Case Report

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Simoni Shah , Zeal Soni , Nilay Suthar , Dev Desai *

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Case Report

Primary Sjogren's Syndrome: A Rare Case of Hypokalemic Periodic Paralysis

Simoni Shah [1], Zeal Soni [2], Nilay Suthar [3] and Dev Desai [4,*]

[1] Narendra Modi Medical College, LG Hospital, Ahmedabad, India; Email Address: - simoni.shah85@gmail.com
[2] Smt. NHL Municipal Medical College, Ahmedabad, India; Email Address: rlzs040@gmail.com
[3] Professor, Department of Medicine, Smt. NHL Municipal Medical College, Ahmedabad, India; Email Address: - nilaysuthar@gmail.com
[4] Smt. NHL Municipal Medical College, Ahmedabad, India; Email Address: devhdesai01@gmail.com

Abstract: A multisystem chronic autoimmune disorder, Sjogren syndrome, is characterized by lymphocytic infiltrates in the exocrine glands. Keratoconjunctivitis sicca and xerostomia are the two pathognomic findings in Sjogren syndrome. Here, we present a case of a 30-year-old an old Indian woman presenting with primary Sjogren syndrome. The clinical features, diagnostic modalities, treatment options, and management of complications have been discussed.

Keywords: Sjogren syndrome; periodic paralysis; Hypokalemia; autoimmine

INTRODUCTION:

Sjogren syndrome, an autoimmune disorder that primarily targets the exocrine glands (most commonly, the salivary glands and the lacrimal glands), presents with the main complaints of dry eyes and dry mouth [1]. The age of presentation is usually after 40 years. Autoantibodies and autoimmune B cells target the exocrine glands, resulting in their hypertrophy [2]. With further disease progression, it also affects other organs such as the liver, kidney, thyroid, vagina, nerves, etc. Diagnosing this condition is difficult because it does not present with evident clinical symptoms. Positive anti-Ro and anti-La antibodies are characteristic [3]. A salivary gland biopsy showing focal lymphocytic infiltrates is the confirmatory test. Treatment includes artificial tear drops, pilocarpine for dry mouth, and other treatments depending on the symptoms. There is no specific cure for it.

CASE PRESENTATION:

A 30-year-old woman came to our hospital for evaluation of sudden onset of quadriparesis for two days. The patient had no history of fever, trauma, chronic diarrhea, increased urine production, palpitations, tremors, excessive sweating, and chronic drug use. There was also no history of double vision, drooping of the eyelids, difficulty in speech or swallowing or food regurgitation, sensory complaints, involvement of the bowel and bladder, difficulty breathing, muscle cramps, joint pain, or variation in weakness in the day.

PAST HISTORY:

The patient has no history of HTN, DM, or tuberculosis. The patient is currently not taking any medication.

PHYSICAL EXAMINATION:

On examination, the patient was conscious and oriented to time, place, and person. Pulse-84 / min regular, B.P 120/80mmhg, Respiratory rate 16 / min. Neurological examination: hypotonia in all

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four limbs. The power in both the upper and lower extremities is (0/5). The deep tendon reflex in both limbs had diminished. Plantar bilateral flexor. The cranial nerve examination was normal.

LAB EXAMINATION:

Table 1. Blood Examination.

Test	Observed value	Reference Range
Haemoglobin	10.3 g/dl	(12-18)
WBC	10.8 kU/L	(5.2-12.4)
RBC	4.67*10^6/ul	(4.5-5.5)
Haematocrit	40.7%	(40-50)
Platelet counts	2.04 kU/L	(130-400)
Neutrophile	85%	(49-74)
Lymphocyte	07%	(26-46)
Monocyte	06%	(2-12)
Eosinophil	02%	(0-5)
Basophil	00%	(0-2)

Table 2. Renal Function Test.

Test	Observed value	Reference Range
Blood Urea	37.0 mg/dl	(15-45)
Creatinine serum	0.9 mg/dl	(0.5-1.1)
Sodium serum	136 mmol/L	(132-146)
Potassium serum	2.8 mmol/L	(3.5-5.5)
Chlorine serum	114 mmol/L	(99-109)

Table 3. Urine Examination.

Test	Observed value	Reference Range
Urine sodium	74.24 mmol/L	30-280 mmol/L
Urine potassium	32.1 mmol/L	40-80 mmol/L
Urine chlorine	50.6 mmol/L	110-250 mmol/L
Urinary pH	7.5	5-8

Table 4. Autoimmune Workup.

Test	Observed value
Rheumatoid factor	64 IU/ml
Antinuclear antibody	+++
SSA/Ro 60 kD	58 U/ml
SSA/Ro 52 kD	89 U/ml
SSB	39 U/ml
Gp210	16 U/ml

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TREATMENT:

The patient was initially treated with an IV potassium supplement and an IV bicarbonate replacement. Within 24 hours, the patient's serum potassium level came within normal range and the patient's clinical condition improved. The patient is then moved to oral potassium and oral bicarbonate supplements and discharged with the same medication. The patient was explained about other features of Sjogren syndrome and asked for close follow-up.

