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Original Article

HYPOKALEMIC PARALYSIS AS PRIMARY PRESENTATION OF SJÖGREN'S SYNDROME – A CLINICAL CASE REPORT

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Abstract

Background

Sjogren's syndrome is an autoimmune rheumatic disease characterized by lymphocytic infiltration of exocrine glands resulting in multi-system presentations including xerostomia and dry eyes.

Case Report

A 47-year-old woman presented with generalized weakness, difficulty in walking, nausea, and pain abdomen. She had reduced muscle power (2/5) with reduced tone, and absent deep tendon reflexes in all four limbs. Further investigation revealed severe hypokalemia with metabolic acidosis. She was treated with intravenous potassium chloride and diagnosed with Sjogren's syndrome based on her serology profile. After treatment, her condition improved.

Conclusion

Distal Renal Tubular Acidosis is a major cause of potassium loss in urine. Sjogren's Syndrome is an important cause of Distal Renal Tubular Acidosis, which can present as severe hypokalemia. Although it commonly presents with symptoms like dry eyes and dry mouth, it can rarely present as Distal Renal Tubular Acidosis and hypokalemia as the primary presentation.

Recommendation

Sjogren's syndrome should be suspected in patients with hypokalemia with features of Distal Renal Tubular Acidosis.

Keywords: *Sjogren's Syndrome, Hypokalemia, Hypokalemic Paralysis, Renal Tubular Acidosis.*
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Introduction

Sjögren's syndrome (SjS) is a chronic autoimmune disorder characterized by lymphocytic infiltration and dysfunction of exocrine glands, predominantly impacting the lacrimal and salivary glands. Clinically, hallmark manifestations include xerostomia (dry mouth) and keratoconjunctivitis sicca (dry eyes), but the disease is also known to have systemic implications. It can significantly compromise other organ systems, including pulmonary, renal, and neurologic functions, leading to a spectrum of complications that require comprehensive management and multidisciplinary care [1]. The female-to-male incidence ratio for the condition is 9:1, while the prevalence ratio stands at 10.72:1 [2]. The disease spectrum ranges from a benign glandular disorder to more aggressive systemic manifestations. Notably, renal involvement can occur in rare cases, sometimes preceding sicca symptoms. This renal involvement typically results in Renal tubular acidosis (RTA), leading to significant hypokalemia, a finding also observed in various autoimmune disorders such as systemic lupus erythematosus and Hashimoto's thyroiditis [3,4]. Distal renal tubular acidosis (dRTA) is characterized by a reduced capacity to maximally acidify urine (pH < 5.5) in the context of systemic acidosis, which is present in approximately 25% of patients [5,6]. This report describes a case of Sjögren's syndrome complicated by hypokalemic periodic paralysis due to dRTA, which was initially mismanaged as systemic lupus erythematosus (SLE).

Case Presentation

A 47-year-old Indo-Aryan Hindu female patient presented to the Emergency department of BMIMS, Pawapuri, Nalanda, with a 7-day history of bilateral generalized limb weakness, fatigue, and difficulty in walking. She reported associated symptoms including nausea and generalized abdominal pain for the past 2 days. The onset of generalized weakness was gradual and progressive, exhibiting symmetrical involvement that hindered daily activities such

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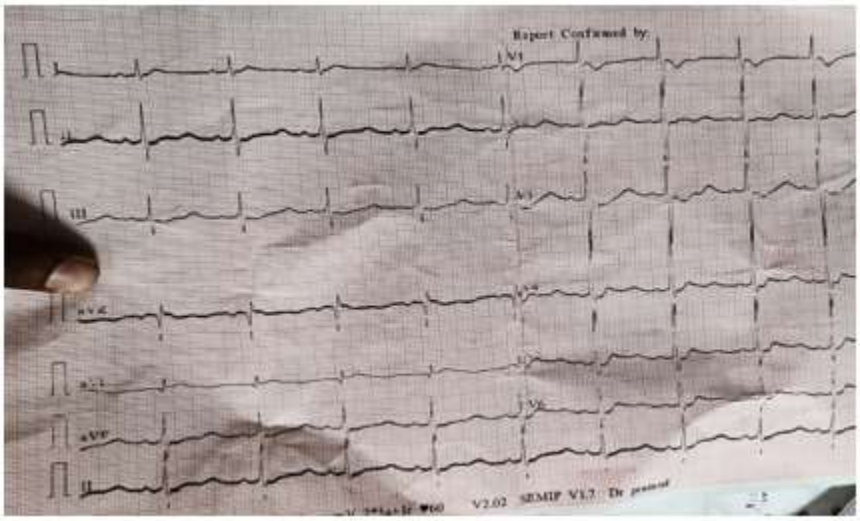
as combing hair, climbing stairs, and prolonged standing or walking. Notably, there were no myalgias, arthralgias, or signs of muscle atrophy.

