CASE REPORT
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Respiratory arrest following severe muscle weakness as initial presentation of Sjögren’s syndrome: diagnostic and therapeutic strategies: a case report

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ABSTRACT

Background: Sjögren’s syndrome refers to an autoimmune disorder defined by chronic inflammation as a consequence of the exocrine gland’s lymphocytic infiltration. Patients may present with heterogeneous clinical symptoms involving impairment in the lungs, kidneys, and nervous system. The most common renal involvement is tubulointerstitial nephritis (TIN) responsible for 20% of renal tubular acidosis. We report a case of respiratory failure resulting from severe hypokalemia in a patient with Sjögren syndrome.

Case presentation: A 46-year-old female who experienced progressive quadriplegia leading to respiratory distress caused by severe hypokalemia. Laboratory findings showed: sodium 128 mmol/L, potassium 1.1 mmol/L, Chloride 93 mmol/L, Calcium 7.3 mg/dL (corrected 7.5 mg/dL), albumin 3.3 g/dL, Magnesium 1.8 mg/dL with amylase 244 U/L, lipase 4168 U/L. The ANA test increased slightly and Ro-52, PM-Scl100 showed strong positive results, finally the patient was diagnosed with Sjögren's syndrome with renal tubular acidosis (RTA). She was later diagnosed with renal tubular acidosis secondary to Sjögren’s syndrome and treated using methylprednisolone, potassium, and alkali administration. Later, her potassium levels reached normal, muscle weakness improved, and she was extubated.

Conclusion: This case emphasizes the concept of an uncommon form of Sjögren’s syndrome in the form of severe refractory hypokalemia due to renal tubular acidosis accompanied by acute pancreatitis disguised in patients without SICCA complaints. Renal tubular acidosis is a rare case, but Sjögren’s syndrome is a disease that must be considered in all patients who present with paralysis due to refractory hypokalemia so that there is no misinterpretation and management.

Keywords: Autoimmune disease, Renal tubular acidosis, Hypokalemia, Quadriplegia, Respiratory arrest, Sjögren’s syndrome.


INTRODUCTION

Sjögren’s syndrome refers to an autoimmune disorder defined by chronic inflammation as a consequence of the infiltration of lymphocytes into the exocrine glands. This condition can be categorized into primary or secondary.¹² Primary Sjögren’s syndrome occurs without other autoimmune diseases presenting with dry eyes (keratoconjunctivitis sicca) and dry mouth (xerostomia).¹³ On the other hand, Sjögren’s syndrome may develop in conjunction with other autoimmune disorders as a secondary type, examples include systemic lupus erythematosus (SLE), rheumatoid arthritis (RA), and systemic sclerosis (scleroderma).³ There exists heterogeneity in the prevalence rate and incidence of Sjögren’s syndrome due to variances in its classification criteria and study design. Globally, Sjögren’s syndrome occurs in 61 out of 100,000 people, only about 3% of those aged 50 years and over, with an average age of 56 years old. Most cases occur in those aged 20 to 40 years. Women to men ratio is on average between 9:1 to 19:1. Sjögren’s syndrome is typically identified during the 5th decade of life, with an average age range between 51.6 (±13.8) to 62 (±13) years, nonetheless initial symptoms may manifest years prior to a confirmed diagnosis.⁴

Tubulo-interstitial nephritis and/or renal tubular acidosis are present in about 25–40% of individuals afflicted with Sjögren’s syndrome.⁵⁶ Extremities paralysis is a well-known hypokalemia complication as a consequence of renal tubular acidosis. This sustained condition of severe hypokalemia may lead to a progression of muscle weakness, which may potentially result in a respiratory arrest as a result of muscle paralysis. We report a case of respiratory failure resulting from severe hypokalemia in a patient with Sjögren syndrome.

CASE REPORT

A 46-year-old woman was admitted to the emergency department with progressive limb weakness and respiratory distress. No history of upper respiratory tract, bowel, or bladder infection, or recent travels abroad was reported. There were no records of patients taking medications.
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received methylprednisolone 62.5mg/24 hour combined with continuous intravenous administration of 25% potassium-chloride with a total of 1,200 mEq in 5 days eventually bringing potassium levels within normal limits, muscle weakness improves and the patient was extubated, shown in Table 1.

DISCUSSION

Sjögren’s syndrome is extremely rare, mainly due to its nonspecific manifestations, which frequently go undiagnosed or overlay with other autoimmune disorders. In this exceptionally unique case, the patient exhibited erratic renal symptoms with primary Sjögren’s syndrome, including diabetes insipidus, renal tubular acidosis, TIN, and nephrolithiasis (kidney stones). These clinical findings are not typically correlated with primary Sjögren’s syndrome. The simultaneous presence of all the aforesaid clinical manifestations in this patient has led to an unconventional diagnosis of primary Sjögren’s syndrome. On average, Sjögren’s syndrome affects middle-aged women, and the sex-adjusted prevalence value is a range of 2.2-10.3 for every 10,000 individuals.

The diagnosis of primary Sjögren’s syndrome normally relies on the American-European Consensus Group (AECG) criteria. Other commonly used criteria that were based on AECG include the Copenhagen criteria, the European classification criteria, and the International Collaborative Clinical Alliances Cohort. The AECG criteria include the presence of ocular and oral manifestations, lymphocytic sialadenitis on minor salivary gland (typically obtained and observed through biopsy), and the presence of specific autoantibodies (i.e., anti-Ro/SSA and –La/SSB). Patients are typically required to meet at least four out of the six listed criteria for a diagnosis. However, these diagnostic criteria are less comprehensive when discussing juvenile cases, consequently making diagnoses in adolescents with atypical symptoms more perplexing. Tabea Seeliger et al. describe the initial symptoms of Sjögren’s syndrome in a study involving 184 patients who indicated limb weakness and found a significant association between severe

On admission, a physical examination revealed a decrease in muscle strength in all extremities scoring a grade of 2/5, her speech was feeble. Her eyes displayed a complete range of movement and no sign of sensory loss. No abnormalities of cardiac or pulmonary signs were detected. However, she abruptly experienced respiratory failure, leading to emergency intubation for mechanically-assisted ventilation, then immediately followed by her transfer to the intensive care unit. Head CT scan and cardiac ultrasound showed no abnormalities (Figure 1).

