

Case Report:

Isolated extra pontine myelinolysis – a rare imaging appearance of osmotic demyelination syndrome

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ABSTRACT

Rapid correction of hyponatraemia leads to serious neurological complications, like osmotic demyelination syndrome (ODS). In ODS, magnetic resonance imaging (MRI) often reveals features of pontine myelinolysis, that may occur in isolation or may, sometimes be associated with extrapontine myelinolysis. Isolated extrapontine myelinolysis is rare. We report the case of a 53-year-old lady brought to the emergency service with vomitings, and altered sensorium. She was found to have profound hyponatraemia (serum sodium 110 meq/L). Correction of hyponatremia was done with slow intravenous infusion of 3% sodium chloride. However, inadvertant, concomitant oral administration of salt led to overcorrection with serum sodium going upto 150 meq/L. She developed quadriplegia, depressed level of consciousness and respiratory failure and required ventilatory support. MRI brain showed features of isolated extrapontine myelinolysis.

Key words: *Hyponatremia, Osmotic demyelination syndrome, Extrapontine myelinolysis*

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INTRODUCTION

Osmotic demyelination syndrome (ODS) is a relatively uncommon neurological complication produced by rapid correction of hyponatraemia. It leads to damage to myelin of pontine and extrapontine structures.¹ Extrapontine myelinolysis is even rare compared to pontine myelinolysis. ODS presents with quadriparesis, changes in neurocognitive function and characteristic lesions in magnetic resonance imaging (MRI) of brain.²

CASE REPORT

A 53-year-old lady was brought to the emergency service with the complaints of vomiting and altered sensorium of 3 days duration. Illness started with non-bilious, non-projectile vomiting. There was no history of diarrhoea, pain in abdomen, seizures or

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haematemesis. Her menstrual cycles were regular till she had undergone hysterectomy 20 years ago. She was a mother of 4 children. On clinical examination she was drowsy, but arousable and responding to verbal commands. She was afebrile. Pulse was 88 beats/min, blood pressure was 150/80 mm of Hg, respirations were 14/min and jugular venous pulse was not elevated. There was no evidence of pedal oedema or ascites. There was no focal neurological abnormality. Rest of the physical examination was unremarkable.

Her haemoglobin was 10 g/dL, total leukocyte count 19,200/mm³ and erythrocyte sedimentation rate (ESR) was 52 mm at the end of the 1st hour. Serum biochemistry showed osmolality 286 mosm/Kg of water, sodium 110 mEq/L, potassium 2.1 mEq/L, creatinine 0.61 mg/dL, urea 16 mg/dL, plasma glucose 153 mg/dL, magnesium 2.6 mg/dL, albumin 4.4 g/dL,

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calcium 10.3 mg/dL, and phosphorous 2.2 mg/dL. Liver function tests were within the normal limits. Urine osmolality was 486 mosm/Kg. Serum thyroid stimulating hormone levels (TSH) was 8.9 μ IU/mL (normal 0.5 – 4.0 μ IU/mL), T4 was undetectable (normal 55 – 135 ng/mL) and T3 was 0.7 ng/mL (normal 0.8 - 2.0 ng/mL), suggestive of sick euthyroid syndrome. Stimulated serum cortisol was greater than 60 μ g/dL at the end of both 30 minutes and 60 minutes, suggestive of normal response. Chest radiograph and abdominal ultrasonography revealed no significant findings. Computed tomography (CT) of brain showed lacunar infarcts in bilateral lentiform nuclei with mild cerebral atrophy.

Normal saline infusion at the rate of 75 mL/hour and intravenous potassium chloride were started. Repeat serum sodium and potassium measured after 6 hours of above mentioned treatment was 108 mEq/L and 2.8 mEq/L respectively. As the patient had profound hyponatraemia and hypokalaemia with

depressed level of consciousness, intravenous administration of 3% NaCl was started at rate of 10 mL/hour. Potassium supplementation was continued. Over the next 36 hours, serum sodium rose to 115 meq/L with improvement in sensorium.

As the patient became fully conscious and oriented in time, place and person, hypertonic saline infusion was stopped and she was allowed intake orally. Oral salt and oral potassium supplementations were continued. After the next 12 hours patient became excessively drowsy and gradually became comatose.