The abdominal pain emerged suddenly, was described as burning, non-radiating, and was relieved by rest but exacerbated by exertion, with a pain severity rated at 5/10. She experienced associated odynophagia and constipation. The patient had a previous diagnosis of systemic lupus erythematosus (SLE) for 5 years, based on a positive antinuclear antibody (ANA) test. Her current regimen for SLE included hydroxychloroquine 200 mg twice daily. Additionally, she has a past medical history of recurrent hypokalemic limb weakness, for which she required potassium chloride administration during episodes, with no identifiable underlying cause; although initially responsive, symptoms were recurrently reported. No surgical interventions were performed, and the family's medical history was unremarkable.

Upon examination, the patient appeared ill but was alert and fully oriented. Clinical findings included pallor with no icterus, cyanosis, clubbing, or edema. Vital signs were stable, except for tachycardia at 118 beats per minute. The neurological assessment revealed muscle power graded at 2/5 in all four limbs, diminished tone, and absent deep tendon reflexes, while sensory function remained intact. Chest examination showed normal vesicular breath sounds and cardiac auscultation was unremarkable aside from normal heart sounds without murmurs. Abdominal palpation demonstrated generalized tenderness and distension.

Laboratory results indicated severe hypokalemia, with serum potassium levels at 1.6 mEq/L. The ECG exhibited ST segment depression, prolonged QT interval, and U waves, findings consistent with severe hypokalemia (Figure 1). The patient was admitted to the intensive care unit for electrolyte repletion and monitoring, receiving potassium chloride infusions along with supportive care.

Figure 1: ECG Findings Consistent with Severe Hypokalemia



Biochemistry		
	Serum	Urine
Arterial pH	7.31	6.5
Sodium(mEq/L)	137	87
Potassium(mEq/L)	1.6	32
Chloride (mEq/L)	110	112
Bicarbonate(mEq/L)	16	
Anion gap (mEq/L)	11	6
BUN (mg/dL)	11	
Creatinine (mg/dl)	0.8	
Serology		
ANA	Positive (1:160 by IFA)	
Anti-Ro (SSA)	Positive (241 AU/ml)	
Anti-La (SSB)	Positive (86 AU/ml)	

Table 1: Lab Parameters of The Patient

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Initially, ANA was positive while anti-dsDNA was negative. Subsequently, an Extractable Nuclear Antigen (ENA) panel was ordered, revealing positive anti-Sjögren's-syndrome-related antigen A (SSA) and anti-Sjögren's-syndrome-related antigen B (SSB), confirming a diagnosis of SjS (Table 1). The treatment regimen included intravenous methylprednisolone (1 g daily for 3 days), meropenem (1 g thrice daily), methotrexate (7.5 mg weekly), folic acid (5 mg 6 days weekly), and hydroxychloroquine (200 mg daily), potassium chloride syrup (15 ml thrice daily) were prescribed until her follow-up.

Following treatment, the patient's generalized weakness improved significantly, with motor power in all four limbs improved to 4/5 and normalization of tone and reflexes, including a normal plantar reflex response. After a successful ICU course, she was transferred to the general ward for continued management of SjS. Upon follow-up 15 days later, she denied any residual symptoms and was advised to return for evaluation in 2 months.

Discussion

Hypokalemia is characterized by plasma potassium levels below 3.5 mmol/L [7]. In clinical practice, renal manifestations of Sjögren's syndrome can lead to hypokalemia, frequently causing diagnostic challenges [8]. This case report aligns with the Surgical Case Report (SCARE) guidelines [9]. Hypokalemia can result from inadequate potassium intake or excessive loss, with the latter being more common. Potassium loss may occur via urinary or gastrointestinal (GI) routes, or through shifts into cells. GI losses typically arise from diarrhea or vomiting, while intracellular shifts can be triggered by insulin therapy, sympathetic overactivity, or thyrotoxicosis. Urinary potassium loss is often linked to conditions such as primary hyperaldosteronism, Cushing's syndrome, and RTA [10].

