Marchiafava–Bignami Disease Presenting as Acute Psychosis

Sir,

Marchiafava–Bignami disease (MBD) is a rare neurological disorder of unknown etiology, afflicting mostly middle or elderly males with a history of chronic alcohol consumption or chronic malnutrition.[1-3] There are <300 described cases in the literature and many cases go undiagnosed.[4] Its pathological hallmark is symmetrical demyelination and necrosis of the central part of the corpus callosum.[5] The disease has varied neuropsychiatric manifestations which include agitation, confusion, delirium, rigidity, dysarthria, mutism, split-brain syndrome, incontinence, dementia, psychotic symptoms, and gaze palsy.[6,7]

We report a rare presentation of MBD in a 32-year-old male having a history of heavy alcohol consumption who presented with persecutory delusion, auditory hallucinations, dysarthria, and urinary incontinence and whose psychotic symptoms responded to olanzapine.

CASE REPORT

A 32-year-old unmarried Indian male presented with complaints of suspiciousness, decreased sleep, and violent behavior for 15 days and episodes of urinary incontinence with difficulty in speaking for 10 days. He feared being killed and poisoned, so he confined himself to his room and refused to eat. The patient had a history of heavy alcohol consumption since the age of 15 (750 ml whiskey daily, 42% alcohol by volume). There was no significant medical or psychiatric history and no significant family history. Two months before the presentation, the patient was admitted for alcohol detoxification and he was abstinent since then. On examination, he was malnourished, disheveled, verbally abusive, and anxious. Persecutory delusions and auditory hallucinations were present. He was conscious, alert, and oriented to time, place, and person. There were no signs of meningeal irritation, and the vitals were stable. Neurological evaluation revealed dysarthria and hypertonicity of limbs. He had deranged liver transaminase enzymes (aspartate transaminase – 143.2 IU/L, alanine transaminase – 65.7 IU/L), macrocytosis (MCV – 108 fL), reduced serum Vitamin B1 level, and Grade 1 fatty liver.

Neurology consultation was sought, and a noncontrast computerized tomographic scan of the brain revealed hypodensity in the region of the corpus callosum. Magnetic resonance imaging scan showed a T2-hyperintense lesion, with restricted diffusion of the corpus callosum, without swelling or enhancement. A diagnosis of MBD was made. He was started on tablet olanzapine 5 mg twice daily, 300 mg oral thiamine, 5 mg of oral folic acid, and tablet lorazepam 2 mg at night.

After 4 days, the patient was no longer violent, sleep was improved, and delusion and hallucination subsided. However, dysarthria, episodes of urinary incontinence, and hypertonicity of limbs persisted. Over the next 10 days, the patient showed improvement, tablet lorazepam was stopped, and olanzapine was reduced to 5 mg once daily at night. Subsequently, he was discharged and followed up for 6 months, during which he was maintaining well on 5 mg olanzapine once daily and 300 mg oral thiamine and had no psychotic phenomena, but his neurological symptoms persisted.

DISCUSSION

MBD is a rare disorder generally associated with chronic alcohol consumption and has a varying clinical course. Its acute form can present with impaired consciousness, seizure, and coma while its subacute form presents with mental confusion, emotional distress, behavioral abnormality, and psychosis.[5] Augusto *et al*.[8] reported visual hallucinations in a 52-year-old woman with chronic alcoholism and MBD, and Hui *et al*.[9] described, in a 41-year-old man, persecutory delusion and auditory hallucinations which did not respond to 15 mg of haloperidol or 6 mg of risperidone. Our patient had a relatively early onset of MBD. His clinical presentation was dominated by the presence of psychotic phenomena which improved considerably on 10 mg of olanzapine and was well maintained on 5 mg of olanzapine in the 6-month follow-up. He tolerated olanzapine without significant adverse effects. Olanzapine has been shown to be particularly useful in psychosis with comorbid substance abuse, and these patients have better outcomes and low risk of extrapyramidal adverse effects.[10]

Since early recognition and treatment may provide a favorable outcome, MBD should be included in the differential diagnosis of patients with heavy alcohol consumption presenting with psychotic phenomena. Although there is no established treatment of MBD, the use of thiamine and folic acid is recommended.[5] While earlier studies have questioned the role of antipsychotics in managing the psychotic phenomena in MBD, our patient benefited considerably, possibly because of early detection.

Declaration of patient consent

The authors certify that they have obtained patient consent form. In the form, the patient has given his consent for all of his clinical information to be reported in the journal. The patient understands that his name and initials will not be published and due efforts will be made to conceal his identity, but anonymity can not be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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