Sjogren’s Syndrome Presenting with Hypokalemic Periodic Paralysis


Abstract
We report a rare case of a 38 year old female who presented with sudden onset flaccid quadriplegia and respiratory arrest with no significant past clinical history. She was later found to have hypokalemia due to distal renal tubular acidosis and further diagnosed as case of Sjogrens Syndrome.

Discussion
There have been few case reports where hypokalemic periodic paralysis has been investigated and has led to the diagnosis of Sjogren Syndrome. The first such case report is as early as 1981.¹ India’s first such case was reported in1996 in JAPI by Thomas et al. from CMC Vellore.² Rao et al 2006 studied 31 cases of hypokalemic periodic paralysis where 3 cases had Sjogren Syndrome.³ This indicates that 10% of people with hypokalemic periodic paralysis can have Sjogren syndrome which if diagnosed early has more management options available.

Our patient, an otherwise healthy female, presented with acute onset flaccid quadriplegia without sensory involvement. A differential Diagnosis of AIDP and HPP was considered. Hypokalemia favored the second diagnosis which was strengthened by immediate improvement of respiratory arrest by potassium supplementation. Further, metabolic acidosis and raised TTKG indicated towards Distal RTA.

Distal RTA may be primary or more commonly secondary to paraproteinemia, medullary sponge kidney, nephrocalcinosis, obstructive uropathy and autoimmune disease.⁴ Hypokalemia due to dRTA is usually a late manifestation. However it has been reported in < 2% cases of Sjogren syndrome (SS) as a presenting feature has been documented only in 4 cases.⁵-⁷ Our case lacked ocular and oral dryness which may develop subsequently.⁸ The systemic manifestations necessitate corticosteroids, cytotoxic agent or both. Our patient is presently receiving low dose of steroid therapy and is currently asymptomatic.

We report this case to highlight the fact that patients presenting with HPP can be due to early SS which needs to be investigated and started on appropriate line of management.

References

38 year female with no significant past and family history was brought to the emergency ward with sudden onset weakness since one day. She was afebrile with a pulse rate of 78/min in sinus rhythm and BP 120/80 mm/Hg. Within minutes of arrival, she developed sudden respiratory failure and was intubated. She was shifted to intensive care unit where respiratory rate was 14/min on ventilator support with IPPV mode. There were no enlarged lymph nodes or parotid glands or thickened nerves.

She had bilateral symmetrical flaccid quadriplegia with weakened neck muscles, diminished reflexes and mute planters. There was no cranial nerve or sensory involvement. All this was highly s/o a AIDP. However, EMG/ NCV ruled out AIDP.

Serum sodium was 150mEq/L and serum potassium was 2.5 mEq/L. ECG also showed prominent U waves. She was in metabolic acidosis with pH was 7.085 and a normal anion gap. Liver and Renal functions were normal. Transtubular potassium gradient (TTKG) = 11 indicated towards Renal Tubular acidosis (RTA) Hypokalemia was corrected with i.v. potassium chloride. On 5th day respiration improved and she was weaned off the ventilator.

Autoantibody screen revealed positive antinuclear antibody titer of 1:320 with speckled pattern. Anti dsDNA, anti-Sm antibodies, rheumatoid factor and anti U1snRNP were negative. Thyroid function tests were normal. Anti-Ro (SS-A) and anti-La (SS-B) antibody were strongly positive in very high titer of 1:320 with speckled pattern. Anti dsDNA, anti-Sm and anti U1snRNP were negative.

A salivary gland biopsy demonstrating multiple foci of lymphocyte and plasma cell infiltrate with ducts revealing intraepithelial lymphocyte infiltrate further corroborated Sjogren’s syndrome (Figure 1). Patient could not afford salivary gland scintigraphy and sialography. Renal biopsy was refused by patient.

Patient has been maintained on potassium chloride solution, sodium bicarbonate capsules, low dose prednisolone and is on regular outpatient follow up.


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**Fig. 1:** Multiple foci of periductal and intra epithelial lymphoplasma cell infiltrate are seen forming lymphoepithelial islands.