A case of Sjogren's syndrome presenting with recurrent hypokalemia

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Abstract

We report a case of a 26-year old lady who presented with a history of several episodes of limb weakness requiring repeated hospitalization over the last 12 years and about 6 years back, she also developed features of sicca complex. Further investigations revealed hypokalemia, distal renal tubular acidosis and bilateral extensive nephrocalcinosis. Finally, a diagnosis of Sjogren's syndrome was made. Hypokalemia may be the presenting feature of Sjogren's syndrome. Sjogren's syndrome may be suspected in patients with recurrent hypokalemia and renal tubular acidosis.

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Introduction

Sjogren's syndrome is a rare systemic autoimmune condition with chronic inflammation of exocrine glands. It typically involves the lacrimal and salivary glands, causing dry eyes and dry mouth respectively [1]. In this disorder, kidneys are also involved due to autoimmune tubulointerstitial nephritis and distal renal tubular acidosis (RTA) [2]. Distal RTA is characterized by inability to acidify the urine in the distal parts of the nephron [3]. Though distal RTA is common in Sjogren's syndrome, it usually remains asymptomatic [4]. However, left untreated, it can lead to marked acid-base abnormalities like hyperchloremic metabolic acidosis and severe hypokalemia [3]. Hypokalemia is the most common electrolyte abnormality in distal RTA and may present earlier than typical glandular symptoms [5].

Here, we present a case of young lady with several episodes of weakness due to hypokalemia, who was subsequently diagnosed with primary Sjogren's syndrome. Though, Sjogren's syndrome is a recognized cause of distal RTA, its presentation as hypokalemic paralysis has not been widely reported in clinical practice.

Case report

A 26-year-old woman with a 12 year history of recurrent limb weakness presented to our institution with epigastric pain, vomiting and profound weakness for 5 days. Soon after admission, pain and vomiting subsided with conservative treatment but the weakness persisted. On query, she gave history of several episodes of limb weakness requiring repeated hospitalization over the last 12 years. Each episode resolved after potassium supplementation. She was labeled as a case of hypokalemic periodic paralysis and precluded further work up in primary health care centre. During review of her symptoms, she mentioned dry mouth with oral ulceration, dry eyes, dyspareunia, hair fall and multiple inflammatory small joint pain for the last 6 years. For the last two years she noticed marked tooth erosion (Figure-1) and unintentional weight loss. There was no significant family history of note.

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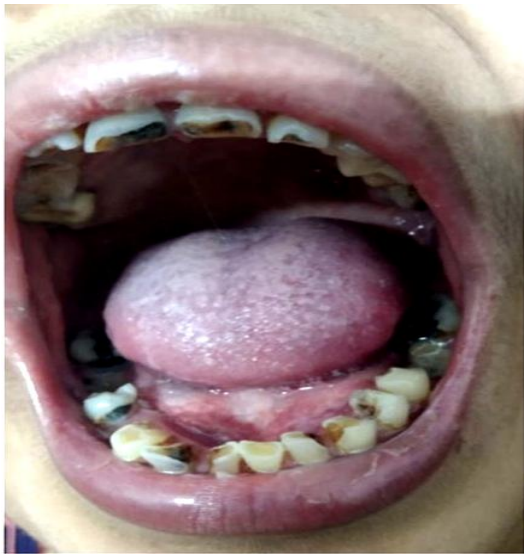

Figure-1: *Photograph showing presence of dental erosion*

On examination, there was xerosis of eyes and mouth, dental erosions and a positive Schirmer's test (<10mm of wetting in 5min). She was mildly anemic and had an enlarged (1x1 cm), non-tender, firm right supraclavicular lymph node. Nervous system examination revealed 3/5 muscle weakness, flaccid reflexes, flexor planters and no sensory deficit. Examination of all other systems was unremarkable.

