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Phenotypic Manifestations in Two Cases of Osmotic Demyelination Syndrome

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Abstract: Osmotic demyelination syndrome (ODS) is an emergency acute neurological condition that usually occurs secondary to rapid correction of chronic hyponatremia (low concentration of sodium in the blood). We present two cases: one with typical ODS following rapid sodium correction, showing classic MRI findings in the pons and basal ganglia; and another with atypical ODS presenting with cranial nerve palsy and ataxia, despite normal sodium levels, and diffuse hyperintensities in the pons and medulla on MRI. These cases emphasize the clinical and radiological variability of ODS, highlighting the importance of careful monitoring and gradual correction of electrolytes, particularly sodium, in high-risk patients.

Keywords: osmotic demyelination syndrome, hyponatremia, neurology, neuroimaging

Introduction

Osmotic demyelination syndrome (ODS) is an acute, non-inflammatory demyelinating disorder of the central nervous system characterized by symmetric destruction of myelin and oligodendrocytes with preservation of neurons and axons. It most commonly affects the pons, known as central pontine myelinolysis (CPM), but may also involve extrapontine regions such as the basal ganglia, thalamus, midbrain, cerebellum, or cortex, referred to as extrapontine myelinolysis (EPM).[1,2] Originally described by Adams et al in 1959 in alcoholic and malnourished patients,[3] ODS has since been widely associated with the rapid correction of chronic hyponatremia, which remains its most well-recognized trigger. It can also develop due to hypernatremia and hyperosmolar syndrome, hyperglycemia in association with concomitant acidosis, hyperemesis, severe diarrhea, cirrhotic liver disease, chronic alcoholism and organ transplantation, where patients have an inherent tendency for hyponatremia.[4] Importantly, ODS can occur even with a seemingly "adequate" correction of hyponatremia or in the absence of any measurable electrolyte imbalance, including normonatremic cases.[5]

Recent literature has expanded our understanding of ODS pathogenesis, suggesting that astrocyte dysfunction may precede demyelination and that hypokalemia, hypophosphatemia, and systemic illnesses may also serve as independent or additive risk factors.[6] Despite this, clinical recognition remains challenging due to the condition's broad spectrum of neurological presentations and radiologic findings.[7]

The current scientific gap lies in the under-recognition of atypical or normonatremic presentations of ODS, particularly those not driven by rapid sodium shifts. This case report presents two patients with distinct radiological and clinical profiles: one with classic post-correction ODS and one with normonatremic, atypical ODS. These cases aim to emphasize the phenotypic diversity of ODS, support broader diagnostic vigilance, and highlight the importance of comprehensive risk assessment beyond sodium correction alone.

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Case Description

Case 1

A 67-year-old patient was admitted to the neurological ICU department with acute confusion, general stiffness, left-sided weakness, inability to swallow, and slurred speech. Two weeks earlier, the patient experienced prolonged diarrhea, vomiting, and hypotension secondary to gastroenteritis, prompting hospitalization ten days after the onset of symptoms. The patient received fluids and electrolyte replacement. About 72 hours after that, he developed hypomimia, severe dysarthria, dysphagia with drooling, dysphonia, positive oral automatism signs, generalized muscle rigidity and bradykinesia. These extrapyramidal signs and bulbar symptoms are commonly seen in ODS which affects the pons and basal ganglia. Neurological examination also revealed that muscle strength in the left extremities was diminished to 3 points with hyperreflexia and left-sided plantar extension, consistent with corticospinal tract involvement seen in ODS.

Brain MRI performed 24 hours after neurological symptoms onset, showed symmetric hyperintensity on T2/FLAIR imaging in the pons, midbrain and basal ganglia with no diffusion restriction (Figure 1A–C).

Serum electrolytes were within normal range, except mild hypernatremia 148 mmol/L (normal 133–145 mmol/L). Further medical documentation evaluation revealed that at the time of first hospital admission, three days prior to fluid and electrolyte replenishment, the patient's serum sodium level was 102 mmol/L, potassium 3.0 mmol/L (normal 3.5–5.2 mmol/L), and both were rapidly corrected within 24 hours.

After two weeks of symptomatic treatment, the patient's confusion resolved, and left-sided weakness, dysphagia, and dysarthria partially improved. As a result of ODS, the patient developed parkinsonism, with symptoms effectively managed by levodopa therapy.

