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A CASE OF RENAL TUBULAR ACIDOSIS DUE TO SJOGREN's SYNDROME

Dr Basavesh Gongadashetti*	Dr D Y Patil Medical College, Nerul , Navi Mumbai *Corresponding Author
Dr Smita Patil	Dr D Y Patil Medical College, Nerul , Navi Mumbai
Dr Swetabh Roy	Dr D Y Patil Medical College, Nerul , Navi Mumbai
Dr N D Moulick	Dr D Y Patil Medical College, Nerul , Navi Mumbai
Dr Harsh Patel	Dr D Y Patil Medical College, Nerul , Navi Mumbai
Dr Sravanth Gandavarapu	Dr D Y Patil Medical College, Nerul , Navi Mumbai

ABSTRACT

Sjogren syndrome is a systemic autoimmune disorder commonly presenting with dryness involving the eyes and mouth due to inflammation and resultant pathology of the lacrimal and salivary glands. Renal tubular acidosis (RTA) is seen in one-third of the cases. RTA with hypokalemic paralysis as a presenting features of Sjogren syndrome is described in few case reports in literature. In this case report, we discuss a 36 year old female with distal renal tubular acidosis (RTA), who had experienced severe hypokalemic episode, the patient was eventually diagnosed with Sjogren's syndrome.

INTRODUCTION

Renal tubular acidosis (RTA) is characterized by renal tubular impairement in balancing physiologic acid base. It is often results from a defect in tubular transporters, which participate in the secretion or uptake of specific ions, due to congenital causes, exposure to nephrotoxic drugs, diuretic abuse, autoimmune disease or malignancy(e.g multiple myeloma).there are 3 major types of RTA: distal or type 1, proximal or type 2 and hyperkalemic or type 4. All three types of RTA are characterized by a positive urine anion gap, hyperchloremic non-anion gap metabolic acidosis, alkalotic or acidotic urine Ph and serum potassium derangements (hypo- or hyperkalemia).

Distal RTA (type 1 or classic RTA), which is the focus of this case report, can be further defined by an alkalotic urinary pH(>5.5) and profound hypokalemia(<3.0). Distal RTA can be a rare complication of Sjogren's syndrome in adloscents, although studies have demonstrated that approximately a greater proportion of adult patients develop defects in distal tubular acidification due to tubulointerstitial nephritis. In this report, we discuss a case of distal RTA secondary to Sjogren's syndrome and her management with potassium and alkali repletion.

CASE STUDY

A 36year old female presented to casualty with weakness in all 4 limbs and also with complaints of fever and multiple episodes of vomiting since 5 days. She had experienced cramping of bilateral lower limb a day back followed by weakness in all 4 limbs since morning. She gives history of multiple hospital admissions for generalised weakness and acute febrile illness. There was no family history of autoimmune diseases.

Except for bradycardia (heart rate of 56bpm),her vital signs were within normal limits and on CNS examination she had hypotonia and power was 1/5 in all four limbs, rest of CNS and other systemic examination was unremarkable. Results of initial lab tests were sodium- 132, potassium- 1.8, chloride- 119, bicarb- 8.9 and venous blood gas pH- 7.3. Urine sodium- 36, urine potassium- 1.8, urine Chloride- 46 with anion gap of 4.61. Urinalysis showed pH of 6.5 without blood or protein. The ultrasonography of kidney did not show nephrolithiasis or any abnormalities. ECG showed sinus bradycardia and U waves.

She was hydrated properly and was given potassium chloride and sodium bicarbonate, which corrected both her hypokalemia and hyperkalemic non-anion gap metabolic acidosis. She also had gain in her power.

The serum potassium started decreasing with subsequent monitoring even with oral potassium chloride. Repeated Urine spot potassium was 29 and venous blood gas was suggestive of non-anion gap metabolic acidosis. Urinalysis showed pH of >5.5 and positive urinary anion gap. She was diagnosed distal RTA with these reports.

Her autoimmune panel was positive for antinuclear antibodies (ANA 1:1000) and anti-Ro/SSA and anti-La/SSB (200 and 33.9 respectively). A presumptive diagnosis of Sjogren's syndrome was made. She was started on tablet hydroxychloroquine 400mg OD and was advised to follow up with nephrology and rheumatology in outpatient setting.

DISCUSSION

Sjogren's syndrome is a chronic, slowly progressing autoimmune disease characterized by lymphocytic infiltration of the exocrine glands resulting in xerostomia and dry eyes. The syndrome has unique features since it presents with a wide clinical spectrum from organ-specific autoimmune exocrinopathy to systemic disease. The disease can present as an entity alone or in association with other autoimmune diseases.

The majority of patients have symptoms related to impaired lacrimal and salivary gland function. The disease evolution is slow and in the majority of patients runs a benign course.

The principle oral symptom is dryness(xerostomia). Patients report difficulty in swallowing dry food, a burning mouth sensation, an increase in dental caries. Physical examination shows a dry, erythematous, sticky oral mucosa. Enlargement of parotid or other salivary glands occurs in two-thirds of patients with primary Sjogren's syndrome but is uncommon in those in association with rheumatoid arthritis.

Ocular involvement is the other major manifestation of

Sjogren's syndrome. Patients usually describe a sandy or gritty feeling under the eyelids. Other symptoms include burning, accumulation of secretions in thick strands at inner canthi, decreased tearing, redness, itching, eye fatigue, and increased photosensitivity.

Extraglandular (systemic) manifestations are seen in one-third of patients which include cardiac involvement leading to prolonged QT, pulmonary disease, neurologic involvement and tubulointerstitial disease (e.g. tubulointerstitial nephritis, RTA, Fanconi syndrome, and glomerulonephritis). The true mechanism behind RTA is not well known. Available data point to reduced hydrogen electrolyte secretions consequently due to immune-mediated injury of the hydrogen-ATPase pump of the alpha-intercalated cells or autoantibody directed against carbonic anhydrase II.

CONCLUSION:

The patient in the present case was treated for hypokalemia and based on her medical history, examination findings, and investigations, she was diagnosed as Sjogren's syndrome. Although Sjogren's syndrome is characteristically associated with symptoms of dry eyes and mouth, extraglandular renal manifestations are not uncommon presentations. The most common renal manifestation of Sjogren's syndrome is tubulointerstitial nephritis, which can be manifested as renal tubular acidosis.

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