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Case Report

Male Sjogren's syndrome presenting with respiratory paralysis due to hypokalemia

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ABSTRACT

Respiratory paralysis due to hypokalemia is a rare entity in Sjogren's syndrome. Apart from distal renal tubular acidosis (RTA) and hypokalemia the clinical symptoms like dryness of eye, mouth and parotid swelling were absent in our case. Due to this rarity in this pattern of presenting the symptoms, the differential diagnosis of autoimmune disease is often missed and it will eventually end up in a fatal condition. Hypokalemia causes muscle weakness gradually and there will be sudden onset of respiratory paralysis which could be dangerous to the individual. Although there is the presence of rare combination of symptoms clinical history, blood gas analysis, urine analysis and ANA profile will help in the appropriate diagnosis. Here we report a rare case of male Sjogren's syndrome presenting with respiratory paralysis due to hypokalemia.

Keywords: Respiratory paralysis, Hypokalemia, Sjogren's syndrome

INTRODUCTION

Hypokalemic weakness and paralysis is often noted in distal renal tubular acidosis (dRTA).[1] Potassium depletion can result in several muscular related complications.[2] Hypokalemia can hyperpolarize the skeletal muscle cells, impairing their ability to develop depolarisation necessary for muscle contraction.[3] Profound potassium deficiency can cause paralysis of respiratory muscles too. Hypokalemia and an increased urine pH confirms the diagnosis of distal RTA. RTA is one of the major causes of acquired hypokalemic paralysis. Distal RTA is the common pathway for potassium loss in variety of autoimmune diseases including sjogren's syndrome.[4,5] Renal tubular acidosis causes impairment of urinary acidification. Distal RTA is the most common form and it is associated with autoimmune diseases such as Sjogren's syndrome and systemic lupus erythematosus.[6,7] Several studies have already suggested that autoantibodies against CA II enzyme or the acid base transporters are involved in the pathogenesis of dRTA in autoimmune diseases.[8,9].

CASE REPORT

A 33 years old male presented to emergency department with history of weakness of both the lower limbs followed by upper limbs since 2 days. The weakness was of sudden in onset and gradually progressive in nature. He had vomiting since 2 days, 4-6 episodes per day. He reported that he had difficulty in breathing since last four hours. There was no history of fever, loose stools, abdominal pain, chest pain, palpitation, cough, headache, seizure, joint pain or rashes. He was non-alcoholic and non-smoker.

On examination the patient was conscious, oriented, febrile and dehydrated. Vital signs were of blood pressure (BP) - 110/90 mmHg, pulse rate (PR) - 104 bpm, respiratory rate (RR) - 40 breaths per min, temperature - 100.8° F and SpO_2 on room air was 88%. On systemic examination hypotonia of both the upper and lower limbs were found. Power in both upper and lower limbs was 2/5. Knee jerk and ankle jerk were diminished and plantar reflex was flexor on both sides.

Arterial blood gas analysis was done. Arterial blood gas (ABG) was suggestive of normal anion gap with metabolic acidosis. pH was 7.17 with HCO_3 7.3, potassium – 1.4 mmol/l and anion gap 11 mEq/l. Electrocardiogram showed ST depression with U waves (Figure 1). Within few minutes of arrival in view of tachypnea and respiratory distress patient was intubated and put on mechanical ventilation.

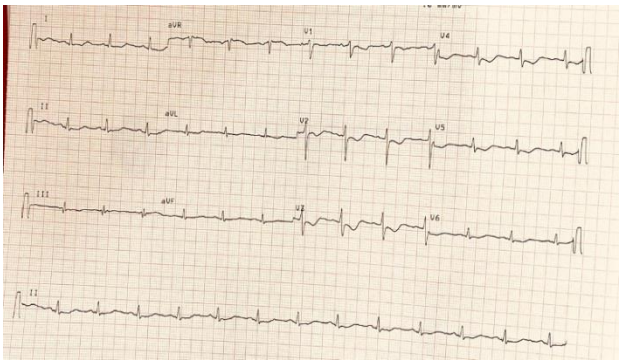

Figure 1: ECG of ST depression with u waves in lead v1 to v5.

Laboratory tests revealed slightly elevated total white blood cell (WBC) count of 14,900 cells/mm^3 and decreased Hb of 9.8 gm/dl. Erythrocyte sedimentation rate (ESR) was raised to 55 mm/hour. The serum urea 29 mg/dl, serum creatinine 1.4 mg/dl, serum potassium 1.6 mEq/l, serum magnesium 2.8 mg/dl were noted. Urinary analysis showed urinary loss of potassium with urinary spot k 20.7 mEq/l. Urinary pH was 7 and trans-tubular potassium gradient (TTKG) was 7.5.

