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Sjogren's syndrome Associated with Type 1 Renal Tubular Acidosis and Severe Hypokalemia: A Case Study

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Abstract

Sjogren's syndrome (SS) is a chronic autoimmune inflammatory condition that primarily involves exocrine glands like the glands of saliva and tears. SS also involves extra-glandular involvement with renal involvement in the form of tubulointerstitial nephritis, followed by glomerulonephritis, distal renal tubular acidosis (RTA), and Fanconi syndrome. We report a case of type 1 RTA with severe hypokalemia caused by SS. ECG showed Sinus Bradycardia with QT prolongation with U waves. USG's abdomen showed nephrolithiasis of size 4 mm in the left kidney. Diagnosis of distal RTA type 1 associated with SS was based on the above findings. This case emphasizes the need to evaluate for autoimmune conditions when a patient presents with Distal RTA.

Keywords: Distal Renal Tubular Acidosis; Hypokalemia; Sjogren's Syndrome

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Introduction

Dry eyes and xerostomia are two symptoms of the chronic, slowly progressing autoimmune condition known as Sjogren's syndrome (SS) [1]. Additionally, it affects lacrimal, salivary, and exocrine glands as well as extracorporeal organs such as the liver and kidney [2]. The clinical signs of SS are varied, making it simple to misdiagnose or even go unnoticed [2]. Gradually, it may cause serious acid-base imbalances, including hyperchloremic metabolic acidosis and fatally low levels of potassium [3]. Tubulointerstitial Nephritis (TIN) is the most frequent histological abnormality, with Glomerulonephritis as second, according to biopsy data available in the literature [4]. Although TIN is the most frequent histology result, hypokalaemia with biochemical evidence of distal renal tubular acidosis (RTA) has very occasionally been described [4].

Case Report

A 23-year-old female came with chief complaints of vomiting and weakness of bilateral upper and lower limb weakness from day 1. The patient had a history of vomiting for the past 2 days. History of fever since day 2. History of weakness in limbs and sudden onset progressed and reached grade 2 power at the time of presentation. No history of dry mouth (xerostomia) and dry eyes (xerophthalmia). The patient had a similar history of complaints in the past. The patient has a history of Guillain-Barre Syndrome. General Examination was unremarkable except for other neurological systems showing the motor weakness of grade 2 power with hyporeflexia with no sensory involvement as well as no bowel and bladder involvement.

Renal function test for serum electrolytes showed: Sodium-142, Potassium-2.7, and Chloride-106. Serum Osmolality was 295 with pH 7.21 and bicarbonate values at 11.4. Complete urine analysis showed urine pH at 7.0. The occurrence of calcium oxalate crystals was observed during urine analysis. A complete antinuclear antibody (ANA) test profile showed: SS-A and SS-B were strongly positive. Electrocardiography (ECG) showed Sinus Bradycardia with QT prolongation with U waves (Figure 1). Ultrasonography (USG) of the abdomen showed nephrolithiasis of size 4 mm in the left kidney. Diagnosis of distal RTA associated with SS was based on the above findings. The patient was promptly treated with intravenous normal saline and intravenous potassium chloride was infused, after which the patient showed signs of recovery.

Discussion

SS is an autoimmune disease of exocrine glands with extra glandular involvement involving the renal, cardiac, and pulmonary systems [5]. SS is characterized by lymphocytic infiltration of the exocrine glands [6]. Sjogren's syndrome generally affects middle-aged women [7]. Renal involvement of SS primarily includes TIN. Clinicians find it difficult to

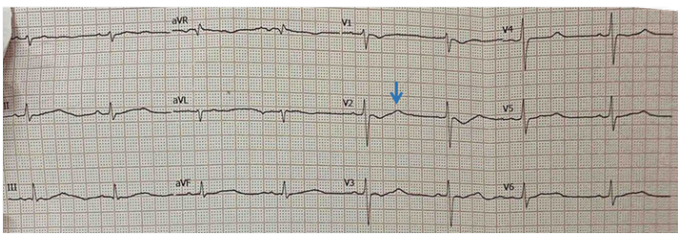

Figure 1: Evidence of U waves with QT prolongation in ECG.

Table 1: Criteria to diagnose SS based on AECG [4,7].

Parameter	Criteria	Remark(s)
Ocular symptoms	• Dry eyes > 3 months • Eye itching • Usage of artificial tears > 3 times per day	Need at least one sign
Oral symptoms	• Dry mouth > 3 months • Swollen salivary glands • Cannot swallow dry foods	Need at least one sign
Ocular signs	• ≤ 5 mm per 5 min Positive vital dye staining (van Bijsterveld ≥ 4) • Schirmer's test	Need at least one sign
Histopathology	Lip biopsy to determine focal lymphocytic sialoadenitis	focus score ≥ 1 per 4 mm^2
Oral signs	• The unstimulated flow of saliva (≤ 1.5 mL under 15 min) • Abnormal salivary scintigraphy • Abnormal parotid sialography	Need at least one sign
Auto-antibody test	• Anti-SSA (Ro) • Anti-SSB (La)	Need at least one sign

diagnose SS, especially when the initial presentation is different from the exocrine expression of dry mouth and eyes [4]. American-European Consensus Classification (AECC) criteria were used to diagnose SS (Table 1) [4,7]. But AECG or American College of Rheumatology (ACR) guidelines have limited scope to diagnose juvenile cases. Sicca Symptoms are one of the presenting conditions generally included in diagnostic criteria [4].

Distal RTA presents with hypercalciuria, hypocitraturia, and alkaline urine [7]. Based on either biochemical or biopsy results available in the literature, the incidence of renal involvement in SS ranges from 0.3% to 27% [4]. Rarely, the only significant symptom of SS associated with hypokalemic paralysis is what prompts a patient to see a doctor [6]. Hypokalemia is one of the most common electrolyte abnormalities presenting clinically. Hypokalemia causes can be due to most common causes like renal losses, intracellular shifts, or insufficient potassium intake [2]. Renal potassium loss includes causes like primary aldosteronism, Bartter syndrome, RTA, Liddle syndrome, and adrenal hyperplasia [2]. Even though SS-related distal RTA has not been considered in the differential diagnosis of hypokalemic paralysis, it is important to be aware of the probable possibility.

RTA is a group of clinical syndromes with tubular defects which may include hydrogen secretion or carbonate reabsorption defects. RTA is of four distinct types, Type 1 RTA (distal RTA), Type 2 RTA (Proximal RTA), Type 3 RTA (Mixed RTA), and Type 4 RTA (Hyperkalemic RTA) [2]. Type 1 RTA is the most common. Autoimmune diseases are the most common causes of distal RTA in adults, for example, Systemic sclerosis, rheumatoid arthritis, systemic lupus erythematosus, and SS [4]. Additionally, familial reasons (Wilson's disease and medullary sponge kidney), hypercalciuric conditions (sarcoidosis, vitamin D intoxication, and hyperparathyroidism), and medications (ibuprofen, lithium carbonate, amphotericin B, and ifosfamide) can also cause distal RTA [4].

It has been noted that corticosteroids combined with other immunosuppressants can decrease the progression of renal impairment in SS. However, the effectiveness of the therapy has generally only been shown in patients with fast aggravating renal disease or insufficiency [6]. However, the patient's symptoms significantly improved with potassium replacement, and she is currently still taking oral potassium and bicarbonate supplements.

In conclusion, a prompt and early diagnosis will help prevent serious complications. Treating the disease early can avoid permanent end-organ damage and t is of utmost importance to consider SS in patients with hypokalemia with metabolic acidosis.

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