Case Report

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Central Pontine Demyelination: A Case Report

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Central Pontine Demyelination is a neurological disease caused by rapidly fluctuating serum osmolality resulting in severe damage of the myelin sheath of nerve cells in the brainstem, more precisely in the area of pons. This condition is associated with electrolyte disorders, especially severe hyponatremia and its rapid correction. Its clinical course is characterized by alterations in the mental status to debilitating neurological status i.e. coma. Chronic hyponatremia and its correction, with or without evaluating safe limit could result in pontine demyelination. Demyelination might also occur with normal sodium levels, and even if serum sodium levels are corrected within safe limits. The objective of this case report is to give a broad perspective on Central Pontine Demyelination and to discuss about the different factors contributing to the demyelination and the various causes, pathophysiology and the management of this condition.

Key Words: central pontine demyelination, cerebral edema, hyponatremia, pons

Central Pontine Demyelination is a neurological disease caused by rapidly fluctuating serum osmolality resulting in severe damage of the myelin sheath of nerve cells in the brainstem, occurs more precisely in the area of pons[2]. It occurs sporadically in all ages, equally affecting both males and females.[6]Pontine lesions may be associated with extrapontine myelinolysis (EPM), which can symmetrically or asymmetrically affect cerebellar peduncles, caudate nucleus, putamen, frontal and temporal white matter, fornix, external and extreme capsules, claustrum, thalamus, subthalamic nucleus, internal capsule, amygdaloid nucleus, lateral geniculate nucleus, deep layers of cerebral cortex, hippocampus and corpus callosum.[2,5,9]

Disease is associated with electrolyte disorders, especially severe hyponatremia and its rapid correction. Some disorders predispose to this condition such as liver failure, liver transplantation, pituitary tumor resection, severe burns,chronic alcoholism, chronic renal failure, hemodialysis,lymphoma,carcinoma, malnutrition, severe bacterial infections, dehydration,electrolytic disorders(hyponatremia, hypernatremia, hyperglycemia, hypokalemia), acute hemorrhagic pancreatitis.[2,3,5,7,9]

Its clinical course is characterized by acute mental status changes with depressed level of consciousness, locked in syndrome, coma, quadriparesis, pseudobulbar palsy-develops 2 to 7 days after correction of hyponatremia. [2,5]

Brain MRI is diagnostic for pontine demyelination. Lesions are symmetric or asymmetric and hypointense on T1-weighted images, typically sparing the periphery of pons and hyperintense on T2 weighted and FLAIR images.[1,6,7,9] In subacute and chronic phases, extrapontine lesions become smaller in size and more clearly defined[2]. The early identification of patient at the risk of disease is first step for its treatment.[3]

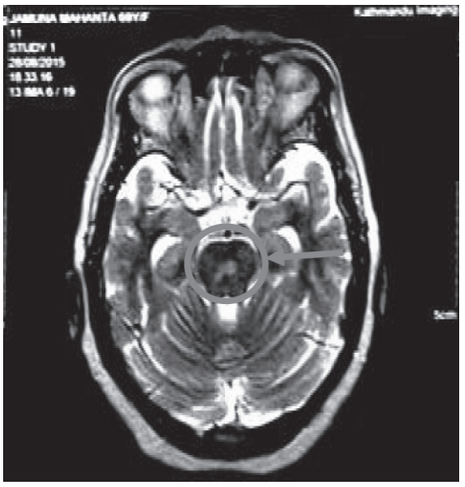

Figure 1: Hyperintense area seen in pons area in axial view

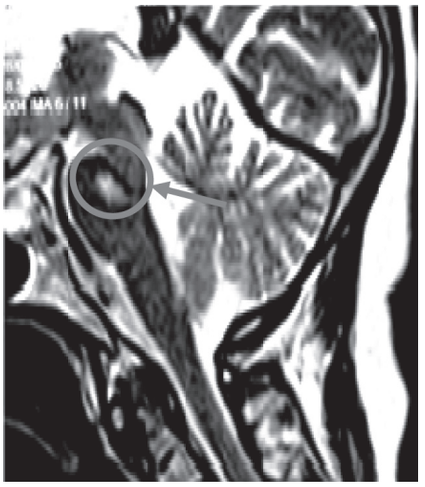

Figure 2: Hyperintense area seen in pons area in sagittal view

We report a case of central pontine myelinolysis, a rare neurological disease, associated with correction of hyponatremia.

Case Report

A previously healthy 54-year-old lady, hypertensive under medication for 12 years was admitted to our hospital following 3 days' history of weakness of both upper and lower limbs and slurring of speech along with drooling of saliva from left side of her mouth for 2 days.

