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Case Report

Quadriparesis as a Presentation of Sjögren Syndrome-Induced Distal Renal Tubular Acidosis with Hypokalemia and Osteomalacia

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Abstract

Sjögren syndrome is a systemic autoimmune disease primarily affecting the salivary and lacrimal glands, commonly presenting with dry eyes and dry mouth. While exocrine gland involvement is typical, extraglandular manifestations such as renal tubular acidosis (RTA) can occur, leading to metabolic disturbances. Distal RTA is the predominant renal complication in Sjögren syndrome and may cause hypokalemia severe enough to result in neuromuscular symptoms like quadriparesis. Additionally, chronic RTA contributes to osteomalacia, characterized by impaired bone mineralization and the development of pseudofractures (Looser zones). We report the case of a 31-year-old female with a 10-year history of recurrent hypokalemia and episodes of quadriparesis who presented with thigh pain and difficulty walking. Clinical evaluation revealed sicca symptoms, positive anti-Ro/SSA antibodies and laboratory evidence of distal RTA with hypokalemia and metabolic acidosis. Radiological studies showed bilateral subtrochanteric impending fractures consistent with osteomalacia. She underwent prophylactic orthopedic fixation and was treated with potassium and alkali supplementation, along with immunosuppressive therapy for Sjögren syndrome. This case highlights the diagnostic challenges of atypical presentations of Sjögren syndrome and underscores the importance of considering systemic complications such as distal RTA and osteomalacia. Early recognition and comprehensive management are crucial to prevent severe morbidity.

Keywords: Sjögren syndrome, distal renal tubular acidosis, hypokalemia, quadriparesis, osteomalacia, Looser zones

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Introduction

Sjögren syndrome is a long-term autoimmune condition primarily affecting the salivary and lacrimal glands, leading to dry mouth and dry eyes (sicca symptoms) [1]. The disease was first described in the early 20th century by Swedish physician Henrik Sjögren and is now recognised as a systemic disorder with potential involvement of multiple organ systems [2]. Although its hallmark features involve exocrine gland dysfunction, Sjögren syndrome can extend beyond these glands. It is often associated with other autoimmune conditions, such as rheumatoid arthritis and systemic lupus erythematosus [2,3]. Around half of affected individuals may develop extraglandular complications, which can impact the skin, lungs, kidneys, joints or nervous system [1,2]. Because of its systemic nature, virtually any organ can be affected.

Among the less commonly discussed complications is renal involvement, particularly renal tubular acidosis (RTA). About 9% of patients with Sjögren syndrome develop RTA, and of those, approximately 97% have the distal (type 1) form [4,5]. This variant is characterised by a failure of the distal tubules to adequately secrete hydrogen ions, impairing the kidney's ability to acidify urine and leading to metabolic acidosis. Severe hypokalemia can lead to quadriparesis by disrupting normal neuromuscular function, as low serum potassium levels impair membrane excitability and muscle contraction, resulting in generalised muscle weakness that can progress to paralysis [4,5]. Secondary or acquired forms of distal RTA are frequently associated with conditions characterised by elevated globulin levels, including autoimmune diseases. A significant consequence of long-standing distal RTA is osteomalacia, a bone disorder characterised by inadequate mineralisation [6,7]. Although this bone disease can occur in distal RTA even without Sjögren syndrome, it is more commonly associated with immune-mediated causes, including Sjögren syndrome. One of the classic radiographic findings in osteomalacia is the presence of pseudofractures, also known as Looser zones [6,7]. These are partial cortical breaks that arise due to bone weakening and are typically seen on imaging in patients with chronic acid-base disturbances.

Herein, we report a rare and clinically significant case of quadriparesis as the initial manifestation of Sjögren syndrome-induced distal RTA, presenting with severe hypokalemia and associated osteomalacia. This case highlights the importance of recognising atypical neuromuscular and skeletal presentations in systemic autoimmune disorders, which may precede classic sicca symptoms and lead to delayed diagnosis if not considered.

Case Presentation

A 31-year-old female presented with complaints of thigh pain and limping, with a history of progressive difficulty in walking over the past three years. She had been diagnosed with hypothyroidism in 2016 and was on a regular dose of oral thyroxine 100 mcg daily. Notably, she had a 10-year history of recurrent hypokalemia characterised by episodes of quadriparesis, which resolved promptly after potassium replacement at a local healthcare facility. She also reported loss of multiple teeth and had undergone dental implantation in 2022. On further evaluation, she revealed a history of dry mouth and dry eyes, suggestive of sicca symptoms. However, there was no history of skin rashes, photosensitivity, oral or genital ulcers, alopecia, joint pains, fever or abdominal or urinary symptoms.

