Sjögren’s syndrome presenting with hypokalemic paralysis

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ABSTRACT

Introduction: Acute systemic weakness is a common reason for emergency department visit. Acute hypokalemic paralysis is one of its clinical presentation forms. It is a rare but treatable cause of acute weakness. We report a case that presented with hypokalemic paralysis in emergency room. Case Report: We report a rare case of a 36-year-old female presented with sudden onset flaccid left hemiparesis with no significant past clinical history. Initially the patient was considered as an ischemic stroke, but she was later found to have hypokalemia due to distal renal tubular acidosis and further diagnosed as case of Sjögren’s syndrome. Conclusion: Acute flaccid neuromuscular weakness is a common presenting symptom in emergency room. Hypokalemia is an important differential diagnosis. The cause of hypokalemia should be sought. The diagnosis of distal renal tubular acidosis must be considered in patients presenting with hypokalemic paralysis and renal stones, and may be the first clue in the identification of an underlying autoimmune disorder, particularly Sjögren’s syndrome.

Keywords: Hypokalemia paralysis, Neuromuscular weakness, Renal tubular acidosis, Sjögren’s syndrome

INTRODUCTION

Acute systemic weakness is a common reason for emergency department visit. The etiologies of this syndrome are very diverse including neurologic, metabolic, and infectious causes. Among its clinical presentation forms, we find the acute hypokalemic paralysis, which characterized by acute systemic weakness and low serum potassium. It is a rare but treatable cause of acute weakness. However, treatment modalities will differ in each of the above-described etiologies and it requires careful evaluation of patient to reach the final diagnosis. We report a case that presented with hypokalemic paralysis in emergency room. Initially the patient was considered as an ischemic stroke, but by step-wise approach, we finally reached the diagnosis.

CASE REPORT

A 36-year-old female was admitted in emergency room with sudden onset left hemiparesis. She has a history of recurrent urinary tract infections with left
nephrectomy for renal stones and pyonephrosis, polyuric-polydipsic syndrome for eight years with normal blood sugar level and hypertension.

The patient did not have history of nausea, vomiting, diarrhea or use of any drugs. Her family history was insignificant. On examination, she was afebrile, pulse was 86/min, regular, blood pressure 130/80 mmHg. Patient looked pale and dehydrated. Cardiac examination was normal. Examination of lungs and abdomen were also normal. Neurological examination revealed flaccid muscle weakness involving all four limbs primarily in the left side with areflexia without sensory involvement. Plantar reflexes were flexor bilaterally.

The electrocardiogram has shown flattened T waves, ST depression and presence of subtle U waves (Figure 1). Laboratory results are summarized in Table 1. HBsAg, HCV RNA, HIV were negative.

The diagnosis of ischemic stroke was suspected in a history of hypertension and sudden onset. A brain scan was normal, which is not against the diagnosis. However, renal tubular acidosis (RTA) was suspected in the presence of hyperchloremic metabolic acidosis, a urine pH greater than 7 and negative urine culture after 48 hours with no history of digestive disorders or diuretic usage. Autoantibody screen revealed positive antinuclear antibody (640), anti-SSA and anti-RO52 antibodies. Anti dsDNA, anti-Sm, anti-SSB, anti-SCL70 and anti-JO1 antibodies, rheumatoid factor and CIC were negative. These reports and distal RTA raised a high index of suspicion of Sjögren’s syndrome.

To confirm the diagnosis, a labial biopsy was performed showing a lymphoplasmacytic infiltrate of grade IV according to Chisholm’s classification, which is in favor of Sjögren’s syndrome.

Patient received a potassium supplementation at the dose of 1 g per hour and alkalinization by sodium bicarbonate at the dose of 100 mEq in the emergency department. She was treated by prednisolone at the dose of 0.5mg/kg/day for six weeks with good outcome. She has a regular nephrology follow-up.

**DISCUSSION**

Distal RTA is a condition characterized by an inability of the distal nephron to acidify the urine. It is characterized by hyperchloremic hypokalemic, metabolic acidosis with a normal serum anion gap and urine pH greater than 5.5 [1].

