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Marchiafava-Bignami Disease: Clinical and MRI Findings in Two Patients

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= Abstract = **We herein report two alcoholic patients with highly suspicious Marchiafava-Bignami disease (MBD) who developed acute neurologic dysfunction and showed characteristic abnormalities in the corpus callosum on the brain MRIs: focal low signal lesion(s) in the genu or splenium of the corpus callosum, and diffuse callosal atrophy on the sagittal T1-weighted images.**

Key Words : *Marchiafava-Bignami disease, Brain MRI, Low signal lesion, Callosal atrophy, Corpus callosum.*

INTRODUCTION

Marchiafava-Bignami disease (MBD) is a rare disorder characterized by the degeneration of the corpus callosum. Before the era of magnetic resonance imaging (MRI), the diagnosis of MBD was usually made on autopsy because of the wide spectrum of clinical manifestation (dementia, frontal lobe releasing signs, inability to walk, hemiparesis, dysarthria, dysphagia, spasticity, and altered consciousness).

Nowadays we can diagnose MBD antemortem using MRI machinery. To our knowldge, there have been less than 10 cases of MRI-supported MBD reported worldwide. We herein present two cases with MBD supported by the characteristic lesions in the corpus callosum on the MRIs.

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Patient 1

A 52-year-old man was admitted to the hospital because of poor verbal output, dysphagia, free voiding and inability to walk of sudden onset.

Four years prior to admission, he had developed sudden dysarthria, left hemiparesis, and disorientation, which improved after several days on admission to the clinic of herbal medicine. Four months prior to admission, he developed emotional lability, dysarthria, and abnormal behavior of subacute onset over a week. Thereafter, the symptoms persisted without significant improvement. He was a chronic heavy alcoholic for about 25 years, although he lessened his alcohol consumption during recent four years.

Mental status examination revealed disorientation, poor memory, emotional lability, right-left disorientation, global aphasia, apraxia, and abu-

Fig. 1. (patient 1): Thin section of sagittal T1-weighted image shows callosal atrophy and a small focal low signal lesion in the genu of the corpus callosum

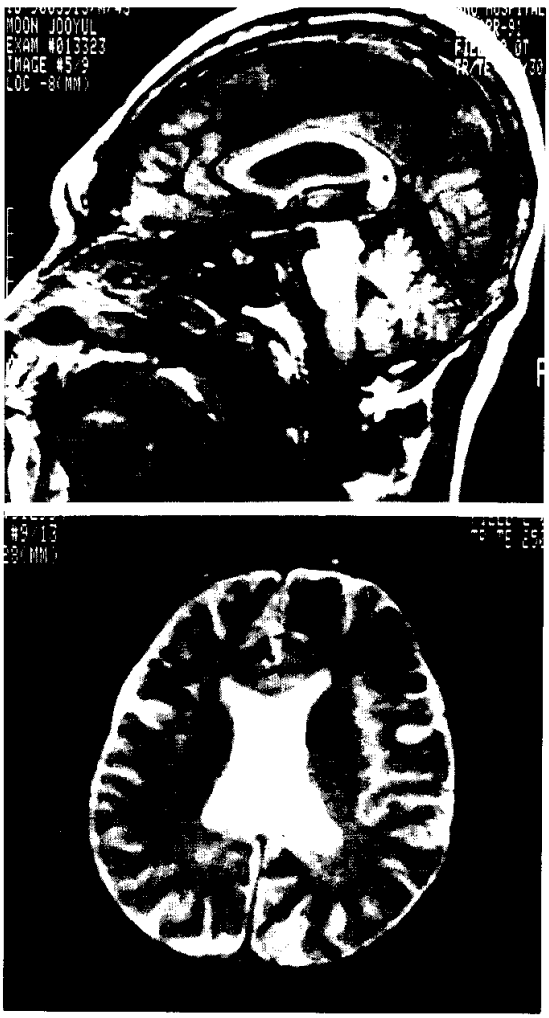

Fig. 2. A, B (patient 2): Sagittal T1-weighted image shows focal low signal lesions in the splenium of corpus callosum (Fig. 2-A). Axial T2-weighted image reveals high signal lesions in both anterior and posterior commissures (Fig. 2-B)

lia. The score of the minimental status examination was about 10/30. However there was no certain evidence of disconnection syndrome. There were marked frontal lobe releasing signs, severe dysarthria, bilateral clumsiness of hands, bilateral dysmetria, and increased bilateral deep tendon reflexes. The motor power, sensory examination, and the ocular motility were normal. During the admission he became able to walk and other neurological symptoms improved remarkably except for dementia. Studies on cerebrospinal fluid and visual and brainstem-auditory evoked potentials were normal. Removal of the cerebrospinal fluid failed to improve his neurological symptoms. Initial conventional sections of sagittal T_1-weighted images showed callosal atrophy, but failed to reveal any focal lesion in the corpus callosum. Two days later we performed a follow-up MRI which revealed a focal low signal lesion in the genu of the corpus callosum on the thin sections of sagittal T_1-weighted images (Fig. 1).

