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Systemic Sclerosis with Interstitial Lung Disease: A Case Report

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Abstract: ***Background: A 45 - year - old female presented with breathlessness (MMRC grade 2), fatigue, and generalized myalgia for 10 days. Initial investigations indicated normocytic hypochromic anemia, elevated ESR and CRP levels, moderate pleural effusion on 2D echocardiography, and subpleural fibrosis with ground - glass opacities on HRCT. Cardiology and pulmonology consultations suggested the possibility of interstitial lung disease (ILD). Given the suspected systemic autoimmune involvement, rheumatology and ophthalmology evaluations were conducted, leading to further diagnostic testing. Method: A comprehensive diagnostic workup included blood tests, imaging studies, and specialist consultations. Hemoglobin levels were measured, and a peripheral blood smear was analyzed. ESR and CRP levels were assessed to determine inflammation. Echocardiography and HRCT were performed to evaluate cardiac and pulmonary involvement. Ophthalmological examination included Schirmer's test for Sjögren's syndrome. Rheumatological evaluation involved ANA with ANA profile, LDH, and serum ferritin measurements. Results: The patient was found to have normocytic hypochromic anemia (Hb: 9.9), elevated ESR (71), and CRP (9.6). Echocardiography revealed moderate pleural effusion and a normal ejection fraction (65%). HRCT showed subpleural fibrosis and ground - glass opacities. Schirmer's test for Sjögren's syndrome was negative. Rheumatological tests revealed positive ANA with a cytoplasmic pattern and SSA/Ro - 52kD antibodies, elevated LDH (457), and serum ferritin (617). The patient was diagnosed with systemic sclerosis with ILD. Conclusion: The patient was treated with corticosteroids, immunosuppressive agents, calcium supplements, antivertigo medications, and other supportive treatments, resulting in symptomatic improvement. This case highlights the importance of a multidisciplinary approach in diagnosing and managing systemic sclerosis with ILD.***

Keywords: Systemic sclerosis, Interstitial lung disease, ANA profile, Ground - glass opacities, Pleural effusion, Immunosuppressive therapy, Multidisciplinary approach.

1. Introduction

Systemic sclerosis (SSc) is a chronic connective tissue disease characterized by widespread microvascular damage and fibrosis of the skin and internal organs. Interstitial lung disease (ILD) is a common and serious complication of SSc, often leading to significant morbidity and mortality. This case report discusses the presentation, diagnostic challenges, and management of a patient with SSc complicated by ILD.

2. Case History

A 45 - year - old female presented with complaints of breathlessness (MMRC grade 2), fatigue, and generalized myalgia for 10 days. Her past medical history was unremarkable. Initial physical examination revealed pallor but no other significant findings.

Laboratory investigations showed a hemoglobin level of 9.9 g/dL with a peripheral blood smear indicating normocytic hypochromic anemia. The erythrocyte sedimentation rate (ESR) was markedly elevated at 71 mm/hr, and C - reactive protein (CRP) was 9.6 mg/L, suggesting systemic inflammation. A 2D echocardiogram revealed moderate pleural effusion with a preserved ejection fraction of 65%. High - resolution computed tomography (HRCT) of the chest demonstrated subpleural fibrosis and ground - glass opacities in the bilateral lung fields.

Given the findings, a cardiology consultation was obtained, which recommended further evaluation by rheumatology and pulmonology specialists. The pulmonology department suspected the development of ILD and initiated further workup. Ophthalmological evaluation, prompted by the suspicion of Sjögren's syndrome, included a Schirmer's test which was negative.

The rheumatology assessment raised the possibility of systemic sclerosis. Further serological tests included an antinuclear antibody (ANA) test, which was positive with a cytoplasmic pattern. The ANA profile identified SSA/Ro - 52kD antibodies. Additionally, lactate dehydrogenase (LDH) was elevated at 457 U/L, and serum ferritin was significantly increased at 617 ng/mL.

Based on these findings, the patient was diagnosed with systemic sclerosis with interstitial lung disease. She was treated with corticosteroids, immunosuppressive agents, calcium supplements, antivertigo medications, and other supportive treatments. Over the course of her hospital stay, she showed significant symptomatic improvement and was discharged with a comprehensive management plan.

3. Discussion

Systemic sclerosis is a multisystem autoimmune disorder that often presents diagnostic and therapeutic challenges. The presence of ILD in SSc patients is a critical determinant of prognosis. Early diagnosis and intervention are essential to manage symptoms and slow disease progression.

The patient's presentation with breathlessness, fatigue, and generalized myalgia, combined with laboratory findings of anemia, elevated inflammatory markers, and imaging studies indicating pulmonary involvement, pointed towards a systemic inflammatory process. The diagnosis was further supported by serological tests showing a positive ANA with a specific cytoplasmic pattern and the presence of SSA/Ro - 52kD antibodies.

The management of SSc with ILD typically involves immunosuppressive therapy to control the underlying autoimmune process and reduce inflammation. Corticosteroids are often used initially for their potent anti - inflammatory effects. This patient's treatment regimen, which included steroids and immunosuppressive agents, led to significant symptomatic relief.

This case underscores the importance of a multidisciplinary approach involving rheumatologists, pulmonologists, cardiologists, and other specialists. Such collaboration is crucial for comprehensive care and optimal patient outcomes.

4. Conclusion

This case report highlights the complexity of diagnosing and managing systemic sclerosis with interstitial lung disease. A multidisciplinary approach and prompt, appropriate treatment can lead to significant symptomatic improvement and better patient outcomes. Early recognition and management of ILD in SSc are vital to improve quality of life and prognosis.

References

[1] Denton CP, Khanna D. Systemic sclerosis. Lancet.2017; 390 (10103): 1685 - 1699.
[2] Varga J, Trojanowska M, Kuwana M. Pathogenesis of systemic sclerosis: recent insights of molecular and cellular mechanisms and therapeutic opportunities. J Scleroderma Relat Disord.2017; 2 (3): 137 - 152.
[3] Hoffmann - Vold AM, Fretheim H, Midtvedt Ø, et al. Tracking impact of interstitial lung disease in systemic sclerosis in a complete nationwide cohort. Am J Respir Crit Care Med.2019; 200 (10): 1258 - 1266.
[4] Khanna D, Mittoo S, Aggarwal R, et al. Connective tissue disease - associated interstitial lung diseases (CTD - ILD) - report from OMERACT CTD - ILD working group. J Rheumatol.2015; 42 (11): 2168 - 2171.
[5] Tashkin DP, Roth MD, Clements PJ, et al. Mycophenolate mofetil versus oral cyclophosphamide in scleroderma - related interstitial lung disease (SLS II): a randomised controlled, double - blind, parallel group trial. Lancet Respir Med.2016; 4 (9): 708 - 719.