

## Hypokalemic Paralysis as a Presenting Manifestation of Primary Sjogren's Syndrome – A Case Report.”



### Medical Science

**KEYWORDS :** Hypokalemic paralysis, Sjogren's syndrome.

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### ABSTRACT

*Sjogren's syndrome is a chronic slowly progressive autoimmune disease characterised by lymphocytic infiltration of exocrine glands resulting in xerostomia and dry eyes with systemic manifestations. The disease may present alone (primary sjogren's syndrome, pSS) or in association with other autoimmune rheumatic diseases (secondary sjogren's syndrome). It primarily affects middle aged women. We herein report a case who presented with hypokalemic paralysis which on further work up found to be secondary to pSS.*

### INTRODUCTION:

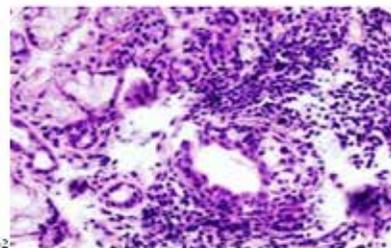
- Primary sjogren's syndrome (pSS) is a chronic autoimmune disease characterized by a progressive lymphocytic infiltration of exocrine glands with systemic involvement. Renal tubular acidosis (RTA) caused by tubulointerstitial nephropathy is a common extraglandular manifestation of pSS. We herein present a case of pSS manifesting as hypokalemic paralysis caused by distal RTA. Therefore pSS should also be suspected in cases of hypokalemic paralysis.

### CASE REPORT :

- A 30yr old female, who is farmer by occupation presented with sudden onset weakness of all four limbs since 1 day with history of similar complaints 4yrs back after which she recovered in 2days of hospitalization. History of dryness of mouth, excessive hair loss since 4yrs. She was afebrile with pulse rate 88/min in sinus rhythm and BP 110/80 mm of Hg. Examination revealed hypotonia in all limbs with power of 0/5 in all limbs with significant weakness in trunk and abdomen muscles. Deep tendon reflexes are absent with normal superficial reflexes and flexor plantars.
- Lab investigation showed Sr.Potassium-1.52meq/L, ABG (Arterial blood gas analysis) suggestive of metabolic acidosis (PH 7.146)  $\text{HCO}_3^-$ :7.8meq/l,  $\text{Pco}_2$ —23mmofHg, Anion gap- 10(Normal), ECG showing U waves. EMG/NCV is normal (AIDP was ruled out). 24hrs urine potassium level 84 meq/day indicating renal loss, TTKG (Trans-tubular potassium gradient):6 indicating Renal tubular acidosis, urinary citrate :147 mg per day indicating distal RTA. Autoantibody screening revealed ANA positive with florescence pattern, Serum anti-Ro (SS-A) & anti-La (SS-B) antibody levels were strongly positive. Schirmer's test positive (Figure 1). Buccal and Mucosal biopsy demonstrating multiple foci of lymphocyte and plasma cell infiltrate with ducts revealing intraepithelial lymphocytic infiltrate further corroborated Sjogren's syndrome. (Figure 2). Patient could not afford salivary gland scintigraphy and sialography. Renal biopsy was refused by patient.
- Patient improved after treating with i.v. and oral potassium supplements and sodium bicarbonate. Power improved from 0/5 to 4/5 within 2 days and at the time of discharge patient had normal power. Patient was discharged on oral potassium and alkali supplementation and was diagnosed as Primary Sjogren's Syndrome with Distal Renal Tubular Acidosis with Hypokalemic paralysis.



• FIGURE-1



• FIGURE-2

### DISCUSSION :

- Cases of pSS manifesting for the first time as hypokalemic paralysis caused by distal RTA have been rarely reported. Our patient presented with acute onset flaccid quadriplegia without sensory involvement. A differential diagnosis of AIDP and HPP was considered but hypokalemia and normal NCV favoured hypokalemic periodic paralysis. Further metabolic acidosis and raised TTKG indicated towards distal RTA. Distal RTA may be primary or secondary to autoimmune disease, nephrocalcinosis, obstructive uropathy, paraproteinemia and medullary sponge kidney. The symptoms of dryness of mouth and excessive hair loss points towards autoimmune disease which on further evaluation (autoantibody screening, schirmer's test, buccal biopsy) favoured sjogren's syndrome.
- pSS is a disease of exocrine glands presenting as dry eyes and dry mouth. Non exocrine organ system involvement includes skin, lung, GIT, CNS-PNS, Musculo-skeletal and kidney. The spectrum of renal disease includes interstitial nephritis which can manifest as distal or proximal RTA, tubular proteinuria, nephrogenic DI, glomerular diseases or renal failure. The most common manifestations are tubular dysfunction with chronic interstitial nephritis with distal RTA with hypokalemia d/t decreased distal tubular sodium delivery, sec-

ondary hyperaldosteronism ,defective H-K ATPase and bicarbonaturia.

- We report this case to highlight the fact that patients presenting with hypokalemic periodic paralysis can be due to sjogren's syndrome which need to be investigated and started on appropriate line of management.

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