Primary Sjögren’s Syndrome Presenting with Distal Renal Tubular Acidosis and Rhabdomyolysis

EBS Prakash*, ME Fernando**, Malathi Sathiyasekaran***, RM Bhoopathy****, JJ Jayanth+, J Samuel++

Abstract
Primary Sjögren’s syndrome (PSS) is rare in India. Clinically manifest renal disease in PSS is uncommon and is usually an autoimmune tubulointerstitial nephritis presenting with distal renal tubular acidosis (dRTA) or a urinary concentrating defect. Hypokalemic paralysis due to dRTA is rare but well documented in medical literature. Rhabdomyolysis as a consequence of hypokalemia in PSS is exceptional. We report a case of PSS with dRTA and rhabdomyolysis causing prolonged respiratory failure and quadriplegia.

INTRODUCTION
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rimary Sjögren syndrome is rare in India and is more common in females than in males.1 It was described in 1933 by Sjögren as a triad of rheumatoid arthritis, dry eyes and dry mouth. It is characterised by inflammation and autoimmune destruction of exocrine glands. Renal involvement occurs in about 30% of cases2 and is often an autoimmune tubulointerstitial nephritis causing distal renal tubular acidosis (dRTA) or a urinary concentrating defect. However, renal involvement is usually subclinical and clinically overt renal disease is rare.2 Hypokalemic paralysis due to dRTA seen in Sjögren’s syndrome is rarely reported from India3 though well documented in western literature.4-6 Rhabdomyolysis in primary Sjögren’s syndrome is very rare. We report a case of primary Sjögren’s syndrome with renal tubular acidosis and hypokalemia-induced rhabdomyolysis.

CASE REPORT
A 49 years old female was admitted with difficulty in breathing and inability to use all four limbs for seven days and history of weight loss and recurrent fever for three months. On examination she was conscious, oriented, febrile. Conjunctiva; was dry and sticky secretions were seen at the inner canthi. Blood pressure was 140/80 mmHg and pulse rate was 82/min. Respiratory rate was 33/min and respiration were shallow. She was unable to move her head and only eyelid blinking was present. Muscle power was 0/5 in all groups and deep tendon reflexes were absent but sensations were well preserved. There was no muscle wasting or tenderness. Plantars were not elicitable. Rest of systemic examination was unremarkable. She was intubated and given ventilator support.

Investigations showed TLC 5,800/mm³ with neutrophils 57%, lymphocytes 36%, monocytes 6%, and eosinophils 1%, haemoglobin 11.4 gms%, platelets 4,45,000/mm³, ESR 130mm/hr serum potassium 1.5 mEq/L, sodium 143 mEq/L, chloride 114 mEq/L, bicarbonate 14.6 mEq/L, blood sugar 71 mg/dL, urea 20 mg/dL, creatinine 1.1 mg/dL, creatinine kinase was 1430 IU/L [ref. 24-190 IU/L] and increased over the next few days to 11020 IU/L. Quatitative buffy coat examination for malaria and plasmodium LDH assay were negative. IgM ELISA and MAT for leptospirosis were negative. ECG showed prominent U waves. Chest X-ray and ultrasonogram of the abdomen were normal.

Urinary potassium excretion was high- 174 mEq/day (ref. 25-100 mEq/day). A positive urinary anion gap despite low serum bicarbonate established the diagnosis of renal tubular acidosis (RTA). Normal urinary bicarbonate excretion following correction of acidosis and urinary pH of 7.0 confirmed diagnosis of distal (Type 1) renal tubular acidosis (dRTA).

A diagnosis of Sjögren’s syndrome was considered.
A review of the patient’s history revealed dry eyes and dry mouth of 2 years’ duration. Schirmer’s test was consistent with Sjögren’s syndrome. ANA was 4.8 OD Index (ELISA) [ref. < 1.0 OD Index] and RA factor was 32 IU/ml [ref. < 10 IU/ml]. Anti SS-A (Ro) and SS-B (La) antibodies were positive. Anti U1 RNP and dsDNA were negative. Muscle biopsy showed rhabdomyolysis and myophagocytosis (Fig.1). A final diagnosis of primary Sjögren’s syndrome with distal renal tubular acidosis and hypokalemia induced rhabdomyolysis causing quadriplegia was made.

Pulse methylprednisolone 1 gm IV was given for three days. Patient transiently improved but later developed pneumonic consolidation in the left lower lobe. Broncho alveolar lavage and blood culture grew Klebsiella sensitive to amikacin and ciprofloxacin. Amikacin 250 mg q8hrs and ciprofloxacin 500 mg q 12 hrs were initiated. Prednisolone dose was reduced to 30 mg/day and stopped after 3 days. Urine culture showed no growth. CT scan of the abdomen and brain and CSF analysis were normal. There were no pressure sores. Fungal culture was negative.

On the 12th day of hospitalisation BP began to fall and inotropes were started. On the 14th day abnormal liver and renal parameters were noted and a diagnosis of MODS was made. Muscle power remained 0/5 and patient remained ventilator dependent. The patient succumbed to septic shock on the 16th day.

**DISCUSSION**

Sjögren’s syndrome classically presents with the sicca syndrome. Cases presenting with hypokalemic paralysis is rare (<2% of cases) but well documented. Almost all recover with correction of hypokalemia. Hypokalemia can cause rhabdomyolysis which can occasionally be severe and widespread causing prolonged and irreversible paralysis. Death can occur from respiratory paralysis and myoglobinuric renal failure.

This patient had hypokalemic paralysis and renal potassium wasting. Primary mineralocorticoid excess and hyperreninemia were excluded due to absence of hypertension and metabolic alkalosis. Hypokalemic hyperchloremic metabolic acidosis with a normal serum anion gap suggested renal tubular acidosis. Further investigations confirmed distal (type 1) renal tubular acidosis and Sjögren’s syndrome. There was no neurological recovery despite correction of hypokalemia and a progressive rise in creatinine kinase level was observed. This prompted a muscle biopsy which revealed rhabdomyolysis.

Rhabdomyolysis is frequently reported to occur from direct or ischemic muscle injury, seizures, strenuous exercise, viral or autoimmune polymyositis, snake bite, liquorice fibrates, statins, hypocalcemia, malignant hyperpyrexia and neuroleptic malignant syndrome. In these situations it is accompanied by hyperkalemia. A creatinine kinase level in excess of 10,000 U/L is common. Severe myoglobinuria often causes acute tubular necrosis. Hypokalemia *per se*, could cause rhabdomyolysis and in this instance a paradoxical combination of hypokalemia and elevated creatinine kinase will be seen.

Hypokalemia-induced rhabdomyolysis causing flaccid paralysis in Sjögren’s syndrome is exceptional. We report this case to highlight the fact that Sjögren’s syndrome could have a fatal outcome at presentation due to rhabdomyolysis and prolonged respiratory insufficiency.

**REFERENCES**