CASE REPORT

Central Pontine Myelinolysis: A Case Report

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Abstract

Central pontine myelinolysis (CPM) classically occurs due to rapid rise in serum osmolarity. Most cases have been associated with a history of chronic alcohol abuse, malnutrition, diuretic abuse, and hyponatremia. The pathological process of CPM starts in the central pons near median raphe and spreads out "like a brush Fire" into the surrounding basis pontis. Extrapontine sites such as internal capsule, basal ganglia, cerebellum, and cerebrum can also be affected. We report a case of 60-year-old male with history of chronic alcoholism who presented to us with severe neurological deficits 10 days after his episode of severe hyponatremia.

Keywords: Central pontine myelinolysis, Chronic alcoholic, Hyponatremia.

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Introduction

Central pontine myelinolysis was first described by Adams and colleagues in 1959 as a disease affecting alcoholics and malnourished.[1] It is a clinical condition characterized by rapid destruction of myelin sheath of oligodendritic cells caused by rapid rise in serum osmolarity.[2] Osmotic demyelination syndrome (ODS) usually presents as a biphasic clinical course characterized by initial phase of encephalitis with or without seizure from hyponatremia, then normonatremia is restored and this is followed by second phase of deterioration several days later.

Clinical presentation of CPM includes dysarthria, dysphagia (secondary to corticobulbar fiber involvement), flaccid quadriparesis (secondary to corticospinal track involvement), which later becomes spastic, all from involvement of basis pontis. Pupillary, oculomotor abnormalities may occur if lesion extends into tegmentum of pons. Large lesion in pontine side can present as change in consciousness level reflecting the "locked-in syndrome."[3]

The exact incidence is unknown but the outcome of CPM is very poor with the mortality rates as high as 40–50%.[3] In India, a retrospective study by Rao et al. reported vegetative state in 18% patients and mortality of 12%.[4] We report a case of 60-year-old male who was diagnosed with CPM and was rehabilitated successfully.

Case Presentation

A 60-year-old chronic alcoholic male presented to medical casualty with history of self-induced vomiting (5–6 episodes per day), cough with expectoration, hiccups, altered behavior, and dizziness since the past 3 days followed by alleged history of fall in washroom one day before. He had past history of admission for symptomatic hyponatremia (serum sodium: 123 mEq L^{-1}, serum potassium: 4.9 mEq L^{-1}); he was managed with 3% hypertonic saline. His serum sodium was 129 mEq L^{-1} within 12 hours (rate of correction was maintained to be 0.5 mEq L^{-1} per hour); he improved symptomatically and was discharged home on sixth day of hospital stay.

The patient presented to neurology department after 10 days of discharge with complain of imbalance, slurred speech, drowsiness, and fever with cough since the past 2 days. A systemic examination was remarkable which included Glasgow Coma Scale (GCS) $E_4V_1M_1$, hypertonic bilateral upper and lower limbs, hyperreflexia, bilateral extensor plantar, nystagmus, and restricted extraocular muscle movements. The patient was shifted to intensive care unit (ICU) for further management. On receiving in ICU, the patient was intubated in view of poor GCS and aspiration risk. An appropriate treatment in the form of antibiotics, antimalarials, stress ulcer prophylaxis, deep venous thrombosis prophylaxis, injection thiamine, and intravenous fluids were started.

Differential diagnosis of CPM and extrapontine myelinolysis, subdural hemorrhage, associated sepsis with delirium, hepatic encephalopathy, Wernicke–Korsakoff syndrome and autoimmune encephalitis were made. All routine blood, urine and radiological investigations were within normal limits. Magnetic resonance imaging (MRI) brain on coronal flair T2-weighted images showed hyperintensities in the central part of pons with relative sparing of peripheral pons, bilateral corticospinal tracts, and some of the transverse pontine fibers. The hyperintensities were seen to form a trident pattern in the upper pons with involvement of tegmentum of midbrain, bilateral thalamic, and some parts of globus pallidus persistent with CPM (Figs 1 and 2).

The patient received supportive management. Tracheostomy was done on seventh day of ICU stay in view of poor cough reflex and absent gag reflex. The patient was gradually weaned from ventilator and put on T-piece. He was managed with a multidisciplinary approach involving various specialties. Neurology,

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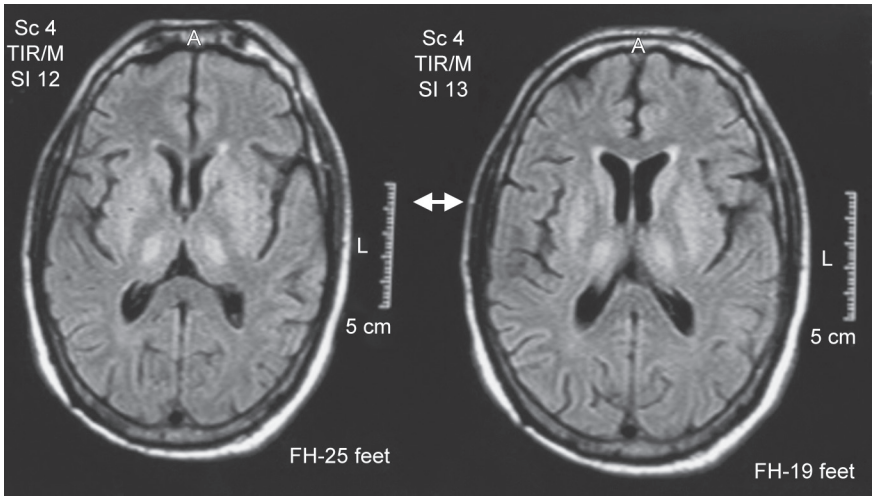

