INTERNATIONAL JOURNAL OF PHARMACEUTICAL SCIENCES
[ISSN: 0975-4725; CODEN(USA): IJPS00]
Journal Homepage: https://www.ijpsjournal.com

OPEN ACCESS

Case Study

Primary Sjogren's Syndrome Presenting as Life-Threatening Hypokalemic Paralysis: A Diagnostic Challenge

Rohmotullah mondal*[1], Mohd Aheteshamuddin[2], Praveen kumar[3], Rohith A.[4]

[1,2]*Pharm D Intern, Aster CMI hospital*
[3]*Head clinical pharmacologist, Aster CMI hospital.*
[4]*Clinical pharmacologist, Aster CMI hospital*

ARTICLE INFO

Published: 16 Feb. 2025
Keywords:
Sjogren's syndrome, hypokalemic periodic paralysis, renal tubular acidosis, electrolyte imbalance, autoimmune disease, metabolic acidosis, neuromuscular disorder
DOI:
10.5281/zenodo.14878541

ABSTRACT

Sjögren's syndrome (SS) is an autoimmune disorder primarily affecting exocrine glands, leading to sicca symptoms such as dry eyes and dry mouth. However, extraglandular involvement, including renal tubular dysfunction, can significantly impact patient outcomes. One such renal complication is type 1 renal tubular acidosis (RTA), which can result in severe hypokalemia and lead to hypokalemic periodic paralysis (HPP). HPP is a rare neuromuscular disorder characterized by episodic muscle weakness or paralysis, often triggered by electrolyte imbalances. We report a case of a 36-year-old female presenting with progressive limb weakness, which was later diagnosed as HPP secondary to distal RTA associated with SS. Laboratory investigations confirmed severe hypokalemia, metabolic acidosis, and positive anti-SSA and anti-SSB antibodies. The patient responded well to potassium and bicarbonate replacement therapy, with complete resolution of symptoms. This case highlights the importance of recognizing SS as a potential cause of recurrent hypokalemia and neuromuscular dysfunction. Early diagnosis and intervention are crucial to preventing complications such as HPP and long-term renal damage. Clinicians should consider autoimmune disorders in patients with unexplained metabolic acidosis and neuromuscular symptoms.

INTRODUCTION

Sjögren's syndrome (SS) is a chronic autoimmune disorder characterized by lymphocytic infiltration of exocrine glands, leading to xerophthalmia and xerostomia {1}. The disease primarily affects middle-aged women and can occur as a primary disorder or secondary to other autoimmune diseases such as rheumatoid arthritis and systemic lupus erythematosus {2}. Beyond its classic glandular involvement, SS frequently presents

***Corresponding Author:** Rohmotullah mondal
Address: *Pharm D Intern, Aster CMI hospital*
Email ✉: rohmatullahmondal6000@gmail.com

Relevant conflicts of interest/financial disclosures: The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

with systemic manifestations, including vasculitis, interstitial lung disease, and neurological dysfunction {3}. Among these, renal involvement is a significant but often underrecognized complication {4}. Renal manifestations in SS include interstitial nephritis, nephrogenic diabetes insipidus, and, most commonly, type 1 renal tubular acidosis (RTA) {5}. Type 1 RTA, also known as distal RTA, results from impaired hydrogen ion secretion in the distal tubules, leading to hyperchloremic metabolic acidosis with inappropriately high urinary pH {6}. This dysfunction contributes to systemic acid-base imbalance and significant electrolyte disturbances, particularly hypokalemia {7}. Severe hypokalemia can precipitate hypokalemic periodic paralysis (HPP), a condition characterized by transient episodes of muscle weakness or paralysis due to potassium shifts affecting membrane excitability {8}. HPP in the context of SS is rare but can be a life-threatening presentation if unrecognized {9}. Patients with undiagnosed distal RTA and SS may present with recurrent episodes of weakness, fatigue, or even complete paralysis {10}. Given the potential severity of these complications, early identification and management of SS-related RTA and HPP are critical to preventing long-term morbidity {11}.

This case highlights a rare presentation of SS with HPP as the initial manifestation, emphasizing the importance of recognizing renal tubular dysfunction in autoimmune diseases {12}.

