Annals of Medicine and Medical Sciences (AMMS)
Volume 04, 2025, *Page No.: 881 to 886*
Available at: *http://ammspub.com*

Case Report

OPEN ACCESS

Paralysis Unveiling Autoimmunity: A Case of Hypokalemic Paralysis due to Distal Renal Tubular Acidosis as Presenting Manifestation of Sjogren's Syndrome

Aditya Kalwaghe [1], Rishikesh Kololikar [2], Manjit Sisode [3], Nirmalkumar Rawandale [4], Shubham Gedam [5]

[1]Junior Resident, Department of Internal Medicine, Government Medical College, Miraj, India.
[2]Senior Resident, Department of Internal Medicine, Government Medical College, Miraj, India.
[3]Assistant Professor, Department of Internal Medicine, Shri Bhausaheb Hire Government Medical College, Dhule, India.
[4]Professor & Head of Department, Department of Internal Medicine, Shri Bhausaheb Hire Government Medical College, Dhule, India.
[5]Junior Resident, Department of Internal Medicine, Shri Bhausaheb Hire Government Medical College, Dhule, India.

*Corresponding Author: Aditya, Kalwaghe; *aditya.kalwaghe211@gmail.com*

Abstract

Sjogren's syndrome is a chronic autoimmune disorder primarily affecting exocrine glands, but it can also present with extra-glandular manifestations, including renal and neurological complications. Distal renal tubular acidosis is a rare but significant renal manifestation of Sjogren's syndrome, often asymptomatic but can lead to severe hypokalemia. We report a case of a 44-year-old woman who presented with acute flaccid quadriparesis due to severe hypokalemia. Initial evaluation revealed normal central nervous system imaging. Laboratory investigations revealed hyperchloremic normal anion gap metabolic acidosis, elevated urine potassium-creatinine ratio, and alkaline urine, leading to a diagnosis of distal renal tubular acidosis. Further workup demonstrated positivity for SS-A/Ro and SS-B/La antibodies, along with clinical features of dry eyes and dry mouth, confirming Sjogren's syndrome as the underlying etiology. Patient was treated with intravenous potassium followed by long-term potassium citrate, sodium bicarbonate, and immunosuppressive therapy. With appropriate management, she showed complete resolution of symptoms and remained stable during follow-up. This case highlights the importance of recognizing renal involvement as an early or atypical presentation of Sjogren's syndrome, particularly in patients without overt sicca symptoms, and emphasizes the need for heightened clinical suspicion for systemic autoimmune disorders in patients presenting with unexplained hypokalemic paralysis.

Keywords: ***Distal renal tubular acidosis, electrolyte imbalance, extra-glandular manifestations, hypokalemic paralysis, sjogren's syndrome.***

Introduction

Sjogren's syndrome (SS) is a chronic systemic autoimmune disorder primarily affecting exocrine glands, characterized by lymphocytic infiltration and impaired gland secretion [1]. SS typically presents with hallmark clinical features of dry eyes (xeropthalmia) and dry mouth (xerostomia), collectively referred to as sicca syndrome [2]. However, in rare cases, it may present with non-sicca symptoms affecting other organ systems. While SS primarily targets salivary and lacrimal glands, it may also involve many extra-glandular systems, including joints, skin, nervous system, lungs, kidneys, etc[3].

The renal system is less commonly involved in SS, but is clinically significant. The typical renal complication is tubulointerstitial nephritis (TIN), caused due to infiltration by inflammatory cells and deposition of immune complexes [4]. This inflammation and the autoantibodies can cause various renal abnormalities, clinically presenting as Fanconi syndrome, diabetes insipidus, distal renal tubular acidosis (dRTA), Bartter syndrome, Gitelman syndrome, and nephrolithiasis [3]. Among them, dRTA is the most common renal manifestation [1]. It is frequently asymptotic or presents with minimal symptoms, often leading to delayed or missed diagnoses. However, in some cases, dRTA can cause significant disturbance in acid-base and electrolyte homeostasis, including metabolic acidosis and hypokalemia [5]. This imbalance

Received: July 02, 2025; **Revised:** July 22, 2025; **Accepted:** July 26, 2025

can manifest as symptoms ranging from muscle cramps to severe complications like hypokalemic paralysis, cardiac arrhythmias, and even respiratory arrest [6].

