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Case Report

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HYPOKALEMIC RESPIRATORY PARALYSIS UNMASKING SJOGREN'S SYNDROME IN MALE

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ABSTRACT

Sjogrens syndrome is an autoimmune disease with female predominance characterised by exocrine involvement. Extraglandular manifestations are reported in upto 10% of cases and kidneys are most commonly involved. However, presentation in form of hypokalemic paralysis is rarely reported and even few cases are reported in males. We present a case of hypokalemic paralysis that eventually unmasked periodic paralysis in a male patient.

KEYWORDS: Sjogrens syndrome, Distal Renal tubular acidosis, Paralysis, Hypokalemia.

INTRODUCTION

Sjogren's syndrome (SS) is an autoimmune lymphocytic infiltrative disease that can result in chronic inflammatory changes to exocrine glands and extra-glandular systemic organs. Overall prevalence is reported as 0.1-4.8% and females are affected more frequently than males.[1] The prevalence of SS among women amounted to 0.31% in comparison to men with reported prevalence of 0.07%. [2]

Extra glandular manifestation of SS can involve any organ and renal involvement is reported in up to 10% of cases.[3] Distal renal tubular acidosis (dRTA) is the most common renal manifestation. Complete dRTA (urine acidification defect with acidosis) is less commonly reported than incomplete dRTA (urine acidification defect without acidosis).[4] Hypokalemic paralysis is a well known, albeit rare, complication of dRTA from any cause.[5,6] In a cross-sectional retrospective study from database maintained at Centre for Rheumatic Diseases, Pune; only 16 out of 50000 patients were documented to have Hypokalemic paralysis.[7]

In this case report, we discuss an adult male with hypokalemic flaccid paralysis who was eventually diagnosed as Sjogren's syndrome.

CASE REPORT

A 51 year old male presented to the emergency department (EMD) of a tertiary care hospital with history of weakness in all four limbs for 8 hours. The weakness was progressive with complete paralysis of the upper and lower extremities. His attendant also reported that the patient was having difficulty in breathing for past 1 hour and he was immediately intubated in EMD in view of respiratory distress and tachypnea. There was no history of fever, headache, seizure, animal bite or trauma to head or neck. There were no complaints of preceding diarrhoea or vomiting. He was a smoker and non-alcoholic.

On examination the patient was drowsy and not oriented with a GCS of E2V1M1. Vital signs were recorded as Blood pressure (BP) - 114/76 mmHg, Pulse rate (PR) - 98 bpm, Respiratory rate (RR)- 38/min, Spo2 on room air was 78%. Power in bilateral upper and lower limbs across all the joints, neck flexor and trunk was 0/5. Deep tendon reflexes were absent in bilateral biceps, triceps, knee and ankle with mute plantars. Cardiovascular, respiratory and gastrointestinal examinations were unremarkable.

Arterial blood analysis (ABG) was suggestive of normal anion gap metabolic acidosis. Ph was 7.14 with HOC3 - 8.1, Potassium – 1.42 mmol/l and anion gap of 10meq/l. Electrocardiogram(ECG) was suggested of widespread ST depression and T wave flattening with prominent U wave (best seen in precordial lead V3) with apparent QTc interval of 494 msec(figure-1).

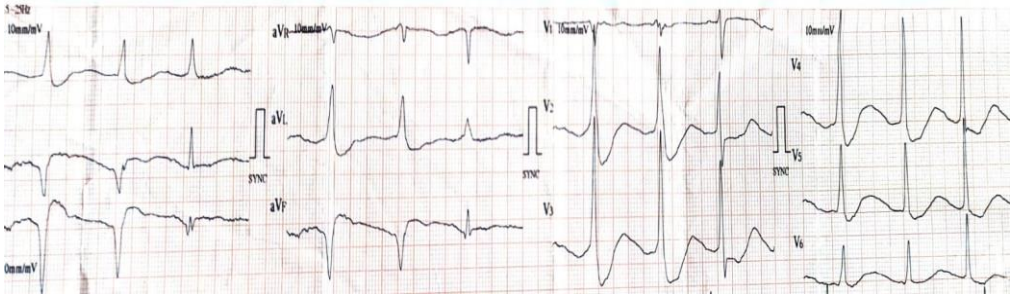

Figure 1: ECG suggested of widespread ST depression and T wave flattening with prominent U wave (Best seen in precordial lead V3).

Hemogram was done which showed Hb- 15.7g/dl, TLC-9300/ mm^3, platelets - 2.37L/mm^3. Complete biochemical blood panel was done which revealed severe hypokalemia- 1.6(consistent with ABG), other reports were as follows (table-1): Sodium- 141mmol/l, Chloride-106 mmol/l, Calcium/phosphorus- 8.4/2.8 mg/dl, Magnesium- 2.2 meq/l, urea/Creatinine- 22/0.9mg/dl. Liver function test, thyroid function test and intact-PTH were normal.

Urine analysis was suggestive of Ph- 6.8, no cells or cast; 24hours urinary loss of Potassium (K^+) was 75mmol and trans-tubular potassium gradient (TTKG) was 12. This decrease in pH, along with an elevated TTKG ushered in the diagnosis of dRTA, with hypokalemic paralysis as its complication.

Antinuclear antibody (ANA) Immunofluorescence assay revealed speckled pattern cytoplasm and ANA immunoblotting showed anti- SSA and anti- SSB anti bodies to be strongly positive(Table-1).

Table 1: Patients laboratory results.