There was a history of 17 – 20 episodes of repeated vomiting 12 days back followed by muscle weakness. She had h/o erosive gastritis since 20 years. Her family member took her to nearby hospital for repeated vomiting and weakness. All baseline investigations were sent. Baseline parameters were normal except her sodium level, which was found to be decreased down to 110 mmol/L. She then had sodium correction using 3% NaCl. On first day, her sodium level elevated upto 118mmol/l, 2nd day-125mmol, 3rd day-131 mmol/l and 135mmol/l in the 4th day. After the sodium level was back to normal, she got discharged.

During presentation in our hospital, she was anxious and she did not follow command. Her Glasgow Coma Scale was E4V5M5. Her neurological examination revealed slurring of speech and ataxia but no abnormalities were noted in cranial nerves and meningeal signs. Her motor power in bilateral upper and lower limbs were 4/5. Sensation was intact and reflexes were normal. Other systemic examinations showed no significant abnormalities.

Initial investigations showed normal Complete Blood Count, Random Blood Sugar and Urine Routine Microscopy. Her Na/K level was 138/4mmol/l. Her Urea, Creatinine, Lipid Profile, TFT, Chest X-ray and Echocardiography had normal findings. Plain CT head was done-normal scan. MRI Brain revealed Central Pontine Myelinolysis **(Figure 1,2)**.

Patient was kept under close monitoring in Intensive Care Ward and managed with steroids, Carbonic anhydrase inhibitors and Citicoline. She was hospitalized for 8 days, advised for long term physiotherapy and discharged.

Discussion

Central Pontine Myelinolysis (CPM) and Extra Pontine Myelinolysis (EPM) belong to the osmotic demyelination syndrome (ODS) and are frequently related to rapidly corrected hyponatremia.[3,5,8] Demyelination might also occur with normal sodium levels, and even if serum sodium levels are corrected within safe limits. Central basis pontis is more susceptible to acute metabolic stress.[2]

Patient described here had correction of hyponatremia, from 110 to 139mmol/l in 4 days. Her hyponatremia was corrected within safe limits but she developed pontine demyelination. It indicates demyelination can result from chronic hyponatremia, even if hyponatremia is corrected with much precautions. The initial intensity of hyponatremia and the corresponding absolute increase in final serum sodium levels determine its prognosis.[2,7,8]Here, patient had chronic hyponatremia due to erosive gastritis with multiple episodes of vomiting. She developed weakness of upper and lower limbs, along with slurring of speech 8 days after her sodium correction. Chronic hyponatremia and its correction, with or without evaluating safe limit could result in pontine demyelination.

The fall in serum tonicity in patients with hypotonic hyponatremia promotes water movement into the brain and if the hyponatremia is acute and severe, can lead to

cerebral edema and neurologic symptoms. In response to hyponatremia, the brain makes adaptations that lowers the cerebral volume toward normal and reduce the likelihood of these complications. However, brain adaptations that reduce the risk of cerebral edema make the brain vulnerable to injury if chronic hyponatremia is corrected. The neurologic manifestations associated with overly rapid correction results into potentially incurable disease central pontine demyelination.

Avoidance of CPM/EPM is dependent upon recognizing those patients with conditions pre-disposing them to osmotic myelinolysis and then moderating the rate of normalization of the electrolyte imbalance. Possible mechanisms include a hyper osmotically induced demyelination process resulting from rapid intracellular/ extracellular to intravascular water shifts producing relative glial dehydration and myelin degradation and/or oligodendroglial apoptosis.[6]Another possible explanation of central pontine demyelination is that the brain begins to adapt to hypotonicity almost immediately after a fall in serum sodium, and the adaptation is complete within two days. The rapid correction thus results into this disease. This disease is characterized by extremely poor prognosis. If patient develops locked in syndrome, its fatal. Prognosis is somewhat better during childhood.[2]

Conclusion

Central Pontine Demyelination is a neurological disease caused by rapidly fluctuating serum osmolality resulting in severe damage of the myelin sheath of nerve cells in the brainstem, occurring more precisely in the area of pons. It may or may not be associated with rapid correction of hyponatremia as seen above in the case presented, in which the correction of sodium was done within the safe limit, however the demyelination was noted. It may also be associated with normal level of serum sodium. The prognosis of the disease is poor. If left untreated, it may lead to severe neurological deficit or coma once patient develops locked in syndrome. So, the key point is the physician's knowledge regarding the correction of sodium and even after the proper doing so, the timely detection of the condition once patient presents with the symptoms. This might be crucial in the early and effective management of the patient.

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