The nervous system is also less commonly affected by SS but can present as peripheral neuropathy, cognitive dysfunction, autonomic dysfunction (e.g. orthostatic hypotension), etc [7]. Additionally, it may be secondarily involved due to electrolyte disturbances resulting from renal involvement, such as hypokalemia which can cause muscle cramps, weakness and even flaccid paralysis in severe cases.

Here, we report a case of a middle-aged female with hypokalemic paralysis caused due to distal renal tubular acidosis (dRTA), which is an atypical initial presentation of primary Sjogren's syndrome (pSS). Severe hypokalemia can lead to life threatening complications. Hence this case underscores the importance of recognizing systemic complications in SS to facilitate timely diagnosis and management.

Case Description

A 44-year-old women presented to the emergency department with complaints of extreme generalized muscle weakness. She started experiencing muscle weakness about four days ago which was progressive and progressed to a point of difficulty getting out of bed and complete loss of muscle power. She denied having experienced any similar episode before. The patient also mentioned a history of foreign body sensation in both eyes and dryness of mouth since past 1 year, for which she had not sought any medical care. She was cachexic and had insignificant past medical or family history.

On physical examination, she was afebrile and vitally stable. Her neurological examination revealed symmetric flaccid quadriparesis, and revealed no contraction or movements in all four limbs (motor power of 0/5 as per the Medical Research Council (MRC) Muscle Strength Grading Scale), without any sensory deficits, cranial nerve involvement, or altered mental status. All deep tendon reflexes were absent. Patient denied experiencing diplopia, dysphagia, slurring of speech, bladder involvement, or convulsions. Her cardiovascular, respiratory and gastrointestinal, and thyroid examinations were otherwise unremarkable. Her ECG showed changes consistent with hypokalemia, including presence of prolongation of QT interval, U-waves, ST segment depression, and T-wave inversion (Fig. 1.).

Laboratory investigations from admission blood work revealed severe hypokalemia (serum potassium of 1.7 mEq/L) (Table I). CNS imaging studies of brain and spinal cord ruled out any central nervous system pathology (Fig. 2.). Diagnosis of hypokalemic paralysis was made owing to the hypokalemia and normal CNS imaging.

Arterial blood gas analysis from admission blood work revealed an anion gap of 10 and metabolic acidosis (pH of 7.12). Urine analysis revealed pH of 7.2 and a urine potassium-creatinine ratio of around 25.79 mEq/mmol (Table I). Urinalysis was suggestive of urinary potassium loss. Patient denied any history of vomiting, diarrhea, diuretic or laxative use. In view of hypokalemic hyperchloremic normal anion gap metabolic acidosis, elevated urine potassium-creatinine ratio, and alkaline urine, a diagnosis of Type I or Distal renal tubular acidosis (dRTA) complicated with hypokalemia was made. Additionally, there were no signs indicating proximal tubular dysfunction.

Further workup regarding the cause of dRTA was performed. The administration of ammonium chloride at a dose of 0.1 g/kg induced metabolic acidosis, however her urinary pH remained above 6.5, this result supported the diagnosis of dRTA. She was not using any relevant medication causing dRTA, like lithium, amphotericin B, and NSAIDs. Thyroid profile was within normal limits, and viral markers were also negative (HIV, hepatitis B, and hepatitis C). All these findings pointed to an autoimmune origin of dRTA. Ultrasound of abdomen and pelvis was normal and excluded nephrocalcinosis or obstructive uropathy.

