Case Report

Cognitive Dysfunction associated with Marchiafava Bignami Disease treated with Cerebroprotein Hydrolysate

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ABSTRACT

Marchiafava-Bignami disease (MBD), also known as Bignami syndrome, is a rare neurological disorder primarily characterized by degeneration of the corpus callosum, a structure that connects the two hemispheres of the brain. It is most often associated with chronic alcohol abuse and malnutrition. A variety of psychiatric and neurological disorders. We report a case of the disorder that responded to cerebroprotein hydrolysate.
Keywords: corpus callosum, Marchiafava-Bignami disease, alcohol abuse.

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INTRODUCTION

Marchiafava-Bignami disease (MBD) is characterized by primary degeneration of the corpus callosum associated with chronic alcohol consumption [1]. However, it can also occur in patients who do not use alcohol [2]. The main hypothesis for its pathogenesis is that the disease is a result of B vitamin deficiency. Although many patients may improve after administration of vitamin B, others do not. In these cases, morbidity and mortality are relatively high [3], as of 2004, approximately 250 patients had been described, 200 had died, 30 suffered from severe dementia, and only 20 experienced positive outcomes. It seems that with alcoholism, the prognosis is worse. Most patients are male, are between 40 and 60 years of age, and have a history of chronic alcoholism and malnutrition [1-2]. The diagnosis is difficult and generally is a result of neuroimaging, particularly magnetic resonance imaging (MRI) [4-5]. The treatment of MBD is still controversial and shows variable results. The implication of nutritional factors, like Wernicke's encephalopathy, suggests that replacement of B vitamins is beneficial. However, with the variable response to this type of therapy, corticosteroids and amantadine have been proposed by some authors [6-7].

CASE REPORT

A 45-year-old Hindu, right handed, educated till 5th standard, plumber ,married male had presented to our outpatient department with complaints of sleep disturbances, tremulousness of hands, suspiciousness, fearfulness and abusive and aggressive behaviour since 2 days. As per the informant, patient has been consuming alcohol for 20-25 years. But in the last 10-15 years he has been drinking in daily pattern and would consume around 3-5 litres of country liquor per day. He spends almost 50 percent of his income in buying alcohol. It was noted that he spends most of his time procuring, consuming, and recovering from alcohol use. Whenever he is unable to consume alcohol he develops physical withdrawal symptoms in the

form of sleep disturbances, tremulousness of hands, fearfulness. Patient had frequent fights with his wife and use to get irritable towards children on trivial issues.

Around 2 days back he had his last drink of 90 ml of country liquor since then behavioural disturbances in the form of irritability, abusive and aggressive behaviour towards family members, suspiciousness that people are coming to harm him and could ask family members to close the doors. Hence patient was brought for further management. As patient was not manageable by family members was admitted in psychiatric ward.

There was no history suggestive of any other psychopathology. There was no history suggestive of any medical/surgical comorbidity or family history of psychiatric illness. On examination of mental status, the patient was ill-kempt and unshaven, conscious, not cooperative and non-communicative. Patient was physically restrained in all four limbs, getting irritable and trying to remove restraints. Eye to eye contact was not initiated, rapport was not established, and affect was perplexed. A diagnosis of Alcohol withdrawal was made per International Classification of disease; eleventh revision and the patient were started on tablet Lorazepam 10 mg and Inj. thiamine 300 mg along with Inj. lorazepam 4mg SOS.

Patient exhibited improvement in sleep disturbances, suspiciousness, aggressive and abusive behaviour. But on serial MSE it was noted that patient had difficulty communicating. Patient was only able to tell his name and would identify his family members by pointing figure at them but could not recall their names, along with poverty of speech and poor registration and recall in immediate memory as well as recent and remote memory. Even after 10 days patient had minimal improvement, his MMSE scores were 5-6 over the period and did not improve. Hence MRI was done for organic causes, and MRI was suggestive of few patchy T2, FLAIR hyperintense and T1 hypointense signal intensity noted involving genu, body and splenium of corpus callosum predominantly involving its central layer with relative sparing of dorsal and ventral layers with patchy areas of restricted diffusion along genu and splenium. Features were suggestive of demyelinating aetiology like Marchiafava-Bignami disease.

Following which neurology reference was taken and patient was started with inj. Thiamine 500 mg TDS. But patient had minimal improvement in symptoms. Hence a trial of Cerebroprotein hydrolysate was decided to be given after relatives' consent. Hence patient was started with Inj. Cerebroprotein hydrolysate 90 mg after sensitivity testing and was given consecutively for 20 days. Patient as well as family members reported about 50 percent improvement in memory disturbances.

DISCUSSION

There is no prototypical clinical presentation of MBD. Subtle clinical signs such as reduced consciousness, emotional and psychotic symptoms, depression and apathy, aggression, seizures, hemiparesis, ataxia, and apraxia may be present as initial manifestations [4]. However, their development can lead to coma and death [4, 8-9]. It is worth noting that both acute and subacute MBD can produce an extensive list of neurological manifestations that are frequently observed in the ICU. The chronic form, which is less common, is characterized by mild dementia, but it can be progressive. Until recently, a definitive diagnosis of MBD could only be made at autopsy. However, in the era of modern neuroimaging, it is possible to confirm the diagnosis based on a typical clinical profile with a history of alcoholism and a CT scan or MRI demonstrating specific pathological lesions in the corpus callosum [4].

As the aetiology of this disease remains uncertain, specific and well-defined therapy is not available. Treatment with thiamine and other B vitamins, including vitamin B-12 and folic acid, has been used in many patients who have recovered. However, therapeutic failure is not uncommon, even if treatment is started at the beginning of symptoms [10]. Some patients who survive acute or subacute MBD have subsequent dementia, but partial or complete recovery is occasionally observed.

Use of Cerebroprotein has been tried in cases of dementia. Cerebroprotein hydrolysate is a newer pharmacological agent which is reported to be neurotrophic in nature and manufactured synthetically by the standardized enzymatic breakdown of lipid-free porcine brain proteins [11]. It has reported that cerebroprotein via certain mechanisms enhances neurogenesis, neuronal survival, and neuronal plasticity while also having a neuro-immunotrophic mechanism of action [12]. There are anecdotal case reports in the

literature where cerebroprotein has been reported to be of some benefit in traumatic brain injury [13], certain forms of dementias [8] and extrapontine myelinosis [14].

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