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A Case Report of Renal Tubular Acidosis Type 1 without Glomerular Disease in an Adolescent with Pediatric-onset Systemic Lupus Erythematosus

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ABSTRACT

Introduction: Between 50-75% of children and adolescents with systemic lupus erythematosus (SLE) experience kidney involvement within the first year of diagnosis. The gold standard for diagnosing renal involvement in SLE is a renal biopsy. It is uncommon for SLE to cause isolated tubular involvement without any glomerular disease.

Case Presentation: We report an adolescent girl with a known history of systemic lupus erythematosus who presented to the emergency department with progressively worsening weakness. The diagnosis revealed that she had distal renal tubular acidosis (RTA) without any glomerular disease. Her history of nephrocalcinosis and kidney stones on renal ultrasound is most consistent with distal renal tubular acidosis diagnosis.

Conclusion: This case highlights the importance of considering renal tubular acidosis in lupus patients who experience recurrent hypokalemic episodes. When a patient presents with a normal anion gap metabolic acidosis and hyperchloremia, without evidence of gastrointestinal HCO3 loss or absorption of exogenous acid, renal tubular acidosis (RTA) should be considered.

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Introduction

The prevalence of kidney involvement in SLE is 50%-75% in children [1]. The kidneys are typically affected either at the time of diagnosis or within the first year [2]. It is rare for SLE to only affect the renal tubulointerstitium. It typically involves the glomerulus (known as "lupus nephritis") [1]. Although the prevalence of renal tubular acidosis (RTA) during SLE is not well-researched, Based on the available literature, the probability of RTA is very low [3].

When a patient with no signs of gastrointestinal HCO3 losses or absorption of exogenous acid presents with a normal anion gap metabolic acidosis plus hyperchloremia, RTA should generally be considered [4]. RTA type 1 or distal RTA is characterized by alkaline urine, low potassium levels, nephrolithiasis, and nephrocalcinosis [5].

Here we report an adolescent girl with a definite diagnosis of Systemic Lupus Erythematosus (SLE), based on the American College of Rheumatology (ACR) classification criteria, who developed acute quadriplegic weakness due to severe hypokalemia caused by distal Renal Tubular Acidosis (RTA).

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Case Presentation

The patient, an 18-year-old female with a history of SLE, presented to the emergency department in October 2023 with 10 days of progressive weakness.

She initially experienced general malaise, nausea, and lethargy. This was followed by muscle cramps and weakness in her right upper extremity, which progressively worsened. Over the next three days, she experienced weakness on her right side, including her lower extremity and pelvic muscles. She described poor appetite and persistent nausea but did not report any fever, diarrhea, or symptoms of a viral respiratory illness.

Three years ago, she was diagnosed with SLE after presenting with symptoms such as malar rash, photosensitivity, polyarthritis, fatigue, alopecia, oral ulcers, and positive anti-dsDNA and ANA antibodies. A year before her SLE diagnosis, the patient reported experiencing headaches, hair loss, and oral ulcers during the review of systems. Over the past three years, she has been hospitalized three times due to her systemic lupus erythematosus (SLE). She experienced hypokalemia during two admissions.

In October 2021, she was admitted for generalized muscle weakness and was noted to be hypokalemic with a potassium level of 2.4 mEq/L. She has been diagnosed with a flare-up of systemic lupus erythematosus (SLE) and post-viral myositis.

In February 2023, she was hospitalized again for muscle weakness. Her potassium on admission was critically low at 2.13 mEq/L but improved to 4.37 mEq/L by discharge one week later. The patient had a renal ultrasound during hospitalization which revealed medullary nephrocalcinosis. Additionally, a kidney stone measuring 5.3mm was incidentally observed.

After being discharged, the patient underwent two follow-up ultrasounds at two-month intervals. The first ultrasound, performed in May 2023, revealed the presence of microlithiasis and a 5.3 mm lower calyx stone in the right kidney. A follow-up ultrasound in October 2023 showed worsening nephrocalcinosis and new renal calculi. The radiologist suggested they were consistent with RTA or medullary spongy kidney.

During the presentation, the patient remained alert and oriented. Vital signs were stable with a blood pressure of 118/72 mm Hg, heart rate of 70 beats/min, respiratory rate of 17 breaths/min, and oxygen saturation of 98% on room air. The patient's temperature was 37.2°C. A physical examination revealed reduced muscle strength in the right upper and lower extremities. Cranial nerves, sensation, and reflexes were intact. She had scarring alopecia but no active rash or oral ulcerations.

