Magnetic Resonance Imaging Findings in a Patient with Acute Marchiafava-Bignami Disease: Case Report

Akut Marchiafava-Bignami Hastalığı Olan Bir Hastada Manyetik Rezonans Görüntüleme Bulguları

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ABSTRACT Marchiafava-Bignami disease or syndrome (MBD) is a rare disorder that all cases had a history of chronic alcohol abuse. It is thought to be due to vitamin B complex deficiency. All cases had a symmetrical lesions in the corpus callosum and causes progressive demyelination and necrosis. It can spread to white matter of the neighboring tissues and rarely to the subcortical areas. Clinical symptoms include not only dysarthria, ataxia, tetraparesis, disphagia, dementia as a clinical outcome, cognitive deficits but also disturbed consciousness, seizure and coma. Some of them show asymmetrical hypointense areas in multiple regions on susceptibility-weighted imaging. Here, we report a 25-year-old man had progressive ataxia, dysarthria, and disphagia. This is a case of MBD that was diagnosed by magnetic resonance imaging (MRI).

Key Words: Alcoholism; magnetic resonance imaging; Marchiafava-Bignami disease

ÖZET Marchiafava-Bignami Hastalığı (MBH) nadir bir bozukluk olup vakaların hepsinde kronik alkol kullanım öyküsü mevcuttur. Hastalığın B kompleks vitaminlerinin eksikliğine bağlı olduğu düşünülmektedir. Vakaların hepsinde korpus kallozumda simetrik lezyonlar mevcut olup ilerleyici demiyelinizasyona ve nekroza neden olmaktadır. Lezyon komşu dokulardaki beyaz cevhere nadiren de subkortikal alanlara yayılabilmektedir. Dizartri, ataksi, tetraparezi, disfaji ve demans gibi klinik bulguların yanında bilişsel defisitler, bilinçte bozulma, nöbet ve koma da görülebilir. Bazı vakalarda görüntüleme testlerinde birçok bölgede asimetrik hipointens alanlar mevcuttur. Bu vaka takdiminde ilerleyici ataksi, dizartri ve disfajisi olan 25 yaşındaki erkek hasta sunulmaktadır. Bu vakada MBH tanısı manyetik rezonans görüntüleme (MRG) yöntemiyle konmuştur.

Anahtar Kelimeler: Alkolizm; manyetik rezonans görüntüleme; Marchiafava-Bignami hastalığı

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Marchiafava-Bignami disease (MBD) is a rare disease defined by characteristic demyelination of the corpus callosum usually seen in chronic alcoholics in their middle or late ages.[1,2] Italian pathologists, Amico Bignami and Ettore Marchiafava, described this syndrome in the postmortem study of an Italian alcoholic in 1903. There have only been about 250 cases reported in the literature.[3-6] There are two main subclinical types: the first is the severe type which affects consciousness level and causes coma; and the second one is the less severe type which can cause similar symptoms as in alcohol dependency.[7]

Postmortem histopathological studies reveal, tissue loss and colour change (from yellow to red) especially in the middle area of corpus callo-

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sum. Microscopic study of the tissues shows macrophages with digested lipid particles and demyelination. Inflammatory changes are not observed. These findings can rarely be seen in areas other than corpus callosum such as anterior and posterior commissure.[1,3,6-10]

The pathophysiology of MBD is still unknown.[3] MBD can be treatable by correction of vitamin deficiency and supportive treatment.[3,7,9] Seizure, stupor and coma may develop in advanced cases.

CASE REPORT

A twenty five year old male patient who had been consuming about 5000 mL/day alcohol for 10 years, had blurry vision for two months; muscle weakness and balance disturbance for over a month. A week before admittion to emergency room, he had difficulty of swallowing and changes in mental status. When brought to emergency room, he was unconscious (Glasgow coma scale: 5/15), and had deep respiratory failure. The patient was accepted to intensive care unit, intubated urgently and mechanically ventilated for three weeks. In the laboratory studies total protein and albumin levels were low, suggesting malnutrition. Cerebrospinal fluid examination wasn't performed.

