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Case Report

Osmotic Demyelination Syndrome : A Case Report

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Abstract

We report a case of a middle aged lady who presented with alteration of consciousness and dysphasia. She was found to have hyponatremia which was corrected rapidly. After initial improvement, she subsequently developed marked deterioration of conscious level with upper motor sign signs in all four limbs. Osmotic demyelination syndrome was diagnosed by MRI. Severe hyponatremia carries a risk of cerebral edema with a significant mortality, but correcting it too rapidly can result in even more disastrous condition- osmotic demyelination syndrome.

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Introduction

Central pontine myelinolysis (CPM) was first described by Adams et al. in 1958[1]. These authors observed that alcoholic or malnourished patients developed spastic quadriplegia, pseudobulbar palsy and encephalopathy or coma from non-inflammatory demyelination in the basis pontis. It is now accepted that CPM may occur as a complication of severe hyponatremia, especially if this is prolonged, or if it is corrected rapidly [2]. CPM is defined as concentrated, frequently symmetrical, non-inflammatory demyelination within the central pons. However, in 10% of cases demyelination also occurs in extrapontine regions including the midbrain, thalamus, basal nuclei and cerebellum. The pathophysiology is not clear. It has been suggested that, in areas of interdigitation of white and grey matter, cellular edema caused by fluctuations in electrolyte forces results in compression and subsequent demyelination of fiber tracts[2,3]. The term 'osmotic demyelination syndrome' (ODS) has been coined as it is better suited than CPM for cases, especially those with extra-pontine lesions that result from correction of hyponatremia[2, 4].

Alcoholism is the most common predisposing factor for CPM and ODS. Among others hyponatremia, malnutrition, prolonged diuretic use and psychogenic polydipsia are common. The incidence is unknown. The most common clinical findings are of pseudobulbar palsy, spastic quadriplegia and delirium. Other features may include horizontal and vertical gaze palsies, coma or locked-in syndrome. The diagnosis is usually made clinically. T2-weighted magnetic resonance imaging (MRI) images may demonstrate hyperintense areas of demyelination.

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Case Report

A 55-year-old hypertensive, non-diabetic house wife hailing from Paba, Rajshahi, suddenly became drowsy and confused and unable to speak. There was no definite weakness of any limb. With these complaints she was admitted into hospital and was diagnosed as a case of hypertension with diuretic induced electrolyte imbalance and was treated accordingly. She was improved with treatment and discharged after 5 days. But on the next day the patient again become unconscious and got admitted into hospital. There was no history of fever, headache, convulsion or head-injury. She was taking β blocker and diuretic for last 2 years.

On examination, she was found unconscious. Her pulse was 80 bpm, BP- 95/60 mm of Hg, respiration rate–18 /m and temperature was normal.

On Glasgow Coma Scale her Level of consciousness was 07; Pupils– normal, reacting to light; Fundus– normal; Doll's eye movement– present; All reflexes are bilaterally exaggerated; Clonus- absent; Planter reflex – equivocal

Investigations

On 1st Admission:

On 27/03/09: WBC-11500 /mm3; Hb%-10.3 gm/dl; ESR-24mm in 1st hr; N-80; E-02; L-16; M-02; RBS- 6.3mml/l; S. Creatinine- 1.1 mg/dl; S. Electrolyte- Na+ -95.1 mmol/L; K+ -2.52 mmol/L; Cl- -61.7 mmol/L; HCO3- -23.6 mmol/L; Urine RE – Normal; S. Bilirubin – 1 mg/dl.

On 28/03/09: S. Electrolyte- Na+ -118.3 mmol/L; K+ -2.7 mmol/L; Cl- 87.3 mmol/L; HCO3- -23 mmol/L.

On 30/03/09: S. Electrolyte- Na+ -130 mmol/L; K+ -2.5 mmol/L; Cl- 98.7 mmol/L; HCO3- -23 mmol/L.

On 02/04/09: CT Scan of Brain- Normal finding.

On 2nd Admission:

On 06/04/09: S. Electrolyte- Na+ -133 mmol/L; K- -4.44 mmol/L; Cl- 94.3 mmol/L; HCO3- -27.2 mmol/L; ECG-Old anterior myocardial infarction, Chest X-ray PA view- cardiomegaly, Echocardiography- Anterior MI with fair LV function.

On 07/04/09: MRI of Brain- Symmetrical hyperintensity of T2 weighted and FLAIR images are noted in both basal ganglia and in the pons which is hypointense in T1 weighted image. Features are suggestive of Central Pontine Myelinolysis.

