Case Report

Citation: Kashyap, A., Gosavi, S., Savanth, S. and Mehta, S.R., 2020. Primary Sjogren's Syndrome Manifesting as Recurrent Hypokalemia with Distal Renal Tubular Acidosis. Sri Lanka Journal of Medicine, 29(2), pp 77-79
DOI: <http://doi.org/10.4038/sljm.v29i2.227>

Primary Sjogren's Syndrome Manifesting as Recurrent Hypokalemia with Distal Renal Tubular Acidosis

Ajeya Kashyap, Siddharth Gosavi, Sharath Savanth and Sudha Rani Mehta

Department of General Medicine, JJM Medical College, Davanagere

Correspondence: Siddharth Gosavi
Postgraduate Resident, Department of General Medicine, JJM Medical College, Davanagere.
Email: ramshyamsid@gmail.com

<https://orcid.org/0000-0001-7072-2154>

Abstract

Sjogren's syndrome is an autoimmune disease with both glandular and extraglandular manifestations.

We report a rare case of primary Sjogren syndrome without sicca symptoms presenting with renal tubular acidosis and recurrent hypokalaemia.

Keywords: *Sjogren's Syndrome, Hypokalaemia.*

INTRODUCTION

Sjogren's syndrome is defined as a chronic, slowly progressing autoimmune disease characterized by lymphocytic infiltration of exocrine glands. Purpura, Raynaud's phenomenon and cutaneous vasculitis are very uncommon features of this condition. It may be associated with myopathy, arthralgia, thyroiditis, depression, immune mediated interstitial nephritis, hypergammaglobulinemia, and cryoglobulinemia. Common serological findings include Anti SS-A, Anti SS-B and Rheumatoid factor [1].

It most commonly presents as xerostomia and dry eyes called keratoconjunctivitis sicca.

CURRENT CASE

Ethical clearance was obtained from the institutional ethics committee and informed written and oral consent was obtained from the patient for publication of clinical details.

A 45year old female presented to the emergency ward of Chigateri General Hospital attached to JJM Medical college, Davangere, complaining of weakness of bilateral upper and lower limbs for 6 weeks. Similar episodes associated with myalgia and joint pains during the past 3 months, had

been treated for hypokalemia with clinical improvement each time. There was no history of altered sensorium, seizures, altered bowel and bladder movements, dryness of eyes, vomiting, diuretic/alcohol/laxative abuse or any major comorbidities. Examination revealed hypotonicity of both lower limbs, weakness of both proximal and distal muscles with power of 2/5 in the lower limbs and 3/5 in upper limbs. Deep tendon reflexes were diminished with bilateral plantar reflex mute. The Schirmer test was negative. There was no salivary gland enlargement.

The serum potassium was 1.9mmol/l. The full blood count, renal function tests, liver function tests, thyroid function tests, serum magnesium levels (2mg/dl) and serum calcium (8.8mg/dl) levels were within normal limits. Tests for HIV, HBsAg, and HCV were negative. The urinary sediment and cultures were negative. The C-Reactive protein level was 72mg/dl. Arterial blood gas (ABG) and urine electrolytes showed metabolic acidosis with hyperchloremia with high urine anion gap. Distal renal tubular acidosis was diagnosed. Serum anion gap was 7.8 mmol/L and urine anion gap was 16 mmol/L. ANA screening and profile revealed Antinuclear antibodies (ANA) were positive at titers of 1:160. Anti SS-A and Anti SS-B were positive. Rheumatoid factor was negative.

Potassium correction was started from the day of admission. After investigation, the patient was started on Hydroxychloroquine, and intra-venous and oral potassium, and sodium bicarbonate. As the patient had joint pain and myalgia, with raised C reactive protein, she was started on high dose oral prednisolone of 1mg/kg/day. The potassium level increased from 1.9 to 2.7mmol/l, and at discharge was 3.8mmol/l. A lip biopsy showed salivary lobules of sero-mucinous glands and acini with one significant periductal lymphocytic infiltrate (>50 lymphocytes/hpf) with a few plasma cells. A dense plasmacytic infiltrate was seen elsewhere (not included in estimated focus score). Minimal fibrosis was noted. Estimated glandular area was 1.272mm² while plasma cells (in the estimated foci) were <10%. These histological findings were compatible with Sjogren's disease with a focus score of 3.14. There was no facility in our hospital to conduct IgG4 staining.

DISCUSSION

This patient presented with a history of repeated hospitalization due to hypokalemia, with the last episode associated with quadriparesis, lower limb weakness predominating. Myasthenia gravis, compressive myelopathy, transverse myelitis, and drug induced neuropathy were ruled out. Our case had refractory hypokalemia causing acute flaccid paralysis. As the patient had histological features of Sjogren's syndrome along with positive anti SS-A and anti SS-B without sicca symptoms, a diagnosis of undifferentiated connective tissue disorder or incomplete Sjogren's syndrome was made [2].

Distal renal tubular acidosis is reported in less than 2% of patients with primary Sjogren's syndrome [3].

This patient presented with hypokalemic paralysis due to distal renal tubular acidosis as the first and only clinical manifestation of Sjogren's syndrome.

She showed good response to potassium replacement therapy, and sodium bicarbonate. There is a need to consider Sjogren's syndrome with distal renal tubular acidosis as a cause in patients with recurrent unexplained hypokalemia even in the absence of sicca symptoms.

CONCLUSION

Hypokalaemic periodic paralysis due to distal renal tubular acidosis could be the first manifestation of Sjogren syndrome. Diagnosis of Sjogren's syndrome in a patient without sicca symptoms will alter the management of patient. Monitoring of the patient and recognition of other manifestations of Sjogren Syndrome that is amenable to immunomodulatory therapy (e.g. inflammatory arthritis, interstitial lung disease), or overlap autoimmune syndromes (e.g. rheumatoid arthritis, SLE) is important. Patients with Sjogren's Syndrome have a significantly elevated risk of lymphoma, especially GI lymphomas, for which they need to be periodically monitored in the long term.

Author declarations

Acknowledgements

No acknowledgements to disclose

Author contributions

AK developed the concept of the case and acquired the necessary clinical data. SG contributed to the design of the report. SS helped in drafting the case report. SRM revised the report critically for intellectual content. AK and SG approved the final version of the case report.

Funding sources

There was no source of funding.

Availability of data and materials

Clinical data is available with all the authors.

Ethics approval and consent to participate

Ethical clearance was obtained from the institutional ethics committee and informed written and oral consent was obtained from the patient for publication of clinical details.

Competing interests

None conflicts of interest to disclose.

REFERENCE

1. Baer A (2020) Clinical Manifestations of Sjogren Syndrome exocrine gland disease. Uptodatecom. Available: <https://www.uptodate.com/contents/clinical-manifestations-of-sjogrens-syndrome-exocrine-gland-disease>. Accessed 4 October 2020.
2. Jonsson R, Brokstad K, Jonsson M, Delaleu N, Skarstein K (2018) Current concepts on Sjögren's syndrome – classification criteria and biomarkers. European Journal of Oral Sciences 126: 37-48. DOI: <https://doi.org/10.1111/eos.12536>.
3. Sarma A (2020) Hypokalemic paralysis due to primary Sjogren syndrome. Ijemin. Available: <http://www.ijem.in/text.asp?2018/22/2/287/232393>.