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Interhemispheric disconnection due to Marchiafava–Bignami disease

HMMTB Herath *MD*, Sudath Ravindra *MD*, Chulika Makawita *MD*, Anomali Vidanagamage *MD MRCP*, Bimsara Senanayake *MD FRCP*

Institute of Neurology National hospital of Sri Lanka, Colombo, Sri Lanka

Marchiafava–Bignami disease (MFBD) was first described by Italian pathologists Amico Bignami and Ettore Marchiafava in 1903 in an Italian Chianti wine drinker. Clinical presentation is variable, and include impaired consciousness, disorientation, aggression, seizures, depression, hemiparesis, ataxia, apraxia, psychosis, personality changes and coma.[1] Magnetic Resonance Imaging (MRI) is the most sensitive diagnostic tool for MFBD and reveals corpus callosal demyelination, necrosis and subsequent atrophy. No specific treatment is available but thiamine, folate, and other B vitamins (especially vitamin B12) are commonly used with some success. We report here a man with possible MFBD with features of interhemispheric disconnection on serial MRIs. Written informed consent was obtained from the patient's legal guardian for publication of this report.

CASE REPORT

A 65 years old male, an alcoholic, presented with gradual onset confusion over 2 weeks. He was a previously well, unmarried engineer with a good income, and had no vascular risk factors, fever or other constitutional symptoms. He was consuming alcohol for the past 20 years, roughly four units per day, and mainly consisted of beer, wine and spirits. On examination the patient was well build without any evidence of nutritional deficiencies. He was disorientated. Neurological exam revealed ataxia but with preserved motor power. There were no cranial nerve or peripheral sensory involvement. Examination of the higher cortical functions were abnormal with ideomotor apraxia and apraxia of the left (non- dominant) hand. He was unable to imitate hand gestures such as 'waving good bye' and could not mime tools such as a toothbrush and a pen on verbal command. He was also unable to perform tasks from left hand as well, such as picking up object from left hand and dressing. These findings were suggestive of possible interhemispheric disconnection. The rest of his examination was normal with no evidence of hepatic encephalopathy. Basic investigations including serum electrolyte, blood glucose, full blood count, renal functions, liver functions were normal. We were unable to measure serum B12, folate and thiamin levels. Magnetic resonance imaging (MRI) brain showed lesions involving the corpus callosum mainly posteriorly (Figure 1). One-month later MRI brain revealed anterior extension of the lesions involving the body and the head of the corpus callosum with necrosis of the posterior part (Figure 2). Lesions were demyelinating in nature. Cerebrospinal fluid examination was normal including testing for oligoclonal bands. His clinical presentation with abnormalities in higher cortical function, chronic alcoholism and the MRI findings pointed towards a diagnosis of MFBD. He was treated with high doses of vitamins, mainly vitamin B complex, thiamine and showed gradual improvement. In the MRI done 2 months later oedema of the corpus callosum has reduced and it appeared atrophic (Figure 3). Although ataxia and confusion improved significantly apraxia improved only slightly.

DISCUSSION

MFBD is a rare disease, characterized by demyelination and later necrosis of the corpus callosum. With the development of new imaging techniques such as MRI, more cases of MBD are diagnosed. Heinrich *et al*. analyses 50 cases of MFBD and describes two clinic-radiologic subtypes.[2] Type A – Clinically major impairment of consciousness with coma, stupor and poor outcome. Radiologically here is involvement of the entire corpus callosum with swelling and hyper intensities in T2.

