Electromyographic and histological features of postpartum hypernatremic rhabdomyolysis

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Abstract

Introduction: Rhabdomyolysis results from many causes including hypernatremia. Postpartum hypernatremia with osmotic cerebral demyelination is a rare cause of reversible rhabdomyolysis. Electromyographic studies in postpartum hypernatremia have not been reported. Materials and Methods: Electromyography (EMG) was performed in five women with postpartum hypernatremia and muscle biopsy was performed in one of them. Results: Among the five women presenting with postpartum hypernatremia associated with marked elevation of serum creatine kinase, four had quadriplegia. All had varying degrees of encephalopathy at admission and recovered without residual deficits after gradual correction of hypernatremia. Needle EMG revealed fibrillations with positive sharp waves in five patients and myotonic discharges in three patients. Serial EMG in one patient revealed the occurrence of transient fibrillations, positive sharp waves and myotonic discharges. Muscle biopsy revealed extensive rhabdomyolysis in one patient. Conclusion: EMG in hypernatremic rhabdomyolysis revealed spontaneous activity including fibrillations, positive sharp waves and myotonic discharges along with myopathic potentials. Electromyographic findings depend on the interval from the onset and the degree of rhabdomyolysis.

Key Words
Electromyography, hypernatremia, myotonic discharges, rhabdomyolysis, postpartum

Case Reports

Five patients with postpartum hypernatremic encephalopathy admitted under neurological services underwent nerve conductions and concentric needle electromyography (EMG). Their salient clinical, biochemical and EMG findings are shown in the Table 1. All patients had poor water intake during the postpartum period preceding the symptom onset. They had dehydration, hyperchloremic hypernatremia, azotemia, moderate thrombocytopenia and elevated serum creatine kinase (CK) levels at admission. Hypernatremia was gradually corrected with hypotonic intravenous and enteral fluids. Nerve conductions were normal in all patients. Cerebral magnetic resonance imaging revealed features of extrapontine myelinolysis with involvement of the corpus callosum and internal capsule in four patients; three had hyperintensity in hippocampus.

Patient 1
A 20-year-old lady was admitted 2 weeks after first delivery with progressively declining word output for 10 days. She was disoriented for 3 days before admission with oliguria. She had normal tone and power with brisk tendon reflexes. EMG of right upper and lower limb muscles revealed fibrillations and positive sharp waves. Muscle power remained normal during...
Table 1: The clinical features, biochemical and electromyographic findings in patients with hypernatremic rhabdomyolysis

| Patient 1 | Patient 2 | Patient 3 | Patient 4 | Patient 5 |
|-----------|-----------|-----------|-----------|-----------|
| Onset after delivery (days) | 5 | 10 | 14 | 8 |
| Symptom duration (days) | 5 | 10 | 1 | 7 |
| Encephalopathy (days) | 5 | 3 | 10 | 4 |
| History of weakness (days) | None | 5 | 10 | None |
| Follow-up duration (months) | 19 | 18 | 8 | 7 |
| Glasgow coma score | 14 | 11 | 10 | 4 |
| Tone | Normal | Normal | Reduced | Normal |
| Power Proximal | 5/5 | 1/5 | 1/5 | 1/5 |
| Distal | 5/5 | 3/5 | 3/5 | 2/5 |
| Tendon reflexes | Brisk | Normal* | Sluggish* | Normal* |
| Release reflexes | Present | Present | Present | Absent |
| Urea (mg/dl) | 46 | 61 (36) | 167 (30) | 53 (14) |
| Creatinine (mg/dl) | 0.8 (0.5) | 1.5 (0.56) | 3.4 (1.3) | 2.1 (0.4) |
| Sodium (mEq/L) | 154 (147) | 172 (143) | 192 (139) | 158 (137) |
| Potassium (mEq/L) | 3.9 | 3.6 | 3.4 | 3.0 |
| Fibrillation | + | ++ | + | + |
| Positive sharp waves | ++ | +++ | + | ++ |
| Myotonic discharges | 0 | + | + | 0 |
| Motor unit potentials | Normal | Myopathic | Myopathic | Myopathic |

*Tendon reflexes became brisk during recovery of quadriparesis, **During second EMG study 13 days after symptom onset, †Lost from follow-up. The biochemical values in parenthesis are at discharge and the durations are in days. Normal value for serum sodium=135-145 mEq/L, Potassium=3.0-5.0 mEq/L, Urea=15-40 mg/dl, Creatinine=0.4-1.0 mg/dl

hospita; serum CK was 6,139 units/l at admission (normal range: 50-200. units/l); 4,175 units/l on the day of EMG and declined to 149 units/l at 1 month follow-up. Subsequently, she had uneventful second pregnancy and puerperium 1 year later.

Patient 2
A 25-year-old lady was admitted 3 weeks after fourth delivery with 5 days progressive quadriparesis and disorientation with drowsiness for 3 days. She was bedbound and incontinent for 2 days and had symmetric proximally dominant quadriparesis and during recovery was found to have ataxia. Serum CK was 58,587 units/l at admission and reduced to 917 units/l on 15 days from the onset of symptoms when needle EMG was performed. EMG revealed moderate amount of fibrillation and positive sharp waves, which was prominent in proximal muscles. Myotonic discharges were seen in biceps brachii, quadriceps and tibialis anterior [Figure 1]. Left biceps brachii muscle revealed features of extensive rhabdomyolysis, a few regenerating fibers and areas of perivascular infiltration as shown in Figure 2. Clinical evaluation was normal at 1 month follow-up and serum CK level was 90 units/l.

