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Original Research Paper

Rheumatology

A RARE CASE OF SJOGREN'S SYNDROME PRESENTED WITH HYPOKALEMIC QUADRIPARESIS.

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ABSTRACT Sjögren's syndrome (SS) is a chronic autoimmune disorder primarily affecting exocrine glands, leading to dryness of the eyes and mouth. However, it can also involve multiple organ systems, including the renal tubules, resulting in renal tubular acidosis (RTA). Hypokalemic periodic paralysis (HPP) is a rare but serious manifestation of RTA in SS, caused by excessive urinary potassium loss. We report a case of a patient with primary Sjögren's syndrome presenting with acute flaccid paralysis due to severe hypokalemia. Laboratory findings confirmed distal RTA with metabolic acidosis and hypokalemia. Prompt potassium and bicarbonate supplementation led to symptom resolution. This case highlights the importance of considering SS in patients with unexplained hypokalemia and paralysis, as early recognition and management of RTA can prevent life-threatening complications. Clinicians should be aware of this rare but reversible association to ensure timely diagnosis and treatment.

KEYWORDS :

INTRODUCTION

Sjögren's syndrome (SS) is a chronic inflammatory systemic autoimmune disorder that primarily affects lacrimal and salivary glands due to lymphocytic infiltration. SS can lead to renal complications even before the onset of sicca symptoms. The most common renal manifestation of SS is chronic interstitial nephritis. Clinical presentations vary and may include Fanconi syndrome, distal renal tubular acidosis (dRTA), nephrogenic diabetes insipidus, or mild asymptomatic hypokalemia. Patients with dRTA often have few or no symptoms, leading to delayed diagnosis. If untreated, dRTA can result in serious acid-base imbalances, including hyperchloremic metabolic acidosis and critically low potassium levels.

CASE REPORT

A 25-year-old female presented to the emergency department with complaints of acute-onset bilateral upper and lower limb weakness for two days. She denied dysphagia, slurred speech, or diplopia. Her history revealed occasional swelling over the cheek region for the past two years and symptoms of gritty sensation in the eyes, photophobia, and itching for two months.

On examination, she was conscious and oriented. Central nervous system (CNS) examination revealed acute flaccid paralysis of all four limbs, reduced muscle tone, muscle power of 2/5, bilateral areflexia, and flexor plantar response.

ECG Findings: ST depression, T-U complex formation, and prolonged QT interval.

Arterial Blood Gas Analysis:
pH: 7.22, Bicarbonate: 6.2 mmol/L, pO2: 116 mmHg, pCO2: 15 mmHg, SpO2: 97%.

Electrolyte Panel:
- Serum Sodium: 143.3 mEq/L
- Serum Potassium: 1.63 mEq/L
- Serum Chloride: 127 mEq/L
- Serum Calcium: 8.3 mg/dL
- Serum Osmolality: 265 mOsm/kg

Urine Analysis:
- Urine Sodium: 22 mEq/L
- Urine Potassium: 10.3 mEq/L
- Urine Chloride: 20 mEq/L
- Urine Creatinine: 16 mg/dL
- Urine Urea: 76 mg/dL
- Urinary Anion Gap: 12.3
- Urine Osmolality: 330 mOsm/kg
- Urinary Osmolar Gap: 101 mOsm/kg

Additional Investigations:
- ANA profile: Positive for Anti-SS-A and Anti-SS-B antibodies.
- Ultrasound (USG) Abdomen and Pelvis: Normal findings.
- USG of Salivary Glands: Bilateral parotid and submandibular glands were smaller with multiple hypoechogenic areas, suggestive of parotitis.
- Schirmer's Test: Positive.

Findings confirmed hyperchloremic normal anion gap metabolic acidosis with a positive urinary anion gap, normal urinary osmolar gap, and urinary pH of 7, indicating distal renal tubular acidosis (dRTA) secondary to primary Sjögren's syndrome.

DISCUSSION AND CONCLUSION

Sjögren's syndrome (SS) is a systemic autoimmune disease that primarily targets exocrine glands but can also involve the kidneys, leading to distal renal tubular acidosis (dRTA). The prevalence of complete dRTA in patients with primary SS is reported to be approximately 5%. The hallmark of dRTA is the inability of the distal nephron to acidify urine, resulting in metabolic acidosis and hypokalemia.

Hypokalemia caused by dRTA can present as hypokalemic periodic paralysis (HPP), a severe condition characterized by sudden-onset muscle weakness. If left untreated, HPP can lead to respiratory muscle involvement and life-threatening complications.

Diagnosis of SS-related dRTA requires a high degree of clinical suspicion. Key diagnostic findings include metabolic acidosis with normal anion gap, urine pH > 5.5, positive urinary anion gap, and electrolyte imbalances. Autoimmune markers (anti-Ro/SSA, anti-La/SSB) support the diagnosis of SS.

Management includes potassium and bicarbonate supplementation to correct metabolic imbalances and prevent recurrent episodes of paralysis. Immunomodulatory therapy may be necessary in severe cases to control underlying autoimmune activity.

This case underscores the need for increased awareness of SS-related dRTA as a cause of HPP. Early recognition and prompt intervention can prevent complications, improve

patient outcomes, and highlight the systemic nature of SS beyond its classical sicca symptoms.

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