Hypokalaemia Induced by Renal Tubular Acidosis in a Patient with Sjogren's Syndrome: A Case Report

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ABSTRACT

In this case study, a 38-year woman was diagnosed with distal renal tubular acidosis (RTA) secondary to Sjogren's Syndrome (SS) after presenting with recurrent episodes of sudden-onset quadriparesis, hypokalaemia (serum potassium: 1.07 mmol/L), and metabolic acidosis (HCO_3^-: 15.9 mmol/L). Positive anti-SSA/Ro antibodies (55.96 U/mL), an elevated erythrocyte sedimentation rate (ESR: 65 mm/1st hour), and autoimmune serology results confirmed the diagnosis even in the absence of characteristic sicca symptoms. Distal RTA, a recognised SS complication, was confirmed by the patient's alkaline urine (pH 7.0) and high spot urine potassium-to-creatinine ratio (43 mEq/g). Potassium supplements, sodium bicarbonate for acidosis, and immunosuppression with steroids, hydroxychloroquine, and azathioprine were all part of the treatment. In addition to highlighting the significance of autoimmune examination in patients with unexplained hypokalaemia and RTA, this case also shows the diagnostic difficulties of SS in the absence of classic glandular signs. To avoid complications and improve the outcomes, it is essential to identify and treat SS-related renal and electrolyte abnormalities as soon as possible.

Key Words: *Sjogren's Syndrome, Renal tubular acidosis, Hypokalaemia, Autoimmune disease, Anti-SSA/Ro antibodies.*

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INTRODUCTION

Sjogren's Syndrome (SS) is a chronic autoimmune condition characterised by lymphocytic infiltration of the exocrine glands, leading to typical symptoms such as xerostomia (dry mouth) and keratoconjunctivitis sicca (dry eyes).[1] SS is a systemic illness that can affect the kidneys as well as other organs. Although less frequent, renal symptoms include renal tubular acidosis (RTA), glomerulonephritis, and interstitial nephritis.[2] In the context of SS, hypokalaemia frequently occurs as a result of RTA, where the kidneys struggle to eliminate hydrogen ions and reabsorb bicarbonate, resulting in metabolic acidosis and the subsequent loss of potassium.[3] Patients with SS may exhibit hypokalaemia in a variety of ways, from asymptomatic to severe, requiring immediate diagnosis and treatment. These symptoms may include fatigue, muscle weakness, cardiac arrhythmias, and even respiratory failure.[4] The autoimmune-mediated destruction of the renal tubules, specifically the distal nephron's intercalated cells, that regulates acid-base homeostasis, is the pathophysiology linking SS to RTA and hypokalaemia.[5]

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The intricate mechanisms underlying this connection are not yet fully understood, hindering timely diagnosis due to the ambiguous symptoms and the necessity for heightened clinical vigilance.

This case report outlines a specific clinical situation involving a patient with SS who experienced hypokalaemia as a result of RTA, highlighting the diagnostic and treatment difficulties faced. Furthermore, a thorough literature review is included to examine the epidemiology, pathophysiology, clinical characteristics, and management approaches for RTA and hypokalaemia in SS.

CASE REPORT

A 38-year married female presented with complaints of intermittent, abrupt-onset quadriparesis for six months. The weakness was exacerbated by household tasks and was temporarily alleviated by intravenous potassium supplementation. She experienced several instances of vomiting over four days, along with extreme weakness (muscle power 2/5 proximally and distally). She denied having diarrhoea, rash, diplopia, or indications of sicca. Significant hypokalaemia (K^+ 1.07 mmol/L), metabolic acidosis (HCO_3^- 15.9 mmol/L, pH 7.40), and a high spot urine potassium-to-creatinine ratio (43 mEq/g) were found (Table I). Distal RTA was suspected during acidosis, as the urine pH remained inappropriately alkaline (7.0).

Autoimmune serology showed positive antinuclear antibodies (1+ speckled), anti-SSA/Ro (55.96 U/mL), and anti-dsDNA (55.00 IU/ml), along with elevated erythrocyte sedi-

mentation rate (ESR: 65 mm/1st hour) and rheumatoid factor (25.9 IU/mL). Anti-SSB/La, Sm, Scl-70, and U1-RNP antibodies were negative, ruling out lupus nephritis (LN), scleroderma, and mixed connective tissue disease (Table I). Abdominal imaging revealed mild gallbladder distension and a calcified adnexal lesion, most likely incidental. Thyroid function tests (TSH, free T4) were within normal limits excluding autoimmune thyroid involvement.

Due to metabolic abnormalities, positive anti-SSA/Ro antibodies, and the elimination of other causes, the diagnosis of hypokalaemic distal RTA secondary to SS was made, despite the absence of characteristic sicca symptoms. The patients was treated with sodium bicarbonate for acidosis, vigorous potassium replacement, and immunosuppression with hydroxychloroquine (HCQ), azathioprine, and corticosteroids. The difficulties in treating autoimmune-mediated RTA were highlighted by the patient's inconsistent progress. This case emphasises the significance of autoimmune assessment even without typical glandular signs, highlighting SS as a rare but significant cause of recurrent hypokalaemic paralysis in young girls.