Case Description

A 36-year-old female presented to the emergency department with complaints of progressive weakness in both lower limbs, which had worsened over the past 24 hours. She also reported pain in her thighs and hands, difficulty walking, and recent onset of slurred speech. There was no history of fever, seizures, loss of consciousness, or gastrointestinal symptoms. She reported dry eyes and dry mouth, which she had experienced intermittently over the past few months. On examination, the patient was alert and oriented, with stable vital signs (temperature 98°F, heart rate 81 bpm, respiratory rate 12 bpm, blood pressure 142/96 mmHg, and SpO2 100%). Cardiovascular and pulmonary examinations were unremarkable. Neurological evaluation revealed significant limb weakness but preserved deep tendon reflexes and intact sensation. Laboratory findings included serum potassium of 2.1 mmol/L (normal: 3.5–5.0 mmol/L), serum bicarbonate of 14 mmol/L, and arterial blood gas showing metabolic acidosis with a normal anion gap. Urinalysis revealed a urine pH of 7.1, consistent with impaired acidification. An autoimmune workup was positive for anti-SSA and anti-SSB antibodies, confirming SS. MRI of the spine showed mild degenerative changes but no acute abnormalities. The patient was diagnosed with HPP secondary to SS-related distal RTA. She was treated with intravenous Magnesium sulphate, potassium chloride and sodium bicarbonate, Tab HCQ, resulting in gradual improvement in muscle strength. Oral potassium and bicarbonate therapy were continued upon discharge, and she was advised to follow up with rheumatology and nephrology specialists.

DISCUSSION

Renal involvement in SS is an often overlooked yet significant complication that can present with a range of symptoms, from mild electrolyte imbalances to life-threatening metabolic derangements {1}. Distal RTA, the most common renal manifestation of SS, is characterized by an inability to excrete hydrogen ions in the distal tubules, leading to metabolic acidosis {2}. The condition is typically associated with hyperchloremia and inappropriately high urinary pH (>5.5) {3}. One of the most serious consequences of distal RTA is hypokalemia, which can trigger periodic paralysis, particularly in susceptible individuals {4}. Hypokalemic periodic paralysis (HPP) is a disorder

characterized by transient episodes of severe muscle weakness due to potassium shifts affecting membrane excitability {5}. In autoimmune conditions like SS, the immune-mediated destruction of renal tubules can exacerbate potassium losses, predisposing patients to HPP {6}. The pathogenesis of SS-associated distal RTA involves lymphocytic infiltration and immune complex deposition in renal tubules, leading to tubulointerstitial nephritis {7}. This autoimmune process results in reduced hydrogen ion secretion, impaired potassium reabsorption, and chronic acid-base imbalance {8}. The combination of metabolic acidosis and potassium depletion can lead to severe neuromuscular symptoms, as seen in our patient {9}. Management of SS-related distal RTA involves correcting electrolyte disturbances with potassium and bicarbonate supplementation {10}. Long-term immunosuppressive therapy, including corticosteroids and hydroxychloroquine, may be required to control the underlying autoimmune process {11}. Regular monitoring of renal function and electrolyte levels is essential to prevent recurrent episodes of HPP and progressive renal dysfunction {12}. This case emphasizes the need for heightened awareness of SS-related renal complications, particularly in patients presenting with unexplained hypokalemia and neuromuscular symptoms {13}. Early diagnosis and appropriate management can prevent severe complications and improve patient outcomes {14}.

CONCLUSION

Sjogren's syndrome can present with a variety of systemic complications, including distal RTA leading to severe hypokalemia and periodic paralysis. This case illustrates the importance of recognizing renal involvement in SS, as early diagnosis and intervention can prevent life-threatening electrolyte disturbances. Clinicians should maintain a high index of suspicion for SS in patients with recurrent hypokalemia and neuromuscular symptoms. Timely electrolyte correction, immunosuppressive therapy, and regular follow-up are crucial in preventing disease progression and improving long-term outcomes.

