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Case Report

Proximal renal tubular acidosis in Sjögren's syndrome: diagnostic challenge and successful management

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ABSTRACT

Proximal renal tubular acidosis (RTA) is a rare but important extra-glandular manifestation of Sjögren's syndrome, often presenting diagnostic challenges due to its overlapping features with other renal and autoimmune disorders. This case report describes a 44-year-old woman with classic sicca symptoms and constitutional complaints including significant weight loss, prolonged fever, chronic cough, and oral dryness. She was found to have proteinuria, elevated creatinine, metabolic acidosis, severe hypocalcaemia, and electrocyte imbalances, markers suggestive of renal involvement. Serological work-up confirmed Sjögren's syndrome, while detailed laboratory analysis recognized proximal (type 2) RTA accompanied by Fanconi syndrome, based on the detection of proteinuria, hypocalcaemia, hypokalaemia, and vitamin D deficiency. Differential diagnosis initially favoured plasma cell dyscrasias; however, polyclonal rather than monoclonal gammopathy, with a normal kappa-lambda ratio, established the autoimmune etiology. Hospital management included intravenous calcium gluconate, ongoing calcium and vitamin D supplementation, corticosteroids, azathioprine, sodium bicarbonate infusion, and potassium repletion, resulting in rapid symptomatic and metabolic improvement. The case highlights the complexity of distinguishing isolated proximal RTA from Fanconi syndrome and underscores the importance of early recognition and immunosuppressive management to prevent osteomalacia and progressive renal dysfunction. Regional and ethnic variability in RTA types further complicates diagnosis, confirming the need for high clinical suspicion and multidisciplinary evaluation. This report seeks to inform clinicians about the nuances of Sjögren's-related renal disease and emphasizes individualized therapy tailored to the underlying autoimmune pathophysiology.

Keywords: Sjögren's syndrome, Proximal renal tubular acidosis, Hypocalcaemia, Polyclonal gammopathy, Azathioprine, Corticosteroid therapy, Electrolyte imbalance

INTRODUCTION

Sjögren's syndrome is a chronic autoimmune disorder that primarily targets exocrine glands, leading to classic sicca symptoms, but it can also involve extra-glandular organs such as the kidneys. Renal involvement most commonly presents as distal renal tubular acidosis (type 1 RTA), while pure proximal RTA (type 2) is rare.[1] The mechanism involves autoimmune lymphocytic infiltration and immune-mediated injury to renal tubules; in uncommon cases, this affects the proximal tubule, resulting in impaired bicarbonate reabsorption and development of type 2 RTA.[2]

CASE REPORT

A 44-year-old female patient presented with a history of constitutional symptoms, including significant weight loss (BMI 17 kg/m²), low-grade fever for the past 6 months, chronic cough, and dryness of mouth. She had no known comorbidities and no family history of tuberculosis. She was initially evaluated on an outpatient basis and was found to have elevated serum creatinine, leucocytosis, and

anaemia. Due to abnormal laboratory findings, she was admitted for further evaluation. On admission, her vital signs were stable: blood pressure 110/70 mmHg, pulse rate 78/min, respiratory rate 24/min, and oxygen saturation 97% on room air. She was conscious, coherent, and oriented. Laboratory investigations revealed abnormal complete blood profile indicating anaemia and leucocytosis, along with abnormal urine analysis showing proteinuria.

Table 1: Investigations confirming Sjögren's syndrome with renal involvement.

Parameter/test	Result	Interpretation
Haemoglobin	8.4 g/dl	Anaemia, likely chronic disease or inflammatory anaemia, with anisopoikilocytosis, microcytic hypochromic RBCs, due to chronic inflammation, renal involvement, or nutritional deficiency in chronic illness like Sjögren's
Total leukocyte count	23,000 cells/mm³	Leucocytosis; suggests active inflammation
Absolute neutrophil count	9.91×10⁹/l	Absolute neutrophilia; supports acute inflammatory process
Serum calcium	6.2 mg/dl	Hypocalcaemia, due to vitamin D deficiency or RTA
Serum potassium	2.8 mmol/l	Hypokalaemia consistent with renal potassium wasting typical in type 2 RTA.
Serum chloride	126 mmol/l	Hyperchloremia, a common electrolyte disturbance in renal tubular acidosis
Carbon dioxide – total serum/plasma	8 mmol/l	Low bicarbonate reflecting metabolic acidosis
Serum albumin	2.6 g/dl	Hypoalbuminemia caused by chronic inflammation, possible renal loss
Vitamin D – serum/plasma	<3.4 ng/dl	Severe vitamin D deficiency, contributing to hypocalcaemia and osteomalacia risk often associated with RTA
Albumin to globulin ratio	0.4 g/dl	Reversed ratio typical of autoimmune conditions
Serum anti-SS-A native	125	Positive auto anti-bodies supporting diagnosis of Sjögren's syndrome
Serum anti-Ro-52	127	
Serum anti-SS-B (La)	115	
KAPPA free light chains - serum	203.4	Elevated kappa, lambda light chains consistent with polyclonal gammopathy
LAMBDA free light chains - serum	235.22	
Serum creatinine	1.8mg/dl	Elevated indicating impaired kidney function likely secondary to autoimmune tubulointerstitial nephritis or renal involvement in Sjögren's
Urinalysis (CUE)	Proteinuria – 96 md/dl, no haematuria	proteinuria suggestive of autoimmune renal involvement
Urine pH	5.2	Acidic urine pH <5.5 consistent with type 2 RTA when plasma bicarbonate is depleted, indicating distal nephron acidification is intact despite proximal bicarbonate wasting
Renal ultrasound	Raised parenchymal echotexture (both kidneys)	Suggests chronic tubulointerstitial nephritis, possibly autoimmune (Sjögren's/RTA II)
Chest X-ray	Fibrotic bands of opacities noted in right upper and left mid lung zones, faint interstitial opacities noted in right lower lung zone	Chronic lung involvement possibly related to autoimmune disease or past infection

