

Magnetic Resonance Finding of Acute Marchiafava-Bignami Disease with Diffuse Involvement: A Case Report

광범위한 침범을 보였던 급성 Marchiafava-Bignami 병의 Magnetic Resonance 소견: 1 증례

Young Jin Heo, MD, Hae Woong Jeong, MD, Hyun Sin In, MD

Department of Radiology, Busan Paik Hospital, Inje University School of Medicine, Busan, Korea

Marchiafava-Bignami disease (MBD) is a rare toxic disorder strongly associated with chronic alcoholism (1-3, 6, 7). It is characterized by progressive demyelination and necrosis of the corpus callosum. The process may extend to neighboring white matter and subcortical regions. We report a case of MBD in which fluid-attenuated inversion recovery and diffusion-weighted imaging revealed symmetrical hyperintense lesions with diffuse involvement of the corpus callosum, white matter, corticospinal tract, internal capsule, and middle cerebellar peduncle (3, 4, 8).

Index terms

Marchiafava-Bignami Disease
Corpus Callosum
Diffusion-Weighted Imaging

INTRODUCTION

Marchiafava-Bignami Disease (MBD) is a disease observed in chronic alcoholic patients which results from the symmetric demyelination of the corpus callosum (1). The clinical diagnosis of MBD can be difficult. Recent magnetic resonance (MR) imaging including diffusion-weighted imaging (DWI) have been helpful in analyzing the distribution of lesions as well as establishing the diagnosis. We report here on a case of MBD with symmetric involvement of the corpus callosum, white matter, corticospinal tract, internal capsule, and middle cerebellar peduncle.

CASE REPORT

A 49-year-old man was referred to the emergency department with dizziness, trembling hands and weakness in both legs for a day. The patient had been an alcoholic with a daily intake of alcohol. DWI obtained at admission showed symmetrical hyperintense lesions involving the corpus callosum, white

matter, corticospinal tract, internal capsule, and middle cerebellar peduncle (Fig. 1). Fluid-attenuated inversion recovery (FLAIR) and T2 weighted images (T2WI) showed subtle high signal intensity and apparent diffusion coefficient mapping, which showed relatively high hypointensity in those lesions. No further enhancement of the lesions was observed after gadolinium. On the basis of the clinical history and imaging findings, MBD was diagnosed. The patient was admitted and given intravenous thiamine, which resulted in an almost complete remission of the symptoms related to the disease. Follow-up MR imaging performed 11 months after the onset showed the disappearance of signal-intensity abnormalities on FLAIR, T2WI, and DWI (Fig. 2).

DISCUSSION

MBD is a rare form of toxic demyelination and necrosis of the corpus callosum associated with chronic alcohol consumption. This disease is occasionally observed in non-alco-

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Corresponding author: Hae Woong Jeong, MD
Department of Radiology, Busan Paik Hospital,
Inje University School of Medicine, 633-165
Gaegeum-dong, Busanjin-gu, Busan 614-735, Korea.
Tel. 82-51-890-6549 Fax. 82-51-896-1085
E-mail: hwjeong2000@lycos.co.kr