Distal RTA is rare and it may be primary or secondary resulting from various disorders like autoimmune diseases, kidney transplantation, nephrocalcinosis, medullary sponge kidney, chronic obstructive uropathy, drugs (mainly amphotericin B, ifosfamide, and lithium).

### Table 1: Laboratory findings of the patient

| Parameter          | Value       | Normal Range |
|--------------------|-------------|--------------|
| Serum Sodium       | 146 mEq/l   | 138–142      |
| Serum Potassium    | 1.16 mEq/l  | 3.5–5        |
| Serum Chlorine     | 122 mEq/l   | 100          |
| Serum Calcium      | 1.9 mmol/l  | 2.2          |
| Serum Phosphorus   | 0.45 mmol/l | 0.8          |
| Serum Urea         | 5.36 mmol/l | <7.5         |
| Creatinine         | 86 μmol/l   | <100         |
| ASAT               | 8.9 IU/l    | 40           |
| ALAT               | 6.3 IU/l    | 40           |
| TBil/C             | 4/1 mg/dl   | <5           |
| C-reactive Protein | 2.13 mg/l   | <4           |
| Serum pH           | 7.26        | 7.38–7.42    |
| PaO2               | 77.7 mmHg   | 100          |
| PaCO2              | 25.9 mmHg   | 38–42        |
| HCO3-              | 11.9 mmHg   | 22–26        |
| SaO2               | 93.7%       | 100          |
| Serum Lactate      | 1.8 mmol/l  | <2           |
| Serum Protein      | 74 g/l      | 60           |
| Urine Sodium       | 180 mmol/24 h | >40/24 h    |
| Urine Potassium    | 55 mmol/24 h | >20/24 h    |
| Urine Chlorine     | 6.1 mmol/24 h | >20/24 h    |
| Urine pHe          | 7           | 4.6–8        |
| Urine Protein      | Trace       | 0            |
| Hematuria          | Trace       | 0            |
| Serum anion gap    | 12 mEq/l    | 12–16        |
| Urine anion gap    | 22 mEq/l    | -10 to 10    |
| Urine culture      | Negative    | Negative     |
| Hemoglobin         | 12.8 mg/dl  | 12–16        |
| WBC                | 15.4x10^9/L | 4–9x10^9/L   |
| RBC                | 5.1x10^12/L | 4.7–6.1x10^12/L |
| Platelet Count     | 243x10^9/L  | 150–400x10^9/L |
| INR                | 1.1         | 0.9–1.2      |
Among the autoimmune diseases that may be associated with this syndrome, we find the Sjögren’s syndrome in which, the clinical presentation can be associated with distal acidification in up to 25% of patients, a mild metabolic acidosis and in some case, hypokalemic paralysis can be its presenting symptom [2, 3]. The mechanism by which Sjögren’s syndrome leads to distal RTA is incompletely understood. It may be due to high levels of anti-carbonic anhydrase anti-bodies, which affect the function of carbonic anhydrase in cortical collecting ducts [4] or a lack of intact H+ ATPase pumps in the intercalated cells [5]. Usually, patients with distal RTA present with symptoms of nephrolithiasis, but the symptoms of hypokalemia and hypocalcaemia may be the first abnormalities, like in our case. The clinical manifestations of potassium depletion vary between individual patients, and their severity depends on the degree of hypokalemia. It can range from fatigue, myalgia and muscular weakness to complete paralysis. Correction of hypokalemia and alkaline replacement are the standard therapy of distal RTA. Hypokalemia should be corrected first because alkaline replacement can aggravate hypokalemia resulting in dangerous consequences. Steroid therapy should be considered in presence of severe RTA and hypokalemia paralysis in Sjögren’s syndrome to prevent relapses and to improve the response to replacement therapy [6, 7–11].

**CONCLUSION**

Acute flaccid neuromuscular weakness is a common presenting symptom in emergency room. Hypokalemia is an important differential diagnosis besides primary neuromuscular diseases. The cause of hypokalemia should be sought. The diagnosis of distal renal tubular acidosis must be considered in patients presenting with hypokalemic paralysis and renal stones, and may be the first clue in the identification of an underlying autoimmune disorder, particularly Sjögren’s syndrome.

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