The laboratory results indicate a significant drop in potassium levels to 2.0 mEq/L. additionally, the patient's serum bicarbonate level is 13 mEq/L, serum sodium is 142 mEq/L, chloride is 121 mEq/L, pH is 7.31, and anion gap is 10 mEq/L. Urine pH was 8 with no proteinuria. Ultrasound showed medullary nephrocalcinosis.

The patient was admitted to the ICU for cardiac monitoring due to severe hypokalemia. A nephrologist was consulted for suspected renal tubular acidosis. The patient was prescribed oral potassium citrate and spironolactone to take. After her serum potassium level was returned to 4.1 mEq/L and her muscle weakness improved, she was transferred to the rheumatology service in stable condition after 4 days.

The patient's recurrent hypokalemia and acidosis, history of kidney stones, and nephrocalcinosis on renal ultrasounds were most consistent with distal renal tubular acidosis secondary to SLE. She is currently taking oral potassium citrate, spironolactone, oral prednisolone, Mycophenolate mofetil, and hydroxychloroquine to maintain her SLE and distal RTA.

Discussion

Renal tubular acidosis (RTA) is a type of normal anion gap metabolic acidosis that can be classified into three subtypes based on the underlying cause. Type 1 or distal RTA is associated with limited urinary hydrogen secretion, type 2 or proximal RTA is characterized by decreased bicarbonate reabsorption, and type 4 RTA is caused by hypoaldosteronism. These subtypes can occur as a primary disorder or as a secondary condition due to other underlying medical conditions. [6–7]. In contrast to type 4, types 1 and 2 are characterized by hypokalemia due to excess renal loss of potassium. [8]. Type 1 RTA is commonly caused by autoimmune diseases such as SLE, Sjogren syndrome, rheumatoid arthritis, systemic sclerosis, thyroiditis, hepatitis, and primary biliary cirrhosis. This type of RTA can lead to nephrolithiasis, nephrocalcinosis, and bone disease [4, 9].

A renal biopsy is the gold standard for diagnosing kidney involvement in SLE [10]. A biopsy was not performed due to the absence of glomerular involvement symptoms, such as proteinuria and hematuria. Although glomerular involvement cannot be ruled out in the absence of symptoms, a renal biopsy was included in the patient's follow-up plan. Established the diagnosis of distal RTA with normal anion gap hyperchloremic metabolic acidosis, inappropriately alkaline urine (pH >5.5), and other symptoms [11].

Prompt potassium repletion rapidly improved weakness and nausea. However, severe hypokalemia can cause life-threatening

arrhythmias and arrest with potassium below 2.5 mmol/L. [12]. During hospitalization, potassium levels were closely monitored daily. follow-up testing was conducted after discharge to prevent recurrent hypokalemia. [13].

Hypokalemia accompanying distal RTA may be reversible by addressing acidosis alone. Patients may experience recurrent hypokalemia and require potassium supplements. Potassium citrate is the most effective method for alkalinizing urine in distal RTA. This provides both potassium and bicarbonate to resolve issues of acidosis and hypokalemia. [14–15]. Spironolactone has been shown to effectively correct hypokalemia [16]. Combining spironolactone and potassium citrate reduces doses and combats urinary potassium losses and acidosis in distal RTA. Upon discharge, our patient received spironolactone and potassium citrate in addition to previous lupus treatments.

Conclusion

This case emphasizes the importance of recognizing distal RTA as a complication of SLE, even when presenting solely as hypokalemia and paralysis. Maintaining a high index of suspicion for renal tubular acidosis (RTA) in lupus patients with recurrent hypokalemic episodes can enable early diagnosis and prevent life-threatening complications.

Authors' contributions

All authors have contributed to the manuscript. Conception and design: AM (Abdolreza Malek) & MV (Mahdieh Vahedi). Data collection: AB (Asma Batouri) & AK (Amir Muhammad khuban). Manuscript writing and review: MV & SS (Sepideh Seyedkaboli) & AK. All authors read and approved the final manuscript.

Ethics approval and consent to participate

In compliance with the Helsinki Declaration, informed assent or consent was obtained from the patient's parents.

Consent for publication

Consent was obtained from the parents of the patient. Our study doesn't include personal data.

Availability of data and materials

You can request the study's data from the corresponding author.

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Conflict of interest

The authors declare no competing interests.

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