Case Report

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RENAL TUBULAR ACIDOSIS DUE TO SJOGREN'S SYNDROME PRESENTING AS HYPOKALEMIC QUADRIPARESIS: A CASE REPORT

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ABSTRACT

Sjögren's syndrome is characterized by dry mouth and eyes symptoms caused by lymphocyte infiltration of the exocrine glands as it is an autoimmune condition. Nonetheless, this condition can lead to symptoms and various illnesses. Renal tubular acidosis (RTA) occurs when the kidney tubules are unable to regulate acid-base balance. This study tracked a 40-year-old female who initially showed symptoms of hypokalemic stroke and normal anion gap acidosis and was later determined to have Sjögren's syndrome. The patient complains of weakness in the legs, vomiting, abdominal pain and dry cough. Like other Sjögren's symptoms, dry eyes and mouth have been present for years. Electrolyte abnormalities and lymphocytic infiltration in the lymph nodes can be confirmed by physical examination and laboratory tests. Positive antibodies, Schirmer test, and histology results indicate the diagnosis. Treatments include potassium replenishment, artificial tears, and the use of coconut oil. Early diagnosis and treatment can help prevent the condition from worsening and improve patient outcomes. It also emphasizes the importance of completing a comprehensive evaluation that includes organ system and specific markers of disease.

KEYWORDS: Sjogren Syndrome, Renal Tubular Acidosis, Hypokalemic Paralysis..

Sjögren's syndrome is a condition where there is an immune system attack on the the exocrine glands, which cause symptoms like dry eyes and mouth, along with potentially affecting other organs in the body. Approximately 33 percent of people with Sjogren's syndrome encounter systemic symptoms. (2) RTA is characterized by renal tubular failure within the context of normal acid-base balance. Tubular transporters, which play a role in secreting or absorbing specific ions, are frequently lacking because of various factors such as autoimmune disorders, cancer (like multiple myeloma), genetic disorders, exposure to toxic drugs affecting the kidneys, diuretic use, and other illnesses. RTA can be classified into three main categories: hyperkalemic (type 4), types 2 and 1, or proximal and distal. Differential blood potassium levels (high or low), irregular urine pH, and either the three types of RTA are characterized by hyperchloremic non-anion gap metabolic acidosis or a positive urine anion gap (3, 11). Renal involvement is prevalent and may initially manifest as RTA-I, with hypokalemia and normal anion gap acidosis. Here, we provide a case study of a woman in her 40s who Originally had normal anion gap acidosis and hypokalemic paralysis, but was subsequently diagnosed with Sjogren's syndrome because of the condition's wide range of clinical symptoms.

CASE PRESENTATION

A 40-year-old female presented to the emergency department complaining of all four limb weakness for 12 hours. It was both abrupt and gradual in its beginning. It began in the lower limbs and progressed to include bilateral upper limbs. Prior to the presentation, she reported repeated instances of vomiting including food particles. Vomiting was coupled with nausea and mild stomach discomfort. She denied any history of diabetes, hypertension, loose stools, night sweats, lymphadenopathy, or weight loss.

Further inquiry showed that she had dry eyes and a dry mouth (figure 01) for the last 6 years, with no/minimal secretions. She also complained of slight vaginal dryness, which caused dyspareunia. It was linked to

trouble swallowing and a gritty feeling in both eyes.

A systemic evaluation revealed no diarrhea, uncomfortable swelling stiff joints, pruritis, epistaxis, hair loss, or a history of abortion. It is important to note that she had no history of upper respiratory tract infection, trauma, spinal injury, diarrhea, hypokalemia, surgery, recent vaccination, a high carbohydrate diet, or recognized carbohydrate and lipid abnormalities.

Her medical background was not very noteworthy. No family or personal history of autoimmune disease was present.

Upon presentation, her vital signs were as follows: heart rate: 150/90 mmHg, blood pressure:

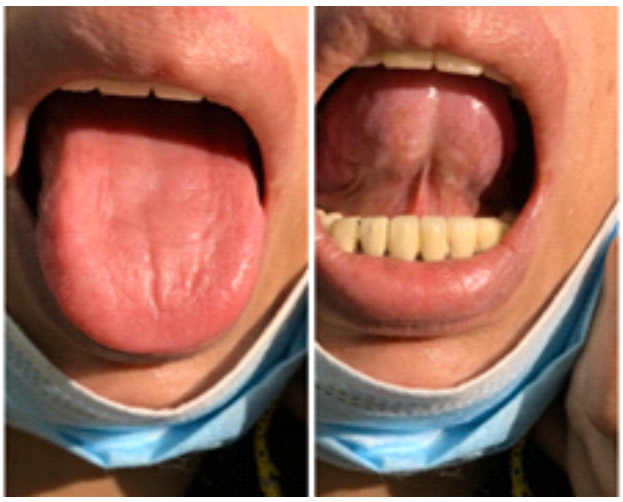

Fig. 1 a). Dry Tongue with loss of shine and saliva. Depapillation over dorsal surface of anterior 2/3rd of tongue. b). Dry Ventral Surface of tongue

100/min, respiratory rate: 20/min, temperature: 98°F, blood sugar: 210 mg/dl, SpO2: 98% at room air.

On physical testing, she was determined to have a GCS of 15/15 and all cranial nerves were intact. Her plantars were downgoing bilaterally, and her pupils were equivalent and responding to light on both sides.

Other neurological examination was as follows:

	Upper limb	**Upper limb**	**Lower limb**	**Lower limb**
	Right	Left	Right	Left
Bulk	normal	normal	normal	normal
Tone	decreased	decreased	decreased	decreased
Power	2/5	2/5	2/5	2/5
Reflexes	normal	normal	Knee jerk present Ankle jerk absent	Knee jerk present Ankle jerk absent

Table 1: Neurological Exam Findings of the patient.

