Case Report

Metabolic mimic of neurological emergency: Hypokalemic quadriparesis due to distal renal tubular acidosis in Sjögren syndrome

Nivetha kaliyamoorthy1*, Ramanathan2

[1]Junior resident, Department of emergency medicine, Kauvery Hospital, Tennur, Trichy, Tamil Nadu
[2]HOD, Department of emergency medicine, Kauvery Hospital, Tennur, Trichy, Tamil Nadu
*Correspondence

Abstract

Background: Distal renal tubular acidosis (dRTA) is an uncommon but important cause of severe hypokalemia and non-anion-gap metabolic acidosis. When due to Sjögren syndrome, the presentation may be subtle—or dramatically life-threatening.

Key words: Distal renal tubular acidosis (dRTA); Sjogren syndrome; Tubulointerstitial nephritis

Case presentation: A 40-year-old woman presented in a gasping state with sudden-onset quadriparesis and inability to lift her neck. Initial evaluation revealed profound hypokalemia (1.2 mmol/L), severe non-anion-gap metabolic acidosis (pH 6.87; HCO_3 9.8–15 mmol/L), type II respiratory failure, and urinary potassium wasting with alkaline urine and positive. urinary anion gap. Autoimmune evaluation showed positive ANA, strongly positive anti-Ro and anti-SSB, with xerostomia and xerophthalmia, confirming primary Sjogren syndrome. She required emergency intubation and aggressive potassium correction (up to 380 mEq/day). Sodium bicarbonate was withheld initially to prevent worsening hypokalemia. Clinical recovery was rapid, with extubation on day 3 and complete reversal of neuromuscular weakness.

Conclusion: Sjögren syndrome–associated distal RTA can present catastrophically with hypokalemic paralysis and respiratory failure. Early recognition and correction of electrolyte imbalances, ongoing potassium losses with Supplementation helps in early recovery.

Citation: Nivetha kaliyamoorthy, Ramanathan. Metabolic mimic of neurological emergency: Hypokalemic quadriparesis due to distal renal tubular acidosis in Sjögren syndrome. Kauverian Med J. 2026;3(4):44-50.

Academic Editor: Dr. Venkita S. Suresh

ISSN: 2584-1572 (Online)

1. Introduction

Sjögren syndrome is a systemic autoimmune disorder with well-recognized extraglandular manifestations, including renal involvement. Tubulointerstitial nephritis is the most common renal pathology, and distal (type 1) renal tubular acidosis is a classical functional consequence of tubular injury.

Distal renal tubular acidosis results from impaired hydrogen ion secretion in the collecting duct, leading to non-anion gap metabolic acidosis, inappropriately alkaline urine, and renal potassium wasting. Although hypokalemia in this setting is usually mild, severe potassium depletion can rarely lead to acute neuromuscular paralysis.

Life-threatening presentations with respiratory muscle involvement are uncommon and may mimic acute neurological emergencies. This case highlights a dramatic initial presentation of previously undiagnosed Sjögren syndrome manifesting as severe hypokalemic quadriparesis and type II respiratory failure due to distal renal tubular acidosis.

2. Case Presentation

A 40-year-old woman with known hypothyroidism (Thyronorm 50 μg/day) and chronic polyarthralgia, dry mouth, dry eyes presented to the emergency department in a gasping state.

3. History of illness

At 12 AM, she developed sudden weakness of both upper and lower limbs, progressing over a few hours to involve neck muscles, leading to inability to lift her head by early morning. She remained conscious and able to vocalize initially. She was evaluated at a local hospital where hypokalemia was noted but not treated. During transport to our hospital she became unresponsive and breathless. No drug exposure, diarrhoea, vomiting, or toxin use was reported.

On arrival

	On arrival to ER	After stabilization
Airway	Gasping; not verbalising	Intubated with 7.5 mm ETT
Breathing	Inadequate rate and depth	98% with 100%fio2 on MV
Circulation	BP:60/ HR:56/min	130/90 mmhg with noradrenaline, HR: 87 BPM, CVS: S1,S2 heard.
Disability	E1V1M1, Bilateral pupil 3mm SRTL	E1VTM1, Bilateral pupil 2mm,RTL
Exposure	Temp: normal, CBG:304mg/dl,Alopecia, dry mouth, caries of incisors, xerostomia	-

CNS examination	
On arrival to ER, before kcl correction	Next day, after kcl correction:
E1vtM1, Pupil ERTL, Tone: normal, DTR: Areflexia, Bilateral plantar mute. Power, cerebellar signs, sensations: could not be assessed.	E4VtM6 Bilateral pupil ERTL No CN palsy Tone: normalpower:4/5 in all 4 limbs(very Minimal proximal muscle weakness)DTR: 2+ in all 4 limbs Plantar: b/l flexorc No cerebellar signs ,gain could not be assessed