Based on her clinical presentation, she was evaluated to find out the cause of recurrent hypokalemia. Laboratory investigations are shown in Table-1. Investigations revealed hypokalemia, normal anion gap metabolic acidosis and raised urine pH. Urine pH remained high (>5.3) after acid load test. These findings were consistent with distal RTA. She had evidence of microcytic hypochromic anemia with normal iron profile. Hemoglobin (Hb) electrophoresis showed evidence of beta-thalassemia trait. Liver function, renal function and calcium profile were normal. Her autoantibody screen revealed positive anti-nuclear (ANA), anti-Sjogren's syndrome type A (anti SS-A) and anti-Sjogren's syndrome type B (anti SS-B) antibodies. All other autoantibodies including anti-double stranded DNA, anti-Scl 70 (topoisomerase I), anti –ribonucleoproteins (anti-RNP), anti-Jo 1, anti-smooth muscle (anti- Sm) were negative. Imaging

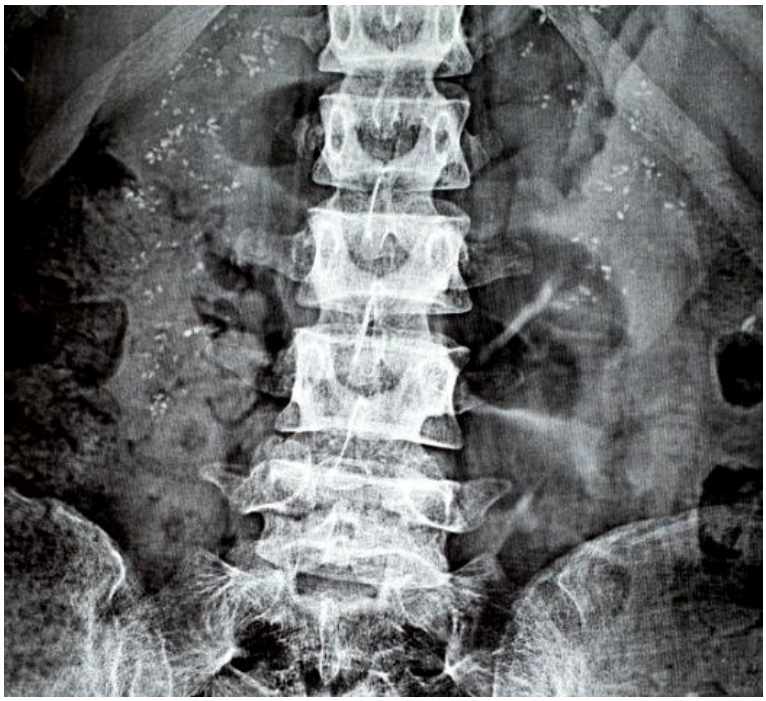

Figure-2: *Imaging of the abdomen showing extensive bilateral nephrocalcinosis*

Table-1: *Investigation results of the patient*

Parameter	Observed value	Reference range
Hb	9.8 gm/dl	12-15.5 gm/dl
MCV	74 fl	92-96 fl
MCH	24 pg	27.5-31.5 pg
MCHC	32 g/dl	31.5-34.5g/dl
RDW	21%	10-14 %
Hb electrophoresis	Beta-thalassemia trait	
S. electrolyte		
S. sodium	145.8 mmol/l	136-148
S. potassium	2.73 mmol/l	3.5-5.2
S. chloride	110 mmol/l	98-108
Arterial blood gas analysis		
pH	7.277	7.35-7.45
S. bicarbonate	15.8 mmol/L	22-28 mmol/L
PCO2	33.9 mmHg	35-45 mmHg
PO2	150.1 mmHg	>80 mmHg
S. creatinine	0.9 mg/dl	0.7-1.2 mg/dl
Urine pH	7.5 [basal] 7.0 [after acid load test]	
ANA	5.57 S/C	<0.8 S/C
Anti-dsDNA	12.87 IU/ml	<24 IU/ml
Anti-SSA	140 U/ml	<18 U/ml
Anti-SSB	40.7 U/ml	<18 U/ml
Anti- Sm	5.4 U/ml	<18 U/ml
Anti-RNP	6.3 U/ml	<18 U/ml
Anti-scl 70	4.8 U/ml	<18 U/ml
Anti-Jo 1	3.7 U/ml	<18 U/ml
Ultrasonogram of whole abdomen	Bilateral nephrocalcinosis	