Screening for autoimmune diseases revealed strongly positive for SS-A/Ro60, SS-A/Ro52, and SS-B/LA, while rheumatoid factor, anti-CCP, anti-ds DNA, Anti-histones, Anti-SmD1, Anti-PNCA, Anti-Scl-70, Anti-U1-snRNP, Anti-AMA-M2, Anti-Jo-1, and Anti-PM-Scl antibody were all negative. Ophthalmology consultation was ordered, examination revealed dryness in both eyes, with a positive schirmer test (3 mm in the left eye, 4 mm in the right eye). Ocular staining with fluorescein and lissamine green was done (total score of 10). Following this, salivary gland biopsy was considered, which was refused by the patient. The autoantibody pattern and clinical findings of sicca symptoms pointed towards a diagnosis of primary Sjogren's syndrome (score of 5 according to the ACR/EULAR 2016 classification criteria). This case was considered primary SS, since patient's clinical and laboratory results did not meet the full diagnostic criteria for secondary SS associated with conditions like systemic lupus erythematosus, rheumatoid arthritis, scleroderma, and primary biliary cirrhosis.

Finally, after considering the patient's history, clinical findings, laboratory investigations, autoantibody profile, and after ruling out other potential causes of dRTA, a final diagnosis of hypokalemic paralysis secondary to distal renal tubular acidosis due to primary Sjogren's syndrome was made.

She was admitted to intensive care unit in view of severe hypokalemia for continuous cardiac monitoring and electrolyte replacement. Potassium correction was initiated and was treated with infusion of intravenous (IV) potassium in normal saline. Her potassium levels were closely monitored to assess for potential rebound hyperkalemia. By admission day 4, patient's potassium was completely corrected (serum potassium of 3.9 mEq/L) (Fig. 3.) and remained stable between 3.5 - 4.0 mEq/L, leading to resolution of her paralysis (motor power of 5/5 in all her limbs). In few days the patient was able to walk independently and regained full strength in all limbs.

She was discharged with oral potassium citrate 1080 mg tablet thrice daily and oral sodium bicarbonate (NaHCO3) 650 mg tablet twice daily. Considering the dRTA associated with SS, the patient was also started on a daily dose of 60 mg prednisone for 4 weeks, followed by tapering to 10 mg per day, 50 mg azathioprine, and 200 mg hydroxychloroquine. She was advised and counseled for regular diet, regular monitoring, and artificial tears for her ocular problems.

At 1-, 3-, and 6- months follow-up, she is currently asymptomatic, with serum potassium levels of 3.9, 3.7, and 4.1 mEq/L respectively. Potassium citrate and NaHCO3 doses were adjusted according to repeat laboratory testing. Patient maintains good compliance with her prescribed treatment and is scheduled for regular follow-ups.

Table I: Laboratory investigations - on admission and at discharge values

Lab Investigations	On admission	At discharge	Reference range
Sodium	140	144	132 – 156 mEq/L

Potassium	1.7	3.9	3.5 - 5.5 mEq/L
Calcium	9.1	9.6	8.6 - 10.2 mg/dL
Magnesium	1.9	1.8	1.7 - 2.2 mg/dL
Chloride	121	109	104 – 112 mEq/L
Urea	26	32	19 – 48 mg/dL
Creatinine	0.9	1.1	0.7 - 1.3 mg/dL
Albumin	3.9	3.7	3.2 - 4.8 mg/dL
SGOT	35	37	8 – 40 U/L
SGPT	34	29	7 – 56 U/L
Anion gap	10	9	<12
pH	7.12	7.39	7.38 - 7.42
HCO_3	10	30	22 – 26 mEq/L
PCO_2	39	42	35 – 45 mmHg
PO_2	91	98	75 – 100 mmHg
Urine pH	7.2	-	<5.5 - Acidic >7.0 - Alkaline
Urinary sodium	12.5	-	45 - 106 mEq/L
Urinary potassium	143	-	25 – 125 mEq/L
Urinary creatinine	5.54	-	4 – 11 mmol/L
Urine potassium-creatinine ratio	25.8	-	< 1.5 mEq/mmol
TSH	3.2	-	0.4 – 4.0 mIU/L
fT3	2.83	-	2.14 – 4.09 pg/mL
fT4	0.98	-	0.89 – 1.76 ng/dL

Abbreviation 1: SGOT - Serum Glutamic Oxaloacetic Transaminase, SGPT - Serum Glutamic Pyruvic Transaminase, HCO_3^- - Bicarbonate, PCO_2 - Partial Pressure of Carbon Dioxide, PO_2 - Partial Pressure of Oxygen.