4. POCUS

- Adequate LV function
- IVC diameter 1.79 cm
- IJV collapsible
- Lungs: Bilateral A-profile
- Abdomen: No free fluid
- DVT scan: Negative

ABG

pH	6.87
Po2	137
PCo2	8
Lac	1.4
Na	142
K	1.2
Cl	114
Hco3	15.5

5. Investigations

CBC

Hb	10
TC	13000
Plt	235000
RFT	Urea/creatinine:33.5/0.86
LFT	Normal
Electrolytes	148/1.2/121/13
HbA1C	5.0
p.acetone	negative
Cpk	52U/L
Procal	0.34
TSH	5.21
CT brain	Normal

Echocardiography

- Ostium secundum ASD (22 mm)
- RA/RV dilation
- Mild pulmonary hypertension (TRPG 30 mmHg)
- Ultrasound abdomen:
- Liver hemangioma
- 4 mm calculus in left lower calyx

Urine analysis

- Urinary k+ 12.5 mmol/L
- Urinary Na+ 75 mmol/L
- Urinary Cl- 63 mmol/L
- Urinary anion gap +24.5
- Urine pH alkaline (>5.5)

The patient was treated with multiple high dose IV pottassium chloride, her weakness got improved, but her serial serum electrolyte analysis showed persistent metabolic acidosis.

Na	K	Cl	HCO_3	AG
142	1.2	121	9.8	9
150	4.9	130	12	8
157	5.3	138	10	11
138	3.5	116	20	2

Autoimmune workup

- ANA strongly positive (nuclear pattern)
- Anti-Ro (SSA): 54 (positive)
- Anti-SSB: 35 (positive)
- Anti-dsDNA negative
- Schirmer test: Almost absent tear secretion

6. ***Differential diagnosis***

S No	Diagnosis	Comment
1	Hypokalemic periodic paralysis	Would have normal AG, normal urine acidification, no acidosis, no autoimmune features.
2	Guillain–Barré syndrome	Would show sensory/autonomic involvement; reflexes absent but acidosis and hypokalemia unexplained.
3	Thyrotoxic periodic paralysis	Mild elevation of TSH ; no acidosis.
4	Toxic ingestion (e.g., toluene)	No exposure history; autoimmune markers strongly positive.
5	Distal renal tubular acidosis (autoimmune)	Fits all findings: alkaline urine, positive UAG, hypokalemia, NAGMA, urinary K wasting, Sjögren features- xerostomia, xerostomia, incisor caries,Schirmer positive, anti RO,anti SSB-positive.

Final diagnosis

Primary Sjögren syndrome presenting with distal (type 1) RTA causing severe hypokalemic quadriparesis and respiratory failure.

7. Treatment

On presentation, the patient was managed as a medical emergency because of gasping respiration, altered sensorium, and hemodynamic instability. Immediate airway protection was achieved with endotracheal intubation and mechanical ventilation. Circulatory support was initiated with intravenous fluids followed by noradrenaline infusion, resulting in stabilization of blood pressure.

Given the presence of profound hypokalemia with acute neuromuscular paralysis and respiratory muscle involvement, aggressive potassium replacement was prioritised as the definitive therapy. A central venous catheter was inserted to permit safe administration of high-concentration potassium chloride and enable close biochemical monitoring. Potassium was replaced using a combination of intravenous potassium chloride infusions and oral supplementation, titrated according to frequent serum potassium measurements. The patient required exceptionally high doses, receiving approximately 380 mEq of potassium on the first day, consistent with severe total body potassium depletion due to renal potassium wasting.

Despite the severity of metabolic acidosis, sodium bicarbonate therapy was deliberately withheld during the acute phase. Alkali administration was avoided because it can exacerbate hypokalemia by shifting potassium intracellularly and increasing renal potassium losses, thereby worsening neuromuscular weakness and increasing the risk of cardiac arrhythmias. Once serum potassium levels stabilised, potassium-based alkali therapy in the form of oral potassium citrate was initiated to facilitate gradual correction of the metabolic acidosis.

Supportive care included ventilatory support until respiratory muscle strength recovered, artificial tear supplementation for ocular dryness, and close intensive care monitoring. No immunosuppressive therapy was initiated during the acute phase, as the patient's presentation was driven by a reversible metabolic derangement. Plans were made

for rheumatology follow-up for further evaluation and long-term management of primary Sjögren syndrome.

