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Case Report

Secondary Sjogren's syndrome presenting with hypokalemic periodic paralysis

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Renal tubular acidosis (RTA) may develop in a large population of patients with Sjogren's syndrome (SS), but most of the subjects are asymptomatic. Here, we report a patient with known rheumatoid arthritis and symptoms of xerostomia, xerophthalmia and periodic paralysis. SS should be considered as a cause of RTA. The treatment of the underlying disorder may ameliorate the symptoms.

INTRODUCTION

Renal tubular acidosis (RTA) develops in a large population of patients with Sjogren's syndrome (SS), but most of the subjects are asymptomatic [1]. We report a patient with known rheumatoid arthritis (RA) who was suffering from xerostomia, xerophthalmia and periodic paralysis; admitted with complaint of progressive weakness.

CASE REPORT

A 47-year-old woman was admitted to the emergency department of Imam Khomeini Hospital Complex with progressive weakness. Present illness started 5 days before admission. She was a known case of RA with positive rheumatoid factor (RF) and involvement of wrists, metacarpophalangeal and proximal interphalangeal joints for >3 years. She had a history of foreign body sensation in her eyes, dry mouth and recurrent muscular weakness for the past year. The patient denied vomiting and intake of diuretics, alcohol or laxatives.

On physical examination, vital signs were in normal range and significant findings were deformities in the small joints of both hands, weakness of proximal and distal muscles of all extremities (2/5 and 3/5, respectively). Patellar and Achilles reflexes were slightly diminished. Laboratory results are summarized in Table 1.

Sinus bradycardia with decreased amplitude in T waves, mild ST depression and presence of subtle U waves were apparent on the electrocardiogram (Fig. 1).

Hyperchloremic metabolic acidosis in our patient who had a urine pH greater than 7 and negative urine culture with no history of diarrhea, vomiting or diuretic usage suggested RTA. In regard to the history of xerostomia and xerophthalmia from 1 year ago in a known case of RA, we investigated for SS. Anti-nuclear antibody was 1/320 (positive), RF = 100 U/ml (normal range: 40–60 U/ml), anti-SSA/Ro autoantibodies >200 RU/ml (normal value <20 RU/ml) and anti-SSB/La autoantibodies >181 RU/ml (normal value <20 RU/ml). The Schirmer test was 2.5 mm in 5 min (positive result). Based on revised international classification criteria for SS [2] and negative HCV and HIV antibodies, no past history of head and neck irradiation, lymphoma, sarcoidosis and anticholinergic drug usage, diagnosis of SS was confirmed. Our patient refused salivary gland biopsy, but her unstimulated total salivary flow was 1 ml in 15 min (positive result). In regard to her past medical history of RA and clinical and serologic findings, secondary SS was diagnosed.

She was taking prednisolone 5 mg daily, hydroxychloroquine 400 mg daily and MTX 7.5 mg weekly. Potassium

Table 1: Laboratory findings of the patient

Serum Na	146 (135–145) mEq/l	Creatinine	1.1 (0.6–1.1) mg/dl
Serum K	2.7 (3.5–5) mEq/l	Erythrocyte sedimentation rate	21 mm/h
Serum Ca	8.8 (8.6–10.2) mg/dl	Hemoglobin	13.6 mg/dl
Serum P	3.4 (2.5–4.5) mg/dl	Albumin	4.5 g/dl
Serum Mg	2.3 mg/dl	Alanine aminotransferase	34 (20–40) IU/l
		Thyroid stimulating hormone	2 (0.5–4.5) mIU/l
Serum pH	7.26	Serum Cl	122 (96–106) mEq/l
		Urine Cl	28 (95–105) mEq/l
PCO_2	26 mmHg	Urine Na	23 (>20) mEq/l
		Urine K	27 (25–100) mEq/l
Serum HCO_3	11.7 mmol/l	Urine pH	7
PO_2	90 mmHg	Urine osmolality	320 mOsmol/l
O_2 sat	93%	Urine culture	Negative
		Serum anion gap[a]	12.3 (7–13) mEq/l
Serum urea	22 mg/dl	Urine anion gap[b]	22 (20–90) mEq/l

[a]Serum anion gap = Na − (Cl + HCO_3).
[b]Urine anion gap = (Na + K) − Cl.

