OSMOTIC DEMYELINATION SYNDROME - A CASE SERIES

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ABSTRACT Osmotic Demyelination Syndrome is a rare neurological condition caused by a number of clinical conditions and with a wide spectrum of clinical features. This case series describes three cases admitted to our hospital and looks at their clinical profile, including aetiology, clinical features, imaging findings and outcomes after treatment. Rapid correction of sodium and alcoholism were found to be the main etiologies. Clinical presentation included altered sensorium, dysarthria, extrapyramidal symptoms, pyramidal involvement and movement disorders. Imaging done showed symmetrical lesions involving the pons and the basal ganglia. The treatment was supportive, and the outcomes were good in most cases. Early identification of this condition and treatment is key to good long term outcomes.

KEYWORDS Osmotic Demyelination Syndrome, Central Pontine Myelinolysis, Extrapontine Myelinolysis, Hyponatremia, Alcoholism

Introduction

Osmotic demyelination syndrome (ODS) is a relatively rare entity that was initially described by Adams et al. in 1959 as a condition involving the pons secondary to a demyelinating aetiology. This syndrome includes two main subtypes - central pontine myelinolysis (CPM) and extrapontine myelinolysis (EPM)[1]. The pons is the main site involved though other areas, including the basal ganglia, thalamus, cortex, and the subcortical white matter, were also involved in extrapontine cases[2]. Rapid correction of hyponatremia is the main cause detected, especially with rates exceeding 12 mmol/L per day[3]. Other associations include alcoholism, psychogenic polydipsia, diuretic use and chronic malnutrition. The exact pathogenesis is unclear, but it is postulated that the main changes observed in ODS were due to fluctuating osmotic changes causing pressure on the tract fibres and leading to the subsequent demyelination of these fiber tracts.

The most common clinical findings are pseudobulbar palsy, spastic quadriplegia, extrapyramidal features and altered mental status[4]. Other features may include horizontal and vertical gaze palsies, coma or locked-in syndrome and also akinetic mutism. The diagnosis is usually made by a combination of clinical suspicion and MRI imaging studies.

MRI scans show hyperintensities involving the central pons or other brain regions like the basal ganglia and the thalamus on T2-weighted (T2W) and fluid-attenuated inversion recovery (FLAIR) images [5]. Diffusion-weighted imaging (DWI) also showed increased signal intensity in some cases. These demyelinated lesions involving the central pons are trident in shape, sparing corticospinal tracts travelling in the ventral pons. Radiological changes tend to persist for months in spite of clinical improvement. The treatment of ODS cases is only supportive in nature[6]. Prevention of secondary complications is important for a good long term outcome. Rehabilitation therapy will be required in most cases.

We present a series of three cases of osmotic demyelination diagnosed and treated here in our hospital. We aim to highlight the likely etiologies, clinical presentation, imaging features and outcomes in these cases.
Case report

Case 1

This 62-year-old patient - a known case of diabetes and hypertension had a two-week history of generalized tiredness and slowness of activities. He was evaluated outside and detected to have hyponatremia with a presenting sodium level of 107 mmol/L. This was corrected with hypertonic saline infusion to 138 mmol/L within two days. This was followed by the patient developing stiffness of all four limbs, decreased speech output and tremors of all four limbs. He was referred here for further evaluation. Clinical examination revealed abulic phenotype with decreased responses and significant extrapyramidal features in the form of rigidity involving all the limbs, bradykinesia, tremors and gait imbalance. Lab values showed normal sodium reports in our hospital. MRI brain (Figure 1) - showed symmetrical hyperintensities on T2 weighted images involving the thalami, basal ganglia and also central pons with restriction on diffusion-weighted images and also the involvement of frontal lobes bilaterally. He was managed symptomatically with levodopa, pramipexole and physiotherapy. He had significant improvement in symptoms at discharge and is under follow up.

