Case Report

Autoimmune thyroiditis associated with primary Sjögren's syndrome complicated with distal renal tubular acidosis and hypokalaemic paralysis: A case report

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Abstract

Primary Sjögren's syndrome (pSS) is known to present with distal renal tubular acidosis (dRTA), which can manifest as hypokalaemic paralysis. The main involvement of the kidneys in pSS is tubulointerstitial nephritis. Furthermore, pSS is an autoimmune rheumatological condition that rarely co-exists with autoimmune thyroiditis.
We report a 42-year-old hypothyroid female, who complained of a one-day history of severe proximal muscle weakness in all four limbs and a two-month history of left ankle joint pain and swelling. Her serum potassium level was 1.7 mEq/L, with biochemical evidence of normal anion gap metabolic acidosis with positive urinary anion gap suggesting dRTA. As she complained of gritty eyes and recurrent stomatitis a diagnosis of Sjögren's syndrome was sought after. Her serology revealed a high rheumatoid factor titre of 378 (<20) IU/mL with negative anti-cyclic citrullinated peptide levels (<0.5 IU/mL). Positive antinuclear antibodies (1:640), anti-Sjögren's syndrome-related antigen A (SS-A) antibodies and anti-Ro-52 antibodies in the absence of other extractable nuclear antigen antibodies suggested pSS. This was confirmed with labial biopsy which revealed focal lymphocytic infiltration. Magnetic resonance imaging revealed the "salt and pepper" appearance of bilateral parotid glands with glandular atrophy. Even though she was clinically and biochemically euthyroid, given the background of thyroid dysfunction, we investigated for autoimmune thyroiditis (AIT) which was confirmed with imaging and a high anti-thyroid peroxidase antibody titre 532.71 IU/mL (<5.61). Her functional status markedly improved following the correction of serum potassium values. She was started on long term hydroxycholoroquine and oral potassium citrate.
This is a rare case of AIT associated with pSS complicated with dRTA presenting with hypokalaemic paralysis. This case illustrates the importance of screening for pSS among patients with dRTA and the rare coexistence of pSS and AIT.

Keywords: Primary Sjögren's syndrome, distal renal tubular acidosis, Autoimmune thyroiditis

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Introduction

Primary Sjögren's Syndrome (pSS) is known to present with distal renal tubular acidosis (dRTA), which could manifest as hypokalaemic paralysis. The main involvement of kidneys in pSS is tubulointerstitial nephritis. Furthermore, pSS is an autoimmune rheumatological condition that rarely co-exists with autoimmune thyroiditis [1].

Case report

We report a 42-year-old female patient with hypothyroidism, who presented with a one-day history of severe flaccid proximal weakness in a background of left ankle joint pain and swelling for six weeks. One day prior to the weakness she had taken her afternoon meal, following which she had developed the weakness.

Initial examination revealed lower limbs to be bilaterally flaccid with a proximal power of 2/5 (muscle movement with gravity elimitated) and a distal power of 4/5 (muscle movement against gravity and resistance). Upper limb examination revealed normal tone with a proximal power of 2/5 and a distal power of 4/5, bilaterally. All tendon reflexes were absent. Initial blood gas analysis revealed hyperchloraemic metabolic acidosis with a normal anion gap (pH=7.29, Na^+=133.3 mmol/L, K^+=1.42 mmol/L, Cl^-=113.1 mmol/L, HCO_3^-14.2 mmol/L, anion gap=7.4 mmol/L).

The serum potassium level was 1.7 mEq/L and the urinary pH was 7.2 (high). The acid loading test with ammonium chloride revealed a positive urinary anion gap and persistently high urinary pH of 7.1 suggestive of dRTA. She was resuscitated with intravenous potassium chloride and, on day three, her reflexes returned to normal and the power of the limbs improved.

As she complained of gritty eyes and recurrent stomatitis for six months a provisional diagnosis of Sjogren's syndrome was made.

Her serology revealed a high rheumatoid factor titre (378 (<20) IU/mL), positive antinuclear antibodies (1:640), positive anti-Sjögren's syndrome related antigen A (SS-A) antibodies and positive anti-Ro-52 antibodies with no other extractable nuclear antigen antibodies. This confirmed the diagnosis of pSS. The diagnosis was further established with labial biopsy which revealed foci of aggregates of lymphocytes and periductular lymphocyte infiltration (Figure 1A). A magnetic resonance imaging revealed the "salt and pepper" appearance of bilateral parotid glands with glandular atrophy (Figure 1B).

