

# Cerebral encephalopathy with extrapontine myelinolysis in a case of postpartum hypernatremia

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## Abstract

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. We report a rare case of post-partum hypernatremic encephalopathy associated with serum sodium levels as high as 200 mEq/l on presentation. Although the serum sodium levels were brought down gradually, subsequent imaging showed progression of demyelination and deterioration of the neurological status.

**Key words:** Demyelination; encephalopathy; extrapontine myelinolysis hypernatremia; postpartum

## Introduction

Hypernatremia is a potentially lethal condition, and can cause encephalopathy, rhabdomyolysis, and osmotic demyelination.<sup>[1]</sup> 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. We report a rare case of post-partum hypernatremic encephalopathy associated with serum sodium levels as high as 200 mEq/l on presentation.

## Case Report

A 26-year-old female delivered a healthy child by normal vaginal delivery. Postpartum, she was in good health for

2 weeks, when she started having complaints of generalized weakness, inability to walk, and altered sensorium which persisted for 2-3 days and gradually progressed till she presented to the hospital. On admission, she was drowsy, agitated, and had tachycardia. A possibility of abdominal infection with post-partum sepsis related encephalopathy was thought till her serum sodium level in arterial blood gas (ABG) was 204 mEq/l. ABG analysis was repeated, which again showed serum sodium of 204 mEq/l. Subsequently, magnetic resonance imaging (MRI) brain was advised.

MRI of the brain revealed confluent symmetrical abnormal increased T2-weighted (T2W) and fluid attenuated inversion recovery (FLAIR) signal involving cerebellar white matter, middle and superior cerebellar peduncles, cerebral peduncles, medial midbrain, posterior limb of internal capsule, external capsules, corona radiata, medial thalami, and dorsal aspect of pons [Figure 1]. Involvement of bilateral posteromedial temporal lobes, mid body, and splenium of the corpus callosum extending into the forceps major bilaterally was seen. Restricted diffusion was noted within the lesions with hypointensity on apparent diffusion coefficient (ADC) images [Figure 2]. No evidence of hemorrhage or mass effect was seen. Contrast enhanced MR venography was also done which did not reveal any evidence of venous sinus thrombosis.

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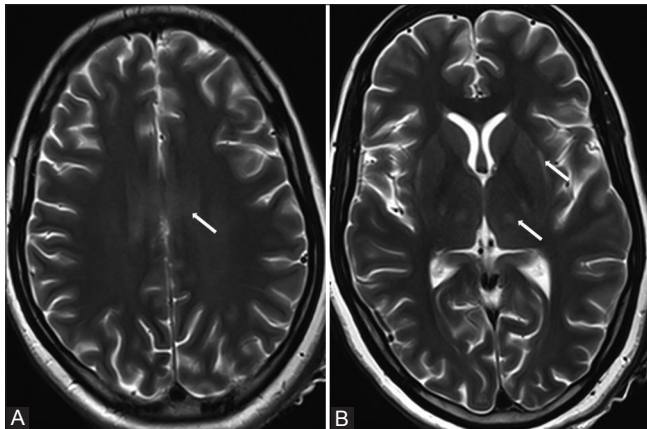
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She developed breathlessness with oxygen desaturation and was put on mechanical ventilation. Her creatinine phosphokinase (CPK) levels were high suggesting rhabdomyolysis in the given setting. Subsequently, gradual correction of hypernatremia was done. Her sodium level after 7 days of IV fluid therapy was 149 mEq/l, but sensorium had deteriorated. A repeat MRI was done after

