

MR IMAGING IN ACUTE MARCHIAFAVA-BIGNAMI DISEASE: A CASE REPORT

IMAGING IN RISONANZA MAGNETICA NELLA MALATTIA DI MARCHIAFAVA-BIGNAMI IN FORMA ACUTA: CASE REPORT

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CASE REPORT

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Case Report

Here we describe the clinical case of a 57-year-old male patient, with a 30-year history of alcohol (wine). On examination, the patient was confused. He showed gait disturbance and lack of motor coordination. The patient was examined by a 1.5 T superconducting MR scanner (Toshiba ExcelArt™). The MRI protocol included axial fast spin-echo T2-weighted sequences (TR/TE range, 4,000-4,900/90-120); field of view [FOV], 18-22 × 22 cm; matrix, 160-192 × 256-384; number of excitations [NEX], 1-2; axial contrast-enhanced spin-echo T1-weighted sequences (450-540/10-15; FOV, 18-22 × 22 cm; matrix, 160-176 × 258-384; NEX, 1-

Abstract.

Marchiafava-Bignami disease (MBD) is a rare complication of chronic alcoholism. MBD is characterized by primary demyelination of the corpus callosum. We report one case of MBD in which MR imaging showed high T2 FLAIR and diffusion-weighted signal intensity of the corpus callosum and in the deep and shallow white matter, in particular, of the temporo-occipital and fronto-parietal lobes. Here we describe the clinical case of a 57-year-old male patient, with a 30-year history of alcohol (wine) brought to us in a state of mental confusion. He showed gait disturbance and lack of motor coordination.

KEYWORDS: Marchiafava-Bignami disease, Diffusion MRI, Corpus Callosum.

Riassunto.

La malattia di Marchiafava-Bignami (MBD), è una rara complicanza dell'alcolismo cronico. La MBD è caratterizzata dalla demielinizzazione primaria del corpo calloso. Riportiamo un caso di MBD in cui le immagini di risonanza magnetica mostrano iperintensità di segnale in T2 FLAIR (fluid-attenuated inversion recovery) e in diffusione del corpo calloso e della sostanza bianca, profonda e superficiale, dei lobi occipito-temporale e fronto-parietale. Descriviamo un caso clinico di un paziente di 57 anni, alcolista da 30 anni (vino), giunto alla nostra attenzione in stato di confusione mentale. Presenta disturbi della coordinazione motoria e del linguaggio.

PAROLE CHIAVE: Malattia di Marchiafava-Bignami, Sequenze RM pesate in diffusione, Corpo Calloso.

2); axial FLAIR sequences (8,000-10,000/105-120; FOV, 18 × 22; matrix, 160-192 × 256-320; NEX, 1-2; inversion time, 2,300-2,600 msec); and axial single-shot spin-echo echo-planar diffusion-weighted sequences with b values of 0 and 1,000 s/mm² along all three orthogonal axes (4,000-8,000/95-120; FOV, 22-25 × 26-30 cm; matrix, 128 × 128; NEX, 1). FLAIR images showed abnormal hyperintensity in the corpus callosum (Fig. 1) and weak hyperintensity of the temporo-occipital cortices (Fig. 2), while diffusion-weighted images showed symmetrical hyperintense lesions with relatively reduced ADC symmetrical hyperintense lesions in the fronto-parietal (Fig. 3) and in the temporo-parietal

cerebral cortices (Fig. 4). The patient was admitted and given intravenous vitamin B complex, which resulted in a nearly complete settlement of the clinical manifestations of the disease.

