Cortical Involvement in Marchiafava-Bignami Disease Can Be a Predictor of a Poor Prognosis: A Case Report and Review of the Literature

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Abstract

Marchiafava-Bignami disease (MBD) is a rare alcohol-associated disorder characterized by demyelination and necrosis of the corpus callosum. We herein present the case of a 56-year-old man with chronic alcoholism who was admitted to our hospital in a coma without focal or lateralizing neurological signs. MRI revealed a callosal lesion consistent with MBD and additional bifrontal linear cortical lesions. The callosal lesion completely disappeared with intravenous administration of high-dose multivitamins and corticosteroids, although the patient remained in a vegetative state. This case further supports the notion that cortical involvement in patients with MBD is a predictor of a poor prognosis.

Key words: Marchiafava-Bignami disease, alcoholism, vitamin B1, cortical lesion, Morel’s laminar sclerosis, prognosis

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Introduction

Marchiafava-Bignami disease (MBD) is a rare neurological disorder associated with chronic, heavy alcohol consumption and/or malnutrition leading to confusion, seizures and frequently death. It is pathologically characterized by demyelination and necrosis of the central layer of the corpus callosum. The prevalence of this disease is unclear. A review of the literature in 2001 found only approximately 250 cases since the first report by Marchiafava and Bignami in 1903. Over 90% of the patients exhibited a poor prognosis (1). The increased availability of MRI in daily clinical practice, however, makes early in vivo diagnosis of this condition and the discovery of mild cases possible. With adequate therapy, these patients can recover completely with disappearance of the callosal lesions on serial MRI (2). In contrast, recently reported cases of MBD with predominant bifrontal cortical involvement (3-9) in which the patients exhibited very poor prognoses imply that cortical lesions may be a marker of poor outcomes. We herein present a similar case of MBD with cortical involvement in which a poor prognosis resulted from a cortical not a callosal lesion.

Case Report

A 56-year-old man who had suffered from chronic alcoholism for over 30 years was admitted in a coma following generalized fatigue and a loss of appetite that lasted for two weeks. He had a history of partial gastrectomy due to a gastric ulcer in his thirties. He had been admitted to a regional hospital approximately once a year due to appetite loss. Each time of hospitalization, intravenous infusions over several days improved his condition. On admission, a physical examination revealed a remarkably emaciated comatose man with slow and deep breathing, sinus tachycardia, normal blood pressure and normal body temperature. There were no focal or lateralizing neurological signs. The laboratory data revealed macrocytic anemia and hypoproteinemia. The patient’s liver and renal functions were almost normal; however, his serum level of vitamin B1 was low (12 ng/mL: normal range 20-50), although the level of vitamin B12 (361.1 pg/mL) was in the normal range (180-914 pg/mL). The protein level in the cerebrospinal fluid was 76 mg/dL.

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nous hyperalimentation was initiated with high-dose multivitamins accompanied by a three-day administration of high-dose methylpredonisolone (10). His condition gradually improved. He opened his eyes on the fifth day of hospitalization and did not show any meaningful responses. Reintubation was decided and he was transferred to a sanatorium. Three months later, still in a vegetative state, he was transferred to a sanatorium.

Figure. MRI of the patient (1): A: Axial DWI (TR 1,000, TE 80). B: Axial FLAIR (TR 14,000, TE 100). C: Axial T2 (TR 5,000, TE 80). D: Axial T1 (TR 400, TE 20). E: Axial T1 contrast enhanced (TR 400, TE 20). F: Axial T1 FLAIR (TR 8,000, TE 200, 80 mg of Gd-DTPA). G: Axial T2 FLAIR (TR 4,000, TE 120, 80 mg of Gd-DTPA).

The patient was diagnosed as having MBD, and intravenous hyperalimentation was initiated with high-dose multivitamins. On admission, he was still completely mute with tetraplegia and did not show any meaningful responses. Reintubation was decided and he was transferred to a sanatorium.