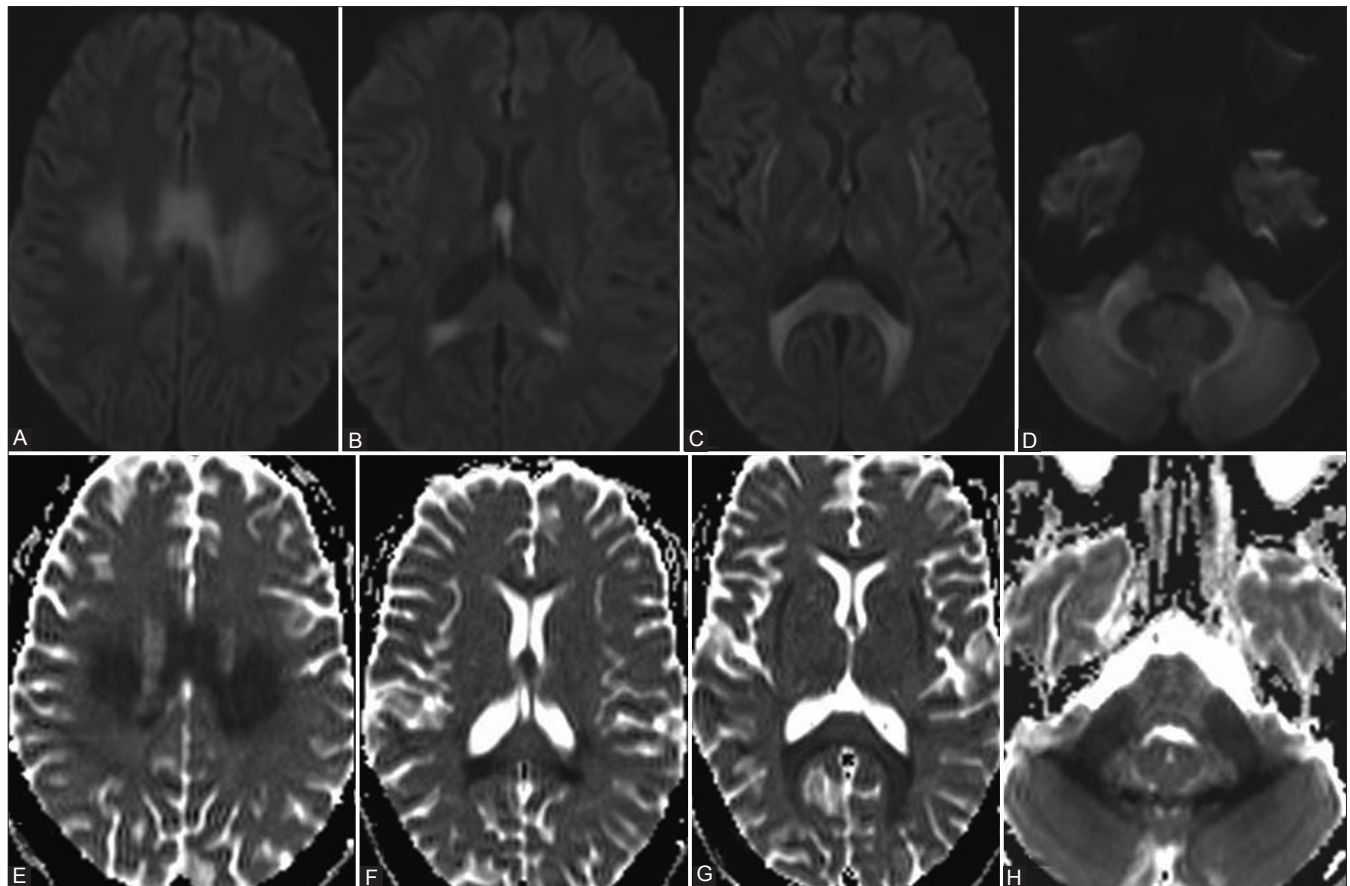
1 week when serum sodium was brought to normal level. It revealed progression of the lesions with involvement of cortex and subcortical white matter in the bilateral frontoparietal regions with persistent diffusion restriction [Figure 3]. The patient stayed in the hospital for 3 weeks without any clinical improvement and got discharged against medical advice.

