Hypokalemic Paralysis as an Initial Presentation of Sjogren Syndrome

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Abstract

Sjogren syndrome (SS) is a systemic autoimmune disorder with predominant exocrine gland involvement leading to sicca symptoms. Among extraglandular manifestations, renal disease is the most common. Tubular interstitial nephritis and renal tubular acidosis (RTA) are the common presentations. Mild hypokalemia associated with distal RTA is common in SS, however, severe hypokalemia causing paralysis is unusual. We report the case of a 26-year-old female who presented with hypokalemic paralysis. On evaluation, distal RTA was diagnosed. Further evaluation showed positive SS-a/SS-b antibodies in high titer, which confirms the diagnosis of primary SS. Our report illustrates that SS is a rare but important cause of hypokalemic paralysis.

Keywords: Hypokalemic paralysis, renal tubular acidosis, Sjogren syndrome

Résumé

syndrome de Sjogren (SS) est une maladie auto-immune systémique avec une atteinte prédominante des glandes exocrines entraînant des symptômes de sicca. Parmi manifestations extraglandulaires, la maladie rénale est la plus courante. La néphrite interstitielle tubulaire et l’acidose tubulaire rénale (RTA) sont les présentations. Une hypokaliémie légère associée à un RTA distal est courante dans les SS, cependant, une hypokaliémie sévère provoquant une paralysie est inhabituelle. Nous rapportons le cas d’une femme de 26 ans qui présentait une paralysie hypokaliémique. À l’évaluation, un RTA distal a été diagnostiqué. Plus loin l’évaluation a montré des anticorps SS-a / SS-b positifs à titre élevé, ce qui confirme le diagnostic de SS primaire. Notre rapport montre que SS est un cause rare mais importante de paralysie hypokaliémique.

Mots-clés: paralysie hypokaliémique, acidose tubulaire rénale, syndrome de Sjogren

Introduction

Chronic interstitial nephritis is the most common renal presentation in Sjogren syndrome (SS). The clinical manifestations of interstitial nephritis are variable, which include Fanconi syndrome, distal renal tubular acidosis (RTA), nephrogenic diabetes insipidus, or mild asymptomatic hypokalemia. RTA in SS is usually mildly symptomatic, observed in up to 25% of patients.[1] Our patient had one episode of similar weakness with mild hypokalemia which recovered with potassium supplementation, and the patient was diagnosed as hypokalemic periodic paralysis. Renal involvement can occur in SS even before sicca symptoms. Although hypokalemic paralysis as a complication of RTA is rare, we highlight hypokalemic paralysis as an initial presenting symptom in a patient with SS.

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CASE REPORT

A 26-year-old female presented to the emergency department with chief complaints of acute-onset weakness of all the four limbs for the last 3 days. The weakness was progressive, and she was not able to walk at the time of hospitalization. There was one episode of similar weakness 15 days back, for which she was admitted and treated with potassium supplementation. There was no other significant illness in the past. There was no history of vomiting, diarrhea, fever, or altered behavior. Drug history was not significant. General physical examination revealed mild pallor. Neurological examination revealed motor weakness (power 2/5 in the lower limbs and 3/5 in the upper limbs). Deep tendon reflexes were diminished in all the four limbs. There was no sensory deficit. Rest of the systemic examination was unremarkable. Her laboratory analysis revealed normal anion gap hyperchloremic metabolic acidosis [Table 1].

In the absence of gastrointestinal loss or diuretic use, we suspect RTA as a probable cause of metabolic acidosis. Urinalysis revealed pH 7.45, which was failed to lower with ammonium chloride test (0.1 g/kg), which further consolidates our diagnosis of distal RTA. Ultrasonography abdomen was unremarkable, and there was no evidence of obstructive uropathy. On further evaluation of distal RTA, her autoimmune profile showed positive antinuclear antibody and anti SS-a/SS-b antibodies in high titer. Thyroid functions were normal, and viral markers were also negative (hepatitis B, C, and HIV). After ruling out the other causes, the possibility of SS was considered. Our patient was admitted to have dry eyes and dry mouth for the last 6 months. Schirmer test was also positive (4 mm in the right eye/5 mm in the left eye). A final diagnosis of hypokalemic paralysis with distal RTA was made. She was started on potassium chloride (intravenous) and sodium bicarbonate. After 1 week, her muscle power recovered completely with normalization of all laboratory parameters. At discharge, aldosterone activity secondary to volume contraction results in increased renal potassium excretion.