Further evaluation included an ANA profile, which was strongly positive for anti-Ro (SSA) and anti-La (SSB) antibodies. Collectively, the findings from Table 1 confirmed the diagnosis of Sjögren's syndrome, type 2 renal tubular acidosis (RTA) and hypocalcaemia.

During hospitalization, the patient developed tetanic spasms and was found to have low serum calcium. She received intravenous calcium gluconate followed by ongoing calcium and vitamin D supplementation. She was initiated on azathioprine and prednisolone (Omnacortil) for immunosuppression. Supportive management included

intravenous fluids for hydration, pantoprazole, cephalosporin antibiotics, oral ondansetron, spironolactone, sodium bicarbonate, and intravenous potassium chloride for metabolic acidosis.

The patient showed clinical and symptomatic improvement with treatment. At the time of discharge, she was hemodynamically stable and was advised to continue prednisolone, azathioprine, calcium and vitamin D supplementation, spironolactone, and potassium–magnesium citrate syrup on an outpatient basis with regular follow-up.

DISCUSSION

Proximal RTA is often part of a broader proximal tubular dysfunction known as Fanconi syndrome, characterized by bicarbonate, phosphate, glucose, amino acid, and uric acid wasting. Acquired causes of Fanconi syndrome include autoimmune diseases such as Sjögren's, malignancies, and drugs like cisplatin or tenofovir. Therefore, the coexistence of Sjögren's syndrome with type 2 RTA and Fanconi syndrome, as seen in our patient, reflects a rare but clinically significant manifestation of proximal tubular injury in autoimmune disease.[3]

Literature supports the rarity of proximal RTA in Sjögren's, only about 2% in large systematic reviews, although some cohorts have reported higher proportions, possibly due to regional differences. Several Indian and Asian cohorts report that 11–25% of patients with primary Sjögren's develop RTA of any type, while a Chinese study reported rates as high as 73%, likely reflecting inclusion of subclinical cases.[4,5] In a large systematic review of 440 SS-RTA cases, only 8 patients (≈2%) had pure type 2 RTA, whereas 388 had distal RTA and 38 had mixed features.[6] However, some regional data, such as Ram et al reported a higher proportion of proximal involvement, up to 42.3%, suggesting possible ethnic or geographic variability.[1]

In this patient, the first differential diagnosis considered was multiple myeloma, as it is a well-known cause of proximal tubular dysfunction, Fanconi syndrome, and type 2 RTA. However, serum free light chain analysis showed elevated kappa and lambda levels with a normal kappa/lambda ratio, indicating polyclonal gammopathy rather than a monoclonal pattern seen in plasma cell dyscrasias like multiple myeloma or monoclonal gammopathy of undetermined significance (MGUS). Polyclonal gammopathy is typically associated with chronic inflammation, autoimmune diseases (such as Sjögren's syndrome), infections, or chronic liver disorders. In this case, liver function tests were normal, further excluding liver-related causes. Therefore, the normal kappa/lambda ratio was the key factor that differentiated Sjögren's-related immune activation from multiple myeloma, allowing us to rule out malignancy and liver dysfunction.[7-9]

It is important to distinguish isolated proximal RTA from Fanconi syndrome. All patients with Fanconi syndrome have proximal (type 2) RTA, but not all patients with type 2 RTA have Fanconi syndrome. Isolated proximal RTA involves selective bicarbonate wasting, whereas Fanconi syndrome is a generalized proximal tubular defect, leading to loss of multiple solutes such as glucose (glycosuria), amino acids (aminoaciduria), phosphate (hypophosphatemia), uric acid, and proteinuria due to low-molecular-weight protein loss. In this patient, the presence of proteinuria along with disturbances in calcium and vitamin D metabolism strongly supports Fanconi syndrome rather than isolated pRTA.[10,11]

