



ORIGINAL RESEARCH PAPER

General Medicine

A CASE REPORT EVALUATING DUAL PULMONARY INVOLVEMENT (INTERSTITIAL LUNG DISEASE AND PULMONARY ARTERIAL HYPERTENSION) IN A PATIENT WITH SYSTEMIC SCLEROSIS AT A TERTIARY CARE CENTRE

KEY WORDS:

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BACKGROUND

Systemic sclerosis is a chronic autoimmune connective tissue disorder characterized by immune dysregulation, progressive fibrosis, and widespread microvascular damage affecting the skin and multiple internal organs. Among its systemic manifestations, pulmonary involvement remains the most significant determinant of morbidity and mortality. Interstitial lung disease (ILD) and pulmonary arterial hypertension (PAH) are the two most serious pulmonary complications associated with systemic sclerosis. Individually, each carries a poor prognosis; however, their coexistence represents a particularly severe disease phenotype with higher mortality rates and limited therapeutic options.

ILD in systemic sclerosis results from progressive fibrosis of the pulmonary interstitium, leading to restrictive lung disease and impaired gas exchange. PAH, on the other hand, develops due to obliterative vasculopathy of the pulmonary arterioles, resulting in elevated pulmonary artery pressures and right heart dysfunction. The presence of both conditions poses significant diagnostic challenges, as symptoms such as dyspnea and fatigue may overlap, often leading to delayed diagnosis. Furthermore, management becomes complex, as immunosuppressive therapy for ILD and vasodilator therapy for PAH must be carefully balanced.

Early detection through vigilant screening and a multidisciplinary approach has been shown to improve outcomes. Reporting such cases is essential to enhance clinical awareness, promote early diagnosis, and highlight the importance of regular pulmonary evaluation in patients with systemic sclerosis. This case report aims to contribute to the existing literature by emphasizing the clinical significance of dual pulmonary involvement in systemic sclerosis.

Aim:

To present a case of systemic sclerosis complicated by interstitial lung disease and pulmonary arterial hypertension and to discuss its diagnostic and management challenges.

Objectives:

1. To describe the clinical presentation and systemic features of systemic sclerosis.
2. To evaluate pulmonary involvement using radiological and cardiac investigations.
3. To highlight the coexistence of ILD and PAH as a severe disease subset.
4. To emphasize the importance of early diagnosis and multidisciplinary management in improving patient outcomes.

Material and Methods

This study is a descriptive case report of a patient diagnosed with systemic sclerosis who presented to a tertiary care

hospital with respiratory complaints. A detailed clinical history was obtained, focusing on symptoms suggestive of connective tissue disease and pulmonary involvement. Thorough physical examination was conducted to identify cutaneous and systemic manifestations of systemic sclerosis. Laboratory investigations included routine hematological and biochemical tests along with an autoimmune profile. Immunological workup comprised antinuclear antibody (ANA) testing and disease-specific antibodies relevant to systemic sclerosis. Pulmonary evaluation was carried out using high-resolution computed tomography (HRCT) of the chest to assess interstitial lung involvement. Cardiac assessment included echocardiography to evaluate pulmonary artery pressures. Additional supportive investigations were performed as required.

The patient's treatment plan, response to therapy, and clinical course were documented. Data was analysed descriptively to correlate clinical findings with diagnostic results.

RESULTS

The patient presented with progressive dyspnea on exertion, fatigue, and features suggestive of a connective tissue disorder. Cutaneous manifestations consistent with systemic sclerosis were observed on physical examination, along with features of peripheral vasculopathy. Pulmonary examination revealed findings suggestive of restrictive lung disease.

HRCT of the chest demonstrated radiological features consistent with interstitial lung disease with subpleural sparing-NSIP. 2D echocardiography revealed elevated pulmonary artery pressures, indicative of pulmonary arterial hypertension. The autoimmune profile supported the diagnosis of systemic sclerosis.

- ANA by IF-3.93
- Anti Scl-positive

Based on clinical, radiological, and immunological findings, a diagnosis of systemic sclerosis complicated by ILD and PAH was established. The patient was initiated on immunosuppressive therapy with Mycophenolate mofetil for ILD along with targeted pulmonary vasodilator therapy for PAH-Tadalafil. Supportive management was provided. The patient showed symptomatic improvement and clinical stabilization.

DISCUSSION AND CONCLUSION

Pulmonary involvement is the leading cause of mortality in systemic sclerosis, with ILD and PAH being the most critical complications. The coexistence of both conditions signifies advanced disease and is associated with a poorer prognosis compared to either condition alone. Early symptoms may be nonspecific, often resulting in delayed recognition. Therefore, a high index of suspicion and regular screening using HRCT echocardiography are essential in patients with systemic sclerosis.

Management of patients with both ILD and PAH remains challenging and requires a multidisciplinary approach involving rheumatologists, pulmonologists, and cardiologists. Early initiation of appropriate therapy can slow disease progression and improve quality of life.

This case highlights the importance of comprehensive pulmonary evaluation in systemic sclerosis and underscores the need for early diagnosis and prompt management of dual pulmonary complications to improve patient outcomes.

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