Laboratories	Patient values	Reference ranges
Complete Hemogram		
Hemoglobin (Hb)	15.7	13-15g/dl
Total leukocytes count(TLC)	9.3	4.0-10.0*10^3/uL
Platelets	237	150-400*10^3/uL
Serum Biochemistry		
Sodium	141	135-145
Potassium	1.6	3.5-5.0
Chloride	106	90-105
Urea	22	17-43
Creatinine	0.9	0.6-1.2
Calcium	8.4	8.2-10.2
Phosphorus	2.8	2-5-4.0
Magnesium	2.2	1.8-2.6
TSH	2.93	0.30-5.5 uiU/ml
T3	0.99	0.7-2.4 ng/ml
T4	6.39	5.0-14.1 ug/dl
Ipth	54	15-65ng/l
Urine Electrolytes(24 hours)		
Ph	6.9	4.6-8.0
Sodium	184	30-300mmol/24hrs
Potassium	75	25-125mmol/24hrs
Chloride	224	110-250mmol/24hrs
Creatinine	61.25	Undefined
Arterial Blood Gas		
Ph	7.14	7.35-7.45
HCO3	8.1	22-26 mmol/l
Anion gap	10	<12meq/l
Autoimmune panel		
ANA	Positive	-
Primary dilution	1:80	-
Primary intensity of IF	3+	-
End point titre	1:320	-

Pattern	Speckled	-
Anti-Ro(SSA)	Strong positive +++	-
Anti-La(SSB)	Strong positive +++	-
Anti-U1-RNP	Positive ++	-

ANA: antinuclear antibody, anti- SSA/B: anti sjogren's syndrome-related antigen A/B; anti-U1-RNP: anti-ribonucleoprotein antibody

For objective evidence of glandular involvement, Schirmer's test was demonstrated to be positive. Diagnosis of Sjogren's Syndrome was made based on American College of Rheumatology and European League Against Rheumatism (ACR/EULAR) Classification score of 4(anti-SS-A/Ro positive, schirmer's in atleast 1 eye).

Patient was started on intravenous potassium chloride infusion along with bicarbonate supplementation and mechanical ventilation was continued along with continuous cardiac monitoring. On day 2, patient was weaned off from mechanical ventilation. His muscle power resumed to 5/5 and patient was subsequently discharged on potassium citrate, low dose steroids and hydroxychloroquine.

DISCUSSION

Sjogren's syndrome (SS) is an autoimmune exocrinopathy with variable systemic involvement caused by lymphocytic infiltration. It is usually suspected when patient presents with sicca syndrome (dry eyes, dry mouth). Although the involvement of minor salivary gland is present in most patients, 62% of patient lacked clinical signs and symptoms of salivary gland involvement.[8] SS may occur in isolation (Primary SS) or more commonly in context of any underlying autoimmune disease (Secondary SS) like systemic lupus erythematosus (SLE), rheumatoid arthritis or scleroderma. SS often affects middle-aged women with mean age of presentation of primary SS (pSS) in general population being 52.7 years.[9,10]

Presence of renal involvement in SS has been known since 1960s, usually in the form of tubulointerstitial nephritis and distal RTA (dRTA). Most cases, however, are either mild or asymptomatic.[11] SS manifesting for the first time as paralysis caused by dRTA is a very rare presentation and has been reported in <2% cases.[12, 13]

Distal renal tubular acidosis (type 1 RTA) is a disorder of distal nephron, which cannot lower the urine pH normally. dRTA can be inherited of acquired. Among acquired, autoimmune disease like SS, sarcoidosis and SLE are common. dRTA is associated with hypokalemia and normal anion gap metabolic acidosis although pathophysiology is not clearly understood; however, multiple theories have been put forward. The distal segment and collecting tubule is the site of final regulation of urinary acid excretion, and type A intercalated cells in this segment perform the function of distal H^+ ion secretion and HCO3- reabsorption. The secretion of H^+ is effected by H^+K^+ ATPase pumps abundantly expressed on apical pole of intercalated cells. The proton secretion activity is coupled with HCO3- reabsorption activity carried out by basolateral Cl-/HCO3- exchanger. The possible mechanism is absence or defect in this H^+K^+ ATPase pumps. This has been backed up by several case reports with immunocytological analysis on renal biopsy which demonstrated complete absence of this pump in affected patients. How the immune injury leads to loss of H-ATPase activity is not known. Autoantibody directed against carbonic anhydrase II has been proposed as another mechanism of distal RTA in pSS. More research is needed to establish a definitive mechanism and cause-effect relationship, however in a case series of 60 patients presenting as symptomatic hypokalemia (40% without clinical symptoms), only two patients relapsed after receiving therapy against SS during a mean follow up of 16 months following stoppage of potassium supplementation.

Our patient presented with hypokalemic paralyses and investigations were suggestive of distal RTA. To look for cause of distal RTA, autoimmune profile was sent keeping a distant possibility of Sjogren's syndrome; as clinically sicca symptoms were absent and there was scarcity of literature on male Sjogren's syndrome presenting as hypokalemic paralysis. Our patient responded well to potassium supplementation and put on low dose steroids and immunomodulators and is currently in follow up with rheumatology outpatient department and doing well.

CONCLUSION

This case report identifies the importance of identifying treatable cause in a life threatening condition like acute flaccid paralysis. We would also emphasize on the fact that hypokalemia should not just be corrected but also investigated. This case was peculiar as this was a male patient presenting with dRTA secondary to SS. SS presenting as hypokalemic paralysis is very rare in males, but this differential should be kept in mind while investigating a case of hypokalemic paralysis secondary to distal RTA.

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