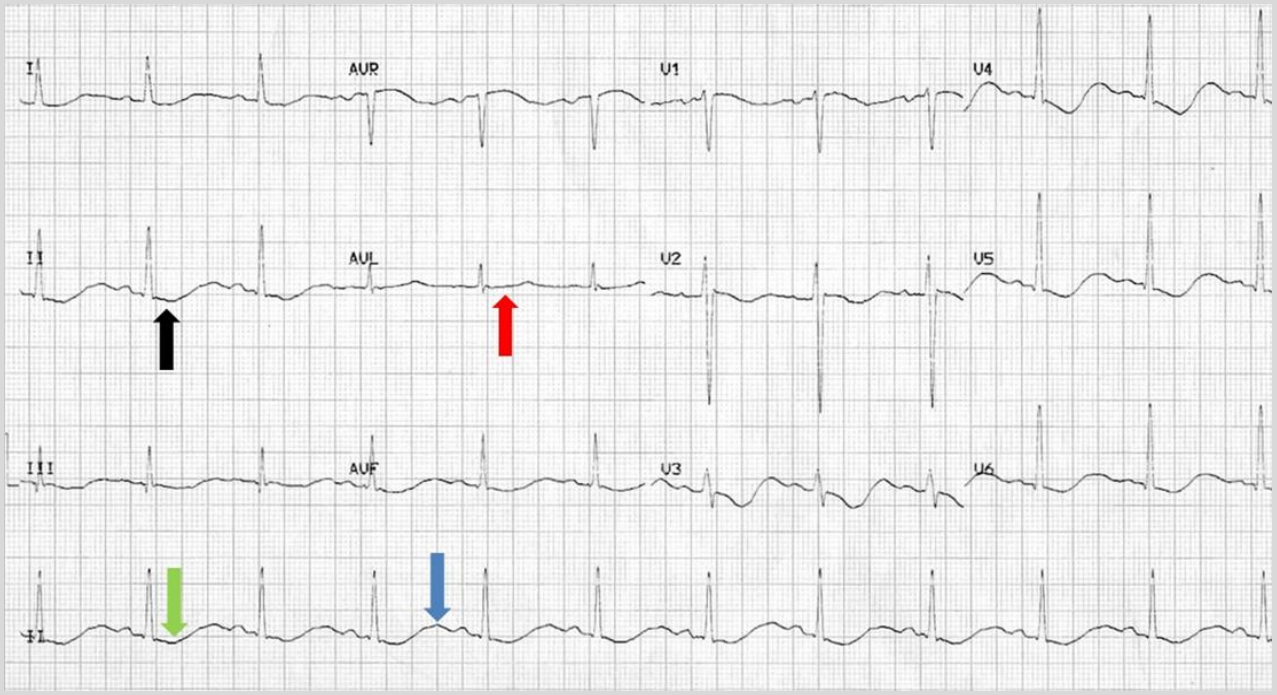


Fig. 1: ECG of patient showing prolongation of QT interval (red arrow), U-waves (blue arrow), ST segment depression (black arrow), and T-wave inversion (green arrow).

