Crit Care Shock (2026) 29:69-72

Severe hypokalemic paralysis as the initial manifestation of distal renal tubular acidosis secondary to primary Sjogren's syndrome

Fathimathu Afrah[1], Pulkit Mehrotra[1], Vengadakrishnan[1]

Abstract

Profound hypokalemia can occasionally present as acute paralysis. If not recognised promptly, the outcome can be catastrophic. Among the several causes of potassium loss, distal renal tubular acidosis (dRTA) is uncommon. It arises as a part of an autoimmune process such as primary Sjögren's syndrome. We describe a fifty-six-year-old woman who came to the Emergency Department with the sudden onset of flaccid quadriparesis and breathlessness for 1 day. Investigations revealed severe hypokalemia (1.7 mmol/l) accompanied by a normal-anion-gap metabolic acidosis. Urinary analysis confirmed dRTA, and further evaluation identified previously undiagnosed primary Sjögren's syndrome. After timely potassium correction, alkali therapy, and short-course corticosteroids, the patient recovered fully neurologically. This case highlighted the importance of evaluating the cause of hypokalemia and considering Sjögren's syndrome even in the absence of other initial manifestations.

Keywords: Hypokalemia, paralysis, renal tubular acidosis, autoimmune disease, Sjögren's syndrome.

Introduction

Acute hypokalemic paralysis is one of those emergencies that remind us how metabolic disorders can masquerade as neurological disease. The differential diagnosis ranges from periodic paralysis and endocrine causes to renal and gastrointestinal potassium loss. (1) When renal wasting is responsible, the culprit is often a tubular acidification defect rather than simple diuretic use.

[1.] Department of General Medicine, Sri Ramachandra Institute of Higher Education and Research, Chennai, Tamil Nadu, India

Address for correspondence:
Dr. Fathimathu Afrah
Department of General Medicine, Sri Ramachandra Institute of Higher Education and Research, Chennai, Tamil Nadu, India
Tel: +918122269555
Email: afrah.rawoof@gmail.com

Distal renal tubular acidosis reflects the kidney's inability to secrete hydrogen ions in the collecting duct. (2,3) The urine, therefore, remains inappropriately alkaline, and potassium is lost, setting the stage for profound hypokalemia. (4)

Sjögren's syndrome, although classically recognised for its exocrine involvement, affects the kidney in nearly one-third of cases. (5) Tubulointerstitial nephritis and distal renal tubular acidosis (dRTA) are the most frequent renal lesions. (5)

The case that follows illustrates this sequence vividly and underscores why electrolyte disturbances in women should prompt an autoimmune screen.

Case presentation

A 56-year-old post-menopausal woman, previously healthy, awoke in the early hours with weakness of both lower limbs, then progressed to both upper limbs, following which she was unable to lift her head from the pillow. The patient had acute-onset breathlessness on admission. There were no sensory symptoms, diplopia, bulbar complaints, or recent illnesses. She denied diarrhoea, vomiting, or drug intake.

On arrival, she was alert and afebrile. Patient was tachypneic with a respiratory rate of 28/min, peripheral oxygen saturation of 95% on room air, pulse 55/min, and blood pressure 120/70 mmHg. Neurological examination showed symmetrical flaccid quadriparesis with power 1/5, absent deep tendon reflexes, flexor plantar responses, and intact sensations. As the patient had progressive respiratory fatigue, she required short-term mechanical ventilation and intensive care unit (ICU) care.

Arterial blood gas analysis showed pH 7.28 with a normal anion gap. The electrocardiogram displayed sinus bradycardia and a first-degree atrioventricular block. Routine investigations are listed in **Table 1**, which revealed a low serum potassium level of 1.7 mmol/l. Patient was treated with calcium gluconate infusion and potassium chloride, cautiously under cardiac monitoring, followed by oral potassium citrate and spironolactone.

Liver and renal function tests were otherwise within normal limits. Initial urine pH remained 7.0 despite systemic acidosis, urine sodium 149 mmol/l, urine chloride 85 mmol/l, urine potassium 9.97 mmol/l, urine bicarbonate 5.7 mmol/l, and urine anion gap was positive. These results confirmed the setting of dRTA.

Autoimmune testing revealed a positive anti-nuclear antibody (ANA) (speckled pattern) and strong reactivity to anti-Sjögren's syndrome-related antigen A (anti-SSA) (Ro), anti-Sjögren's syndrome type B (anti-SSB) (La), and Ro-52 antibodies. Rheumatoid factor and anti-cyclic citrullinated peptide (anti-CCP) were negative. Schirmer's test showed <5 mm wetting in 5 minutes, confirming tear-film deficiency. Salivary-gland ultrasonography demonstrated diffuse heterogeneity without obstruction. The 2016 American College of Rheumatology and the European Alliance of Associations for Rheumatology (ACR/EULAR) classification criteria revealed a score of 4 points, satisfying the threshold for primary Sjögren's syndrome. Collectively, the findings established primary Sjögren's syndrome with dRTA.

