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Complex Case of Systemic Sclerosis-Associated Interstitial Lung Disease: A Multidisciplinary Approach to Diagnosis and Management

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A B S T R A C T

Systemic sclerosis (Ssc), commonly known as scleroderma, is a rare and complex autoimmune disease that can affect multiple organ systems, including the skin and lungs. Interstitial lung disease (ILD) is a frequent and severe pulmonary complication of SSc, often associated with poor outcomes. We present a challenging case of a 42-year-old female with SSc-associated ILD, highlighting the importance of a multidisciplinary approach in both diagnosis and management.

Keyword: Scleroderma; ILD; Systemic Sclerosis

Introduction

Scleroderma, also known as systemic sclerosis (Ssc), is a rare and complex autoimmune disorder that primarily affects the connective tissues of the body. It is characterized by excessive collagen production, resulting in skin thickening and fibrosis. However, SSc is not limited to just the skin; it can also impact various internal organs, with the lungs being a common target. This pulmonary involvement often takes the form of interstitial lung disease (ILD), a condition that can significantly affect patients' quality of life and overall prognosis.[1,2]

ILD is a broad term encompassing various lung disorders characterized by inflammation and fibrosis of the interstitium, the tissue that supports the air sacs in the lungs. Scleroderma-associated ILD (SSc-ILD) is a major pulmonary manifestation of SSc and contributes significantly to the morbidity and mortality associated with the disease. Understanding the relationship between SSc and ILD is crucial for effective diagnosis and management.[2,3]

The exact cause of SSc and SSc-ILD remains unclear, but it is believed to result from a combination of genetic and environmental factors.[4] Autoimmune processes play a central role, leading to excessive collagen production, immune system dysregulation, and chronic inflammation. As a result, the lung's delicate interstitial tissue becomes scarred and stiff, impairing its ability to expand and contract efficiently. This leads to progressive dyspnea, cough, and reduced lung function.[5,6]

Diagnosing SSc-ILD typically involves a combination of clinical, radiological, and serological findings. Patients may present with symptoms such as shortness of breath, cough, and digital ulcers. High-resolution computed tomography (HRCT) scans of the chest can reveal characteristic findings, including ground-glass opacities and reticulation. Serological markers like antinuclear antibodies (ANA), anti-Scl-70 antibodies, and anti-centromere antibodies can be helpful in supporting the diagnosis.[7]

Treatment strategies for SSc-ILD are evolving, but they primarily focus on managing symptoms, slowing disease progression, and improving quality of life. Immunosuppressive agents, such as cyclophosphamide and mycophenolate mofetil, are commonly prescribed to modulate the immune response and reduce inflammation. Glucocorticoids may be used in combination with immunosuppressants for acute exacerbations, although long-term use is discouraged due to potential side effects.[8,9]

Supportive care measures include oxygen therapy, pulmonary rehabilitation, and vaccinations to reduce the risk of infections. In some cases, lung transplantation may be considered for end-stage disease when conservative therapies fail to halt disease progression.

Case Presentation

A 42-year-old woman presented with a 3-year history of Raynaud's phenomenon, skin tightening, and dyspnea on exertion. Physical examination revealed digital ulcers, skin thickening, and fine end-inspiratory crackles on lung auscultation. Pulmonary function tests showed restrictive lung disease, and high-resolution computed tomography (HRCT) demonstrated a reticular pattern with ground-glass opacities consistent with ILD.

Further evaluation included serologic testing, which revealed positive antinuclear antibodies (ANA), anti-Scl-70 antibodies, and anti-centromere antibodies. Echocardiography showed no evidence of pulmonary hypertension. A bronchoalveolar lavage was performed, revealing a lymphocytic predominance.

The patient underwent a multidisciplinary evaluation involving rheumatologists, pulmonologists, and radiologists, with input from dermatologists and pathologists. A diagnosis of SSc-associated ILD was established based on the 2013 ACR/EULAR classif-ication criteria and the American Thoracic Society (ATS)/European Respiratory Society (ERS) classification of ILD.

Treatment and Follow-up

The patient was initiated on a therapeutic regimen that combined immunosuppressive agents (cyclophosphamide) with glucocorticoids. Her Raynaud's phenomenon was managed with calcium channel blockers and prostacyclin analogs. Close monitoring was performed, including regular pulmonary function tests, HRCT scans, and echocardiography.

Despite initial treatment, the patient continued to experience progressive lung involvement, highlighting the need for aggressive therapeutic intervention. Consequently, she was transitioned to mycophenolate mofetil. Additionally, palliative measures such as oxygen supplementation and pulmonary rehabilitation were introduced.