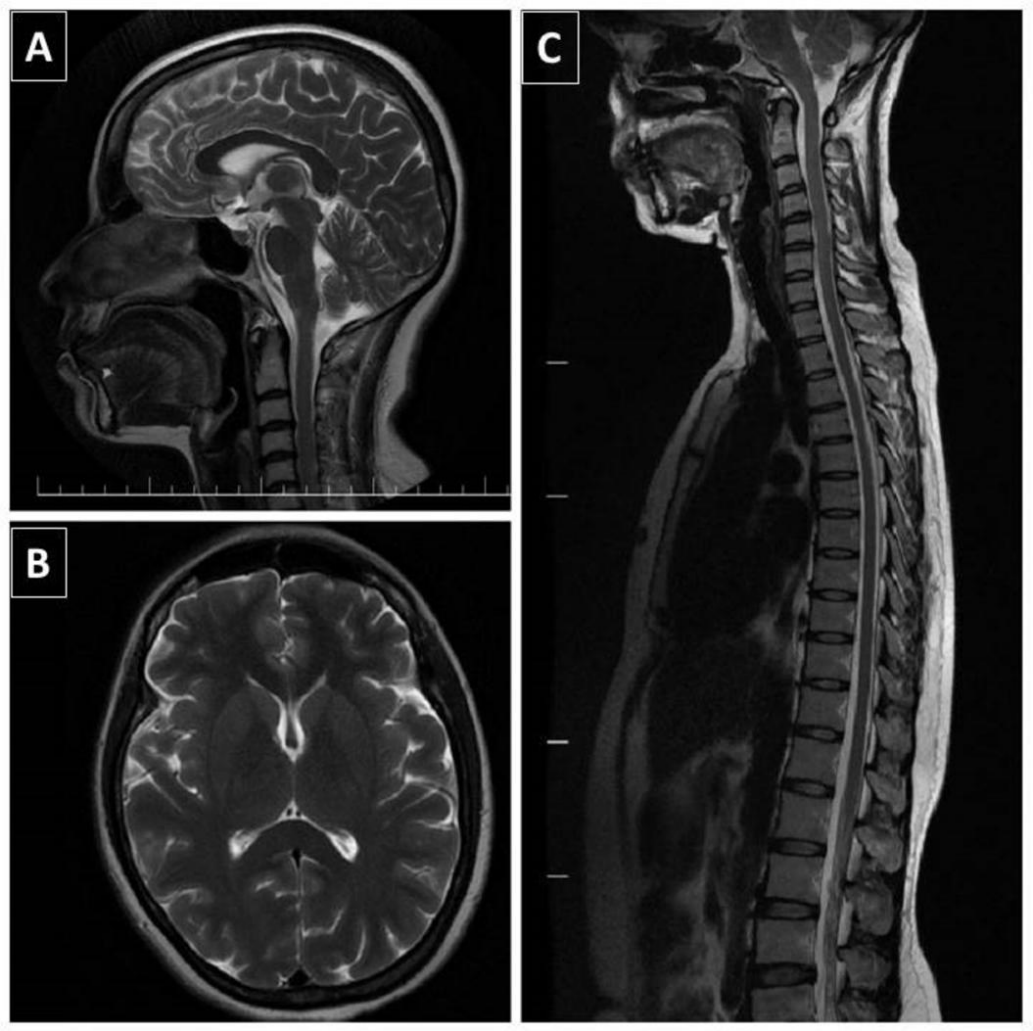
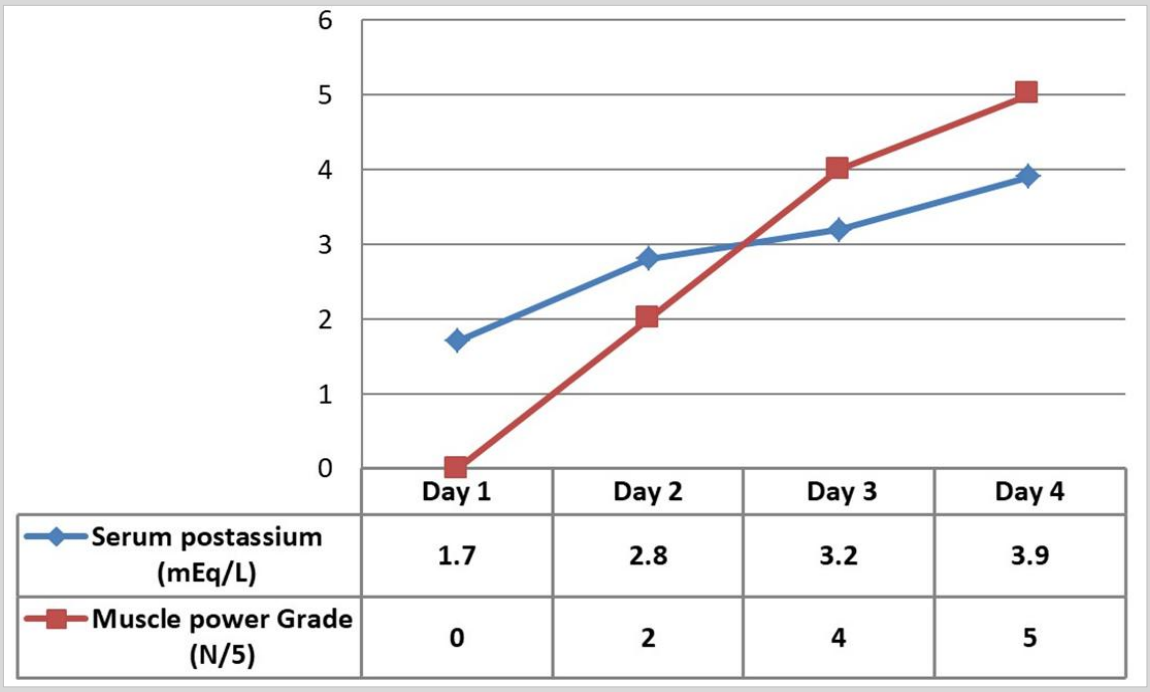


