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Case Report

Antisynthetase Syndrome Revealed by Glomerular Syndrome

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Abstract: Renal involvement in antisynthetase syndrome, although less frequent than other manifestations such as inflammatory myopathy or interstitial lung disease, is an important complication that can present as acute or chronic renal failure as well as significant proteinuria. The exact mechanisms are not fully understood, but various renal lesions have been described. Diagnosis relies on clinical evaluation, laboratory and imaging tests, as well as detection of autoantibodies, notably anti-Jo 1 antibodies. A multidisciplinary approach involving rheumatologists, nephrologists, and other specialists is necessary for comprehensive evaluation and rapid identification of renal complications. Early intervention and close monitoring are crucial to optimize clinical and renal outcomes in patients with this syndrome. As was the case with our 30-year-old patient managed for glomerular syndrome, with a history of early abortions. She presents with symptoms including muscle weakness, joint pain, Raynaud's phenomenon, cough, and general deterioration. Biological examinations show anemia, inflammation, liver and muscle involvement, as well as proteinuria and hematuria. Anti-Jo1 antibodies are positive, confirming autoimmune reactivity. Radiological examinations reveal pulmonary and muscle involvement, corroborating the clinical picture. Treatment with corticosteroids and immunosuppressants is initiated, leading to significant improvement in the patient's condition. Antisynthetase syndrome, although rare, can affect multiple organ systems, including the kidneys. Its diagnosis relies on multidisciplinary evaluation and detection of specific autoantibodies. Treatment aims to control inflammation and prevent complications. Long-term monitoring is necessary to maintain clinical and biological improvements.

Keywords: Antisynthetase syndrome, renal, glomerular syndrome, autoimmune disease.

INTRODUCTION

Although renal involvement in antisynthetase syndrome is less common than other manifestations such as inflammatory myopathy or interstitial lung disease, it remains an important complication requiring special attention. The exact prevalence of renal involvement in antisynthetase syndrome is not fully elucidated due to the rarity of this condition and the lack of specific data, but it is crucial to recognize it due to its impact on overall patient prognosis. Renal involvement can manifest in various ways, including the development of acute or chronic renal failure, as well as significant proteinuria. The exact mechanisms underlying renal involvement in antisynthetase syndrome are not fully understood, but several renal lesions have been described, including immune complex glomerulonephritis, tubulointerstitial, and vascular involvement. The diagnosis of antisynthetase syndrome relies on a combination of clinical manifestations, laboratory and imaging findings, as well as the detection of specific autoantibodies, notably anti-Jo 1 antibodies. Therefore, a multidisciplinary approach involving rheumatologists, nephrologists, and other specialists is necessary to thoroughly evaluate patients presenting with clinical symptoms compatible with this syndrome and to promptly identify potential renal complications. Early intervention and close monitoring are essential to optimize both clinical and renal outcomes for patients with antisynthetase syndrome, as illustrated by the case we report of a young adult female with antisynthetase syndrome.

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OBSERVATION

A 30-year-old woman was admitted to our department for the management of glomerular syndrome. Among her medical history, two early abortions were noted, with the last one occurring one month prior to the onset of symptoms of glomerular syndrome. The patient presents with a symptomatology characterized by muscle weakness, joint pain, Raynaud's syndrome, cough, and general malaise. During the clinical examination, several manifestations were observed in the patient, including hyperthermia at 38°C, edema of the lower limbs extending up to the knees, hyperkeratosis with fissures on the fingertips and lateral aspects of the fingers, also known as "mechanic's hands" (Figure 1), which is a dermatological manifestation often associated with autoimmune diseases such as dermatomyositis. Additionally, a pruritic erythema disappearing upon pressure on the abdomen, back, and limbs (Figure 2), inflammatory arthritis of the wrists and interphalangeal joints of both hands, and muscular weakness at the roots of the limbs, which may be associated with inflammatory myopathy observed in some patients with autoimmune diseases, including antisynthetase syndrome.

