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Case Report

# Hypokalemic Paralysis Leading to Respiratory Failure: An Unusual Presentation of Sjogren's Syndrome

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**Abstract**

Sjogren's Syndrome (SS) is a chronic multisystemic autoimmune inflammatory disorder with predominant exocrine gland involvement resulting in dryness of the eyes and mouth. Among extra glandular manifestations, renal involvement is commonly seen, which can be in the form of Renal Tubular Acidosis (RTA). SS may at times present as mild hypokalaemia with distal RTA but severe hypokalemia resulting in respiratory failure is rare. Here we report a case of twenty-five-year-old female presenting in emergency room with flaccid quadriparesis and hypercapnic respiratory failure. Further evaluation revealed hypokalemia (Potassium ($K^+$) 1.6mmol/L) with distal renal tubular acidosis along with a background of primary Sjogren's Syndrome. We illustrate that the possibility of Sjogren's Syndrome should be considered in a young female who present with rapidly progressive weakness, hypokalemia and distal renal tubular acidosis with respiratory failure.

**Keywords:** Hypokalemic Flaccid Paralysis; Distal renal tubular acidosis; Sjogren's syndrome; Respiratory failure

## Abbreviations

ABG: Arterial Blood Gases; ESSDAI: EULAR Sjogren's Syndrome Disease Activity Index; HPP: Hypokalemic Periodic Paralysis; RTA: Renal Tubular Acidosis; SS: Sjogren's Syndrome; TTKG: Trans Tubular Potassium Gradient

## Introduction

Hypokalemic Periodic Paralysis (HPP) is a rare type of periodic paralysis, with a prevalence of 1 in 100,000 population [1-4]. HPP may be familial with autosomal dominant or acquired inheritance. HPP is usually presented with acute onset of generalized weakness, proximal more than the distal. RTA in SS is usually mildly symptomatic, which is observed in up to 25% of patients [2]. Potassium concentrations less than 2mEq/L should suggest a secondary cause of hypokalemic paralysis, such as that seen in some cases of distal RTA but respiratory failure in these patients is quite rare. The potassium levels remain normal between the acute attacks in primary HPP but is at lower level in secondary HPP. The patient in this case report presented to emergency room with respiratory failure and inability to move all four limbs, which is a rare condition, associated with Sjogren's syndrome.

## Case Presentation

A 25yr old female with no known comorbidities was brought to the Emergency Room (ER) with complaints of sudden onset of weakness in all four limbs. There was a history of fall from bike three days back. She developed swelling in ankle following the fall, no bony injury was reported and there was no loss of consciousness or seizures. She was advised analgesics (Diclofenac 75mg BD for 3 days) and steroid (Deflazacort 30mg once daily for 3 days) by a local practitioner. The patient slept at 10 PM in night with no weakness and woke up in Morning with a difficulty in moving all the extremities. Initially patient was taken to a local hospital where MRI brain & spine was done and found out to be normal. In view of her respiratory distress, she was intubated and referred to a Tertiary Care Centre.

On arrival to ER, the patient was intubated and sedated, but arousable on verbal command. She was hemodynamically stable, not on any vasopressors. Neurological examination revealed lower motor neuron lesion with flaccid paralysis, power of 2/5 in upper limbs and 1/5 in lower limbs, neck flexors and extensors 0/5, deep tendon reflexes absent and bilateral plantar reflex was mute. The patient was shifted to ICU for further management. In ICU, point of care testing Arterial Blood Gas (ABG) analysis showed pH 7.06, $PaO_2$ 162mmHg on $FiO_2$ 0.4, $PaCo_2$ 60mmHg, Bicarbonates 13.8mmol/L, Sodium 136mmol/L, Potassium 1.6mmol/L, Chloride 110mmol/L with normal anion gap metabolic acidosis along with respiratory acidosis. Her random blood sugar was 346 mg/dl and urine ketones were negative. Fluid resuscitation was done in ICU using ringer lactate along with i.v. potassium chloride supplementation. The serum $K^+$ level was achieved more than 3.5mmol/L in next 24 hours. The patient's symptom improved drastically, she regained power of 5/5 with sustained neck holding for more than five seconds. The patient was extubated next day. Further investigations were performed to elucidate the etiology of hypokalemia (Table 1). In the absence of any history of gastrointestinal loss or diuretic use, the possibility of RTA was suspected as a probable cause of normal anion gap metabolic acidosis. Urinalysis revealed pH 6.0, positive urinary anion gap (59mmol/L) and Trans Tubular Potassium Gradient (TTKG) of 17, which further consolidated our diagnosis of distal RTA [3]. After extubation, the patient gave the history of dryness in eyes and mouth for last 1yr with occasional joint pains. After ruling out other causes of dryness, the possibility of SS was considered and the patient was further evaluated. Shimmers test result showed 4mm tear flow at 5min and antibodies to Sjogren's Syndrome A (SSA Ro), Sjogren's Syndrome B (SSB La) were strongly positive. The working diagnosis of