Laboratory investigations revealed mild anaemia with a haemoglobin level of 9.4 (12.5-15.0 g/dl), normal total leukocyte count of 6.6 × 10^3 (4-11 × 10^3/µL) and thrombocytopenia with a platelet count of 74,000 (150,000-400,000/µL). Renal function tests showed elevated blood urea 48 (5-20 mg/dL) and serum creatinine 1.7 (0.6-1.1 mg/dL). Urinalysis revealed a urinary pH of 6.1. Electrolyte analysis demonstrated normal sodium 142 (135-145 mmol/L), low potassium 3.0 (3.5-5.0 mmol/L), elevated chloride 121 (96-106 mmol/L), low calcium 8.0 (8.5-10.5 mg/dl), low phosphate 0.4 (0.9-1.4 mmol/L), elevated alkaline phosphatase 368 (44-148 IU/L) and markedly reduced bicarbonate 4.6 (22-28 mmol/L). Serum albumin, 25-hydroxyvitamin D and intact parathyroid hormone were within normal limits at 3.7 (3.5-5.5 g/dl), 65 (50-100 ng/ml) and 32 (10–65 ng/L), respectively. Inflammatory markers were elevated, with a C-reactive protein level of 47.3 (0-5 mg/L) and an ESR of 56 (10-30 mm/hour). Immunological testing revealed a positive antinuclear antibody with a speckled pattern at a high titer of 1:128,0, and the extractable nuclear antigen panel showed strongly positive anti-Ro/SSA antibodies (>70 IU/mL), confirming the diagnosis of Sjögren syndrome. Radiological examination of the femurs demonstrated bilateral subtrochanteric unicortical impending fractures, consistent with Looser zones and indicative of osteomalacia (Figure 1). A dual energy X-ray absorptiometry (DEXA) scan further revealed a T-score of -2.0 and a Z-score of -2.1, indicating low bone mineral density.

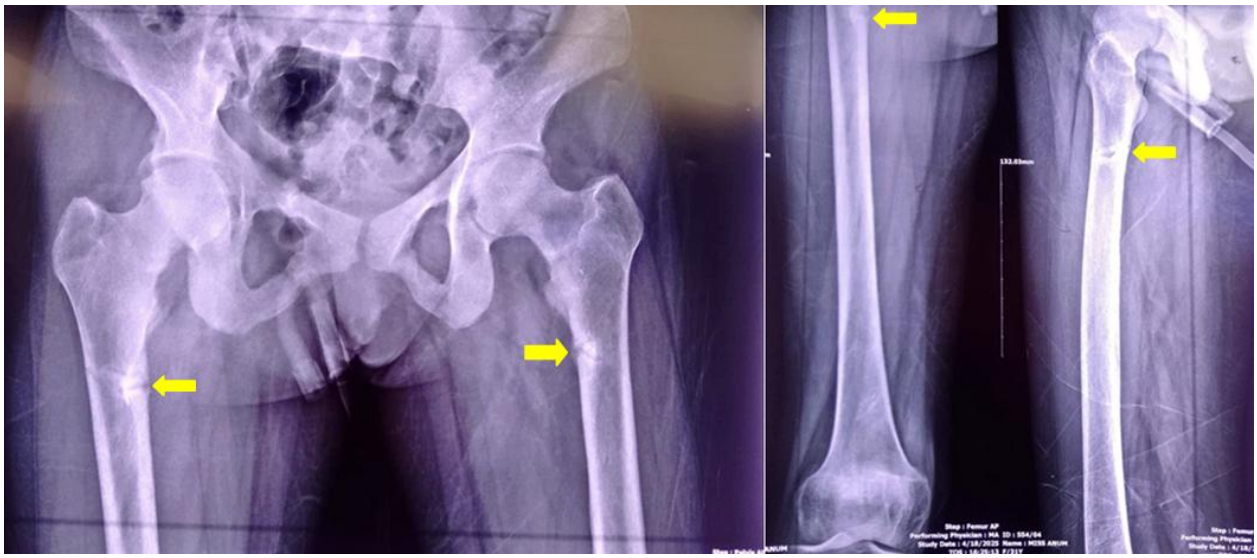

Figure 1: X-ray demonstrating bilateral subtrochanteric unicortical impending fractures in the femurs consistent with Looser zones of osteomalacia

Based on her clinical presentation, investigations and radiographic findings, she was diagnosed with Sjögren syndrome complicated by distal renal tubular acidosis, hypokalemia and osteomalacia. For the bilateral subtrochanteric impending fractures, the patient underwent prophylactic fixation with bilateral proximal femur nail anti-rotation under the care of the orthopaedic surgery team. To manage Sjögren syndrome, she was started on azathioprine along with symptomatic treatment using artificial tears and saliva substitutes. Her distal renal tubular acidosis was treated with oral potassium supplementation and sodium bicarbonate to correct metabolic acidosis and maintain electrolyte balance. After stabilisation, the patient was discharged with instructions for regular follow-up in the out-patient department (OPD) to monitor her clinical status, response to treatment and any adverse effects.