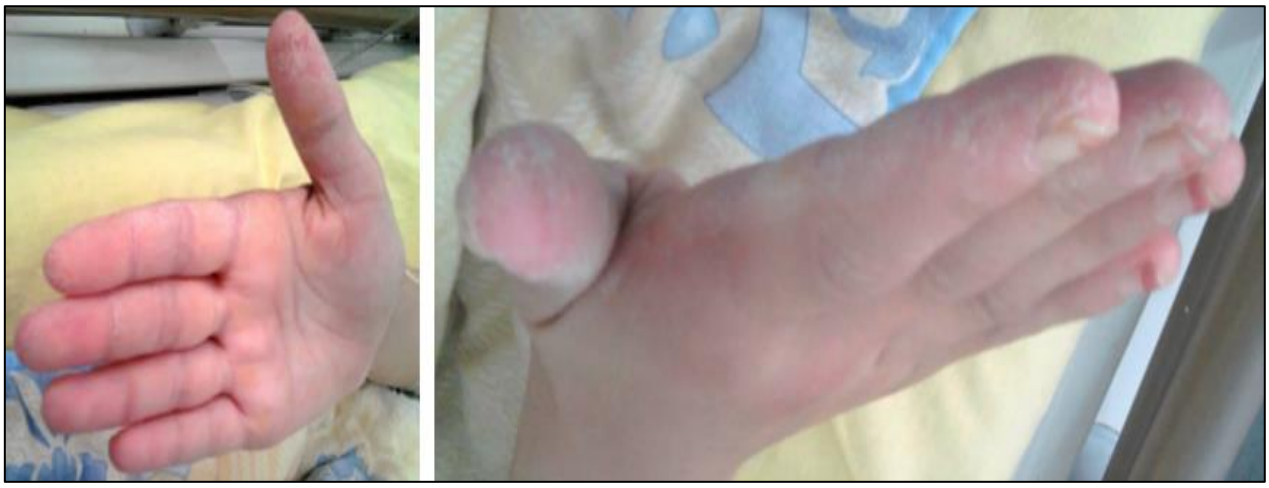

Figure 1: Hyperkeratosis with fissures on the fingertips "mechanic's hands."

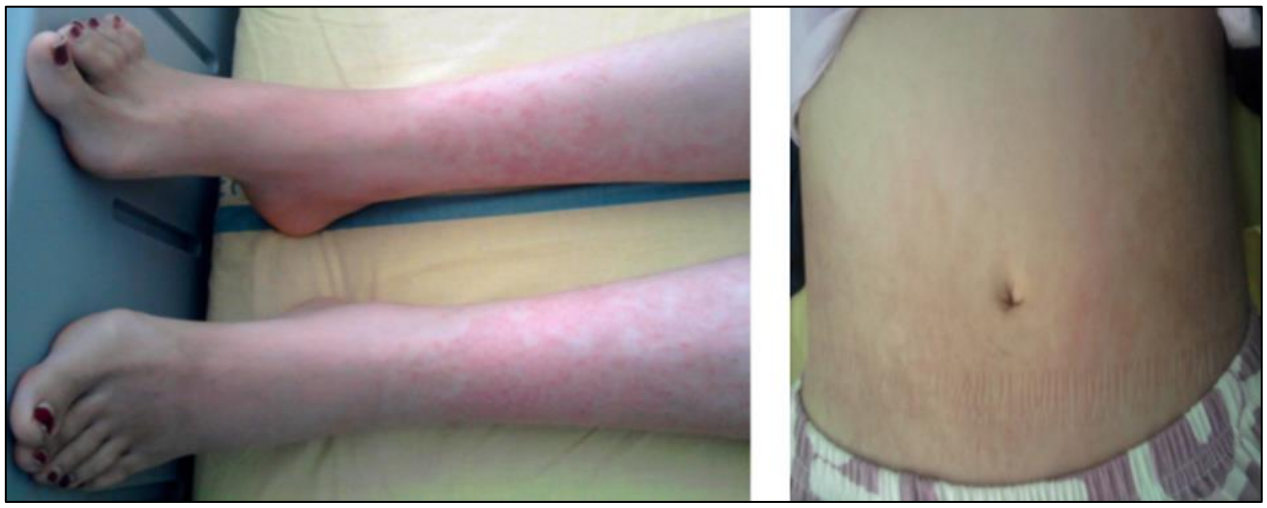

Figure 2 : Pruritic erythema on the abdomen and limbs

The results of the laboratory tests revealed several abnormalities, including: Anemia with a hemoglobin level of 10 g/dl and lymphopenia at 1200/mm3. An elevated sedimentation rate and positive C-reactive protein, suggesting inflammation. Elevated transaminases at 20 times the normal level, indicating liver involvement. High levels of creatine phosphokinase (CPK) at 3489 IU/L and lactate dehydrogenase (LDH) at 1497 IU/L, indicating muscle involvement. Blood cultures returned negative, ruling out systemic bacterial infection. Urine chemistry showed 2+ proteinuria and 4+ hematuria. Twenty-four-hour proteinuria was quantified at 1.26 g/24h, confirming significant protein leakage in the urine. Cytobacteriological examination of urine was negative, excluding urinary tract infection. Immunologically, anti-Jo1 antibodies were positive, indicating autoimmune reactivity against synthetases, while antiphospholipid antibodies were negative, ruling out their involvement in the observed pathology.

