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Hypokalemicquadriparesis as the Presenting Manifestation of Sjogrenssydrome with Autoimmune Thyroid Disease (Aitd)

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ABSTRACT
Sjogrens syndrome is an autoimmune disease which mainly affects the exocrine glands and is characterised by sicca symptoms. Renal involvement can be glomerular or involvement of the tubulointerstitial compartment. Overlap between various autoimmune diseases is known. Sjogrens can likewise be associated with autoimmune thyroid disease. Here we describe the case of a 45 year old lady who presented with hypokalemicquadriparesis and was subsequently found to have both Sjogrens syndrome and autoimmune hypothyroidism She recovered completely with treatment.

Key words : Sjogrens syndrome, distal RTA, Autoimmune thyroid disorders

I. INTRODUCTION

Sjogren syndrome is a connective tissue disease which typically presents with involvement of eyes and lacrimal glands. It is characterised by T lymphocyte mediated destruction of peri ductal epithelial cells. Renal tubular acidosis (RTA) is seen in 9% of cases with Sjogrens syndrome; distal RTA being the most common.[1] Severe hypokalemia in Sjogrens as the presenting manifestation without any other obvious symptom is rare. Hypokalemia is also observed as part of autoimmune thyroid disease (AITD) and is more commonly seen in Graves disease. Here we present a lady who presented with hypokalemicquadriparesis and was diagnosed to have both autoimmune hypothyroidism and Sjogrens syndrome .

II. CASE REPORT

A 45 old lady with history of untreated hypothyroidism, presented with sudden onset gradually progressive flaccid quadriparesis initially involving both lower limbs and later progressing to the upper limbs. There was no history suggestive of sensory, cranial nerve, autonomic, bowel or bladder involvement. There was no history of fever, recent immunization, diarrhoea or vomiting preceding the event. At time of admission ,she was conscious and oriented. Neurological examination was remarkable for bilateral lower limb power 2/5 and upper limb power 3/5 with decreased tone and absent reflexes. Cranial nerves and sensory system were normal. Mouth appeared dry. Initial laboratory values shown in Table 1 was significant for severe hypokalemia (S. potassium 1.8meq/L), Further evaluation showed normal anion gap metabolic acidosis with a positive urinary anion gap and increased potassium creatinine ratio indicating renal potassium loss. Other electrolyte levels were within normal limits. The investigation reports were suggestive of renal tubular acidosis and hence fractional excretion of bicarbonate was done which was low and a diagnosis of distal renal tubular acidosis (dRTA) was made. In view of of age and gender, a diagnosis of Sjogrens syndrome was considered as etiology of dRTA. On reassessing the history, patient apparently had been having symptoms of dry mouth which she had attributed to warm climate. She had no symptoms of decreased lacrimation or dry skin. ANA and autoimmune antibody profile showed strong positivity for anti Ro/SSA and anti LA/SSB. Lip biopsy done showed lymphocytic infiltration consistent with the diagnosis of Sjogrens syndrome. She was also diagnosed to have autoimmune hypothyroidism in view of elevated thyroid stimulating hormone and thyroid peroxidase antibody [Anti TPO].

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Table.1 below showing the initial blood investigations

Table 1

Test name	Results
Hemoglobin	11gm/dl
Total count	9480cells/microlitre
ESR	80mm/hour
S. Sodium	134mEq/l
S. Potassium	1.82mEq /l
S. Creatinine	0.8mg/dl
S . Bicarbonate	8.35mEq/L
S chloride	120mEq/L
S calcium	8.2mg/dl
ABG	pH 7.1
Anion gap	7.47
TSH	15.96mIU/L
FREE T3	3.3 p mol/L
FREE T4	9.5 p mol/L
Anti TPO	367.1 IU/ml
Urine ph	6.5
Urine spot potassium	14mEq/l
Potassium creatinine ratio	61.1
Urine creatinine spot	22.9 mg/l
Albumine	3.8g/dl