Respiratory examination showed no significant findings.

She had tenderness in pre-auricular region with mild swelling. Initial lab investigations revealed; serum sodium : 141 mmol/L (136-145mmol/L), potassium : 1.71 mmol/L (3.5-5.0 mmol/L), chloride : 113 mmol/L (98-106 mmol/L), Calcium : 9.2 mg/dL (9-10.5mg/dL), Phosphorus : 6.1 mg/dL (3-4.5mg/dL) and Bicarbonate : 15.2 mmol/L (23-28mmol/L). The pH on venous gas was 7.21 (7.38-7.44). Her Hb at presentation was 11.4g/dl , TLC : 14.27x10^3/uL , Platelets : 333x10^3/uL , Urea : 57mg/dl (8-20mg/dL) and Creatinine : 1.47mg/dl (0.7-1.3mg/dL). Urine pH was found to be 6.5 a8-1069nd serum albumin was 3.2g/dL (3.5-5.5g/dL).

Potassium replacement was done according to the potassium deficit ; which helped the patient in regaining power in all four limbs.

The renal ultrasonography scan indicated bilateral nephrocalcinosis. Her ENA profile showed positive antigen-A linked to Sjögren's (anti-SSA) 60kD: 78U/ml (<25U/ml), anti-SSA 52kD: 17U/ml (< 10 ml), and anti-Sjögren's related antigen B: 35U/ml (0-73U/ml) in the presence of sicca symptoms, which was further supported by positive Schirmer's test, i.e. Right Eye= 3mm/5min and Left Eye= 4mm/5min. The saliva flow rate was less than 0.1ml/min (10 mm/5 min). The initial buccal mucosa biopsy was inconclusive due to the lack of salivary gland tissue, and she declined a second biopsy.

As the patient was not having any characteristic complaints of peripheral autoimmune features (like arthralgias, myalgias, arthritis) she was not prescribed

Anti-SSA	**Positive**
Anti-SSB	Positive
Anti-smith	Negative
Anti RNP	Negative
Anti Jo1	Negative
Anti Scl-70	Negative

Table 2: Serological Investigation Results in Patient.

DMARDs. Patient's complaints of dry eyes and dry mouth improved with artificial tears, sugar free gums, application of coconut oil in oral cavity. She was advised to decrease intake of coffee, nicotine, herbal tea and cola in her daily life routine. On follow up sicca symptoms improved gradually. She managed to carry out her daily life activities without any deterioration of health.

DISCUSSION

A type of autoimmune disease is Sjögren's syndrome, marked by the presence of lymphocytes invading the exocrine glands, which cause dry skin and mouth symptoms. It can also manifest as exocrinopathy specific to certain organs or as a systemic disease affecting the skin, lungs, kidneys, and gastrointestinal tract. Type 1 RTA is frequently caused by autoimmune diseases such as Wilson's disease, hyperparathyroidism, vitamin D intoxication, and sarcoidosis; medications such as amphotericin B, ibuprofen, and ifosfamide; and rheumatic arthritis SLE, or systemic lupus erythematosus, and Sjögren's syndrome (5, 6, 7).

Sjögren's syndrome's hallmark sicca symptoms are not necessarily the presenting complaint. Some studies have observed muscular weakness, joint discomfort, and hypokalemic paralysis as a result of RTA complications. Thus, RTA symptoms might lead to the identification of Sjögren's syndrome. (12). Doctors' diagnosis of Sjögren's syndrome has been complicated by the focus placed on symptoms of dry mouth and eyes, particularly when the symptoms at presentation are not the same as those of dry mouse and eyes. Any four of the six criteria—oral and ocular symptoms, ocular signs, histology, involvement of the salivary glands, and serum antibodies to Ro/SSA or La/SSB—must be met in order to diagnose primary Sjögren's syndrome. (4, 8, 9)

The basis for our Sjögren's syndrome diagnosis was the fulfillment of all six criteria in our patient. While renal ultrasonography results related to nephrocalcinosis and renal tubular acidosis type I may not definitively point to Sjögren's syndrome, they do provide additional evidence for the diagnosis and show renal involvement. The primary renal problem associated with Tubulointerstitial nephritis is what Sjögren's syndrome is, which in adulthood can result in RTA. On the other hand, RTA is rare in children, accounting for a tiny portion of instances with hypokalemic paralysis or renal potassium wasting (3, 10). As in our case, lymphocytic infiltration of the nephron might emerge as hypokalemia, acidosis, and RTA (predominantly type I).lvement, which ranges from 0.3% to 27% in the literature. When there is renal tubular acidosis in Sjögren's syndrome, the histological findings of the salivary glands, lacrimal glands, and kidneys show similar lymphocytic infiltration in all three tissues. (11) In the beginning, our patient had hypokalemic periodic paralysis and renal tubular acidosis indicating the existence of lymphocytic infiltration of renal tubules, which impaired glomerular filtration. Dry eyes and mouth are symptoms of lymphocytic infiltration of the lacrimal and salivary glands.

CONCLUSION

Sjögren's syndrome can affect the kidneys, most often causing tubulointerstitial nephritis. It can result in RTA, as demonstrated in this situation. The lymphocytic infiltration detected in the renal tubules might disrupt glomerular filtration and produce electrolyte imbalances. This example underlines the need of considering all possible diagnoses, including Sjögren's disease, in individuals who come with unexplained RTA. Early identification and care can aid in the prevention of disease development and the improvement of patient outcomes. Clinicians can treat patients with rapid and effective results when they treat patients with unexplained RTA by considering Sjögren's disease as a differential diagnosis.

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