In neurological examination, he had bilateral vertical and horizontal nystagmus; diminished reflexes, decreased response to painful stimuli, and rigidity in all extremities. Optic nerve examination was normal. Blood glucose, vitamin B_{12}, and folic acid levels were low. Liver and kidney function tests were in normal ranges. Cranial computerized tomography was normal. However, magnetic resonance imaging (MRI) revealed common signal changes presenting as areas of low signal intensity on T_1W images and high signal intensity on T_2W and fluid attenuated inversion recovery (FLAIR) images (TR-8800, TE-98, TI-2200), hyperintensity on diffusion-weighted images (DWI) and relatively decreased signals on apparent diffusion coefficient (ADC) mapping in the following areas of the brain: anterosuperior pons, right and left cerebral pedincles, bilateral internal capsule back leg and lateral thalamus, corpus posterior and splenium of corpus callosum, bilateral periventricular white matter areas and centrum semiovale (Figure 1).

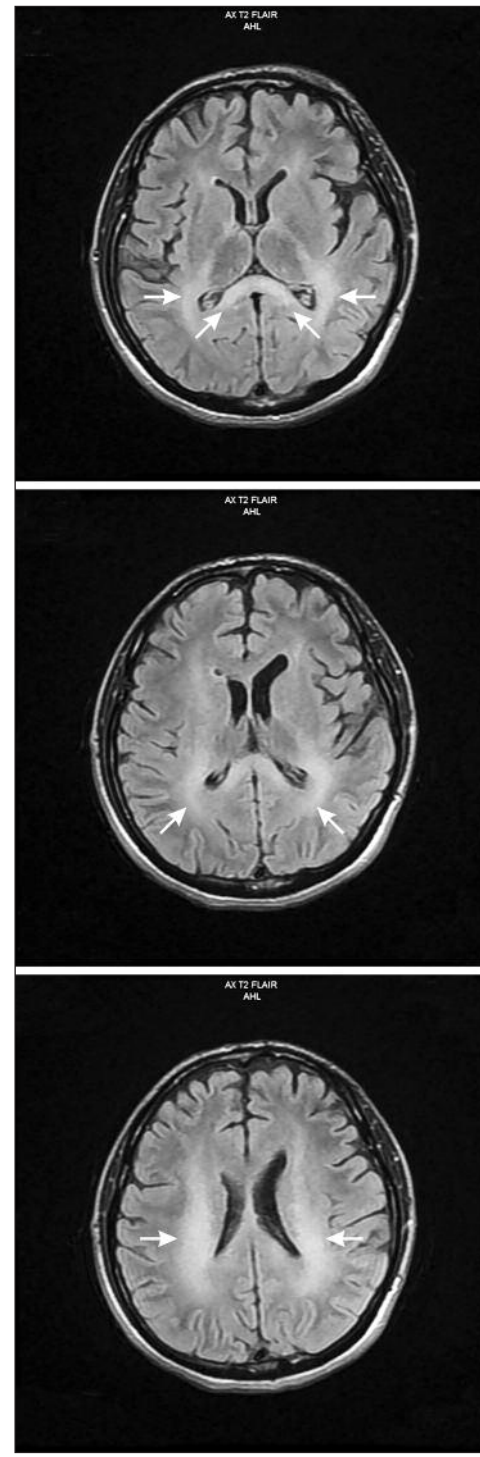

FIGURE 1: Axial flair images on admission show hyperintensity in the corpus callosum and bilateral internal capsule back legs and lateral thalamus.

Treatment of MBD is supportive. The patient was given intravenous thiamine treatment. A month later although the MRI findings didn't change on T_2W and FLAIR images, but on DWI shows decreased hyperintensity on corpus callosum and other sites. The patient was clinically better; awake, cooperated and fully oriented; speech and swallowing reflexes were sufficient. He presented distinctive recovery at the time he was discharged. The clinical signs and radiologic findings of the patient were evaluated as MBD (Figures 2 and 3). The patient was discharged and referred to a physical rehabilitation center for recovery. Informed consent was obtained from the patient's mother for publication.

