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A CASE REPORT ON HYPOKALEMIC PARALYSIS AS A RARE PRESENTATION OF KIDNEY INVOLVEMENT IN PRIMARY SJOGREN'S SYNDROME.

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ABSTRACT

Background:

Primary Sjogren's syndrome (pSS) is an autoimmune disorder that primarily affects exocrine glands, commonly causing dry eyes and a dry mouth (sicca component). It can also involve non-exocrine organs, including the skin, lungs, gastrointestinal tract, central and peripheral nervous system, musculoskeletal system, and kidneys.

Case presentation:

A 21-year-old female presented with sudden quadriparalysis and normal vital signs, ruling out common causes. Clinical examination revealed muscle weakness and absent reflexes. Laboratory findings included low serum potassium, ECG changes indicative of hypokalemia, and non-anion gap metabolic acidosis. A urine spot test confirmed inappropriate renal potassium loss, diagnosing type 1 distal renal tubular acidosis. Positive autoimmune markers led to the diagnosis of primary Sjogren's syndrome. Treatment with sodium bicarbonate, prednisolone, and oral potassium supplementation resulted in improved muscle power.

Conclusion:

Kidney involvement in primary Sjogren's syndrome (pSS) can present uncommonly as hypokalemic paralysis, sometimes occurring before the onset of sicca symptoms. Physicians should consider pSS in patients presenting with hypokalemic paralysis and renal tubular acidosis (RTA), even in the absence of sicca symptoms. Early recognition and appropriate treatment are essential to manage potentially life-threatening hypokalemia and improve renal outcomes.

Keywords: *Hypokalemic Paralysis, Primary Sjogren's syndrome, Renal Tubular Acidosis*

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Introduction

Primary Sjogren's syndrome (pSS) is an autoimmune disorder characterized by a diverse range of clinical manifestations, with its hallmark being the involvement of exocrine glands leading to the common presentation of dry eyes and a dry mouth, often referred to as the sicca component [1]. While pSS is predominantly associated with exocrine gland dysfunction, it is important to recognize that this autoimmune condition can extend its impact to various non-exocrine organs, leading to a wide spectrum of clinical symptoms and complications.

Beyond the characteristic sicca symptoms, pSS may manifest in the skin, lungs, gastrointestinal tract, central and peripheral nervous system, musculoskeletal system, and even the kidneys, among other non-exocrine organs [2]. This systemic nature of pSS can result in significant morbidity and impaired quality of life for affected individuals.

Understanding the diverse clinical presentations and organ involvement in pSS is of paramount importance for both clinicians and researchers. Therefore, this study aims to shed light on the less common but clinically significant kidney involvement in pSS, which can uncommonly present as hypokalemic paralysis. By exploring this atypical presentation, we hope to contribute to the early recognition and timely management of such cases, which not only addresses the potentially life-threatening hypokalemia but also improves the overall renal outcome in patients with primary Sjogren's syndrome.

The aim of the present study is to explore and document a rare presentation of kidney involvement in Primary Sjogren's syndrome (pSS), specifically focusing on hypokalemic paralysis as an atypical manifestation.

Case Presentation

Patient Profile:

A 21-year-old female presented with sudden quadriparalysis, which had started a day prior to her admission. There was no other significant illness in the past. She had no history of trauma, insect or animal bites,

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diarrhea, exposure to insecticides, recent vaccination, fever, headache, or altered sensorium. She had normal body temperature, heart rate, and blood pressure.

Clinical Examination:

The patient displayed muscle weakness (grade I power) in all four limbs, with absent deep tendon reflexes and superficial reflexes. Her neck tone was normal, and other systemic examinations were unremarkable.

Laboratory Investigations:

Metabolic parameters were normal except for low serum potassium (1.51 meq/l). Electrocardiogram findings indicated ST depression, T wave inversion, presence of U waves, and a prolonged QT interval, suggestive of hypokalemia. Blood gas analysis showed non-anion gap metabolic acidosis with hyperchloremia and hypokalemia. Correction of hypokalemia led to gradual improvement in muscle power by the third day.

