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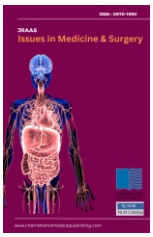

Case Report

Section: General Medicine

Systemic Sclerosis Presenting with Distal Renal Tubular Acidosis, Severe Hypokalemia, & Interstitial Lung Disease: A Rare Multisystem Presentation

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HIGHLIGHTS

- Rare dRTA presentation
- Severe hypokalemic crisis
- Systemic sclerosis overlap
- Extensive multisystem involvement
- Early diagnosis improves survival

Key Words:
Systemic sclerosis
Sjögren syndrome
Distal renal tubular acidosis
Hypokalemia
Interstitial lung disease
Autoimmune disease

ABSTRACT

Introduction: Systemic sclerosis (SSc) is a multisystem autoimmune connective tissue disorder characterized by fibrosis, vasculopathy, and immune dysregulation. Distal renal tubular acidosis (dRTA) is an uncommon manifestation and may rarely present with life-threatening electrolyte abnormalities. **Aim & Objectives:** To report a rare case of systemic sclerosis–Sjögren overlap syndrome presenting with distal renal tubular acidosis and severe hypokalemia. To highlight the importance of early recognition of autoimmune causes of unexplained metabolic acidosis and the role of prompt diagnosis and appropriate immunosuppressive management in improving clinical outcomes. **Case Presentation:** A 58-year-old woman with a history of type 2 diabetes mellitus and hypothyroidism presented with acute respiratory distress, hypoxia, hypotension, and generalized limb weakness. She required intensive care unit admission and ventilatory support. Initial investigations revealed severe hypokalemia and metabolic acidosis. Further evaluation demonstrated a positive urine anion gap consistent with distal renal tubular acidosis. Autoimmune workup showed positivity for antinuclear antibodies, anti-Scl-70, anti-SSA, and Ro-52 antibodies. Skin biopsy confirmed systemic sclerosis with Sjögren overlap syndrome. Additional investigations revealed interstitial lung disease with a nonspecific interstitial pneumonia pattern, esophageal dysmotility, vocal cord paralysis, and proteinuria, indicating extensive multisystem involvement. The patient was treated with correction of electrolyte abnormalities, corticosteroids, immunosuppressive therapy, and supportive care. Significant clinical improvement was observed with normalization of serum potassium levels and recovery from respiratory and hemodynamic compromise. **Results:** A 58-year-old woman with systemic sclerosis–Sjögren overlap syndrome presented with distal renal tubular acidosis, severe hypokalemia, and metabolic acidosis causing respiratory failure and hypotension. Treatment with electrolyte correction, corticosteroids, immunosuppressants, and supportive care resulted in normalization of serum potassium, resolution of acidosis, and significant clinical recovery. **Conclusion:** This case highlights a rare presentation of systemic sclerosis with Sjögren overlap syndrome manifesting as distal renal tubular acidosis and severe hypokalemia. Early recognition of autoimmune causes of unexplained metabolic acidosis and hypokalemia is essential for timely diagnosis and appropriate management.

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INTRODUCTION

Systemic sclerosis (SSc) is a chronic autoimmune connective tissue disorder characterized by immune dysregulation, microvascular injury, and progressive fibrosis affecting the skin and multiple internal organs. Although uncommon, with an estimated prevalence of 50–300 cases per million population worldwide, SSc is associated with significant morbidity and mortality owing to its multisystem involvement [1,2]. The disease manifests across a spectrum ranging from limited cutaneous disease to diffuse systemic involvement affecting the lungs, gastrointestinal tract, kidneys, heart, and musculoskeletal system [3].

