

Extrapontine myelinolysis: rare manifestation of a well-known disorder

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DESCRIPTION

A young alcoholic male patient was referred from another hospital with a history of loose stools and vomitings. There was documentation of hyponatraemia (Na-111 mEq/L) which was treated with intravenous normal saline. Next day, sodium was documented to be 134 mEq/L. On the third day, the patient had worsening of sensorium and was referred to our department for further management.

On presentation, the patient was comatose and had sluggishly reactive pupillary reflexes.

His MRI (figure 1) showed symmetrical hyperintensity in bilateral caudate and putamen nuclei with globus pallidus and pontine sparing consistent with extrapontine myelinolysis. The patient had rapid worsening course in our hospital and expired on the same day.

Osmotic demyelination syndrome (ODS) was first described by Adams *et al*¹ in alcoholics and malnourished patients. In ODS, demyelination is usually present within the central basis pontis (central pontine myelinolysis) or maybe found in the midbrain, thalamus, basal ganglia and cerebellum (extrapontine myelinolysis). Extrapontine lesions may accompany the pontine lesions in 10% of the cases.

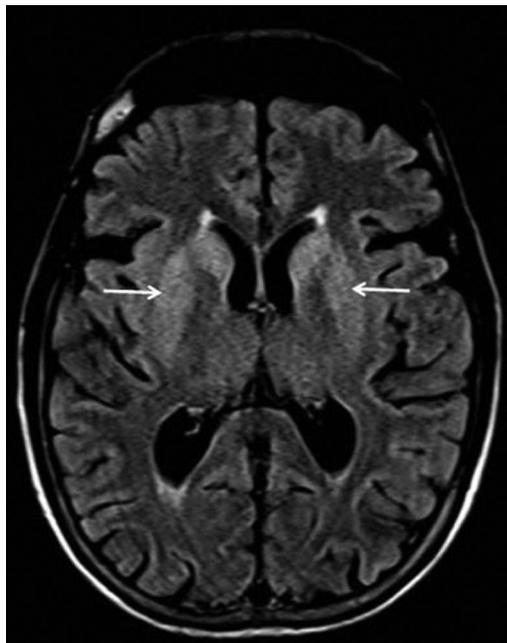


Figure 1 Axial fluid-attenuated inversion recovery MRI showing symmetrical hyperintensity in bilateral caudate and putamen nuclei (arrows) with globus pallidus sparing.

Risk factors associated with ODS include rapid correction of hyponatremia, alcoholism, malnutrition, liver disease and liver transplant.²

As seen in our case, patients with hypovolemic hyponatraemia tend to have a rapid increase in plasma Na⁺ concentration with saline therapy. This happens because of a rapid fall of arginine vasopressin levels with intravenous saline resulting in water diuresis. Hence sodium levels should be monitored frequently (2–4 h) to guide further saline therapy. Furthermore, his alcoholic status might have predisposed him for ODS.

ODS is usually associated with poor prognosis though a few case reports³ have suggested better outcome after IVIG administration.

Learning points

- ▶ Extrapontine myelinolysis is characterised by demyelinating lesions in basal ganglia, thalamus, cerebellum and midbrain with pontine and pallidal sparing.
- ▶ In patients with hypovolaemic hyponatraemia, volume repletion causes a rapid fall in arginine vasopressin levels resulting in large water diuresis, which can lead to a more rapid correction of hyponatraemia than desired.
- ▶ Sodium levels should be frequently monitored (2–4 h) while managing a case of hyponatraemia, and sodium correction should not exceed 8–10 mmol/day.

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Competing interests None.

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