Table. Marchiafava-Binami Disease with Cortical Involvement Proven by MRI Study

<table>
<thead>
<tr>
<th>Author</th>
<th>Year</th>
<th>Age (Y)</th>
<th>Chronic alcoholism (duration)</th>
<th>Consciousness (level)</th>
<th>Seizure</th>
<th>Vit B1</th>
<th>CSF</th>
<th>MRI findings of callosal lesion</th>
<th>MRI findings of cortical lesion</th>
<th>White matter lesion</th>
<th>Therapy</th>
<th>Prognosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Yoshizaki et al [8]</td>
<td>2010</td>
<td>55 M</td>
<td>Yes</td>
<td>Stupor</td>
<td>No</td>
<td>ND</td>
<td>Normal</td>
<td>G, S</td>
<td>Persistent frontoparietal</td>
<td>No</td>
<td>Yes</td>
<td>Memory deficit</td>
</tr>
<tr>
<td>Lee et al [9]</td>
<td>2011</td>
<td>52 M</td>
<td>Yes (30Y)</td>
<td>Confusion</td>
<td>No</td>
<td>ND</td>
<td>S</td>
<td>Resolution</td>
<td>Frontal</td>
<td>No</td>
<td>Yes</td>
<td>Relative improvement</td>
</tr>
<tr>
<td>Present Case</td>
<td>2012</td>
<td>56 M</td>
<td>Yes (&gt;30Y)</td>
<td>Coma</td>
<td>No</td>
<td>12</td>
<td>Protein</td>
<td>S Resolution</td>
<td>Frontal</td>
<td>No</td>
<td>Yes</td>
<td>Vegetative state (&gt;3Mo)</td>
</tr>
</tbody>
</table>

ND: not described, NS: not specified, M: male, F: female, D: day, Mo: month, Y: year, G: genu, B: body, S: splenium

*: They described that "in most cases, the entire corpus callosum was affected".
Discussion

The increased availability of brain MRI has led to the discovery of mild cases of MBD with partial callosal involvement and favorable outcomes (2). It has also led to the discovery of cortical involvement in a few cases of MBD, almost always with a poor prognosis (ref 3-9; Table). Since information about callosal lesions (focal or diffuse, reversible or persistent) is insufficient, the question of whether the unfavorable outcome observed in this case was due to the cortical or the callosal lesion has not been resolved.

It is certain that, in two cases (ref. 9 and ours), the callosal lesions were confined to the splenium and completely resolved. The prognoses in these two cases were very different, however. This difference might be due to the extent of cortical involvement, as the lesion in the patient of Lee et al. (9), who exhibited a relatively favorable outcome, was restricted bilaterally to the precentral gyri, whereas, in our patient, who exhibited a poor prognosis, the lesion extended from the precentral gyri to the superior and middle frontal gyri (Figure). This indicates that extensive cortical lesions in MBD patients can lead to a poor prognosis, even if the callosal lesion is slight enough for complete remission.

These cortical lesions, characteristic of MRI in patients with MBD, are considered to indicate Morel’s laminar sclerosis, a unique pathological feature seen in the brains of chronic alcoholics (3). Such lesions are also observed in cases of Wernicke’s encephalopathy, another complication of chronic, heavy alcohol consumption and/or malnutrition (11). In addition, most cases of Wernicke’s encephalopathy with cortical involvement are also associated with poor outcomes (11). The cortical lesions might not be caused by direct alcoholic toxicity, but rather thiamine deficiency, since over half of patients with Wernicke’s encephalopathy with cortical involvement are not alcoholics (11). Unfortunately, the pretreatment serum thiamine levels have not been reported, except in one case of MBD with cortical involvement (8).

Precisely why frontal cortices are vulnerable to chronic alcoholism and/or thiamine deficiency is not clear. In an animal experiment, however, thiamine deficiency, but not chronic ethanol consumption, was reported to decrease glutamate uptake in the prefrontal cortex, leading elevation of glutamate and neurochemical dysfunction (12). Further accumulation of similar cases with precise clinical and laboratory information is required to elucidate both the clinical influence and pathophysiology of cortical lesions in patients with MBD.

The authors state that they have no Conflict of Interest (COI).

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References