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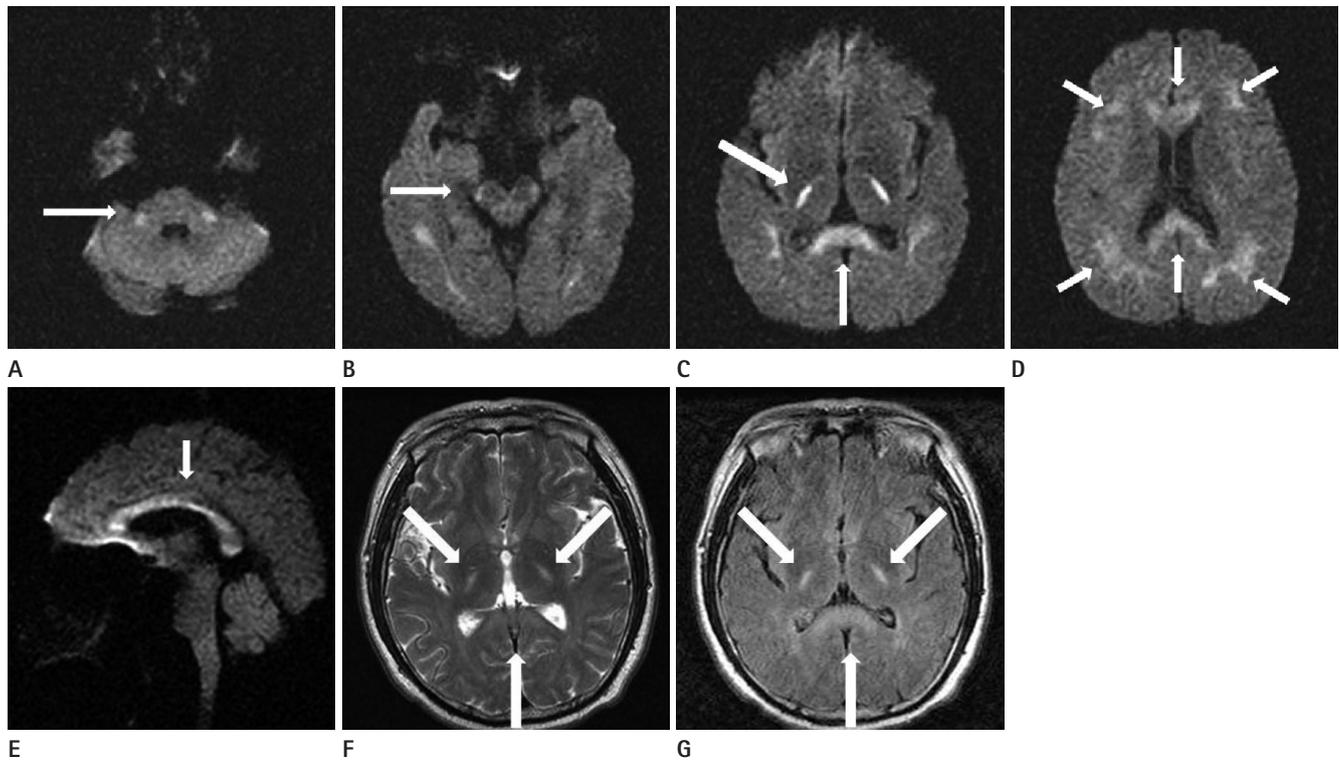


Fig. 1. Axial diffusion-weighted magnetic resonance images obtained upon admission show symmetrical hyperintense lesions involving the middle cerebellar peduncle (A), corticospinal tract (B), posterior limb of internal capsule and the splenium of the corpus callosum (C), as well as the corpus callosum and white matter (D). Sagittal image shows the involvement of the entire corpus callosum (E). Axial T2WI (F) and FLAIR (G) images also showing hyperintense lesions in the posterior limb of the internal capsule and splenium of the corpus callosum. Note.—FLAIR = fluid-attenuated inversion recovery, T2WI = T2-weighted image

