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Postpartum hypernatremic cerebral encephalopathy with osmotic myelinolysis: Report of two cases and review with emphasis on magnetic resonance imaging findings

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ABSTRACT

Postpartum complications are diverse. Electrolyte imbalance with hypernatremia can occur secondary to underlying postpartum complication or can arise de novo. Hypernatremia causes demyelination similar to hyponatremia but predominantly involves extra-pontine structures. Here, we present two cases, one classical and another a variant of a recently described entity called postpartum hypernatremic encephalopathy with osmotic demyelination. The more classical appearance is altered signal intensity changes in the posterior limb of internal capsules, external capsule, crus cerebri, corticopontine tracts, middle cerebellar peduncle, hippocampus, fornix, and cerebellar white matter with classical wine glass appearance on coronal T2-weighted images. The nonclassical case shows a different and atypical imaging finding of the same disease with small focal transient altered signal intensity changes in the splenium of corpus callosum. Both the patients recovered with conservative management of the electrolyte imbalance.

Key words: Demyelination, encephalopathy, hypernatremia

INTRODUCTION

Osmotic demyelination due to rapid correction of the reduced sodium levels (hyponatremia) is a known entity. This includes both pontine and extra-pontine demyelination of which extra-pontine regions predominantly being basal ganglia and thalamic regions. However, even high sodium levels can cause demyelination. Here, we present two cases of encephalopathy due to raised sodium levels in postpartum women, one with classical magnetic resonance imaging (MRI) findings and another with a subtle MRI finding. These cases represent an entity, which has been termed cerebral encephalopathy with extra-pontine myelinolysis in postpartum hypernatremia.

CASE REPORT

Case 1
A 26-year-old female presented with altered sensorium on the 4th postpartum day. On examination, she was deeply comatosed, and there was decreased responsiveness and brisk deep tendon reflexes. She was afebrile, and blood pressure recording was 110/70 mm of Hg. Computed tomography head was reported as normal. Complete blood count, renal function test, thyroid, and liver function tests were normal. Serum sodium levels on arrival were 160 mEq/L and presumed to be due to dehydration. MRI performed on a 1.5 Tesla system (Magnetom Avanto, Siemens, Germany) revealed the following findings as displayed in Figure 1. Bilateral symmetrical T2-weighted/fluid-attenuated inversion recovery sequence (FLAIR) hyperintensity [Figure 1] involving posterior limb of internal capsules, external...
capsule, crus cerebri, corticopontine tracts, middle cerebellar peduncles, hippocampus, fornix, cerebellar white matter, corpus callosum body, and splenium extending into forceps major. These areas showed restricted diffusion [Figure 2] and appeared hypointense on apparent diffusion coefficient (ADC) images. There was no abnormal enhancement of the lesion. There was no venous sinus thrombosis. As the differential of meningitis was not considered, a lumbar puncture was not performed. As the renal function levels were normal, there was no suspicion of rhabdomyolysis and CPK levels were not evaluated. Gradual correction of the hypernatremia was performed with 5% dextrose and free water to reduce sodium at not more than 12 mEq/L/day. No intervention with steroids or immunoglobulins was performed. There was gradual clinical improvement during her hospital stay. She was referred to a higher center where she was reviewed, and the same diagnosis was adhered to. Finally, after 4 weeks of hospital stay she recovered completely.

Case 2
Another young adult female (aged 24) patient presented in the same month with features of altered sensorium and one episode of seizures. She was in her 5th postpartum day. Antenatal and natal periods were uneventful. On admission, she had tachycardia and blood pressure was within normal limits. Complete blood routine and electrolyte workup was done to rule out sepsis and electrolyte imbalance, respectively. The electroencephalogram was normal. MRI performed on a 1.5 Tesla system (Magnetom Avanto, Siemens, Germany) revealed T2-weighted/FLAIR hyperintensity in the splenium of corpus callosum which showed restricted diffusion appearing bright on diffusion b-1000 images and had reduced ADC values [Figure 3]. There was no adjacent mass effect and no postcontrast enhancement. There were no other altered signal intensity changes in the rest of the neuro-parenchyma. As the sodium levels were more than 180 mEq/L, the diagnosis of postpartum hypernatremic encephalopathy was considered. The possibility of the posterior reversible encephalopathy syndrome (PRES) was also considered as a differential. The possibility of neuromyelitis optica or multiple sclerosis was not considered. The patient was treated conservatively, and the sodium levels were monitored and slowly correction of sodium levels was done in the same manner as the previous case. The patient improved symptomatically. A repeat MRI was done 7 days later, which showed resolution of the signal intensity changes in the splenium of corpus callosum [Figure 4]. The sodium levels at this point of time were 140 mEq/L.

