Central Pontine/Extrapontine Myelinolysis Presenting with Manic and Catatonic Symptoms

Sir,

Central pontine myelinolysis (CPM) is a neurological disorder associated with demyelinating lesions in the central pons. It is usually caused by electrolytic disorders, especially rapid correction of severe hyponatremia.[1] Systemic disorders such as chronic alcoholism, hepatic failure, severe burns, malignant neoplasms, and hemodialysis also predispose to this condition. It may coexist with extrapontine myelinolysis (EPM), where it involves lesions outside pons, i.e., in caudate and lentiform nuclei, putamen, thalami, cerebellum, hippocampus, and cerebral cortex. It often presents with dysphagia, dysarthria, quadriplegia, encephalopathy, or coma. Such cases may additionally develop tremors, dystonia, cogwheel rigidity, ataxia, mutism, myoclonus, etc. Besides neurological symptoms, CPM may also present with neuropsychiatric symptoms such as personality changes, inappropriate affect, emotional lability, disinhibition, catatonia, psychosis, and delirium, as described in some reports.[2-4] None of the previous reports have suggested an occurrence of prominent manic symptoms followed by catatonia in patients with central pontine/extrapontine myelinolysis (CPEM). We describe a unique case of an elderly man with such a presentation after rapidly corrected hyponatremia.

CASE REPORT

A 72-year-old man was admitted to the cardiothoracic surgery unit floor for aortic valve repair and coronary artery bypass graft. He had history of hypertension and coronary artery disease along with rheumatic aortic stenosis for the last 30 years. He had no psychiatric illness and denied any substance use in the past. During preoperative workup, his serum sodium was found to be low (114 mmol/L), but he was asymptomatic. Serum sodium levels repeated over the next couple of days remained unchanged. During surgery, his serum sodium level decreased to 104 mmol/L. As per intraoperative notes, serum sodium was rapidly corrected with intravenous 3% saline. His serum sodium levels were 134 and 139 mmol/L, respectively, on postoperative days 1 and 2. His postoperative course was uneventful from the cardiac surgery perspective. However, on

day 3, he developed manic symptoms in the form of persistently elated mood, decreased need for sleep, perceived increase in physical energy, loud and excessive speech, authoritativeness, sexual disinhibition, and grandiose and persecutory ideas.

A psychiatric consultation was sought for his behavioral problems. On mental status examination, he was alert and fully oriented and had elated mood, grandiose and persecutory ideas, impaired judgment, and absent insight. He denied any hallucinatory experiences. There were no apparent neurological deficits. Oral haloperidol 2 mg/day was started, and his manic symptoms began to improve within the next 2 days. By postoperative day 6, manic symptoms remitted and he was discharged.

He remained well for the next 2 days, after which his family brought him to the emergency department because of decreased food intake and decreased spontaneous movements. He was found to have dysphagia and dysarthria. On examination, he had rigidity and brisk reflexes in all four limbs, along with staring, posturing, and gegenhalten. Although some of the findings (rigidity, brisk reflexes, dysphagia, dysarthria) could be ascribed to pyramidal/extrapyramidal involvement, motor behaviors such as staring, posturing, and gegenhalten were characteristic of catatonia.

We repeated hemogram, serum chemistry, urine routine/microscopy analysis, and serum creatine phosphokinase levels, none of which showed any abnormality. We also performed magnetic resonance imaging (MRI) brain, which showed [Figure 1] central hyperintensities in the pons, bilateral caudate, and lentiform nuclei, as well as the thalami on axial T2-weighted sequence. Therefore, based on the overall history, examination, and neuroimaging findings, we made a diagnosis of CPEM secondary to rapid correction of hyponatremia. Haloperidol was discontinued. He was admitted to Geriatric Medicine unit, managed conservatively with fluid and nutrition management, and given oral lorazepam 1 mg twice a day for catatonic symptoms. He showed mild improvement in the catatonic symptoms over the next 5 days, was able to walk with assistance, and was able to speak and feed himself. Further lorazepam dose escalation could not be done as his family got him discharged due to logistical issues, with a plan to follow-up in the outpatient clinic. Repeat MRI brain could not be done for the above reason as well.

DISCUSSION

In the present case, neuroleptic malignant syndrome (NMS) was the single most obvious differential considered, based on the history of haloperidol use and occurrence of acute-onset generalized rigidity. NMS is characterized by fever, altered mental status, autonomic instability, rigidity, increased serum creatine phosphokinase (CPK) level, leukocytosis, etc. However, it was ruled out clinically because our patient was afebrile and had clear sensorium, no evidence of autonomic instability, and normal complete blood count and serum CPK level. A diagnosis of CPEM was quite evident, with a typical history of rapid correction of low serum sodium, leading to the occurrence of manic symptoms followed by neurological as well as catatonic symptoms along with the development of characteristic brain changes. We ascribe CPEM as the main cause of manic symptoms and catatonia here, based on other factors such as temporal association, the age of presentation, and no past or significant family history of psychiatric illness. Patient's poor outcome could be attributed to the extremely low serum sodium before it was rapidly corrected.[5]

So far, to the best of our knowledge, there has been only one report of manic symptoms occurring in association with CPEM. According to Goggin *et al.*, a patient initially developed delirium that evolved into mania. In that case, the brain lesions were limited to central pons only.[6] In the present case, catatonia emerged later, which has been described in only a few of the earlier reports.[7,8] It was observed that the patient's neurological and neuropsychiatric symptoms were well correlated with the brain lesions observed in MRI. The patient was found to have demyelination in bilateral caudate, lentiform nuclei, and thalami, which could potentially disrupt striatal and thalamic functioning. Frontal–striatal–thalamic circuit (FST) is implicated in regulating mood, reward processing, action selection, strategic planning, and working memory. Previous studies have shown that alteration in the functioning of the striatal–thalamic circuit is implicated in bipolar disorder.[9] Bipolar disorder is also well associated

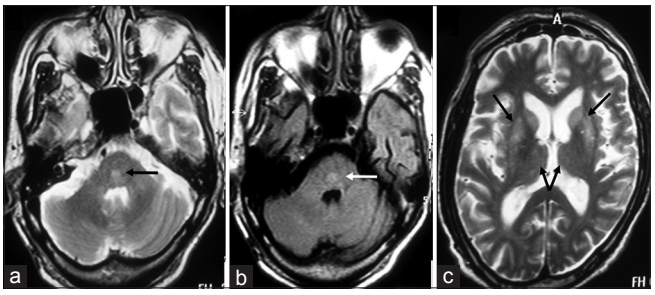

Figure 1: (a-c) There is central hyperintensity in the pons (arrow) on axial T2W (a) and FLAIR (fluid attenuated inversion recovery) (b) MR images of posterior fossa. Axial T2W image at the level of basal ganglia shows hyperintensity in the bilateral caudate and lentiform nuclei, as well as thalami (arrows). Imaging findings in this particular clinical scenario are diagnostic of osmotic demyelination (both central pontine and extrapontine myelinolysis)

with catatonic symptoms[10] seen in our patient. There is a possibility that FST might be involved in the pathophysiology of catatonic symptoms too. We recommend that neuropsychiatric symptoms such as psychotic, mood, or catatonic symptoms developing in the background of rapid correction of low serum sodium should be investigated thoroughly and should not be assumed to be a part of a primary psychiatric disorder. Such rare psychiatric presentations of CPEM could provide insights into the brain areas involved in the occurrence of specific psychiatric symptoms and perhaps, mental disorders.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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