On examination there was up-beating nystagmus, spastic quadriparesis and exaggerated deep tendon reflexes. Serum sodium was found to be 150 mEq/L. Intravenous 5% dextrose infusion was administered. Measures to prevent bed sores like use of waterbed, frequent position changing were provided. Oxygen saturation was maintained with oxygen mask till 7th day of admission. Sodium gradually came

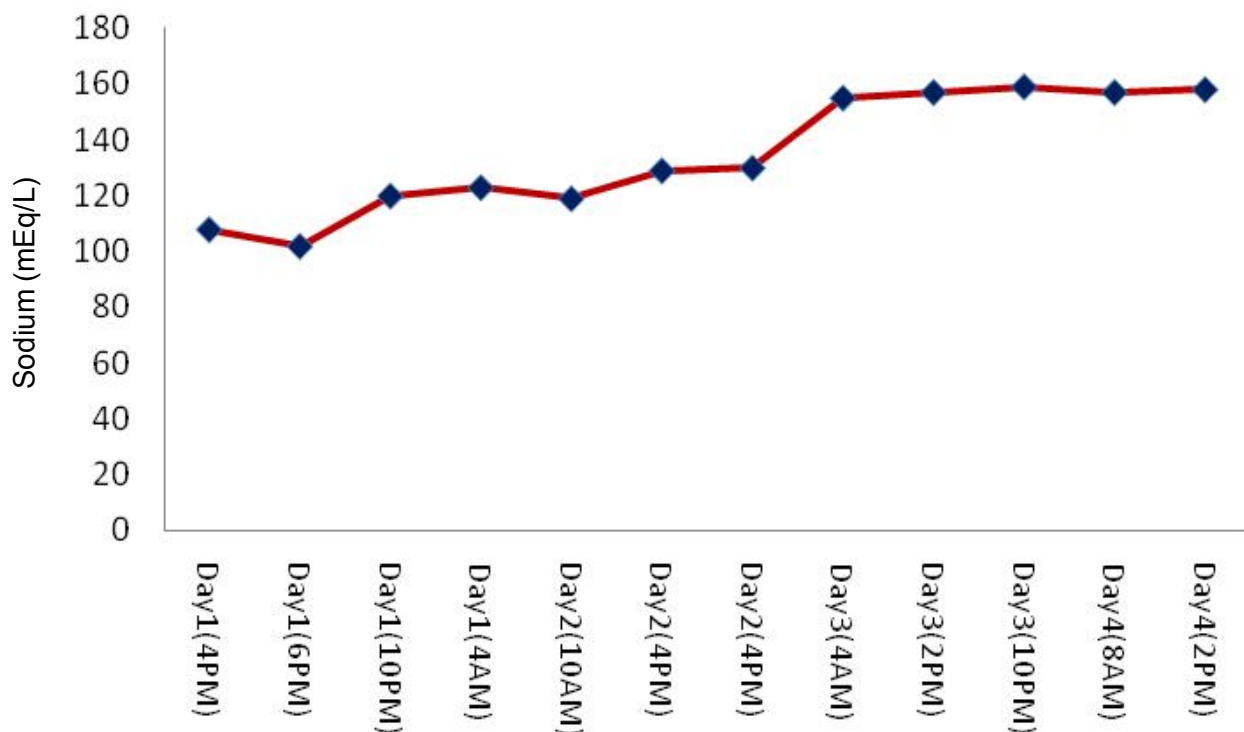


Figure 1: Serum sodium (mEq/L) levels during first four days of in-hospital stay

down to 130 meq/L. On the 7th day patient developed excessive oral secretions and was not able to maintain O₂ saturation with face mask. Assisted mechanical ventilatory support was instituted. In spite of these efforts, the patient died on the 26th day of admission.

Change in serum sodium for the first four days is shown in the Figure 1. Magnetic resonance imaging (MRI) of brain obtained on 5th day of admission showed changes of extra pontine myelinolysis in diffusion weighted images (Figure 2).

DISCUSSION

In hospitalized patients hyponatremia is the most commonly encountered serum electrolyte imbalance. The non-osmotic release of anti-diuretic hormone (ADH) from supraoptic and paraventricular nuclei causes free water retention and dilutional hyponatraemia.³ Severe hyponatraemia (serum sodium < 120 meq/L) is a serious electrolyte disorder associated with life-threatening neurological complications.⁴ Acute hyponatraemia causes increased intracranial pressure and produces cerebral oedema. At the same time rapid correction of hyponatraemia leads to disastrous consequences like ODS.⁵ So hyponatraemia should be treated very judiciously, weighing the risks and benefits.