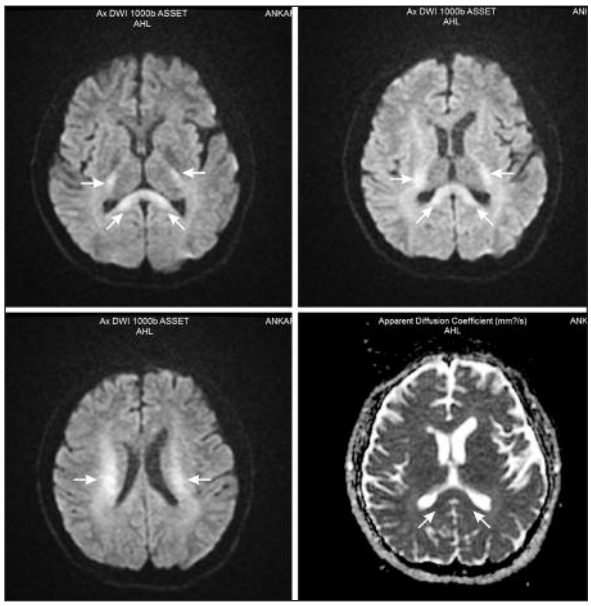

FIGURE 2: Diffusion weighted images (DWI) also shows hyperintensity in these regions with relatively decreased signal on apparent diffusion coefficient (ADC) mapping.

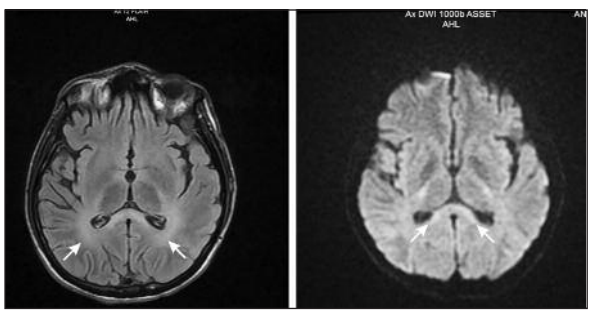

FIGURE 3: Diffusion weighted images (DWI) shows decreased hyperintensity on corpus callosum after treatment.

DISCUSSION

Diagnosis of MBD is made by radiologic imaging in addition to clinical findings.[1,3,5-8] According to timing, there are three types of the disease. In sub-acute type, patients suffer from dementia, dysarthria, and rigidity of muscles, but they can live for years. Although acute type is easy to differentiate from Wernicke Encephalopathy (WE) radiologically, it may be difficult to differentiate clinically. Some cases may even be accompanied by WE. This form is severe, causing seizure and unconsciousness which can lead to death. Typically, cranial MRI revealed high signal intensity on T_2 and FLAIR sequences in the corpus callosum. In WE, MRI revealed abnormal signal intensity and contrast in mamillary body, periaqueductal area and third ventricle wall. Chronic MBD is characterized with dementia and it can be mixed with Alzheimer's disease, multienfarct dementia, or Pick disease. MRI findings of acute pontin myelinolysis, which develops due to rapid treatment of hyponatremia, and it should be kept in mind in differential diagnosis.

T_2 signal abnormalities in corpus callosum is diagnostic for MBD. Lesions in MBD are large, often symmetrically in the midline in the splenium and don't reach the edge of corpus callosum. In our patient MRI showed iso-low signal on T_1- weighted images (TR/TE 500/15 ms), high signal on T_2- weighted images (TR/TE 4500/101 ms) and FLAIR (TR/TE8402/101 ms) sequences and hyperintensity on DWI with relatively low signal on ADC mapping on pons anterosuperior right-left, cerebral pedincules, bilateral internal capsule back legs and lateral thalamus, corpus posterior and splenium of corpus callosum, bilateral periventriculer white matter areas and centrum semiovale. When both MR diffusion images were compared, the signal intensity of corpus callosum was more intense in the former MRI. The other sequences were identical in both MRI.

It is not known how MBD affects commissural structures, but it is thought that cortical laminar sclerosis which is replacement of third level neurons with glial tissue, develops secondary to corpus

callosum damage. Since the incidence of the disease is very low among alcohol dependents, it is doubtful that direct toxic effects of alcohol is responsible for the etiology of the disease. It has been reported that rarely, similar clinical symptoms have encountered in patients who never drink alcohol. Hence, it is thought that alcohol isn't single reason for MBD.[1,2]

In conclusion cranial MRI especially in diffusion sequences is very helpful to make early diagnosis and follow-up in MBD. With DWI the rapid resolution of the callosal and extracallosal hyperintensities may demonstrate early despite the other sequences don't change. When clinical signs are not specific and severe, diagnosis can be made based on the imaging findings.

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