Discussion

MB disease or syndrome is a primary degeneration of the corpus callosum associated with chronic alcohol consumption, but is occasionally seen in non-alcoholic patients (1, 2). The main pathologic change associated with Marchiafava-Bignami disease is a degeneration of the corpus callosum with different degrees of damage, from demyelination with preservation of axons to necrosis accompanied in few cases by bleeding during the subacute phase. The genu of the corpus callosum is more frequently involved, but the degeneration can extend to the entire corpus callosum, preferentially involving fibers of the central portion. Necrosis produces cystic lesions within the corpus callosum, mainly in the genu and splenium with gliotic walls and content of foamy macrophages (3). It is generally accepted that the disease is mainly due to a deficiency in the vitamin B complex, and although many patients improve following administration of these compounds, others do not, and some die from the disease (1, 4). At first, MB was thought to be particular to individuals living in the central region of Italy and consuming large amounts of inexpensively manufactured Chianti red wine. It is now known that MB occurs worldwide and that all alcoholic

beverages are implicated. Most patients are male, between 40 and 60 years of age, and have a history of chronic alcoholism and malnutrition (1, 5, 6). Other white matter tracts such as the anterior and posterior commissures and the cortico-spinal tracts may be involved. Lesions may be also found in the hemispheric white matter and in the middle cerebellar peduncles. Diagnosis is made on the basis of clinical findings in combination with imaging features (7, 8). Patients present acutely with mental confusion, disorientation, neurocognitive deficits, and seizures. Muscle rigidity and facial trismus may be severe. Most patients presenting with the acute type of MB will go into coma and eventually die, although as illustrated in this report, some survive. Acute MB may be difficult to distinguish from Wernicke encephalopathy and may occur together with it. Patients with Wernicke encephalopathy have ataxia, ophthalmoplegia, nystagmus, and confusion. Patients with MB may also present with hyponatremia. It is well known that rapid correction of the hyponatremia may lead to osmotic myelinolysis, and because 10% of all patients with pontine myelinolysis will have extrapontine lesions, some have suggested that MB may be yet another manifestation of the osmotic myelinolysis syndromes (5, 7, 8).

Conclusions

Acute MB is a disorder found in chronic alcoholics that should be differentiated from other alcohol-induced lesions. Increased T2 signal inten-

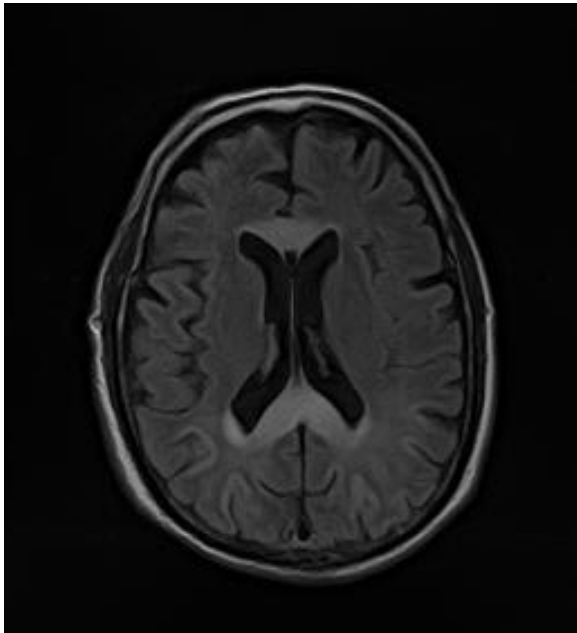


Fig. 1: Axial T2 FLAIR (fluid attenuation inversion recovery) MR image. Axial T2 weighted MR image shows hyperintense signal in the corpus callosum.

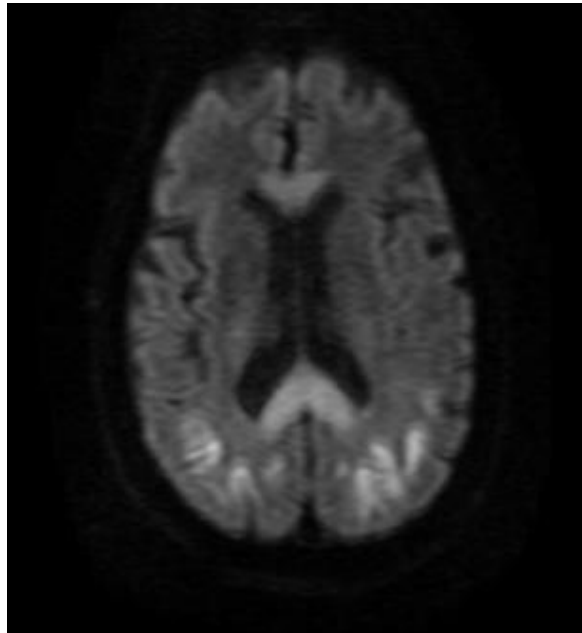


Fig. 2: Axial DWI MR image. The diffusion-weighted imaging (DWI) sequence shows a hyperintense signal in the temporo-occipital regions and in the corpus callosum.