Complete laboratory tests were taken and the result as follows: blood gas analysis pH 7.02, PaCO2 42, pO2 187, HCO3- 11.4, BE -20, SO2 99% with nasal Canul, serum sodium 138 mmol/L, potassium 1.1 mmol/L, chloride 93 mmol/L, calcium 7.3 mg/dL, magnesium 1.8 mg/dL, creatinine 2.5 mg/dL, blood urea nitrogen 31 mg/dL, blood glucose 150 mg/dL, leukocyte count 36,870/uL, hemoglobin 11.9 gr/dL, platelets 591,000/uL. Urinalysis showed urinary pH 7, haematuria 2+, and proteinuria 1+. Relatively high excretion of urinary potassium was observed at 70 mmol/day, despite extreme hypokalemia. To rule out other etiology for hypokalemia, we ran thyroid function tests and turned out normal. Considering the gathered available data, we concluded that severe hypokalemia induced by renal tubular acidosis was the likely cause of the aforementioned respiratory arrest and quadriplegia experienced by the patient.

Abdominal USG showed bilateral parenchymal kidney diseases with nephrolithiasis. Further investigation in immunology revealed that C3 and C4 were within normal limits. Anti-dsDNA antibody and anti-Sm antibody were negative, with strong positive titer in anti-Ro and PM100. Then the patient was diagnosed with Sjögren’s syndrome and

<table>
<thead>
<tr>
<th>Day</th>
<th>Potassium (mmol/L)</th>
<th>Replacement (mmol/24 hours)</th>
</tr>
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<tbody>
<tr>
<td>Day 1</td>
<td>1.1</td>
<td>250</td>
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<tr>
<td>Day 2</td>
<td>2.2</td>
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<tr>
<td>Day 3</td>
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<td>250</td>
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<tr>
<td>Day 4</td>
<td>2.5</td>
<td>250</td>
</tr>
<tr>
<td>Day 5</td>
<td>3.0</td>
<td>200</td>
</tr>
</tbody>
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Table 1. Potassium level and potassium replacement requirement

Table 2. Initial Symptoms of Sjögren’s Syndrome

Paraesthesia (isolated), n (%) 17 (39%) Paraesthesia with paresis, n (%) 6 (14%) Paresis (isolated), n (%) 11 (25%) Gait disturbance, n (%) 5 (5%) Bladder dysfunction, n (%) 2 (2%) Dyspnoea, n (%) 1 (2%) Facial palsy, n (%) 1 (2%)

Figure 1. Head CT without contras.

Table 2. Initial Symptoms of Sjogren’s Syndrome

Tabea Seeliger et al.
neuropathy and Sjögren's syndrome. This can be seen in detail in Table 2. This suggests that serious neuropathy and limb weakness can frequently be directly linked to Sjögren's syndrome in some patients.9

In our case, no sicca symptoms were found. Her previous medical history showed no similar complaints, and she didn't take any medications. However, she came with a chief complaint of muscle weakness and soon progressed to respiratory failure. Later, laboratory findings at admission showed severe hypokalemia (1.1 mEq/L) with metabolic acidosis (pH 7.02) and urinary pH at 7 suggesting that the renal tubular acidosis is a distal type. Under these circumstances, the rapid use of sodium bicarbonate for alkalization purposes exacerbates the rapid use of sodium bicarbonate for alkalization purposes exacerbates the rapid use of sodium bicarbonate for alkalization purposes exacerbates the rapid use of sodium bicarbonate for alkalization purposes exacerbates the rapid use of sodium bicarbonate for alkalization purposes exacerbates the rapid use of sodium bicarbonate for alkalization purposes exacerbates the rapid use of sodium bicarbonate for alkalization purposes exacerbates the rapid use of sodium bicarbonate for alkalization purposes exacerbates the rapid use of sodium bicarbonate for alkalization purposes exacerbates the rapid use of sodium bicarbonate for alkalization purposes exacerbates the rapid use of sodium bicarbonate for alkalization purposes exacerbates the rapid use of sodium bicarbonate for alkalization purposes exacerbates the rapid use of sodium bicarbonate for alkalization purposes exacerbates.

The occurrence of distal RTA in individuals with Sjögren's syndrome has been documented at an estimated 25-40% value. Tsuboi et al. noted that periodic paralysis was present in nearly 40% of Sjögren's syndrome cases linked to distal RTA, with all cases, including our own, being females in gender.10-12 These cases exhibited a favorable prognosis, with rapid respiratory support and the administration of potassium and alkali, leading to an almost instant positive response.

CONCLUSIONS
While respiratory arrest correlated to Sjögren's syndrome is a highly unusual occurrence, this complication is remarkably critical and potentially lethal. Therefore, when an adolescent presents severe neuropathy and renal manifestations such as renal tubular acidosis, it is obligatory to conduct further diagnostic testing to eliminate the possibility of Sjögren's syndrome. Simultaneously, these individuals should obtain adequate treatment with respiratory support, potassium, and alkali repletion therapy to enable rapid clinical recovery and potentially fatal sequelae.

CONFLICT OF INTEREST
There's no conflict of interest in this study.

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REFERENCES