

Case Report

A case of Sjogren's syndrome presenting as hypokalaemia secondary to renal tubular acidosis

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ABSTRACT

Renal tubular acidosis (RTA) is a known but relatively rare presentation of Sjogren's syndrome. It requires a high level of clinical suspicion to be diagnosed. We report a 38-year-old female who presented with respiratory arrest that was later found to be caused by hypokalemia secondary to renal tubular acidosis. Further work up proved a diagnosis of Sjogren's syndrome. The patient improved with correction of electrolytes and hydroxychloroquine. This case highlights the importance of consideration of Sjogren's syndrome as a probable cause of RTA as renal complications of Sjogren's syndrome have a good prognosis if identified and treated promptly.

Keywords: Sjogren's syndrome, Renal tubular acidosis, Hypokalaemia, Respiratory arrest

INTRODUCTION

Sjogren's syndrome is an auto-immune, lymphoproliferative condition that causes inflammation of exocrine glands. Extra-glandular manifestations such as haematological, pulmonary and renal involvement are more common in the Asian population. Renal involvement is a well-recognized in primary Sjogren's syndrome.^{1,2} Here we present a case of distal renal tubular acidosis (RTA) secondary to Sjogren's syndrome that led to hypokalemic paralysis.

CASE REPORT

A 38-year-old female with no known co-morbidities presented with complaints of generalised weakness for 2 days and breathing difficulty since morning. On further questioning, her attenders reported a history of weakness that began in the lower limbs and gradually progressed to involve the trunk and upper limbs. She developed sudden onset difficulty breathing and was brought to the ER. On examination, the patient was unresponsive with a Glasgow coma scale (GCS) of 3/15 and was in a gasping state. Central nervous system examination showed reduced deep

tendon reflexes. Arterial blood gas (ABG) showed a pH of 7.07 and a bicarbonate of 8.1 mmol/l, suggestive of severe metabolic acidosis and her anion gap was normal (10 mmol/l). She was intubated in view of severe respiratory distress. Serum potassium was 1.9 mEq/l. Intravenous potassium correction was started and her power gradually improved. Urine routine was sent and in view of urinary pH of 7.0, a diagnosis of distal RTA was made.

Her magnetic resonance imaging (MRI) brain report also showed a parotid swelling and on further questioning, her attenders gave a history of pain in the parotid region for which she was on symptomatic treatment. The parotid swelling along with a history of fatigue and RTA led to a suspicion of Sjogren's syndrome and an antinuclear antibodies (ANA) profile was positive for anti-SSB antibodies which confirmed the diagnosis.

The patient was treated with T. hydroxychloroquine (HCQ) 200 mg HS, potassium citrate syrup (15 ml TDS) and tablet sodium bicarbonate (500 mg BD). Ophthalmology opinion was obtained and methylcellulose eyedrops were started in view of dryness in the right eye (Schirmer's test – 8 mm).

At the time of discharge, serum potassium was 3.7 mEq/l. Her sensorium and weakness had improved. She was discharged with T. HCQ 200 mg HS, potrate syrup and T. nodosis (1-0-1). She was followed up in the OPD after two weeks and was doing well, she was continued on T. HCQ once daily.

DISCUSSION

Sjogren's syndrome is an autoimmune disorder associated mainly with keratoconjunctivitis sicca and xerostomia. It has a global prevalence of 1%. It also affects other exocrine glands in the larynx (hoarseness), trachea (cough), skin (pruritus), and vagina (dyspareunia). Extra-glandular manifestations range from purpura, urticaria, arthritis, and Raynaud's phenomenon to severe complications such as renal disease, interstitial lung disease, B-cell non-Hodgkin lymphomas and peripheral neuropathy.^{3,4}

Data shows that patients were found to have symptoms for an average of 3.9 years before being diagnosed with Sjogren's syndrome and that systemic manifestations develop usually 10 years after the onset of dry eye symptoms, underscoring the importance of dry eye symptoms as an initial manifestation of the disease.⁵ A study by Komori et al showed that the diagnosis of Sjogren's syndrome was missed or delayed due to variable and non-specific symptomology, initial visits to ophthalmologist for dry eye that is not further investigated, and high seronegative disease incidence (anti-SSA and anti-SSB negative).⁶ Our patient had complaints of fatigue and reported a history of dry eyes only on probing, and was hence not diagnosed with Sjogren's syndrome prior to this admission. Parotid swelling was discovered incidentally on an MRI brain which was done in view of poor GCS.

