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Case Report

Hypokalemic paralysis in a patient with Sjögren's disease: a rare emergency

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Abstract

Renal tubular acidosis (RTA) is the most common renal manifestation in Sjögren's disease (SjD), typically subclinical but occasionally presenting with severe hypokalemia. A 59-year-old woman with SjD, treated with hydroxychloroquine and pilocarpine, was admitted with sudden quadriparesis, hypotonia, and muscle weakness. Tests showed severe hypokalemia (K^+: 1.6 mEq/L), normal anion gap metabolic acidosis, and alkaline urine. ECG revealed U-waves and ventricular bigeminy. Electrolyte replenishment normalized potassium within 48 hours, resolving ECG abnormalities and improving muscle strength. After transfer to the rheumatology department, she developed slurred speech, borderline hypokalemia, and metabolic acidosis. Brain MRI confirmed central pontine myelinolysis (CPM). Kidney biopsy verified RTA due to tubulointerstitial nephritis. Treatment with methylprednisolone (40 mg/day) and cyclophosphamide pulses normalized acid-base status and led to neurological recovery.

In SjD, autoimmune-mediated tubulointerstitial inflammation impairs hydrogen ion secretion, causing metabolic acidosis and potassium wasting. Hypokalemic paralysis from RTA is a rare, life-threatening complication. CPM, typically linked to sodium fluctuations, may also relate to SjD activity and metabolic disturbances, requiring heightened clinical suspicion for early detection and management, especially as RTA is often subclinical but can manifest with significant electrolyte imbalances.

Keywords

central pontine myelinolysis, hypokalemia, quadriparesis, renal tubular acidosis, rheumatic diseases

List of abbreviations	
CPM: central pontine myelinolysis	**MRI**: magnetic resonance imaging
ECG: electrocardiogram	**RTA**: renal tubular acidosis
HE stain: hematoxylin and eosin stain	**SjD**: Sjögren's disease

Introduction

Sjogren's disease (SjD) is a systemic autoimmune disorder primarily affecting exocrine glands, characterized by lymphocytic infiltration leading to xerostomia and keratoconjunctivitis sicca. The reported prevalence of the disease reaches up to 3.5% of the population but varies substantially among studies, depending on the geographical region

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and the methodology used.[1] SjD is associated with a broad spectrum of organ involvement, with renal manifestations occurring in approximately 5% to 30% of cases.[2] A recently published meta-analysis reviewing 440 cases of SjD with renal involvement found that distal renal tubular acidosis (RTA) accounted for the majority of cases.[3] Distal RTA in SjD arises from autoimmune-mediated injury to the distal renal tubules, impairing acid excretion and facilitating urinary bicarbonate loss. This results in a characteristic metabolic profile: hyperchloremic non-anion gap acidosis, hypokalemia, and alkaline urine pH.[4] Distal RTA is often overlooked, as it frequently remains subclinical and is typically identified only through targeted metabolic evaluation.[2] Distal RTA in SjD can present with nephrocalcinosis.[5] However, in rare instances, potassium loss associated with RTA may become profound, resulting in acute and severe hypokalemia.[6] This can lead to neuromuscular complications such as paralysis and cardiac arrhythmias, thereby constituting a medical emergency.[6] Currently, there are no validated guidelines for the management of RTA in SjD. We present herein a striking case of a successfully treated patient who presented with severe hypokalemic paralysis and arrhythmia in the context of underlying SjD-induced distal RTA, further complicated by central pontine myelinolysis (CPM).

