An Unusual Initial Presentation of Sjögren’s Syndrome: Severe Hypokalemic Paralysis Secondary to Distal Renal Tubular Acidosis

Ciddi Hipokalemik Paralizi ve Distal Renal Tubuler Asidoz: Sjögren Sendromunun Olağanlısdı Başlangıç Prezentasyonu

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Abstract
Sjögren’s syndrome is mainly affects the exocrine glands. Patients usually complain of persistent dryness of the mouth and eyes. However, nonexocrine organs such as the kidneys are often affected in these patients. Distal renal tubular acidosis (dRTA) and interstitial nephritis are common in Sjögren’s syndrome. Nonetheless, severe hypokalemia and paralysis secondary to dRTA are unusual initial manifestation of Sjögren’s syndrome. Here, we describe a case of a 48 year old women admitted to the emergency setting with severe hypokalemic paralysis and diagnosed Sjögren’s syndrome.

Key Words: Acidosis, hypokalemic paralysis, sjögren’s syndrome

Özet

Anahtar Kelimeler: Asidoz, hipokalemin paralizi, sjögren sendromu

Introduction
Sjögren’s syndrome (sicca syndrome) is a systemic chronic inflammatory disorder characterized by lymphocytic and plasmacytic infiltrates in exocrine organs, such as salivary, parotid, and lacrimal glands. Nonexocrine organs, including the kidneys, are often affected in Sjögren’s syndrome [1]. Distal renal tubular acidosis (dRTA) and interstitial nephritis are common in patients with Sjögren’s syndrome [2]. Nonetheless, the patients with Sjögren’s syndrome rarely present with severe hypokalemia or paralysis secondary to dRTA.

Case Report
A 48 year old Turkish woman presented with weakness of all four limbs with minimal respiratory distress. Her past history revealed two similar attacks 6 months and 1 year ago. She suffered from respiratory arrest during one of these attacks and was transferred to an intensive care unit for full ventilation after intubation in another hospital. She was given an oral potassium supplement for a few weeks; however, she did not have regular follow-up. Her medical history also revealed a myocardial infarction 6 years ago. She smoked a pack of cigarettes a day.

On admission, physical examination showed a clear consciousness, blood pressure of 120/60 mmHg, heart rate of 70 beats/min, and respiratory rate of 14-24 breaths per minute. The rest of the examination was unremarkable except for a neurologic examination that revealed grade 3/5 strength of the proximal muscles. Electrocardiography showed pathologic Q wave and R wave progression loss on the anterior derivations, most likely due to the previous myocardial infarction. Chest radiography was normal. Initial laboratory analysis revealed the following: urea: 39 mg/dL, creatinine: 1.3 mg/dL, sodium: 144 mEq/L, potassium: 1.9 mEq/L, chloride: 118
mEq/L, calcium: 8.3 mg/dL, and phosphate: 2.1 mg/dL. The serum anion gap was normal (13.2 mEq/L). Arterial blood gas showed a serum pH of 7.20, bicarbonate of 12.2 mmol/L, and PCO₂ of 29.5 mmHg. The patient was diagnosed with hypokalemic hyperchloremic metabolic acidosis; thus, renal tubular acidosis was suspected. Urine analysis showed a pH of 8 and a positive anion gap (+41 mEq/L). Renal ultrasound showed multiple hyperechoic lesions with a post-acoustic shadow compatible with bilateral nephrocalcinosis. These findings confirmed the diagnosis of renal tubular acidosis type 1 (distal renal tubular acidosis-dRTA). We started fluid therapy with saline infusion, including potassium chloride and bicarbonate, once the serum potassium increased to 3 mEq/L.

Further work-up regarding the underlying cause of dRTA was performed. Consultation to the ophthalmologist was made because the patient suffered from a burning sensation in both eyes. Schirmer's test demonstrated a 4 mm/5 min binocularly. Additional laboratory tests showed that she was positive for anti-nuclear (titer >1:1000, speckled and nucleolar pattern) and anti-Ro/SSa antibodies. We also performed salivary gland scintigraphy, which showed a very low uptake of the radionuclide (Figure 1), and this result further supported our diagnosis of Sjögren's syndrome.

This study considered a case of primary Sjögren's syndrome because the patient's clinical and laboratory findings did not completely fulfill the diagnostic criteria of the diseases leading to secondary Sjögren's syndrome, such as systemic lupus erythematosus, rheumatoid arthritis, and primary biliary cirrhosis. The patient was treated with oral prednisolone and hydroxychloroquine. We prescribed K-Shohl solution to the patient for potassium and alkali replacement therapy. Initial and post-treatment laboratory findings are shown in Table 1.

**Discussion**

Most of the patients with Sjögren's syndrome present with sicca symptoms, such as keratoconjunctivitis, xerostomia, and sialoadenitis. Numerous extraglandular features, including renal involvement, may develop during the course of the disease [3].

dRTA, characterized primarily by impaired net acid excretion, is the classical renal lesion in Sjögren's syndrome. Four pathophysiologic mechanisms in dRTA are suggested for the impaired acidification by the distal nephron: backleak, pump failure, voltage defect, and rate defect/voltage defect. Sjögren's syndrome is a secondary cause of hypokalemic dRTA, along with multiple myeloma, medullary sponge kidney or systemic lupus erythematosus [4]. Defective expression of the H⁺-ATPase pump of intercalated cells in the cortical collecting tubules due to the immunologic reaction in the kidney is suggested as a pathogenetic mechanism in Sjögren's syndrome [5].

Our patient did not reveal evidence of the other secondary etiologies of dRTA. A positive Schirmer's test, high titers of antibodies to Ro and positive scintigraphy results were strongly suggestive of Sjögren's syndrome.

Severe hypokalemia and paralysis due to dRTA is an unusual initial presentation of Sjögren's syndrome [6, 7]. Palkar et al. [8] also reported a case of Sjögren's syndrome that presented with acute quadriparesis, bulbar weakness, atrial fibrillation, and ventricular ectopics due to hypokalemic dRTA. Severe hypokalemia may rarely cause sudden life-threatening paralysis [6-10]. Furthermore, if adequate treatment is not received, muscle paralysis may lead to respiratory arrest [9]. Sjögren's syndrome was reported as the underlying etiology of the hypokalemic dRTA of a patient who suddenly developed respiratory arrest and was transferred to the emergency department [7, 8, 10]. Although respiratory arrest associated with Sjögren's syndrome is very rare, this complication is very severe and can be fatal. It is important to notice that muscle paralysis and respiratory arrest associated
with severe hypokalemia may be due to dRTA. Furthermore, an undiagnosed Sjögren's syndrome should be taken into consideration as a possible underlying etiology of muscle paralysis and respiratory arrest.

Conflict of interest statement: The authors declare that they have no conflict of interest to the publication of this article.

References


