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Case Report

Pulmonary Manifestations in Systemic Sclerosis

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ABSTRACT:
Scleroderma, also known as progressive systemic sclerosis (SSc), is a multisystem autoimmune disorder characterized by inflammation and fibrosis involving the skin as well as internal organs such as the vasculature, esophagus, and the respiratory tract. Pulmonary involvement consists most often of interstitial fibrosis and pulmonary vascular disease leading to pulmonary arterial hypertension (PAH). Interstitial lung disease (ILD) is the leading cause of death in patients with SSc. The extent of ILD on HRCT in initial evaluation and decline in PFT during the preceding 12 months are helpful in identifying such patients. PFT should be repeated every 6 monthly for patients at risk.
Key words: Pulmonary, Systemic Sclerosis

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INTRODUCTION

Systemic Sclerosis is a complex and clinically heterogenous disease with protean clinical manifestations causing significant disfigurement and mortality. The two principal forms of lung involvement in systemic sclerosis include:

Interstitial lung disease (ILD) in scleroderma:
SSc-associated ILD consists of various histopathologic subtypes, most commonly nonspecific interstitial pneumonitis and usual interstitial pneumonitis. Pulmonary function tests suggests a restrictive defect in patients with ILD. A decrease in the DLCO may be the first PFT abnormality seen in SSc-associated ILD.[1-3]

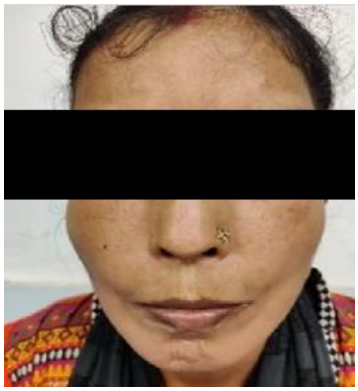
HRCT scanning- The early radiologic changes of SSc-associated ILD are usually found in the dependent lung areas. Among patients with SSc-associated ILD, the most common pathologic pattern, NSIP, is associated with the HRCT finding of ground glass opacities in a peripheral distribution.

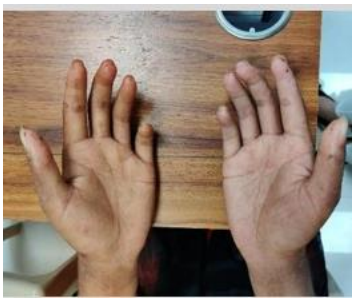
Pulmonary arterial hypertension (PAH) results from vascular remodelling of pulmonary arteries. It occurs as an isolated abnormality or in association with ILD. The prevalence of systemic SSc-related organ injury is challenging to estimate because it occurs early in SSc, such as pulmonary fibrosis in the first two years and renal crisis in the first four years of disease onset, even though patients are often asymptomatic. As patients with organ damage have a poor prognosis, all patients should be carefully evaluated and monitored via follow-up in the initial periods for organ involvement to facilitate the early identification and initiation of appropriate therapy.[4-6]

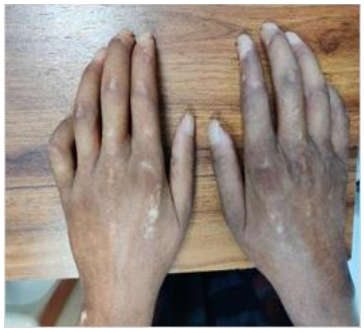
CASE REPORT

35 year old, female came with complain of breathlessness since 1 year , chest pain since 15 days skin tightness in both hands and stiffness of fingers since 15 days, digital ulcers since 15 days.

Patient is a known case of systemic sclerosis since 20 years. Patient had Raynaud's phenomenon and digital ulcers 20 years back during which the patient was diagnosed as systemic sclerosis and spirometry and 2d echo were normal. After 1 year patient had reduced mouth opening and stiffness and developed edema over all fingers. Over 5 years the patient developed new ulcers and contractures of the joints. The patient then had gastro-esophageal reflux disease and pulmonary artery hypertension since last 2 years.





