INTRODUCTION
Hypokalemia is a common disorder in Emergency Department (ED). Hypokalemic paralysis is an important cause of acute flaccid paralysis that may even result in life-threatening cardiac arrhythmia and respiratory paralysis.1 Recognizing the underlying causes of hypokalemic paralysis is essential for the appropriate management of affected patients and prevention of recurrent paralysis attacks.2 Although Renal Tubular Acidosis (RTA) secondary to autoimmune interstitial nephritis develops in a large proportion of patients with Sjogren Syndrome (SS), most of the subjects are asymptomatic.1

Here, we present a case of primary SS without sicca symptoms but with recurrent episodes of hypokalemic paralysis as the initial clinical presentation.

CASE REPORT
A 64-year male patient came with HPP, and was later diagnosed with distal RTA. The patient, who had no xerostomia and xerophthalmia, was diagnosed with primary SS from serologic and histologic findings of minor salivary gland biopsy. The patient recovered after potassium replacement therapy. Renal biopsy was also performed and revealed evidence of tubulointerstitial nephritis. Corticosteroids were administered and there was no recurrence of HPP during a 4-year follow-up period. The case highlights the significance of acute hypokalemia management in emergency department as it can unmask SS even if the SS is not associated with sicca symptoms. Hypokalemic paralysis associated with normal anion gap metabolic acidosis should prompt toward the diagnosis of SS.


Recurrence Hypokalemic Periodic Paralysis Unmasks Sjogren Syndrome without Sicca Symptoms
Yao-Min Hung1,3, Neng-Chyan Huang1,3,4, Shue-Ren Wann1,3, Yun-Te Chang1,3 and Jyh-Seng Wang2,3

ABSTRACT
Hypokalemic Periodic Paralysis (HPP) may occur as a rare complication of Sjogren Syndrome (SS) and Renal Tubular Acidosis (RTA). A 64-year male patient came with HPP, and was later diagnosed with distal RTA. The patient, who had no xerostomia and xerophthalmia, was diagnosed with primary SS from serologic and histologic findings of minor salivary gland biopsy. The patient recovered after potassium replacement therapy. Renal biopsy was also performed and revealed evidence of tubulointerstitial nephritis. Corticosteroids were administered and there was no recurrence of HPP during a 4-year follow-up period. The case highlights the significance of acute hypokalemia management in emergency department as it can unmask SS even if the SS is not associated with sicca symptoms. Hypokalemic paralysis associated with normal anion gap metabolic acidosis should prompt toward the diagnosis of SS.


1 Department of Emergency Medicine / Pathology2, Kaohsiung Veterans General Hospital, Kaohsiung, Taiwan.
3 School of Medicine, National Yang-Ming University, Taipei, Taiwan.
4 Department of Environmental and Occupational Health, National Cheng Kung University, Tainan, Taiwan.

Correspondence: Dr. Jyh-Seng Wang, Department of Pathology, Kaohsiung Veterans General Hospital, 386 Tu-Chung 1st Road, Kaohsiung 813, Taiwan.
E-mail: jswang@isc.vghks.gov.tw

Received: April 17, 2014; Accepted: August 13, 2014.
pattern, and positive anti-La SS(B) antibody. The results of anti-dsDNA antibody, anti-RNP antibody, anticentromere, Scl-70 antibody and anti-Lo SS(A) antibody were all negative. The result of serum complement fractions assay was also normal. The Schirmer’s test for dry eye was positive and sialoscintigraphy with radioisotope Tc-99m-pertechnetate showed class-II xerostomia. Labial gland biopsy showed diffuse lymphocyte infiltration with focal acinar destruction (Figure 1). Renal biopsy confirmed the diagnosis of tubulointerstitial nephritis showing focal interstitial chronic inflammatory cells infiltration associated with tubular atrophy and interstitial fibrosis (Figure 2). The glomerulus was unremarkable.

The diagnosis of primary SS was established and pulse methylprednisolone and cyclophosphamide treatment were prescribed while potassium and alkali supplementation were also given for the distal RTA and symptomatic relief of the hypokalemic paralysis. There was no recurrence of HPP during a 4-year follow-up period.

**DISCUSSION**

Potassium disorders are the most common electrolyte abnormality identified in clinical practice. Presenting symptoms for hypokalemia primarily affect the cardiac, neuromuscular, and gastrointestinal systems. The approach for hypokalemia at ED usually include history taking, physical examination, evaluation of blood pressure, arterial blood gas and urine chemistry analysis. Generally, HPP can occur secondarily to excessive potassium loss. Thyrotoxicosis, diuretic ingestion, hyperaldosteronism, barium poisoning, Gitelman syndrome, and Bartter syndrome are among the disorders causing secondary HPP and enter the list of differential diagnoses. Thyrotoxic HPP has been reported in Asian population. The attack of paralysis is usually precipitated by exertion or heavy meal. Physical examination of patient with thyrotoxic HPP may show normal blood pressure, with thyroid enlargement, thrills, or/hands tremor. However, arterial blood gas will not disclose acidosis or alkalosis. On the contrary, diuretic ingestion will show metabolic alkalosis with volume depletion, and sometimes hypomagnesemia. Clinical presentation of Gitelman syndrome and Bartter syndrome with HPP may include normal blood pressure, hypokalemic alkalosis, hypomagnesemia and secondary hyperaldosteronism. Primary hyperaldosteronism usually manifests in high blood pressure, hypokalemic alkalosis, hyporeninemia, and hyperaldosteronism. Urine analysis will show marked excessive potassium loss.

Recurrent attacks of HPP as initial manifestation of primary SS is rare, but when it occurs it may precede symptoms of dry eyes and dry mouth. HPP is a rare complication of distal RTA and occurrence of HPP during the course of SS has been reported in only a few patients. Soy et al. reviewed 18 cases of SS-associated HPP reported between 1966 - 2004 and found only 4 of them received both renal and lip/lacrimal gland biopsy. We continued a PubMed search about SS-associated HPP reported between 2005 - 2013 which revealed 7 more cases. Including the present case, only 6 cases have biopsy proved tubulointerstitial nephritis. Most of the reported cases received corticosteroids treatment in addition to large doses of potassium chloride supplementation to restore muscle strength.

Among these 26 reported cases of SS-associated HPP, symptoms of HPP were all similar to that of our patient. However, some patients presented with severe respiratory muscle weakness or even paralysis. Interestingly, only 4 (15%) of them (including the present case) were male, with age of 64, 73, 74 and 78 years old respectively. The other 22 (85%) cases were females with age ranging from 15 to 63 years old. Our patient is unique in initial presentation of recurrent HPP without sicca symptoms and finally found to have the diagnosis...
of SS. The present case highlights the significance of acute hypokalemia management in the ED as it can unmask SS even if it is not associated with sicca symptoms.

REFERENCES


