Dreading the Worst: Osmotic demyelination syndrome secondary to rapid correction of hyponatremia

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Introduction

This case reflects the gravity with which perturbations in extracellular [Na+] and their subsequent correction must be approached especially in patients who are at high risk for developing Osmotic demyelination syndrome (ODS).

Case Presentation

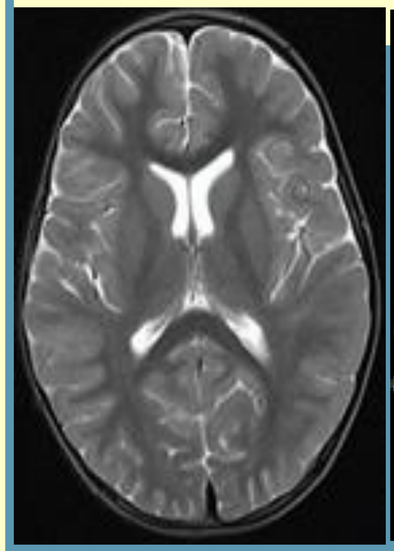
- 52 years old gentleman with Hx of HTN, alcohol use presents to ED with worsening dysphagia and altered mental status.
- Recent hospitalization(8 days ago) for hyponatremia in setting of alcohol use and being on diuretics. He presented at that time with slurred speech and altered mental status. CT and MRI head were unremarkable. SNa was 116 mEq/L on admission. K was 2.2. LFT's were moderately deranged. Serum osmolality was 230.Urine Na was 11 and osmolality was 500 mOsm/L.TSH and cortisol were normal. He was treated with 1L of LR and NS which subsequently dropped his SNa to 101 mEq/L. He was then treated with continuous 3% hypertonic saline infusion. Over the next 24 hours his serum sodium was corrected to 117 mEq/L. Serum Na then gradually increased to of 131 mEq/L by the time of discharge two days later. He continued to have intermittent confusion during hospitalization but remained without focal neurological deficits. He was eventually discharged to home after a repeat MR of the brain was unremarkable
- Home Medications:
  - Hydrochlorothiazide -lisinopril 12.5mg-20mg, 2tab daily, Metoprolol succ 200mg daily

Physical exam and Labs

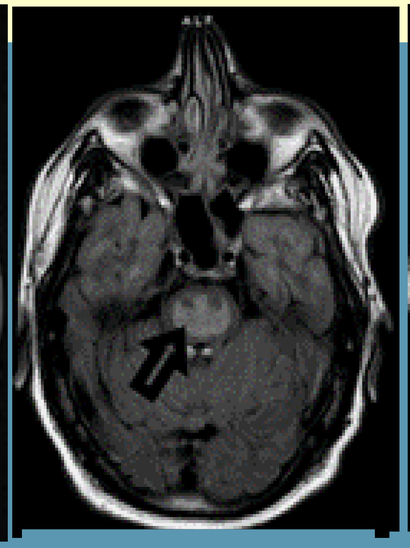
Physical Exam:
Vitals: BP 112/78 | Pulse 84 | Temp 37.6 °C (99.7 °F) (Axillary) | Resp 17 | Ht 1.829 m (6') | Wt 103.8 kg (228 lb 13.4 oz) | SpO2 97% | BMI 31.04 kg/m²
Neuro exam:
Does not follow commands. Does not attempt to speak :Cranial nerves: II-XII Intact. Motor: Brisk withdrawal of both legs with plantar stimulation. Mild withdrawal of both arms with noxious stimulation: Reflexes: 5+ beats of clonus at the right ankle, sustained clonus at the left ankle, Sensory: Grimaces to noxious stimulation in all extremities. Coordination: No observed intentional movements from which to judge coordination. Gait: Unable to walk secondary to altered mental status.
Labs: CBC: Normal: BMP: Normal. AST:45 U/L, ALT: 76 U/L, Bili: Normal, Ammonia: 29
MRI Brain: Hyperintensity in the pons, putamen ,caudate and thalamus bilaterally concerning for ODS.

Hospital Course

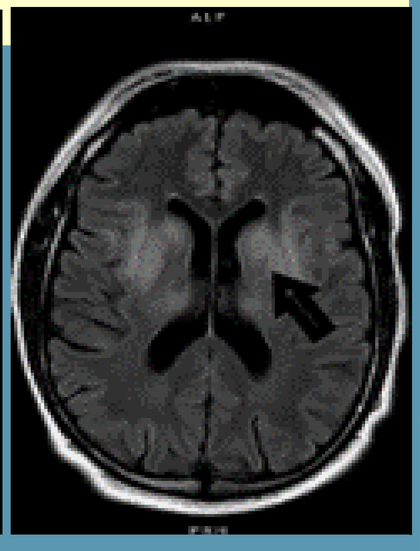
He was admitted to the hospital and due to his neurologic and MR findings concerning for ODS, was treated with 5 sessions of plasmapheresis. His neurological deficits eventually progressed to "locked in" syndrome and repeat MRI showed worsening of the demyelinating lesions. His family made the decision to pursue comfort directed care and he ultimately passed away.



Normal MRI Brain



Pons hyperintensity signal



Basal Ganglia hyperintensity signal

References

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- Sterns RH, Nigwekar SU, The treatment of hyponatremia. Semin Nephrol 29: 282–299, 2009
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Discussion

- Osmotic Demyelination syndrome (ODS) refers to central pontine myelinolysis and extra pontine myelinolysis. The typical clinical course is biphasic. Initially the patient may present with encephalopathic features or seizures due to hyponatremia. As normal sodium concentrations are restored, clinical improvement initially occurs. In the next few days, the patient's condition deteriorates, with flaccid paralysis of all 4 limbs and inability to chew, swallow, or speak. Some patients are left in a state of mutism and paralysis with relatively intact sensation and comprehension (termed pseudo coma or locked-in syndrome.
- High risk patients for ODS are: Severe hyponatremia (SNa <120),chronic alcohol use, malnutrition, prolonged diuretic use, hypokalemia, and liver disease
- Recent American guidelines target replacement rates of 4–6 or 4–8 mmol/L/day in patients at high or low risk for ODS, respectively.
- Treatment of severe hyponatremia with hypertonic saline is necessary and indicated for patients who present with severe neurologic symptoms however the rate of correction should not exceed 6-8 mEq/L and continuous infusion of hypertonic saline should be avoided.
- Reversing rapid correction of hyponatremia with desmopressin and/or hypotonic fluids in osmotic demyelination animal models has been shown to reduce mortality, but data in humans are limited
- Once ODS develops, treatment options are IVIG, High dose steroids and plasmapheresis. The outcome is variable with these treatment modalities.

Conclusions

- It is important to identify patients at high risk for osmotic demyelination syndrome and to correct their hyponatremia appropriately.