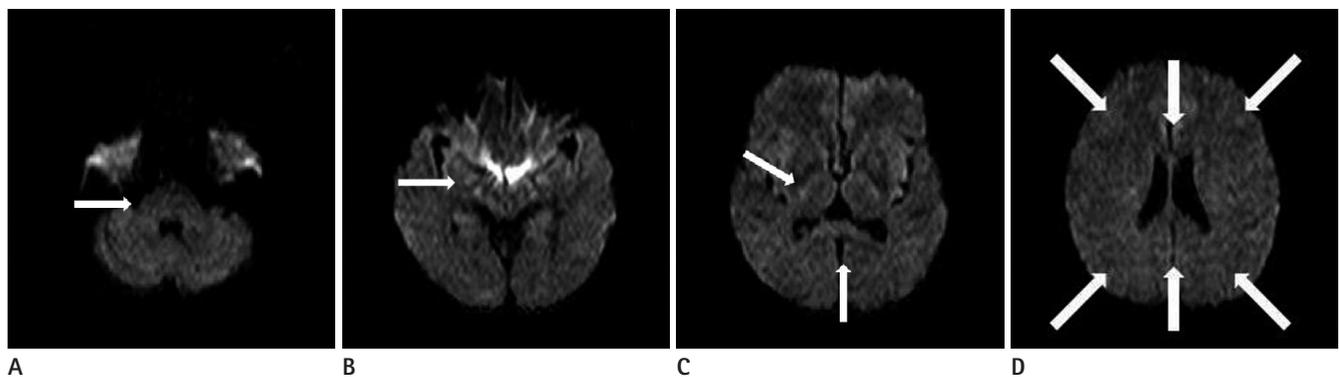


Fig. 2. Follow-up diffusion weighted magnetic resonance images obtained 11 months after the initial study show the disappearance of signal-intensity abnormalities including middle cerebellar peduncle (A), corticospinal tract (B), posterior limb of the internal capsule and splenium of the corpus callosum (C), as well as the corpus callosum and white matter (D).

holic patients as well. It is generally accepted that the cause of the disease is mainly due to the deficiency of vitamin B complex (1, 5, 9). Most patients are male, between 40 and 60 years of age, and have a history of chronic alcoholism with poor oral intake (1, 2).

The main pathologic change associated with MBD is a degeneration of the corpus callosum with different degrees of damage, from demyelination with preservation of axons, to

necrosis. The genu is most frequently involved, but the degeneration can extend to the entire corpus callosum (5, 6). Other white matter tracts such as the anterior and posterior commissures, corticospinal tracts, hemispheric white matter, and middle cerebellar peduncle may also be involved (2). Selective involvement of the entire length of the corpus callosum and chronic alcoholism, other clinical features, and MRI findings support the diagnosis (1). MR imaging is the best technique

for evaluating MBD lesions (3). Characteristic MR imaging findings are symmetric lesions on the corpus callosum; but, the lesions may be also found in the hemispheric white matter, cortex, middle cerebellar peduncles, and internal capsules (4). In the acute phase, the affected areas are hypointense on T1WI and hyperintense on T2WI, due to edematous change with or without demyelination. However, these lesions do not have mass effect and may show peripheral contrast enhancement during the acute stage (2). At 30 hrs after initial imaging, hyperintense signals on DWI appeared which was in contrast to the initial image where the signals did not appear. These findings suggest that the initial change in the corpus callosum was partly attributed to vasogenic edema and that the lesion was then converted into cytotoxic edema. The necrotic lesions in the corpus callosum in chronic stage confirmed the cytotoxic process (7).

Acute MBD is a rare toxic disease, but typical history, clinical manifestation, and MR findings with DWI could facilitate its diagnosis.

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광범위한 침범을 보였던 급성 Marchiafava–Bignami 병의 Magnetic Resonance 소견: 1 증례

허영진 · 정해웅 · 인현신

Marchiafava–Bignami 병은 만성 알코올 중독과 관련성이 많은 드문 독성 장애이다. 이는 뇌들보의 점차적인 탈수초화와 괴사를 나타낸다. 진행과정 중에 인접한 백색질과 피질하의 병변까지 확산될 수 있다. 우리는 액체감식역전회복영상과 확산강조영상에서 뇌들보, 백질, 겉질 척수로, 속섬유막, 양측 소뇌다리 등에 광범위하게 대칭적으로 고신호강도를 보였던 1 증례를 경험하여 보고하고자 한다.

인제대학교 의과대학 부산백병원 영상의학과학교실