Pulmonary complications are among the leading causes of mortality in SSc. Interstitial lung disease (ILD), particularly the nonspecific interstitial pneumonia (NSIP) pattern, is the most frequent pulmonary manifestation and may occur in up to 50–70% of patients during the disease [4,5]. Progressive pulmonary fibrosis contributes substantially to functional impairment and reduced survival. Gastrointestinal involvement is similarly common, with esophageal dysmotility resulting from smooth muscle atrophy and fibrosis occurring in more than 80% of patients [6]. These manifestations often precede the diagnosis and may remain unrecognized until advanced disease develops. Renal involvement in SSc most commonly presents as scleroderma renal crisis; however, tubular dysfunction is increasingly recognized as an important but underdiagnosed complication [7]. Distal renal tubular acidosis (dRTA) is characterized by impaired hydrogen ion secretion by the distal nephron, leading to nonanion gap metabolic acidosis, hypokalemia, nephrocalcinosis & urinary acidification defects [8]. Although dRTA has been described in several autoimmune disorders, including Sjögren syndrome and systemic lupus erythematosus, its occurrence in systemic sclerosis remains rare [9,10]. Severe hypokalemia secondary to dRTA may result in life-threatening complications such as respiratory muscle weakness, paralysis, cardiac arrhythmias, and circulatory collapse [11].

The coexistence of systemic sclerosis and Sjögren syndrome overlap further to increase the likelihood of renal tubular dysfunction because autoimmune-mediated injury to the distal nephron is a well-established feature of Sjögren syndrome [12]. In such patients, dRTA may be the presenting manifestation and can obscure the underlying connective tissue disease [13]. Consequently, recognition of atypical presentations is crucial for timely diagnosis and institution of immunomodulatory therapy. Systemic sclerosis with distal renal tubular acidosis and interstitial lung disease **(Figure 1)**.

We report a rare case of systemic sclerosis with Sjögren overlap presenting initially as shock, respiratory failure, severe hypokalemia, and metabolic acidosis due to distal renal tubular acidosis. The patient additionally demonstrated interstitial lung disease, esophageal dysmotility, vocal cord paralysis, and proteinuria, reflecting extensive multisystem involvement. This case highlights the importance of maintaining a high index of suspicion for autoimmune connective tissue disorders in patients presenting with unexplained electrolyte abnormalities and multisystem manifestations, as early diagnosis & multidisciplinary management can significantly improve outcomes.