Case presentation

A 59-year-old female patient with primary SjD under maintenance therapy with hydroxychloroquine and pilocarpine presented to the emergency department of the General Hospital of Karditsa (Thessaly, Greece), following a sudden and dramatic onset of generalized muscle weakness. Neurological assessment upon admission revealed quadriparesis with generalized hypotonia and markedly reduced muscle strength in both upper and lower extremities. The patient remained hemodynamically stable and alert, and there was no history of recent trauma or preceding infectious illness. Laboratory investigations revealed profound hypokalemia, with serum potassium concentration at 1.6 mEq/L. Notably, the patient had received laxative preparation the previous day in anticipation of a scheduled colonoscopy. Arterial blood gas analysis showed metabolic acidosis (pH=7.23 and HCO_3=13.4 mmol/L) with a normal anion gap, while urinalysis showed an alkaline urinary pH of 8.0, suggesting distal (type 1) RTA.[7] Electrocardiographic evaluation disclosed the presence of U-wave depressions and ventricular bigeminy.[8] The patient was managed within the cardiology department of the hospital, where intensive intravenous potassium replacement therapy was initiated. Within 48 hours, potassium reached the lower limit of the reference range, electrocardiographic abnormalities resolved, and the patient exhibited nearly normal motor function. Given the strong suspicion of distal RTA associated with SjD, she was transferred to the Department of Rheumatology and Clinical Immunology at the University General Hospital of Larissa (Thessaly, Greece). Upon arrival in the clinic, slurred speech emerged as a new neurological symptom. On brain magnetic resonance imaging (MRI), findings consistent with CPM were identified ***(Fig. 1A)****. The patient continued to display a borderline low potassium level and persistent non-anion gap metabolic acidosis despite ongoing aggressive electrolyte supplementation. Renal biopsy revealed histopathological evidence of tubulointerstitial nephritis, which confirmed the renal involvement of SjD* ***(Fig. 2)****. Corticosteroids were initiated (methylprednisolone 40 mg/day), alongside intravenous cyclophosphamide pulses, to halt the renal insult. Following treatment, a steady and sustained im-*

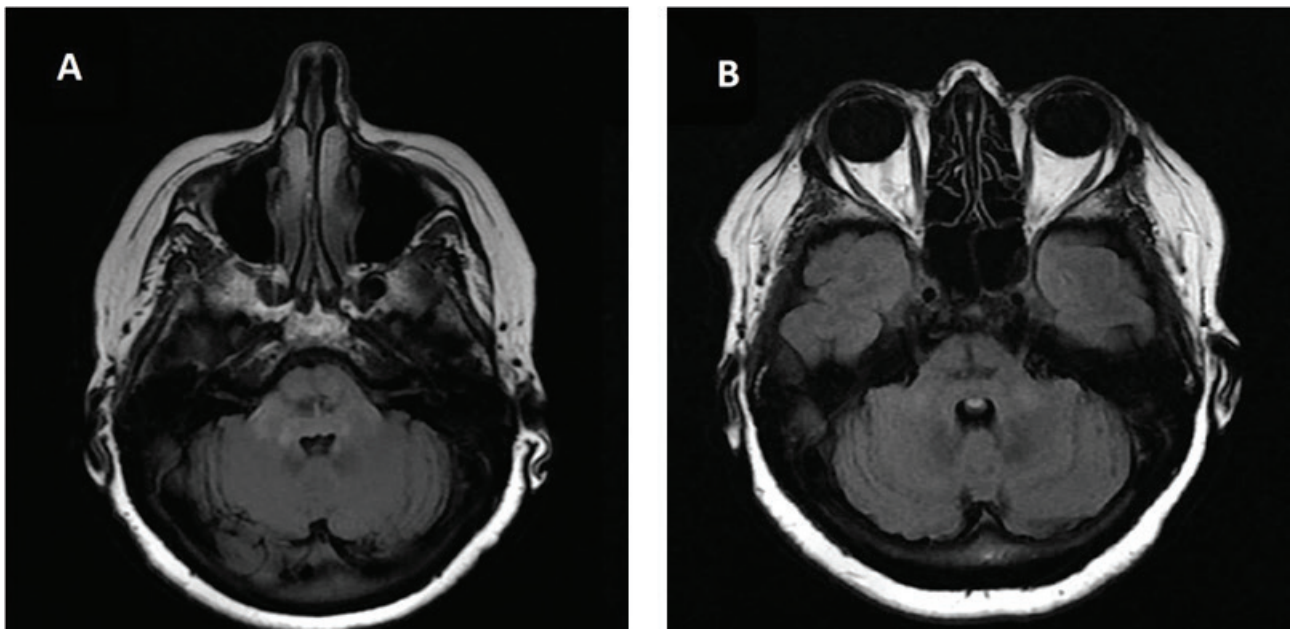

Figure 1. Brain MRI showing **A.** central pontine myelinolysis and **B.** resolution of findings.

