

CASE REPORT

A RARE CASE OF: MARCHIAFAVA-BIGNAMI DISEASE

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HOW TO CITE THIS ARTICLE:

Manisha Panchal, Maulik Jethva, Anjana Trivedi, Pinkal Patel, Manish Yadav. "A Rare Case of: Marchiafava-Bignami Disease". Journal of Evolution of Medical and Dental Sciences 2014; Vol. 3, Issue 41, September 04; Page: 10254-10259, DOI: 10.14260/jemds/2014/3333

ABSTRACT: Marchiafava-Bignami disease is the symmetrical demyelination of the corpus callosum, mostly observed in people with chronic alcoholism & sometimes in people with chronic nutritional deficiency. We have been reported such case of a male patient who had history of chronic alcoholism, different clinical presentation and MRI findings consistent with the diagnosis of Marchiafava-Bignami disease.

KEYWORDS: Marchiafava-bignami syndrome, MBD, Corpus callosum demyelination, chronic alcoholism.

INTRODUCTION: Alcohol abuse and alcohol withdrawal are associated with a variety of neuropsychiatric syndromes, some of which are associated with significant morbidity and mortality. Marchiafava-Bignami disease (MBD) is a rare, alcohol associated disorder characterized by demyelination and necrosis of the corpus callosum.

CASE REPORT: A 36 year male patient was admitted in our medical ward with history of unconsciousness since two days. The patient was chronic alcoholic since last 18 years. There was no history of fever, headache, vomiting, seizures, jaundice, head injury and ear or nasal bleed. Neurologically, no significant past history was present. On admission, neurological examination showed a Glasgow Coma Scale of E2M2V4.

The occulocephalic reflex was normal. Examination of cranial nerves showed normal pupillary size and reaction. The fundus examination was normal. Motor examination did not reveal any focal deficit. No meningeal signs were present. Further physical examination and review of other systems was normal. Laboratory results revealed normal biochemistry profile.

X ray chest and ECG were normal. CSF studies were normal. MRI of the brain, which was performed immediately on admission in the emergency department, showed significant abnormalities: On T2W and FLAIR images – diffuse high signal areas in the genu, body and splenium of corpus callosum (Sandwich sign). On T1W images – hypo intensity noted at the same areas. On diffusion weighted images (DWI) - these areas showed restricted diffusion.

On the basis of history, clinical features and imaging studies the diagnosis of acute form of MBD was made. Patient was treated with vitamin B complex, thiamine & other required nutritional diet during hospitalization. At discharge he showed improvement in his consciousness and there was residual mild aphasia without cranial nerve involvement.

DISCUSSION: Although first described by Carducci (1898) in Italian red wine drinkers, the Italian pathologists Marchiafava and Bignami (1903) described a unique alteration of the corpus callosum in three alcoholic patients who died after having seizures and coma.^[1] The disease affects persons in middle and late adult life.

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With a few exceptions, the patients have been males and severe chronic alcoholics^[2]. The underlying mechanism of the disease is still not understood. It is probably caused by the combination of alcohol abuse and malnutrition, leading to metabolic, toxic and vascular disturbances.^[3] There are no characteristic clinical presentations of Marchiafava-Bignami disease. Clinical clues for the disease are reduced consciousness, psychotic and emotional symptoms, depression and apathy, aggression, seizures, hemiparesis, ataxia, apraxia and frequently leading to coma and death.^[4]

The course of the disease may be acute, sub-acute or chronic and may lead to death within weeks to months. Marchiafava-Bignami disease may present in various clinical forms^[5, 6]. Acute Marchiafava-Bignami disease includes seizures, impairment of consciousness, and rapid death. Subacute Marchiafava-Bignami disease includes variable degrees of mental confusion, dysarthria, behavioral abnormalities, memory deficits, signs of interhemispheric disconnection, and impairment of gait.

Chronic Marchiafava-Bignami disease, which is less common, is characterized by mild dementia that is progressive over years. Until recently, the definite diagnosis was confirmed at autopsy. However, in the era of modern imaging technology, diagnosis could be based on clinical profiles, history of alcoholism, and specific localizations of pathological lesions in the corpus callosum demonstrated by CT and MRI.^[3] The corpus callosum appears hypo attenuated on CT scans.

MRI is currently the most sensitive diagnostic tool. The entire corpus callosum is rarely involved. Similar lesions may also be found in the middle cerebellar peduncles and in the hemispheric white matter involving the centrum semi ovale and extending, in some cases, into the adjacent white matter.