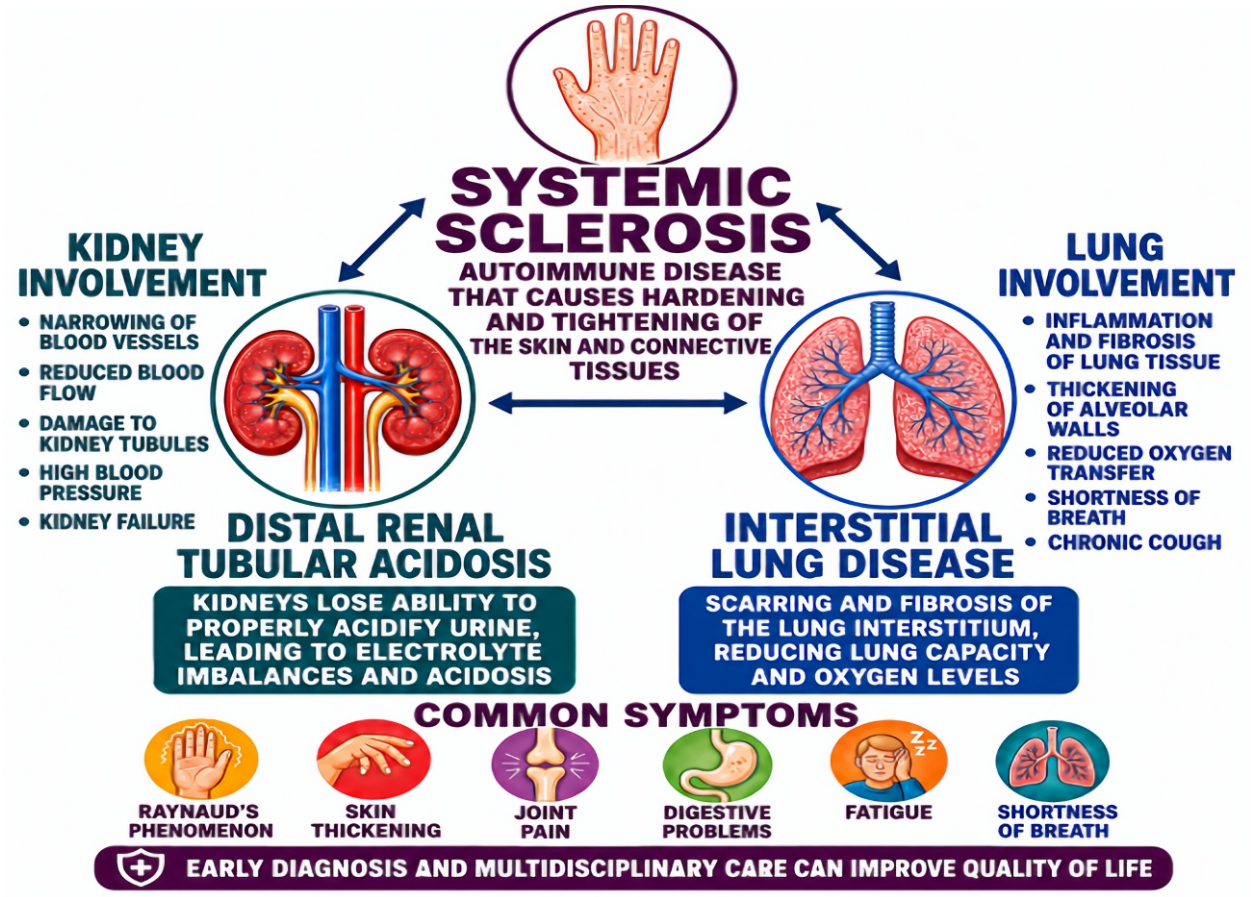

Figure 1: Schematic illustration of systemic sclerosis showing distal renal tubular acidosis, interstitial lung disease, and associated clinical manifestations.

CASE PRESENTATION

A 58-year-old woman with a known history of type 2 diabetes mellitus and hypothyroidism presented with acute respiratory distress, hypoxia, hypotension, and generalized limb weakness. Owing to severe respiratory compromise and shock, she required endotracheal intubation and intensive care unit admission. Initial evaluation revealed severe hypokalemia associated with metabolic acidosis. The patient was managed with ventilatory support, hemodynamic stabilization, and electrolyte correction. However, the persistence of metabolic abnormalities prompted further evaluation for an underlying systemic disorder. During hospitalization, additional features suggestive of multi-system disease were identified. Clinical examination revealed skin changes consistent with systemic sclerosis. The patient also demonstrated evidence of pulmonary & gastrointestinal involvement. Further assessment showed interstitial lung disease and esophageal dysmotility. Vocal cord paralysis was also detected, indicating upper aerodigestive tract involvement. The constellation of cutaneous, pulmonary, gastrointestinal, and renal manifestations raised suspicion for an underlying auto-immune connective tissue disorder. Hand showing cutaneous features of systemic sclerosis **(Figure 2)**.

Investigations

Laboratory evaluation revealed severe hypokalemia with metabolic acidosis. Further assessment demonstrated a positive urine anion gap, consistent with distal renal tubular acidosis (dRTA). Autoimmune workup showed positivity for antinuclear antibodies (ANA), anti-Scl-70, anti-SSA, and Ro-52 antibodies, raising suspicion of an underlying connective tissue disease. Skin biopsy findings were compatible with systemic sclerosis, confirming the diagnosis of systemic sclerosis with Sjögren overlap syndrome. High-resolution computed tomography (HRCT) of the chest demonstrated interstitial lung disease with a nonspecific interstitial pneumonia (NSIP) pattern, while gastrointestinal evaluation revealed esophageal dysmotility. Vocal cord examination confirmed vocal cord paralysis, indicating additional upper aerodigestive tract involvement. Collectively, these findings established the diagnosis of systemic sclerosis with multisystem involvement affecting the renal, pulmonary, gastrointestinal, and integumentary systems. During hospitalization, serial biochemical monitoring demonstrated progressive improvement in metabolic parameters. Serum potassium increased steadily from 1.4 mmol/L on admission to 4.0 mmol/L at discharge, while serum creatinine declined from 2.3 mg/dL to 1.35 mg/dL, reflecting correction of distal renal tubular acidosis and recovery of renal function **(Figure 3)**.