In the cases of life-threatening hypokalemic paralysis, treatment should be focused on potassium correction, rather than metabolic acidosis. In addition, alkali therapy might aggravate the hypokalemia by shifting potassium into the cells and bicarbonaturia. Corticosteroids and other immunosuppressants are reserved for rapidly declining renal function.[3] One previous report had described the recurrence of RTA within 6 months of steroids,[11] however, successful treatment was reported in other case reports.[7,8]

Table 1: Biochemical and hematological investigations

| Test                | Value                          |
|---------------------|--------------------------------|
| CBC                 | Hb - 12.6 g/dL, TLC - 12.3±10/L, Plt - 265×10⁹/L |
| Serum electrolytes  | Sodium - 135 mmol/L, potassium - 1.9 mmol/L, chloride - 115 mmol/L |
| ABG                 | pH - 7.10, pCO₂ -37 mmHg, HCO₃ -10 mmol/L, potassium - 2.01 mmol/L, serum osmolality - 298 mosm/kg |
| Urinalysis          | pH - 7.45, urinary Na⁺ - 144.0 mmol/L, K⁺ - 8.4 mmol/L, urine chloride - 65 mmol/L, urine osmolality - 315.3 mosm/kg |
| Serum anion gap     | 10 mmol/L                      |
| Urinary anion gap   | 89.4 mmol/L (positive)         |
| LFT                 | ALT - 38 U/L, AST - 41 U/L, total protein/albumin - 6.9/3.8 mg/dL |
| KFT                 | Urea - 34 mg/dL, creatinine - 1.12 mg/dL |
| Viral markers       | HIV - negative, HBsAg - negative, Anti-HCV - negative |
| Autoimmune profile  | ANA - 1:1250 fine speckled, anti-SS-a - 489 IU/ml, SS-b - 356 IU/ml |

ANA=Antinuclear antibodies, HB=Hemoglobin, HCV=Hepatitis C virus, AST=Aspartate aminotransferase, ALT=Alanine aminotransferase, CBC=Complete blood count, ABG=Arterial blood gas, LFT=Liver function test, KFT=Kidney function test, Plt=Platelet, TLC=Total leukocyte count, HCV=Hepatitis C virus
CONCLUSION
Mild asymptomatic renal disease is common in SS, although hypokalemic paralysis and at times respiratory failure can be an initial presentation. It is imperative to keep the possibility of SS as a differential while evaluating a patient with hypokalemic paralysis.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

REFERENCES
1. Poux JM, Peyronnet P, Le Meur Y, Favereau JP, Charmes JP, Leroux-Robert C. Hypokalemic quadriplegia and respiratory arrest revealing primary Sjögren’s syndrome. Clin Nephrol 1992;37:189-91.
2. Ren H, Wang WM, Chen XN, Zhang W, Pan XX, Wang XL, et al. Renal involvement and followup of 130 patients with primary Sjögren’s syndrome. J Rheumatol 2008;35:278-84.
3. Maripuri S, Grande JP, Osborn TG, Fervenza FC, Matteson EL, Donadio JV, et al. Renal involvement in primary Sjögren’s syndrome: A clinicopathologic study. Clin J Am Soc Nephrol 2009;4:1423-31.
4. Cohen EP, Bastani B, Cohen MR, Kohner S, Hemken P, Gluck SL. Absence of H(+-)ATPase in cortical collecting tubules of a patient with Sjögren’s syndrome and distal renal tubular acidosis. J Am Soc Nephrol 1992;3:264-71.
5. Bastani B, Haragsim L, Gluck S, Siamopoulos KC. Lack of H-ATPase in distal nephron causing hypokalaemia distal RTA in a patient with Sjögren’s syndrome. Nephrol Dial Transplant 1995;10:908-9.
6. Takemoto F, Hoshino J, Sawa N, Tamura Y, Tagami T, Yokota M, et al. Autoantibodies against carbonic anhydrase II are increased in renal tubular acidosis associated with Sjögren syndrome. Am J Med 2005;118:181-4.
7. Soy M, Pamuk ON, Gerenli M, Celik Y. A primary Sjögren’s syndrome patient with distal renal tubular acidosis, who presented with symptoms of hypokalemic periodic paralysis: Report of a case study and review of the literature. Rheumatol Int 2005;26:86-9.
8. Yılmaz H, Kaya M, Özbek M, ÜUreten K, Safa Yıldırım İ. Hypokalemic periodic paralysis in Sjögren’s syndrome secondary to distal renal tubular acidosis. Rheumatol Int 2013;33:1879-82.
9. Garza-Alpírez A, Arana-Guijarro AC, Esquivel-Valerio JA, Villarreal-Alarcón MA, Galarza-Delgado DA. Hypokalemic paralysis due to primary Sjögren syndrome: Case report and review of the literature. Case Rep Rheumatol 2017;2017:7509238.
10. Sarma A. Hypokalemic paralysis due to primary Sjogren syndrome. Indian J Endocrinol Metab 2018;22:287-9.
11. Sarah S, Lijo G, Sukanya E, Rajasekaran D. Renal tubular acidosis due to Sjögren’s syndrome presenting as hypokalemic quadriparesis: A report of two cases. Indian J Nephrol 2015;25:386-7.