Discussion

The presented case of a 42-year-old woman illustrates the diagnostic and management challenges associated with systemic sclerosis (SSc) and its commonly associated complication, interstitial lung disease (ILD). This complex clinical scenario showcases the importance of a multidisciplinary approach to navigate the intricacies of the disease and improve patient outcomes.

The patient's clinical presentation is indicative of systemic sclerosis, with prominent features being Raynaud's phenomenon, skin tightening, and exertional dyspnea. Raynaud's phenomenon is often an early sign

of connective tissue disease and is characterized by exaggerated vasoconstriction in response to cold or emotional stress. Skin involvement, with digital ulcers and skin thickening, is consistent with SSc. The presence of fine end-inspiratory crackles on lung auscultation and restrictive lung disease on pulmonary function tests raise concern for ILD, a common and severe pulmonary complication in SSc.

The high-resolution computed tomography (HRCT) findings play a critical role in confirming ILD. The reticular pattern with ground-glass opacities seen in HRCT is characteristic of ILD and indicates fibrotic and inflammatory changes in the lung interstitium. These radiological findings support the suspicion of ILD in this patient.

The positive serologic findings of antinuclear antibodies (ANA), anti-Scl-70 antibodies, and anti-centromere antibodies are essential for diagnosis. ANA is a general marker of autoimmune disease, while the presence of anti-Scl-70 antibodies is strongly associated with SSc and, in particular, with a higher risk of ILD. Anti-centromere antibodies are often associated with a distinct subtype of SSc and may indicate a milder form of ILD.

The echocardiography findings indicating no evidence of pulmonary hypertension are reassuring. Pulmonary hypertension is a common complication of SSc and can further complicate ILD. Its absence suggests that the patient's cardiopulmonary status may be relatively stable.

The lymphocytic predominance on bronchoalveolar lavage can be a valuable diagnostic clue. An elevated lymphocytic cell count in BAL is a feature seen in several autoimmune-related ILDs, including SSc-ILD. It underscores the importance of integrating clinical, radiological, and laboratory data for a comprehensive diagnosis.

The collaborative approach involving rheumatologists, pulmonologists, radiologists, dermatologists, and pathologists is essential in cases like this one. SSc-ILD is a complex disease that requires input from various specialists to ensure accurate diagnosis and appropriate treatment. The 2013 ACR/EULAR classification criteria and the ATS/ERS classification of ILD provide valuable guidance for establishing a diagnosis.

Early diagnosis of SSc-ILD is critical, as it allows for timely intervention. In this case, the patient's ILD was diagnosed promptly, and she was initiated on an immunosuppressive treatment regimen. Given the progressive nature of SSc-ILD, close monitoring, adjusting treatment as necessary, and providing supportive care are crucial components of managing this condition. The utilization of pulmonary rehabilitation and oxygen therapy can significantly improve the patient's quality of life.

Conclusion

Systemic sclerosis-associated interstitial lung disease remains a formidable clinical challenge. This case report illustrates the complexities involved in diagnosing and managing SSc-ILD and highlights the necessity of a collaborative approach among healthcare professionals to optimize patient care. As research into SSc and ILD

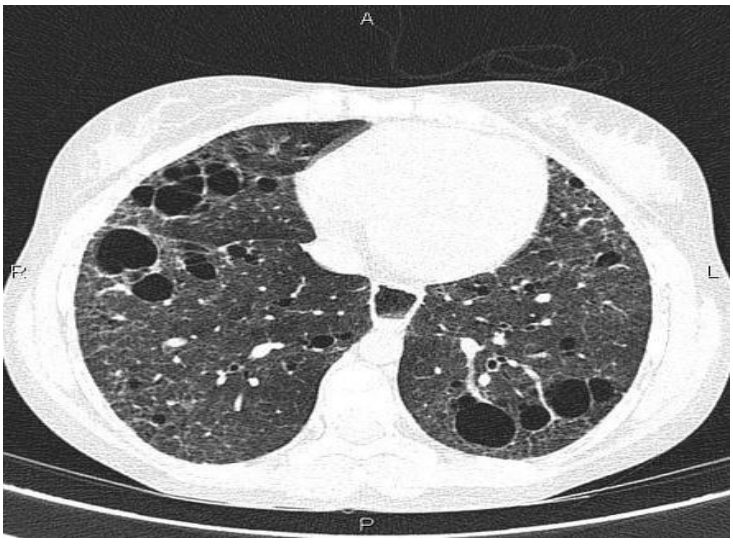

Figure 1. Interstitial lung disease associated with systemic sclerosis

continues, novel therapies and diagnostic strategies may offer hope for better outcomes in the future.

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