The patient was treated with potassium chloride and spironolactone. Prednisolone 40 mg daily and hydroxychloroquine 200 mg at bedtime were introduced, together with pilocarpine and lubricating eye drops. Within two days, serum potassium normalised, and muscle power improved to 4/5. She was extubated on the third day. The patient walked independently on the day of discharge. Six weeks later, she remained symptom-free with stable electrolytes on low-dose steroids and alkali replacement.

Discussion

dRTA represents a failure of the distal nephron to generate a steep hydrogen-ion gradient, so the urine cannot be acidified. (2) This leads to the accumulation of hydrogen ions, resulting in a normal-anion-gap metabolic acidosis. To preserve electrical neutrality, potassium excretion increases, leading to hypokalemia. (6) Over time, the chronic acid retention also favours bone demineralisation and nephrocalcinosis.

In autoimmune disorders such as Sjögren's syndrome, lymphocytic infiltration of the renal interstitium and circulating antibodies against the H^+-ATPase, anion-exchanger 1 proteins disrupt tubular proton transport. (7,8) François and Mariette estimated that roughly one in four patients with primary Sjögren's developed renal involvement, most commonly dRTA. (9) Clinically, the triad of hypokalemia, normal-anion-gap acidosis, and persistently alkaline urine is diagnostic. (2,5) Other causes of hypokalemic paralysis—such as thyrotoxic periodic paralysis, proximal RTA, or gastrointestinal potassium loss—show different biochemical patterns and are easily distinguished once urine studies are obtained.

Management combines rapid potassium repletion, correction of systemic acidosis with alkali salts, and immunosuppression to control the underlying autoimmune inflammation. (2,9) Our patient's swift improvement once therapy began mirrored these observations. Continued alkali supplementation and periodic monitoring help prevent relapse and chronic kidney injury.

Conclusion

When a middle-aged woman presents with acute flaccid paralysis, normal-anion-gap acidosis, and alkaline urine, dRTA must be considered. Autoimmune screening is essential, as Sjögren's syndrome presents initially as hypokalemic paralysis before other manifestations. Recognising this link allows prompt potassium and alkali correction and the initiation of immunotherapy—interventions that can restore complete neuromuscular function and preserve long-term renal health.

Table 1. Routine investigations

Investigation	Values	Reference
Hemoglobin	12.7 g/dl	12–15g/dl
Total white blood cells	10,360 cells/mm^3	4000–11000 cells/mm^3
Platelet count	233,000/mm^3	150,000–450,000/mm^3
Serum sodium	139 mmol/l	136–145 mmol/l
Serum potassium	1.7 mmol/l	3.5–5.5 mmol/l
Serum chloride	11.7 mmol/l	98–107 mmol/l
Serum bicarbonate	10 mmol/l	22–24 mmol/l
Serum calcium	8.9 mg/dl	8.6–10 mg/dl
Serum magnesium	2.2 mg/dl	1.6–2.4 mg/dl

References

1. Fox RI. Sjögren's syndrome. Lancet 2005;366: 321–31.
2. Rodríguez-Soriano J. Renal tubular acidosis: the clinical entity. J Am Soc Nephrol 2002; 13:2160–70.
3. Stinebaugh BJ, Schloeder FX, Tam SC, Goldstein MB, Halperin ML. Pathogenesis of distal renal tubular acidosis. Kidney Int 1981;19:1–7.
4. Comer DM, Droogan AG, Young IS, Maxwell AP. Hypokalaemic paralysis precipitated by distal renal tubular acidosis secondary to Sjögren's syndrome. Ann Clin Biochem 2008; 45:221–5.
5. Maripuri S, Grande JP, Osborn TG, Fervenza FC, Matteson EL, et al. Renal involvement in primary Sjögren's syndrome: a clinicopathologic study. Clin J Am Soc Nephrol 2009;4: 1423–31.
6. Batlle D, Moorthi KMLST, Schlueter W, Kurtzman N. Distal renal tubular acidosis and the potassium enigma. Semin Nephrol 2006;26: 471–8.
7. Routsias JG, Tzioufas AG. Sjögren's syndrome--study of autoantigens and autoantibodies. Clin Rev Allergy Immunol 2007;32:238–51.
8. Fayyaz A, Kurien BT, Scofield RH. Autoantibodies in Sjogren's syndrome. Rheum Dis Clin North Am 2016;42:419–34.
9. François H, Mariette X. Renal involvement in primary Sjögren syndrome. Nat Rev Nephrol 2016;12:82–93.