### Discussion

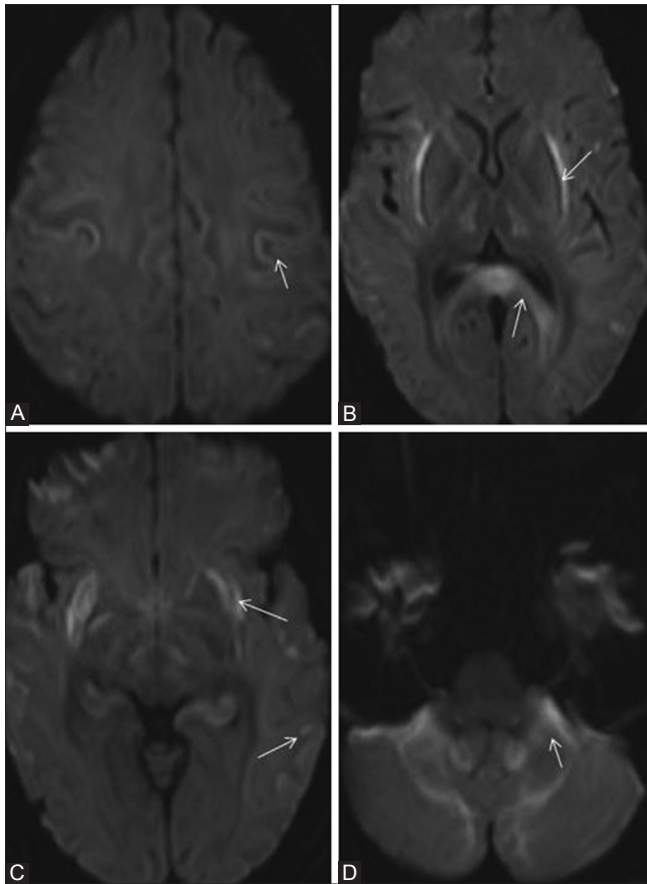
Hypernatremia is quite uncommon in the post-partum period. Hypernatremia, a potentially lethal condition, can cause encephalopathy, rhabdomyolysis, and osmotic demyelination.<sup>[1]</sup> A case series of 11 patients has previously been reported, where serum sodium levels were high. An otherwise healthy female developing such a high sodium level is unusual. Even lowering the sodium at 8-10 mEq/day did not help improve her sensorium. It is known that sudden lowering of sodium (i.e., more than 10 mEq/day) may affect brain function and myelinolysis is expected, but such a high level of sodium once achieved is always detrimental to the brain tissue.<sup>[2-4]</sup> Even lowering sodium at a slow rate would not help the neurological status to improve.<sup>[5]</sup> As in our report, MRI brain revealed progressive



**Figure 1 (A and B):** MRI reveals hyperintense signal involving bilateral para-sagittal parietal regions, caudate nuclei, external capsules and thalami (arrows) on axial T2W Images (1A and B)



**Figure 2 (A-H):** Diffusion weighted images reveal restricted diffusion with hyperintense signal involving bilateral para-sagittal parietal regions, body and splenium of corpus callosum, external capsules, thalami and cerebellar peduncles (2A - 2D). Corresponding ADC images show corresponding decreased signal in the above mentioned lesions (2E-2H)



**Figure 3 (A-D):** MRI done after 1 week, diffusion weighted images reveal progressive restricted diffusion with hyperintense signal (arrows) involving parietal-temporal cortex and subcortical white matter, body and splenium of corpus callosum, external capsules, thalami and cerebellar peduncles. (3A-3D)

lesions with cortical and subcortical demyelination even after the hypernatremic state was corrected. The prognosis remains poor for such patients. EEG findings are suggestive of generalized slowing and decreased amplitude of the waves.

Neurological complaints secondary to extrapontine myelinolysis (EPM) and rhabdomyolysis caused by hypernatremia are infrequently reported. Only a case series with 11 patients has been reported previously in postpartum period.<sup>[1]</sup> Although the exact etiological factor is unknown, a ritual in some communities in North India, that is, to restrict the water intake for females in post-partum period, may exacerbate dehydration.<sup>[1]</sup> This is considered as the cause of hypernatremia. Another case has been reported in literature where hypernatremia secondary to hunger strike resulted in neurological deterioration.<sup>[6]</sup> Patients present with seizures and altered sensorium. Prompt identification and appropriate management can improve outcome in these patients. We acknowledge that information concerning imaging features of EPM remains scarce and prevents accurate diagnosis. Here, we discuss the imaging features of

this rare case of extrapontine demyelination caused secondary to postpartum hypernatremia. MR imaging plays the most important role in the initial diagnosis. We also discuss the differential diagnosis based on the imaging features.