Case 2

A 51-year-old patient presented to the emergency department with a history of one-week high-grade fever, multiple vomiting and diarrhea on the background of alcohol abuse. The patient reported self-administering fluid and electrolyte replacement using over-the-counter oral rehydration solutions. On admission, he complained of diplopia, face asymmetry, weakness and numbness in the right extremities, a decrease in hearing, and severe unsteadiness.

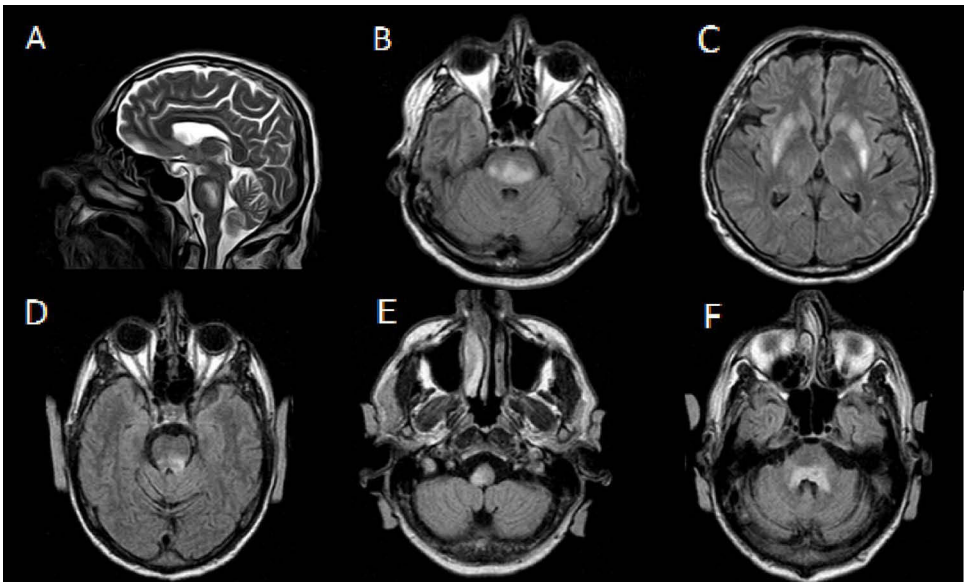

Figure 1 Representative MRI images of the presented cases. Case 1 (**A**–**C**): FLAIR images showed significant hyperintensity involving the pons (**A**), lower midbrain (**B**), and symmetric basal ganglia (**C**), consistent with osmotic demyelination syndrome. Case 2 (**D**–**F**): FLAIR images showed a diffuse area of hyperintensity without distinct margins, in the posterior and left part of the pons (**D**), medulla (**E**) and cerebellar peduncles (**F**).

Neurological status showed left abducent and facial nerve palsy, two-sided hypoacusis, horizontal nystagmus, left soft palate palsy, right-sided central hemiparesis with positive Babinski sign, absent patellar and Achilles reflex bilaterally, left-sided pain, temperature hypoesthesia and atactic gate.

The patient had a history of Hepatitis C, minimally active. During the hospital stay, he was diagnosed with erosive esophagitis due to previous profound vomiting. On admission, serum thiamine and electrolyte levels were within normal limits, except for chloride, which was decreased. Sodium was 136 mmol/L (reference range: 133–145 mmol/L), potassium was 3.8 mmol/L (reference range: 3.5–5.2 mmol/L), chloride was 92 mmol/L (reference range: 95–110 mmol/L), and thiamine was 181 nmol/L (reference range: 75–195 nmol/L).

Brain MRI, performed three days after admission, showed a diffuse area of hyperintensity in T2/FLAIR without distinct margins, in the posterior and left part of the pons, cerebellar peduncles, and medulla (Figure 1D-F). The latter two are uncommon radiological findings for ODS. Although clinical history and radiological findings raised suspicion for both ODS and Wernicke's encephalopathy as initial differential diagnoses, despite atypical neuroradiological features, the decreased chloride level and normal thiamine concentration were more consistent with a diagnosis of ODS.

Treatment was symptomatic, with fluid, glucose, electrolytes, and vitamins maintained. After 2 weeks, a moderate decrease in ataxia was observed, while right face asymmetry, left arm and leg paresis remained unchanged. The patient was discharged for further rehabilitation.