In this case, the patient displayed severe hypokalemia with concurrent ECG abnormalities. Arterial blood gas analysis revealed metabolic acidosis with low bicarbonate levels, indicative of RTA. This may be due to either primary hereditary factors or secondary processes, with SjS being a notable contributor to secondary RTA [11]. SjS typically presents with xerostomia and keratoconjunctivitis sicca, affecting 70-80% of patients, and prompting parotid gland enlargement in about 20% [12]. Systemic manifestations occur in 30-40% of cases, with renal involvement being the most common non-exocrine manifestation, typically presenting as tubulointerstitial nephritis, with distal RTA as the predominant renal presentation [13].

RTA refers to a spectrum of disorders affecting bicarbonate reabsorption and hydrogen ion secretion. This condition is marked by a maintained glomerular filtration rate (GFR) along with a distinct hyperchloremic metabolic acidosis (HMA). dRTA involves impaired hydrogen ion secretion in the distal nephron, and severe forms lead to an inability to excrete acid loads, resulting in progressive hydrogen ion retention and HMA [14]. While diminished H+-ATPase activity is the most frequent defect in dRTA, several other underlying defects can also impair hydrogen ion secretion [15]. dRTA is often associated with renal potassium wasting, occasionally severe enough to precipitate paralysis or respiratory failure.

The lymphocytic infiltration accompanying SjS may lead to further autoimmune complications such as primary biliary cholangitis and obstructive bronchiolitis. Although distal RTA is commonly observed in this context, it frequently remains asymptomatic and undiagnosed. In adults, autoimmune diseases are significant etiologies for dRTA, making a thorough evaluation of these conditions essential, especially in cases of idiopathic dRTA [15,16]. Importantly, hypokalemia is the most frequent electrolyte disturbance linked to distal RTA. In this patient, the absence of classic Sjögren's symptoms like xerostomia and dry eyes, combined with a history of recurrent hypokalemia, highlights the necessity of recognizing systemic complications as critical diagnostic indicators, as demonstrated in this case.

Conclusion

Sjogren's syndrome is an uncommon but significant etiology of severe hypokalemia, particularly in patients diagnosed with distal renal tubular acidosis (RTA). It is critical to consider this syndrome in differential diagnoses, as it may be erroneously identified as systemic lupus erythematosus or other metabolic and rheumatological disorders in individuals presenting with pronounced hypokalemia.

Recommendation

The prompt recognition and diagnosis of Sjögren's syndrome pose significant challenges due to existing diagnostic criteria that may lead to frequent oversight. Patients exhibiting hypokalemic paralysis attributable to renal tubular acidosis (RTA) must undergo a comprehensive evaluation for underlying primary disorders, including Sjögren's syndrome, even in the absence of hallmark symptoms typically associated with the disease.

List Of Abbreviations

SjS: Sjögren's syndrome
RTA: Renal tubular acidosis
dRTA: Distal renal tubular acidosis
GI: Gastrointestinal
SLE: Systemic lupus erythematous
ANA: Antinuclear antibody

Author Contributions

I, Binay Kumar Mahto, served as the lead and corresponding author, overseeing the manuscript's drafting and conducting the data analysis. Aditya Prakash was responsible for table preparation and the design of the analytical framework. Sant Kumar facilitated the data collection and contributed conceptual insights for the article development. Rashmi Kumari, Head of the Department of Medicine at BMIMS Pawapuri Nalanda, provided supervision throughout the manuscript drafting process and granted final approval for publication. All authors reviewed and approved the version of the manuscript submitted for publication.

Data Availability

Information was gathered using a thorough method that involved interviewing patients, reviewing medical records, and assessing laboratory reports. The authors will avail data upon request.

Declaration Of Patient Consent

The authors confirm that all necessary patient consent forms have been secured, and both the patient and their family members have consented to the publication of the clinical information in the journal. It is acknowledged that while the patient's name and initials will remain undisclosed and efforts will be made to protect their identity, complete anonymity cannot be assured.

Conflict Of Interest

The authors declared no conflicts of interest concerning the authorship and/or publication of this case report.

