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Case Report

Presentation of primary Sjogren's syndrome with livedo reticularis and hypokalemia: A case report

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Introduction

Sjogren's syndrome (SS) is a chronic, systemic, autoimmune disease that affects exocrine glands, leading to sicca symptoms. It can also cause extra-glandular manifestations and has a significantly increased risk of lymphoma development [1]. SS can be classified as primary or secondary based on the presence of other connective tissue disorders. The prevalence of SS is 0.1-4.8% in different populations and it primarily affects middle-aged women with a female-to-male ratio of 9:1 [2,3].

Case Report

A 36-year-old female presented to the National Hospital, Kandy with a net-like pigmented rash on the bilateral anteromedial aspect of her legs, suggestive of livedo reticularis, which had been present for one month (Figure 1). The patient did not have any previous skin rashes, exposure to heat, or sources of skin irritation. In the past, she had mentioned experiencing sporadic weakness in her lower limbs, which improved with potassium supplements. Additionally, she had been experiencing dry mouth, dry eyes, and dyspareunia for the past four years, but had no history of thyroid or connective tissue disorders.

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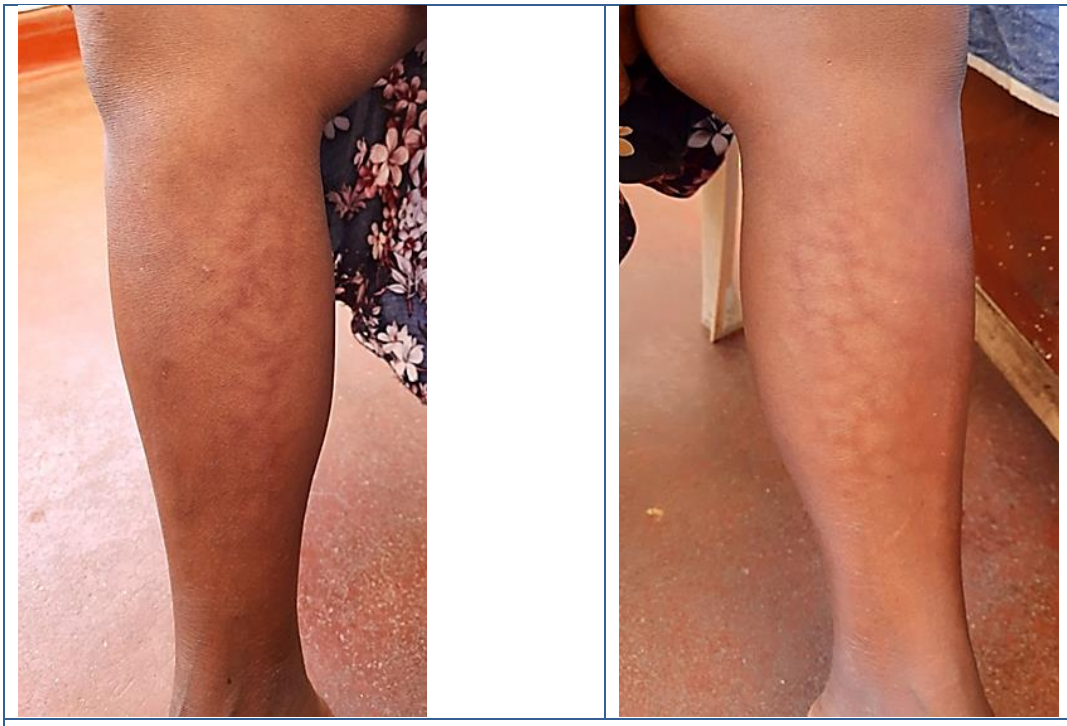

Figure 1: Livedo reticularis over bilateral lower limbs

During her last admission, her serum potassium was 1.7 mmol/L and her ABG showed hyperchloraemic metabolic acidosis with a normal anion gap (serum anion gap -12.2). She had normal serum calcium levels (2.16 mmol/L), and normal renal and liver profiles. She had a high urinary anion gap (24) with a urine pH of 7.62 and evidence of nephrocalcinosis (Figure 2), suggestive of distal renal tubular acidosis.