Case 2

This 35-year-old patient had a history of binge alcohol consumption followed by multiple episodes of vomiting. He was admitted outside for the same and detected to have hyponatremia - sodium levels of 103 mmol/L. He was treated with hypertonic saline and supportives outside, and the sodium was corrected to 134 mmol/L over a span of five days. He presented to our hospital with a history of tremors involving all four limbs, slurring of speech and slowness of all activities of one-week duration. Clinical examination revealed conscious and oriented patient with dysarthria, resting tremors of both upper limbs, decreased blink rate, gait ataxia and bilateral pyramidal signs in both upper and lower limbs with extensor plantar responses. Sodium levels were normal at admission. MRI Brain (Figure 2) - showed symmetric T2 hyperintensities involving central pons, bilateral striatum with restricted diffusion suggestive of osmotic demyelination. He was treated with intravenous thiamine, levodopa, pramipexole and supportive. He had improvement in symptoms and gait on follow up.

Case 3

This 58-year-old patient was admitted to a hospital outside with a history of fever with chills and generalized weakness. He was diagnosed with a urinary tract infection outside and started on antibiotics. He also had significant hyponatremia - sodium level of 107 mmol/L - which was corrected with hypertonic saline to 133 mmol/L over 4 days. He presented to our hospital with a history of altered behaviour, restlessness, irrelevant talk, slurred speech and difficulty in swallowing. He also had one episode of a generalized tonic-clonic seizure. Clinical examination revealed altered behaviour, spastic dysarthria, pseudobulbar palsy and bipyramidal limb signs. Lab values showed normal sodium levels at admission. MRI Brain (Figure 3) - showed symmetrical T2 hyperintensities involving corpus striatum, amygdala and external capsule - representing extrapontine myelinolysis. He was treated with levodopa, bromocriptine, piracetam, baclofen and supportive. He had significant improvement in speech and gait and was started on physiotherapy. He was discharged after stabilization.
of only EPM. Studies looking at the commonest sites involved in ODS have implicated the pons followed by the basal ganglia and the thalamus. Other sites involved include the cerebellum and lateral geniculate body. This has been postulated to be secondary to densely packed fiber tracts in these regions. In addition, rapid surges in osmotic pressure in these regions cause compression of nerve fibers, causing demyelination and lesions described. Pons and basal ganglia were the most common structures in our patients.

Current treatment of ODS is mainly supportive and involves correction of all underlying electrolyte disturbances and treatment of the underlying cause. The sodium correction rate should not exceed 8-12 mmol per day. In the presence of risk factors like alcoholism, malnutrition, or hypokalemia, in addition to hyponatremia, serum sodium should not be increased by more than 8 mmol/l per 24 h. In our patients, symptomatic treatment was carried out with levodopa and pramipexole in extrapyramidal signs and baclofen in cases with spasticity. All patients in our series had good improvement with treatment. Studies have shown that correction of altered sensorium and seizures trended to have better outcomes than those who had dysarthria and extrapyramidal symptoms. This could be explained by the extent of brain damage.

Experimental therapies tried include corticosteroids, plasma exchange and intravenous immunoglobulin[10]. Unfortunately, none of them has shown any significant benefit in trials. Rehabilitation in good centers also helps recover speech, swallowing, and gait.

Conclusion

ODS is a rare neurological syndrome that has a number of different etiologies and clinical manifestations. Imaging findings help to confirm the diagnosis in most cases. Though treatment is mainly supportive, early recognition can result in good long term outcomes. Further studies are required to understand the pathogenesis and management of this condition.

Abbreviations

- ODS-Osmotic Demyelination Syndrome
- CPM-Central Pontine Myelinolysis
- EPM-Extrapontine Myelinolysis
- MRI-Magnetic Resonance Imaging
- DWI-Diffusion Weighted Imaging
- FLAIR-Fluid Attenuated Inversion Recovery

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Conflict of interest

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References