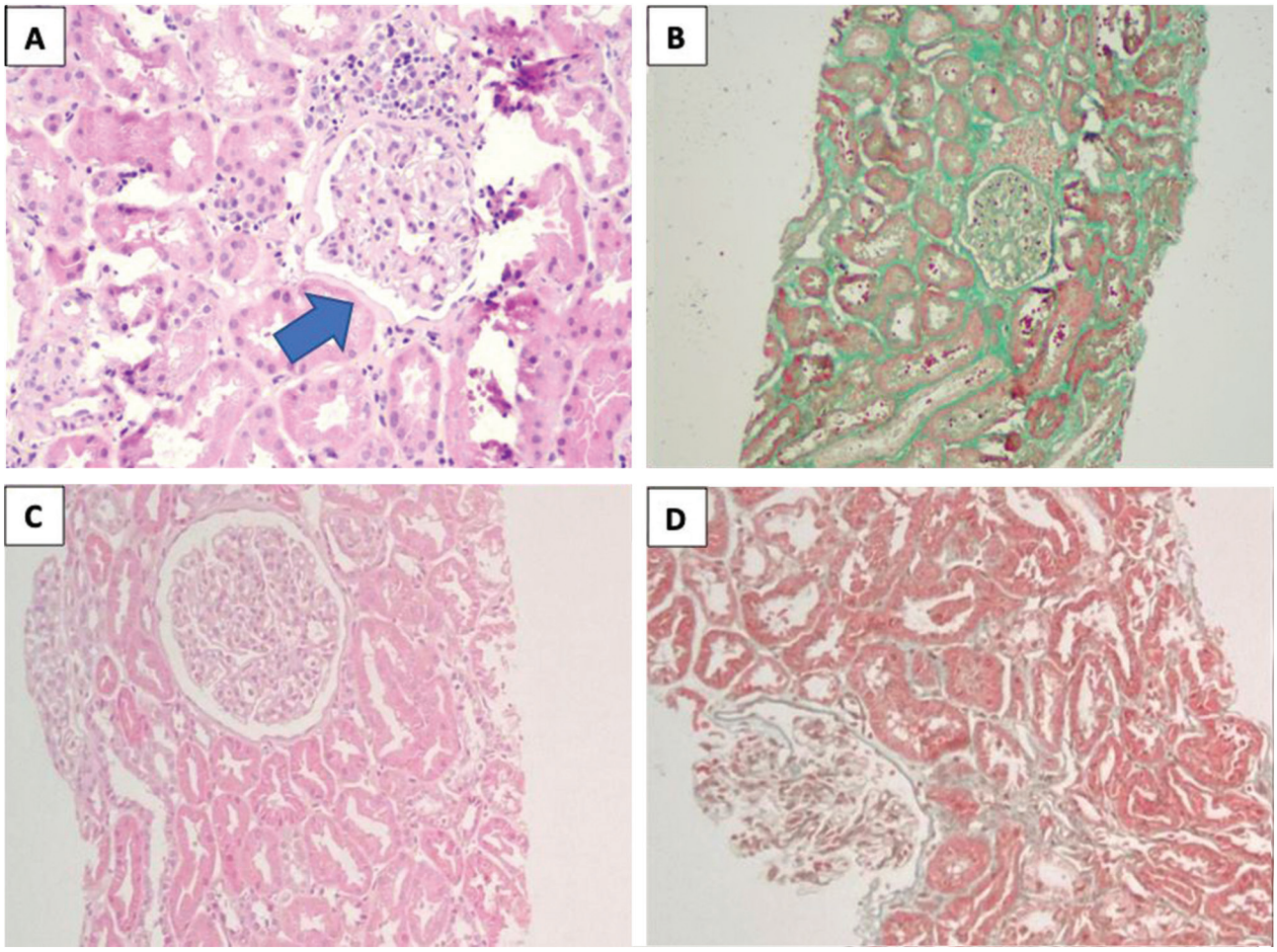

Figure 2. Renal biopsy demonstrating interstitial nephritis with preserved glomerular morphology compared to a normal renal biopsy. **A.** Light microscopy (H&E staining, ×200) demonstrates a morphologically normal glomerulus (arrow), with intact capillary loops and no evidence of mesangial expansion or hypercellularity. The surrounding interstitium shows a patchy infiltrate of mononuclear inflammatory cells, consistent with interstitial nephritis. **B**. Masson trichrome stain (×100) also displaying mild interstitial inflammation along with mild interstitial fibrosis, as indicated by the blue-stained collagen deposition. **C.** Normal renal biopsy (H&E staining, ×200) **D.** Normal renal biopsy (Masson trichrome stain, ×200).

provement was observed in both renal metabolic parameters and neurological symptoms. The patient no longer required electrolyte supplementation, and her serum potassium and acid-base balance normalized. Speech impairment and muscle strength improved significantly, and in a follow-up MRI performed 4 weeks after the initial scan, all abnormalities had resolved ***(Fig. 1B)****.*

