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HYPOKALEMIC PARALYSIS DUE TO DISTAL RENAL TUBULAR ACIDOSIS AS THE INITIAL MANIFESTATION OF PRIMARY SJÖGREN'S SYNDROME: A CASE REPORT

Emergency Medicine

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ABSTRACT

Primary Sjögren's syndrome is a chronic systemic autoimmune disorder that predominantly affects the exocrine glands, producing classical sicca symptoms such as dry eyes and dry mouth. However, extra-glandular manifestations may occur and can involve multiple organ systems including the kidneys, lungs, and nervous system. Renal involvement in Sjögren's syndrome most commonly manifests as tubulointerstitial nephritis and may lead to distal renal tubular acidosis (dRTA). Distal renal tubular acidosis can cause metabolic acidosis and renal potassium wasting, leading to severe hypokalemia. In rare cases, profound hypokalemia may present with acute flaccid paralysis. We report a case of a 34-year-old female presenting to the emergency department with acute quadriparesis secondary to severe hypokalemia. Further investigations revealed normal anion gap metabolic acidosis and urinary potassium wasting suggestive of distal renal tubular acidosis. Autoimmune evaluation demonstrated strongly positive anti-SSA/Ro and anti-SSB/La antibodies, and Schirmer's test confirmed ocular dryness, fulfilling the 2016 ACR/EULAR criteria for primary Sjögren's syndrome. The patient was successfully managed with intravenous potassium replacement followed by immunomodulatory therapy. This case highlights the importance of considering autoimmune causes such as Sjögren's syndrome in patients presenting with hypokalemic paralysis

KEYWORDS

Primary Sjogren's syndrome, Hypokalemic paralysis, Distal renal tubular acidosis

INTRODUCTION

Sjögren's syndrome is a chronic autoimmune disease characterized by lymphocytic infiltration and destruction of exocrine glands, leading to reduced secretory function. The disease typically presents with xerostomia and xerophthalmia, commonly referred to as sicca symptoms(1). It can occur as a primary disorder or secondary to other autoimmune conditions such as rheumatoid arthritis or systemic lupus erythematosus(2).

Although glandular manifestations are the hallmark of the disease, extra-glandular involvement may occur in approximately 30–40% of patients(3,4). Renal involvement is reported in about 4–9% of cases and most commonly presents as tubulointerstitial nephritis(5). One of the important clinical consequences of renal tubular dysfunction is the development of distal renal tubular acidosis(6).

Distal renal tubular acidosis occurs due to impaired hydrogen ion secretion in the distal nephron, resulting in normal anion gap metabolic acidosis and alkaline urine(7,8). This defect often leads to renal potassium wasting, causing severe hypokalemia(9).

Severe hypokalemia may result in neuromuscular manifestations such as muscle weakness or hypokalemic paralysis, which can occasionally be the first presenting feature of Sjögren's syndrome(10). Early recognition is essential because prompt correction of electrolyte imbalance can rapidly reverse neurological deficits(11).

Here we present a case of hypokalemic paralysis due to distal renal tubular acidosis as the initial presentation of primary Sjögren's syndrome.

CASE REPORT

A 34-year-old female presented to the emergency department with complaints of progressive generalized weakness for two days, eventually leading to inability to stand or walk. There was no history of trauma, seizures, loss of consciousness, diarrhea, vomiting, diuretic use, or excessive sweating.

The patient reported chronic dryness of mouth and foreign body sensation in the eyes for nearly one year, which had not been previously evaluated.

PRIMARY SURVEY IN THE EMERGENCY DEPARTMENT

On arrival, the patient was assessed according to standard emergency evaluation principles.

Airway: Patent; the patient was able to speak normally.

Breathing: Spontaneous breathing with symmetrical chest expansion and adequate oxygen saturation.

Circulation: Pulse rate and blood pressure were stable with adequate peripheral perfusion.

Disability: Neurological examination revealed symmetrical flaccid quadriparesis with motor power 0/5 in all limbs and absent deep tendon reflexes. No sensory deficits or cranial nerve abnormalities were noted.
Exposure: No evidence of trauma, dehydration, or skin abnormalities.

PRIMARY ADJUNCTS

Initial investigations included electrocardiography and laboratory evaluation.

Electrocardiogram (ECG) showed classical features of hypokalemia:

ST segment depression, QT interval prolongation, T-wave inversion, Prominent U waves

Laboratory results revealed:
Serum potassium: 1.0 mEq/L

Arterial blood pH: 7.22 , anion gap: 9 (normal anion gap metabolic acidosis)

These findings suggested severe hypokalemia with metabolic acidosis.

Table 1. Laboratory Investigations

Investigation	Result	Normal Range	Interpretation
Serum Potassium	1.0mEq/L	3.5 – 5.0 mEq/L	Severe hypokalemia
Arterial Blood pH	7.22	7.35 – 7.45	Metabolic acidosis
Anion Gap	9	8 – 16	Normal anion gap metabolic acidosis

Urine Potassium/Creatinine Ratio	45 mEq/g	< 13 mEq/g	Urinary potassium wasting
Urine pH	7.2	<5.5 in acidosis	Suggestive of distal renal tubular acidosis
Anti-SSA (Ro) antibodies	Positive	Negative	Suggestive of Sjögren's syndrome
Anti-SSB (La) antibodies	Positive	Negative	Suggestive of Sjögren's syndrome
Schirmer Test	2 mm (left), 3 mm (right)	>10 mm	Severe ocular dryness

PROVISIONAL DIAGNOSIS

Based on clinical presentation and initial investigations, a provisional diagnosis of hypokalemic paralysis due to severe hypokalemia was made.