Potassium is responsible for physiological processes such as nerve impulse conduction, maintaining cell membrane potential, muscle function and acid-base balance. While hypokalaemia is asymptomatic in most patients, it can cause muscle cramps or paralysis. Cardio-respiratory involvement can be life threatening as in our patient.⁷ Fatal complications of hypokalaemia are mainly due to disruption of cardiac electrical activity and paralysis of respiratory muscles.⁸ A study by Hess et al showed that 17% of patients with severe hypokalaemia (less than 2.5 mEq/l) died during hospitalisation.⁹

Tubulointerstitial nephritis (TIN) is the most common renal disease associated with Sjogren's syndrome, accounting for 85% of patients with renal manifestations. Other renal manifestations of Sjogren's syndrome include interstitial glomerulonephritis, nephrogenic diabetes insipidus, nephrolithiasis, tubular dysfunction, immunoglobulin G4-related disease and chronic kidney disease.¹⁰

Though the exact mechanism of RTA in Sjogren's syndrome is unknown, it has been proposed to be due to

absence of H (+)-ATPase in alpha intercalated cells of nephrons, preventing proton secretion. Hypokalaemia in RTA is due to the subsequent increase in distal potassium excretion to maintain electroneutrality.¹¹ Another mechanism for RTA in Sjogren's syndrome is the presence of antibodies against carbonic anhydrase II.¹² In this patient, a normal anion gap metabolic acidosis, alkaline urine and hypokalaemia hinted at a possible distal renal tubular acidosis. A study by Sandhya et al showed a 6.6% prevalence of RTA in patients with primary Sjogren's syndrome.¹³ Another European study showed a 5% prevalence of distal RTA in primary Sjogren's syndrome.¹⁴

Other causes of distal RTA include inherited mutations, drugs like NSAIDs, amphotericin B and lithium, sickle cell disease, tubulointerstitial diseases like chronic pyelonephritis, chronic interstitial nephritis, obstructive uropathy and renal transplant rejection, and hypergammaglobulinemic states like multiple myeloma. It can also be secondary to other autoimmune disease like systemic lupus erythematosus, rheumatoid arthritis, and systemic sclerosis. Sjogren's syndrome is the most common auto-immune disease associated with renal tubular acidosis.¹⁵ A study conducted in India showed that 34.8% of renal tubular acidosis cases were attributed to Sjogren's syndrome. However, this can vary based on geographic and demographic factors.¹⁶

Distal RTA can cause nephrocalcinosis through hypercalcinuria, reduced citrate excretion and reduced proton excretion, resulting in alkaline urine that allows calcium precipitation.¹⁷ Ultrasound was done and our patient had no evidence of nephrocalcinosis.

In our patient, initiation of bicarbonate and potassium correction lead marked symptomatic improvement. Parotid swelling, a positive Schirmer's test and the presence of anti-SSB antibodies confirmed the diagnosis of Sjogren's syndrome.

Renal involvement in Sjogren's syndrome usually has a good prognosis, but 10-20% progress to end stage chronic kidney disease, hence early detection and management can improve the quality of life in these patients.⁷

CONCLUSION

Sjogren's syndrome can sometimes present with predominant extra-glandular manifestations and is a differential to be considered for distal RTA. It can cause hypokalaemic paralysis which is life threatening and requires early and prompt management. Renal involvement in Sjogren's syndrome has a good prognosis. Hence, effective work up and management can be lifesaving.

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