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# An Unusual Case of Acute Progressive Limb Weakness in A 58-Year-Old Female

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**Abstract**

A 58-year-old Asian woman with well controlled hypertension presented to the Emergency Department with acute and progressive bilateral lower limb weakness. Before she could be evaluated for the cause of her symptoms, she went into cardiac arrest. Resuscitation was started immediately and ROSC was attained in about 20 minutes. Investigations revealed severe hypokalemia and metabolic acidosis. Further enquiry revealed a history of 11 kg weight loss over a period of 1 month immediately prior to the acute episode of limb weakness. The constellation of these findings prompted an autoimmune analysis which tested positive for anti SSA/SSB antibodies, confirming her diagnosis of Primary Sjögren's Syndrome (pSS). It was concluded that the distal RTA secondary to Sjogren's syndrome was the cause of severe hypokalemia in our patient. By presenting this case we aim to highlight one of the rare presentations of Sjogren's syndrome.

**Keywords:** Autoimmune, Distal Renal tubular acidosis, Extraglandular, Primary Sjögren's Syndrome

## Introduction

Sjögren syndrome is chronic systemic autoimmune disorder characterized by lymphocytic infiltration of exocrine glands. The disease can present as alone (primary Sjögren syndrome) or associated with other autoimmune conditions such as SLE, rheumatoid arthritis, and scleroderma (secondary Sjögren syndrome). The prevalence of primary Sjögren's syndrome is ~0.5%–1% and middle-aged women (female-to-male ratio is 9:1) are primarily affected.(1) Most patients presents with sicca symptoms such as xerophthalmia (dry eyes), xerostomia (dry mouth), and parotid gland enlargement. In addition, various extraglandular features may develop. This includes arthralgia, arthritis, vasculitis, lymphoma, renal involvement, and Raynaud's phenomenon.(2)Renal involvement includes proximal and distal tubular acidosis, tubular proteinuria, and nephrogenic diabetes

insipidus.[3] The mechanism of hypokalemia in Sjögren syndrome is because of distal RTA brought about by chronic interstitial nephritis which in turn leads to decrease tubular sodium delivery, defective H-KATPase, secondary hyperaldosteronism, and bicarbonaturia.

Hypokalemia is defined as a plasma potassium level below 3.5 mmol/L, is a frequently encountered electrolyte imbalance. Mild cases (3.0–3.5 mmol/L) often show no symptoms, while severe hypokalemia (below 2.5 mmol/L) can lead to paralysis, affecting both limb and respiratory or cardiac system. Distal renal tubular acidosis (RTA), particularly due to Sjögren syndrome, is a key cause of this condition. The following case report depicts a scenario. [4]

**Case Description**

A 58 years old female, known case of systemic hypertension was admitted to the emergency department at a Local hospital with a 3 days' history of rapidly progressing weakness of bilateral lower limbs. She had no back pain, leg numbness, urinary or bowel symptoms. However, within minutes of her presenting to the emergency department, before any further enquiry into the cause of her symptoms could be made, she went into cardiac arrest.

Resuscitation was started immediately and ROSC was attained after 20 minutes. She was put on ventilator support and shifted to the Intensive Care Unit. Investigations showed severe hypokalemia, warranting a diagnosis of hypokalemic periodic paralysis as a cause of her acute limb weakness. Further enquiry revealed a history of 11 kg weight loss over a period of 1 month immediately prior to the acute episode of limb weakness. She had consulted a primary care centre for complaints of loss of appetite and weight loss and was given appetite stimulants. No further investigations were done. She was otherwise well, afebrile and with stable vitals until she presented with acute onset progressive weakness in her limbs. On admission her blood gas showed hyperchloremic metabolic acidosis. As there was no evidence of gastrointestinal loss, and her urine pH remained greater than 5.5 despite systemic acidosis, she was diagnosed with hypokalemic paralysis from a severe distal renal tubular acidosis (RTA).

The findings prompted an immunologic work-up which showed a positive ANA and positive antibodies to SSA and SSB, revealing Sjögren syndrome as the underlying cause of distal renal tubular acidosis.