Fig. 2: Neurological MRI of the patient (A) Sagittal T2-weighted MRI of the brain showing no abnormalities, normal morphology of cerebral hemisphere, and no focal or diffuse region of abnormal signal intensity. (B) Axial T2-weighted MRI of the brain showing normal ventricular size and parenchymal signal intensity. (C) Sagittal T2-weighted MRI of the cervico-thoracic spine showing normal spinal cord morphology and signal intensity.



	Day 1	Day 2	Day 3	Day 4
Serum postassium (mEq/L)	1.7	2.8	3.2	3.9
Muscle power Grade (N/5)	0	2	4	5

Fig. 3: Graphical representation of patient's serum potassium (mEq/L) and muscle power grade (MRC grade - N/5) over a period of first 4 days of admission.

Discussion

Primary Sjogren's syndrome (pSS) is an autoimmune disorder that occurs as an isolated condition, whereas secondary SS occurs in association with other autoimmune disorders like rheumatoid arthritis, systemic lupus erythematosus, or systemic sclerosis [8]. SS typically affects exocrine glands presenting with sicca symptoms such as xerostomia and xeropthalmia, however can present with extra-glandular involvement even before clinically significant sicca symptoms, particularly renal manifestation such as dRTA leading to complications such as hypokalemic paralysis [9].

The age of presentation of our patient is in the early 40s, which was lower than the average age of 52.7 years in a cohort of 400 pSS patients reported by García-Carrasco *et al*.[9]. Additionally, study conducted by Shiozawa *et al*. suggest that patients with renal involvement in pSS tend to be younger at diagnosis [10]. The younger age of presentation in some cases of pSS-associated RTA has been noted in prior studies. Goroshi *et al*. reported a mean age of 33.1 ± 8.22 years for hypokalemic paralysis in pSS, much younger than the typical age of pSS diagnosis (52.7 years) [1]. That suggests that renal manifestation may develop earlier in the disease course, possibly preceding other systemic features. The female predominance of pSS is also notable. Our patient's profile is consistent with the data from García-Carrasco *et al*., where they found a significant female predilection (female-to-male ratio of 14:1) [9].