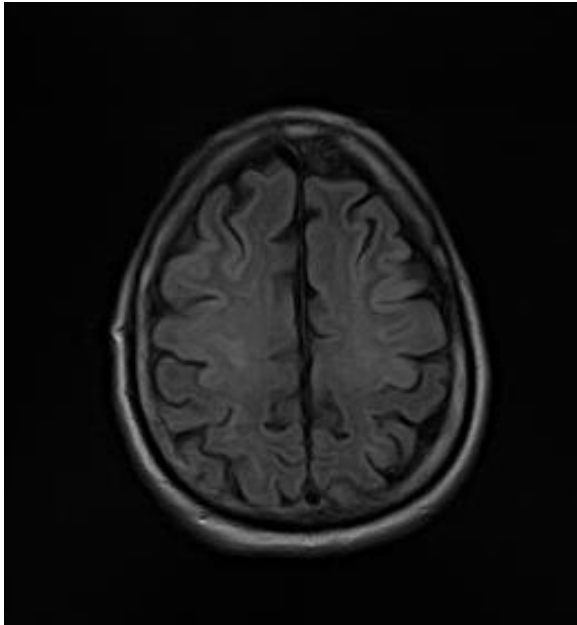


Fig. 3: Axial T2 FLAIR (fluid attenuation inversion recovery) MR image. Axial T2 weighted MR image shows weak and faded symmetrical hyperintense lesions in the fronto-parietal white matter.

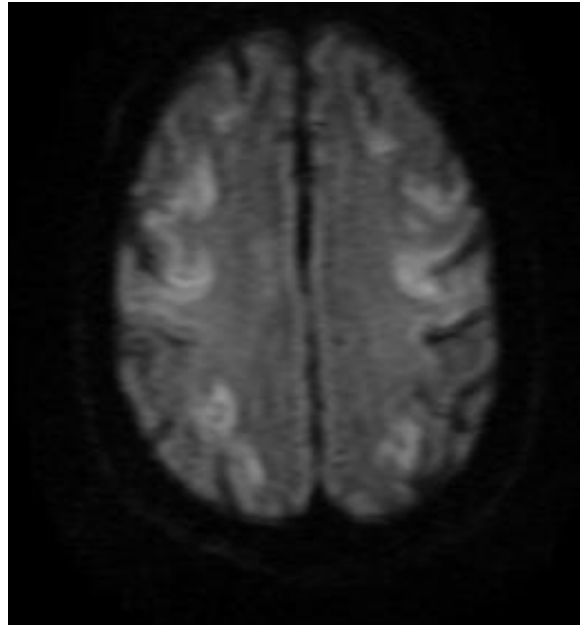


Fig. 4: Axial DWI MR image. The diffusion-weighted imaging (DWI) sequence shows abnormal increased signal intensity in the fronto-parietal white matter.

sity involving the body of the corpus callosum is present, and when identified in the correct clinical setting, the diagnosis of acute MB disease can be made. Prompt therapy may result in survival of these patients. This study confirms that during the course of acute Marchiafava-Bignami disease, MR imaging is the appropriate technique to evaluate morphologic and metabolic changes of the corpus callosum, including their evolution over time and after treatment. DWI sequences may support the pathogenetic theory. The resonance base-sequences may reveal a pathological condition only when this is associated to structural tissue damage. In our experience, DWI MR may integrate standard brain RM testing, with reduced timing, making it possible to detect alterations, which would remain unexposed/unidentified by means of standard base-sequences.

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