

Osmotic Demyelination Syndrome

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ABSTRACT

Formerly known as central pontine myelinolysis, osmotic demyelination syndrome (ODS) is defined by a symmetrical destruction of myelin sheaths involving mainly the central portion of the basis pontis without evidence of vascular involvement. We report the case of a 60-year-old man who presented to the emergency department with a 2-week history of progressive confusion, memory loss, and lower extremity weakness with limited ambulation. A computed tomography scan of the head revealed areas of low attenuation within the pons, and brain magnetic resonance imaging (MRI) confirmed the changes as compatible with ODS.

CASE PRESENTATION

A 60-year-old man with a history of alcoholism and diabetes mellitus type 2 presented to the emergency department with a 2-week history of progressive confusion, memory loss, and lower extremity weakness with limited ambulation. He was unkempt in appearance and oriented to person and place with ataxia, grade 3 horizontal nystagmus, and dysmetria. Muscle strength was reduced symmetrically in both lower extremities. Blood tests were abnormal only for sodium at 120 mEq/L (range 133-144 mEq/L), while all other results, including ammonia level, were normal. Hyponatremia correction was accomplished according to current guidelines¹ over a period of 2 days (Figure 1). A computed tomography (CT) scan of the head revealed areas of low attenuation within the pons (Figure 2). Brain magnetic resonance imaging (MRI) confirmed the changes as compatible with osmotic demyelination syndrome (ODS) (Figure 3). He improved over the course of the next few weeks and was discharged to an alcohol and other drug abuse program for treatment of his alcoholism.

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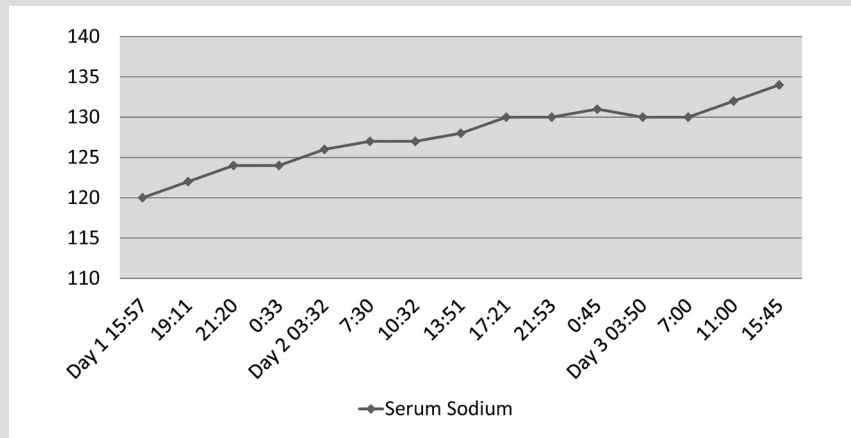
DISCUSSION

Formerly known as central pontine myelinolysis, ODS is defined by a symmetrical destruction of myelin sheaths involving mainly the central portion of the basis pontis without evidence of vascular involvement.^{2,3} The demyelination process usually occurs after rapid correction of chronic hyponatremia.³⁻⁵ ODS is associated with conditions such as alcoholism, malnourishment, diabetes, hepatic failure, liver

transplantation, cirrhosis, chronic renal failure, and malignancy.⁶ The initial diagnosis is made clinically through behavioral disturbances and neurological deficits including confusion, mutism, dysarthria, dysphagia, bulbar and pseudobulbar paresis, hyperreflexia, paraplegia, quadriplegia, and seizures. In severe cases a locked-in state and coma may be seen. Cerebellar ataxia has been reported.⁶⁻⁹

This condition was thought to be uniformly fatal with only postmortem diagnosis, but after the introduction of brain imaging, asymptomatic and milder courses without neurological deficit have been reported.¹⁰ Demyelination lesions occasionally can be detected by CT as low attenuation changes in the pons (Figure 1). The best noninvasive diagnostic technique is brain MRI, which facilitates better anatomical characterization. Typical MRI findings are of a homogeneous, well-defined region in the pons with symmetric hypodensity on T1-weighted images, hyperintensity on T2-weighted (Figure 3A), and Fluid Attenuation Inversion Recovery (FLAIR) images (Figure 3B), with no associated mass effect. In some cases the entire central pons is involved with only a thin rim of normal signal around it. These findings are not specific, and it is their anatomical distribution, combined with suggestive clinical features, that form the basis for the ODS diagnosis. MRI findings may lag behind clinical manifestations by as much as 4 weeks, so an initial negative result does not exclude ODS, and a repeat study in 2 weeks is recommended.^{11,12} The extent of the lesions does not correlate with severity of the manifestations or final outcome; this must be remembered to

Figure 1. Rate of Serum Sodium Correction Over a 48-hour Period



avoid a premature pessimistic prognosis based solely on severity of the radiographic abnormalities.^{7,13,14}

CONCLUSION

The prognosis of ODS is heterogeneous, ranging from complete neurological recovery and resolution of MRI findings to progression of deficits and death. To date, there is no specific treatment available, so efforts to prevent its occurrence remain paramount. An appropriate rate of hyponatremia correction and treatment of comorbid conditions are essential to reduce the risk of suffering this potentially devastating disease.^{7,8,15}

Figure 2. Unenhanced Computed Tomography (CT) Scan of the Head

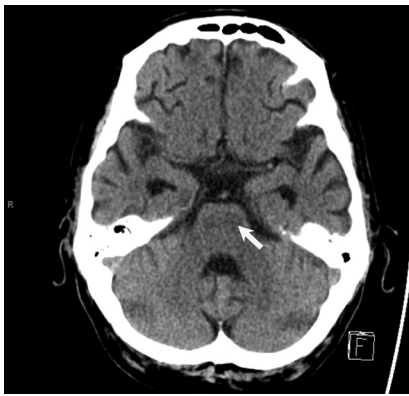


Image shows some patchy low density focus in the central pons (arrow).

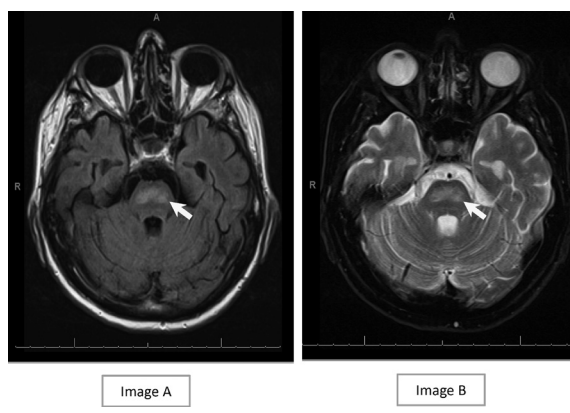
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Figure 3. Magnetic Resonance Images (MRI)



Fluid Attenuation Inversion Recovery (FLAIR) image (A) and T2-weighted magnetic resonance image (B) showing bilateral patchy high signal intensity within the pons (arrows). No mass effect is observed.

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