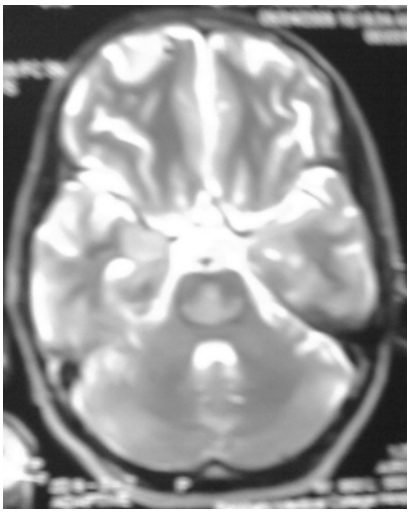

Fig.1

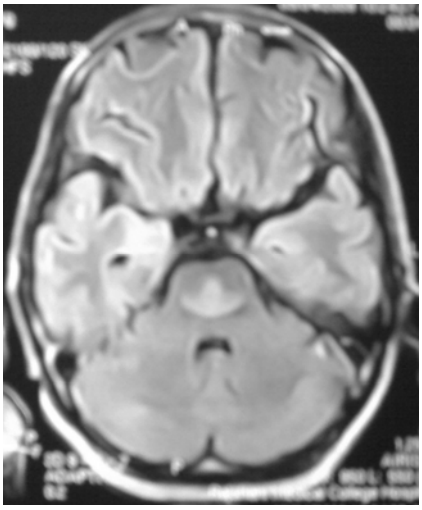

Fig. 2

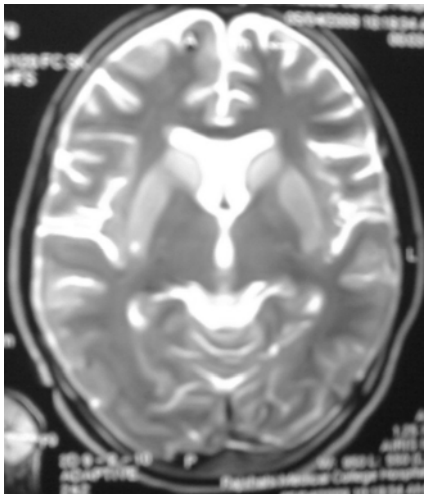

Fig. 3

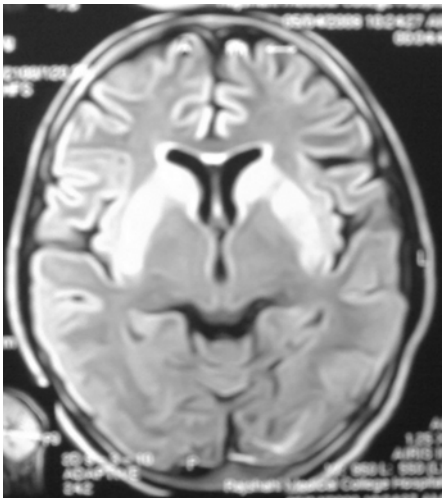

Fig. 4

Figures: MRI of Brain shows symmetrical hyperintensity of T2 weighted and FLAIR images are noted in the pons (Fig. 1 & fig. 2) and in both basal ganglia (Fig. 3 & Fig. 4).

Discussion

Hyponatremia is common in hospital patients, the incidence being 3–5% for a serum level of 130 mmol/L or less [2] and 0.15% for 120 mmol/L or lower [5]. In most series the mortality from severe hyponatremia (< 120 mmol/L) lies between 40% and 50%, except in settings such as intensive care where it ranges from 10% to 20% [6]. Knowledge of its duration and etiology is necessary to guide treatment. Acute hyponatremia, especially with symptoms, carries a risk of cerebral edema and requires prompt action. However, rapid correction in chronic hyponatremia will predispose to osmotic demyelination, so that the difficulty lies in judging whether the hyponatremia is acute or chronic. There is also no consensus about the best rate of correction. Correcting the serum sodium at a rate not greater than 0.5 mmol/L/h is advised. However, if the hyponatremia is known to be chronic, an increase of only 8 mmol/L/day is the target. For acute hyponatremia, initial rates of correction up to 1–2.4 mmol/L/h have been suggested, with the intention of reaching mild to moderate hyponatremia until symptoms resolve. Sodium replacement can then be slowed or stopped [2, 7]. However, Reynolds et al. assert that 'as the duration may be difficult to judge, the presence of symptoms and their severity should guide the treatment strategy' [8]. Hyponatremia is categorized as severe when serum concentration is < 120 mmol/L, moderate when serum concentration is 120-124 mmol/L and mild when serum concentration is 125-134 mmol/L [9].

It is possible that the patient presented here had acute on chronic hyponatremia, of mixed etiology. She was taking diuretic for her hypertension for long time, which might have contributed to this. This patient's sodium levels had been raised from 95.1 mmol/L 118.3 within 24 hours. Probably initial improvement followed by rapid deterioration was due to aggressive correction of hyponatremia.

Other differential diagnoses of her secondary collapse were demyelinating diseases such as multiple sclerosis or acute disseminated encephalomyelitis, infections such as meningoencephalitis, hemorrhagic or thrombo-embolic strokes as well as space-occupying lesions.

Once osmotic demyelination syndrome or central pontine demyelination has been diagnosed, the treatment is supportive[10]. The prognosis is variable but the avoidance of secondary complications is crucial. Those who survive often require

prolonged neuro-rehabilitation. In a study of 34 patients with central pontine myelinolysis, two died, and of the remaining 32, 11 fully recovered, 11 had remaining deficits but were independent and 10 were dependent [11].

Conclusion

Severe hyponatraemia carries a risk of cerebral edema with a significant mortality, but correcting it rapidly can result in disastrous condition like osmotic demyelination syndrome.

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