Differential Diagnosis

The rapid development of hemiparesis, dysphagia, and dysarthria in Case 1 suggested a brainstem stroke, but this diagnosis was ruled out by a brain MRI obtained after the onset of neurologic symptoms. Neuroimaging findings and a history of rapid electrolyte correction prior to the onset of neurologic symptoms suggest that ODS is the most likely diagnosis.

In Case 2, neuroimaging findings resembled various toxic, infectious, and inflammatory disorders. Among these, Wernicke's encephalopathy was considered the most competitive differential diagnosis; however, it was excluded based on decreased blood chloride levels and a normal thiamine concentration. A comprehensive clinical, laboratory, and radiological assessment ruled out other potential causes of the patient's clinical and imaging presentation, leaving ODS as the most plausible diagnosis.

Discussion/Conclusion

ODS has long been recognized as a complication of rapid correction of hyponatremia. However, recent studies have identified various other medical conditions that can also lead to ODS, independent of fluctuations in serum sodium level.[2,6] We described two cases of ODS: one exhibiting a typical presentation and the other demonstrating atypical features.

The patient in Case 1 presented with a history of severe hyponatremia of 102 mmol/L, which was rapidly corrected within 24 hours, triggering ODS with focal neurological symptoms developing three days later. The fact that this patient also had hypokalemia of 3.0 mmol/L is important, as it's an additional risk factor for ODS.[8] The patient in Case 2 appears to be independent of the classic mechanism of rapid correction of hyponatremia. He presented with unusual neurological impairment for ODS, manifesting as inferior medial pontine syndrome and ataxia. Brain MRI showed a diffuse area of hyperintensity in T2/FLAIR without distinct margins involving the posterior and left part of the pons, medulla, and cerebellar peduncles, which are uncommon radiological features of ODS. Involvement of the medulla and cerebellar peduncles is particularly rare, even in extrapontine myelinolysis.[9]

Patients with a high risk of central pontine demyelination syndrome are those with a serum sodium concentration ≤105 mmol/L or those with significant hypokalemia, alcoholism, malnutrition, and advanced liver disease.[2]

In chronic severe hyponatremia, the brain adapts to reduced serum osmolality by the efflux of organic osmolytes such as glutamate, glucose, taurine, and glycine, along with water, from brain cells. This compensatory mechanism reduces cellular swelling. However, rapid correction of hyponatremia can lead to an inability of brain cells, particularly astrocytes, to rapidly reuptake the lost osmolytes and water, resulting in cellular dehydration and apoptosis. Astrocyte damage compromises the integrity of the blood-brain barrier, leading to the release of inflammatory cytokines, which activate microglia and induce an inflammatory response. This cascade of events can lead to demyelination.[6] In cases of severe hyponatremia (serum sodium <120 mmol/L) accompanied by neurological symptoms, the administration of hypertonic saline (3%) is indicated. Co-administration of desmopressin with hypertonic saline has been shown to reduce the incidence of overcorrection.[6] The correction

rate should be carefully controlled, typically aiming for an increase of 6–9 mmol/L over 24 hours and should never exceed 12 mmol/L in the same period.[10] Osmolality should be monitored hourly to ensure effective antidiuretic action and prevent complications associated with rapid overcorrection.[11]

Physicians should be aware of the diverse presentations and underlying mechanisms of ODS, which can arise not only from the rapid correction of hyponatremia but also from other risk factors such as malnutrition, alcoholism, hypokalemia, and chronic diseases. By raising awareness of these presentations and pathogenesis variations, medical practitioners can more effectively prevent and manage ODS, improving patient outcomes.

Key Clinical Message

ODS can result from rapid sodium correction or risk factors like severe hyponatremia (<105 mmol/L), hypokalemia, alcoholism, or malnutrition. To prevent ODS, clinicians should carefully monitor high-risk patients and limit sodium correction to 6–9 mmol/L over 24 hours, avoiding overcorrection to improve outcomes.

Consent

Written informed consent was obtained from both patients for publication of case details and accompanying images. Institutional approval was not required for publication of the case reports.

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Disclosure

The authors declared no potential conflicts of interest with respect to publication of the article.

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