Type B – Clinically mild impairment of consciousness and a favorable outcome. Radiologically there is only partial involvement

Address correspondence to: HMMTB Herath, National hospital of Sri Lanka, Colombo, Sri Lanka. E-mail: tharukaherath11@gmail.com

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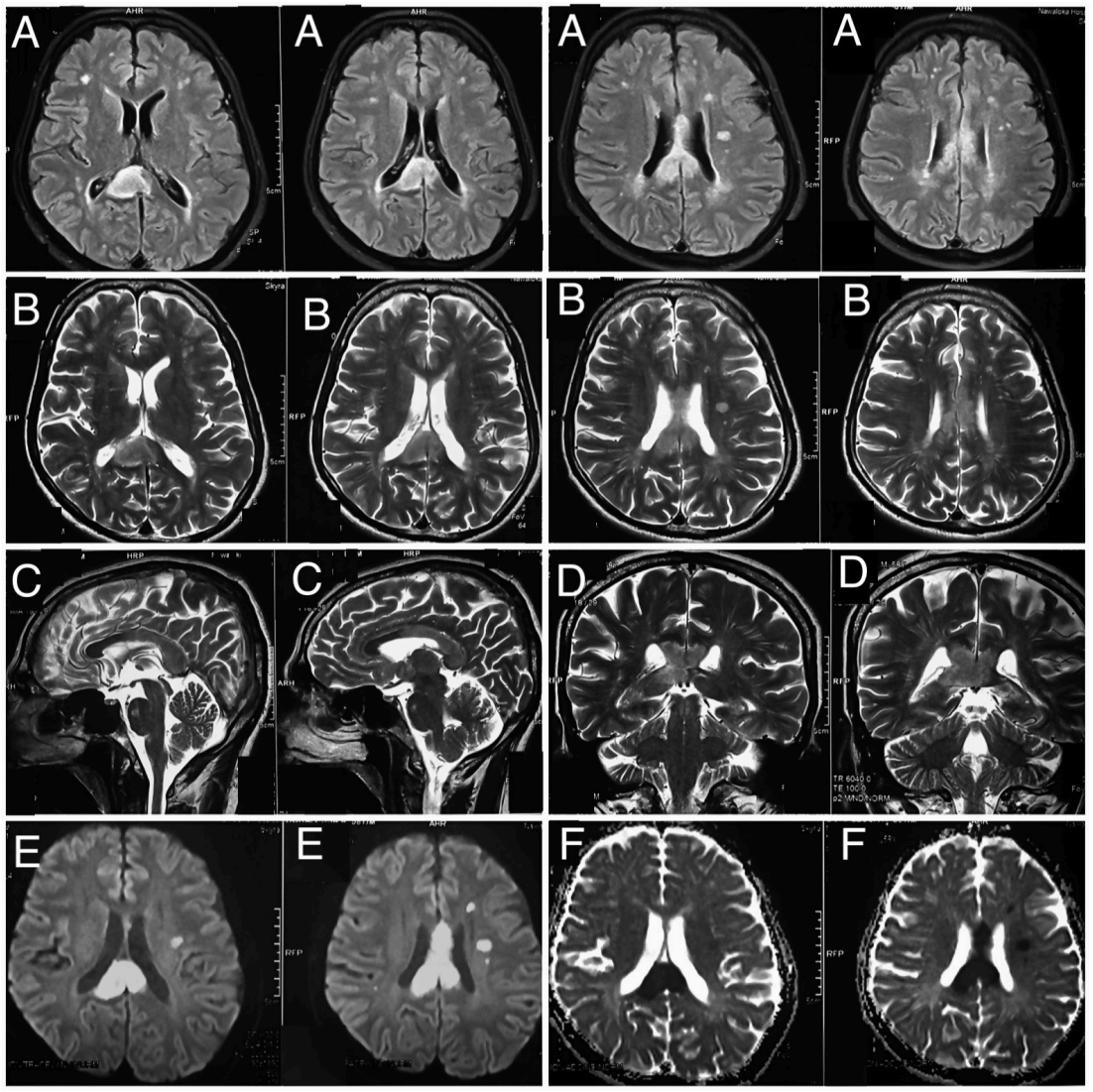

Figure 1. MRI images (A) FLAIR Axial, (B) T2 Axial, (C) T2 sagittal, (D) T2 Coronal demonstrate high signal and swelling of the posterior corpus callosum. There are multiple focal lesions involving the periventricular and deep white matter of the frontal lobes. (E) and (F) DWI and ADC images shows diffusion restriction in the involved corpus callosum.