Discussion

The present case describes a young woman with Sjögren syndrome presenting with recurrent quadriparesis, severe hypokalemia and osteomalacia due to presumptive distal RTA, highlighting a rare combination of neuromuscular and skeletal complications preceding classic sicca symptoms. Although our patient had mildly elevated serum urea and creatinine, the presence of severe hyperchloremic metabolic acidosis, persistent hypokalemia and hypophosphatemia favoured distal RTA rather than advanced chronic kidney disease as the primary aetiology of her metabolic bone disease. The presence of normal anion gap metabolic acidosis with hyperchloremia and inappropriate urinary acidification in the setting of a systemic autoimmune disease further supported the diagnosis of distal RTA. Furthermore, chronic kidney disease-related mineral bone disorder typically presents with hyperphosphatemia in advanced stages, which was not observed in this case. Additionally, serum 25-hydroxy vitamin D and iPTH levels were within normal limits, effectively excluding nutritional vitamin D deficiency and secondary hyperparathyroidism as primary contributors to the bone pathology.

In contrast, serum alkaline phosphatase was elevated, consistent with increased osteoblastic activity and impaired mineralisation seen in osteomalacia secondary to chronic metabolic acidosis. Urinary pH was inappropriately elevated despite systemic metabolic acidosis, supporting impaired distal hydrogen ion secretion consistent with distal (type 1) RTA. Urinary acidification studies were not performed during the acute presentation because of the need to correct severe electrolyte abnormalities, limiting the ability to subtype the renal tubular acidosis.

The overall clinical and laboratory profile in the setting of Sjögren syndrome made distal RTA the most likely diagnosis in our case. The patient had a prior diagnosis of hypothyroidism, raising the possibility of concomitant autoimmune thyroid disease, such as Hashimoto's thyroiditis. Autoimmune thyroid disorders are frequently associated with Sjögren syndrome, and their coexistence may represent a broader autoimmune overlap or, in some cases, polyglandular autoimmune syndrome type III [8,9]. However, thyroid antibody testing was not available to definitively establish the diagnosis of autoimmune thyroiditis in this case. While the coexistence of multiple autoimmune conditions may reflect shared immunopathogenic mechanisms, the RTA and osteomalacia observed in this patient are well-recognised extraglandular manifestations of Sjögren

syndrome. They can occur independently of autoimmune thyroid disease [6,7].

Sjögren syndrome affects between 0.5% and 1.0% of the population and occurs at roughly half the rate of rheumatoid arthritis [1,2]. Estimates indicate that approximately 400,000 to 3.1 million adults worldwide are affected by this condition. Although it can develop at any age, symptoms most frequently appear between 45 and 55 years of age. Around 50% of individuals diagnosed with Sjögren syndrome also have rheumatoid arthritis or other connective tissue diseases such as systemic lupus erythematosus [2,3]. This autoimmune disorder is seen globally, predominantly in adults, and less commonly in children, without any clear racial or geographic differences in incidence. There is a marked female predominance, with women affected about nine times more often than men, a pattern similar to SLE [10,11]. The absence of a standardised, evidence-based screening approach to identify patients with dry eye symptoms who should be evaluated for Sjögren syndrome contributes to many patients being underdiagnosed or referred late [12,13]. This results in a substantial number of cases going unrecognised or diagnosed only after a significant delay.

Extra-glandular involvement occurs in nearly 42% of patients, but the frequent presence of sicca symptoms often complicates diagnosis when patients present primarily with non-sicca symptoms. Our case illustrates this complexity, with the patient presenting with an unusual case of impending femoral fracture caused by RTA secondary to Sjögren syndrome. Chronic RTA can cause bone loss by buffering acidemia through the release of minerals from bone, leading to reduced bone density and decreased levels of phosphate and bicarbonate [6,7]. While RTA is an uncommon complication of Sjögren syndrome, the occurrence of pseudofractures or complete fractures in this setting is even rarer. The most recent case reported in 2024 described a man in his 30s who experienced hip pain and was found to have bilateral pseudofractures [14]. Detailed assessment found underlying RTA and Sjögren syndrome. Similarly, in 2021, a 35-year-old woman suffered a femoral fracture following a minor injury, which was linked to RTA caused by possible incomplete Sjögren syndrome [15].

Conclusion

This case illustrates a rare but recurrent complication associated with Sjögren syndrome complicated by distal RTA with profound hypokalemia and osteomalacia leading to recurrent quadriparesis and impending fractures. The atypical presentation, with neuromuscular weakness and skeletal involvement preceding classic sicca symptoms, contributed to challenges in reaching an accurate diagnosis, underscoring the importance of considering systemic complications in patients with autoimmune disease. It highlights the importance of continued research to enhance the identification, treatment, and overall management of this condition. Early recognition, targeted electrolyte and bone management, and immunosuppressive therapy are essential to prevent long-term morbidity and improve outcomes.

Informed consent

Informed consent: Detailed informed consent was obtained from the patient and her next of kin (husband) prior to data collection and manuscript writing, with assurance to maintain anonymity, confidentiality and privacy.

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