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Case report
Osmotic demylination syndrome

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ABSTRACT

The osmotic demyelination syndrome (ODS) is a complication of rapid correction of hyponatremia causing injury to axons due to rapid changes in osmotic gradients across extracellular compartment of the brain. Features of the syndrome may include dysarthria, dysphonia ,tremors,quadriparesis and neurocognitive changes.MRI shows characteristic lesions in pons and(or) extrapontine locations or both. Treatment is symptomatic and supportive. Slow correction of hyponatremia seems to be the best way to prevent development of the syndrome.

1. INTRODUCTION

Osmotic demylination syndrome occurs due to injury to brain parenchyma due to rapid changes in plasma osmolality. Pons is involved more commonly but other areas like basal ganglia and cerebellum can also be involved. Prognosis is variable. We report a case of osmotic demylination syndrome in which both pons and basal ganglia were involved and patient recovered on conservative and supportive therapy.

2. CASE HISTORY

A 30 year old chronic alcoholic who had recently stopped drinking presented with generalized tonic clonic seizures to a local hospital. Patient was found to be markedly dehydrated and in drowsy state. Laboratory values revealed hyponatremia (serum Na+ 108 mmol).Patient was treated with antiseizure medication and given intravenous fluids to correct dehydration. The following day laboratory investigations revealed serum Na+ 120mmol/,suggestive of rapid correction of hyonatremia. After two more days patient condition worsened and he was referred to our hospital . Patient was conscious, had tremors involving both upper and lower limbs and the tongue. Patient had weakness of all four limbs, difficulty in swallowing and slurring of speech. Neurological examination of the patient revealed him to be conscious ,cooperative, oriented and with normal intelligence. Dysarthria was present. Cranial nerve examination revealed upper motor neuron(UMN)type of paralysis of ninth and tenth nerves. Marked hypertonia and decreased power was noted in all the four limbs. All the deep tendon reflexes were exaggerated .Plantars were bilaterally extensor. Examination of sensory and cerebellar systems was normal. No signs of meningeal irrritation were noted. All routine laboratoty tests were normal. MRI of the brain was performed to find the exact anatomical basis of neurolgical signs in the patient. MRI of the brain revealed pontine and basal ganglia lesions. Pontine lesions were hyperintense on flair and showed diffusion restriction ,sparing of peripheral and corticospinal tract(figure1,2). Basal ganglia lesions were hyperintense on flair and showed mild restriction on DW imaging(figure 3,4).Because of these characteristic pontine and basal ganglia lesions, clinical features and history of rapid correction hyponatremia ,diagnosis of osmotic demylination syndrome was made. Patient was given supportive and symptomatic treatment. There was gradual recovery in his clinical condition over a period of two to three weeks when he was discharged. At 6 weeks follow up, patient showed complete recovery.

3. Discussion:

Osmotic demyelination syndrome(ODS) is an acquired condition that results from an osmotic insult. It is described as central pontine demylination(CPM) when pons is involved, which is the most common site of involvement and extrapontine demylination(EPM) when the lesions are seen in the thalami, basal ganglia, lateral geniculate body, cerebellum, and the cerebral cortex.Regardless of the etiology, CPM is consistently reported to be more common than EPM or CPM/EPM combined[1,5,13].It is most commonly associated with rapid correction of hyponatremia, in which sodium level increases by more than 12 mmol/L/d usually in patients with a history of chronic alcohol abuse and malnutrition who are generally are deficient in organic osmolytes. Additional comorbid conditions that predispose patients to osmotic demyelination syndrome include prolonged use of diuretics, liver failure, liver transplantation and extensive burns. Osmotic demyelination syndrome affects men more often than women, and it is most common in middle-aged patients[6].

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The pathophysiology of ODS has not been well understood. Histologic studies have shown oligodendroglial cells are most susceptible to ODS related osmotic stresses, with the distribution of ODS changes paralleling the distribution of oligodendroglial cells that normally embed large neurons within the central pons, thalamus, cortex, putamen, lateral geniculate, and other extrapontine sites[3,7,8,9,10,11].The main pathologic finding seen with ODS is a symmetric area of myelin disruption in areas with admixed gray and white matter. The demyelinating process is characterized by vacuolization and intramyelinitic splitting with eventual rupture of the myelin sheaths, believed to be caused by osmotic gradient effects[2,3,6].

Clinical Presentation ODS can present in three distinct ways: isolated CPM, isolated EPM, and combined CPM and EPM. Classically, the symptoms of CPM present in a biphasic pattern. Initially, the patient presents with a generalized encephalopathy and electrolytic dysfunction, both of which improve after treatment. After 2-3 days of rapid electrolyte correction, the patient develops neurologic abnormalities associated with myelinolysis, including dysphagia, dysarthria, opthalmoplegia, diplegia, and altered mental status that can eventually progress to coma or death.EPM presents with tremors and rigidity[3,6,7,13].

Imaging findings of osmotic demyelination syndrome typically lag behind clinical symptoms. On DWI, mildly restricted lesions can be detected within 24 hours after onset of symptoms and thus provide the earliest indication of this disease entity.Imaging Findings On CT, ODMS typically manifests as low-density lesions in the pons or other affected regions, and occasionally shows enhancement[4]. In acute CPM, a symmetric hyperintense trident-shaped area in the central pons is a characteristic finding on T2-weighted and FLAIR MR images .In EPM, symmetric signal alterations can be seen in the basal ganglia, thalami, lateral geniculate body, cerebellum, and cerebral cortex[6,7].Lesions show diffusion restriction and typically do not enhance after the administration of contrast material. The differential considerations include pontine infarcts, which can be distinguished by their asymmetric distribution, involvement of the peripheral pontine fibers, demyelinating disease processes, neoplastic involvement of the pons, and metabolic syndromes such as Leigh disease and Wilson disease[6].

Treatment is largely supportive. After ODS is established, dopaminergic medications similar to those used in the treatment of Parkinson's disease are helpful in ameliorating symptoms. Reinduction of hyponatremia and steroid usage has been suggested by some authors but this has not been widely accepted. The prognosis for osmotic demyelination syndrome is variable.The optimal rate of sodium correction to prevent ODS is less than 10 to 12 mEq/L per 24 hours and less than 18 mEq/L in 48 hours. However, they noted that patients with other risk factors for ODS may still be at risk for development of the syndrome at these rates of correction and may require slower rates[12].

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