MR imaging findings of osmotic demyelination syndrome consist of abnormal hyperintensity involving pons and extrapontine sites including basal ganglia, thalami, and cerebral white matter. Symmetric trident-shaped hyperintensity in the central pons is a characteristic finding with sparing of ventrolateral pons and the pontine portion of corticospinal tracts.<sup>[7]</sup> Typical sites of involvement of EPM associated with rapid correction of hyponatremia include caudate nucleus, lentiform nucleus, and thalami. The lesions exhibit minimal to no mass effect and show no enhancement in most of the cases. Restricted diffusion is seen in most of the cases. Atypical sites of involvement include grey matter, white matter, corpus callosum, splenium, cerebellum, hippocampus, and external capsule. Other rare sites include midbrain, subthalamic nuclei, claustrum, hypothalamus, medulla, and amygdala.

One previous study done on postpartum hypernatremic patients showed involvement of corpus callosum in all patients.<sup>[1]</sup> Ten patients underwent MRI which revealed hyperintensity of corpus callosum in all the patients in T2, FLAIR, and diffusion weighted sequences (DWI). Symmetrical hyperintensities were also seen in internal capsule, corona radiata, cerebellar peduncles, and hippocampus in various combinations. Our patient also showed involvement of corpus callosum.

“Wine-glass”-appearance has been described in patients with hypernatremic myelinolysis which consists of extensive symmetrical T2, FLAIR, and DWI hyperintensities of white matter, internal capsule through midbrain and pons to middle cerebellar peduncle.<sup>[8]</sup> It depicts the involvement of corticospinal tract. Symmetrical hyperintensities of internal capsule, crus cerebri, and pons on coronal T2W images are seen. Similar appearance has also been described in entities like primary lateral sclerosis, amyotrophic lateral sclerosis (ALS), and leukodystrophies.

Metrogyl poisoning, an entity described with similar imaging findings, manifests after metronidazole intake for more than 2 weeks. MR imaging of brain demonstrates abnormal symmetrical hyperintensity within cerebral white matter, corpus callosum, and cerebellum.<sup>[9]</sup> Near total resolution of findings is seen on discontinuation of medication.

In contrast, acute disseminated encephalomyelitis [ADEM] presents with lesions predominantly in cerebral or cerebellar cortices, subcortical and parieto-occipital white

matter, centrum semiovale, cerebellar peduncles, and the brainstem.<sup>[10]</sup> The lesions are large, asymmetric, irregular in morphology, and regress dramatically with short course steroids, immunoglobulins, or plasmapheresis. Associated involvement of spinal cord is also seen. Sometimes, a “fried-egg” appearance with central rounded hyperintensity on T2W images corresponding to “egg yolk” is seen.<sup>[10]</sup>

Posterior reversible encephalopathy syndrome (PRES) is a remarkably heterogeneous group of disorders seen in patients with hypertensive encephalopathy, pregnant patients with eclampsia, and post-transplant population on cyclosporin A and tacrolimus.<sup>[11]</sup> It is also seen in patients with uremia, thrombotic thrombocytopenia purpura, and hemolytic-uremic syndrome. CT and MR imaging typically show symmetrically distributed areas of vasogenic edema predominantly within posterior circulation territories involving occipital and parietal lobes; however, involvement of anterior circulation structures is also common. Conversion to irreversible cytotoxic edema has also been described. The abnormalities primarily affect white matter, but cortex may also get involved.<sup>[11]</sup> Localized mass effect and mild enhancement may be associated.

Cerebral venous thrombosis, another common condition associated with peripartum state, has been described as a differential.<sup>[12]</sup> The absence of a flow void and the presence of altered signal intensity in the sinus is a primary finding of sinus thrombosis on MR images. The signal intensity of venous thrombi on MR imaging varies according to the interval between onset of thrombus formation and time of imaging. Parenchymal lesions, focal edema, venous infarcts, and hemorrhages are better depicted and more commonly identified at MR than at CT.

In summary, this rare case of EPM is instructive from the perspective of imaging based primary diagnosis of demyelination.

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