DISCUSSION

Hypernatremia is considered when the sodium levels are >145 mEq/L.\textsuperscript{4} Hypernatremia causes central nervous system insult similar to hyponatremia ranging from cognitive dysfunction to serious autonomic and brainstem injury.\textsuperscript{5} It can cause varied neurological manifestations such as seizures, flaccid quadriapresis, dysarthria, dysphagia, pupillary and oculomotor abnormalities, which indicate pontine involvement. Patients having hypernatremia can also present with features of movement disorders such as parkinsonian features and myoclonic jerks. This type of clinical presentation points toward involvement of extra-pontine structures such as basal ganglia, thalamus, and subthalamic regions.\textsuperscript{1,5}

Figure 1: Magnetic resonance axial T2-weighted images showing bilateral symmetrical hyperintensity involving posterior limb of internal capsules (a: Short black arrows), external capsule (a: Long black arrow), hippocampus, fornix, corpus callosum body, and splenium extending into forceps major (b: Black arrow, c), corticopontine tracts and crus cerebri (d and e: Black arrow), middle cerebellar peduncles (f: Black arrow), cerebellar white matter

Figure 2: Magnetic resonance axial diffusion weighted images showed the corresponding T2-weighted/fluid-attenuated inversion recovery hyperintense areas, which showed restricted diffusion (a-c: White arrows) appearing bright and appeared hypointense on corresponding apparent diffusion coefficient mapping (d-f)
The mechanism by which increased sodium levels cause demyelination has been explained based on the theory that hyperosmolality in the extracellular compartment of the brain causes imbalance in the homeostatic mechanism, thereby altering the intracellular components within the oligodendrocytes rendering them nonfunctional. This leads to myelin degradation.[6]

Electrolyte imbalances in puerperium can occur due to complications such as postpartum hemorrhage, infections, endocrine disturbances such as thyroiditis and reduced water intake. Naik and Saroja published a case series of 11 postpartum patients presenting with encephalopathy secondary to hypernatremia and showed signal intensity changes involving the corpus callosum, internal capsule, corona radiata, and cerebellar peduncles.[3] Similar findings are present in our first case [Figure 1] with classically described “wine glass” appearance being seen on coronal T2-weighted images. This particular imaging finding is seen because of selective involvement of the corticospinal tracts.[7] This type of demyelination appearance has also been described for amyotrophic lateral sclerosis and some of the leukodystrophies where the etiopathology is completely different.[8,9] This combination of clinical history, finding of raised sodium levels, and MRI findings are specific for this entity.

Our second case showed a focal area of T2-weighted/FLAIR hyperintensity with diffusion restriction in the splenium of corpus callosum. There were no other areas of altered signal intensities and corticospinal tracts were spared. This is a variant in the manifestation of hypernatremic encephalopathy. As the patient symptomatically improved, a repeat MRI was done at 7th-day post-admission where there was resolution of the signal intensity changes of splenium [Figure 4]. Hence, it can be implied that follow-up MRI can be also useful in assessing the progression or regression of brain changes due to hypernatremia.

There are many causes of transient lesions in the splenium of corpus callosum, which include hypernatremia among the various metabolic causes.[10] As the sodium levels were very high at the time of admission, a diagnosis of postpartum hypernatremic encephalopathy was diagnosed in this case.

Rapid correction of hyponatremia leads to pontine and extra-pontine myelinolysis,[2] where the typical MRI findings have been commonly reported. Irrespective of dominant hypernatremia or hyponatremia the rapid correction of dyselectrolemias can result in osmotic demyelination. In both the above cases, possibilities such as PRES and cerebral venous thrombosis were ruled out by imaging. Both cases showed normal magnetic resonance venogram. PRES (also known as hypertensive encephalopathy) in pregnant patients is usually a sequel of preeclampsia or eclampsia. The imaging findings in PRES is predominant involvement of posterior occipital and also anterior subcortical and deep white matter with occasional postcontrast enhancement.[11,12] Transient T2-weighted/FLAIR hyperintensity with diffusion restriction in the splenium of corpus callosum is also seen in PRES.[10] However, as the sodium levels were high and the patient (case 2) was normotensive at the time of admission with no history of preeclampsia/eclampsia during the course of pregnancy, diagnosis of hypernatremic encephalopathy with osmotic demyelination is reasonable.
CONCLUSION

Here, we present two cases of a rare entity, documented in recent times as postpartum hypernatremic encephalopathy with osmotic demyelination. The first case demonstrates typical features and second case reveal an atypical imaging aspect of this pathologic condition. As both of our patients recovered completely after conservative management, this treatable ailment should be a differential diagnosis in patients presenting with cerebral symptoms in postpartum status. The MRI findings are typical and point toward this diagnosis.

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