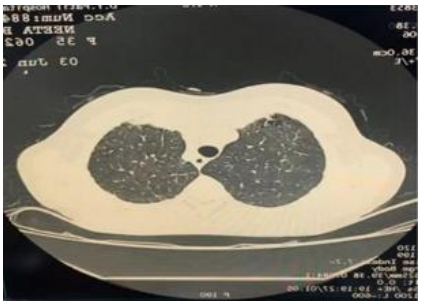


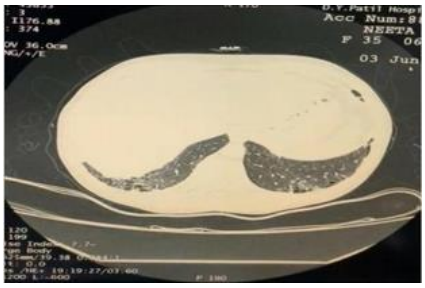
On examination: Skin changes included salt and pepper appearance , vitiligo, thickening of skin and induration. Raynaud's phenomena, reduced oral aperture, joint contracture of the hand were the other findings. On auscultation bilateral breath sounds were reduced and bilateral crepts and loud P2.

ANA by IF	Positive (Homogenous) 1:1000
DNA (Double Strand) Antibody NcX	Negative (33.64)
U1-snRNP IgG	Negative
Jo-1 Antibody IgG	Negative
Centromere Antibody IgG	Negative
Scl-70 IgG	Positive (>200)
SSA-RO IgG	Negative
SSA-La IgG	Negative
Sm Antibody, IgG	Negative
RNP-Sm Antibody IgG	Negative

Investigations: 1) CBC:-Haemoglobin:-10.9 g/dl, Wbc total count:-5,600 ; Platelets:-2,21,000 ; MCV:-84 2) Creatinine:- 0.71 mg/dl 3)SGPT:-32 U/L 4)ESR:-100 mm/hr.

HRCT Thorax:- Macrocystic honeycombing along basal and subpleural regions of bilateral lower lobes suggestive of interstitial lung disease.





Pulmonary Function Test:-Reduced DLCO. Mild restriction and air trapping.
2Decho:- LVEF:-60%,
Normal LV cavity with normal LV function. PASP:-52mmHg.
Treatment given:-
The patient was started on Mycofenolate mofetil (500 mg) twice day, Tadalafil (20) twice a day, Ambrisentan (5 mg) once a day, Diltazem (30 mg) twice a day,Pirfenadone (200 mg) thrice a day, Pentoxyfylline (400mg) thrice a day,T. Methotrexate(7.5 mg), Rabeprazole (20 mg) twice a day, Foracort (Formoterol (6 mcg)+ Budesonide(200mcg)) inhaler 2 puffs twice a day.

DISCUSSION

Scleroderma or systemic sclerosis (SSc) is a heterogeneous disorder characterized by endothelial dysfunction, dysregulation of fibroblasts resulting in excessive production of collagen, and profound abnormalities of the immune system. These changes cause progressive fibrosis of the skin and internal organs, system failure and death. While the etiology of SSc is generally unknown, genetic and environmental factors are thought to contribute to host susceptibility. SSc, whether presenting in the limited or diffuse form, is a systemic disease with the potential for multiple organ system involvement including the gastrointestinal, cardiac, renal, and pulmonary systems. Pulmonary manifestations of SSc include, but are not limited to, pulmonary vascular diseases such as pulmonary arterial hypertension (PAH) and pulmonary veno-occlusive disease (PVOD), interstitial lung disease (ILD), and increased susceptibility to lung neoplasms.[7- 9]

In the present case repot, 35 year old, female came with complain of breathlessness since 1 year , chest pain since 15 days skin tightness in both hands and stiffness of fingers since 15 days, digital ulcers since 15 days. Patient is a known case of systemic sclerosis since 20 years. Skin changes included salt and pepper appearance , vitiligo, thickening of skin and induration. Raynaud's phenomena, reduced oral aperture, joint contracture of the hand were the other findings. The patient was started on Mycofenolate mofetil (500 mg) twice day, Tadalafil (20) twice a day, Ambrisentan (5 mg) once a day, Diltazem (30 mg) twice a day,Pirfenadone (200 mg) thrice a day, Pentoxyfylline (400mg) thrice a day,T. Methotrexate(7.5 mg), Rabeprazole (20 mg) twice a day, Foracort (Formoterol (6 mcg)+ Budesonide(200mcg)) inhaler 2 puffs twice a day.