INITIAL MANAGEMENT IN THE EMERGENCY DEPARTMENT

Because severe hypokalemia carries a risk of life-threatening cardiac arrhythmias, the patient was immediately placed on continuous cardiac monitoring.

Emergency management included:

- Establishment of intravenous access
- Urgent biochemical investigations
- Intravenous potassium chloride infusion at 10 mEq/hour under ECG monitoring

Due to the severity of electrolyte disturbance, the patient was transferred to the intensive care unit for close monitoring.

SECONDARY SURVEY

Further investigations were performed to identify the cause of potassium loss. Urine studies revealed- Urine potassium-to-creatinine ratio: 45 mEq/g, pH: 7.2 These findings indicated renal potassium wasting and distal renal tubular acidosis. Autoimmune evaluation demonstrated Strongly positive anti-SSA/Ro antibodies and anti-SSB/La antibodies, Schirmer's test confirmed severe ocular dryness, measuring 2 mm in the left eye and 3 mm in the right eye.

Based on these findings, the patient fulfilled the ACR/EULAR 2016 classification criteria for primary Sjögren's syndrome.

OUTCOME AND FOLLOW UP

Serum potassium levels normalized within four days of treatment, and the patient showed complete recovery of motor power.

At discharge, the patient was prescribed:

Oral potassium supplementation, tablet Prednisolone (tapering dose), tablet Azathioprine and tablet Hydroxychloroquine. At 1-, 3-, and 6-month follow-up, the patient remained asymptomatic with stable potassium levels.

DISCUSSION

Renal involvement in Sjögren's syndrome is relatively uncommon but clinically important. The most frequently reported renal manifestation is tubulointerstitial nephritis, which can lead to multiple forms of renal tubular dysfunction, including distal renal tubular acidosis (dRTA)[1].

Distal renal tubular acidosis results from defective hydrogen ion secretion in the distal nephron, leading to failure of urinary acidification despite systemic metabolic acidosis. This results in normal anion gap metabolic acidosis, alkaline urine, and renal potassium wasting[2]. Persistent potassium loss can produce severe hypokalemia, which may manifest as muscle weakness, paralysis, or even life-threatening cardiac arrhythmias[3].

Hypokalemic paralysis is a rare but well-recognized complication of dRTA associated with Sjögren's syndrome. Renal involvement occurs in approximately 4–9% of patients with primary Sjögren's syndrome, with distal renal tubular acidosis being the most common renal abnormality[4]. Renal tubular dysfunction may also precede the classical sicca symptoms in some patients, leading to delayed diagnosis of the underlying autoimmune disorder[5].

Several case reports have described hypokalemic paralysis as the initial manifestation of Sjögren's syndrome. Severe hypokalemia due to distal renal tubular acidosis can result in acute flaccid paralysis that resolves rapidly after potassium replacement therapy[5,11]. Electrolyte abnormalities may therefore be the first clinical clue to an underlying autoimmune disease[6].

The mechanism of renal tubular dysfunction in Sjögren's syndrome is believed to involve autoimmune-mediated damage to the distal nephron, particularly the intercalated cells responsible for hydrogen ion secretion. Lymphocytic infiltration and immune-mediated injury lead to impaired urinary acidification and increased potassium excretion[10].

Management of hypokalemic paralysis involves rapid correction of potassium levels, preferably through intravenous replacement under continuous cardiac monitoring. Correction of metabolic acidosis is usually performed after potassium levels are stabilized because bicarbonate therapy can further worsen hypokalemia[14].

Long-term management focuses on treating the underlying autoimmune disease using immunosuppressive therapy, including corticosteroids and disease-modifying agents such as azathioprine or hydroxychloroquine[15]. Early diagnosis and appropriate treatment are essential to prevent recurrence of hypokalemia and progression of renal involvement.

In our case, severe hypokalemia resulted in acute quadriparesis, which improved completely after potassium correction. Subsequent evaluation revealed distal renal tubular acidosis and strongly positive autoimmune markers consistent with primary Sjögren's syndrome. The patient responded well to potassium supplementation and immunomodulatory therapy and remained asymptomatic on follow-up.

This case highlights the importance of considering autoimmune etiologies such as Sjögren's syndrome in patients presenting with unexplained hypokalemia and metabolic acidosis, particularly when accompanied by renal potassium wasting.

CONCLUSION

Hypokalemic paralysis due to distal renal tubular acidosis is an uncommon but important manifestation of primary Sjögren's syndrome. Emergency physicians should consider autoimmune etiologies in patients presenting with unexplained hypokalemia and metabolic acidosis.

Early diagnosis and prompt treatment can result in rapid neurological recovery and prevent potentially life-threatening complications.

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