These lesions usually do not have mass effect and may show peripheral contrast enhancement during the acute phase^[7, 8]. As lesions become chronic, cystic lesions are likely to develop. Pathology may also be seen on diffusion-weighted imaging and ADC as areas of restricted diffusion. Greater the restriction worst the prognosis^[7].

After a few months, signal intensity alterations become less evident but residual atrophy of the involved structure usually is present^[9]. The diagnosis of MBD rests mainly on evidence of these callosal lesions. The corpus callosum may also be affected in other diseases such as ischemic stroke, contusion, multiple sclerosis, and lymphoma. MBD, however, is distinguished from these disorders by the symmetry of the callosal lesions with relative sparing of thin upper and lower edges.

Other neuropsychiatric conditions associated with chronic heavy drinking are: Wernicke's encephalopathy, Korsakoffs psychosis, alcoholic dementia, cerebellar degeneration, central pontine myelinolysis, alcoholic amblyopia, alcoholic pellagra encephalopathy and peripheral neuropathy. Because the etiology of the disease is uncertain, a specific therapy is not available. Cessation of alcohol intake is mandatory.

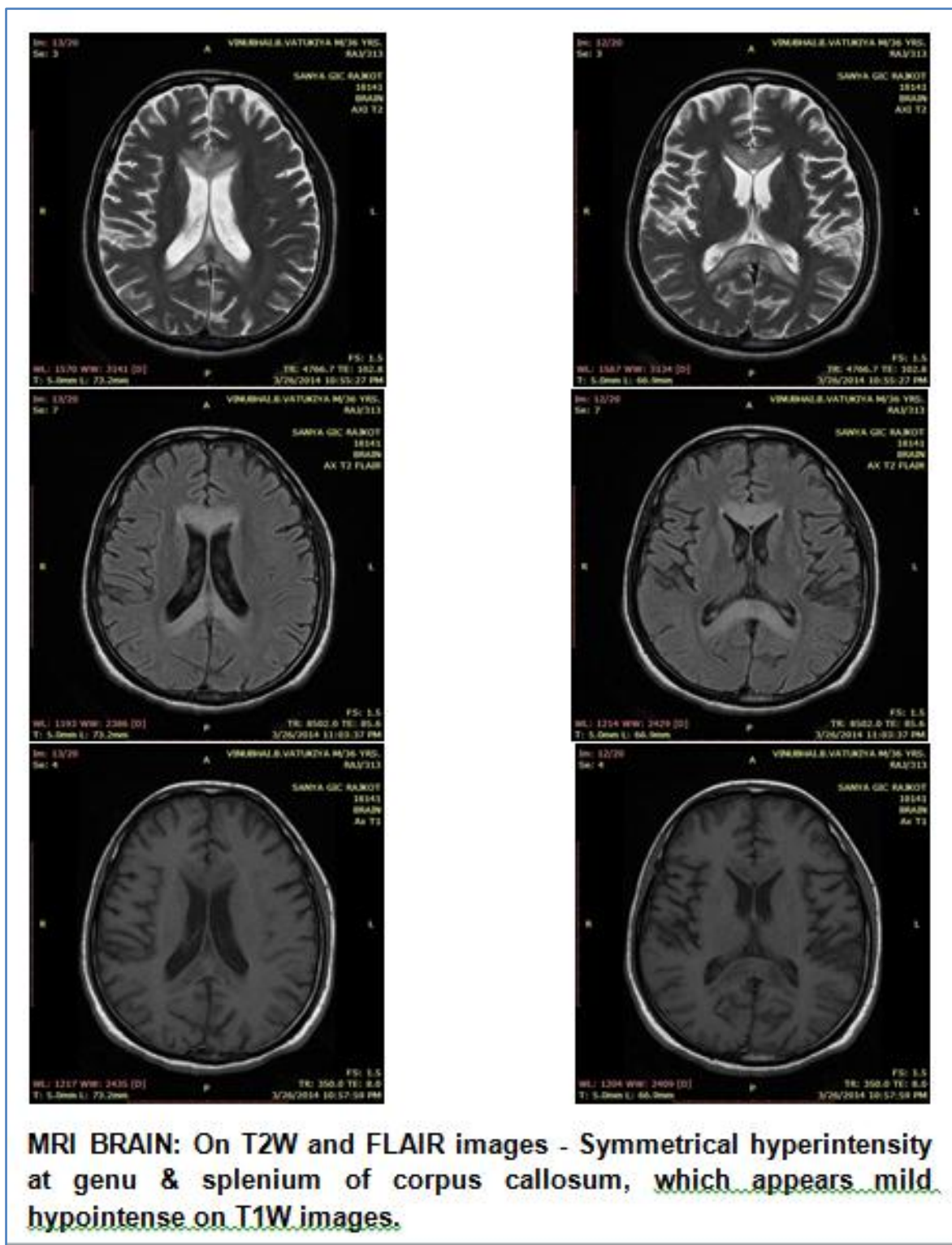
Therapy with thiamine and vitamin B complex, including vitamin B-12 and folate, has been used in many patients who have recovered. However, identical therapy has been used in patients who did not recover. Seizures and coma are treated symptomatically. A favorable response in a demented condition or even partial or complete recovery has been reported after the use of corticosteroids in such patients^[3]. Patients who survive should stop alcohol consumption, receive rehabilitation and nutritional counseling.

