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CASE REPORT

Distal RTA with Hypokalemic Quadriparesis as the Initial Presentation of Primary Sjogren's Syndrome

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Abstract

Sjogren syndrome (SS) is a multisystemic autoimmune disorder with predominant exocrine gland involvement leading to sicca symptoms. Among extraglandular manifestations, renal involvement is commonly seen. Renal involvement can be either tubulointerstitial (TIN) or glomerular, the former being more common. Distal RTA is the most common manifestation of TIN presenting as mild hypokalaemia, metabolic acidosis and rarely with hypokalaemic periodic paralysis. We report a case of a 70-year-old female who presented with hypokalaemic periodic paralysis and metabolic acidosis diagnosed as distal RTA. On further evaluating the cause of distal RTA, the patient was diagnosed with Sjogren's syndrome and managed accordingly after assessing the ESSDAI score. Our report illustrates that Sjogren syndrome is a rare but important cause of hypokalaemic periodic paralysis.

Keywords

Primary Sjogren's syndrome, Distal renal tubular acidosis, Hypokalaemic periodic paralysis

Introduction

Renal involvement in primary Sjogren's syndrome (pSS) is not commonly defined. It is either due to tubulointerstitial involvement or less commonly due to glomerular involvement [1]. Renal tubular acidosis is seen in 9% of the patient suffering from pSS. Distal renal tubular acidosis is the most common presentation of renal involvement in a patient with pSS [2]. Clinical features of distal RTA include generalized weakness, muscle pain, polyuria, polydipsia, fractures, and renal colic. Laboratory derangement commonly includes mild hypokalaemia, metabolic acidosis, hypercalciuria, and proteinuria. Hypokalaemic periodic paralysis as the presenting symptom of pSS is uncommon. Our patient had 2 episodes of hypokalaemic periodic paralysis without renal dysfunction in the past for which she was treated with intravenous potassium and improved, but the cause was not evaluated. She presented to us with severe hypokalaemic periodic paralysis with renal dysfunction (eGFR - 26.5 ml/min/m^2) and metabolic acidosis. The evaluation revealed distal renal tubular acidosis (RTA) and pSS was diagnosed on basis of clinical presentation and presence of antibodies.

Case Report

A 70-year-old female with no known comorbidities presented to the Emergency Department with complaints of acute onset progressive weakness of all four limbs for 7 days, associated with decreased talk and decreased oral intake for 4 days. History of fever, headache, trauma, vomiting, or diarrhoea was absent. The patient has been admitted outside and was referred to our hospital for further management. The patient has had 2 episodes of similar complaints in the past 3 months, for which she was diagnosed to have hypokalaemia, and weakness gradually improved with correction of hypokalaemia.

At the presentation to the ED, the patient was conscious but confused, blood pressure 120/76 mmHg, respiratory rate 20/min, and pulse rate of 98/min. General physical examination revealed pallor, dry eyes, dry oral cavity with dental erosions, and bilateral upper

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Table 1: Lab parameters of the patient.

Lab parameters	Patient	Reference value
Hemoglobin	10.9 gm/dl	11-12 gm/dl
WBCs	16000/µL	< 12000/µL
Platelets	1.9 lacs/µL	1.5-4.5 lacs/µL
Erythrocyte sedimentation rate	3 mm in 1st hour	< 20 mm in 1st hour
HsCRP	38.20 mg/dL	< 6 mg/dL
Serum sodium	141 mEq/L	135-145 mEq/L
Serum potassium	**1.74 mEq/L**	**3.5-5.1 mEq/L**
TSH Free T3 Free T4	1.09 mIU/L 0.57 pg/ml 0.615 ng/dl	TSH - 0.3-4.0 mIU/L Free T3 - 2.2-4.2 pg/ml Free T4 - 0.80-1.70 ng/dl
ABG	pH-7.13, pCO_2- 23 mmHg, HCO_3-6.8 mmol/L, Na-140 mmol/L, K-1.74 mmol/L, Chloride-122 mmol/L, Serum Osmolality-305 mosm/kg	
Urinalysis	pH-7, urinary Na-68 mmol/L, urinary K-37 mmol/l, urinary Chloride-97 mmol/L, urine osmolality-327 mosm/kg	
Serum anion gap	10.8 mmol/L	Normal range 8-12 mmol/L
Urinary anion gap	**8 mmol/L (positive)**	**Positive - in favor of renal potassium loss**
TTKG	**18.45**	**TTKG > 4 - suggestive of renal loss**
Fractional bicarbonate excretion	4.10%	< 5% - in favor of distal RTA
KFT	**Urea-83 mg/dl,** **Creatinine-1.97 mg/dl**	Urea - 17-43 mg/dl Creatinine 0.6-1.1 mg/dl
eGFR	**26.8 mg/min/m²**	< 60 mg/min/m² - renal dysfunction
Viral markers	HIV-negative HBsAg-negative Anti HCV-negative	Negative
Autoimmune profile	**ANA-Positive,** **Anti SS-A-positive,** **Anti SS-B-positive,**	Negative
Schrimmers Test	**3 mm in both eyes**	< 10 mm - suggestive of dry eye
CECT brain	No abnormalities detected	
Ultrasound abdomen	No evidence of nephrocalcinosis	
Buccal mucosa punch biopsy	Mild lymphocytic inflammation and unremarkable minor salivary glands.	

