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Original Research Paper

General Medicine

SJOGREN'S SYNDROME PRESENTING AS HYPOKALEMIC PARALYSIS- A RARE CASE PRESENTATION

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ABSTRACT

Introduction: Sjogren's syndrome is slowly progressive protype autoimmune disease characterized by lymphocytic infiltration of the exocrine glands. Most common manifestation are related to tubular dysfunction resulting from chronic interstitial nephritis and can manifest as distal renal tubular acidosis [dRTA] ,proximal RTA, tubular proteinuria and nephrogenic diabetes inspidus of which RTA is the main clinical manifestation. It is more common in middle aged women and two thirds of them will develop symptoms Hypokalemic paralysis rarely occurs as the first manifestation of renal tubule disorder due to pSS. **Case Report:** A 30 year old female presented to casualty of hind institute of medical sciences, sitapur, uttar Pradesh. With complains of all four limbs weakness which is acute in onset. Patient also gives history of similar complains 15 days back and was diagnosed hypokalaemia and was given potassium correction and symptomatic treatment and discharged with advice of oral potassium supplement which she discontinued after discharge . **Conclusion:** We report this case to highlight the fact that patients presenting with hypokalaemia periodic paralysis can be due to early sjogren's syndrome which might not present with typical sicca symptoms and dryness of mouth, enlargement of parotid gland, which needs to be investigated and started on appropriate line of management.

KEYWORDS :

INTRODUCTION

Sjogren's syndrome is an autoimmune disease with glandular [salivary and lacrimal] and extra glandular [neurologic, renal, hepatic, respiratory, vascular, and cutaneous] manifestations.Tubulointerstitial nephritis [TIN] is the main renal involvement associated with primary Sjogren's syndrome [pSS]. TIN can manifest as distal renal tubular acidosis [RTA], nephro-genic diabetes insipidus, proximal tubular dysfunction, and others [1], of which RTA is the main clinical presentation[2]. RTA has been reported in 4.3 to 9°/o of pss patients; it is more common in middle- aged women, and two-thirds of them will develop symptoms [2, 3]. Hypokalemic paralysis is the initial symptom in seven percent of patients with Sjogren's syndrome [4].

Case Report

1 A 30 years old female presented to casualty of hind institute of medical sciences, sitapur, uttar Pradesh with complains of all four limb weakness which is sudden onset. Patient also gives history of similar complains 15 days back and was diagnosed hypokalemia and was given potassium correction and symptomatic treatment and discharged with advice of oral potassium supplement which she discontinued after discharge. Her vital signs on admission were a temperature of 98.6 F, a heart rate of 58 beats per minute, a respiratory rate of 20 breaths per minute, oxygen saturation of 97% at room air, capillary blood glucose of 108 g/dL, and blood pressure of 100/60 mmHg. On physical examination, the deep tendon reflexes were diminished and had quadriparesis with power of 1/5 in all four limbs. Patient is given i.v potassium chloride for hypokalaemia and oral sodium bicarbonate for metabolic acidosis. The patient was discharged and she was reported to be asymptomatic with the use of potassium citrate only.

DISCUSSION

This female patient presented with hypokalaemia paralysis due to dRTA as the first and only manifestation of Sjogren's syndrome. Although asymptomatic for sicca symptoms. Clinically and serologically, there was no evidence of underlying cause for secondary Sjogren's syndrome. She showed good response to replacement with potassium citrate and has shown almost near resolution of tubular defect after a course of immunosuppressive therapy with steroids. This case highlights the need to consider the possibility of Sjogren's syndrome in a case of RTA even without any other manifestation of the syndrome and response to immuno-suppressive therapy in complete resolution of renal tubular dysfunction. Distal renal tubular acidosis is a disorder of the distal nephron, which cannot lower the urine pH normally. The underlying cause is due to the excessive back-diffusion of hydrogen ions from the lumen of the collecting duct to blood or there is inadequate transport of hydrogen ions. It is diagnosed by a normal anion gap metabolic acidosis with a simultaneous urine pH greater than 5.5.

Hypokalemia may also occur because urinary concentration and potassium conservation also tend to be impaired. Severe hypokalemia causing paralysis as first presentation of dRTA secondary to Sjogren's syndrome is a rare occurrence. RTA is a frequent extraglandular manifestation of primary Sjogren's syndrome with an incidence of about 7.1-19.2%.[5] It may also be the first clue in the identification of an underlying autoimmune disorder, particularly Sjogren's syndrome.[6] The pathogenesis of dRTA in Sjogren's syndrome is not very clear. Patients with Sjogren's syndrome with dRTA have interstitial nephritis with high levels of anti- carbonic anhydrase anti- bodies which affect the function of carbonic

anhydrase in cortical collecting ducts. [7] The acidification defect was the result of a lack of intact Hb ATPase pumps in the intercalated cells.

Table No 1. Investigation

INVESTIGATION	Value	Reference Range
Hb	1 1.9 gm/dl	12-15 gm/dl
TLC	5700 cell/cumm	4500-11000 cell/cumm
PLATELET COUNT	2.2Lac cell/cumm	1.5-4.5 cell/cumm
Serum Na	142mEq/Lit	135-145 mEq/Lit
Serum K	1.9 mEq/Lit	3.5-5.5 mEq/Lit
Serum Cl	118 mEq/Lit	96-109 mEq/Lit
Serum Mg	1.8 mg/dl	1.4-2.1 mg/dl
Serum phosphorous	2.2 mg/dl	3-4.5 mg/dl
Urinary Na	46 mmol/L	40-220 mmol/L
Urinary K	9.8 mmol/L	25-125 mmol/L
Urinary Cl	68 mEq/24	110-250 mEq/24
Urine Ph	7.5	5-7
Serum anion gap	10 mEq/L	8-12 mEq/L
T3	1.7 ng/ml	0.06-1.18 ng/dl
T4	13.1 ng/dl	5.0-12.45 ng/dl
TSH	1.14 uIU/mL	0.35-5.5 uIU/mL

ANA PROFILE is strongly positive for SS-A native and Ro-52 recombinant

CONCLUSION

We report this case to highlight the fact that patients presenting with hypokalaemia periodic paralysis can be due to early Sjogren's syndrome which might not present with typical sicca symptoms and which needs to be investigated and started on appropriate line of management.

Ethical Approval: Not required.
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Consent for Publication

Written informed consent was obtained from the patients' father for publication of this case report.

Declaration of Competing Interest: None.

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