of the corpus callosum with focal lesions. During the acute stage corpus callosum is edematous with T1 hypo intensity and T2 hyperintensity. This acute stage usually involves the splenium and genu of the corpus callosum.[3] In our patient the MRI evolved similarly with time. Initially the lesions were more posterior as shown in Figure 1 and later the body and the head of the corpus callosum were involved as shown in Figure 2. With time when the oedema decrease T2 signal intensity progressively reduces[4] and later when the lesions become chronic, corpus callosum will become atrophic. (Figure 3) Sometimes necrosis will produce cystic cavities within the corpus callosum mainly in the genu and splenium[5], which were also seen in out patient. Fiber tracking using diffusion tensor imaging is useful to increase the sensitivity of MRI in MFB and show the fiber loss throughout the corpus callosum.[6] Hyper intense lesions in the cerebral cortex, particularly in the frontal in T2 and fluid-attenuated inversion recovery (FLAIR) images have also been reported.[7] Similar multiple focal lesions involving the periventricular, deep white matter of the frontal lobes were evident in Figure 1.

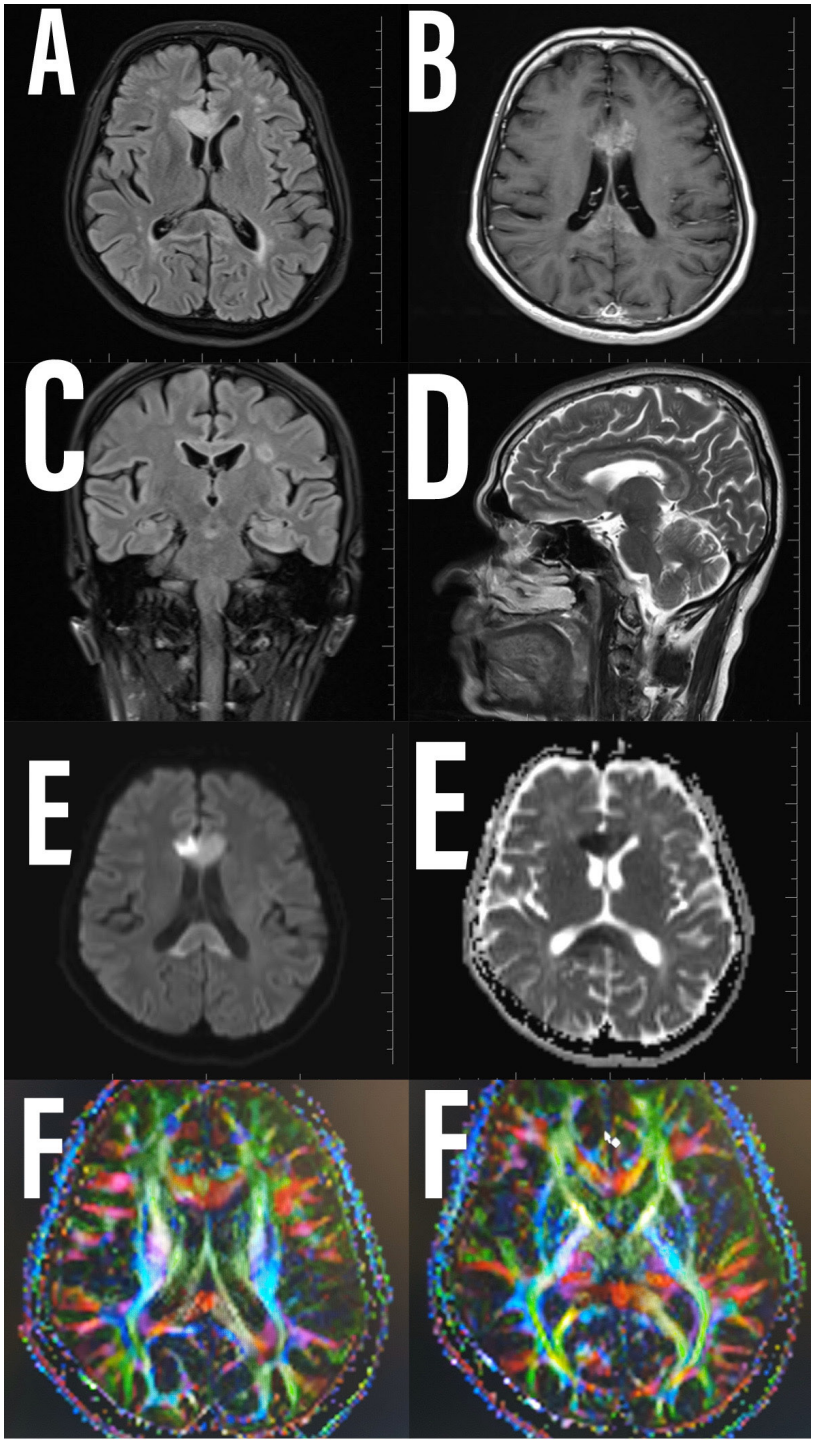

Figure 2. (A) FLAIR Axial (C) FLAIR Coronal (D) T2 sagittal images shows that the involvement of the corpus callosum has extended anteriorly involving the body and the head of the corpus callosum. Previously involved posterior corpus callosum and the splenium show central necrosis. (B) Contrast images reveal mild Gadolinium contrast enhancement of the lesions. DTI images demonstrate loss of integrity and the volume of the transverse fibers of the posterior corpus callosum.

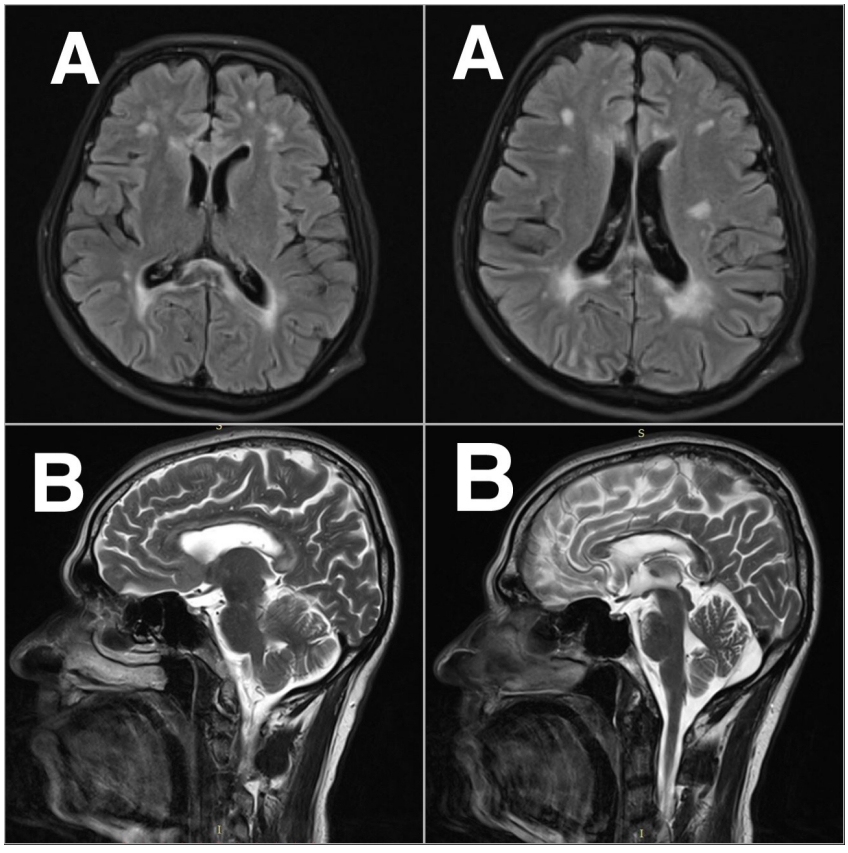

Figure 3. MRI images (A) FLAIR Axial and (B) FLAIR sagittal images showing atrophied corpus callosum and increased FLAIR signals in genu, body and splenium. Diffuse high FLAIR signals are seen in the periventricular white matters bilaterally sparing subcortical U fibers.

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