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Case Report

Overlap syndrome of systemic lupus erythematosus and Sjögren's syndrome presenting with distal renal tubular acidosis

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ABSTRACT

Distal renal tubular acidosis (dRTA) is a rare but clinically significant renal manifestation of autoimmune diseases, particularly overlap syndromes involving systemic lupus erythematosus (SLE) and Sjögren's syndrome. We report the case of a 33-years man who presented with acute limb weakness secondary to severe hypokalaemia. Her biochemical profile revealed normal anion gap metabolic acidosis and elevated urine pH, consistent with dRTA. Immunological testing and histopathology confirmed the diagnosis of overlap syndrome involving SLE and Sjögren's syndrome. She was successfully managed with potassium supplementation, immunomodulatory therapy, and symptomatic treatment. This case highlights the importance of considering autoimmune aetiologies in patients with unexplained hypokalaemia and metabolic acidosis, particularly in the presence of sicca symptoms.

Keywords: Distal renal tubular acidosis, Sjögren's syndrome, Systemic lupus erythematosus

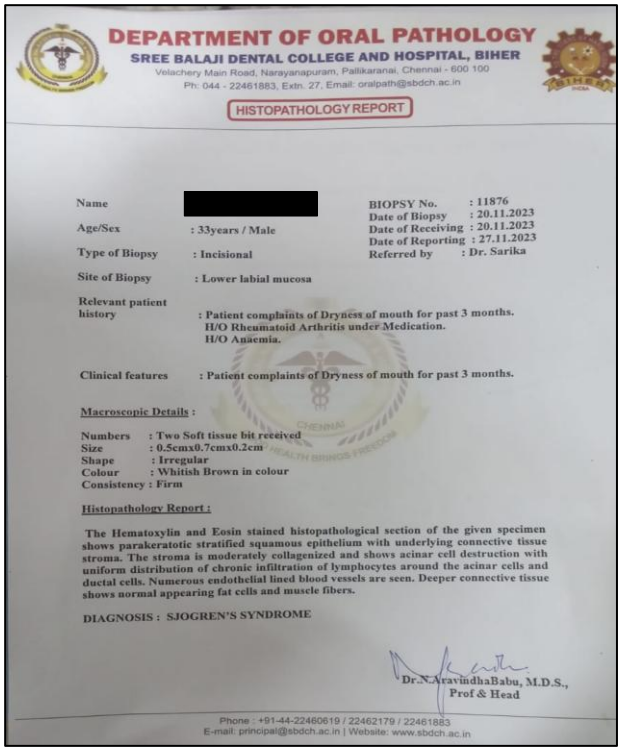
INTRODUCTION

Sjögren's syndrome is a systemic autoimmune disease primarily affecting the exocrine glands, leading to classical symptoms such as dry eyes (keratoconjunctivitis sicca) and dry mouth (xerostomia).[1] In some cases, it occurs in conjunction with other autoimmune conditions, most notably systemic lupus erythematosus (SLE), resulting in an overlap syndrome that combines features of both diseases.[2] This overlap expands the spectrum of clinical manifestations, often including renal involvement, particularly in the form of distal renal tubular acidosis (dRTA). dRTA is characterised by impaired hydrogen ion secretion in the distal nephron, leading to normal anion gap metabolic acidosis, hypokalaemia, and inappropriately high urine pH, despite preserved glomerular function.[3] Despite being uncommon, distal RTA can represent a clinically significant renal manifestation of autoimmune overlap syndromes and must be considered in unexplained cases of muscle weakness or electrolyte imbalance. In autoimmune overlap syndromes, dRTA results from immune-mediated injury to renal tubular cells. Clinically, it may present with muscle weakness, fatigue, or even acute flaccid paralysis due to profound hypokalaemia, along with sicca symptoms attributable to Sjögren's syndrome.[4] Here, we report a case of a 33-years male who presented with acute limb weakness and was subsequently diagnosed with overlap syndrome of SLE with Sjögren's syndrome and dRTA, highlighting the importance of considering dRTA in patients with unexplained hypokalaemia and autoimmune features.

CASE REPORT

A 33-years male patient presented with acute, progressive weakness of both upper and lower limbs, which developed gradually and resulted in difficulty getting out of bed. There was no history of trauma, fever, slurred speech, giddiness, urinary or bowel incontinence, or chronic drug use. She had experienced a similar episode of lower limb

weakness three months prior, which resolved completely after treatment at a local hospital. On examination, the patient was conscious, oriented, and afebrile. Vital signs were stable, with a blood pressure of 110/80 mmHg, pulse rate of 86 bpm, and oxygen saturation of 98% on room air. Neurological examination revealed hypotonia and diminished deep tendon reflexes in all four limbs, while cranial nerve function and sensory examinations were normal.



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HISTOPATHOLOGY REPORT

Name [redacted]
Age/Sex : 33years / Male
Type of Biopsy : Incisional
Site of Biopsy : Lower labial mucosa
Relevant patient history : Patient complaints of Dryness of mouth for past 3 months. H/O Rheumatoid Arthritis under Medication. H/O Anaemia.
Clinical features : Patient complaints of Dryness of mouth for past 3 months.