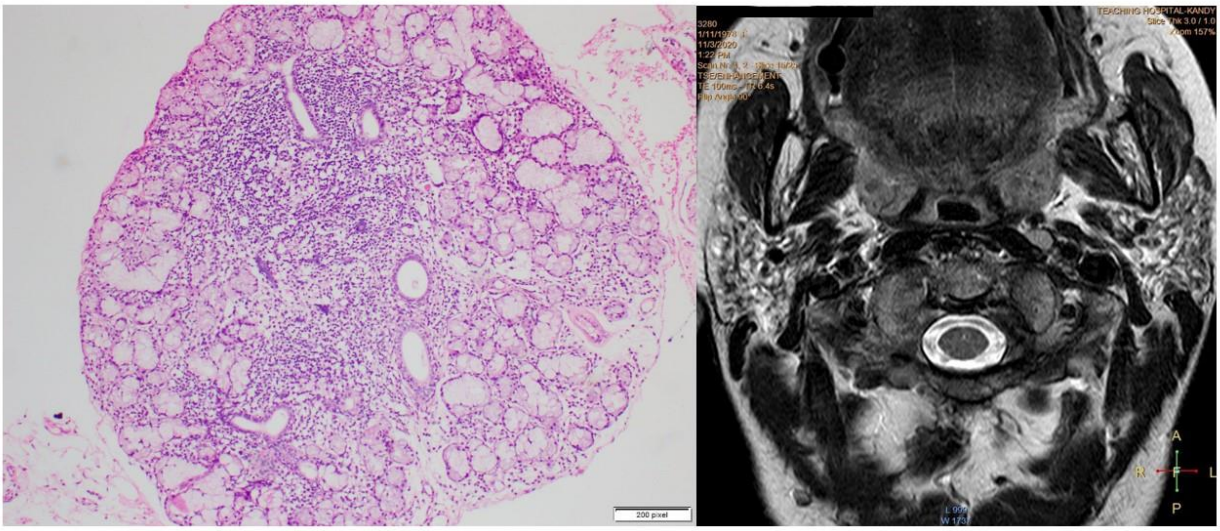

Figure 1A : Salivary gland of the patient showing foci of aggregates of lymphocytes, perpendicular lymphocyte infiltration without noticeable plasma cells; 1B : "salt and pepper" appearance of bilateral parotids with mild glandular atrophy.

Even though she was clinically and biochemically euthyroid, given the background of thyroid dysfunction, we investigated for autoimmune thyroiditis (AIT) which was confirmed with imaging and a high anti-thyroid peroxidase antibody titre [532.71 IU/mL (<5.61)]. Her functional status markedly improved following correction of serum potassium values. She was started on long term hydroxycholoroquine, oral potassium citrate and thyroxine.

Secondary causes of Sjögren's syndrome were ruled out by the absence of systemic symptoms to suggest other autoimmune diseases and the low levels of anti-cyclic citrullinated peptide (<0.5IU/ml), anti-Scl 70 antibodies,

anti dsDNA and normal complement levels [C3=121.2 (90-180), C4=44.9(10-40)].

Discussion

Primary Sjogren's syndrome (pSS) is a multisystemic autoimmune disease with predominant sicca symptoms. Cell-mediated immunity involving the exocrine glands plays a major role in the development of symptoms [1].

Our patient presented with sicca symptoms, arthralgia and severe muscle weakness. Absence of anti-CCP antibodies and high titres of SS-A, SS-B helped to differentiate pSS from secondary Sjogren's syndrome caused by rheumatoid arthritis.

Poor oral hygiene revealed impaired exocrine functions of salivary glands and further imaging showed the characteristic "salt and pepper" appearance and glandular atrophy of the parotid gland. Labial biopsy revealed focal lymphocytic infiltration. Recent evidence revealed plasma cells play a major role in autoimmunity by producing IgG that express CD-138 in the salivary glands which correlates with focus score in pSS [2].

Marked muscle weakness and hypokalaemia guided us for the evaluation of dRTA. Prospective cohort study by Ankit jain *et al*., revealed 50% of pSS patients had renal involvement in the form of renal tubular acidosis (RTA). The most common form of RTA was distal RTA (dRTA) presenting with hypokalemic paralysis [3]. dRTA is a renal potassium wasting disorder characterized by hyperchloraemic normal anion gap metabolic acidosis with persistent alkaline and positive anion gap urine.

History of hypothyroidism raised the possibility of autoimmune thyroiditis (AIT) and high titres of anti-TPO antibodies confirmed this diagnosis. Poly-autoimmunity with coexistence of AIT and pSS has been documented. Urticaria and lymphadenopathy has been documented in patients with AIT-pSS which was not evident in our patient. AIT and pSS are nosologically different autoimmune disorders although they share a common pathophysiological mechanism [4].

Concomitant immunosuppressant and corticosteroid therapy improve the renal functions of patients who are having rapidly declining renal functions [5]. Patients with stable renal functions curative therapy is symptomatic with potassium supplements, preferably potassium citrate.

Conclusion

This is a rare case of AIT associated with pSS complicated with dRTA presenting with hypokalaemic paralysis. This case illustrates the importance of screening for pSS among patients with dRTA and the rare coexistence of pSS and AIT.

Consent for publication

Informed written consent was obtained from the patient and her mother for publication of this case report.

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