Goldin JG et al, summarized that the investigations in systemic sclerosis are pulmonary function test which showed restrictive pattern and high resolution computerised tomography (HRCT) – NSIP pattern is most common. However, a UIP pattern showing peripheral and basilar predominant reticulation and honeycombing without significant ground glass opacities, can also be seen, but is more common in patients with LcSSc. Immunosuppressants are the main stay of treatment for SSc. Cyclophosphamide, Mycophenolate Mofetil, Corticosteroids (usually in combination with the above drugs) are the preferred drugs. Other therapies under investigation are Bosentan (Endothelin receptor antagonist), Imatanib (tyrosine kinase inhibitor), Rituximab (a monoclonal antibody directed against the CD20 antigen on the surface of B lymphocytes) and Stem cell transplant.[10, 11] Sugino, K et al reported the case report of a 65-year-old woman with a 35-year history of limited cutaneous systemic scleroderma was admitted to our hospital complaining of a 3-month history of progressive dyspnoea on exertion. High-resolution CT images of the chest revealed diffuse reticular opacities and traction bronchiectasis predominantly in the bilateral lower lobes of the lung. Specimens obtained during video-assisted thoracic surgery were consistent with fibrocellular non-specific interstitial pneumonia and accompanied by accumulation of lymph follicles within areas of fibrosis. Although the patient received combination therapy with prednisolone and intravenous cyclophosphamide at a dosage of 500 mg/m2 monthly for 5 months, her clinical condition deteriorated gradually. In addition, right heart catheterisation revealed borderline pulmonary arterial hypertension with mean pulmonary artery pressure of 24 mm Hg. Therefore, they initiated a combination therapy of an antifibrotic agent, pirfenidone for 12 months, and the dual endothelin receptor antagonist, macitentan, with prednisolone. As a result, her clinical condition improved dramatically.[12]

CONCLUSION

ILD is the leading cause of death in patients with SSc. The extent of ILD on HRCT in initial evaluation and decline in PFT during the preceding 12 months are helpful in identifying such patients. PFT should be repeated every 6 monthly for patients at risk. Cyclophosphamide has been shown to reduce progression of SSc associated ILD but has potential toxicity. Mycophenolate mofetil is well tolerated and improves both skin induration and ILD. In March 2021 Tocilizumab has been approved by US Food and Drug administration for slowing the rate of decline in pulmonary function in adult patients. In patients who show continuous progression of ILD despite medical therapy, lung transplantation may be considered as a life prolonging procedure.

REFERENCES

1. Hassoun PM. Lung involvement in systemic sclerosis. Presse Med. 2011;40:0.
2. Sharma VK, Trilokraj T, Khaitan BK, Krishna SM. Profile of systemic sclerosis in a tertiary care center in North India. Indian J Dermatol Venereol Leprol. 2006;72:416.
3. Deepa AS, Rachel RP, Ramchandran P, Devaraj U, Arnold SA, Shobha V, D'souza G. Pulmonary involvement in systemic sclerosis: a clinical profile. Lung India. 2016;33:144–147.
4. Mayes MD. Scleroderma epidemiology. Rheum Dis Clin North Am. 2003;29:239–54.
5. Silman A, Jannini S, Symmons D, Bacon P. An epidemiological study of scleroderma in the West Midlands. Br J Rheumatol. 1988;27:286–90.
6. Tamaki T, Mori S, Takehara K. Epidemiological study of patients with systemic sclerosis in Tokyo. Arch Dermatol Res. 1991;283:366–71.
7. Roumm AD, Medsger TA., Jr Cancer and systemic sclerosis. An epidemiologic study. Arthritis Rheum. 1985;28(12):1336–40.
8. Chatterjee S, Dombi GW, Severson RK, Mayes MD. Risk of malignancy in scleroderma: a population-based cohort study. Arthritis Rheum. 2005;52(8):2415–24
9. Olsen NJ, King LE, Jr, Park JH. Muscle abnormalities in scleroderma. Rheum Dis Clin North Am. 1996;22(4):783–96
10. Goldin JG, Lynch DA, Strollo DC, et al. High-resolution CT scan findings in patients with symptomatic scleroderma-related interstitial lung disease. Chest. 2008;134:358–67.
11. Scholand, et al. Interstitial lung disease in systemic sclerosis: diagnosis and management. Rheumatology. 2012;S1:008.
12. Sugino, K., Kabuki, T., Shibuya, K., & Homma, S. (2018). Systemic scleroderma-related interstitial pneumonia associated with borderline pulmonary arterial hypertension. BMJ case reports, 2018, bcr201722175