CASE REPORT

A RARE CASE OF SUB-ACUTE FORM OF MARCHIAFAVA-BIGNAMI DISEASE PRESENTING PREDOMINANTLY WITH PSYCHOTIC SYMPTOMS

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Abstract

Objective: The objective is to present a rare case of sub-acute form of Marchiafava-Bignami disease presenting with psychosis, minimal cognitive impairment and no clinical neurological signs. Methods: This is a case report of a patient with Marchiafava-Bignami disease presenting to a tertiary care psychiatric hospital in Singapore. A review of the literature of the condition is also presented. Results: The patient presented with prominent psychotic symptoms in the context of chronic alcohol abuse. He also had minimal cognitive impairment and clinically no neurological signs. A working diagnosis of alcohol induced psychotic disorder was made. His psychotic symptoms seemed to be resistant to treatment with antipsychotic medications initially and this led to further investigation by MRI scan of brain which revealed atrophy of corpus callosum and no other significant abnormalities. His psychotic symptoms gradually improved with treatment. He is now placed in an intermediate care setting in the hospital while waiting for a suitable community placement. Conclusions: It is rare for Marchiafava-Bignami disease to present with prominent psychotic symptoms with minimal cognitive impairment and no neurological signs clinically. In patients with a history of chronic alcohol abuse presenting with psychotic disorder, Marchiafava-Bignami disease should be suspected as a diagnostic possibility. ASEAN Journal of Psychiatry, Vol. 16 (2): July – December 2015: XXXX.

Keywords: Chronic Alcohol Abuse, Marchiafava-Bignami Disease, Psychotic Symptoms

Introduction

Prevalence of alcohol abuse is common in many societies. Chronic alcohol abuse is associated with multiple systemic complications including neuropsychiatric syndromes. The neurological sequelae of chronic alcohol abuse include peripheral neuropathy, cerebellar degeneration, Wernicke’s encephalopathy, Korsakoff’s syndrome, central pontine myelinosis, alcoholic pellagra and Marchiafava-Bignami disease [1].

Marchiafava-Bignami disease (MBD) is a rare complication of chronic alcohol abuse and its pathogenesis is not clearly understood. It is probably caused by the combination of alcohol abuse and malnutrition and pathologically characterised by corpus callosum demyelination and necrosis [2]. The clinical features of the condition are non-specific and range from reduced consciousness, seizures,
impaired gait, apathy, behavioural changes and cognitive impairment. Psychotic symptoms have been reported in the condition although this is rare. As the clinical features of Marchiafava-Bignami disease are variable, it remains primarily a radiological diagnosis [3]. Here we report a case of Marchiafava-Bignami disease in an individual with chronic alcohol abuse presenting predominantly with psychotic symptoms and no clinical neurological symptoms or signs.

Case Report

A 41 year old male was brought to the emergency services in our tertiary care psychiatric hospital by his family. He presented with increasingly disorganised behaviour, including wandering aimlessly over several weeks, which later escalated to his burning of bed sheets at home, culminating in his first admission to the hospital. On initial assessment it became apparent that such change in behaviour was in the context of psychotic symptoms of the same duration. These symptoms took the form of nihilistic and persecutory delusions as well as auditory hallucinations.

It emerged during the course of this admission that our patient had misused alcohol in excess of twenty five years. He typically drank liquor daily, amounting to an average of 210 units weekly. There was sparse evidence for any periods of abstinence much in excess of several weeks at any time, and overwhelming support for other indicators of dependency including primacy of alcohol use, cravings and withdrawal symptoms. There was no suggestion of previous illicit substance use.

On this occasion, a working diagnosis of alcohol-induced psychotic disorder was made, and he responded well to low dose haloperidol. He was discharged well within one week of admission.

He was readmitted within a couple of weeks, with similar symptoms albeit on this occasion with additional findings of disorientation for time only, as well as patchy amnesia for a two day period when he had reportedly wandered away from home. He had resumed his pattern of significant use of alcohol. His behaviour had become disorganised once again, such as throwing clothes away, also with a re-emergence of the same psychotic symptoms as described.

