Case Reports

Hypernatremic metabolic myopathy due to hypothalamic dysfunction

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ABSTRACT

We report a patient with muscle weakness secondary to elevated serum sodium level. The cause of the elevated sodium level and the mechanism involved in producing muscle weakness are discussed.

Keywords: Hypernatremia, myopathy, hypothalamic dysfunction.

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Metabolic myopathy is muscle weakness due to endocrine disorders or inherited disorders affecting carbohydrates, lipids or mitochondrial function. It may also be due to electrolyte imbalance affecting potassium, magnesium, calcium or phosphorus. Disorders of sodium-hyponatremia and hypernatremia usually produce central nervous system depression with encephalopathy as the major clinical manifestation.[1] However, there have been two recent reports of a myopathy associated with hypernatremia secondary to hypothalamic tumors.[2,3] We report a case of hypothalamic dysfunction associated with hypernatremic metabolic myopathy (HM).

Case Report. A 30 year old British male was admitted with a one day history of difficulty in getting up from sitting, walking and climbing stairs. He used to have similar episodes of weakness lasting several hours associated with dehydration, for 2 years prior to admission but had not consulted any doctor. At the age of 14, he developed seizures and hypothalamic damage which was attributed (but not clearly proven) to accidental toluene exposure present in glue that he was using to make fly fishing hooks. There was no family history of similar illness. He was fully investigated at the Edinburgh Royal Infirmary and found to have central diabetes insipidus, absent thirst sensation and thermoregulatory capacity. His remaining pituitary functions were normal. Attempts to treat him with desmopressin (DDAVP) and a fixed fluid intake initially were complicated by hyponatremia, water intoxication and attacks of hypothermia. He stopped DDAVP after 5 years. He has now been living in Oman for 5 years, leading an active life, controlling his salt and water intake and avoiding exposure to extremes of temperature. Two weeks before admission he had gone scuba diving and was feeling cold after that. Serum sodium done in a nearby hospital was reportedly low and he was advised to take sodium chloride tablets. He had taken around 72 grams daily (1200 millimoles) for ten days prior to admission. At examination he was well built, with normal secondary sexual characteristics, vision and mental functions. He had proximal muscle weakness grade 4/5 power in the upper limbs and 3/5 power in the lower limb. Reflexes and sensations were normal. Investigations revealed a serum sodium of 180 mmol/L, but other electrolytes, calcium, magnesium, urea creatinine, glucose and hematological parameters and magnetic resonance

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Table 1 - Course and investigations.

Day	1	2	3	4	5
Weight (kgs)	83.3	-	-	-	83
Intake (ml)	5000	3000	4000	3000	3000
Temperature (oC)	35	37.5	36	36	36
Muscle Power (MRC grade)					
Upper limbs	4	4	5	5	5
Lower limbs	3	4	5	5	5
Serum sodium mmol/L	180	166	159	149	140
Serum potassium (mmol/L)	3.6	-	3.8	4.3	-
Creatinine kinase (U/L)	734	-	-	-	92

Table 2 - Causes of hypernatremia.

A. Due solely to water loss:

1. Extrarenal
   a. Skin (insensible losses)
   b. Lungs
2. Renal
   a. Diabetes inspidus (central, nephrogenic)
3. Hypothalamic dysfunction

B. Due to water loss associated with sodium loss:

1. Extrarenal
   a. Sweat
2. Renal
   a. Osmotic diuresis (glycosuria, urea)

C. Due to sodium gain:

1. Excessive sodium administration
2. Adrenocortical hyperfunction (hyperaldosteronism, Cushing's syndrome)

images of the brain were normal. Plasma aldosterone was 173 pmol/L (normal > 220). Serum cortisol was 341 nmol/L, serum follicle stimulating hormone 1.7iu/L, serum luteinising hormone 2 iu/l, thyroid stimulating hormone 1.9 miu/L all normal. Electromyogram showed short duration potentials, with excess polyphasics, suggestive of myopathy. He refused muscle biopsy. He was asked to drink around 3 litres of water per day, with which his weakness improved rapidly and the serum sodium and clinical features of myopathy returned to normal over four days.

Discussion.

Our patient had severe hypernatremia, the causes of which are outlined in Table 2. Diabetes insipidus, absence of thirst mechanism, and ingestion of excess sodium chloride, were responsible for his hypernatremia. In the present admission he had proximal muscle weakness, myopathic pattern on EMG and elevated creatinine kinase levels associated with hypernatremia. All these features returned to normal with return of serum sodium to normal levels, attesting to a reversible HM. The two reports on HM were both consequent to hypothalamic tumors and were associated with other feature of hypopituitarism.[2,3] In one of these patients,[2] phosphocreatinine/inorganic phorsphorous ratio was obtained in the resting calf muscle using 31P magnetic resonance spectroscopy. This was found to be low during the state of msucle weakness, but returned to normal after clinical improvement, suggesting that muscle weakness in hypernatremic state was caused by a depletion of muscle energy stores, probably due to an overworking sodium-potassium pump, to correct the intracellular electrolyte imbalance.[3] Since all the reported cases of HM have occurred in association with hypothalamic dysfunction, in this patient selectively affecting osmoregulation and thermo-regulation is not clear. There was no evidence of head injury, basal meningitis, hydrocephalus or tumor. In 30-50% of cases, the condition is idiopathic and usually sporadic, but familial cases are not uncommon. There is some evidence for an autoimmune aetiology for the idiopathic cases.[4] The treatment of our patient and prevention of subsequent episodes of HM requires a tight regulation of fluid and electrolyte intake to ensure normal osmolality and normonatremia. Considering the loss of osmo and thermoregulation, the fluid and electrolyte intake would have to be varied to suit extremes of ambient temperature which occur in the Sultanate of Oman especially in the summer months. Such a regime has been implemented in the patient and on regular follow-up, he has not had relapse of HM.

In conclusion, we draw attention to the rare occurrence of HM due to hypothalamic dysfunction and its management.

References

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