Parameters	Result value
pH	7.28
Serum sodium	142.50 mmol/L
Serum potassium	2.14 mmol/L
Serum chloride	109.80 mmol/L
Blood urea	25.00 mg/dl
Serum creatinine	1.00 mg/dl
Blood sugar fasting	90.00 mg/dl
SGPT/ALT	53.00 U/L
Total WBC Count	11,100.00/Cu mm
Neutrophil	84.00 %
Lymphocyte	12.0 %
Monocyte	1.00%
Urinary K^+ Creatinine Ration	
Urine potassium	33.0 mmol
Urine creatinine	31.6 mg/dl
Potassium creatinine Ratio	11.86%

Diagnosis:

A urine spot test revealed an elevated urinary pH and increased urinary potassium to creatinine ratio, indicating inappropriate renal potassium loss. These findings were consistent with type 1 distal renal tubular acidosis (dRTA). Further investigations revealed strongly positive antinuclear antibodies (ANA), anti-SS-A, anti-Ro antibodies, and anti-

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La antibodies, confirming the diagnosis of primary Sjogren's syndrome (pSS).

Management:

The patient received treatment with sodium bicarbonate (1-3 mmol/kg per day), prednisolone, and oral potassium supplementation. Her muscle power improved to grade IV in both upper and lower limbs.

Discussion

The case report highlights a unique presentation of primary Sjogren's syndrome (pSS) in a 21-year-old female who exhibited sudden onset quadriparalysis without the classical sicca symptoms typically associated with the disorder. Notably, her metabolic investigations were mostly normal except for severe hypokalemia, which led to profound muscle weakness. The diagnostic journey revealed non-anion gap metabolic acidosis with hyperchloremia and hypokalemia, eventually pointing towards distal renal tubular acidosis (dRTA) as the underlying cause. The presence of strongly positive antinuclear antibodies (ANA) and specific anti-SS-A, anti-Ro, and anti-La antibodies confirmed the diagnosis of pSS. Her condition improved significantly with the correction of hypokalemia and management with sodium bicarbonate, prednisolone, and oral potassium supplementation.

The case underscores the atypical manifestation of pSS where renal involvement preceded the typical exocrine symptoms. Hypokalemic paralysis in pSS is an uncommon but critical manifestation that requires early recognition and prompt treatment. The presence of dRTA as a complication of pSS indicates the systemic nature of the disease, affecting not just exocrine glands but also internal organs like the kidneys. The patient's response to treatment highlights the efficacy of correcting the electrolyte imbalance and addressing the autoimmune activity through immunosuppression.

Similar cases have been reported, emphasizing the variability in pSS presentations. A study by [3] discussed the multi-organ involvement in Sjogren's syndrome, highlighting the importance of considering pSS in the differential diagnosis of various nonspecific systemic symptoms [3]. Another study by [4] focused on the renal aspects of pSS, indicating that renal tubular acidosis could be a presenting feature in some patients. Both studies support our findings and the notion that pSS can present primarily with systemic involvement, even in the absence of sicca symptoms.

Sjogren's syndrome (SS) is typically associated with symptoms of dry eyes and mouth, a result of immune cells targeting the glands responsible for moisture. Beyond these common symptoms, SS may also manifest in various systemic issues including arthritis, pulmonary complications, Raynaud's phenomenon (a condition where extremities change color in response to cold or stress), renal complications, hematological disorders, vascular inflammation, and potentially lymphoma. Among the renal manifestations, inflammation in the kidney's filtering segments is frequently observed. A particularly severe and uncommon renal complication related to SS is glomerulopathy, known for its detrimental prognosis [4, 5].

The mechanisms leading to distal renal tubular acidosis (dRTA) in the context of SS are not entirely understood. Hypotheses include the potential absence of a crucial acid pump in the renal tubules or interference by specific autoantibodies that hinder the kidneys' ability to manage acid levels [6]. A notable consequence of dRTA is a significant drop in potassium levels, which, in extreme cases, can cause profound muscle weakness, sometimes resembling paralysis. Intriguingly, such paralytic episodes have occasionally been reported as the initial manifestation of SS [7].

