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Extra pontine myelinolysis in a tetraplegic patient: Case report

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Hyposmolar hyponatremia (serum sodium < 130 mmol/l) is a common phenomenon in the spinal cord injury (SCI) patient population and in most cases, it is of relatively little consequence. However, rapid correction or over correction of hyponatremia (a change in serum sodium > 25 mmol/l within 48 h) has been linked to Central Pontine Myelinolysis (CPM) and Extra Pontine Myelinolysis (EPM), usually along with other recognized predisposing factors. We report the first case of isolated Extra Pontine Myelinolysis in an SCI patient without any of the recognized predisposing factors, following correction of hyponatremia. The signs and symptoms of Extra Pontine Myelinolysis were not very remarkable in our patient because of prior spinal cord injury. The diagnosis was confirmed by the typical finding of myelinolysis in the basal ganglion region on MRI. Hyponatremia occurs frequently in the SCI patient population, thus placing them at increased risk for Extra Pontine Myelinolysis. Therefore, we emphasize the importance of watching for this entity during the management of hyponatremia in the SCI patient population and recommend the use of MRI scans to confirm the clinical diagnosis.

Keywords: extra pontine myelinolysis; central pontine myelinolysis; hyponatremia; tetraplegia

Introduction

Since the first description of central pontine myelinolysis (CPM) in 1959[1] and extra-pontine myelinolysis (EPM) in 1987,[2] the etiology of each has remained uncertain. Rapid correction (a change in serum sodium > 25 mmol/l within 48 h) or over-correction of hyponatremia have been implicated as causes. Moreover, there are usually predisposing conditions such as alcoholism, sepsis, hepatic disease, pneumonia, malnutrition, hemorrhagic pancreatitis, SIADH, and severe burns. Hyposmolar hyponatremia (serum sodium < 130 mmol/l) is common in the spinal cord injury patient population and in most cases, it is of relatively little consequence.[3,4] We report a case of isolated extra-pontine myelinolysis, which occurred following correction of hyponatremia in an SCI patient without any of the recognized predisposing factors.

Case report

The patient was a 61 year old male who became a C6 Frankel D tetraplegic patient following cervical disc surgery. There was no history of alcohol use, cigarette use, or head injury, and he was otherwise in good health. One week post-operatively, he developed a fever secondary to a urinary tract infection and was placed on intravenous ceftazidime (Day 1 on Figure 1). One week into the course of antibiotic therapy, he developed persistent nausea, vomiting, and diarrhea. During the following four days he became increasingly lethargic, and his serum sodium dropped to 101 mmol/l (Day 11 on Figure 1). Laboratory studies showed the following: urine osmolality 346 (normal 390–1090); urine sodium 20 mEq/1; serum osmolality 214 mOsm/kg (normal 278–298); BUN 29 (normal 9–20); creatinine 0.9 (normal 0.8–1.5); TSH 1.01 mIu/ml

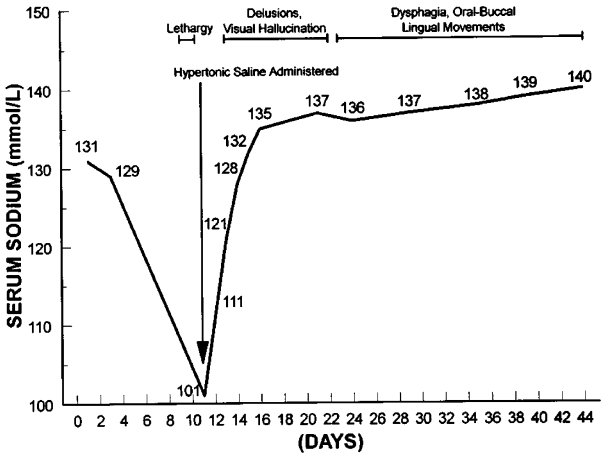

Figure 1 Serum Sodium and Development of Extra-Pontine Myelinolysis. Intravenous hypertonic saline rapidly raised serum sodium levels, leading to altered mental status and neurological changes over a subsequent 3 week period

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(normal 0.49 – 5.66). His serum sodium was raised to 128 over a two day period first with 500 cc of 5% saline at 100 cc/h, then with normal saline at 75 cc/h. During this period, the patient remained normotensive and afebrile.

Over the next week and a half, he was noticed to be intermittently confused and deluded, and had visual hallucinations. He remained afebrile and the serum electrolytes were within normal range. Gradually, he manifested dysphagia and oral buccal-lingual movements without evidence of lower cranial nerve abnormality. An MRI scan of the cervical spine showed no new pathology. Subsequently he developed dyskinesia, including head tremor and a bilateral 4–5 Hz resting hand tremor. A non-contrast enhanced head MRI scan of the cervical spine showed enhancement of the basal ganglia bilaterally and generalized atrophy (Figure 2) Over the next 3 weeks, the patient showed gradual improvement, with resolution of the oral buccal-lingual movements and improvement in mentation. However, the dyskinesia and dysphagia persisted.