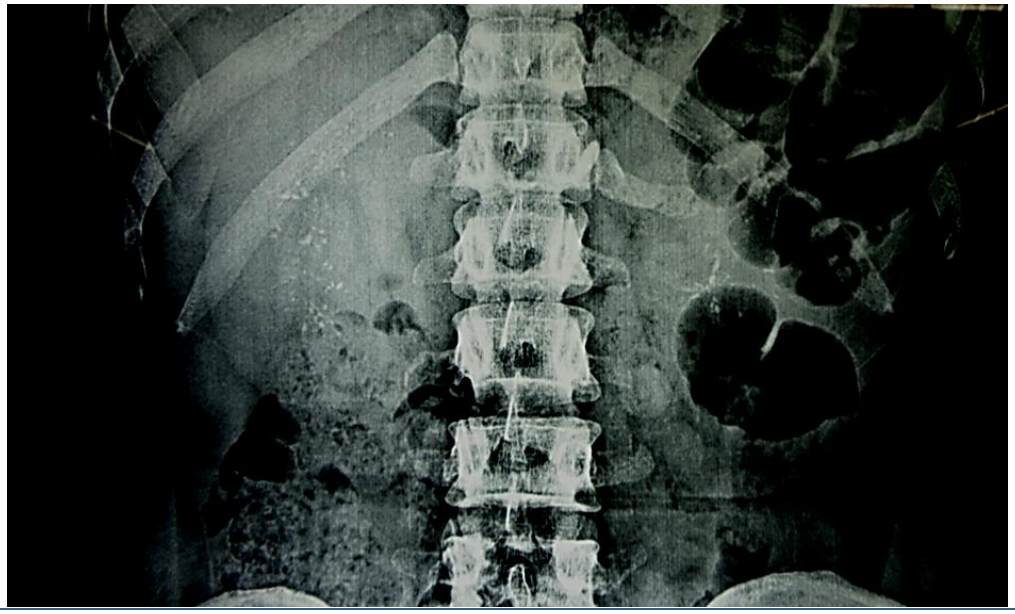

Figure 2: X-ray KUB showing bilateral nephrocalcinosis

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Her autoantibody screening was positive for ANA (fine speckled pattern titre >1:80), Anti-Ro, and Anti-La antibodies. She also had positive TPO antibodies and ultrasound evidence of thyroiditis but was clinically and biochemically euthyroid. A biopsy of her labial gland showed collections of mucinous minor salivary glands infiltrated by a lymphoplasmacytic infiltrate and lymphoid follicle formation with a Focus score >1, consistent with chronic sialadenitis and a diagnosis of Sjogren's syndrome (Figure 3).