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**Table 1:** Investigations of the patient.

| | |
|---|---|
| Complete Blood Picture | Hb - 11.2g/dl, TLC - $17\times10^9$/L, Platelets - $312\times10^9$/L |
| Serum Electrolytes | Sodium - 136mmol/L, Potassium - 1.6mmol/L, Chloride - 110mmol/L |
| Serum Osmolality | 315mosm/kg |
| Urinalysis | pH - 6.0, urinary $Na^+$ - 35mmol/L, $K^+$ - 37mmol/L, urine chloride - 13mmol/L, urine osmolality - 420mosm/kg |
| Urinary Anion Gap | 59mmol/L (positive) |
| TTKG | 17 (Suggestive of renal loss of potassium) |
| Thyroid Function Tests | Within normal limits |
| Kidney Function Tests | Urea - 49mg/dl, Creatinine - 1.19mg/dl |
| Viral Markers | HIV - negative, HBsAg - negative, Anti-HCV – negative |
| Autoimmune Profile | ANA positive |
| | Anti-SS-a strongly positive |
| | Anti -SS-b strongly positive |
| Schrimmers Test | 4mm for 5min in both eyes (<10mm suggestive of dry eyes) |
| MRI Brain & Spine | No abnormalities detected |
| Nerve Conduction Study | Normal |
| Ultrasound Abdomen | No evidence of nephrocalcinosis |

Sjogren's Syndrome was made based on recently developed American College of Rheumatology/European League against Rheumatism (ACR-EULAR) classification criteria for primary Sjogren's syndrome [5]. The final diagnosis of hypokalemic paralysis with distal RTA with Sjogren's syndrome was made. At the time of discharge, the patient was started on oral potassium citrate, artificial tears and advised for oral hygiene. On further follow-up, she has been symptom free with no further episode of hypokalemia.

## Discussion

SS is characterized by chronic lymphocytic infiltration of salivary, lacrimal and parotid glands, leading to sicca symptoms (xerophthalmia and xerostomia). Renal involvement in pSS has a prevalence of around 9% although the first clinical manifestation with renal involvement is a rare phenomenon. Renal involvement in pSS is either due to tubulointerstitial involvement or less commonly due to glomerular involvement. Lymphocytic infiltration of the renal tubules by T cells, B cells, or plasma cells has been hypothesized behind the pathogenesis of renal involvement. The prevalence of distal RTA is reported from 5% to 70% in different studies [6,7]. The exact mechanism by which SS causes distal RTA is incompletely understood. The complete absence of H-ATPase pump in cortical collecting duct is one of the proposed theories described in some studies [8,9]. Another possible mechanism is carbonic anhydrase II inhibition caused by the high titre of autoantibodies, which leads to defective H+ secretion [10]. Hypokalemia is common in RTA, but severe hypokalemia mimicking periodic paralysis with respiratory failure is very rare.

Abbasi et al. reported a similar case of secondary hypokalemic paralysis presented with respiratory failure, in which the association for secondary hypokalemia is thyrotoxicosis [11].

Fujimoto T et al. reported a similar case in 27yrs old female presented with hypokalemic paralysis with respiratory involvement as the presentation of primary Sjogren's syndrome [12], where patient was not intubated but in our case patient developed hypercapnic respiratory failure for which she was intubated and improved drastically with potassium correction.

Renal involvement in Sjogren's syndrome is usually characterized by chronic interstitial nephritis, clinically by a variable but generally mild elevation in the serum creatinine, abnormalities in tubular function (distal renal tubular acidosis), nephrogenic diabetes insipidus, hypokalemia but glomerular involvement is less common in Sjogren's syndrome.

Management of renal involvement is based on the EULAR Sjogren's Syndrome Disease Activity Index (ESSDAI) [13]. Patients with mild ESSDAI scores are managed with correction of hypokalemia/ acidosis and patients with moderate to high ESSDAI scores are treated with glucocorticoids as a first line therapy, if not responding second line therapy includes immunosuppressant agents in moderate ESSDAI and rituximab, cyclophosphamide or plasma exchange in high ESSDAI category patients. Here our patient found to have mild disease activity according to renal domain of ESSDAI scoring, so managed with correction of hypokalemia/ acidosis, after which she improved. Long-term follow-up should monitor electrolytes, renal function and SS symptoms.

## Conclusion

Mild asymptomatic renal disease is common in SS, although Hypokalemic paralysis with renal tubular acidosis as a presenting complaint can be rarely associated with Sjogren's syndrome, which leads to respiratory failure. Proper evaluation, early treatment along with further follow up visit is essential to prevent life threatening respiratory failure associated with secondary hypokalemic paralysis. Our case report strengthens the existing literature regarding life threatening respiratory failure associated with secondary hypokalemic paralysis in-patient with Sjogren's syndrome.

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