INTERNATIONAL JOURNAL OF EPILEPSY 2 (2015) 51–53

Available online at www.sciencedirect.com

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Image of the Month

Marchiafava–Bignami disease – An alcohol related medical emergency

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ARTICLE INFO

Article history:
Received 29 December 2014
Accepted 14 March 2015
Available online 15 May 2015

Keywords:
Marchiafava–Bignami disease
Alcohol
Neurological emergency

ABSTRACT

We report a case of Marchiafava–Bignami disease, an alcohol related neurological emergency.

A 50 years old male presented to the emergency department in an unconscious state. Patient was a chronic alcoholic and was found unconscious on the road. On examination his Glasgow coma scale (GCS) was E2M2V3. Pupils were normal size and reacting. Fundus examination was normal. In view of his poor GCS and inability to protect airway, patient was intubated and connected to ventilator. He was noticed to have tongue bite. In view of tongue bite patient was provisionally diagnosed as status epilepticus and started on Injection Thiamine 100 mg OD and 1 gm of levetiracetam. Hemogram, biochemical investigation, ECG, Chest X-Ray and EEG was normal.

Magnetic resonance imaging with diffusion weighted image showed restricted diffusion with low apparent diffusion coefficient in genu, body and part of splenium (Fig. 1A–F), bilateral middle cerebellar peduncles symmetrically (Fig. 2 A–B) and a small lesion in left frontal white matter. All the lesions were hyperintense on FLAIR and T2 weighted images. The characteristic MRI features with selective demyelination of the central layers of the corpus callosum with sparing the dorsal and ventral layers producing sandwich sign (Fig. 2C) was diagnostic of Marchiafava-Bignami disease (MBD).[1]

Alcohol abuse and withdrawal are associated with various neuropsychiatric manifestations like Wernicke's encephalopathy, Korsakoffs psychosis, alcoholic dementia, cerebellar degeneration, central pontine myelinolysis, delirium tremens and psychosis. MBD is a rare toxic disease most commonly seen in chronic alcoholics and less frequently in malnourished patients. Acute presentation is with confusion, seizures or death; sub acute and chronic forms have various degree of cognitive and behavioral disturbance. The symptoms of MBD are non specific and clinical diagnosis is often difficult. MBD is a radiological diagnosis which helps us in differentiating it from other alcohol related disorders. The disease typically affects the body of the corpus callosum, followed by the genu, and finally the splenium.[2] The entire corpus callosum may be also involved. The central layers of the corpus callosum are

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http://dx.doi.org/10.1016/j.ijep.2015.03.001