Patient 3
This 18-year-old lady was admitted 16 days after her first delivery with progressive quadriparesis and irrelevant speech that started 6 days after delivery. She had symmetrical severe quadriparesis with hypoactive tendon reflexes at admission. Tendon reflexes were noted to be brisk when the weakness improved. Serum CK was 11,288 units/l at admission and declined to 144 units/l on 10th day after symptom onset when EMG revealed fibrillations and positive sharp waves in limb and paraspinal muscles. Myotonic discharges were seen in biceps brachii and extensor digitorum brevis. Evaluation at one month after discharge was normal including serum sodium and CK levels.

Patient 4
A 20-year-old primiparous lady was admitted with status epilepticus, which occurred 2 weeks after uneventful delivery. Blood pressure was 110/80 and the Glasgow coma scale was 4/15. She was found to have proximally dominant flaccid quadriparesis when she became conscious following seizure control. Serum CK was 15,122 units/l on the 2nd day of admission. EMG performed 7 days later revealed moderate amount of fibrillations and positive sharp waves in biceps brachii, quadriceps and tibialis anterior. Limb muscle weakness had become nearly normal at the time of EMG and serum CK level had reduced to 7,559 units. She had mild limb weakness with bilateral cerebellar ataxia at discharge and was lost from follow-up.

Patient 5
This 19-year-old lady presented 15 days after her first delivery with 7 days progressive quadriparesis with subsequent occurrence of altered behavior, impaired memory and disorientation for 4 days. Serum CK was 5,484 units/l at admission and reduced to 1,276 units next day. EMG performed on second hospital day (8 days from the onset of symptoms) revealed short duration motor unit potential (MUPs) with incomplete interference. Second EMG study 5 days later revealed fibrillations and infrequent myotonic discharges in biceps brachii and quadriceps. Third EMG evaluation 5 days after the second study revealed short duration MUPs without spontaneous activity. Neurological examination at discharge and 1 month follow-up were normal. Serum CK was 357 units/l at 1 week after discharge.

Discussion
Rhabdomyolysis is a clinical and biochemical syndrome resulting from necrosis of striated muscle fibers.[1-3] Features of
Serum levels of aminopeptidase of placental origin are elevated during pregnancy and decline to normal levels by compensatory increase in secretion from the pituitary. Relative reduction of activities of daily living may lead to failure to conserve water through excessive renal water loss and hypernatremia. In addition, cerebral osmotic demyelination could have contributed to the presence of reduced thirst sensation despite the presence of hyperosmolar state. The custom of restricting water intake in women during the postpartum period in some parts of Southern India could have contributed to worsening of hypernatremia and was present in all the five women in this study. Though the first patient in the present study did not have weakness, she had markedly elevated CK levels suggesting rhabdomyolysis. While rhabdomyolysis could have been caused by recurrent seizures in the fourth patient, presence of hypernatremia with features of osmotic demyelination syndrome favors hypernatremia as the causative feature for the clinical syndrome. All the five patients in the present report conform to the features of postpartum hypernatremic rhabdomyolysis with osmotic cerebral demyelination.

Sodium and potassium concentrations across the sarcoplasmic membrane are maintained by the energy dependent Na-K Adenosine triphosphatase, which could get disrupted in the presence of hypernatremia. In addition, hypernatremia alters the transmembrane sodium gradient. These factors can increase the intracellular sodium concentration leading to increase the intracellular calcium levels, which increases the intracellular proteolytic activity and lead to rhabdomyolysis.

Electromyographic findings in muscle disorders including dystrophies, inflammatory myopathies, endocrine myopathies, metabolic myopathies, etc., are well-documented. However, the literature on EMG findings in rhabdomyolysis is relatively sparse. EMG was available in 60 of the 475 patients with rhabdomyolysis among whom 33 had fibrillations and positive sharp waves; 22 had “non-irritable myopathy” and five patients had normal EMG study. EMG in a boy with non-traumatic rhabdomyolysis revealed myopathic features without spontaneous activity. Five of 15 patients in the acute phase of rhabdomyolysis had myopathic potentials and none had spontaneous discharges. Fibrillations positive sharp waves, myopathic potentials and myotonic discharges were demonstrated in patients experiencing statin-induced rhabdomyolysis.

All five patients with postpartum hypernatremic rhabdomyolysis studied in second and third week after the onset of weakness had fibrillation potentials and positive sharp waves. Myotonic discharges were seen in three patients and myopathic potentials in five patients. During the serial EMG evaluation in the patient five, fibrillations and myotonic discharges appeared only during the second EMG study performed 13 days after the onset of weakness and disappeared when the weakness improved along with normalization of serum CK level. Timing of EMG study could therefore affect the findings observed in rhabdomyolysis and possibly could have contributed to relatively low frequency of EMG abnormalities reported in some of the earlier studies. Pathogenesis of electromyographic spontaneous activity in rhabdomyolysis could be similar to that seen with inflammatory myopathies wherein there is focal myonecrosis and the electromyographic needle placed away from the endplate zone can record fibrillation potentials and positive sharp waves.
Muscle histology in primary rhabdomyolysis may not reveal specific findings.[2] Muscle biopsy in a boy with non-traumatic rhabdomyolysis revealed myonecrosis, interstitial and perivascular inflammatory cells along with few regenerating fibers similar to the findings in our patient.[4] The presence of inflammatory cells in our patient could be secondary to myonecrosis rather than a primary process.

**Conclusion**

Postpartum hypernatremia is a rare cause of rhabdomyolysis and encephalopathy. Electromyographic evaluation of hypernatremic rhabdomyolysis revealed denervation potentials and myotonic discharges. Further studies are needed to evaluate the pathogenesis of hypernatremic rhabdomyolysis and serial electromyographic findings in hypernatremic rhabdomyolysis.

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