Biopsy picture shown below

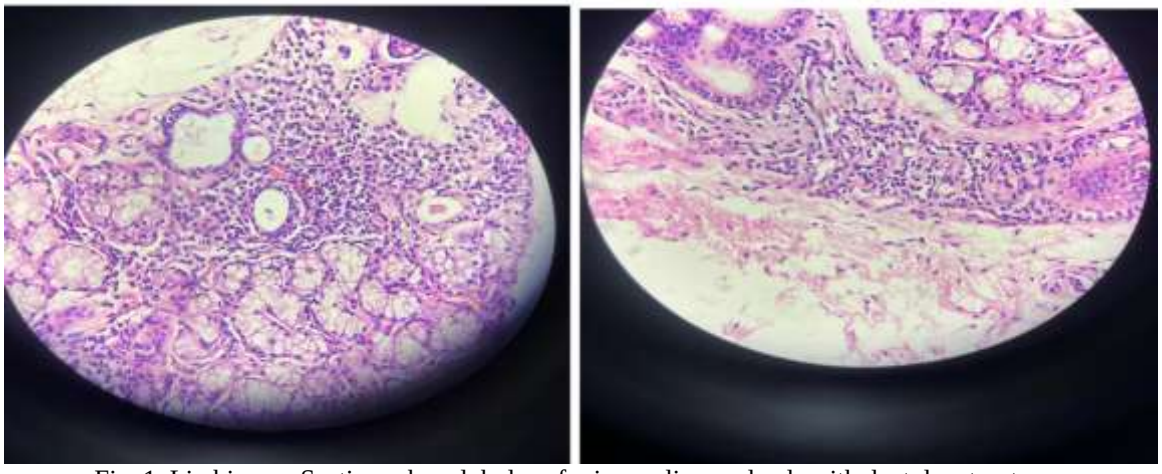

Fig. 1. Lip biopsy -Sections show lobules of minor salivary glands with ductular structures

There is moderate generalised mature lymphoplasmacytic cells infiltrate with focal aggregation of more than 50 of them around the ducts (more than 50 lymphoplasmacytic cells aggregate is known as focus score).[11]Focus score of the given sample is 2.0.

From the above blood investigations and biopsy, she was diagnosed to have both Sjogrens syndrome and autoimmune primary hypothyroidism- Hashimoto thyroiditis

III. MANAGEMENT

Intravenous potassium correction was administered. Following correction of hypokalemia, bicarbonate correction was instituted. Motor weakness completely resolved on correction of electrolyte imbalance. Patient had become ambulant at the time of discharge. Thyroxine supplementation was also initiated.

IV. DISCUSSION

Even though the most common acquired cause of distal RTA is Sjogrens syndrome, its initial manifestation as hypokalemicquadriparesis

is very rare.[2] The pathogenesis of renal involvement in Sjogrens is still not clearly elucidated. One of the mechanisms proposed is that antibodies are formed against vacuolar H+ ATPase anion exchanger 1 and carbonic anhydrase II. [3, 4, 5] Another hypothesis postulated is a defect in the regulation of ATPase of vacuolar and endosomal membranes.[6] Both proximal as well as distal RTA have been described. Hypokalemic paralysis and RTA are also observed in primary hypothyroidism especially with autoimmune primary hypothyroidism.[7, 8]There is also an association between Sjogrens and autoimmune hypothyroidism.[9] Both Hashimotos thyroiditis and Sjogrens syndrome are characterised by lymphocytic infiltration around the ducts and are pathologically similar. Also both diseases can progress to Non Hodgkins Lymphoma.[9, 10] Here hypokalemic paralysis may have been precipitated by autoimmune hypothyroidism in this patient with underlying Sjogrens syndrome.

CONCLUSION

Hypokalemia due to RTA is a rare but life-threatening complication of Sjogrens syndrome. Electrolyte disorders should be kept as differential diagnosis in the evaluation of neurological disorders. Distal RTA in middle aged ladies should prompt a search for underlying autoimmune conditions.

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