FOLLOW UP CASE:

The patient was followed for 6 months and there has been no aggravation of the condition.

DIAGNOSIS:

The patient was given the appropriate treatment and showed improvement afterward. Based on laboratory findings and clinical presentation, the patient is diagnosed with hypokalemic paralysis due to distal renal tubular acidosis caused by primary Sjogren syndrome.

DISCUSSION AND LITERATURE:

Sjogren syndrome is an autoimmune disorder characterized by dry eyes and dry mouth. It is associated with other autoimmune disorders such as rheumatoid arthritis, systemic lupus erythematosus, etc. It can occur at any age, but most often develops after the age of 40 years [4]. Females are more prone to Sjogren's than men. It can be primary or secondary. Primary Sjogren's syndrome is not associated with connective tissue disorders, while secondary Sjogren's has associated comorbidities such as SLE, RA, and Scleroderma.

The pathogenesis of Sjogren syndrome is a multi-step process. Its clinical manifestations develop in the presence of a triggering environmental factor such as infections, immunosuppression, etc. in a genetically susceptible individual [5]. Autoimmunity and chronic inflammation are the two main processes that result in this disease. Autoreactive B cells and autoantibodies first target the salivary and lacrimal glands.

Clinical symptoms vary from person to person. The most common symptoms are keratoconjunctivitis sicca (dry eyes) and xerostomia (dry mouth). The eyes feel gritty as if sand is in the eyes, while the mouth feels full of cotton, making it difficult to speak and swallow. Other extra-glandular features include [6]:

- Raynaud phenomenon: most common extra glandular presentation
- Cutaneous vasculitis
- Arthritis
- Interstitial lung disease
- Non-Hodgkin lymphoma
- Vaginal Dryness

Sjogren syndrome is difficult to diagnose because its presentation varies from person to person. Furthermore, not all tests are sensitive to Sjogren's diagnosis. The different diagnostic tests used are as follows:

1. Schirmer test [7]: Here, a sterile strip of filter paper is placed under the lower eyelid and then the area moistened by the absorbed fluid is measured after 5 min. More than 10mm absorption is considered normal; anything less than that suggests dryness of the eyes.
2. Positive anti-Ro (SSA) and anti-La (SSB) autoantibodies.
3. Salivary gland biopsy [8]: It shows focal lymphocytic sialadenitis. It is the confirmatory diagnostic test.
4. Other tests such as sialogram, salivary scintigraphy, kidney and liver function tests, etc. can be done to aid diagnosis further and look for complications.

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The complications are mainly related to autoimmunity against exocrine glands [9]. These include:

1. Dental caries: A dry mouth (decreased saliva) provides a favorable medium for bacterial overgrowth resulting in caries.
2. Yeast infections, especially oral thrush,
3. Vision problems: Dry eyes result in blurred vision and corneal damage.
4. Non-Hodgkin lymphoma

Other less common complications depend on the organ system affected:

- Lungs: Bronchitis, pneumonia
- Liver: hepatitis, cirrhosis
- Kidney: altered renal function and electrolyte imbalance resulting in distal renal tubular acidosis. Alternate electrolytes can cause serious complications such as hypokalemic paralysis, cardiac arrhythmias, and altered mental status.
- Nerves: peripheral neuropathy

Being an autoimmune disorder, there has not been a definitive cure for Sjogren syndrome. Symptomatic treatment is given [10]:

- Artificial tears for dry eyes.
- Cyclosporine to reduce eye inflammation.
- Pilocarpine to increase saliva production.
- Vaginal lubricants for vaginal dryness.
- NSAIDS for arthralgias/arthritis.
- Antifungal for yeast infections.
- Immunosuppressants such as methotrexate can be administered, having shown some improvement in the patient's condition.
- Surgery is performed in the cases of refractory dry eyes.

CONCLUSION:

Sjogren syndrome, also known as keratoconjunctivitis sicca, is a chronic autoimmune disorder characterized by exocrine and extra-glandular symptoms. Although the most common symptom is dryness of the eyes and mouth, it can also present with fatigue, arthritis, vaginal dryness, etc. There are various diagnostic tests available, but no specific treatment is available. Further research is needed to find a cure for the same. Studies have shown that immunosuppressants have given promising results in treating Sjogren syndrome.

Ethical Statement: Being a case report study, there were no ethical issues and the IRB was notified about the topic and the case. Still, no formal permission was required as this was a record-based case report. Permission from the patient for the article has been acquired and ensured that their information or identity is not disclosed.

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Conflicts of Interest: The authors declare no conflicts of interest.

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