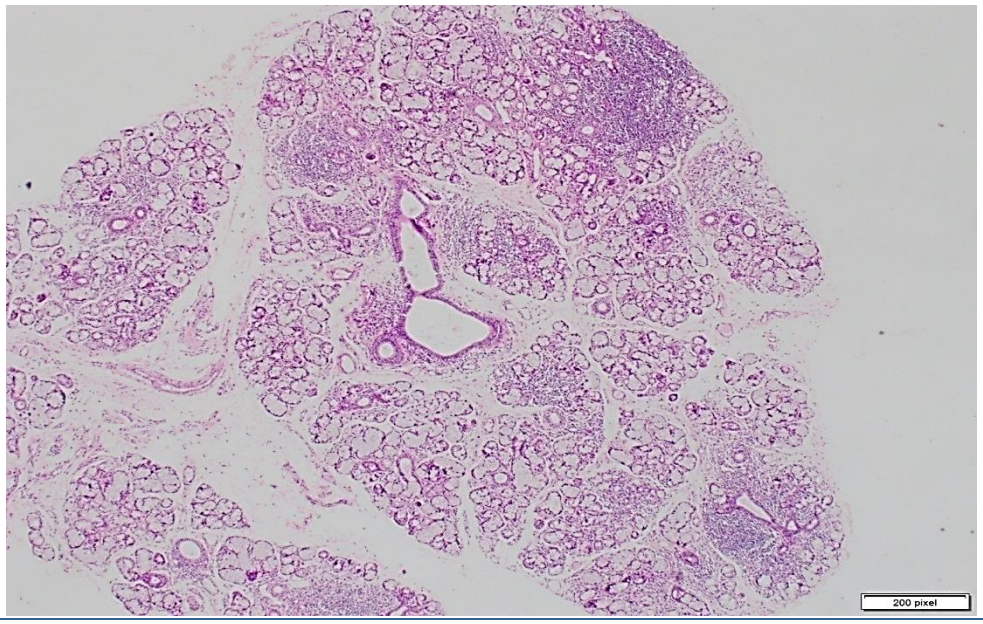

Figure 3: Labial gland biopsy showing mucinous minor salivary glands infiltrated by a lymphoplasmacytic infiltrate and lymphoid follicle formation

Based on clinical features and investigations, we made a final diagnosis of primary Sjogren's syndrome with distal renal tubular acidosis, livedo reticularis, and associated autoimmune thyroiditis. The patient is being closely monitored for associated conditions or complications and is being treated with potassium citrate and symptomatic treatments.

Discussion

Sjögren's Syndrome (SS) is a systemic autoimmune disease affecting the exocrine glands. The diagnosis of the syndrome is typically based on biopsy results of minor salivary glands and/or lacrimal glands, a physical examination of the oral cavity and eyes and the presence of autoantibodies. Distal renal tubular acidosis (dRTA) is a rare condition reported in less than 2% of SS patients [4]. Skin involvement is relatively common in SS, including eyelid dermatitis, xerosis, and cutaneous vasculitis [5]. Other non-vasculitic skin manifestations, such as livedo reticularis, annular erythema, erythema nodosum, and localized nodular cutaneous amyloidosis, have also been reported [6].

Our patient experienced sicca symptoms for 4 years before seeking medical attention. Her first presentation was with hypokalaemic paralysis possibly caused by dRTA. At the time, she also had evidence of nephrocalcinosis but normal renal function. She showed

a positive response to potassium supplements and potassium citrate. The patient's diagnosis was delayed as she defaulted on follow-up appointments due to financial difficulties. Her next presentation was with livedo reticularis but there was no evidence of connective tissue or autoimmune disorders.

According to the 2002 American-European Consensus Group (AECG) criteria, the diagnosis of primary SS should be based on either four of the following criteria, including histopathology or autoantibodies, or three of the four objective criteria in the absence of other connective tissue diseases: (1) ocular symptoms lasting more than 3 months, (2) oral symptoms lasting more than three months, (3) ocular signs (such as Schirmer's test or vital dye staining), (4) histopathology (focal lymphocytic sialadenitis in more than 1 focus per $4mm^2$), (5) oral signs, and (6) the presence of anti-Ro/SSA or anti-La/SSB autoantibodies [1].

In our patient's case, four of the above criteria were fulfilled, and the diagnosis of primary SS with dRTA and associated autoimmune thyroiditis was confirmed. Further autoantibody panels could not be conducted due to financial constraints. However, the patient did not display any clinically significant features of other connective tissue disorders.

Conclusion

Patients with SS may present for the first time with hypokalaemia due to dRTA or with the involvement of extra glandular organs. An accurate diagnosis of SS is crucial for proper management as it allows for monitoring and recognition of other manifestations that may be amenable to immunomodulatory therapy or for the detection of overlapped autoimmune diseases and the development of lymphoma. Clinicians should actively look for systemic complications of SS and tailor their management approach to the individual patient's presentation. Patients and their caregivers should be educated on lifestyle modifications for effective management of the disease, which may require a multidisciplinary approach in case of new symptoms or organ involvement.

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