A Case of Severe Hypernatremic Myopathy by Primary Hypodipsia, Hyperglycemic Hypertonic State in a 17-Year-Old Patient With Mental Retardation

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Abstract

Although severe hypernatremia, defined as a sodium concentration > 180 mEq/L, is associated with a high mortality rate, particularly in adults, little is known about severe hypernatremia in patients with diabetic ketoacidosis (DKA). Hypernatremic myopathy has been reported in patients with severe dehydration, acute kidney injury, and rhabdomyolysis. Dehydration in a diabetic patient with mental retardation may be caused by osmotic diuresis and exacerbated by hypodipsia, which requires the patient to be educated about regular oral hydration. Here, we describe a 17-year-old patient with reversible severe hypernatremia (serum corrected sodium, 180 mEq/L) and primary hypodipsia, but without DKA. The patient visited the emergency room due to agitation and weakness in both lower extremities. Initial laboratory tests showed hyperglycemia, acute kidney injury, acute hepatitis, and myopathy. Following rehydration with one-quarter isotonic saline and intensive insulin therapy, the patient’s symptoms (including leg weaknesses) and laboratory findings improved.

Keywords: Severe hypernatremia; Myopathy; Hypodipsia; Hyperglycemia

Introduction

Severe hypernatremia, defined as a sodium concentration > 180 mEq/L, is associated with a high mortality rate, particularly in adults. Moreover, children with hypernatremia are estimated to carry a 15-fold higher risk of death than those with no hypernatremia [1]. Severe and extreme hypernatremia are defined as serum sodium concentrations > 160 mmol/L and > 190 mmol/L, respectively [2]. There are few studies reporting severe hypernatremia in patients with diabetic ketoacidosis (DKA) or myopathy associated with hypernatremia. Hypernatremic myopathy is associated with severe dehydration, acute kidney injury, and rhabdomyolysis [2]. In diabetic patients with mental retardation, dehydration can be caused by osmotic diuresis and is exacerbated by hypodipsia. Here, we describe a 17-year-old patient with reversible severe hypernatremia (serum corrected sodium, 180 mEq/L) with primary hypodipsia and myopathy, but without DKA.

Case Report

A 17-year-old patient presented to the emergency room of our hospital with a 3 day history of agitation and weakness in both legs. He had been admitted twice before due to similar recurrent symptoms. During his most recent admission, he was diagnosed with mental retardation, dysgenesis of the...
corpus callosum (Fig. 1), and complete cleft palate; however, he had a normal 46XY karyotype. Two years earlier, he was diagnosed with type 2 diabetes mellitus, but his parents stopped his oral antidiabetic drugs because they caused significant discomfort. Upon physical examination, he was alert but uncooperative due his mental problems. He showed grade III motor weakness with a positive Gowers’ sign, as well as decreased deep tendon reflexes at both knees. He did not complain of thirst.

Initial laboratory findings showed hyperglycemia (serum glucose, 695 mg/dL), acute kidney injury (serum creatinine, 1.86 mg/dL; estimated GFR, 50.54 mL/min), acute

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<th>Serum Na (mM/L)</th>
<th>Corrected Na (mM/L)</th>
<th>Serum osm (mOs/Kg)</th>
<th>Urine osm (mOs/kg)</th>
<th>Serum CK (IU/L)</th>
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HD, hospital day; Na, sodium; osm, osmolarity; CK, creatine phosphokinase.

Figure 2. After hydration (from HD1 to HD10), the concentrations of Na and CK in the patient’s serum and osmolarity improved. HD, hospital day; Na, sodium; osm, osmolarity; CK, creatine phosphokinase.
Severe Hypernatremic Myopathy with HHS

Hypernatremic myopathy may be diagnosed from typical myopathic symptoms and sustained hypernatremia, which show prompt symptomatic improvement when the hypernatremia is controlled [5]. Other compatible clinical and laboratory findings may help the diagnosis (but are not necessary), and head CT/MRI may be performed to identify any hypothalamic lesions [5].

Adipsia can aggravate the dehydration and influence the occurrence of hypernatremia. Adipsic hypernatremia is associated with several intracranial pathologies, mostly in or around the hypothalamus [8]. One study summarized the findings in four patients with adipsic hypernatremia associated with the abnormal development of the corpus callosum [6]. Chromosomal abnormalities are associated with dysgenesis of the corpus callosum and cleft palate, but no chromosomal abnormality was detected in our patient [9-11].

Treatment for hypernatremic myopathy involves rehydration and treatment of the underlying disease [5]. Following hydration with one-quarter isotonic saline and intense insulin therapy, the current patient’s symptoms resolved completely and the laboratory findings were normal.

In summary, dehydration in a diabetic patient with mental retardation may be caused by osmotic diuresis and be exacerbated by hypodipsia, resulting in severe hypernatremia. It is important that these patients receive intensive education about regular oral hydration.

Conflicts of Interest

There were no conflicts of interest.

References