Radiological examinations were performed, including an abdominopelvic ultrasound, which showed no notable abnormalities. However, chest X-ray revealed bilateral reticular and micronodular images, indicating diffuse pulmonary involvement (Figure 3). Thoracic computed tomography revealed scattered micronodular infiltrates in both lungs, accompanied by multiple calcified mediastinal lymph nodes (Figure 4). Further investigations, such as pulmonary function tests (PFTs), confirmed the presence of a restrictive syndrome characterized by impaired lung capacity. Electromyography (EMG) of the four limbs showed signs suggestive of muscle involvement of myogenic origin, corroborating the clinical picture of muscle weakness. Muscle biopsy performed on the patient confirmed the diagnosis of dermatomyositis, revealing characteristic alterations in muscle tissue. Capillaroscopy revealed organic microangiopathy, suggesting impairment of blood microcirculation. Lastly, a renal biopsy was performed to evaluate renal involvement, but the results are currently pending. These radiological and functional findings confirm the presence of systemic involvement in the patient, affecting the lungs, muscles, and potentially the kidneys.

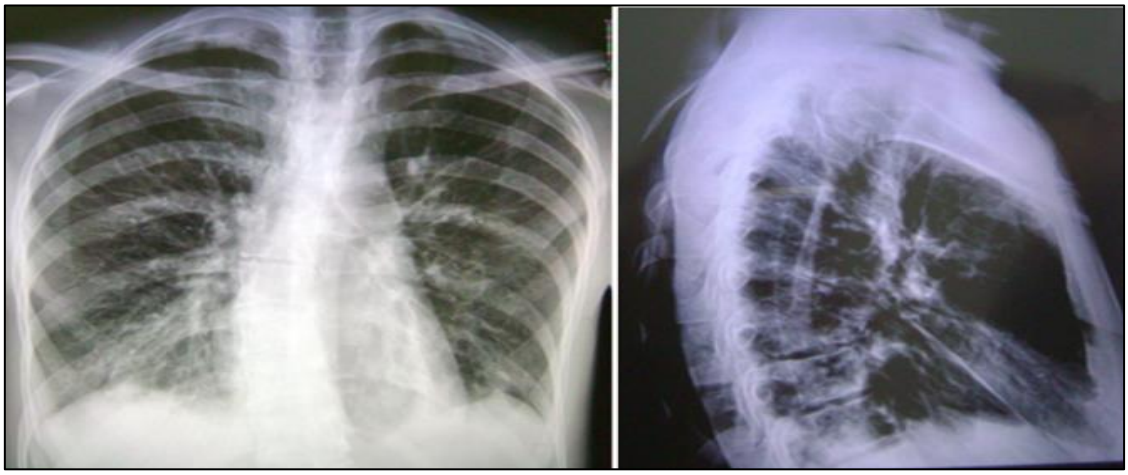

Figure 3 : Bilateral reticular and micronodular images on chest X-ray

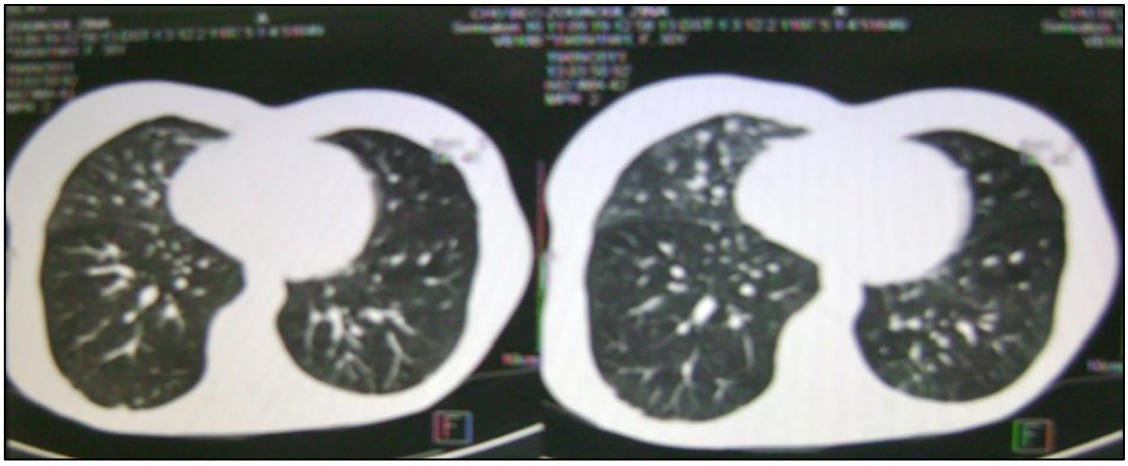

Figure 4 : Nodal nodules on CT scan

With the aim of treatment, the patient was initiated on corticosteroids at a dose of 1 mg/kg/day, followed by a gradual tapering. This treatment was complemented by the prescription of an immunosuppressant (Immurel) at a dosage of 100 mg per day. Under this therapy, the patient's clinical evolution rapidly improved. A significant regression of muscle weakness was observed, along with the disappearance of respiratory symptoms and an improvement in skin condition with the resolution of skin eruptions. Additionally, hepatic parameters returned to normal and signs of inflammation decreased. On the biological level, hematuria and proteinuria resolved, leading to the presence of microalbuminuria measured at 80 mg/24 hours. This positive response to treatment underscores the effectiveness of the therapeutic approach chosen in managing the glomerular syndrome associated with dermatomyositis in this patient. However, close and continuous monitoring will be necessary to ensure the maintenance of this long-term clinical and biological improvement.

DISCUSSION

