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Hypokalemic Periodic Paralysis Secondary to Type 1 Renal Tubular Acidosis in Sjögren Syndrome: A Case Report

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Abstract

Background

Hypokalemic periodic paralysis (HPP) is an uncommon but potentially life-threatening clinical condition characterized by episodic muscle weakness associated with severe hypokalemia. Distal renal tubular acidosis (dRTA) is a recognized but rare cause of HPP and may occur secondary to autoimmune disorders such as Sjögren syndrome.

Case Presentation

We report the case of a 40-year-old female who presented with sudden onset quadriparesis. Clinical examination revealed bilateral upper and lower limb weakness with diminished reflexes. Arterial blood gas analysis demonstrated severe metabolic acidosis with hyperchloremia and profound hypokalemia. Laboratory investigations revealed urine pH of 6.5, serum potassium of 1.43 mEq/L, bicarbonate of 7.2 mEq/L, and positive anti-Ro/SSA and anti-Ro52 antibodies. MRI brain imaging was unremarkable. Nerve conduction studies suggested sensory motor axonal polyneuropathy. A diagnosis of hypokalemic periodic paralysis secondary to distal renal tubular acidosis associated with Sjögren syndrome was established. The patient improved significantly following aggressive potassium and bicarbonate replacement therapy.

Conclusion

Distal renal tubular acidosis secondary to Sjögren syndrome should be considered in patients presenting with unexplained hypokalemic paralysis and normal anion gap metabolic acidosis. Early diagnosis and prompt electrolyte correction are essential to prevent morbidity and mortality.

Keywords: Hypokalemic periodic paralysis, Distal renal tubular acidosis, Sjögren syndrome, Hypokalemia, Autoimmune disease

Introduction

Hypokalemic periodic paralysis (HPP) is characterized by episodic flaccid muscle weakness caused by severe hypokalemia. It may be primary (familial) or secondary to underlying endocrine, renal, gastrointestinal, or autoimmune disorders. Among the secondary causes, distal renal tubular acidosis (dRTA) is an important but relatively uncommon etiology.

Distal renal tubular acidosis results from impaired hydrogen ion secretion in the distal nephron, leading to non-anion gap metabolic acidosis, hypokalemia, and alkaline urine. Autoimmune disorders, particularly Sjögren syndrome, are among the common acquired causes of dRTA. Sjögren syndrome is a chronic autoimmune disease primarily affecting exocrine glands, resulting in symptoms such as xerostomia and xerophthalmia. However, extra-glandular manifestations including renal involvement may occasionally be the initial presentation.

Hypokalemic paralysis due to dRTA associated with Sjögren syndrome is rare and may delay diagnosis if not considered early. We present a case of severe hypokalemic periodic paralysis secondary to type 1

renal tubular acidosis associated with Sjögren syndrome in a middle-aged female.

Case Report

A 40-year-old female presented to the emergency department with sudden onset weakness of all four limbs for one day. The weakness was progressive and predominantly involved proximal muscles. There was no history of fever, diarrhea, vomiting, trauma, seizures, bowel or bladder involvement, or toxin exposure.

On examination, the patient was conscious and oriented. Neurological examination revealed bilateral upper and lower limb weakness with muscle power graded 0/5 in the lower limbs and significantly reduced power in the upper limbs. Deep tendon reflexes were diminished bilaterally, and plantar reflexes were flexor. Sensory examination was normal. There were no cranial nerve deficits.

Initial laboratory investigations demonstrated severe hypokalemia and metabolic acidosis. Arterial blood gas analysis showed:

1. pH: 6.91
2. pCO2: 22.7 mmHg
3. pO2: 229.2 mmHg
4. HCO3−: 7.2 mEq/L
5. SO2: 98.8%

Serum electrolyte values were:

1. Serum sodium: 137 mEq/L
2. Serum potassium: 1.43 mEq/L
3. Serum chloride: 117 mEq/L
4. Serum calcium: 7.2 mg/dL
5. Serum creatinine: 1.21 mg/dL
6. Anion gap: 8.63

Urine investigations revealed:

1. Urine sodium: 65 mEq/L
2. Urine potassium: 17.1 mEq/L
3. Urine chloride: 37 mEq/L
4. Spot urine potassium: 16.5 mEq/L
5. Urine pH: 6.5
6. Urine osmolality: 215 mOsm/kg
7. Spot urine creatinine: 38.4 mg/dL

Electrocardiography showed hypokalemic changes including prominent U waves and T-wave flattening. MRI brain imaging was normal. Nerve conduction velocity testing suggested sensory motor axonal polyneuropathy.

Further evaluation for secondary causes revealed positive antinuclear antibody (ANA) with fine speckled pattern. Anti-Ro/SSA and Anti-Ro52 antibodies were strongly positive. Thyroid profile demonstrated FT3 of 1.5 pg/mL, FT4 of 1.17 ng/dL, and TSH of 0.36 µIU/mL. Serum CRP was 6.68 mg/L and procalcitonin was 0.84 ng/mL.

Based on the clinical features and laboratory findings, a diagnosis of hypokalemic periodic paralysis secondary to distal renal tubular acidosis associated with Sjögren syndrome was made.

The patient was treated with aggressive intravenous potassium supplementation along with bicarbonate correction and supportive management. Significant improvement in muscle power was observed following correction of hypokalemia and acidosis.

Discussion

Hypokalemia is among the most common electrolyte abnormalities encountered in clinical practice. Severe hypokalemia can manifest as muscle weakness, respiratory compromise, arrhythmias, and paralysis. Distal renal tubular acidosis is an important renal cause of hypokalemia resulting from impaired distal acidification.

The pathophysiology of dRTA in Sjögren syndrome involves autoimmune-mediated tubulointerstitial nephritis leading to dysfunction of hydrogen ion transporters, including H+-ATPase and H+/K+-ATPase pumps in the collecting duct. This results in impaired urinary acidification, metabolic acidosis, and potassium wasting.

Renal involvement in Sjögren syndrome is relatively uncommon compared with glandular manifestations, but dRTA remains the most frequent renal manifestation. In some patients, hypokalemic paralysis may be the initial presenting feature before the classical sicca symptoms become evident.

The diagnosis of dRTA is based on the presence of normal anion gap metabolic acidosis, inappropriately elevated urine pH (>5.5), and hypokalemia. Positive autoimmune markers such as ANA and anti-Ro antibodies support the diagnosis of Sjögren syndrome.

Electrocardiographic manifestations of severe hypokalemia include ST-segment depression, T-wave

Volume 9, Issue 3; May-June 2026; Page No 376-378

flattening, U waves, and arrhythmias. Early recognition is critical because severe hypokalemia can be fatal if untreated.

Management involves prompt potassium correction followed by alkali therapy to correct acidosis. Long-term management includes treatment of the underlying autoimmune disease and monitoring for recurrent electrolyte disturbances.

This case highlights the importance of considering Sjögren syndrome-related distal renal tubular acidosis in patients presenting with unexplained hypokalemic paralysis.

Conclusion

Hypokalemic periodic paralysis secondary to distal renal tubular acidosis is a rare but important manifestation of Sjögren syndrome. Patients presenting with acute flaccid paralysis and severe hypokalemia should undergo evaluation for renal tubular acidosis and autoimmune disorders. Early diagnosis and timely correction of electrolyte imbalance can significantly reduce morbidity and prevent life-threatening complications.

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Volume 9, Issue 3; May-June 2026; Page No 376-378