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The piglet and the trident sign in osmotic demyelination syndrome

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A 28-year-old man with potomania was referred to the emergency department for confusion. On physical examination he was stuporous and had mild pyramidal syndrome. His blood sodium level was 109 mmol/L. After an overly rapid correction of hyponatremia (122 mmol/L, i.e, plus 13 mmol/L, in the first 12 hours), he had reduced alertness and finally developed coma on day 5. He was intubated and mechanically ventilated. Brain magnetic resonance imaging showed prominent central pontine FLAIR-hyperintensity with peripheral and corticospinal tract sparing (Fig.1). These findings are typical of osmotic demyelination syndrome (ODS) and have been compared with a pig's snout or a trident sword (piglet sign or trident sign). One month later, he was alert with spastic paraparesis, still under mechanical ventilation.

Hypoosmolality leads to the loss of protective osmolytes that regulate cell size and prevent pathological swelling. ODS occurs after rapid osmolar shift: if the rise of serum tonicity exceeds that of intracellular space, a cell shrinking can occur, and consequently myelin membrane wrapping can separate from axons. The pons is the most prone region to ODS. To avoid this issue, targeted rate of correction should not exceed 8 mmol/L per day on any day of treatment.

Figure Legend

Figure 1:

Brain magnetic resonance imaging; the largest image is a T1 sagittal sequence demonstrating rounded hypointensity in the central pons: **A.** Axial T1 and FLAIR sequence showing a central pontine lesion with the appearance of a pig's snout (piglet sign). **B.** Coronal T1 and FLAIR sequence showing a pontine lesion with a three-pronged lance appearance (trident sign). The trident sign is classically described in the axial plane but can be easily seen also in coronal plane.

Consent for publication

Informed written consent was obtained from the patient

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