Management and Outcome

The patient was treated with intensive supportive care, including mechanical ventilation, correction of hypokalemia, and management of shock. Following identification of distal renal tubular acidosis, electrolyte abnormalities and metabolic acidosis were addressed with appropriate replacement therapy. After confirmation of the underlying autoimmune disorder, corticosteroids and immunosuppressive therapy were initiated. Multidisciplinary management was provided for pulmonary and gastrointestinal manifestations. The patient showed significant clinical improvement with stabilization of hemodynamic status, correction of metabolic abnormalities, improvement in respiratory function, and successful recovery from the acute illness. She was subsequently discharged on continued immunosuppressive therapy and follow-up care.

RESULTS

A 58-year-old woman was diagnosed with systemic sclerosis with Sjögren overlap syndrome after presenting with distal renal tubular acidosis, severe hypokalemia, and normal anion-gap metabolic acidosis causing generalized weakness, acute respiratory failure, and hemodynamic instability. Autoimmune evaluation revealed positive ANA, anti-Scl-70, anti-SSA, and Ro-52 antibodies, while skin biopsy confirmed systemic sclerosis. Further investigations demonstrated extensive multisystem involvement, including interstitial lung disease with a nonspecific interstitial pneumonia (NSIP) pattern, esophageal dysmotility, vocal cord paralysis, and proteinuria. Following correction of electrolyte abnormalities, intensive supportive care, corticosteroid therapy, and immunosuppressive treatment, the patient showed normalization of serum potassium levels, resolution of metabolic acidosis, marked improvement in respiratory and hemodynamic status, and significant overall clinical recovery.

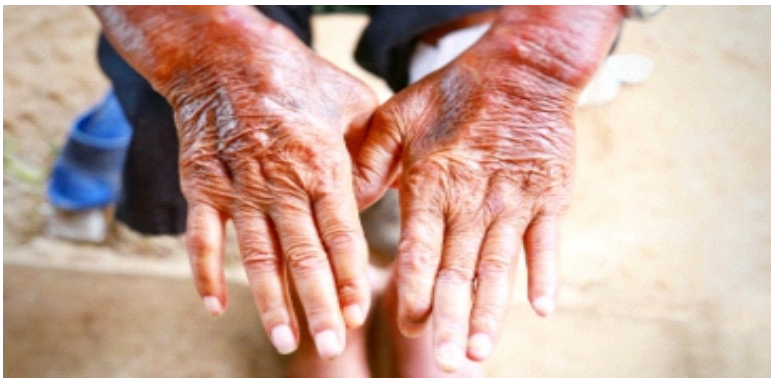

Figure 2. Clinical photograph of the hands showing cutaneous manifestations of systemic sclerosis. The image demonstrates diffuse skin tightening, shiny skin texture, and loss of normal skin folds over the dorsum of both hands, findings characteristic of sclerodermatous skin involvement. The fingers appear tapered with reduced skin elasticity, consistent with chronic cutaneous fibrosis.

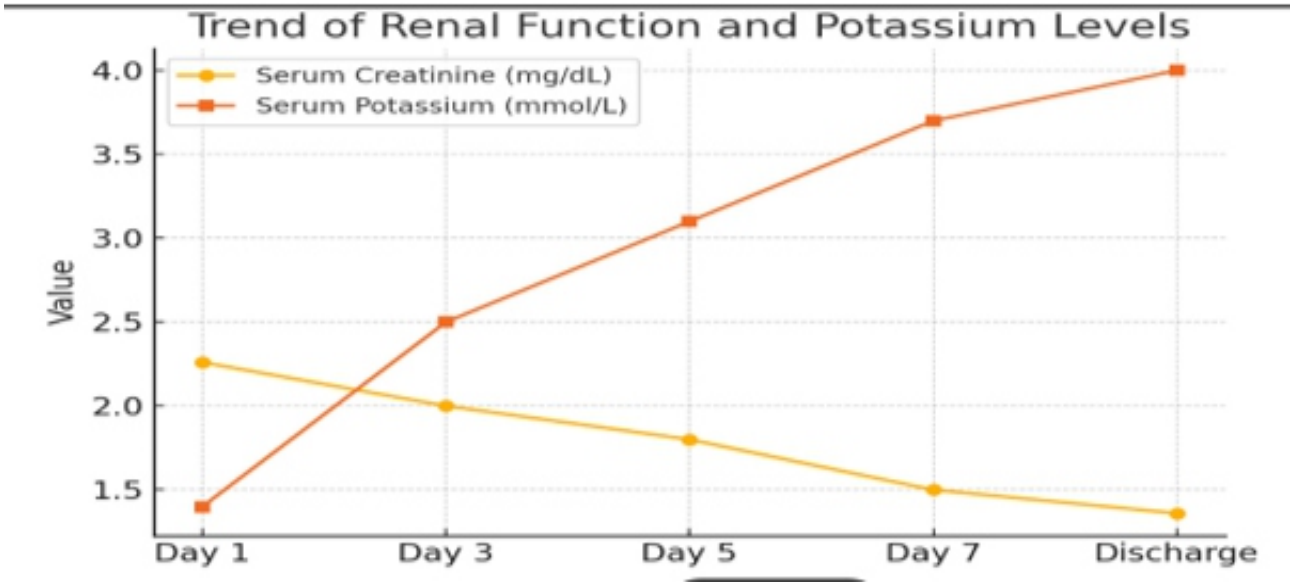

Figure 3. Trend of renal function and serum potassium during hospitalization. The graph demonstrates progressive correction of severe hypokalemia following treatment, with serum potassium improving from approximately 1.4 mmol/L on admission to 4.0 mmol/L at discharge. Simultaneously, serum creatinine decreased from 2.3 mg/dL to 1.35 mg/dL, indicating improvement in renal dysfunction and overall metabolic status.

DISCUSSION

Systemic sclerosis (SSc) is a complex autoimmune connective tissue disorder characterized by immune dysregulation, microvascular injury, and progressive fibrosis affecting multiple organ systems [1-3]. Although cutaneous manifestations are often the most recognizable feature, internal organ involvement significantly contributes to morbidity and mortality. Pulmonary involvement, particularly interstitial lung disease (ILD), remains one of the leading causes of death in patients with SSc [4,5]. Gastrointestinal manifestations, especially esophageal dysmotility, are also frequently encountered and may precede the diagnosis of systemic disease [6].

The present case is noteworthy because the patient initially presented with severe hypokalemia, respiratory failure, and shock rather than classical rheumatological symptoms. Renal involvement in systemic sclerosis is traditionally associated with scleroderma renal crisis; however, tubular dysfunction represents a less commonly recognized manifestation [7]. Distal renal tubular acidosis (dRTA) results from impaired hydrogen ion secretion in the distal nephron, leading to metabolic acidosis and potassium wasting [8]. The profound hypokalemia observed in our patient likely contributed to the generalized weakness, respiratory compromise, and hemodynamic instability at presentation. Similar presentations have been described in autoimmune disorders, where severe electrolyte disturbances may overshadow the underlying connective tissue disease [9-11].

An important aspect of this case is the coexistence of systemic sclerosis with Sjögren overlap syndrome. Sjögren syndrome is a well-recognized cause of distal renal tubular acidosis due to autoimmune-mediated tubulointerstitial injury [9,10]. The presence of anti-SSA and Ro-52 antibodies, along with systemic sclerosis-specific anti-Scl-70 positivity, strongly supported an overlap syndrome.

Previous studies have shown that renal tubular dysfunction may be one of the earliest manifestations of Sjögren syndrome and may occasionally present as hypokalemic paralysis or severe metabolic acidosis before the diagnosis of the underlying autoimmune condition is established [12,13]. Therefore, clinicians should maintain a high index of suspicion for autoimmune etiologies in patients presenting with unexplained hypokalemia and normal anion-gap metabolic acidosis.

The multisystem nature of disease involvement in this patient further emphasizes the complexity of systemic sclerosis. In addition to dRTA, the patient exhibited ILD with an NSIP pattern, esophageal dysmotility, vocal cord paralysis, and characteristic cutaneous changes. NSIP is the most common radiological pattern seen in SSc-associated ILD and is associated with progressive pulmonary impairment if left untreated [4,5]. The simultaneous involvement of pulmonary, gastrointestinal, renal, and integumentary systems highlights the importance of comprehensive evaluation once an autoimmune disorder is suspected. Early recognition and prompt treatment contributed significantly to the favorable outcome in this case. Correction of electrolyte abnormalities, intensive supportive care, and initiation of corticosteroids and immunosuppressive therapy resulted in marked clinical improvement. Current therapeutic approaches emphasize individualized immunomodulatory treatment and multidisciplinary management to prevent irreversible organ damage and improve long-term outcomes in systemic sclerosis [14,15]. This case highlights the importance of considering systemic autoimmune diseases in patients presenting with unexplained hypokalemia, metabolic acidosis, respiratory failure, and multisystem manifestations. Recognition of rare presentations such as dRTA-associated systemic sclerosis can facilitate timely diagnosis and appropriate treatment, thereby reducing morbidity and improving patient outcomes.

CONCLUSION

This case describes a rare presentation of systemic sclerosis with Sjögren overlap syndrome manifesting as distal renal tubular acidosis, severe hypokalemia, respiratory failure, and shock. The presence of interstitial lung disease, esophageal dysmotility, vocal cord paralysis, and characteristic cutaneous findings reflected extensive multisystem involvement. Distal renal tubular acidosis is an uncommon but potentially life-threatening manifestation that may precede the diagnosis of an underlying autoimmune disorder. Early recognition of unexplained hypokalemia and metabolic acidosis, coupled with comprehensive autoimmune evaluation, was crucial in establishing the diagnosis. Prompt correction of electrolyte abnormalities and initiation of immunosuppressive therapy resulted in significant clinical improvement. This case highlights the importance of maintaining a high index of suspicion for connective tissue diseases in patients with unexplained metabolic and multisystem manifestations.

LIMITATIONS & FUTURE PERSPECTIVES

The study's limitations include a single-centre setting, a relatively small sample size, and a short study duration, which may limit the broader applicability of the results. Future studies should incorporate multicentre designs with larger populations to enhance validity, assess long-term outcomes, and investigate advanced diagnostic & management approaches. Such efforts will improve overall patient care & help minimize complications.

CLINICAL SIGNIFICANCE

The clinical significance of this study lies in its potential to bridge the gap between research findings and practical healthcare applications. It emphasizes the importance of translating scientific observations into meaningful improvements in patient care, diagnosis, and treatment outcomes. By highlighting real-world relevance, the study contributes to evidence based medical practice and supports informed clinical decision making. Ultimately, the findings aim to enhance patient quality of life, optimize therapeutic strategies, and promote better disease management in clinical settings.

ABBREVIATIONS

ANA: Antinuclear Antibody
dRTA: Distal Renal Tubular Acidosis
ICU: Intensive Care Unit
ILD: Interstitial Lung Disease
NSIP: Nonspecific Interstitial Pneumonia
SSc: Systemic Sclerosis
T2DM: Type 2 Diabetes Mellitus

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AUTHOR CONTRIBUTIONS

All authors significantly contributed to the study conception and design, data acquisition, or data analysis and interpretation. They participated in drafting the manuscript or critically revising it for important intellectual content, consented to its submission to the current journal, provided final approval for the version to be published, and accepted responsibility for all aspects of the work. Additionally, all authors meet the authorship criteria outlined by the International Committee of Medical Journal Editors (ICMJE) guidelines.

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CONFLICT OF INTEREST

Authors declared that there is no conflict of interest.

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DATA AVAILABILITY

All data generated and analyzed are included within this research article. The datasets utilized and/or analyzed in this study can be obtained from the corresponding author upon a re-asonable request.

USE OF ARTIFICIAL INTELLIGENCE (AI) & LARGE LANGUAGE MODEL (LLM)

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