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A case of Renal tubular acidosis as an acute presentation of Sjogren Syndrome with Hyperthyroidism

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ABSTRACT

We present a case of 26 year old female with a presentation of hypokalemia and distal renal tubular acidosis and normal anion gap acidosis. All basic **b**lood investigations were normal and Anti Nuclear Antibody screening was done which was grade IV positive. Subsequently ANA profile was done which was positive was Ssa/Ro 60k Ab titer, Ssa/Ro 52kd Ab titer and Ssb Ab titer. Thyroid profile showed TSH 0.33 and T3 and T4 were 3.1 and 4.9 respectively. Urine anion gap was positive and urine pH was 6.8. The patient was diagnosed as Sjogren syndrome with hyperthyroidism and type 1 Renal Tubular Acidosis. This case is unique as the initial presentation of this patient was RTA and the patient had hyperthyroidism. Most patients of Sjogren have hypothyroidism and not hyperthyroidism.

Keywords: Sjogren syndrome, hypothyroidism, renal tubular acidosis, ANA screen

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INTRODUCTION

Sjogren syndrome is a rare autoimmune disorder in which there is lymphocytic infiltration of the exocrine glands[1]. Most common glands affected are lacrimal glands and salivary glands and most common manifestations are dry eyes and dry mouth[2]. However, extraglandular involvement is not uncommon. Renal involvement is quite commonly seen in patients with Sjogrens syndrome with type 1 Renal Tubular Acidosis being the most common involvement. In this case report, we present a 26 year old female who presented with quadriparesis due to hypokalemia and was eventually diagnosed as primary Sjogren syndrome with Hyperthyroidism.

CASE REPORT:

A 26 year old female with no previous medical illness presented to us with complaint of abdominal pain and generalized weakness for a period of 15 days. The patient denied any complaint of fever, tingling/numbness, nausea, vomiting, focal neurological deficit or changes in mental status or sensorium. The patient had f*laccid quadreparesis.* Muscle power was 3/5 in both upper and lower limbs. Muscle tone was reduced in all four limbs. Deep tendon reflexes were absent in all limbs. Sensations were intact in all the *limbs.* A differential diagnosis of Guillan Barre syndrome and Hypokalemia Paralysis was considered. Serum Potassium level revealed a low Potassium level of <2.5mg/dl. Intravenous Potassium correction was started via a peripheral line. After 12 hours, neurological examination was repeated and there was an improvement in muscle power of all the limbs. The muscle power was 5/5 in all the limbs. Arterial blood gas analysis was sent which showed normal anion gap metabolic acidosis with respiratory compensation. Potassium level was repeated after the potassium drip was completed. It was still low (<2.5mEq/l). Repeat ABGA showed normal anion gap metabolic acidosis. Urine pH was 6.8 and urine anion gap was positive. The patient was diagnosed to have Type 1 Renal Tubular Acidosis.

ANA screening showed 1:1000 dilution positive. Intensity was 4+. Pattern was nuclear speckled. Anti-nuclear antibody profile was done which showed Ssa/Ro 60k Ab titer of 97IU/ml. Ssa/Ro 52kd Ab titer was 100IU/ml. Ssb Ab titer was 92IU/ml. Sjogren syndrome was diagnosed. Direct Coombs test was Grade 2 positive. Since thyroid dysfunction is common in patients with Sjogren syndrome, Thyroid profile was done. TSH was 0.33. T3 and T4 were 3.1 and 4.9 respectively. Anti Thyroperoxidase antibody was normal. Tablet Carbimazole 20mg TDS was started. Ophthalmology reference was done for Schiemer test. Test 1 was 20mm in Right eye and 20 mm in left eye. In step 2, It was 15mm in Right eye and 15mm in left eye. Patient was started on Syrup Potchlor. Tablet Prednisolone 7.5 mg OD and Tablet HCQ 200 BD were started as per the advise by Rheumatologist. The patient

showed significant clinical improvement in terms of fatigue and weakness after a period of 15 days. Potassium was repeated after 15 days and it was 3.5mEq/l.

DISCUSSION

Sjogren disease is a complex and heterogeneous condition in which typical presentation is with sicca symptoms i.e. dry eyes and dry mouth[3]. Salivary glands, lacrimal glands and parotid glands are most commonly affected. However this immune process can also affect non-exocrine organs such as kidneys, gastrointestinal tract, skin and lungs. Sicca symptoms may not always be present. In the case series by Shioji et al, only two patients out of four had dry mouth and none had xerosis. All 4 patients presented with muscle weakness or arthralgia [4]. A number of cases have been reported with muscle weakness, hypokalemic paralysis and pathological fractures as the initial presentation of Sjogren syndrome. Therefore, Sjogren syndrome should strongly be suspected in patients with RTA, symptoms related to RTA even in absence of sicca symptoms [4-7].

The diagnosis of Sjogren syndrome is challenging for clinicians as many of the cases of Sjogren syndrome do not have the typical presentation of sicca syndrome. The diagnosis of Sjogren is generally made by the American-European Consensus Criteria which requires four out of six criteria: ocular or oral symptoms; ocular or oral signs; presence of autoantibodies and histopathology from a lip biopsy[8].

Renal involvement in Sjogren syndrome varies from 0.3% to 27% based on biopsy findings or changes in biochemical profile. The most common renal manifestation of Sjogren syndrome is tubulointerstitial inflammation[9-11]. The clinical manifestation depends on the segment involved by the lymphocytic infiltration. Common manifestations are distal RTA, hypokalemia, Gitelman syndrome, Fanconi syndrome and rarely Diabetes Inspidus[12]. Nephrocalcinosis has also been reported in many patients[13].

The association between autoimmune dysfunction and Sjogrens syndrome has been well documented[14]. Sjogren Syndrome is the most frequently autoimmune disease associated with autoimmune thyroid dysfunction[14].

Our patient had Sjogren syndrome with Hyperthyroidism and the initial presentation of the disease was atypical presenting as renal tubular acidosis [1].

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