ODS causes damage to myelin sheath without affecting axons and neurons. No inflammatory infiltrate is seen.⁶ In a pathological case series (n=58) isolated pontine lesions were seen in 50% of cases. Pontine and extra-pontine involvement was evident in 30% cases and isolated extrapontine involvement was seen in 20% cases.² In the present case, there were changes suggestive of extra-pontine myelinolysis only. ODS has a peak incidence in adults aged 30 to 60 years with male predominance.^{7,8} The clinical presentation of ODS is characterized by changes in the cognitive function, quadraparesis with dysphagia, dysarthria and pseudobulbar symptoms, among others.⁷

MRI has got a greater sensitivity and specificity than CT in diagnosing changes of myelinolysis. In ODS, MRI typically reveals trident shaped, symmetrical signal change within the pons on T2-weighted images. Transverse fibres are most affected with relative sparing of peripheral tissue and descending tracts. Contrast enhancement is unusual. Some uncommon manifestations include extrapontine changes, described in the basal ganglia, white matter and cerebellum. Pallidal sparing is usually evident. Diffusion weighted imaging (DWI), which is sensitive to the motion of water is helpful in diagnosing the lesions in ODS during the early clinical phase when conventional MRI imaging

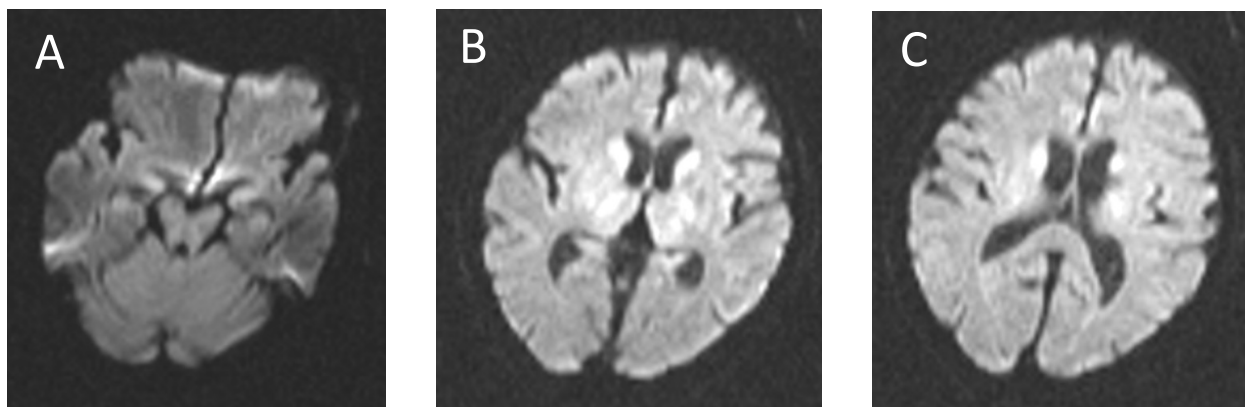


Figure 2: Axial diffusion weighted MRI images showing normal brainstem (A); hyperintensities in bilateral caudate nucleus and thalami (B); and hyperintensities in bilateral corona radiata and caudate nuclei (C)

MRI = magnetic resonance imaging

may be negative. Restricted diffusion has been described as the first imaging manifestation of ODS in the setting of otherwise normal conventional MRI sequences within 24 hours of onset of tetraplegia.⁹

Treatment of hyponatraemia should be individualized. Factors taken into consideration when making a treatment decision for a hyposmolar patient are, the severity, duration of the hyponatraemia and the patient's neurologic status. Patients with acute hyponatraemia (≤ 48 hours duration) are usually symptomatic if the hyponatraemia is severe (i.e., < 125 mEq/L). They are at greatest risk for neurologic complications from the hyponatraemia, but seldom develop demyelination, possibly because sufficient brain volume adaptation has not yet occurred. Patients with chronic hyponatraemia (> 48 hours duration) have minimal neurologic symptoms and are at little risk from complications of hyponatraemia itself but can develop demyelination after rapid correction. In patients who are minimally symptomatic, the lower recommended rate of correction, 0.5 mmol/L or less is preferred. If neurologic symptoms are severe, an initial correction at a rate of 1 to 2 mmol/L per hour may be appropriate. Intensive treatment should be interrupted once the patient becomes asymptomatic or serum sodium levels greater than 125 mmol/L are achieved or a total magnitude of correction of 18 mmol/L is achieved.

Hypoosmolar hyponatraemia can be treated with either isotonic or hypertonic saline infusions, depending on the cause of the hypoosmolality. Patients with hypoosmolality due to volume depletion respond well to isotonic saline infusion. ODS has been reported even after correction of hyponatraemia with normal saline. The present patient developed ODS in spite of very slow rate of hypertonic saline administration. The enthusiastic

additional oral salt administration and associated hypokalaemia are probably the causes for ODS in the patient. The predictive factors for poor outcome in ODS include severe hyponatraemia (≤ 115 mEq/L), associated hypokalaemia and low Glasgow Coma Scale (GCS) at presentation.¹⁰ The best way to prevent ODS is judicious, regulated administration of normal or hypertonic saline.

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