A case of acquired Gitelman syndrome presenting as hypokalemic paralysis

M. Kulkarni, P. Kadri, R. Pinto
Department of Nephrology, Father Muller Medical College, Mangalore, Karnataka, India

ABSTRACT

We report a case of a young female patient who presented with weakness of upper and lower limbs. On evaluation, she had hypokalemia, hypomagnesemia, metabolic alkalosis and hypocalciuria. Anti-Ro (SSA) antibody was positive. She had an acquired Gitelman syndrome due to primary Sjögren's syndrome (SS). SS presenting with features of Gitelman syndrome is very rare.

Key words: Acquired Gitelman syndrome, hypokalemia, Sjögren's syndrome

Introduction

Renal tubular involvement in Sjögren's syndrome (SS) usually manifests with Fanconi syndrome, distal (type 1) renal tubular acidosis (RTA), nephrogenic diabetes insipidus, and hypokalemia. SS presenting with features of Gitelman syndrome is rare.

Here, we describe a case a young female with SS who presented with features of Gitelman syndrome and hypokalemic paralysis.

Case Report

A 29-year-old female presented with weakness of both upper and lower limb weakness since 15 days. She denied any history of weakness or muscle cramps before this episode. Past medical history was unremarkable. There was no precedent history of diarrhea and vomiting. There was no history of any diuretic or laxative use. She had a history of dry eyes and dry mouth for the past 6 months.

On examination, her blood pressure was 110/70 mm of Hg. Laboratory evaluation showed hemoglobin - 9.5 g/dl, platelet 2.95 lakh/mm^3. Blood urea nitrogen was 7 mg/dl, serum creatinine was 0.65 mg/dl, serum sodium was 134 mEq/L, serum potassium was 1.81 mEq/L and serum chloride was 84.3 mEq/L. Arterial blood pH was 7.68; serum bicarbonate was 49.5 mEq/L. Serum magnesium was 1.2 mg/dl. Urine routine microscopy was normal. Urine protein creatinine ratio was 0.1. Urine chloride (spot) was 36.7 mEq/L, urine creatinine (spot) 23.5 mg/dl, and urine calcium (spot) 1.23 mg/dl. Ultrasound abdomen was within normal limits. Anti-Ro(SSA) antibody was strongly positive. Schirmer test was 3 mm in 5 min. This patient satisfied the American European consensus criteria for diagnosis of primary Sjögren's syndrome (SS).[1]

She was treated with intravenous potassium, magnesium supplements and steroids. Her symptoms gradually improved. She was also started on spironolactone. Ten days after admission, she was discharged on spironolactone and prednisolone. On follow-up 15 days later, patient was normokalemic and was continued on spironolactone and steroids.

Discussion

This patient presented with muscle weakness and her laboratory reports showed hypokalemia and metabolic alkalosis with normal blood pressure. Her urinary chloride levels were above 20 mEq/L, which ruled out extra-renal causes of metabolic alkalosis. She did not give any history

Address for correspondence:
Dr. Manjunath Kulkarni,
Department of Nephrology, Father Muller Medical College,
Mangalore, Karnataka, India.
E-mail: drmjkulkarni@gmail.com

Access this article online	
Quick Response Code:	Website: www.indianjnephrol.org
	DOI: 10.4103/0971-4065.146031

of diuretic use. The presence of hypomagnesemia and absence of hypercalciuria (urinary calcium creatinine ratio <0.2) ruled out Bartter syndrome. Hypokalemic alkalosis, high urinary chloride, hypomagnesemia, absence of hypercalciuria in background of no history of diuretic use favored the diagnosis of Gitelman syndrome. Her clinical and laboratory data suggested presence of SS as well.

Tubular involvement in SS is usually distal tubular dysfunction, type I (distal) renal tubular acidosis and nephrogenic diabetes insipidus. Proximal tubular abnormalities are less frequent, and rarely Fanconi's syndrome has been reported in patients with SS.[2,3]

Gitelman syndrome is usually an inherited disorder. Acquired Gitelman syndrome is relatively rare. To the best of our knowledge, only five cases of acquired Gitelman syndrome have been reported in English literature so far. Among these five cases, four had SS, one was a case of chronic sialoadenitis.[4-8]

Acquired Gitelman syndrome secondary to SS presenting with hypokalemic weakness is very rare. Only two cases have been reported so far.[5,7]

Though Gitelman syndrome is an inherited disorder, it can be acquired in patients with autoimmune disorders. Given the paucity of reports, we believe that SS presenting as acquired Gitelman syndrome may be relatively rare. Acquired Gitelman syndrome should be considered in differential diagnosis of renal involvement in patients with SS.

References

1. Vitali C, Bombardieri S, Jonsson R, Moutsopoulos HM, Alexander EL, Carsons SE, *et al.* Classification criteria for Sjögren's syndrome: A revised version of the European criteria proposed by the American-European Consensus Group. Ann Rheum Dis 2002;61:554-8.
2. Lin DF, Yan SM, Zhao Y, Zhang W, Li MT, Zeng XF, *et al.* Clinical and prognostic characteristics of 573 cases of primary Sjögren's syndrome. Chin Med J (Engl) 2010;123:3252-7.
3. Aasarød K, Haga HJ, Berg KJ, Hammerstrøm J, Jørstad S. Renal involvement in primary Sjögren's syndrome. QJM 2000;93:297-304.
4. Casatta L, Ferraccioli GF, Bartoli E. Hypokalaemic alkalosis, acquired Gitelman's and Bartter's syndrome in chronic sialoadenitis. Br J Rheumatol 1997;36:1125-8.
5. Chen YC, Yang WC, Yang AH, Lin SH, Li HY, Lin CC. Primary Sjögren's syndrome associated with Gitelman's syndrome presenting with muscular paralysis. Am J Kidney Dis 2003;42:586-90.
6. Schwarz C, Barisani T, Bauer E, Druml W. A woman with red eyes and hypokalemia: A case of acquired Gitelman syndrome. Wien Klin Wochenschr 2006;118:239-42.
7. Kim YK, Song HC, Kim WY, Yoon HE, Choi YJ, Ki CS, *et al.* Acquired Gitelman syndrome in a patient with primary Sjögren syndrome. Am J Kidney Dis 2008;52:1163-7.
8. Hinschberger O, Martzolff L, Ioannou G, Baumann D, Jaeger F, Kieffer P. Acquired Gitelman syndrome associated with Sjögren's syndrome and scleroderma. Rev Med Interne 2011;32:e96-8.

How to cite this article: Kulkarni M, Kadri P, Pinto R. A case of acquired Gitelman syndrome presenting as hypokalemic paralysis. Indian J Nephrol 2015;25:246-7.

Source of Support: Nil, **Conflict of Interest:** The results presented in this paper have not been published previously in whole or part, except in abstract format.