The annual incidence of antisynthetase syndrome is estimated to be 1.25 to 2.5 cases per million inhabitants, with a prevalence of 1.5 cases per 100,000 people. Although this makes it a rare disease, its prevalence may be underestimated due to diagnostic challenges [1]. Antisynthetase syndrome primarily affects adults, with a slight female predominance. The exact pathogenesis of antisynthetase syndrome remains largely unknown, although immune dysregulation is implicated. It is believed that autoantibodies, especially antisynthetases such as anti-Jo-1 antibodies, play a key role in the inflammatory cascade observed in antisynthetase syndrome [2]. These autoantibodies target aminoacyl-tRNA synthetases, leading to immune activation and systemic inflammatory response.

Clinical manifestations of antisynthetase syndrome are varied and can affect multiple organ systems. In addition to the classic triad of inflammatory myopathy, interstitial lung disease [3], and arthritis, patients may present with Raynaud's phenomenon, hyperkeratotic fissures of the finger pads (mechanic's hands), and in some cases, renal involvement in the form of glomerulonephritis [4]. Although less frequent than other manifestations, renal involvement in antisynthetase syndrome is an important complication. Data on the prevalence of renal involvement in the syndrome remain limited, but it can manifest as acute or chronic renal failure, as well as significant proteinuria (> 0.3 g/24 hours). The exact mechanisms of renal involvement in antisynthetase syndrome are not fully understood, but a variety of renal lesions, including immune complex glomerulonephritis, tubulointerstitial, and vascular involvement, have been described. Diagnosis of antisynthetase syndrome relies on a combination of clinical manifestations, laboratory and imaging findings, as well as detection of specific autoantibodies such as anti-Jo-1 antibodies [5, 6]. Management of antisynthetase syndrome is multidisciplinary, involving rheumatologists, pulmonologists, dermatologists, and sometimes nephrologists. Treatment aims to control inflammation and prevent complications, often with corticosteroids, immunosuppressants, and sometimes biologic agents [7, 8].

CONCLUSION

Antisynthetase syndrome (ASS) can present in various clinical ways, making it essential to consider its diagnosis in atypical presentations such as polyarthritis, pulmonary fibrosis, or hyperkeratosis of the hands. Early management is crucial, especially due to the often unfavorable prognosis associated with interstitial lung involvement. Early screening and appropriate treatment can help prevent or delay the development of pulmonary fibrosis, underscoring the importance of prompt recognition of ASS. In cases where glomerulonephritis is associated, immunosuppressive intervention may be necessary to prevent progression to chronic renal failure. In summary, early diagnostic and therapeutic approaches are critical for improving the prognosis and quality of life of patients with antisynthetase syndrome.

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