mg: milligram; dL: decilitre; mmol: millimole; nmol: nanomole; L: liter; µL: microliter; U: units; µmol: micromole; mm: millimeter; µg: microgram; gm: gram; mosm: milli osmoles

limb swelling and redness suggestive of upper limb cellulitis. Neurological examination revealed areflexic flaccid paralysis with the power of 2/5 in all limbs and bilateral plantar reflex was mute. No cranial nerve deficit, autonomic disturbance, or involvement of bladder and bowel. The rest of the systemic examination was within normal limits.

After the history and clinical examination, we had differentials like Guillain-Barre syndrome, hypokalaemia, and transverse myelitis. Random blood glucose was 122 mg/dl, arterial blood gas showed pH 7.13, bicarbonate 6.8 mmol/L, pCO_2 23 mmHg, sodium 140 mmol/L, potassium 1.74 mmol/L and chloride 122 mmol/L. Other blood investigations are tabulated in Table 1. So after

getting the ABG report diagnosis of normal anion gap hyperchloremic metabolic acidosis with severe hypokalaemia was made. The patient was started on IV potassium chloride supplementation and IV fluids.

Further evaluation including urinary potassium, fractional bicarbonate excretion revealed distal renal tubular acidosis. After 3 days of intravenous potassium chloride infusion patient's power and consciousness improved following which she informed regarding the history of dry mouth and grittiness in eyes for around 6 months, along with occasional joint pain. Schirmer's test showed a 3 mm tear flow at 5 minutes. Diagnosis of pSS was made based on Newly devised Sjogren's International Collaborative Clinical Alliance (SICCA) guidelines 4 of 6 criteria were positive along with positivity to antibodies to SSA and SSB [3]. The final diagnosis of hypokalaemic periodic paralysis with distal RTA with renal dysfunction secondary to pSS was made and found to have moderate disease activity in the renal domain according to ESSDAI scoring [4]. The patient was started on bicarbonate supplementation and oral glucocorticoids after which patient's hypokalaemia and metabolic acidosis improved.

Discussion

Sjogren's syndrome is a multisystemic autoimmune disorder commonly presenting as lacrimal and salivary gland dysfunction. Renal involvement in pSS has a prevalence of around 9%, although the first clinical manifestation with renal involvement is a rare phenomenon. Renal involvement in pSS is either due to tubulointerstitial involvement or less commonly due to glomerular involvement. Lymphocytic infiltration of the renal tubules by T cells, B cells, or plasma cells has been hypothesized behind the pathogenesis of renal involvement. Other causes include decreased hydrogen ion secretion secondary to the absence of a vacuolar H+ ATPase pump in the distal tubules and antibodies to thiazide sensitive NaCl cotransporter (NCCT) [5]. The presence of anti-SSA, anti-SSB, and hypergammaglobulinemia are the only risk factors predisposing to renal involvement in pSS [1].

Clinical features of renal involvement include mild hypokalaemia, diabetes insipidus, renal tubular acidosis, severe hypokalaemic periodic paralysis, and Fanconi syndrome. Glomerular involvement manifested as proteinuria, haematuria and severe renal dysfunction is uncommon. Renal tubular acidosis with severe hypokalaemia causing paralysis is the most common manifestation of renal involvement in Sjogren's syndrome, although it is underdiagnosed [6].

Management of renal involvement is based on the site of kidney involvement, whether tubular or glomerular, and the ESSDAI score. Patients with mild ESSDAI scores are treated for hypokalaemia or acidosis, moderate ESSDAI is treated with oral glucocorticoids or immunosuppressant agents, and patients with severe ESSDAI are treated with glucocorticoids, rituximab, cyclophosphamide, and sometimes with plasma exchange. [2]. Our patient was treated with IV potassium supplementation along with steroids, after which she improved.

Conclusion

Hypokalemic periodic paralysis as a presenting complaint of Sjogren's syndrome is an uncommon entity. Sjogren's syndrome should be considered as a strong differential diagnosis while evaluating hypokalemic periodic paralysis associated with metabolic acidosis. Early treatment along with proper evaluation is essential to prevent recurrence of episodes of hypokalemic periodic paralysis which can be life-threatening. Our case report adds to the existing literature on renal involvement in a patient with Sjogren's syndrome.

Consent

Informed written consent was taken from the patient's son for case report writing.

Conflict of Interest

There is no conflict of interest between the authors.

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Nil.

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