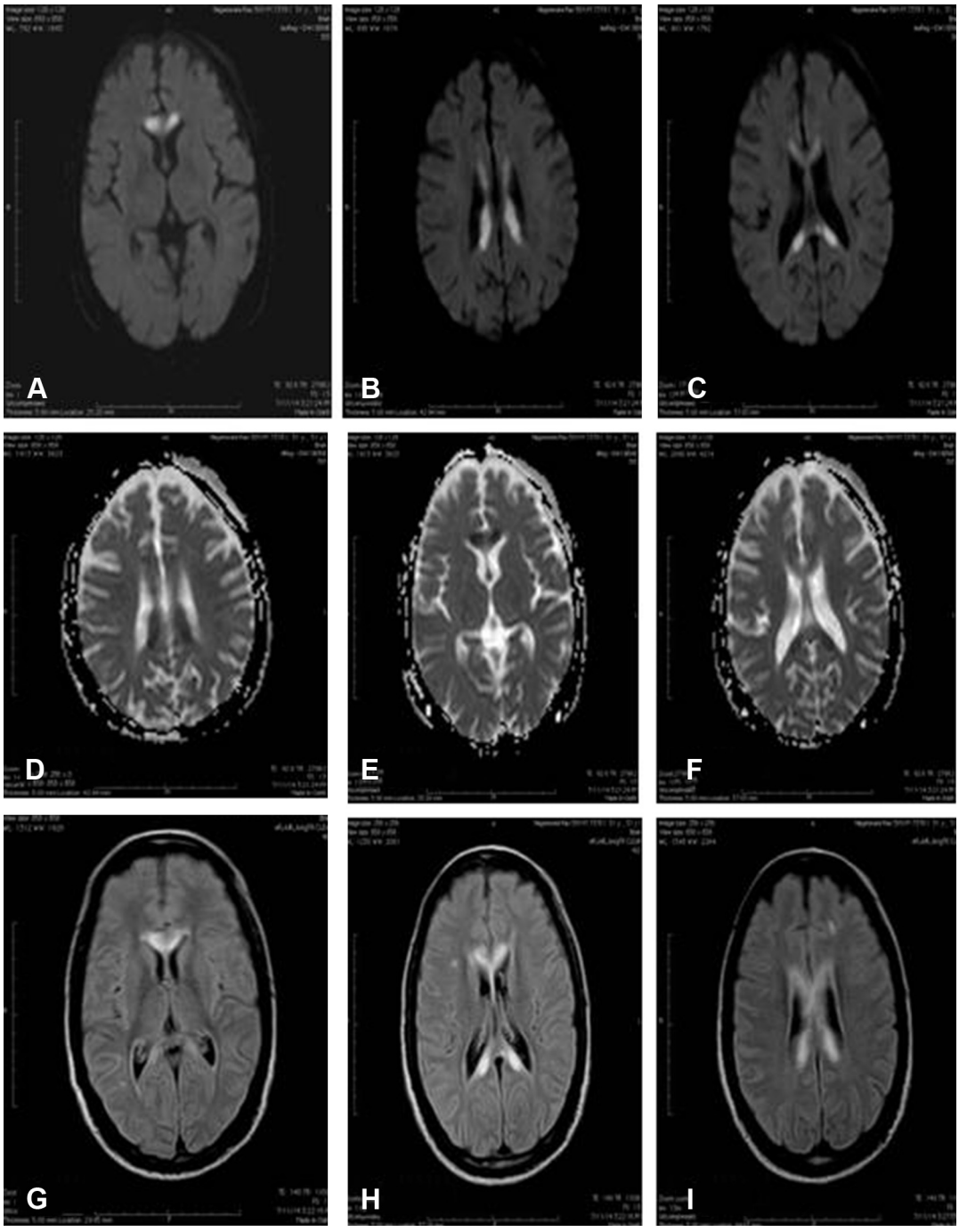

Fig. 1 – A–C: MRI brain axial diffusion weighted images show restricted diffusion in the genu, body and splenium of corpus callosum. D–F: Corresponding ADC images show low apparent diffusion coefficient values. G–I: Axial FLAIR sequences show hyperintensities in the genu, body and splenium of corpus callosum.

affected, with sparing of the dorsal and ventral layers producing sandwich sign.[1] Other white matter tracts such as the anterior and posterior commisures and the cortico-spinal tracts may be involved. Lesions may be found in the hemispheric white matter and in the middle cerebellar peduncles. The subcortical U-fibers tend to be spared.[2–4] Heinrich et al described 2 clinical subtypes of MBD in 2004. Type A is a severe form in which patient is in stupors or comatose with high prevalence of pyramidal-tract symptoms; radiologic features include involvement of the entire corpus callosum. In Type B, the patient s characterized by normal or mildly impaired mental status; radiologic features are partial or focal callosal lesions.[5]

Treatment of MBD is usually empirical and consists of thiamine and supportive care. Our patient gradually improved and was extubated on day 5. Patient is doing well on 1 month follow up. Marchiafava – Bignami disease is a condition associated with neurological emergency. However the

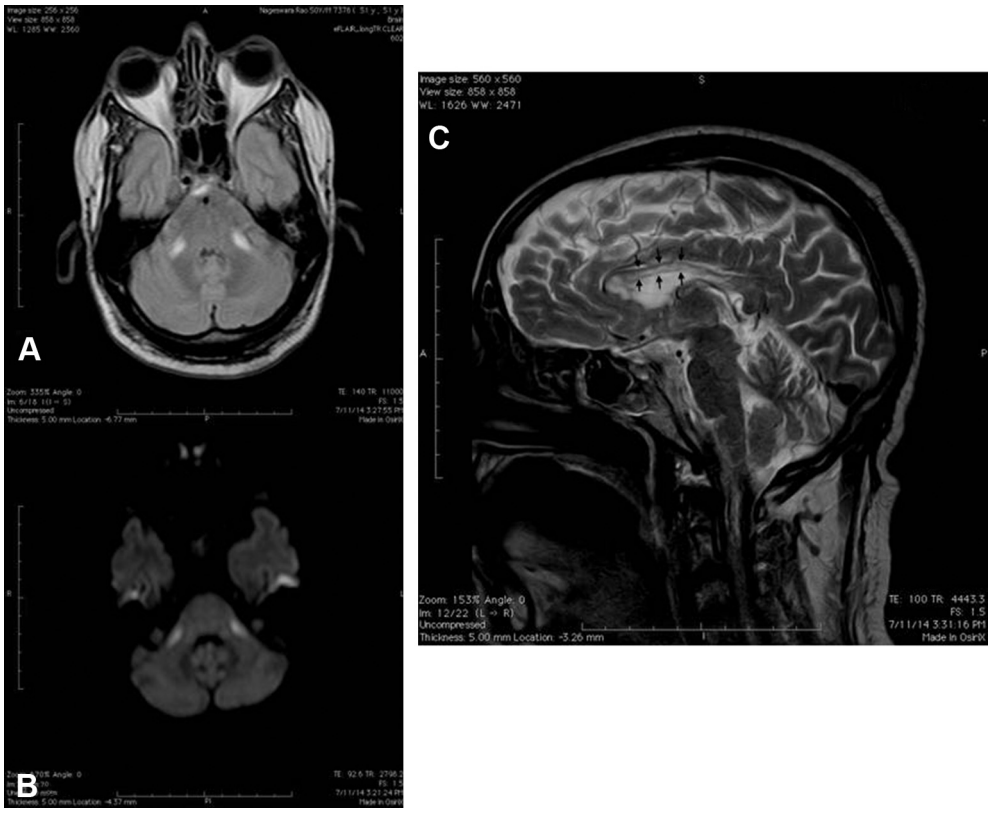

Fig. 2 – A–B: MRI brain axial flair images and DWI showing bilateral symmetric hyper intensities and restricted diffusion in bilateral middle cerebellar peduncles. C: MRI brain sagittal T2WI shows hyper intensity in the middle fibres of corpus callosum (arrows) with relative sparing of dorsal and ventral fibres showing the classic "Sandwich sign''.

outcome of MBD is variable. Although many patients may improve after administration of B vitamins, others do not. In these cases, morbidity and mortality are relatively high.[6,7]

Our case highlights the importance of early diagnosis and prompts appropriate management with vitamin B replacement which is critical in reversing the underlying pathophysiology in the early stage.

Conflicts of interest

All authors have none to declare.

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