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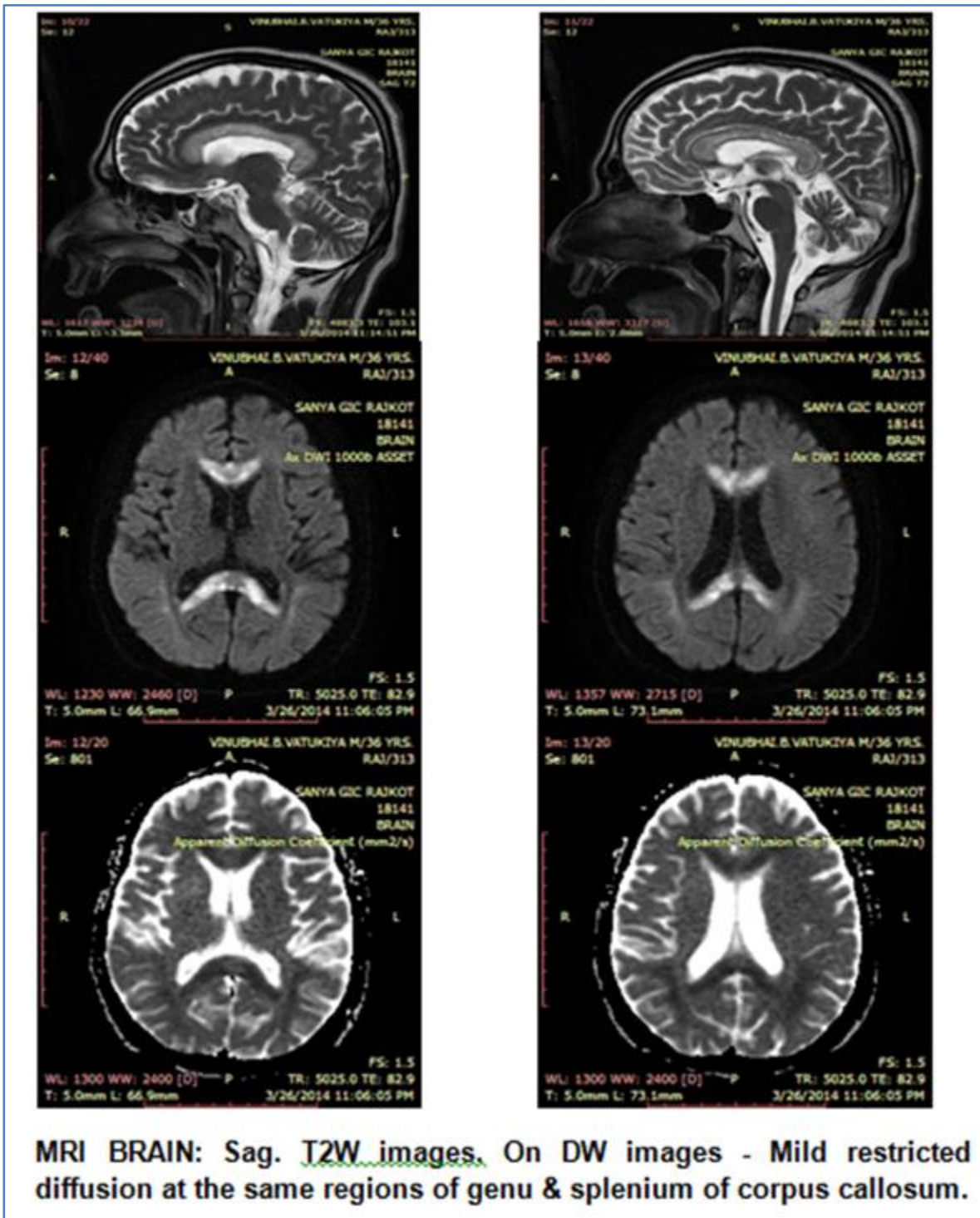
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Date of Submission: 07/08/2014.
Date of Peer Review: 08/08/2014.
Date of Acceptance: 21/08/2014.
Date of Publishing: 02/09/2014.