REFERENCES

1. Fox RI. * Sjögren's syndrome: Diagnosis and treatment. Lancet. 2005;366(9482):321-31. doi:10.1016/S0140-6736(05)66990-5. [PubMed](https://pubmed.ncbi.nlm.nih.gov/16039337/)
2. Mavragani CP, Moutsopoulos HM.* The geoepidemiology of Sjögren's syndrome. Autoimmun Rev. 2010;9(5): A305-10. doi: 10.1016/j.autrev.2009.12.002. [PubMed](https://pubmed.ncbi.nlm.nih.gov/19932205/)
3. Siamopoulos KC, Elisaf M, Moutsopoulos HM.* Renal tubular acidosis in primary Sjögren's syndrome. Clin Rheumatol. 1992;11(2):226-30. doi:10.1007/BF02207372. [PubMed](https://pubmed.ncbi.nlm.nih.gov/1613700/)
4. Lin SH, Lin YF, Halperin ML.* Hypokalemia and paralysis: A practical approach to diagnosis and management. QJM. 2001;94(3):133-9. doi:10.1093/qjmed/94.3.133. [PubMed](https://pubmed.ncbi.nlm.nih.gov/11256897/)
5. Siamopoulos KC, Elisaf M, Drosos AA, Mavridis AA, Moutsopoulos HM.* Hypokalemic paralysis as the presenting manifestation of primary Sjögren's syndrome. Nephrol Dial Transplant. 1994;9(8):1176-8. [PubMed](https://pubmed.ncbi.nlm.nih.gov/7970070/)
6. Fujimoto T, Shiiki H, Takahi Y, Dohi K.* Primary Sjögren's syndrome presenting as hypokalemic periodic paralysis and respiratory arrest. Clin Rheumatol. 2001;20(5):365-8. doi:10.1007/s100670170050.

[PubMed](https://pubmed.ncbi.nlm.nih.gov/11642520/)

7. Pun KK, Wong CK, Tsui EY, Ho CS, Tam SC, Wang C.* Hypokalemic periodic paralysis due to Sjögren's syndrome in Chinese patients. Ann Intern Med. 1989;110(5):405-6. doi:10.7326/0003-4819-110-5-405. [PubMed](https://pubmed.ncbi.nlm.nih.gov/2913910/)
8. Tu WH, Shearn MA, Lee JC, Hopper J Jr.* Interstitial nephritis in Sjögren's syndrome. Ann Intern Med. 1968;69(6):1163-74. doi:10.7326/0003-4819-69-6-1163. [PubMed](https://pubmed.ncbi.nlm.nih.gov/5721980/)
9. Talal N, Zisman E, Schur PH.* Renal tubular acidosis, glomerulonephritis, and immunologic factors in Sjögren's syndrome. Arthritis Rheum. 1968;11(6):774-86. doi:10.1002/art.1780110604. [PubMed](https://pubmed.ncbi.nlm.nih.gov/4176463/)
10. Oh MS, Carroll HJ. * Value and determinants of urine anion gap in metabolic acidosis. Nephron. 2002;90(2):252-5. doi:10.1159/000054679. [PubMed](https://pubmed.ncbi.nlm.nih.gov/12187067/)
11. Kurtzman NA.* Disorders of distal renal acidification: Pathogenesis, diagnosis, and management. Kidney Int. 1990;38(5):720-7. doi:10.1038/ki.1990.266. [PubMed](https://pubmed.ncbi.nlm.nih.gov/2258600/)
12. Raynal C, Durupt S, Durieu I, Villard F, Berthoux F, Sepulchre J. Quadriparesis hypokaliémique avec acidose tubulaire distale révélatrice d'un syndrome de Gougerot-Sjögren primitif. Presse Med. 1999;28(35):1935-7. [PubMed](https://pubmed.ncbi.nlm.nih.gov/10573712/).

HOW TO CITE: Rohmotullah mondal*, Mohd Aheteshamuddin, Praveen kumar, Rohith A., Primary Sjogren's Syndrome Presenting as Life-Threatening Hypokalemic Paralysis: A Diagnostic Challenge, Int. J. of Pharm. Sci., 2025, Vol 3, Issue 2, 1281-1284. https://doi.org/10.5281/zenodo.14878541