A Case of Marchiafava-Bignami Syndrome

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ABSTRACT

Drinking alcohol is a serious social and health problem in our country and throughout the world. It causes varied diseases in different organs of the body, which many do not care to know about. Marchiafava-Bignami disease is one such rare but serious disorder. We are presenting this case to show the serious disorders associated with alcoholism.

Keywords: Corpus callosum, alcohol, mutism, alcohol dehydrogenase, neurological deficit

Marchiafava-Bignami syndrome is one of the central nervous system (CNS) complications of chronic alcohol intake. There is degeneration of the corpus callosum similar to a disconnection syndrome. It is possibly due to B complex deficiencies related to chronic alcohol intake. The prognosis is guarded as most patients present with coma.

CASE REPORT

A 50-year-old male, chronic alcoholic, laborer was admitted to Guru Gobind Singh Hospital, Jamnagar, Gujarat. He presented with fever, vomiting, altered mental status, disorientation to time-place-person and first episode of generalized tonic-clonic convulsions.

On examination, he presented with mutism, altered sensorium, no ophthalmoplegia, no nystagmus, inability to move all limbs, bilateral extensor plantars with semi-dilated reactive pupils bilaterally. Neck stiffness was not present. No focal neurological signs were present. Fundus examination showed no signs of papilledema. A clinical diagnosis of meningoencephalitis or hepatic encephalopathy was made. Meanwhile, contrast-enhanced computed tomography (CECT) brain was done to look for secondary causes of convulsions. Magnetic resonance imaging (MRI) cuts were taken in addition to CT which showed "Ill-defined non-enhancing hypodense area in middle part of corpus callosum, possibly degeneration due to Marchiafava-Bignami syndrome" (Fig. 1 a and b). Hence, diagnosis of "Marchiafava-Bignami syndrome" was made. Patient received injections (cefotaxime, metronidazole, mannitol, thiamine, vitamin B_{12}, hydrocortisone, sodium valproate, IV glucose, amino acids, soybean oil and lecithin) and supportive treatment. On Day 3, he regained consciousness. On Day 4, he became oriented to surroundings. Later he was able to walk with support.

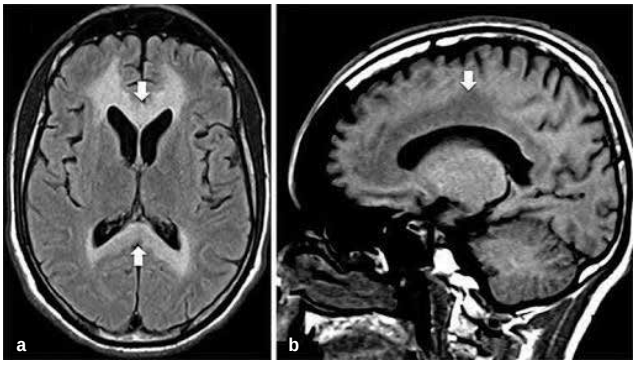

Figure 1. High signal intensity noted in axial FLAIR image with involvement of anterior and posterior portion of corpus callosum and periventricular white matter (**a**). Sagittal T1-weighted image with low signal intensity seen in entire length of corpus callosum shown with arrow (**b**).

DISCUSSION

Marchiafava-Bignami disease is a rare and toxic encephalopathy seen mostly in alcoholics due to progressive demyelination and necrosis of corpus callosum, which may extend to adjacent regions and even up to subcortex. It was first described by Italian

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pathologists, Marchiafava and Bignami, in alcoholic patients who died after having seizures and coma. Most accepted cause for the pathogenesis of this disorder is deficiency of multiple vitamins of vitamin B complex. It can be divided into two subgroups as type A having altered consciousness, stupor, coma and upper motor neuronal signs with involvement of entire corpus callosum and type B having mild impairment of consciousness with small callosum lesion. Type B has good prognosis. Altered mental status, impaired walking and loss of consciousness is seen in more than 50% of patients with this disease.

Other symptoms are dysarthria, impaired memory, signs of disconnection, pyramidal signs, seizures, primitive reflexes, rigidity, hemi-/tetraparesis, incontinence, nystagmus, etc. Mutism, sensory symptoms and gaze palsy are seen in only 10% of patients, but when present, can be useful in differentiating this syndrome from other disorders. Hence, they are more specific for this disease. Diagnosis of this disease is made by history, presentation, examination and radio-imaging modalities such as CT scan and more preferably MRI brain, which shows hypodense lesion due to degeneration in corpus callosum. No specific treatment is available but treatment with parenteral thiamine within 15 days of presentation for 15 days along with other vitamins, proteins and high dose steroids shows better prognosis as compared to controls in previous studies. Antiparkinsonian drug amantadine showed improvement as per one study. Rest and supportive treatment should be given. Prognosis of this disease varies as some patients improve and some deteriorate to even death in spite of treatment.

Certain risk factors predispose to alcohol toxicity:

- Pattern of drinking - more duration means more toxicity; continuous drinking is more dangerous than intermittent; drinking more than 80 g/day is dangerous.
- Women achieve higher blood levels of alcohol than men.
- Certain genes namely HLA-BB and ADH gene 2 are considered to increase alcohol toxicity.
- On the other hand, presence of food in the stomach, especially proteins, decreases alcohol toxicity.

Other than Marchiafava-Bignami syndrome, alcohol can cause other CNS effects - acute intoxication (from euphoria to coma), withdrawal syndrome, alcoholic dementia, cerebrovascular accidents, alcoholic cerebellar degeneration, central pontine myelinolysis, peripheral neuropathy, Saturday night palsy, etc. Other than effects on the brain, alcohol has toxic effects on liver, blood cells, fetus, gastrointestinal tract, cardiovascular system, genitourinary system, skeletal system, endocrine system, respiratory system, etc. Addiction, social and psychological problems are also threats associated with alcohol intake. In addition, alcohol interacts with a lot of commonly used medications. Alcohol toxicity varies from gastric ulcers, which is easily treatable, to even malignancies which have serious prognosis.

In our case, the patient presented in a coma like state. On examination, it was not a deep coma but patient had altered sensorium with mutism. Hence, there was no verbal output. Mutism is one of the rare presentations in this disorder. There was significant improvement with parenteral vitamin replacement although the prognosis is not good in most cases.

CONCLUSION

This case has been presented to highlight the various serious CNS complications in chronic alcoholics. Because it is uncommon, it is rarely diagnosed clinically. This disease should be kept in the mind in chronic alcoholics presenting with coma. Thankfully, our patient recovered in spite of the guarded prognosis of the disease.

"Think before you drink, because if you drink, you may not be able to think again." (Alcoholism can cause brain degeneration)

"As there is no intervention, start with prevention."

SUGGESTED READING

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