We report neuroimaging studies of Marchiafava-Bignami disease (MBD) diagnosed on a 39-year-old alcoholic man with acute clinical deterioration. Initially, cranial CT and MRI demonstrated diffusely edematous change from the genu to the splenium of the corpus callosum (CC) characteristic for acute stage of MBD. Decreased blood perfusion in bilateral parietal and occipital lobes was also noted in SPECT study. Prompt treatment with thiamine and vitamin B complex resulted in clinical improvement. 8 months later, follow-up MR imaging showed typical features of chronic MBD—atrophy with cystic change in the CC.

To our knowledge, this is the first case of MBD reported in Taiwan literature.

Key words: Alcoholism; Brain, corpus callosum; Brain, CT; Brain, MRI; Brain, SPECT; Marchiafava-Bignami disease

Marchiafava - Bignami disease (MBD), pathologically characterized by necrosis and demyelination within the medial layer of the corpus callosum (CC), is a rare disorder observed in people with chronic alcoholism and/or malnutrition [1-3]. In the past, the diagnosis of MBD was only made in the postpartum exam. However, after the introduction of CT and especially MRI, more cases were reported in its early stage and more chronic subtype were diagnosed in living people. Nevertheless, efficient treatment has not established, the prognosis is still pessimistic [1, 4].

CASE REPORT

A 39-year-old right-handed locksmith had a 27-year history of alcohol dependency, varied in amount and kinds. He often drinks all day with irregular diet. About 2 weeks before, he experienced sudden onset of slurred speech and mentality disturbance in the morning that never happened before. On admission, he was drowsy with poor attention. Wide-based, ataxic gait with deviation to right was noted. He had normal muscle power with mild hyperreflexia bilaterally. EEG finding and the results of laboratory examinations were unremarkable.

Cranial CT reveals decreased attenuation in the genu, body, and splenium of the CC (Fig. 1A). Magnetic resonance imaging demonstrated diffuse swelling of the CC from the genu to the splenium. This lesion appeared low signal intensity on T1-weighted image (Fig. 1B), turned bright on T2-weighted & fluid-attenuated inversion recovery (FLAIR) images (Fig. 1C), and strongly enhanced after administration of gadolinium (Fig. 1D). No definite signal abnormalities were seen in the cerebellum, subcortical white matter, or centrum semiovale (Fig. 1). Under the diagnosis of MBD, the treatment with intravenous infusion of thiamine and oral vitamin B complex was started.

Technetium\(^{99m}\) hexylmethylpropylene aminoxime –single photon emission computed tomography (\(^{99m}\)Tc HMPAO-SPECT) scan disclosed decreased perfusion in the regions of bilateral occipital-parietal cortices.
Neurobehavioral examination was arranged 4 days after admission. There was no obvious evidence of double hemianopia, left side hemialexia, or significant left side idiomatic apraxia. He could name object by right side nostril, write words by left hand, and copy drawing by right hand. The disconnection signs he showed were difficulty in cross replication hand posture and poor intermanual point localization which indicated disconnection in both hemispheric somatosensory fibers. He got clinical improvement during 12 days’ hospitalization and discharged with mildly unsteady gait and dysarthria. Eight months later, follow-up MR image showed atrophy of the CC with extensive cystic change in the middle layer (Fig. 3). On neurological examination he had definite improvement in gait, speech, and memory with some sequelae.

**DISCUSSION**

In 1903, Ettore Machiafava and Amico Bignami, two Italian pathologists, presented their autopsy findings of three Italian male alcoholics who died following seizure and coma. Since then, fewer than

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**Figure 1.** CT and MR images on admission. a. Axial cranial CT showed focal low-density areas in the genu and splenium of the CC. b. Axial T1-weighted MR images (SE 631/15/1) of brain revealed extensive lesion, of which the signal intensity the same as that of the gray matter, in the CC. c. Axial FLAIR MR images (SE 9000/105/1) of brain demonstrated high-signal-intensity lesion in the CC from the genu to the splenium. Note absence of definite white-matter disease within the subcortical white matter and centrum semiovale. d. Gadolinium-enhanced sagittal T1 weighted MR images (SE 650/17/1) revealed intense enhancement of callosal lesion.
200 cases of MBD have been reported in the medical literatures throughout the world and the majority of MBDs were diagnosed on autopsy [1, 4, 5]. Because of the high lethality and the scarcity of diagnosed cases, it is difficult for us today to realize the full view of MBD.

