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Case Report

A Case of Marchiafava-Bignami Disease with Complete Recovery: Sequential Imaging Documenting Improvement of Callosal Lesions

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TOBITA, M., MOCHIZUKI, H., TAKAHASHI, S., ONODERA, H., ITOYAMA, Y. and IWASAKI, Y. *A Case of Marchiafava-Bignami Disease with Complete Recovery: Sequential Imaging Documenting Improvement of Callosal Lesions.* Tohoku J. Exp. Med., 1997, **182** (2), 175-179 —— Serial CT and MR imaging findings in a 44-year-old woman with Marchiafava-Bignami disease (MBD) are reported. In the acute stage, CT studies disclosed subtle hypodensity in the splenium, and T2-weighted MR images revealed apparent high signal intensity of the splenium and the central portion of the corpus callosum. Treatment with vitamin B complex resulted in complete recovery. T2-weighted MR images obtained three weeks after admission revealed dramatic resolution of imaging abnormalities, with only faint high signal intensity remaining in the splenium. The sequential changes observed on CT and MR images provided early diagnosis of MBD and the resolution of the lesion considered as brain edema, which suggested that edema might, in addition to demyelination or necrosis, be involved in the acute progression of MBD. ———— Marchiafava-Bignami disease; corpus callosum; alcoholism; MRI; CT

Marchiafava-Bignami disease (MBD), characterized by primary degeneration of the corpus callosum (Marchiafava and Bignami 1903), is a rare complication of chronic alcoholism. This disease may present in acute, subacute or chronic courses (Brion 1976). In the acute stage of MBD, the clinical features are so varied that most reported cases have been diagnosed at autopsy. Recently, CT or MR imaging has enabled early diagnosis, but since treatment has not been established, the prognosis is still poor.

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We present herein a patient with MBD who completely recovered from this disease. Serial observation by CT and MR imaging documented the improvement of the lesions.

Case Report

A 44-year-old Japanese woman experienced severe nausea and developed gait disturbance, dysarthria, and confusion. One week later, she experienced tonic-clonic convulsions and was transferred to our hospital. She had a long history of alcoholism, and her diet was poor and irregular. She consumed 1000 to 1500 ml of shochu (a Japanese spirit made from sweet potatoes, alcohol concentration of 25 to 35%) every week. During the month prior to her admission, her daily alcohol intake increased to 500 ml. On admission, she was in a confused and delirious state. Gaze-evoked horizontal nystagmus, cerebellar ataxia, and scanning and explosive dysarthria were noted. All four limbs were hypertonic and showed hyperreflexia without Babinski signs. CT on admission disclosed subtle hypodensity of the splenium. Conventional spin-echo T2-weighted MR images revealed abnormal high signal intensity in the splenium, the central zone of the corpus callosum, and the deep white matter (Fig. 1), while other portions of the brain, including the mammillary bodies, thalami and brainstem, were all intact. On laboratory examination, macrocytic anemia and mild liver dysfunction were noted. Her level of vitamin B_1 was 24 ng/ml (normal range: 18–53 ng/ml), and data for cerebrospinal fluid including cell count and levels of glucose, total protein, Cl, LDH, and myelin basic protein were normal.

Based on her clinical and MR imaging findings, she was diagnosed as MBD. Treatment with intravenous infusion of vitamin B complex (daily doses: B_1, 1225 mg; B_2, 1075 mg; B_6, 10.75 mg) was started within 24 hours after admission and continued for one week. It resulted in marked clinical improvement. On the

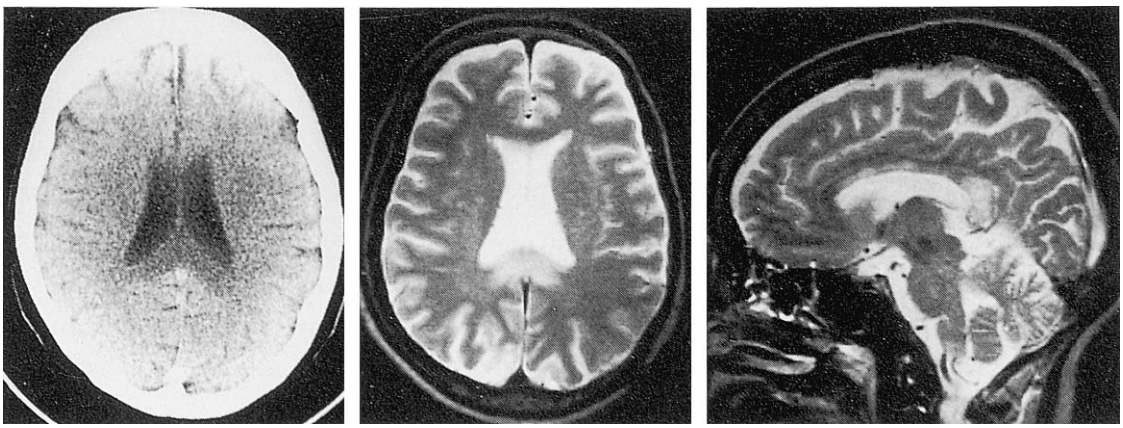

Fig. 1. CT and MR images on admission. CT disclosed abnormal hypodensity of the splenium. Axial and sagittal T2-weighted MR images (SE 2500/90/1) revealed abnormally high signal intensity in the splenium, in the central zone of the truncus of the corpus collosum and in the deep white matter. Swelling of the splenium was also noted.

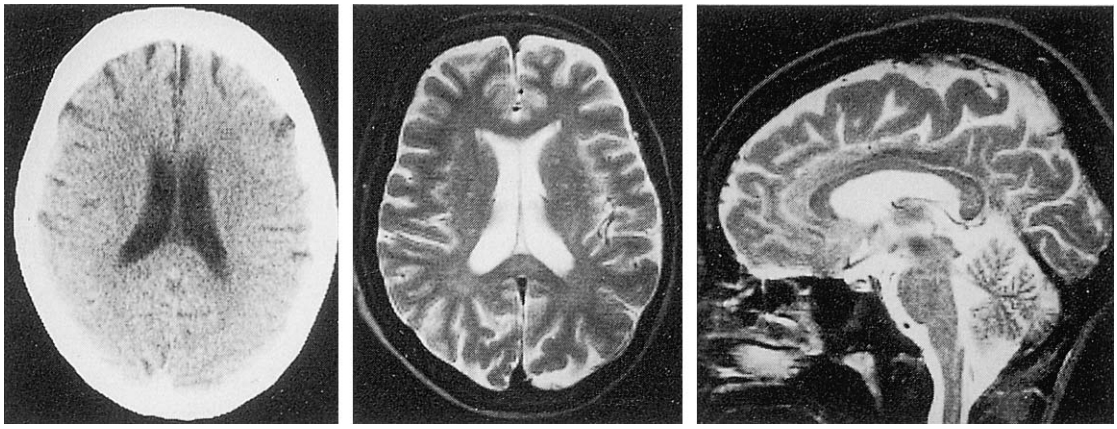

Fig. 2. CT and MR images obtained 3 weeks after admission. CT disclosed no abnormality. Axial and sagittal T2-weighted MR images (SE 2500/90/1) demonstrated regression of both callosal hyperintensity and swelling, with only subtle hyperintensity of the splenium remaining.

second day, marked improvement of consciousness was noted, and two weeks after admission, complete recovery was noted with normal neurological findings. She exhibited no signs of interhemispheric disconnection syndrome during her clinical course. T2-weighted MR images obtained three weeks after admission disclosed subsidence of the swelling and high signal intensity of the corpus callosum with residual subtle high signal intensity in the splenium. CT examination at the same time disclosed no abnormalities (Fig. 2).

Discussion

We have herein reported the case of a patient who completely recovered from MBD. The unique aspect of this case is that serial observation with CT and MR imaging showed resolution of the lesions. From her clinical signs, Wernicke's encephalopathy was also suspected. However, her vitamin B_1 level was not decreased, and she had none of the findings on imaging studies reported to be characteristic of Wernicke's encephalopathy (Donnal et al. 1990; Gallucci et al. 1990; Victor 1990). On the other hand, the lesions in the central portion of the corpus callosum are well known as "sandwich sign," which is characteristic of MBD. From these findings, MBD was diagnosed, although her chronic alcoholism might have partially modified her clinical symptoms. In MBD, neurological complications seen with chronic alcoholism were also apparent (Sato et al. 1981).

It is commonly assumed that Wernicke's encephalopathy is due to vitamin B_1 deficiency. On the other hand, although chronic alcoholism and malnutrition appear to be the most consistent extrinsic factors associated with MBD, its pathogenesis remains controversial. In a recent review, Victor (1994) speculated that some metabolic abnormalities induced by alcohol may be a causative factor. In addition, several investigators have suggested that brain edema plays an important role in the acute progression of MBD (Seitelberger and Berner 1955). In some neuropathological studies, edematous changes occurred in the callosal

lesions of the acute stage of MBD (Ishizaki et al. 1970; Kosaka et al. 1984). Bracard et al. (1986) and Chang et al. (1992) described CT and MR imaging findings obtained in the acute phase of MBD, and these findings suggested that edema of the corpus callosum played an important role in the acute progression of MBD. Kamaki et al. (1996) reported callosal bleeding thought to be due to edema in their case of MBD. The callosal lesion observed during the acute phase of disease in our case, which featured high signal intensity on T2-weighted MR images and hypodensity on CT scans accompanied by swelling, partially regressed within three weeks of initiation of treatment. This reversible change appears to represent edema, while the remaining or irreversible change, the subtle hyperintensity in the splenium, may represent demyelination or necrosis. As callosal hyperintensity decreased, clinical recovery was noted. Thus edematous change in the corpus callosum appears to have played an important role in the acute development of the patient's symptoms in our case. Since MBD is known to occur after long periods of alcoholism or malnutrition, progression and regression of edema in the corpus callosum may be related to the onset and prognosis of MBD.

No specific therapy has been established for MBD. However, treatment with thiamine or vitamin B complex has been reported to have resulted in clinical recovery in several cases (Leventhal et al. 1965; Baron et al. 1989; Humbert et al. 1992; Izquierdo et al. 1992), suggesting that impairment of the mechanism of vitamin B utilization may underlie the pathogenesis of MBD. In most of the reported cases in which treatment was precisely described, as well as our own, such therapy was begun within 1 week after the onset of MBD. These findings suggest that early diagnosis with CT or MR imaging followed by prompt administration of vitamin B complex may improve the prognosis of MBD. However, the mechanism of the favorable effect of vitamin B complex on MBD is obscure. Further study is necessary to clarify the pharmacological action of vitamin B complex.

Although initial CT examination also disclosed hypodensity in the splenium in our case, sagittal T2-weighted MR images clearly demonstrated not only the presence of a splenial lesion but also the existence of small lesions in the body of the corpus callosum. Furthermore, subsequently obtained MR images displayed subtly hyperintense lesions no longer detectable with CT. With MR imaging, therefore, MBD can more readily be diagnosed in the acute stage, enabling early initiation of treatment.

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