Renal involvement in pSS has been well documented, with tubulointerstitial nephritis being the most common underlying pathology. In the cohort of 400 pSS patients by García-Carrasco *et al*. reported renal dysfunction in 6% patients [9]. Distal RTA is the most frequently reported renal dysfunction, followed by nephrogenic diabetes insipidus and proximal RTA [1]. Several studies have assessed the prevalence of renal involvement in pSS. The study by Ren *et al*. found that among 130 patients with pSS, 73.1% developed RTA, and nine presented with hypokalemic paralysis [11]. The findings of Ram *et al*. further support the strong association between pSS and RTA, with 34.8% of RTA cases in their cohort being attributed to pSS [5]. Also Shiozawa *et al*. reported that patient with renal tubular defects may have longer disease duration compared to those without renal abnormalities [10]. These findings suggest that dRTA-induced hypokalemia is a significant but under-recognized extra-glandular manifestation of pSS.

The exact mechanism underlying dRTA in Sjogren's syndrome is still not fully understood. Immunocytochemical studies of renal biopsy samples in several patients have demonstrated total lack of the H-ATPase pump in the intercalated cells of the collecting tubules, leading to defective acid secretion, as described by DeFranco and *et al*.[12]. However, the process by which immune-mediated damage of the H-ATPase function is still not well understood. Additionally, the development of hypokalemia in dRTA is linked to multiple factors, including autoantibodies targeting carbonic anhydrase II, impaired H-K ATPase function, secondary hyperaldosteronism, and excessive urinary bicarbonate loss [13,14].

The link between hypokalemic paralysis and pSS has been documented in multiple studies. Case series reported by Goroshi *et al*. found that 68% of patients with pSS were diagnosed only after experiencing hypokalemic paralysis, while just 38% of patients initially presented with sicca symptoms [1]. Khandelwal *et al*. emphasized that SS should be considered in any patient presenting with hypokalemic paralysis from RTA, even in the absence of sicca symptoms [2]. Our patient, who initially denied dryness symptoms but later admitted to neglected sicca complains upon further evaluation, exemplifies this pattern. These studies underscore the importance of considering pSS in patients with dRTA and hypokalemia, even in the absence of classic sicca symptoms.

In our case, the patient presented with acute flaccid quadriparesis due to severe hypokalemia, with dRTA being identified as the underlying cause. After detailed evaluation and patient lacking clinically significant sicca symptoms and any other autoimmune condition, the diagnosis of pSS was made. This pattern was also observed in the study done by Nahar *et al*. that many pSS patient presented with hypokalemic paralysis, and found that 61.5% patient had recurrence of hypokalemic paralysis with patients experiencing multiple episodes, and emphasized the importance of proper treatment and regular monitoring of serum potassium levels in these patients [15].

In conclusion, our case reinforces the notion that primary Sjogren's syndrome can present with severe electrolyte disturbances and neuromuscular complication before the onset of sicca symptoms. The clinical implications are significant: clinicians must maintain a high index of suspicion for pSS in patients with unexplained hypokalemic paralysis, particularly those with concurrent metabolic acidosis and alkaline urine. Timely recognition and treatment of pSS-associated dRTA can prevent recurrent paralysis, chronic kidney injury, and systemic disease progression. Future research should focus on the pathophysiological mechanisms linking autoimmunity to renal tubular dysfunction and identifying biomarkers for early detection of renal involvement in pSS.

Declarations

Ethical Clearance

Written informed consent was obtained from the patient. Ethics committee approval was not required.

Acknowledgements

None

Conflict of interest

None

Funding/ financial support

None

Contributors

Aditya Kalwaghe, Junior Resident, Department of Internal Medicine, Government Medical College, Miraj, India
Rishikesh Kololikar, Senior Resident, Department of Internal Medicine, Government Medical College, Miraj, India
Manjit Sisode, Assistant Professor, Department of Internal Medicine, Shri Bhausaheb Hire Government Medical College, Dhule, India
Nirmalkumar Rawandale, Professor & Head of Department, Department of Internal Medicine, Shri Bhausaheb Hire Government Medical College, Dhule, India
Shubham Gedam, Junior Resident, Department of Internal Medicine, Shri Bhausaheb Hire Government Medical College, Dhule, India