[Hb = Hemoglobin, MCV= Mean corpuscular volume, MCH = Mean corpuscular hemoglobin, MCHC=Mean corpuscular hemoglobin concentration, RDW= Red blood cell distribution width,
ANA = Anti nuclear antibody, anti ds DNA = anti-double stranded DNA, anti-SSA = anti-Sjogren's syndrome type A, anti- SSB = anti-Sjogren's syndrome type B, anti-Sm = anti-smooth muscle antibody, anti- RNP = anti-ribonucleoprotein (Anti-RNP), anti-scl 70 = anti- topoisomerase I]

of the abdomen showed extensive bilateral nephrocalcinosis (Figure-2). Excision biopsy of the supraclavicular lymph node showed reactive lymphadenitis.

A presumptive diagnosis of primary Sjogren's syndrome was made based on the presence of three (classic sicca symptoms, positive Schirmer's test, positive anti-Sjogren's syndrome antibodies) out of six American-European Consensus classification criteria. [1]. Renal complications included distal RTA and nephrocalcinosis. Accordingly, she was prescribed six cycles of pulse cyclophosphamide and prednisolone to treat the primary disease and halt further renal progression.

Potassium, spironolactone and sodium bicarbonate were given for RTA. She was discharged in apparently good health and advised for regular follow up.

Discussion

Sjogren's syndrome is a rare autoimmune condition with chronic inflammation of exocrine organs such as lacrimal and salivary glands typically resulting in the characteristic symptoms of dry eyes and dry mouth. Extraglandular manifestations of this immune process can affect the kidneys, liver, lungs, pancreas, nervous system and skin [5]. Though renal involvement in Sjogren's syndrome is rare, it is one of the most commonly confronted extraglomerular manifestations. Whilst tubulointerstitial nephritis (TIN) is the most common histological finding in Sjogren's syndrome [6], distal RTA presenting as hypokalemia has been scarcely reported to date.

Renal involvement is uncommon in Sjogren's syndrome. When present, it is mainly due to TIN [6] and manifests as isolated hypokalemia, isolated acidosis, both proximal and distal RTA and nephrocalcinosis [7]. A study done by Ren et al on 130 patients with primary Sjogren's showed that distal RTA was present in as many as 70% of the patients [8]. Although RTA is associated with Sjogren's syndrome, hypokalemia due to RTA is rarely the main and sole presenting feature of this disorder. Furthermore, RTA is usually asymptomatic in patients with Sjogren's syndrome [3]. A case report published by Rajput et al showed that nephrocalcinosis can also be the rare presenting manifestation of Sjogren's syndrome [2].

Our patient presented with a long history of recurrent hypokalemic paralysis, which preceded typical symptoms of Sjogren's syndrome. She also had nephrocalcinosis. Despite these renal involvements, her serum creatinine level was normal. She was finally diagnosed with primary Sjogren's syndrome based on the presence of sicca symptoms and antibodies, which developed six years after her initial presentation with hypokalemia.

This case demonstrates that symptomatic hypokalemia due to RTA can be a presenting feature of Sjogren's syndrome, even before the appearance of typical sicca syndrome. To date the association between hypokalemic RTA and Sjogren's syndrome has not been emphasized enough. Since renal involvement is rare and not included in the diagnostic criteria; and majority of patients present with typical sicca symptoms, diagnosis of Sjogren's syndrome in patients presenting with RTA may be missed. Thus individuals with RTA should be investigated for Sjogren's syndrome.

Author contributions

SI, NN, SKS, MFUR, KMM and MAKA diagnosed and managed the case. SI, NNA and TH wrote the manuscript. TH and MAKA edited the manuscript.

Informed consent: Patient provided consent for publication.

Conflict of interest: The authors did not have any conflict of interest.

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