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References

1. Brito-Zerón P, Baldini C, Bootsma H, Bowman SJ, Jonsson R, Mariette X, Sivils K, Theander E, Tzioufas A, Ramos-Casals M (7 July 2016). "Sjögren syndrome". Nature Reviews Disease Primers. 2 (1): 16047. https://doi:10.1038/nrdp.2016.47
2. Aiyegbusi, O., McGregor, L., McGeoch, L. et al. Renal Disease in Primary Sjögren's Syndrome. Rheumatol Ther 8, 63–80 (2021). https://doi.org/10.1007/s40744-020-00264-x
3. Siddamreddy S, Dandu VH. Thyrotoxic Periodic Paralysis. [Updated 2023 Jul 24]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2024 Jan-. Available from: https://www.ncbi.nlm.nih.gov/books/NBK560670/
4. Jung, S. W., Park, E. J., Kim, J. S., Lee, T. W., Ihm, C. G., Lee, S. H., Moon, J. Y., Kim, Y. G., & Jeong, K. H. (2017). Renal Tubular Acidosis in Patients with Primary Sjögren's Syndrome. *Electrolyte & blood pressure : E & BP*, *15*(1), 17–22. https://doi.org/10.5049/EBP.2017.15.1.17
5. Giglio, S., Montini, G., Trepiccione, F. *et al.* Distal renal tubular acidosis: a systematic approach from diagnosis to treatment. *J Nephrol* **34**, 2073–2083 (2021). https://doi.org/10.1007/s40620-021-01032-y
6. Poux JM, Peyronnet P, Le Meur Y, Favereau JP, Charmes JP, Leroux-Robert C. Hypokalemic quadriplegia and respiratory arrest revealing primary Sjögren's syndrome. Clin Nephrol. 1992;37(4):189-191.
7. In: Kasper DLD, Fauci ASA, Hauser SLS, et al, eds. *Harrison's Manual of Medicine*. McGraw Hill Inc.; 2020. https://harrisons.unboundmedicine.com/harrisons/view/Harrisons-Manual-of-Medicine/623468/all/HYPOKALEMIA. Accessed December 26, 2024.
8. Sedhain A, Acharya K, Sharma A, Khan A, Adhikari S. Renal tubular acidosis and hypokalemic paralysis as a first presentation of primary Sjögren's syndrome. Case Reports in Nephrology. 2018 Oct 16;2018:1–4. https://doi.org/10.1155/2018/9847826
9. Agha RA, Franchi T, Sohrabi C, Mathew G, Kerwan A, Thoma A, et al. The SCARE 2020 Guideline: Updating Consensus Surgical CASE Report (SCARE) guidelines. International Journal of Surgery. 2020 Dec 1;84:226–30. https://doi.org/10.1016/j.ijsu.2020.10.034
10. Kardalas E, Paschou SA, Anagnostis P, Muscogiuri G, Siasos G, Vryonidou A. Hypokalemia: a clinical update. Endocrine Connections. 2018 Apr 1;7(4):R135–46. https://doi.org/10.1530/EC-18-0109
11. Zhou Y, Huang N, Cao G, Luo Y. Type I renal tubular acidosis caused by Sjögren's syndrome with hypokalemia as the first symptom: a case report. Journal of International Medical Research. 2020;48(2). https://doi:10.1177/0300060519880765
12. Mortazavi, H., Baharvand, M., Movahhedian, A., Mohammadi, M., & Khodadoustan, A. (2014). Xerostomia due to systemic disease: a review of 20 conditions and mechanisms. Annals of medical and health sciences research, 4(4), 503–510. https://doi.org/10.4103/2141-9248.139284

13. Duan, N., Li, Z., Fan, Y. et al. Related factors of renal injury in primary Sjögren's syndrome. Immun Ageing 20, 48 (2023). https://doi.org/10.1186/s12979-023-00375-3
14. Bagga A, Sinha A. Evaluation of renal tubular acidosis. Indian J Pediatr. 2007 Jul;74(7):679-86. doi: 10.1007/s12098-007-0120-0. PMID: 17699978.
15. Sengul E, Bunul F, Yazici A, et al. An Unusual Initial Presentation of Sjögren's Syndrome: Severe Hypokalemic Paralysis Secondary to Distal Renal Tubular Acidosis. Eurasian J Med. 2013;45(3):218-221. Doi:10.5152/eajm.2013.43
16. Krishna Bahadur Sodari, Pragyan Basnet, Krishna Adhikari. Sjogren's Syndrome in the Mask of Hypokalemia: A Case Report. J Nepal Med Assoc 2024;62(280):841-3. Doi:10.31729/jnma.8836

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