Recognizing Fanconi syndrome in the context of Sjögren's is vital, as it carries a higher risk of complications such as osteomalacia, bone pain, fractures, electrolyte imbalances, and progressive renal dysfunction. Early identification allows targeted management, electrolyte and bicarbonate supplementation, vitamin D and phosphate replacement, immunosuppressive therapy to control the underlying autoimmune activity, and monitoring for bone health. This differentiation between isolated pRTA and Fanconi syndrome is crucial for prognosis, therapeutic strategy, and accurate interpretation in the context of Sjögren's-related renal disease.[2,12,13]

During hospitalization, the development of tetanic spasms experienced by the patient were correlated with hypocalcaemia, a recognized complication of Fanconi syndrome due to phosphate wasting and impaired vitamin D metabolism leading to osteomalacia. Immediate administration of intravenous calcium gluconate effectively addressed acute calcium deficiency and neuromuscular irritability.

Corticosteroids remain first-line agents in controlling inflammation and preventing further renal damage; azathioprine acts as a steroid-sparing agent to maintain remission while minimizing steroid-related adverse effects. This immunomodulation addresses the pathogenesis rather than just the symptoms, aligning with best practices for treating autoimmune renal involvement. Intravenous fluids ensured adequate hydration and intravascular volume maintenance, important in patients with electrolyte losses. Spironolactone served as a potassium-sparing diuretic to support potassium balance, counteracting hypokalaemia common in proximal RTA. Sodium bicarbonate was administered to correct metabolic acidosis by replenishing plasma bicarbonate levels, a cornerstone in managing RTA to restore acid-base homeostasis. The intravenous potassium chloride supplemented potassium loss critical to preventing cardiac arrhythmias and muscle weakness.[2]

CONCLUSION

This case demonstrates that proximal RTA and Fanconi syndrome, though rare in Sjögren's syndrome, require prompt diagnosis and targeted management to prevent

severe complications like osteomalacia and chronic renal impairment. Multidisciplinary care and immune-suppressive therapy are pivotal for optimal patient outcomes in complex autoimmune renal presentations.

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REFERENCES

1. Ram R, Swarnalatha G, Dakshinamurty KV. Renal tubular acidosis in Sjögren's syndrome: A case series. Am J Nephrol. 2014;40(2):123-30.
2. Jung SW, Park EJ, Kim JS, Lee TW, Ihm CG, Lee SH, et al. Renal tubular acidosis in patients with primary Sjögren's syndrome. Electrolyte Blood Press. 2017;15(1):17-22.
3. Reddy S, Kamath N. Clinical approach to renal tubular acidosis in children. Karnataka Pediatr J. 2021;35(88):88–94.
4. Sandhya P, Jeyaseelan L, Scofield RH, Danda D. Clinical characteristics and outcome of primary sjogren's syndrome: A large Asian Indian cohort. Open Rheumatol J. 2015;9(1):36-45.
5. Coradin R, Lopes MLL, Goldani JC, Ventura PE, Keitel E. Hypocitraturia as a biomarker of renal tubular acidosis in patients with Sjögren's disease. Adv Rheumatol. 2024;64(1):45.
6. Prabhu N, Sankarlingam R, Chilukuri B, Sudan A, Tiwari VP, Muniraju T. A clinical and serological profile of renal tubular dysfunction in autoimmune disorders in A tertiary care centre in south India. J Contemp Clin Pr. 2025;11:1-7.
7. Kumar S, Larson DR, Dispenzieri A, Therneau TM, Murray DL, Leif Bergsagel P, et al. Polyclonal serum free light chain elevation is associated with increased risk of monoclonal gammopathies. Blood Cancer J. 2019;9(6):49.
8. Singh G. Serum free light chain assay and κ/λ ratio performance in patients without monoclonal gammopathies: High false-positive rate: High false-positive rate. Am J Clin Pathol. 2016;146(2):207-14.
9. O'connell T, Horita T, Kasravi B. Understanding and interpreting serum protein electrophoresis. Am Fam Physician. 2005;71(1):105-12.
10. Kashoor I, Batlle D. Proximal renal tubular acidosis with and without Fanconi syndrome. Kidney Res Clin Pract. 2019;38(3):267-81.
11. Matarneh AS, Matarneh B, Rout P, Bokhari SRA. Fanconi syndrome. In: StatPearls. Treasure Island (FL): StatPearls Publishing. 2025.
12. Shi M, Chen L. Sjögren's syndrome complicated with Fanconi syndrome and Hashimoto's thyroiditis: Case report and literature review. J Int Med Res. 2016;44(3):753-9.
13. Gutierrez JO, Zurita MF, Zurita LA. Sjögren's syndrome associated with Fanconi's syndrome and osteomalacia. Am J Case Rep. 2018;19:392-6.

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