The clinical features of MBD varied widely. There were often nonspecific neurological deficits, such as motor (apraxia, dysarthria, gait disturbance) or cognitive (agraphia, alexia, disorientation, memory disturbance) dysfunction, reported. Some of them were associated with hemispheric disconnection syndrome caused by damage to the fibers of the CC. The others may be resulted from other alcohol-related neurologic disorders [1, 2]. MBD, based on the rate of clinical progression, had been divided into acute, subacute, chronic subtypes. Patients with acute or subacute MBD had sudden onset conscious deterioration, dementia, seizures, or coma. Acute MBD with poor prognosis resulted in death within a short time after onset. The subacute form with a more protracted clinical course usually led to death in a few months. The other patients with slowly evolving cognitive impairment over several years were categorized into chronic MBD. Before advent of CT and MRI, this form constituted only 10% of cases with MBD, but recently the majority of reports are on patients diagnosed with chronic MBD [1, 6].

The pathophysiologic process of MBD still remains unknown. However, some published functional neuroimaging studies, including SPECT scan and $^{18}$F]-2-fluoro-2-deoxy-D-glucose positron emission tomography, provided interesting information about MBD, and revealed hypoperfusion and hypometabolism in focal regions of the brain. The authors suggested that these functional impairments would account for cognitive deficits and may be caused by lesions in the CC [1, 7, 8].

The diagnosis of MBD in vivo mainly depends on the neuroimaging evidences rather than the variable clinical features. During acute phase of this disease, on CT findings, there would be low-density and edematous changes in the CC with enhancement after contrast injection. On MR images, the corresponding area would appear low signal intensity on T1WI, high signal intensity on T2WI & FLAIR images and strong enhancement on post-contrast T1WI [1, 4, 9]. When it processes into postacute or chronic stage, the edematous change will be subsided and substituted with necrosis, demyelination, and cystic degeneration of the CC. Typical imaging findings in the postacute and chronic stages are atrophy of the CC with focal or

**Figure 2.** $^{99m}$Tc HMPAO-SPECT scan disclosed decreased perfusion in the regions of bilateral occipital-parietal cortices and brain atrophy.

**Figure 3.** 8 months later, follow-up MR images (SE 525/15/1) (SE 3500/96/1) showed severe atrophy of the CC with severe cystic degeneration in the middle layer, characteristic “sandwich sign” of MBD.
diffuse presence of hypointensity on T1WI and hyper-intensity on T2WI, particularly in the medial layer, that makes characteristic “sandwich sign” of MBD [1, 4, 9]. Extracallosal lesions such as in subcortical white matter, centrum semiovale, or anterior/posterior commissures have also been described [1, 2, 6].

Another specific process related to chronic alcoholism is Wernicke encephalopathy (WE) which mainly involves the periventricular regions, the medial thalamic nuclei, third ventricular floor, and mammillary bodies. Although WE with similar clinical manifestations as MBD, image study can make well differentiation between them. According to above-mentioned findings, we thought that our patient was subjected to acute stage of MBD when initial studies performed with typical imaging pictures of this stage, and this disease progressed into chronic stage proved by characteristic findings on follow-up images.

In recent years several reported cases, as well as our own, developed acute or subacute MBD. They were admitted to a medical ward due to sudden onset or rapid deterioration of neurological symptoms. Based on imaging studies, especially MRI, precise diagnosis was made immediately. Although there is no specific therapy established for MBD, subsequent treatment with thiamine or vitamin B complex resulted in good recovery [3-7]. Our findings, combined with other reports [3-7], suggest that acute or subacute MBD may not have rapid course leading to death, and early diagnosis with CT and MRI followed prompt treatment with thiamine or vitamin B complex appears to improve the prognosis of MBD.

**REFERENCE**

Marchiafava-Bignami Disease: 電腦斷層、磁振造影、單光子射出電腦斷層的影像表現

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我們提出 Machiafava-Bignami Disease (MBD) 神經影像上的發現，MBD 在一位三十九歲酗酒的男性被診斷出來，他呈現急性臨床變化。最初，頭部的電腦斷層和磁振造影影像指出從胼胝體的膝部到喙部產生了瀰漫水腫樣的病變，此為急性期特殊表現。單光子射出電腦斷層檢查指出在兩側大腦頂葉及枕葉有血流灌注減少的情形。即刻以 Thiamine 和維他命 B 群治療使他在臨床上逐漸地好轉。8 個月後，追蹤的磁振造影影像表現出慢性 MBD 典型特徵—胼胝體萎縮合併胼胝體內層囊狀病變。依我們所知，這是台灣文獻中有關 MBD 的第一篇個案報告。

關鍵詞：Marchiafava-Bignami Disease；大腦，胼胝體；大腦，電腦斷層；大腦，磁振造影；大腦，單光子射出電腦斷層掃描；酗酒