References

[1] Goroshi M, Khare S, Jamale T, Shah NS. Primary Sjogren's syndrome presenting as hypokalemic paralysis:

A case series. J Postgrad Med 2017; 63: 128-131. DOI: 10.4103/0022-3859.194224

[2] Khandelwal D, Bhattacharya S, Khadgawat R, Kaur S, Tandon N, Ammini AC. Hypokalemic paralysis as a presenting manifestation of primary Sjögren's syndrome: A report of two cases. Indian J Endocrinol Metab 201; 16: 853-855. DOI: 10.4103/2230-8210.100684

[3] Aiyegbusi O, McGregor L, McGeoch L, Kipgen D, Geddes CC, Stevens KI. Renal Disease in Primary Sjögren's Syndrome. Rheumatol Ther 2021; 8: 63-80. DOI: 10.1007/s40744-020-00264-x

[4] Tu WH, Shearn MA, Lee JC, Hopper J Jr. Interstitial nephritis in Sjögren's syndrome. Ann Intern Med 1968; 69: 1163-1170. DOI: 10.7326/0003-4819-69-6-1163

[5] Ram R, Swarnalatha G, Dakshinamurty KV. Renal tubular acidosis in Sjögren's syndrome: a case series. Am J Nephrol 2014; 40: 123-130. DOI: 10.1159/000365199

[6] François H, Mariette X. Renal involvement in primary Sjögren syndrome. Nat Rev Nephrol 2016; 12: 82-93. DOI: 10.1038/nrneph.2015.174

[7] Perzyńska-Mazan J, Maślińska M, Gasik R. Neurological manifestations of primary Sjögren's syndrome. Reumatologia 2018; 56: 99-105. DOI: 10.5114/reum.2018.75521

[8] Peri Y, Agmon-Levin N, Theodor E, Shoenfeld Y. Sjögren's syndrome, the old and the new. Best Pract Res Clin Rheumatol 2012; 26: 105-117. DOI: 10.1016/j.berh.2012.01.012

[9] García-Carrasco M, Ramos-Casals M, Rosas J, Pallarés L, Calvo-Alen J, Cervera R, *et al*. Primary Sjögren syndrome: clinical and immunologic disease patterns in a cohort of 400 patients. Medicine (Baltimore) 2002; 81: 270-280. DOI: 10.1097/00005792-200207000-00003

[10] Shiozawa S, Shiozawa K, Shimizu S, Nakada M, Isobe T, Fujita T. Clinical studies of renal disease in Sjögren's syndrome. Ann Rheum Dis 1987; 46: 768-772. DOI: 10.1136/ard.46.10.768

[11] Ren H, Wang WM, Chen XN, Zhang W, Pan XX, Wang XL, *et al*. Renal involvement and followup of 130 patients with primary Sjögren's syndrome. J Rheumatol 2008; 35: 278-284.

[12] DeFranco PE, Haragsim L, Schmitz PG, Bastani B. Absence of vacuolar H(+)-ATPase pump in the collecting duct of a patient with hypokalemic distal renal tubular acidosis and Sjögren's syndrome. J Am Soc Nephrol 1995; 6: 295-301. DOI: 10.1681/ASN.V62295

[13] Lin SH, Cheema-Dhadli S, Gowrishankar M, Marliss EB, Kamel KS, Halperin ML. Control of excretion of potassium: lessons from studies during prolonged total fasting in human subjects. Am J Physiol 1997; 273: F796-800. DOI: 10.1152/ajprenal.1997.273.5.F796

[14] Pertovaara M, Bootorabi F, Kuuslahti M, Pasternack A, Parkkila S. Novel carbonic anhydrase autoantibodies and renal manifestations in patients with primary Sjogren's syndrome. Rheumatology (Oxford) 2011; 50: 1453-1457. DOI: 10.1093/rheumatology/ker118

[15] Nahar N, Nahar P. Hypokalemic Paralysis Due to Primary Sjögren's Syndrome: Case Series. J Assoc Physicians India 2022 ; 70: 11-12.