

Adipsic Hypernatremic Myopathy

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Chronic hypernatremia due to adipsia is very rare and occasionally presents with muscle weakness and rhabdomyolysis. We report a patient with chronic hypernatremia without thirst sensation who presented with muscle weakness and was treated successfully with prescribed water intake.

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INTRODUCTION

Chronic hypernatremia due to a defect in thirst mechanism or in osmoregulation of arginine vasopressin secretion is very rare.¹ Chronic hypernatremia due to a selective defect in osmoregulation of thirst could be secondary to structural lesion in hypothalamus such as tumor, cyst, or granulomas, and less commonly without demonstrable structural lesions.² This is often asymptomatic but may present with neurologic signs such as delirium, lethargy, and less commonly, muscle weakness.³ We report a patient who presented with muscle weakness and rhabdomyolysis due to an isolated defect in osmoregulation of thirst without evidence of a structural lesion.

CASE REPORT

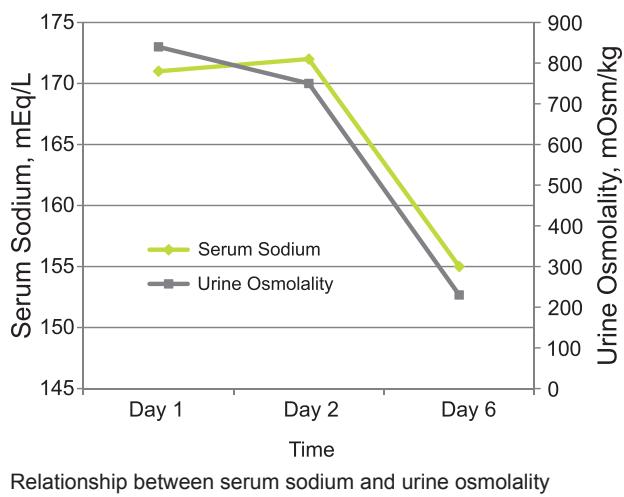
A 27-year-old man presented with progressive

muscle weakness, for the past 3 years. Physical examination was unremarkable except for decreased muscle forces (4/5) especially in the proximal muscles of the lower limbs.

Laboratory examinations showed elevated serum sodium levels, between 165 mEq/L and 180 mEq/L, on multiple measurements over the past several months. He, however, denied any thirst sensation. His 24-hour urine output measured while hospitalized was below 3 L with urine osmolality of 650 mOsm/kg to 800 mOsm/kg. Other laboratory tests showed mild increase in creatine phosphokinase (CPK) and lactate dehydrogenase as well as pancytopenia and elevated erythrocyte sedimentation rate (Table). Bone marrow aspiration and biopsy were, however, unremarkable. The central nervous system imaging studies including brain magnetic resonance imaging with and without gadolinium were unremarkable.

Laboratory Findings

Parameter	Test Date		
	Admission	6 Days After Admission	6 Month After Discharge
Fasting blood glucose, mg/dL	98
Serum urea, mg/dL	54	34	35
Serum creatinine, mg/dL	1.3	1.1	1
Serum sodium, mEq/L	171	154	157
Serum potassium, mEq/L	4.2	4	4.2
Serum calcium, mg/dL	9.3
Serum uric acid, mg/dL	8
Creatine phosphokinase, IU/mL	3435	...	350
Lactate dehydrogenase, IU/mL	774	...	220
Aldolase, IU/mL	50
Urine osmolality, mOsm/kg	840	285	...



Relationship between serum sodium and urine osmolality

He was treated with oral water intake of 100 mL on hourly basis and his serum sodium decreased slowly to 155 mEq/L over a 4-day period. He, however, developed classical symptoms of cerebral edema, including headache and vomiting. Interestingly, his urine osmolality at this time decreased to a nadir of 250 mOsm/kg (Figure). His hourly water intake was decreased to 70 mL to 80 mL and his serum sodium increased to 157 mEq/L and all symptoms of cerebral edema disappeared.

In 6-month follow-up, the patient felt well without muscle weakness and returned to work. He continued on water intake of 70 mL/h, while awake. He denied any thirst sensation. His latest laboratory examination showed a sodium level of 157 mEq/L, urine osmolality of 350 mOsm/kg, and a CPK of 350 IU/L (Table).

DISCUSSION

Our patient presented primarily with a 3-year history of muscle weakness treated unsuccessfully with variety of medications for primary muscular disorders, including polymyositis. His muscle weakness resolved with improvement in his serum sodium. Myopathy caused by hypernatremia is a very rare complication of this disorder,^{4–6} and as a result, it is not included in differential diagnosis of myopathic syndromes.⁷ In previously reported cases of hypernatremic myopathy, serum sodium levels were greater than 160 mEq/L; however, CPK levels were only mildly to moderately elevated and rarely greater than 3000 IU/L.^{4,5} As the Table shows, CPK in our patient was greater than 3000 IU/L and returned to normal levels with treatment

of hypernatremia.

Pathogenesis of hypernatremic myopathy is not fully elucidated. In 1 patient, Hiromatsu and colleagues showed that muscle weakness was caused by depletion of intramuscular energy stores, probably due to the overloading sodium-potassium pump to correct the intracellular electrolyte imbalance.⁵ In addition, this is associated with decreased total body exchangeable potassium due to severe intracellular dehydration.^{8,9}

The etiology of chronic hypernatremia is most commonly structural lesions involving hypothalamus and is often associated with a defect in arginine vasopressin secretion.^{5,10,11} In a few rare cases, such as in our patient, there is no demonstrable structural lesion and arginine vasopressin secretion is intact. In a single case reported recently, the etiology was thought to be development of autoantibody to brain sodium level sensor of body fluids.¹² Most cases are, however, idiopathic. Some of these patients have reset osmostat demonstrated by normal urinary dilution during water loading test.³ In our patient, however, attempts to decrease serum sodium led to development of cerebral edema at the time that serum sodium was 157 mEq/L and urine osmolality, although lower at 250 mOsm/kg, did not reach a level below 100 mOsm/kg to allow the diagnosis of reset osmostat.

Treatment of these patients is very challenging on the basis of water intake. In our patient, such a prescription resulted in a decrease in serum sodium with resolution of all his symptoms. Nonetheless, the influence on mortality and morbidity is not clear.

CONFLICT OF INTEREST

None declared.

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