**Discussion**

Sjögren's disease (SjD) is a chronic autoimmune inflammatory disorder characterized by diminished lacrimal and salivary gland function with resultant dryness of the eyes and mouth. In addition, a variety of other disease manifestations affecting multiple organs and organ systems may occur, and the clinical features of Sjögren's disease can be divided into the two broad categories of exocrine glandular features and extraglandular features. [5] Primary Sjögren's syndrome (pSS) primarily involves exocrine glands. A small group of patients have primarily extraglandular manifestations and positive testing for Ro/SSA antibodies without significantly dry eyes or dry mouth. The reported prevalence of kidney involvement in pSS varies widely, ranging from 1 to 33 %. Most studies observed kidney manifestations in approximately 5 to 14 % of patients with pSS. The most common manifestations are interstitial nephritis and cryoglobulinemia-related membranoproliferative glomerulonephritis (MPGN). In one study that reported kidney biopsy results from 95 patients with pSS and kidney disease, the predominant finding was tubulointerstitial nephritis in 73 patients (77

%) and glomerular disease in 22 patients (23 %). [6] The defects in renal tubular function that lead to acid-base and electrolyte abnormalities in patients with pSS generally occur in conjunction with tubulointerstitial nephritis. The term "renal tubular acidosis" (RTA) refers to a group of disorders in which, despite a relatively well-preserved glomerular filtration rate, metabolic acidosis develops because of defects in the ability of the renal tubules to perform the normal functions required to maintain acid-base balance. All forms of RTA are characterized by a normal anion gap (hyperchloremic) metabolic acidosis. [7] This form of metabolic acidosis usually results from either the net retention of hydrogen chloride or a salt that is metabolized to hydrogen chloride (such as ammonium chloride) or the net loss of sodium bicarbonate or its equivalent. . Distal RTA, frequently seen in pSS, is most commonly due to selective failure of activity or expression of the H+-ATPase. The decreased transit through the proton pump inhibits urine acidification and reduces the electrical dissipation of the membrane potential. [8] The latter has been suspected to be a driving force for K+ secretion and eventual potassium wasting. Renal Tubular Acidosis with hypokalemic paralysis as a presenting feature of pSS, without any exocrine gland involvement is described in very few case reports in literature.

**Conclusion**

When a patient presents with hypokalemic paralysis, it is important not to immediately attribute it to periodic paralysis without considering other possible causes. A comprehensive history, detailed clinical examination, and careful evaluation of arterial blood gases and urine analysis are essential to ensure that conditions like Sjögren syndrome are not overlooked. Renal tubular dysfunction can be the presenting manifestation of Primary Sjögren's syndrome and it is important to consider the possible presence of this disorder in adults with otherwise unexplained distal RTA or hypokalemia.

**References**


1. Fox RI. Sjögren's syndrome. Lancet. 2005;366 (9482): 321-331. doi :10. 1016/ S0140-6736(05) 66990-5
2. Bossini N, Savoldi S, Franceschini F, et al. Clinical and morphological features of kidney involvement in primary Sjögren's syndrome. Nephrol Dial Transplant. 2001;16(12):2328-2336. doi:10.1093/ ndt/ 16.12.2328
3. Maripuri S, Grande JP, Osborn TG, et al. Renal involvement in primary Sjögren's syndrome: a clinicopathologic study. Clin J Am Soc Nephrol. 2009;4(9):1423-1431. doi:10.2215/CJN.00980209
4. Sarma A. Hypokalemic Paralysis Due to Primary Sjogren Syndrome. Indian J Endocrinol Metab. 2018;22(2):287-289. doi:10.4103/ ijem.IJEM_ 666_ 17
5. Mihai A, Caruntu C, Jurcut C, et al. The Spectrum of Extraglandular Manifestations in Primary Sjögren's Syndrome. J Pers Med. 2023;13(6):961. Published 2023 Jun 7. doi:10.3390/jpm13060961
6. Talal N. Sjögren's syndrome: historical overview and clinical spectrum of disease. Rheum Dis Clin North Am. 1992;18(3):507-515.
7. Vivino FB. Sjögren's syndrome: Clinical spectrum and management. Rheumatic Disease Clinics of North America. 2016;42(4):419–437.
8. Martinho AL, Capela A, Duarte F. Hypokalemic Paralysis: The First Presentation of Primary Sjögren's Syndrome. Acta Médica Portuguesa. 2012 Mar-Apr;25(2):122–4.