ASIAN JOURNAL OF PHARMACEUTICAL AND CLINICAL RESEARCH

INNOVARE ACADEMIC SCIENCES
Knowledge to Innovation

Vol 9, Issue 4, 2016

Online - 2455-3891
Print - 0974-2441

Case Report

HYPOKALEMIC PERIODIC PALSY AS THE PRIMARY PRESENTATION OF SJOGREN'S SYNDROME

SRI RAMULU KADILAYA[1], KARTHIK RAO N[1], NAVIN PATIL[2]*, RAGHAVENDRA RAO[1], TALHA AHMED[3], ANUSHA VENKATESH[3], GEORGE VARGHESE[3]

[1]Department of Medicine, Kasturba Medical College, Manipal University, Manipal, Karnataka, India. [2]Department of Pharmacology, Kasturba Medical College, Manipal University, Manipal, Karnataka, India. [3]Undergraduate, Kasturba Medical College, Manipal University, Manipal, Karnataka, India. Email: navin903@gmail.com

Received: 10 May 2016, Revised and Accepted: 23 May 2016

ABSTRACT

The most common presentations of Sjogren's syndrome include dryness of eyes, oral cavity, and features of systemic scleroderma. When a patient walks in with such classical features, it becomes easy for the clinician to diagnose and treat the patient. However, Sjogren's syndrome may present atypically as experienced in the present case. Here, the authors present a case of Sjogren's syndrome, which presented as hypokalemic periodic palsy, secondary to distal renal tubular acidosis.

Keywords: Scleroderma, Potassium, Sicca syndrome, Renal tubular acidosis.

INTRODUCTION

Sjogren's syndrome is a chronic autoimmune disorder with systemic involvement. It is believed to be 9-10 times more common in the female sex. Worldwide prevalence lies between 0.1% and 4.8%. Although the disease most commonly affects the exocrine glands, almost any systemic organ may be affected. The sicca (dryness) component is the most frequent presentation, although it is mild and clinically insignificant in many an occasion [1].

Rare and more severe presentations include vasculitis, glomerulonephritis, and neurological involvement [2]. A very rare presentation that has reported a few times in literature is distal renal tubular acidosis (RTA) that can occur as a complication of Sjogren's syndrome. Distal RTA that may lead to hypokalemia, which could result in periodic paralysis, is a severe presentation that needs intensive care and management [3].

Hypokalemia is very common among elderly and hospital-bound patients [4]. Hypokalemic periodic paralysis (palsy) (HPP) usually presents as an acute flaccid paralysis secondary to decreased serum potassium levels. This decrease may be due to the intracellular shift of potassium ions or due to loss of potassium ions (either through renal or gastric routes). There are two types: Familial and acquired. HPP has been known to be associated with RTA, thyrotoxicosis, dengue fever, etc. [5].

CASE REPORT

A 30-year-old female patient (written informed consent obtained) who presented with polyarthritis was admitted in the emergency ward as she complained of sudden onset weakness of all four limbs (without sensory involvement) since the previous day. On further elicitation of her past history, it was revealed that the patient had had four episodes of severe flaccid muscle paralysis involving both proximal and distal muscle groups, in the last 10 years. During each episode of weakness, her serum potassium levels were found to be low. Between the paralytic episodes, the patient functioned well without any therapy and had normal potassium levels. There was no family history of muscle diseases.

During the current admission, the patient's vital signs were within normal limits. Differential diagnoses of acute inflammatory demyelinating polyradiculoneuropathy and HPP were under consideration. The patient's serum potassium level was 2.4 mmol/L, which is well below normal. Furthermore, her electrocardiography (ECG) showed prominent U-waves, which is classically seen in cases of hypokalemia. Her serum sodium level was normal (141 mmol/L). Further, an arterial blood gas analysis revealed a blood pH of 7.1, signifying acidosis. This decrease in pH, along with an elevated transtubular potassium gradient ushered in the diagnosis of distal RTA, with HPP as its complication.

On further evaluation, it was discovered that the patient had a mild sicca component associated, in the form of mild keratoconjunctivitis sicca and xerostomia. The data presented in this report support the view that the paralytic episodes were due to hypokalemia secondary to RTA associated with Sjogren's syndrome. A final diagnosis of complicated Sjogren's syndrome was made. The patient was started on low dose oral corticosteroids and potassium supplements, following which she has improved symptomatically.

DISCUSSION

In our patient, the presentation was classical of HPP, as evidenced by the serum levels, ECG and response to potassium supplementation. The etiology of HPP was the brainteaser in the case. Hypokalemia paralysis is a well-known, albeit rare, complication of severe distal RTA from any cause [6,7]. Distal RTA may be primary or more commonly secondary to paraproteinemia, medullary sponge kidney, nephrocalcinosis, obstructive uropathy, and autoimmune disease. Hypokalemia due to distal RTA is usually a late manifestation [8].

Sjogren's syndrome manifesting for the first time as HPP caused by distal RTA is a very rare presentation but has been reported in <2% cases of Sjogren's syndrome as a presenting manifestation in the available literature. Management includes potassium supplements, corticosteroids, and cytotoxic agents, if necessary [3,9,10].

CONCLUSION

To conclude, we would like to emphasize on the fact that hypokalemia should not just be corrected but also investigated for the cause. Sjogren's syndrome presenting as HPP is very rare, but it should be borne in mind when investigating a case of HPP secondary to distal RTA.

REFERENCES

1. Mavragani CP, Moutsopoulos HM. Sjögren syndrome. CMAJ 2014;186(15):E579-86.
2. Patel R, Shahane A. The epidemiology of Sjögren's syndrome. Clin Epidemiol 2014;6:247-55.
3. Ataoglu EH, Demir B, Tuna M, Cavus B, Cetin F, Temiz LU, *et al.* Sjögren syndrome presenting with hypopotassemic periodic paralysis due to renal tubular acidosis. Am J Case Rep 2012;13:187-90.
4. Patil N, Rao NK, Balaji O, Hande HM, Rao R, Varghese G. Indapamide induced SIADH with hypokalaemia: A case report. Res J Pharm Biol Chem Sci 2016;7(3):2693-6.
5. Garg RK, Malhotra HS, Verma R, Sharma P, Singh MK. Etiological spectrum of hypokalemic paralysis: A retrospective analysis of 29 patients. Ann Indian Acad Neurol 2013;16(3):365-70.
6. Rowbottom SJ, Ray DC, Brown DT. Hypokalemic paralysis associated with renal tubular acidosis. Crit Care Med 1987;15(11):1067-8.
7. Chang YC, Huang CC, Chiou YY, Yu CY. Renal tubular acidosis complicated with hypokalemic periodic paralysis. Pediatr Neurol 1995;13(1):52-4.
8. Bossini N, Savoldi S, Franceschini F, Mombelloni S, Baronio M, Cavazzana I, *et al.* Clinical and morphological features of kidney involvement in primary Sjögren's syndrome. Nephrol Dial Transplant 2001;16(12):2328-36.
9. Khandelwal D, Bhattacharya S, Khadgawat R, Kaur S, Tandon N, Ammini AC. Hypokalemic paralysis as a presenting manifestation of primary Sjögren's syndrome: A report of two cases. Indian J Endocrinol Metab 2012;16(5):853-5.
10. Palkar AV, Pillai S, Rajadhyaksha GC. Hypokalemic quadriparesis in Sjogren syndrome. Indian J Nephrol 2011;21(3):191-3.