Osmotic demyelination syndrome without a smoking gun

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Osmotic demyelination syndrome (ODS) is classically described as a complication of rapid correction of severe hyponatremia. Currently there are no approved treatments other than supportive care for this rare and devastating condition. We report a case of osmotic demyelination syndrome without serum changes in sodium.

We present a case of a 44-year-old alcoholic woman who initially presented to an outside facility for a two week history of weakness, dysphagia and behavioral changes. She was treated with thiamine, however her weakness and mental status continued to worsen. She was transferred to our hospital for higher level of care. On presentation her exam acutely deteriorated requiring intubation for airway protection. On neurological exam, she did not follow commands, had intact brain stem reflexes, extensor posturing in upper extremities and abnormal flexion in lower extremities. Review of laboratory studies during the entire hospitalization did not reveal any serum sodium changes, hyponatremia or hypernatremia. MRI brain showed DWI and T2 signal changes in the central pons, bilateral thalami, caudate, and putamina with relative sparing of the globus pallidi, consistent with pontine and extrapontine myelinolysis. Based on published case reports1,2, we treated her with a five day course of IVIG followed by a five day course of steroids. The patient progressed to locked-in syndrome. After a few weeks, she was able to move her extra-ocular muscles, mouth and toes. She had tracheostomy and PEG placed and was discharged to a skilled nursing facility.

This case illustrates that central pontine/extra pontine myelinolysis should be considered as a differential diagnosis in chronically ill patients with classic symptoms even in the absence of serum sodium changes. Chronic alcohol abusers are an at risk group as they are exposed to direct toxic effects of alcohol, excess production of free radicals, nitric oxide metabolic effects and may at baseline be more susceptible to osmotic stress, predisposing them to ODS 3,4. Treatment with immunomodulators is controversial and further studies are required.

References:


