Extrapontine Myelinolysis: A Rare Case Report

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Abstract: Extrapontine myelinolysis is an acute demyelination involving other than central pontine regions due to rapidly fluctuating serum osmolality. Here we report a case 7 months old child presented as severe hypernatremic dehydration, resulting extrapontine myelinolysis during the correction.

Key words: Demyelination, Hypernatremic dehydration, MRI, ADC.

I. Introduction:

Extrapontine myelinolysis (EPM) is an acute, rare, demyelinating process in children caused by rapidly fluctuating serum osmolality, resulting in symmetric demyelination⁴. EPM is sporadic in nature affects male and female equally. Extrapontine myelinolysis (EPM), which can symmetrically affect the cerebellar peduncles, caudate nucleus, putamen, frontal and temporal white matter etc ⁵.

Hypernatremic dehydration is an uncommon but serious condition in newborns and young infants. Complications of hypernatremic dehydration include seizures, cerebral edema, venous sinus thrombosis, pontine myelinolysis, permanent brain damage, disseminated intravascular coagulation, acute renal failure, and even death. It is usually caused by diarrhea, improperly prepared infant formula and decreased fluid intake during diarrheal episodes⁶.

Here we present a 7 mo old baby with extrapontine myelinolysis seen during hypernatremic dehydration correction. This was confirmed by brain MRI, which is suggestive of EPM without central pontomyelinolysis. The baby recovered completely without sequelae at the time of discharge and at 1 and 3 months follow-up.

II. Case report:

A 7 months old female child presented with 5 episodes of vomiting containing indigested milk and 10-12 episodes of watery stools since one day.

She was previously healthy. There was no history of fever, blood and mucus stained stools, decreased urine output, seizures and altered sensorium. The baby was born to 2nd degree consanguineous married primi mother with uneventful antenatal, natal and post natal period. She was on mixed feed (breast milk and formula milk) with improper dilutions of formula milk since 3 months of age.

At admission, the baby was dull and lethargic. Vitals signs revealed low volume pulse with heart rate of 190/min, respiratory rate 72/min with sub-costal retractions, and SpO2 of 90% at room air. She had sunken eyes, dry mucus membrane, cool extremities, delayed skin turgor and depressed fontanel. Chest examination showed bilateral vesicular breath sounds. CVS and abdominal examination was normal.

Her serum glucose was 82mg/dl. Arterial blood gas showed p⁰ ⁹ 7.1, PaCO₂ 16.4 mmhg and HCO₃ ⁹ 5.1. Serum electrolytes revealed Na⁺ 162meq/L, K⁺ ⁹ 4.0 meq/L and Sr creatinine 1.8 mg/dL.

She was diagnosed as acute gastroenteritis with severe hypernatremic dehydration with severe metabolic acidosis. Shock was corrected with bolus of isotonic saline. Later, ½ DNS was administered for slow correction of hypernatremia.

On day 2, child had 2 episodes seizure involving right upper limb followed by lower limbs. Seizures were treated with IV Phenytoin. There was no post ictal drowsiness. Lumbar puncture was performed and CSF studies revealed normal.

MRI brain revealed altered signal intensity involving bilateral periventricular deep whitematter, centrelsemiovale, corona radiate also involving fronto-temporoparietal and left occipital subcortical U fibres with hyperintensity on DWI (figure 1 & 2) and restriction and hypointensity in ADC these features suggestive of extrapontine myelinolysis as shown in (figure 3).

Repeat MRI revealed normal which is done after 1 month and 3 months show
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III. Discussion:

Osmotic demyelination syndrome (ODS) is one of the encephalopathies that result from extreme fluctuations in serum sodium concentration and plasma osmolality. It includes both central pontine myelinolysis (CPM) and extrapontine myelinolysis (EPM).

In 1959, Adams et al. first described the pontine myelinolysis and he proved this condition to be associated with either rapid or over-correction of hyponatremia. Tomlinson in 1976 is generally credited with the suggestion that the rapidity of correction of Na+ was the aetiological factor. Brown WD et al. reported three children with severe hypernatremia and extrapontine myelinolysis involving various combinations of thalamus, basal ganglia, external and extreme capsules, and cerebellar vermis. Al Orainy IA et al. reported bilateral, symmetrical changes presumed to indicate extrapontine myelinolysis in the thalamus and globus pallidus in an infant with hypernatremic dehydration without involvement of pons.

Hypernatremia is defined as a serum sodium level greater than 145 mmol/L [mEq/L]. Hypernatremic dehydration is a rare but serious clinical condition in newborns and small infants. It is usually caused by diarrhea, improperly prepared infant formula, decreased fluid intake, or exclusive breastfeeding.

In acute hypernatremic dehydration, the extracellular fluid space becomes hypertonic relative to the intracellular space and draws fluid across the osmotic gradient out of the cells. With a prolonged elevation of plasma osmolality, however, most excess electrolytes in the brain are replaced by organic solutes called as “idiogenic osmoles”. The most important organic osmoles in the mammalian brain include myo-inositol, taurine, glycerylphosphorylcholine, and betaine.

Brain shrinkage induced by hypernatremia can cause vascular rupture, with cerebral bleeding, subarachnoid hemorrhage, and permanent neurologic damage or death. This may predispose to vascular stretching and subsequent rupture of meningeal vessels with potential risk of cerebral or subarachnoid hemorrhage and neurological deficit.

Common symptoms in infants include hyperapnea, muscle weakness, restlessness, a characteristic high-pitched cry, insomnia, lethargy, and even coma. Convulsions are not a common feature, but seizure can be consequences of rapid correction of serum sodium level in these patients in whom the rate of fluid and sodium administration are inappropriate.

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During rapid rehydration with relatively hypotonic intravenous fluids, excess water enters the cerebral cells leading to rebound cerebral edema which may result in permanent cognitive impairment, cerebral dysfunction, spastic paralysis, and seizure disorders. Restoration of serum tonicity is the primary goal in the treatment of hypernatremia. Child with shock should be treated with 0.9 % isotonic saline @ 20ml/kg over 30 minutes. If the child is not in shock, then rehydration may be commenced intravenously using 0.9 % saline at 100 ml/kg/day, followed by slow correction of serum sodium by 10-15 mEq/L/day with 0.45 % saline in 5%-10% dextrose. Underlying cause should be identified and treated accordingly. Oral rehydration solutions should be given along with breast feeds, if child was previously on breast feeding.4,14.

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References: