POST PARTUM DEMYELINATING EXTRAPONTINE ENCEPHALOPATHY SECONDARY TO HYPERNATREMIA

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Abstract
A 25-year-old female post-partum day 15 presented with high grade fever, generalized weakness, vomiting, disoriented to time place and person. On examination, patient was anaemic, febrile and stuporous. Investigations revealed hypernatremia, hyperuricemia, acute kidney injury and features suggestive of extra-pontine myelinolysis on magnetic resonance imaging (MRI) of brain. After correcting hypernatremia, there was a gradual improvement in neurological symptoms of the patient. Extrapontine myelinolysis, a fairly common metabolic disorder, is associated with neurological complications. Central pontine and extrapontine myelinolysis are commonly recognized with rapid correction of sodium. Myelinolysis, however, has rarely been described with hypernatremia. Neuroimaging findings can be crucial in diagnosing hypernatremic encephalopathy in the postpartum period.

Keywords: Demyelination, encephalopathy, extrapontine myelinolysis, hypernatremia, postpartum dehydration.

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Introduction

Hypernatremia is an electrolyte disorder where serum sodium is higher than 145 mmol/l.[1,2,3] Hypernatremia occurs due to various causes like inadequate water intake, excessive sodium intake, gastrointestinal loss, renal loss, certain drugs(like osmotic diuretics, corticosteroids),skin loss. Patient with hypernatremia presents with diverse neurological manifestations, predominantly encephalopathy which can range from impaired cognition to deep coma[4].Rapid correction of hyponatremia can lead to central pontine and extra-pontine myelinolysis with neurological complications. Myelinolysis, however, uncommonly occurs due to hypernatremia [5]. Osmotic myelinolysis due to spontaneous postpartum hypernatremia has been reported where predominantly extra-pontine lesions were observed on magnetic resonance imaging (MRI) [6]. Distribution of these MRI abnormalities is different from the osmotic demyelination syndrome due to hyponatremia.[7] We report a rare case of postpartum hypernatremic encephalopathy associated with serum sodium levels as high as 171 mmol/l which developed during the postpartum period.

Case report

A 25-year-old female with full term normal vaginal delivery live birth post-partum day 15 presented with high grade fever, generalized weakness, vomiting, stupor, irrelevant talking, not oriented to time, place and person from 1 day which was not associated with chills, rigors, abdominal pain, headache, seizures, visual disturbances, substance abuse. Patient had no history of comorbidities or major surgery in the past. The antenatal period was uneventful.

On general physical examination the patient had pallor, was febrile, had tachycardia and hypotension. She was conscious, stuporous, pupils were reactive, bilateral plantar reflex was extensor, hypertonia in all 4 limbs, muscle power 3/5 in all 4 limbs, with brisk(+1) biceps jerk, triceps jerk, supinator jerk, knee jerk, ankle jerk. Her higher mental functions could not be assessed. Her chest sounds were clear, no murmurs were heard, abdomen was soft, non-tender, and maintained saturation of 98% at room air. A possibility of abdominal infection with post-partum sepsis related encephalopathy was thought till her serum sodium level in arterial blood gas (ABG) was 170 mEq/l. Arterial blood gas analysis was repeated, which again showed serum sodium of 168 mEq/l. Subsequently, magnetic resonance imaging (MRI) brain was advised.

Her laboratory investigations showed Hb was low (3.6gm/dl), total counts were normal(4800cells/cu.mm), platelets were low(50000/cu.mm) with raised C reactive protein is 70mg/L and normal erythrocyte sedimentation rate with microcytic hypochromic anaemia on peripheral smear, no blood parasite were seen with a reticulocyte count of 1%.

She presented with elevated serum sodium levels-177mmol/L, potassium was 3.8mmol/L, chloride was 131mmol/L with hypoproteinemia, hypocalcemia, raised serum urea 192mg%, creatinine 3.6, raised liver enzymes SGOT -160 SGPT-84, raised serum LDH (1810), negative indirect Coomb's test, normal total bilirubin levels. Her uric acid was markedly elevated(16mg/dl). Her urinary electrolytes showed low sodium and chloride and normal potassium levels. Her coagulation profile was deranged. Patient was euthyroid with negative for HIV, HBsAg, HCV, HAV, HEV, Dengue IgM, IgG. Her urine routine showed albuminuria, with pus cells, RBCs, epithelial cells. Her ANA was negative. No organisms were isolated from her blood and urine cultures.

On ultrasound of abdomen and pelvis post-partum bulky uterus was noted. Her chest Xray showed clear lung fields.

Her Cerebrospinal fluid on analysis had sugar-121mg/dl, protein-161/dl, ADA-08u/L, LDH-90u/L, Total count of 2 cells/cu.mm with 100% lymphocytes.

Her MRI brain shows lesions appearing hyperintense suggestive on T2 and FLAIR seen in fornix, tail of hippocampus and few areas of bilateral cerebellar hemisphere with few areas showing on blooming on GRE involving bilateral cerebellar hemisphere. Subdural hemorrhage (2.5mm) in right posterior cerebral convexities but no history of head trauma. MRI cervical spine with whole spine screening suggestive of loss of cervical lordosis, Sacralization of L5 vertebral body noted.

On further evaluation a normal awake EEG study was recorded.

Her differential diagnosis could be Cerebral venous thrombosis, Posterior reversible encephalopathy syndrome (PRES), acute disseminated encephalomyelitis [ADEM], primary lateral sclerosis, amyotrophic lateral sclerosis (ALS), leukodystrophies, Metrogyl poisoning.

In view of MRI findings and hypernatremia, management was initiated on line of extra-pontine myelinolysis secondary to postpartum hypernatremia. Total fluid intake was kept between 2.5 and 3 litres per day. Serum sodium levels fell down from 162.44 mmol/l to 153 mmol/l over next 24 hours (day 1) and to 147 mmol/l over the next

24 hours (day 2) and her urine output increased up to 4 litres/day, though there was no improvement in the symptoms. Over this period of time, gradual correction in her serum sodium levels had also been achieved and value of 139.82 mmol/l was obtained (day 8). Patient was then discharged in a clinically stable condition. Patient visited the Medicine OPD after 15 days of discharge. At that time, her serum sodium and potassium levels were normal (Na 137.8 mmol/l, K 4.18 mmol/l) and there was no neurological deficit.

Discussion

Hypernatremia is not common in the postpartum period. Hypernatremia can lead to serious complication which can lead to encephalopathy, rhabdomyolysis and osmotic demyelination. Only 12 patients have been reported previously with postpartum hypernatremia and extra-pontine myelinolysis.*[1,13] A female developing such a high sodium level during postpartum period is uncommon in post partum period. The metabolic clearance of arginine vasopressin (AVP) is increased three- to fourfold due to placental production of a N-terminal peptidase and decreased secretion of AVP or increased clearance of AVP can lead to Diabetes insipidus which leads to hypernatremia. Our patient had limited water intake during postpartum period which is a ritual in some rural areas of India,[14] but is not recognized as a factor leading to hypernatremia. The neurological features of hypernatremia are majorly confusion, seizures, delirium and varying degrees of depression of consciousness ending in deep coma.[15] Our patient had confused state and bradykinesia.

Hypernatremia induced during experimental work in laboratory animals has been shown to lead to cellular damage and myelinolysis.[16,17] Cerebral MR spectroscopic study performed in a patient with hypernatremia showed an elevated in the osmolytes containing myo-inositol, choline, creatine and glutamate.[8] The concentration of these osmolytes reduced with gradual fall in serum sodium levels. Rapid correction of hypernatremia in the presence of increased cerebral organic osmolytes can lead to rapid intracellular shift of water and cerebral oedema causing clinical deterioration.[9]

Singular corpus callosum hyperintensities were described in rotavirus infection,[10] epilepsy, encephalitis, malignancies,[11] status epilepticus.[12] Hyperintensity of the splenium has been seen in encephalitis, epilepsy, hypernatremia and systemic malignancy. 'Wine-glass' appearance has been described in patients with hypernatremic myelinolysis which comprises extensive symmetrical T2, FLAIR and DWI hyperintensities of white matter, internal capsule through midbrain and pons to middle cerebellar peduncle.[5,8] Our findings of bilateral symmetrical hyperintensities in bilateral cerebellar hemisphere, fornix, tail of hippocampus region on MRI scan were suggestive of extra-pontine myelinolysis in our patient and are in agreement with previous studied reports. Extra pontine myelinolysis in our case might occurred due hyperosmolarity secondary to postpartum hypernatremia.

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