Patient 2

A 43-year-old heavy alcoholic suddenly became stuporous with a subsequent fluctuating level of consciousness. Neurologic examination revealed double homonymous hemianopsia, right hemiparesis, right facial palsy of central type and slight limitation of the extraocular movement with the gaze-evoked nystagmus. Mirror writing of the left hand suggested disconnection

syndrome. Studies on cerebrospinal fluid including IgG index were normal. Nerve conduction study revealed polyneuropathy involving the distal lower extremities. brain MRI revealed focal low signal lesions in the splenium of corpus callosum on T_1-weighted images (Fig. 2-A) and commissural high signal lesions on T_2-weighted images (Fig. 2-B).

After several days of multivitamins therapy, his consciousness and subsequently other neurological symptoms improved remarkably.

DISCUSSION

The principal pathological change of the Marchiafava-Bignami disease (MBD) is the necrosis and demyelination of the corpus callosum. This may involve the whole anteroposterior extent of the corpus callosum or only in part, and the central fibers are mainly affected with preservation usually of upper and lower rims. Macroscopically the lesion appears as a greyish softened band which histologically shows glial and vascular remnants. Other regions of white matter sometimes involved include the cerebellar peduncles and the centrum semiovale in the frontal and parietal lesions in the lateral extent of the corpus callosum. The anterior commissure and optic chiasm are less often affected (Duchen and Jacobs, 1992).

The symptom onset is usually rapid, occurring within a few days as in our two cases. It consists of the sudden appearance of neuropsychiatric disorders which include changes in mood with irritability, aggression, intellectual impairment, transient coma, convulsions, and ambulation disorders of astasia-abasia type. The established disease may present an acute, subacute, or chronic course (Brio, 1976). Differentiation from infarction of the corpus callosum may be difficult as in patient 1. However, selective involvement of the entire length of the corpus callosum and focal cystic necrosis confined to its central layer, with sparing of the ventral and dorsal layers, appear more likely to be due to MBD. In the patient 2, demyelinating disorders involving both the corpus callosum and the white matter, especially multiple sclerosis, could be differentiated on the basis of clinical, radiological, electrophysiological and other laboratory findings.

Both of the patients in this report revealed focal low signal lesion(s) in the the corpus callosum and diffuse callosal atrophy on the sagittal T1-weighted images. Hitherto, five articles (Chang *et al.*, 1992; Clavier et al., Bracard *et al.*, 1986; Mayer *et al.*, 1987; Kawamura *et al.*, 1985) have reported MRI findings in seven patients with MBD. Chang et al (1992) described follow-up MRIs of two patients: diffuse swelling of the corpus callosum in acute stage, and atrophy with focal low signal lesions in the corpus callosum in chronic stage. Other MRI reports of MBD (Clavier *et al.*, Bracard *et al.*, 1986; Mayer *et al.*, 1987; Kawamura *et al.*, 1985) are similar to our results. In the patient 1, conventional sagittal T1-weighted images did not reveal a focal low signal lesion in the corpus callosum. However, thin sagittal images with two milimeters slice did. Therefore we think that thin sagittal sections of the brain MRI are needed to find out small focal lesion(s) in the corpus callosum in patients with suspicious MBD.

REFERENCES

Bracard S, Claude D, Vespignani H, Almeras M, Carsin M, Lambert H, Picard L: Computerized tomography and MRI in Marchiafava-Bignami disease. J Neuroradiol 13:87-94, 1986

Brio S: Marchiafava-Bignami syndrome. In: Vinken PJ and Bryun GW, eds. Handbook of Clinical Neurology Elsevier/North-Holland biomedical press, Amsterdam 28:317-330, 1976

Chang KH, Cha SH, Park SH, et al.: Marchiafava-Bignami disease: Serial Changes in Corpus Callosum on MRI. Neuroradiology 34:480-482, 1992

Clavier F, Thiebot J, Delangre T, Hannequin D, Samson M, Benozio M: Marchiafava-Bignami disease. A case studied by CT and MR imaging. Neuroradiology 28:376, 1986

Duchen LW and Jacobs JM: Nutritional deficiences and metabolic disorders. In: Adams JH and Duchen LW, eds. Greenfield's Neuropathology, 5th ed. Edward Arnold, London 818-820, 1992

Kawamura M, Shiota J, Yogishita T, Hirayama K: Marchiafava-Bignami disease: Computed Tomographic Scan and Magnatic Resonance Imaging. Annals of Neurology 18(1):103-104, 1985

Mayer JM, De Liege P, Netter JM, Danz F, Reizine D: Computerized tomography and nuclear magnetic resonance imaging in Marchiafava-Bignami disease. J Neuroradiol 14:152-158, 1987