Materials and methods

The primary tissue stain used in histopathology is hematoxylin and eosin stain, which is frequently shortened to H&E stain or HE stain. Hematoxylin stains nucleic acids through a complicated, poorly understood reaction, giving them a deep blue-purple hue. Eosin is pink and does not specifically stain proteins. In a typical tissue, nuclei are stained blue, whereas the cytoplasm and extracellular matrix have varying degrees of pink staining. Masson's trichrome is a three-color staining procedure used in histology. It is a histological staining method used to differentiate between cells and collagen fibers in tissue samples. Typically, nuclei are stained blue or black, cytoplasm and muscle red, and collagen blue. Masson's trichrome staining is a widely used histological technique for evaluating kidney biopsies, particularly for assessing the extent of glomerular and interstitial fibrosis.

Discussion

This case delineates a rare but clinically significant complication of SjD, wherein distal RTA presents with acute hypokalemic paralysis, compounded by the occurrence of CPM. The sudden onset of severe hypokalemia in our patient reflects an exacerbation of underlying electrolyte disturbances due to distal RTA, precipitated by the use of laxatives. Hypokalemic paralysis may be the first presentation of

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SjD, even preceding classic sicca symptoms, underscoring the need for clinicians to consider autoimmune causes in unexplained metabolic derangements.[5] In our case, renal involvement in the context of SjD was confirmed through biopsy.

Kidney biopsy should typically be performed when the histopathological findings are anticipated to modify therapeutic decision.[2] The histological examination may be useful in distinguishing inflammatory from fibrotic lesions. As fibrosis is considered a manifestation of chronic tissue scarring, anti-inflammatory or immunosuppressive therapies are unlikely to improve tissue dysfunction and may pose additional toxicity risks. In such cases, patient management would be limited to alkali therapy and potassium supplementation. The kidney biopsy of our patient demonstrated both interstitial inflammation and interstitial fibrosis. The patient was given immunosuppressive therapy to reduce the inflammation, which might not have worked if interstitial fibrosis had been the only discovery. To date, evidence-based recommendations for the management of RTA in SjD are lacking. Current therapeutic strategies include the use of corticosteroids, mycophenolate mofetil, cyclophosphamide, and B-cell depletion therapies such as rituximab.[2] A similar case regarding a patient with SjD and interstitial nephritis with fibrosis and inflammation findings in a renal biopsy was treated successfully with steroids and cyclophosphamide.[9] Our patient initially received moderate-to-high doses of corticosteroids, which resulted in a reduced need for electrolyte supplementation. Following the initiation of cyclophosphamide pulse therapy, we were able to taper the corticosteroid dosage and ultimately discontinue electrolyte supplementation.

The occurrence of CPM in this patient likely resulted from rapid osmotic fluctuations during aggressive correction of electrolyte abnormalities. Although more commonly associated with rapid sodium correction, CPM has also been reported in the context of SjD activity and systemic inflammation, suggesting that the autoimmune milieu may have synergistically contributed to the demyelinating process.[10] Very few cases of both CPM and RTA as presentations of SjD have been described in the literature so far. The reported data raise concerns that osmotic demyelination can be precipitated even without rapid sodium correction, particularly when autoimmune and metabolic stressors coexist, and draw a direct link between systemic autoimmunity and pontine vulnerability.[10-15] The identification of CPM in our case highlights not only the importance of cautious electrolyte correction strategies but also the potential vulnerability of the central nervous system in the context of systemic autoimmune diseases.

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Additional information

Ethical statement

- The authors declared that no clinical trials were used in the present study.
- The authors declared that no experiments on humans or human tissues were performed for the present study.
- The authors declared that no informed consent was obtained from the humans, donors or donors' representatives participating in the study.
- The authors declared that no experiments on animals were performed for the present study.
- The authors declared that no commercially available immortalized human and animal cell lines were used in the present study.

Conflict of interest

The authors have declared that no competing interests exist.

Artificial Intelligence (AI) use

The authors accept full responsibility for the content of the manuscript, including the disclosure of any use of AI. No AI tools were used in the preparation of this manuscript.

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Author contributions

NML and AG wrote the main manuscript and created/edited the figures. FK assessed kidney biopsy and provided the respective figure. CK and TS corrected the manuscript. CK, TS, and DB conceived and supervised the study. All authors reviewed the manuscript.

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Data availability

All of the data that support the findings of this study are available in the main text.