Extrapontine Myelinolysis: A Case Report

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Osmotic demyelination usually occurs in patients with rapid correction of hyponatremia. It is a disease of the central nervous system and can involve the pontine or extrapontine areas. It is usually irreversible and can only be managed by prevention. The rate of increase in the plasma sodium level per hour and within twenty four hours are the key points for preventing osmotic demyelination. We report a 37-year old man in whom osmotic demyelination occurred in the extrapontine area (extrapontine myelinolysis) after rapid correction of hyponatremia.

Key words: extrapontine, hyponatremia, osmotic demyelination

Case Report

A 37-year-old man with a history of depressive disorder was sent to our emergency room because of vomiting and drowsiness for 3 days. He did not have other systemic diseases. He had taken antidepressants for the depressive disorder for about one decade. Physical examination revealed mild drowsiness but he could be aroused (GCS E3V4M4). The upper and lower limbs had generalized mildly decreased muscle power and mildly decreased deep tendon reflexes. The brain stem reflexes were all normal. His skin was dry and his ankles were not edematous. His blood pressure was 134/82 mmHg, pulse rate 92 beats per minute, respiratory rate 19 per minute, and body temperature 36.8°C. No other abnormalities were found on physical examination. The laboratory data were blood hemoglobin 12.6 gm/dl, hematocrit 36.4%, white cell count 13,900 cells/μL, blood urea nitrogen 3 mg/dl, creatinine 0.7 mg/dl, sodium 110 mmol/L, potassium 3.0 mmol/L, chloride 73 mmol/L, and osmolality 230 mosmol/kg. The
initial impression was hypo-osmolar hyponatremia.

Because of drowsiness, 3% saline was injected in at a rate of 30 ml per hour to correct hyponatremia. He became alert after 3 hours. The ensuing plasma sodium levels were 119 mmol/L 6 hours later, 127 mmol/L 15 hours later, 126 mmol/L 20 hours later, 135 mmol/L 36 hours later, and 130 mmol/L 48 hours later. (Table) The 3% saline was discontinued in 6 hours and was changed to normal saline and 5% dextrose water according to the plasma sodium level, to lower the rate of increase of the sodium level.

After 3 days, he became stuporous (GCS E2V3M4) and had seizures. The seizures began with convulsions first in the left limbs, and then in the right limbs 1 minute later. He had 3-4 seizures per hour, each lasting about 3 minutes. Between seizures, he was stuporous and could not be aroused. Brain stem reflexes were intact and his vital signs were within normal limits. Computed tomography of the brain showed no abnormalities and a cerebrospinal fluid study was normal. The seizures decreased gradually over the ensuing 3 days and no seizures were noted after 5 days. However, he was still stuporous. A T1-weighted magnetic resonance image of the brain showed normal signal intensities in the striatum, thalamus, and cerebral cortex bilaterally but no abnormalities in the brain stem including the pons (Figure). The differential diagnosis from the magnetic resonance imaging included extrapontine myelinolysis with cortical laminar necrosis, hypoxic-ischemic encephalopathy, or encephalitis.

The clinical course and radiological findings were compatible with extrapontine myelinolysis.

After 2 months, he gradually became more alert, but he could only respond to some visual and vocal stimulation and had abnormal hyperkinetic involuntary movements in the face and limbs.

**Discussion**

CPM was described by Adams and colleagues in 1959. In 1976, pontine and extrapontine myelinolysis were first found to be associated with rapid correction of low serum sodium levels\(^{(1-2)}\).

The sequelae of rapid correction of hyponatremia usually follow a biphasic clinical course, with initial encephalopathy or seizures from hyponatraemia, followed by a rapid recovery as normonatremia is restored, with deterioration several days later. In CPM, the initial signs include dysarthria and dysphagia (secondary to corticobulbar fiber involvement), and flaccid quadripleague (from corticospinal tract involvement) which later becomes spastic, all from involvement of the basis pontis. If the lesion extends into the tegmentum of the pons, pupillary and oculomotor abnormalities may occur. There may be changes in the level of consciousness\(^{(4)}\). EPM has identical pathological findings but the involved areas are extrapontine, so there are different clinical manifestations\(^{(5)}\). Because EPM is rare, its manifestations continue to attract study. It may manifest as postural limb tremors, myoclonic jerks, a parkinsonian picture, catatonia, dystonia, or

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<th>Time (hours)</th>
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pyramidal dysfunction (secondary to the different areas involved). These may resolve completely or partially over months or they can become permanent. The most common causes of hyponatremia are therapy with thiazides, syndrome of inappropriate secretion of antidiuretic hormone in postoperative state, polydipsia in psychiatric patients, gastrointestinal fluid loss, ingestion of dilute fluid, and accidental ingestion of excessive water. These cases are mostly hypotonic hyponatremia, as seen in our patient. The most important aspect of hyponatremia management is a proper rate of correction. In the emergency setting, no matter what the cause of hyponatremia, the rate of correction depends on the absence or presence of neurologic dysfunction. When hyponatremia develops rapidly and is accompanied by neurologic symptoms, more rapid correction is required. When hyponatremia develops slowly, slow correction is needed. In general, acute symptomatic hyponatremia should be treated by hypertonic saline, and the plasma sodium level should be raised by only 1-2 mmol/L/hour and no more than 8 mmol/L during the first 24 hours. In chronic asymptomatic hyponatremia, the plasma sodium level should be raised more slowly, and no more than 8 mmol/L during the first 24 hours. The rate of increase in the sodium level is usually rapid if 3% saline is used intravenously, but it can also occur rapidly with only intravenous normal saline if the initial sodium level is profoundly low. If the sodium level increases too rapidly, intravenous dextrose water or oral water can be administered to re-lower the sodium level to a “safe” level. In our patient, the rate of correction was 1.5 mmol/L/hour in the initial 6 hours and around 20 mmol/L in the first 24 hours; both were beyond the safe level. Although re-lowering the sodium level was attempted, osmotic demyelination still occurred.

In conclusion, hyponatremia is a manifestation of various disorders that are faced and treated in everyday clinical practice. However, if attention is given to the benefits and harm of management of hyponatremia, osmotic demyelination is preventable. We suggest that correction of hyponatremia should not exceed a rate of 1-2 mmol/L/hour and no more than 8 mmol/L/day. During treatment for hyponatremia, especially when using intravenous 3% saline, the plasma sodium level should be checked frequently.
If the sodium level increases too rapidly, water can be administered intravenously or orally to re-lower the sodium level.

References


橋腦外滲透性脫髓鞘症：一病例報告

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滲透性脫髓鞘症一般是發生在低血鈉症矯正過快所致，此病症通常影響中樞神經系統，可分為橋腦及橋腦外兩種。此症一但發生，一般為不可逆性，且無特殊有效治療。因此，預防其發生甚為重要。每一個小時血鈉上升的速度及前二十四小時血鈉上升的速度為預防脫髓鞘症發生的主要關鍵。我們報告一位三十七歲男性因為血鈉矯正過快所導致的橋腦外滲透性脫髓鞘症。

關鍵詞：橋腦外，低血鈉症，滲透性脫髓鞘症