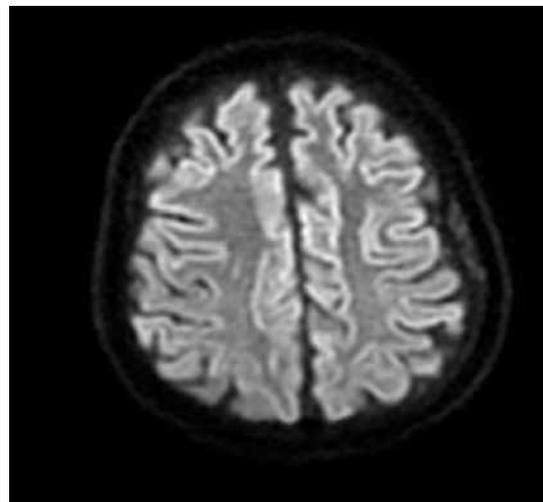


Figure 10. Isointensity in right hemispheric (DWI)

the corpus callosum. Because clinical signs are non-specific, MRI is essential to confirm the diagnosis. However, conventional MRI may miss the diagnosis of MBD. Sugeno *et al.* [1] reported a patient who presented with alcohol-withdrawal delirium. The DWI demonstrated a high intensity area in the corpus callosum. But T2-weighted MRI did not clearly show the lesion. In our case, we found hyperintensity at the corpus callosum (splenium), right frontal lobe and right hemispheric white matter on DWI. The ADC mapping showed relative hypointensity in the aforementioned lesions. T1 and T2-weighted MRI did not clearly show the lesions. Studies [2–6] using DWI have shown a low ADC value, which has been interpreted as demonstrating the presence of cytotoxic oedema in the corpus callosum. These results demonstrate that pathologic change in acute MBD is due in part to cytotoxic oedema of the cortex, and reflect the differing degrees of damage, from demyelination to

necrosis. In the early stage, cytotoxic oedema could only be detected by the DWI. This may explain why conventional MRI cannot reveal the lesions.

Interestingly, our patient showed asymmetric extracallosal lesions and complete resolution of the MRI abnormalities. To date, no case of MBD with extracallosal lesions confined to one side of brain has been reported. However, the reasons are still unclear. Seung *et al.* [7] reported a patient with MBD who received perfusion weighted MRI (PWI) and MR spectroscopy (MRS). The PWI measures of each lesion revealed that the cerebral blood flow and cerebral blood volume of the splenium decreased initially and subsequently increased 10 days later. Therefore, they speculated that ischemia may contribute to the cortex lesions. Hypoperfusion and ischemia of one side of the brain may be the cause of pathogenesis.

It is important for us to know how to treat the disease. Matti *et al.* [8] reviewed 122 reports con-

taining data on 153 subjects with confirmed MBD. They observed a significant trend for a better overall outcome in subjects who were treated with thiamine compared with those who remained untreated. Early thiamine treatment significantly reduced the risk of a poor outcome. Some papers [9–11] have reported improvements after steroid treatment. The administration of thiamine may have become a treatment option. Whether to give corticosteroid therapy is still controversial. It is also interesting that our patient received thiamine and obtained complete resolution of the MRI abnormalities. But the patient was still apathetic, dysarthric and tetraparetic. These indicators can decide the prognosis of patients with MBD.

The outcome of MBD may be variable. Menegon *et al.* and Heinrich *et al.* [12, 13] stated that involvement of the entire callosum and of the cortex was a poor prognostic factor. Some papers [3, 5, 14, 15] reported patients with MBD who had extracallosal lesions and a favourable course. In Seung *et al.*'s [7] research, MRS showed persistent neuronal dysfunction after resolution of the lesions on conventional MRI, and this finding correlated well with the clinical status. Sebastián *et al.* [16] reported two cases of MBD studied with MRI, including diffusion-tensor imaging (DTI) and fibre tractography. The FA values were initially similar to those of a healthy age-matched control. However, at 4-month follow-up, FA values were lower, and the decreases were more prominent in the areas with initial T2, FLAIR, and DWI signal alterations. The DTI findings suggest that corpus callosum white matter fibres are initially intact and are damaged later during the course of MBD. Multimodal MRI, such as MRS and DTI, may explain the inconsistent results. Although imaging lesions disappeared, there was still abnormal metabolism of brain tissue and fibre integrity was broken in the areas with abnormal signal alterations. These may explain the patient with lesions restored but still paralyzed. It was regretted that our patient did not receive MRS examination.

In conclusion, DWI and ADC are necessary in the diagnosis of MBD. The MRS and DTI may be superior in prognosis prediction of patients with MBD.

Conflict of interest

The authors declare no conflict of interest.

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