Table I: Laboratory findings and autoimmune profile.

Tests	Results	Normal ranges
Serum potassium (K^+)	1.07 mmol/L	3.5–5.0 mmol/L
pH	7.40	7.35–7.45
$HCO3^-$	15.9 mmol/L	22–28 mmol/L
PCO2	24 mmHg	35–45 mmHg
Spot urine K/Cr ratio	43 mEq/g	<13 mEq/g (normal)
ESR	65 mm/hour	<20 mm/hour
Rheumatoid factor	25.9 IU/mL	<14 IU/mL
Anti-CCP	Negative	Negative
ANA	1+ speckled	Negative
Anti-SSA/Ro	55.96 U/mL	<20 U/mL
Anti-dsDNA	55.00 IU/mL	<30 IU/mL

DISCUSSION

This case revealed that the patient was diagnosed with hypokalaemia secondary to RTA caused by SS after experiencing recurring bouts of sudden-onset quadriparesis, hypokalaemia, and metabolic acidosis. The diagnosis of SS, a systemic autoimmune illness marked by exocrine gland dysfunction and systemic symptoms, was strongly supported by the patient's clinical presentation, laboratory results, and positive autoantibodies (SS-A/Ro). Moreover, the patient's initial symptoms of hypokalaemia, vomiting, and intermittent weakness improved with potassium treatment. Gastrointestinal potassium loss was ruled out because there was no evidence of diarrhoea, constipation, or other digestive symptoms. A normal anion gap, alkaline urine, and metabolic acidosis suggested distal RTA (Type 1 RTA). RTA is a well-known complication of SS, characterised by impaired acid excretion and reduced potassium reabsorption due to autoimmune-mediated damage to the renal tubules, which is consistent with findings from previous studies.[1]

However, the immune complex deposition or direct targeting of intercalated cells in the collecting ducts are two ways whereby autoantibodies such as anti-SSA/Ro and anti-dsDNA, which were present in this patient, are linked to tubular damage.[5] The diagnostic problem was highlighted by the fact that this patient did not exhibit any symptoms of sicca. Similar cases have been documented when renal symptoms appeared years before any signs of glandular involvement.[6]

Positive SS-A/Ro antibodies and negative SS-B/La, Sm, Scl-70, and U1-RNP antibodies were found during the patient's autoimmune workup. Up to 65% of individuals with SS have SS-A/Ro antibodies, which are very indicative of the diagnosis, especially when other specific autoantibodies are absent.[7] SS-B/La antibodies are frequently linked to SS; however, as they are found in only 60-70% of patients, their absence does not rule out the diagnosis.[8]

The coexistence of anti-SSA/Ro, anti-dsDNA, and ANA raised a differential diagnosis of the SS and systemic lupus erythematosus (SLE). Several factors favour the diagnosis of SS in this case. Notably, the patient lacked clinical features typically required for an SLE diagnosis, such as malar rash, photosensitivity, arthritis, serositis, or haematological abnormalities, as outlined in ACR/EULAR classification criteria.[1] Moreover, RTA is a classical extraglandular manifestation of the SS but is exceedingly rare in SLE, where renal involvement usually manifests as glomerulonephritis. Although anti-dsDNA is considered specific for SLE, it can occasionally appear in SS patients, especially those with extraglandular features, and the absence of other SLE-specific antibodies, such as anti-Sm, supports this.[7] Therefore, there was no evidence of hypocomplementaemia, which is typically observed in active LN. The presence of strongly positive anti-SSA/Ro, along with a clinical phenotype dominated by renal tubular dysfunction without systemic lupus features, aligns more closely with SS. The cases of SS presenting initially with renal involvement in the absence of sicca symptoms have been previously reported, emphasising the need for a high clinical suspicion even in atypical presentations.[5]

An autoimmune aetiology is further supported by the patient's positive ANA (1+ speckled) and increased ESR (65 mm/1st hour). Rheumatoid arthritis was ruled out as the primary cause due to the lack of anti-CCP antibodies and a normal rheumatoid factor (25.9 IU/ml). Anti-dsDNA antibodies can be seen in overlap syndromes and SLE, although they are less common in SS. However, SS was the most likely diagnosis because it lacked additional SLE-specific antibodies (such as Sm) and clinical characteristics.[9]

HCQ is an essential component in the treatment of SS, as it reduces systemic inflammation and regulates the immunological response.[10] Patients with RTA and SS usually have a good prognosis if they receive the right care. However, in order to treat possible side effects such as chronic kidney disease or the development of lymphoma—an uncommon but dangerous SS complication—long-term monitoring is necessary.[11]

The significance of taking SS into account when patients exhibit hypokalaemia and metabolic acidosis, especially when gastrointestinal symptoms are not present, is demonstrated by this

example. The diagnosis was highly supported by RTA and the presence of SS-A/Ro antibodies. These patients can have far better outcomes if they are identified early and treated with potassium supplements, sodium bicarbonate, and immunomodulatory therapy.

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