On mental state examination, he was noted to be poorly kempt. Other findings were normal with the exception of psychotic symptoms as reported and evidence for cognitive impairment without confabulation. The mini-mental state examination indicated disorientation for time, poor attention and concentration, but no difficulties with registration or recall were then evident. Other bed-side cognitive testing did not indicate any other cognitive difficulties except for categorical fluency. Despite his significant use of alcohol over many years, laboratory investigations were noted to be essentially normal with the exception of mildly deranged liver transaminase enzymes and macrocytosis. Furthermore, physical examination, inclusive of a full neurological evaluation, was noted to be entirely normal. The haloperidol was re-instated and gradually titrated to 15 mg daily with only modest effect. This was therefore cross-tapered with an alternative antipsychotic medication, namely risperidone, which was subsequently titrated to 6 mg daily. At that time, he was also prescribed multivitamins, vitamin B complex, as well as additional thiamine 250 mg daily and folic acid 10 mg daily. His symptoms showed very minimal response to risperidone after adequate duration of treatment with the medication. In view of our patient’s minimal response to an adequate trial of antipsychotic medication, a Magnetic Resonance Imaging (MRI) brain scan was arranged. This investigation revealed mild atrophy of the corpus callosum.

Discussion

The first case of MBD was described in 1898 by Carducci, in an alcoholic man who developed sudden seizures, loss of consciousness, coma and eventual death. Autopsy findings in this case revealed necrosis of corpus callosum. In 1903, Italian pathologists Marchiafava and Bignami reported similar findings in an autopsy series of three Italian wine drinkers who were found to have necrosis and cystic degeneration involving the middle layer of the corpus callosum.
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callosum. Since then, over 200 cases of MBD have been reported in the literature [4]. The incidence may be higher than generally believed [5].

As mentioned earlier, the clinical features of MBD may present with varying clinical course. In the acute form, MBD presents with seizures, severe impairment of consciousness, coma and death, the sub acute form presents with ataxia, dysarthria, memory deficits and behavioural abnormalities. The chronic form presents with progressive dementia [6].

In the case we are reporting, the patient presented with psychotic symptoms in the form of delusions and auditory hallucinations, odd behaviour associated with dysphoric mood in the context of chronic heavy alcohol abuse. He did not appear to be in alcohol withdrawal or delirium at the time of admission. His Mini Mental State Examination (MMSE) did not show significant cognitive impairment as the score fluctuated between 23 and 25 out of 30 in the test. A wide range of differential diagnoses were considered initially in view of late onset psychotic symptoms. An unremarkable clinical neurological examination excluded the possibility of central nervous system lesions such as tumours, infection and vascular events contributing to the clinical presentation. It was noted our patient’s age and overall presentation, and in particular the recent cognitive impairment described, would not be typical for first onset of a functional psychotic disorder namely schizophrenia. In addition there was a lack of family history for such.

Alcohol withdrawal syndrome or Wernicke’s encephalopathy was unlikely in view of the absence of withdrawal symptoms or neurological signs such as ataxia or ophthalmoplegia. With regards to his progress, although there was slight improvement in the beginning, his psychotic symptoms persisted despite adequate treatment with antipsychotic medications. The minimal response to treatment led up to further investigation by MRI scan of the brain which, as mentioned earlier, revealed multiple focal areas of high T2 and low T1 signal involving the genu, anterior body and splenium of the corpus callosum. This was associated with mild atrophy of the corpus callosum (Figure 1). Multiple focal areas of high T2 and low T1 signal are seen involving the genu, anterior body and splenium of the corpus callosum associated with atrophy (thinning) of the corpus callosum in these areas.

Figure 1. MRI of Brain – sagittal view, revealed multiple focal areas of high T2 and low T1 signal involving the genu, anterior body and splenium of the corpus callosum
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There were no other abnormalities reported in the scan. In view of the radiological findings of corpus callosum atrophy the diagnosis was revised to Marchiafava-Bignami disease which was congruent with the clinical presentation. Psychosis may occur in Marchiafava-Bignami disease in the context of delirium in the acute form and dementia in the chronic form [7]. However, this presentation of sub-acute form with predominant psychotic symptoms as in our case is rare.

He was continued on risperidone 6 mg daily and he made slow progress in his psychotic symptoms and he was transferred to an intermediate care setting to continue his progress. A recent clinical review of the patient showed no psychotic symptoms. There was no further decline in his cognitive impairment. The family has agreed to take him home on trial leave and the success of which would help us plan his discharge from the hospital.

Our case highlights the rare presentation of a sub-acute form of MBD with prominent psychotic symptoms, minimal cognitive impairment and absence of neurological signs. MBD should be suspected in patients presenting with alcohol related psychotic disorder with persisting psychotic symptoms.

References


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