Day	IV kcl	Oral kcl	Total	Potassium mrng/ Eve	Potassium after correction
Day 1	360 mEq	(Kcl)15ml, 15ml=1.5gm=20mEq	380	1.1 2.1	4.9(stopped) 3.6
Day 2	40 mEq	(Kcitrate)30ml 1 ml=2mEq	100	3.6	5.3
Day 3	160 mEq	90 ml (180mEq)	340	5.3(stopped)	3.5
Day 4	-	30 ml (60mEq)	60	3.5, 4.4	4.4 5.3
Day 5	-	30 ml	60	-	5.0
Day 6	-	30 ml	60	-	4.6
Day 7	-	30 ml	60	-	4.4

8. Outcome and follow-up

With aggressive potassium correction, the patient demonstrated rapid neurological improvement. Limb strength improved within 24 hours, deep tendon reflexes returned, and neck muscle power normalised. Respiratory effort improved in parallel, allowing successful extubation on the third day of hospitalisation. There was no residual motor deficit at discharge. Metabolic acidosis improved gradually with potassium-based alkali therapy. The patient was advised regular follow-up with rheumatology and nephrology for ongoing management of Sjögren syndrome and 5distal renal tubular acidosis.

9. Discussion

Sjogren syndrome is a systemic autoimmune disorder in which renal involvement represents an important but often under-recognised extraglandular manifestation. Tubulointerstitial nephritis is the predominant renal pathology, with distal (type 1) renal tubular acidosis being a classical functional consequence. Autoimmune-mediated injury to α-intercalated cells in the collecting duct leads to impaired hydrogen ion secretion, failure of urinary acidification, and persistent non-anion gap metabolic acidosis.[1,2]

Defective proton secretion in distal renal tubular acidosis results in an inappropriately alkaline urine despite systemic acidosis. Concurrent dysfunction of H^+-ATPase and H^+/K^+-ATPase channels causes renal potassium wasting, predisposing patients to hypokalemia.[3] Although mild hypokalemia is frequently encountered, severe potassium depletion leading to acute neuromuscular paralysis and respiratory failure is distinctly uncommon.[4] In the present case, profound hypokalemia precipitated abrupt quadriparesis with involvement of respiratory muscles, culminating in type II respiratory failure requiring emergency ventilatory support.

The rapid progression of limb weakness to neck and respiratory muscle paralysis posed a diagnostic challenge and closely mimicked acute neurological emergencies such as Guillain–Barré syndrome, myelopathy, or hypokalemic periodic paralysis. However, the presence of severe non-anion gaps metabolic acidosis, persistently alkaline urine, positive urinary anion gap, and significant urinary potassium wasting strongly favoured distal renal tubular acidosis over primary neuromuscular disorders. [5] The rapid and

complete reversibility of weakness following potassium correction further supported a metabolic etiology.

Autoimmune evaluation was pivotal in identifying the underlying cause. Chronic sicca symptoms, poor dentition, alopecia, markedly reduced tear secretion on Schirmer testing, and strongly positive anti-Ro and anti-SSB antibodies established the diagnosis of primary Sjogren syndrome. Renal involvement may precede overt sicca features or remain clinically silent for years, and in some patients, life-threatening metabolic derangements may represent the initial presentation of disease.[6,7]

Management of severe hypokalemia in distal renal tubular acidosis requires prompt and aggressive potassium replacement with close monitoring. Despite the presence of severe metabolic acidosis, sodium bicarbonate was deliberately withheld during the acute phase, as alkali therapy can exacerbate hypokalemia by promoting intracellular potassium shifts and increasing renal potassium losses. Potassium-based alkali therapy was introduced only after stabilization of serum potassium levels. Rapid neurological recovery following potassium correction in this case highlights the reversibility of hypokalemic paralysis when promptly recognized and appropriately treated.

This case underscores the importance of considering autoimmune distal renal tubular acidosis in patients presenting with sudden paralysis and metabolic acidosis. Early recognition of this potentially fatal yet reversible condition enables targeted therapy, avoids unnecessary neurological investigations, and significantly improves clinical outcomes.

References

[1] Maripuri S, Grande JP, Osborn TG, et al. Renal involvement in primary Sjögren's syndrome: a clinicopathologic study. Clin J Am Soc Nephrol. 2009; 4:1423–31.

[2] Bossini N, Savoldi S, Franceschini F, et al. Clinical and morphological features of kidney involvement in primary Sjögren's syndrome. Nephrol Dial Transplant. 2001; 16:2328–36.

[3] Walsh SB, Shirley DG, Wrong OM, Unwin RJ. Urinary acidification assessed by simultaneous furosemide and fludrocortisone treatment. Clin J Am Soc Nephrol. 2007; 2:98–103.

[4] Lin SH, Lin YF. Proximal renal tubular acidosis with hypokalemic paralysis. Am J Kidney Dis. 2001; 37:658–62.

[5] Rodríguez Soriano J. Renal tubular acidosis: the clinical entity. J Am Soc Nephrol. 2002; 13:2160–70.

[6] Goules AV, Tatouli IP, Moutsopoulos HM, Tzioufas AG. Clinically significant renal involvement in primary Sjögren's syndrome. Clin Exp Rheumatol. 2013; 31:20–7.

[7] Pertovaara M, Korpela M, Kouri T, Pasternack A. The occurrence of renal involvement in primary Sjögren's syndrome. Clin Nephrol. 1999; 51:11–7.