Discussion

With wider availability of MRI, central pontine myelinolysis and extra-pontine myelinolysis are being increasingly identified. Often EPM occurs in conjunction with CPM. However, it is unusual for EPM to occur as an isolated condition.[5] In our patient, manifestations of CPM, such as coma, locked-in syndrome, flaccid tetraplegia, pseudobulbar palsy, nystagmus, and cranial nerve palsies[6–8] were absent. However, we observed variable mental status, decreased speech, tremor, and oral buccal-lingual movements. This constellation of findings, which may be attributed to striatal dysfunction, has been described in a case of isolated EPM.[9] EPM may involve the internal and external capsules, basal ganglia, thalamus, cerebellum, and subcortical white matter.[5] Within the zone of demyelination, blood vessels, neurons, and axis cylinders are largely spared, and inflammation is absent.[6]

In previous reports, the onset of CPM and EPM was linked to a rapid correction of serum sodium to normal, or a change in serum sodium greater than 25 mmol/l within 48 h or hypoxia/anoxia. More recent reports have pointed toward hyperosmolality and rapid osmotic shifts, with resultant hypernatremia, hyperglycemia, and azotemia, rather than to absolute serum sodium level alone as causes of CPM and EPM.[10,11] Symptoms and radiographic changes typically occur between 7 and 14 days after an acute osmotic shift. The term 'osmotic myelinolysis' has also been applied to better describe the structural involvement in CPM and EPM. Because axons are typically spared, some improvement in neuronal function may occur with resolution of edema or early re-myelination of neurons. This may explain our patient's clinical improvement in mentation.

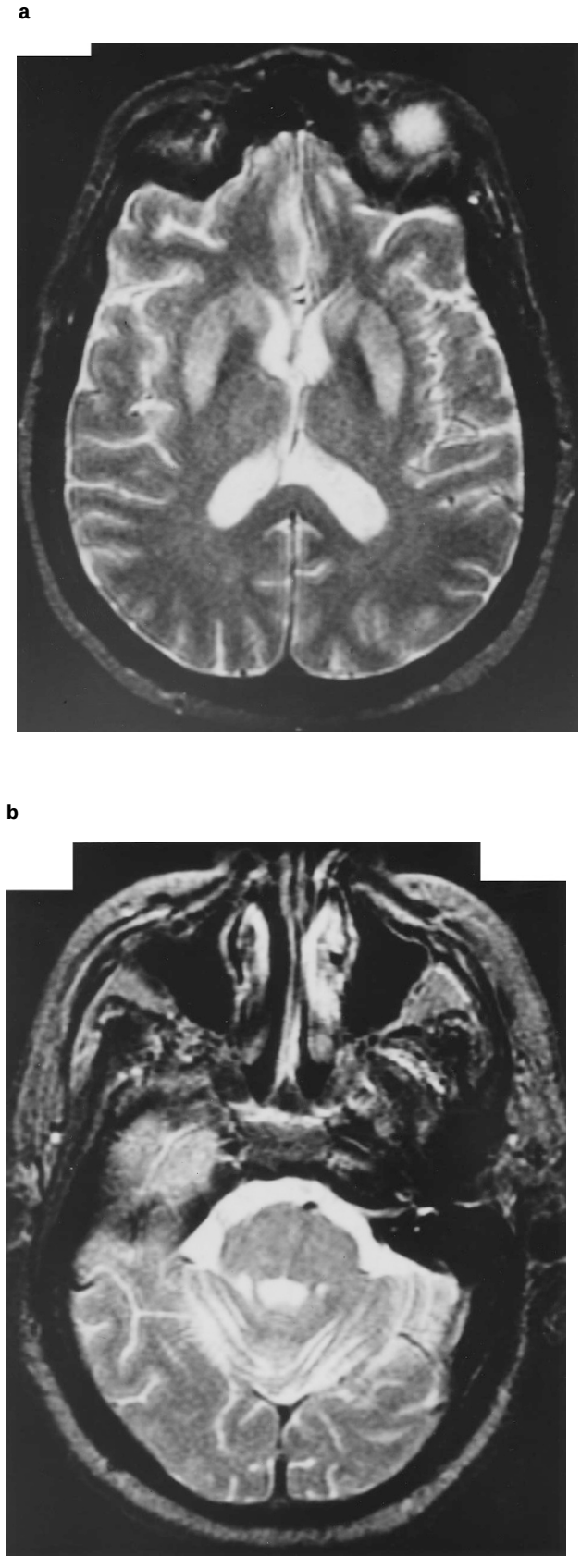

Figure 2 MRI T2 weighted images showing (a) enhancement in regions affected by EPM; (b) lack of involvement of pons

Hyponatremia frequently occurs in SCI patients, often with little or no clinical consequence. It has been estimated that hyponatremia occurs in 10–15% of 'well' SCI patients, compared to 1–2% in the general population of hospitalized patients.[4] The reasons for its occurrence are not fully understood. However, it has been suggested that there is a resetting of the osmotic threshold at which anti-diuretic hormone (ADH) is released.[12,13] This has been proposed to stem from several factors. First, a lack of sympathetic tone allows pooling of blood in the peripheral venous system. Second, an abnormal orthostatic reflex results in orthostatic hypotension with postural changes. These factors combine to create a decreased effective central blood volume, decreased kidney perfusion, and increased anti-diuretic hormone release. Another factor specific to SCI patients, especially those with high cervical injury, is excessive free water intake. There are additional factors such as the use of diuretics, the infusion of intravenous hypotonic saline and chronic renal disease. All of these factors may result in hyposmolar hyponatremia. In our patient, hyponatremia was first detected 1 week post-operatively when the patient developed a fever. At such a time in the post-operative course, the phenomenon of cerebral salt wasting must be considered along with abnormalities of ADH.[14] However, the absence of brain injury, plus mild natriuresis and dilute urine, points to excessive water intake/retention as opposed to excessive salt excretion. Salt or mineralocorticoid supplementation, along with leg stockings and careful monitoring of free water intake may help alleviate the tendency toward low serum sodium.

Conclusion

Hyponatremia occurs frequently in the SCI patient population, thus increasing their risk for extra-pontine myelinolysis. Hyponatremia should be corrected slowly and EPM should be watched for in SCI patients. MRI may be useful in confirming the diagnosis.

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