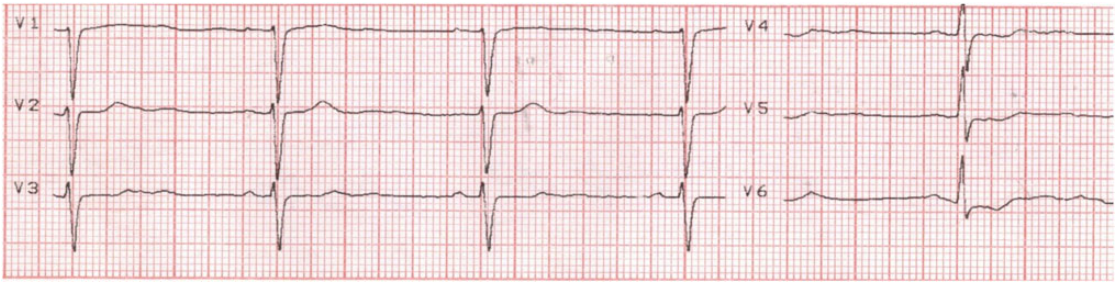

Figure 1: Electrocardiogram changes in the patient.

chloride two tablets daily were added to the above regimen and the dose of prednisolone was increased to 15 mg daily to control the RTA. The level of serum potassium was increased and her weakness was gone after the replacement therapy.

DISCUSSION

The presentation of hypokalemic periodic paralysis, hyperchloremic metabolic acidosis and alkaline urine pH all pointed to the diagnosis of distal RTA.

Distal RTA can occur as a primary disorder or a secondary complication of other medical disease, such as autoimmune diseases, chronic hepatitis and transplant rejection. In autoimmune diseases, relationship between SS with distal RTA is well established [3]. Our patient suffered from RA for >3 years, but the hypokalemic paralysis and symptoms related to SS begun later (almost at the same time period). We concluded that RTA was secondary to SS.

Patients with distal RTA commonly present with symptoms of renal stones, but the first abnormality might be the symptoms of hypokalemia and hypocalcemia. Thirty to 40% of patients with SS have symptomatic and asymptomatic renal involvement [4]. However, severe symptomatic hypokalemia is rare [5].

The common cause of distal RTA is diminished H-ATPase activity [6], as a result urine pH is persistently >5.5. Hypokalemia in RTA is correlated with reduced distal proton (H+) secretion [7]. The presence of alkaline urine in our patient enhanced potassium excretion. Immunohistochemical analysis of renal biopsy in patients with SS showed complete absence of H-ATPase pumps in the intercalated cells; however, the immunologic mechanism of this injury is unknown [8]. The pathophysiology of RTA in SS has been debated in the literature. Although the most common histological renal lesion is interstitial nephritis, it is unclear whether renal tubular defects are the direct result of the interstitial inflammatory process or not. Studies have suggested that lymphocytic and plasma cell infiltrates surrounding renal tubules are associated with and may cause a renal tubular defect [9]. It is thought that hypergammaglobulinemia also might cause distal renal tubular dysfunction [10]. As our patient did not have any renal dysfunction, renal biopsy was not performed.

The presence of severe RTA causing hypokalemic periodic paralysis in SS reflects that the severity of the underlying autoimmune tubulointerstitial nephritis and steroids are used to prevent relapses of hypokalemic paralysis [11]. Steroid therapy, which was successful in a few cases, should be considered in hypokalemic paralysis cases who are nonresponsive to replacement therapy and in those with frequent attacks [1].

Our patient had symptomatic improvement with steroid and potassium replacement therapy, and symptoms did not recur after 1-year follow-up.

CONCLUSION

Autoimmune tubulointerstitial nephritis might lead to overt or latent RTA, which is a frequent extraglandular finding in SS patients [11]. As in some previously reported cases, hypokalemia led to the diagnosis of RTA and SS, although in most of them, some glandular manifestations of SS were presented [1]. The reported case presented with symptoms of hypokalemic periodic paralysis and sicca symptoms for a long time, not severe enough to make her to seek medical care. Therefore, autoimmune disorders, especially SS, should be considered as a cause of distal RTA.

CONFLICT OF INTEREST STATEMENT

None declared.

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