Osmotic demyelination syndrome

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DESCRIPTION

A 25-year-old man presented with progressive impairment of consciousness, ocular bobbing and tetraparesis. Since the age of 3 he had received substitution therapy for central diabetes insipidus after surgical treatment of a craniopharyngioma. Accidental disruption of his intranasal desmopressin treatment 1 day before admission led to an increase of serum sodium of 29 mmol/L in less than 24 h. Initial MRI and cerebrospinal fluid examination were unobtrusive. Despite extensive symptomatic treatment the patient developed signs and symptoms of severe encephalopathy. Follow-up MRI 5 days later revealed widespread grey matter oedema with diffuse high signal and swelling of the cortex (figure 1A, asterisk) and caudate nuclei (figure 1A, arrowhead) on fluid attenuation inversion recovery sequences. Moreover, diffusion weighted imaging showed restricted diffusion in the entire cortex and internal capsule (figure 1B, C) consistent with extrapontine myelinolysis, an osmotic demyelination syndrome that may follow rapid correction of sodium in hyponatraemic patients. Usually, regions of high grey and white matter apposition show focal and bilateral affection.[1 2] Prevention is essential owing to the lack of established treatment options.[2] Since neither clinical nor radiological features are predictive, other factors determine individual prognosis: low Glasgow Coma Scale at hospital entry and during the course of disease, severe hyponatraemia and additional hypokalaemia indicate a poor outcome.[3]

Learning points

- It is important to identify patients at risk from osmotic demyelination syndrome and to correct their hyponatraemia appropriately.
- Although it is recognised that osmotic demyelination syndrome is a rare disease, its dramatic clinical course points out the importance in different disciplines.
- Since osmotic demyelination syndrome is an iatrogenic disease its early recognition and effective prevention is mandatory.

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Competing interests None.

Patient consent Obtained.

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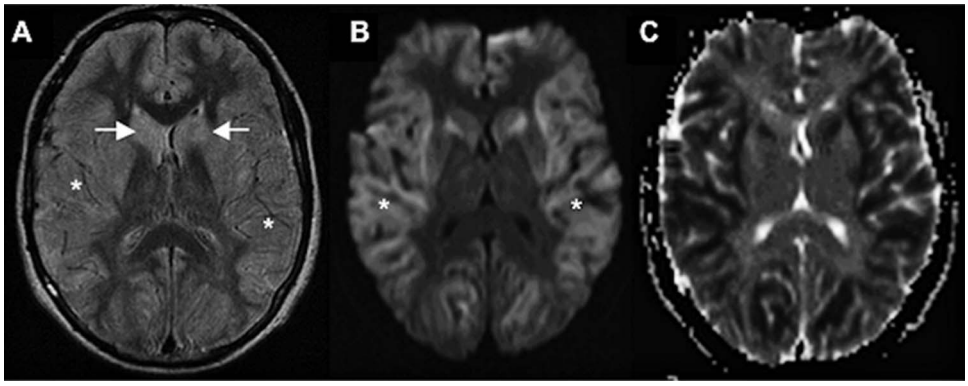

Figure 1 MRI 5 days after hospitalisation: grey matter oedema ((A) fluid attenuation inversion recovery sequences) and restricted diffusion ((B+C) diffusion weighted imaging) characteristic for extrapontine myelinolysis.

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