BIOPSY No. : 11876
Date of Biopsy : 20.11.2023
Date of Receiving : 20.11.2023
Date of Reporting : 27.11.2023
Referred by : Dr. Sarika

Macroscopic Details :
Numbers : Two Soft tissue bit received
Size : 0.5cmx0.7cmx0.2cm
Shape : Irregular
Colour : Whitish Brown in colour
Consistency : Firm

Histopathology Report :
The Hematoxylin and Eosin stained histopathological section of the given specimen shows parakeratotic stratified squamous epithelium with underlying connective tissue stroma. The stroma is moderately collagenized and shows acinar cell destruction with uniform distribution of chronic infiltration of lymphocytes around the acinar cells and ductal cells. Numerous endothelial lined blood vessels are seen. Deeper connective tissue shows normal appearing fat cells and muscle fibers.

DIAGNOSIS : SJOGREN'S SYNDROME

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Figure 1: Histopathological report.

Investigations

Initial laboratory tests revealed severe hypokalaemia (serum potassium 1.44 mmol/l), normal sodium (Na^+ 145.3 mmol/l), and hyperchloremia (Cl^- 115 mmol/l), with normal renal function (serum urea 16 mg/dl and creatinine 0.8 mg/dl). Arterial blood gas (ABG) analysis revealed metabolic acidosis with a pH of 7.121, HCO_3^-12.2 mmol/l, PCO_2 30 mmHg, and a normal anion gap of 10.2, consistent with normal anion gap hyperchloremic metabolic acidosis. CNS imaging, including an MRI of the brain and dorsal spine, revealed no abnormalities. Urine analysis showed a high pH (8.0), urine osmolality of 339.7 mOsm/kg, urinary sodium 146.0 mmol/l, urinary potassium 32 mmol/l, and urine chloride 63 mg/dl. The urinary anion gap was calculated as positive ($Na^+ + K^+ - Cl^- = 178 - 63 = 115$ mmol/l). The urine protein-creatinine ratio was 92 mg/dl.

Immunological and pathological investigations

The Anti-nuclear antibody (ANA) profile was positive for extractable nuclear antigens (U1-70, Sm, snRNP, and SS-A), which is reported in connective tissue disease, most consistent with Sjögren's syndrome with possible SLE overlap. An incisional biopsy of the lower labial mucosa revealed stratified squamous epithelium with an underlying moderately collagenized connective tissue stroma. Microscopic examination revealed acinar cell destruction with lymphocytic infiltration around the minor salivary gland ducts and acini, consistent with chronic sialadenitis. Numerous dilated blood vessels, fat cells, and muscle fibres were also observed. These findings are characteristic of Sjögren's syndrome.

Treatment

The patient received potassium supplementation (Syrup Kesol 15 ml three times daily with fruit juice), dietary modifications (coconut water, bananas, dry fruits), hydroxychloroquine 200 mg on alternate days, refresh tear eye drops, and lacrimal eye ointment.

DISCUSSION

SLE and Sjögren's syndrome are autoimmune diseases that may coexist, resulting in an overlap syndrome characterised by combined clinical features, lymphocytic infiltration of exocrine glands, causing sicca symptoms, but also involves extra-glandular organs, including the kidneys, where dRTA is a notable complication.[5] SLE, a multisystem autoimmune disease, frequently produces diverse autoantibodies and may overlap with Sjögren's syndrome, presenting with glandular and systemic involvement.[6] dRTA arises due to immune-mediated damage to the distal tubules, impairing hydrogen ion secretion and leading to metabolic acidosis with hypokalaemia and alkaline urine. These biochemical abnormalities, including hypokalaemia, high urine pH, and a positive urinary anion gap, are diagnostic features of dRTA that can occur without classic sicca symptoms, underscoring the importance of a thorough evaluation in patients with unexplained metabolic disturbances.[7] This corresponds with the findings in our case, where high urine pH, hypokalaemia, and a positive urinary anion gap confirmed dRTA.

The diagnosis of SLE–Sjögren's overlap syndrome with dRTA requires the integration of clinical, serological, and histopathological data. Positive ANA profiles and extractable nuclear antigen (ENA) testing support the diagnosis of autoimmune diseases. Histopathological confirmation by lip biopsy remains the gold standard for diagnosing Sjögren's syndrome.[8] Additionally, neuroimaging may be employed to rule out neurological causes of weakness in patients presenting with hypokalaemia paralysis. The long-term prognosis in such cases often depends on timely diagnosis and continued monitoring of renal and systemic involvement. Management targets both the maintenance of chemical balance and the underlying autoimmune process. Immediate potassium replacement is critical for preventing cardiac arrhythmias and paralysis. Long-term

immunomodulation with agents such as hydroxychloroquine and corticosteroids is recommended to control systemic disease and prevent progression.[1,9] Supportive treatments for glandular symptoms and close monitoring for renal function deterioration are also essential.[9]

The overlap of SLE and Sjögren's syndrome should alert clinicians to the possibility of renal tubular involvement, and a thorough workup including serological and pathological evaluation is needed. The findings are consistent with those of previous reports by Hara et al and Ram et al highlighting the need for further research, clinical guidelines, a high index of suspicion, and a multidisciplinary approach to enhance patient outcomes.[11]

CONCLUSION

This case demonstrates that distal renal tubular acidosis can be an initial manifestation of autoimmune overlap syndromes such as SLE and Sjögren's syndrome. Early recognition and targeted treatment led to complete clinical recovery and electrolyte correction, with no recurrence. This outcome underscores the importance of considering autoimmune causes of hypokalaemia paralysis and highlights the role of timely, multidisciplinary management in achieving positive outcomes. Routine autoimmune screening should be considered in patients presenting with hypokalaemia paralysis, even in the absence of full-blown sicca symptoms.

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