In individuals with dRTA, the body's struggle to maintain acid-base balance results in marked potassium depletion, exacerbated by conditions that enhance potassium discharge in the urine. In instances of life-endangering muscle weakness caused by low potassium levels, the immediate medical response involves the elevation of these levels [8]. While treatments aimed at correcting acidosis are beneficial, they must be administered judiciously, as they may inadvertently further reduce potassium levels. Immunosuppressive therapies, including corticosteroids, are typically reserved for situations where there is a rapid decline in renal function. Nevertheless, these interventions require vigilant monitoring due to the potential for re-emergence of renal issues [9]. Despite these challenges, there have been reports of successful management of these severe conditions, offering a glimmer of hope for affected individuals.

Key issues:

In the present case of a 21-year-old female with Primary Sjogren's Syndrome (pSS), key issues included its atypical presentation, diagnostic challenges, and the systemic involvement of pSS. The patient presented with sudden onset quadriparalysis, a rare manifestation absent of classic sicca symptoms, leading to diagnostic difficulties. Laboratory findings revealed severe hypokalemia and non-anion gap metabolic acidosis, highlighting the need for

comprehensive laboratory investigations including autoimmune markers. This case emphasized the systemic nature of pSS, affecting not just exocrine glands but also internal organs like the kidneys, with distal renal tubular acidosis (dRTA) as a significant complication. Treatment involved addressing both the electrolyte imbalance and the autoimmune activity, requiring careful monitoring. The case illustrates the complexity of pSS, underscoring the importance of early recognition, comprehensive diagnosis, and a multidisciplinary approach in management to prevent severe, potentially life-threatening complications.

Generalizability:

The case report on a rare presentation of Primary Sjogren's Syndrome (pSS) with kidney involvement manifesting as hypokalemic paralysis offers valuable insights for generalizing to other settings. It emphasizes the importance of recognizing atypical presentations in differential diagnoses, thereby enhancing prompt and accurate detection of complex autoimmune diseases like pSS. The findings underscore the necessity of a multidisciplinary approach in healthcare, highlighting the need for collaboration among various specialists for improved patient outcomes. This case report serves not only as an educational resource for healthcare professionals but also underscores the significance of patient education and public awareness about the systemic nature of autoimmune diseases. Furthermore, it has implications for global health, particularly in resource-limited settings, by guiding general practitioners in early referral and management, potentially reducing complications. This study can also influence research, clinical practice, and healthcare policy by integrating rare presentations into the broader clinical understanding of diseases like pSS, ensuring comprehensive healthcare planning and resource allocation.

Clinical Practice Guidance:

Clinicians should maintain a high index of suspicion for pSS in patients presenting with unexplained hypokalemic paralysis and metabolic acidosis. Early and comprehensive laboratory investigation, including ANA and specific autoantibodies, is crucial for timely diagnosis. Understanding the potential for non-exocrine organ involvement in pSS is essential for guiding appropriate management strategies. Prompt correction of hypokalemia and addressing the underlying autoimmune process can lead to significant improvements and prevent potentially life-threatening complications. Regular monitoring and a multidisciplinary approach are advisable due to the systemic nature of the disease.

By integrating these insights, clinicians can better recognize and manage atypical presentations of pSS, improving patient outcomes through early intervention and comprehensive care.

Conclusion

In conclusion, our case and the reviewed literature emphasize that pSS is a systemic autoimmune disorder with a wide range of manifestations. Clinicians must be vigilant for atypical presentations like severe hypokalemic paralysis, especially in young patients without significant past medical history. Early recognition and appropriate management of these severe complications can prevent life-threatening outcomes and improve the overall prognosis for patients with pSS. This case serves as a reminder of the importance of considering a broad differential diagnosis in cases of unexplained hypokalemic paralysis and underscores the need for a multidisciplinary approach to the care of patients with complex systemic diseases like pSS.

Recommendations:

It is recommended to enhance medical education on autoimmune diseases' varied presentations, advocate for a multidisciplinary management approach, and encourage research on atypical pSS manifestations. Developing comprehensive guidelines, improving patient education, adapting global health policies, and establishing early screening protocols are also crucial. These measures aim to improve understanding and management of complex autoimmune cases, ensuring effective healthcare delivery.

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Ethical considerations:

The study protocol was approved by the Ethics Committee and written informed consent was received from the patient participating.

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Conflict of interest:

The authors have no competing interests to declare.

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