Since ABG showed normal anion gap metabolic acidosis with hypokalemia, so distal RTA was suspected. Renal loss of potassium was confirmed by urinary spot k and TTKG. Antinuclear antibody (ANA) immunofluorescence assay revealed speckled pattern cytoplasm (Figure 2) and ANA immunoblotting was done which showed anti SSA and anti SSB were found to be strongly positive.

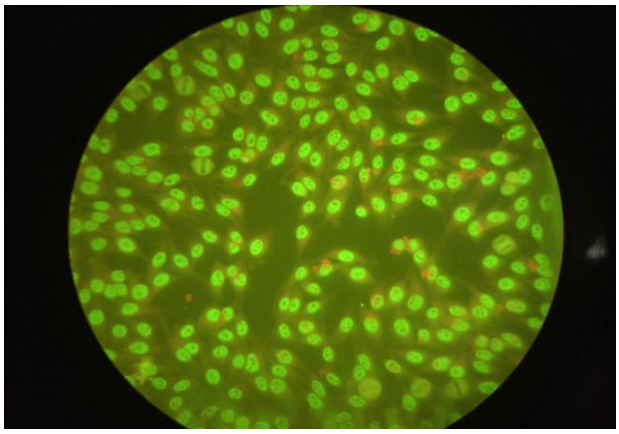

Figure 2: ANA IFA of speckled pattern cytoplasm.

Patient was started with intravenous (IV) potassium chloride 220 mEq for 3 days along with 100 mEq bicarbonate. IV antibiotic was also given. On day 3, patient was weaned off from mechanical ventilation and the muscle power resumed back to 5/5. Patient was discharged with potassium citrate syrup and now in regular follow up with rheumatology outpatient department.

DISCUSSION

Sjögren syndrome is chronic systemic autoimmune disorder characterized by lymphocytic infiltration of exocrine glands. Sjogren's as such has the overall prevalence of 0.1-4.8% and female to male ratio is 9:1. Females are affected more frequently than males.[10] The prevalence of SS among women amounted to 0.31%, while for men it is 0.07%.[11]

Most patients presents with sicca symptoms such as xerophthalmia (dry eyes), xerostomia (dry mouth), and parotid gland enlargement. In addition, various extraglandular features may develop. Renal involvement includes proximal and distal tubular acidosis, tubular proteinuria, and nephrogenic diabetes insipidus.[12]

In distal RTA the nephrons lack the ability to secrete H+ ions. Acquired causes for RTA include autoimmune diseases such as thyroiditis, Graves' disease and Sjogren's syndrome. Sudden life threatening hypokalaemia with muscle paralysis is the most serious clinical consequence of distal RTA.[13] The incidence of RTA patients with Sjogren's is around 34.8%.[14] The largest case series maintained at centre for rheumatic disease, Pune where only 16 sjogren's syndrome patients were documented to have hypokalemic periodic paralysis among a database of 50000 patients.[15]

The possible mechanism of dRTA is due to an absence of H+ - ATPase pump in collecting tubules or defective H+ - ATPase pump.[16] There have been cases reported to have hypokalaemia paralysis in sjogren's syndrome from India and abroad previously. In 1981 the first such case was reported.[17] Although hypokalemia is frequent sequel of RTA, a severe symptomatic decrease in serum potassium concentration has been described in a few cases only. Hypokalemic paralysis is a rarely encountered complication of RTA secondary to primary Sjogren's syndrome.

In our case, the patient had no Sicca symptoms and there was absence of parotid swelling too. Arterial blood gas analysis along with urinary spot k helps in the diagnosis of Sjogren's syndrome. Along with that the presence of antibodies anti SSA and anti SSB confirmed our diagnosis. The patient went into the rare complication of Sjogren's that is hypokalemia induced respiratory paralysis. Since the complication of respiratory paralysis is very rare, the rate of mortality is still unrecognised. Immediate treatment with potassium chloride and

antibiotics saved the patient from the deteriorating condition and mortality. The prognosis of our patient improved after timely diagnosis and prompt treatment. So the patient is discharged with the supplementation of potassium citrate syrup and regular follow up in rheumatology department.

CONCLUSION

When a patient presents with hypokalemic periodic paralysis with distal RTA diagnosis of autoimmune disorders should not be missed. Even though there is no actual standard symptoms, clinical history, ABG analysis and ANA profile will help us in timely diagnosis and early treatment of Sjogren's syndrome.

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