Fig. 1: Brain MRI on coronal flair T2-weighted images showing hyperintensities in thalamus and basal ganglia

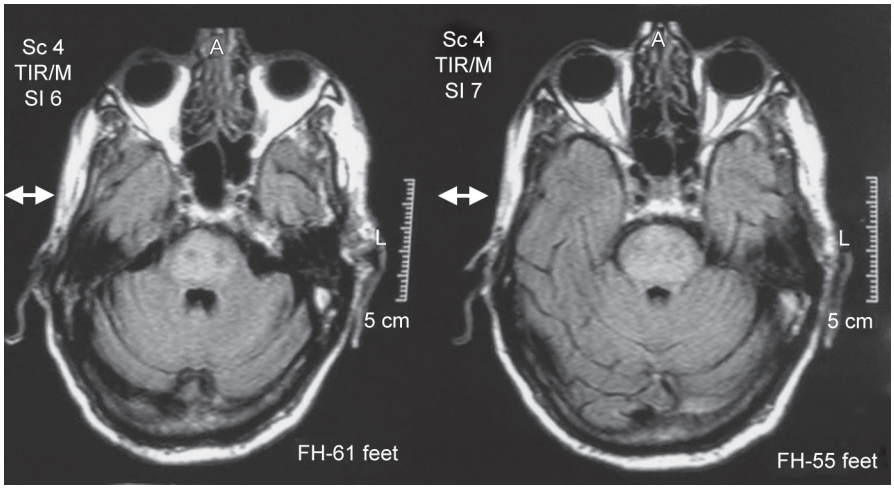

Fig. 2: The "trident pattern" in the upper pons with involvement of tegmentum of midbrain, bilateral thalamus, and relative sparing of peripheral pons and corticospinal tracts

ophthalmology, psychiatry, physical medicine rehabilitation and medicine opinions were taken and treatment was guided accordingly. During the course of ICU stay, the patient eventually became conscious oriented (GCS – $E_4V_TM_6$), all vitals were stable; rigidity and hyperreflexia persisted in all four limbs. The patient was shifted from ICU and thereafter followed up until discharge. After the continuous cycles of physical rehabilitation, the patient was able to walk with support over a period of 2 months.

Discussion

We present here a case of hypo–osmolar hyponatremia in a patient with history of alcohol dependence, complicated by CPM, despite slow correction of plasma osmolarity. The case reports of ODS following slow correction of electrolytes and serum osmolarity has been reported earlier.[5,6]

As blood–brain barrier and cell membrane transport water freely across it, hyponatremia causes free entry of water inside the cell and brain swelling. Brain cells have a protective mechanism which comes into play during hypo-osmolarity to maintain cell volume termed as "regularity volume decrease." This protective mechanism involves forcing out the interstitial sodium-rich fluid into cerebrospinal fluid (CSF) as a result of hydrostatic pressure. Other solutes (organic osmoles), such as taurine, glutamate, and myo-inositol, are lost over a day to few days rendering the cells isotonic to the extracellular fluid and maintaining the cell volume. Animal studies suggest that this process completes in 48 hours. The reaccumulation of electrolytes lost in response to a hypertonic environment is not the same process "in reverse" as their loss. Once inorganic ions shift has been exhausted, if the rate of rise of tonicity is faster than the rate at which organic osmoles can be synthesized and/or transported into the cells, the cell will shrink. It appears that oligodendrocytes are especially vulnerable to death preassembly from volume loss. It is, perhaps, the nutritional status of the patient plays its part, impairing the ability to regenerate the organic osmoles.[7] The process of maintaining iso osmolar environment needs energy reserves for functionality of Na^+/K^+ ATPase pump which is lacking in malnourished and alcoholics. Additionally, alcoholics have decreased glucose uptake in brain due to associated thiamine deficiency. Decreased glucose uptake further impairs energy production. This energy supply–demand imbalance leads to activation and pro-apoptotic drive and neuron cell death.[8]

Another important factor in alcoholics is antidiuretic hormone suppression, cerebral salt wasting syndrome, hypovolemia, and persistence of chronic hyponatremia which independently increase myelin damage due to the impaired sodium and water regulation.[9,10] The patient was also chronic alcoholic and severely dehydrated on the day of presentation.

Now, the management dilemma is balancing between the incidence of CPM and the mortality of hyponatremic brain edema in those patients treated rapidly vs very slowly, respectively.

In acute hyponatremia, cells have insufficient time to adapt to hypotonic extracellular environment; this can lead to cerebral edema, while in chronic hyponatremia, adaptation has already occurred, and a rapid treatment can cause ODS by rapidly increasing extracellular tonicity. [11,12] Therefore, for each patient with profound hyponatremia, cerebral edema or ODS should be suspected.

Guidelines for correction of hyponatremia in both the United States of America and the European countries have reached consensus that the limit (not the goal) should be around 10 mmol L^{-1} per day for both acute and chronic hyponatremias.[13,14] The guidelines of the United States of America recommend a lower limit of 8 mmol L^{-1} per day if there is high risk of ODS such as in patients with hypokalemia, alcoholism, malnutrition, or liver disease.[14,15]

Conclusion

The rapid correction of chronic hyponatremia could lead to an alarming complication of CPM which is life-threatening as well as irreversible in nature, especially in patients with multiple risk factors such as alcohol abuse and malnutrition.

Since there is no clear cutoff for minimal rate of correction of hyponatremia, we should weigh the risk of ODS vs cerebral edema especially in high-risk patients. Keeping this in mind, the treatment of hyponatremia should be done cautiously by slowly correcting it within adaptable limit of the body physiology.

This case is noteworthy as it shows that in alcoholics, hyponatremic correction should be done very slowly; it also shows that ODS can have favorable outcome with timely diagnosis and management.

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