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scleroderma - Pulmonary manifestations

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Dr. N. Gopichand	Dr. N Snigdha
Associate Prof of Pulmonology ACSR Government Medical college. Nellore	Post graduate student Pulmonary medicine Siddhartha medical college Vijayawada

ABSTRACT *Scleroderma is a chronic disease of unknown etiology, usually classified with the collagen diseases. The term diffuse or progressive systemic sclerosis has been introduced to describe those cases showing visceral as well as cutaneous manifestations. The essential pathological features are swelling and disintegration of the collagen tissue accompanied by cellular infiltration and fibrosis; the process may also involve the intima of small vessels with fibrinoid necrosis and occlusion. In this article we would like to present two cases of systemic sclerosis. Case 1: A 30 year old lady showed early changes suggestive of systemic sclerosis along with marked skin changes. Her ANA profile supported Diffuse systemic sclerosis .Case 2:A 64 year old female complaining of severe dyspnoea presented with marked skin changes which were pathognomic of scleroderma .On evaluation findings supported diffuse systemic sclerosis and radiologically showed late changes in respiratory system. We intend to report these two cases to show two different ends in the spectrum of presentation of pulmonary manifestations of systemic sclerosis. Case 1 had less symptomatology compared to Case 2.*

Introduction:

Lung involvement in systemic sclerosis was first described at the end of the last century (Lewin and Heller, 1894), and there have been several more recent reports (Lloyd and Tonkin, 1948; Shuford, Seaman, and Goldman, 1953; Opie, 1955).

Scleroderma or systemic sclerosis (SSc) is a clinically heterogeneous, multi-system autoimmune disorder characterized by endothelial dysfunction, deregulation of fibroblasts resulting in excessive production of collagen and autoantibody formation with profound abnormalities of the immune system. These changes cause progressive fibrosis of the skin and internal organs, system failure and death [1]. Patients with SSc may exhibit proliferative small artery and obliterative micro vascular disease, plus inflammation and fibrosis affecting the skin, oesophagus, respiratory tract and other target organs [2].

Pulmonary involvement is second in frequency only to oesophagus involvement as a visceral complication of systemic sclerosis and has surpassed renal involvement as the most common cause of death. Interstitial lung disease (ILD) and pulmonary vascular disease, particularly pulmonary arterial hypertension (PAH), are the most commonly encountered types of lung involvement [3] Incidence of scleroderma is about 9-19 per million population. The symptoms are usually breathlessness on exertion and cough producing scanty mucoid sputum. Disease course, severity, and organ involvement are variable from patient to patient, and overlap syndromes with other autoimmune conditions can occur [4] . Pulmonary involvement like Interstitial Lung Disease (ILD) and Pulmonary arterial hypertension (PAH) are common and occurs in all SSc subsets. Progression of lung disease in SSc is variable and difficult to predict.

Case Report 1 : A 30 year old female home-maker presented with shortness of breath and dry cough since 3 months. On general examination she had pallor, and showed shiny ,light hide bound skin over extensor aspect of forearm and front of legs. Salt and pepper pigmentation over both ears, microstomia, digital pitting scars were seen. Respiratory examination revealed bilateral normal vesicular breath sounds with basal fine end inspiratory crepitations. Electrocardiography showed sinus tachycardia. Chest X-ray PA view,2-D Echo were normal. High Resolution computed Tomography showed peripheral ground glass pattern in basal segments and bilateral lower zones which is consistent with nonspecific interstitial pneumonia(NSIP). Spirometry showed restrictive pattern. Complete hemogram showed microcytic hypochromic anemia.She was positive for serum antinuclear antibodies , anti scl70 antibody and negative for anticentromere antibody. Endoscopy showed erosive antral gastritis. Barium swallow showed delayed filling of barium in lower 1/3 of oesophagus. Liver Function Test's and Renal Function Test's were within normal limits. She was diagnosed to have diffuse type of systemic sclerosis.

Case Report 2 : A 64 year old female, referred from department of dermatology came with complaints of shortness of breath, dry cough dysphagia. She gave a history of skin lesions over back of trunk and forehead since 3 years, thickening of hands which gradually progressed to contractures since 1 year, difficulty in swallowing first to liquids then to solids since 7 months. On physical examination she was pale. Cutaneous pigmentation was seen over forehead, back of upper trunk. Loss of wrinkles over the face. Her scalp showed salt and pepper pigmentation. Digital pitting scars were present. Purse string appearance of the mouth with microstomia was seen. Sclerodactyly with flexion contractures of interphalangeal joints of little and ring fingers, thickening of skin up to middle of forearm, longitudinal ridging over nails was present. Oral and genital mucosa was normal. Reynaud's phenomena could not be elicited. Cardiovascular examination and per abdomen examination was unremarkable. On auscultation of lungs end inspiratory crepitations were present.

Her routine investigations – Complete blood picture, Random blood sugar, Liver function tests, Renal function tests,

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serum proteins were within normal limits. ESR was elevated 45 mm/hr. She was positive for serum antinuclear antibodies , anti scl70 antibody and negative for anticentromere antibody .Chest X-ray showed bilateral reticulonodular opacities in lower zones with obscuration of bilateral costophrenic angles, bilateral mild interstitial thickening was noted in bilateral mid and lower zones. High Resolution Computerised Tomography scan of the chest showed bilateral interlobular and intralobular septal thickening with mild honeycombing in bilateral mid and lower zones ,bronchiectatic changes in right upper lobe suggestive of interstitial lung disease. Electrocardiogram and 2D Echocardiography were normal. Barium swallow showed narrowing of the proximal and distal 1/3 rd of the esophagus.She was too dyspneic to perform spirometry.

Case1

Case 2:

Discussion:

PAH and ILD are most common pulmonary manifestations in Systemic sclerosis, with ILD of NSIP more commonly associated with diffuse subset. PAH can occur primarily in limited form whereas patients develop PAH secondary to ILD in case diffuse subset. Pulmonary complications are common in SSc and, SSc-ILD and SSc-PAH, the leading causes of death [5]. In case1 pulmonary symptoms were not prominent, and patient initially had no abnormal signs in the chest, although chest radiographs were abnormal .Other pulmonary manifestations include pleural involvement, aspiration pneumonia, Alveolar haemorrhage, small airway disease, malignancy, respiratory muscle weakness ,drug induced toxicity, spontaneous pneumothorax and pneumoconiosis.

Exertional dyspnea and dry cough are the most common presenting symptoms in patients with SSc who develop pulmonary involvement. However, dyspnoea may have many causes in patients with SSc and all SSc patients (with or without dyspnoea) should be thoroughly evaluated for the presence of lung involvement, regardless of their characterization as either lcSSc or dcSSc. However the outcome may vary. The identification and staging of pulmonary manifestation is of paramount importance to the management of patients [2].Steroids and antineoplastic drugs like cyclophosphamide, methotrexate, mycophenolate remain main stay of treatment. Anti-fibrotic drugs like D-penicillamine have a role.PAH is managed on drugs like sildenafil or bosentan.

CONCLUSION:

Unfortunately, systemic sclerosis lung disease is often not detected or diagnosed until the late stages, particularly in those who did not develop the classic signs of skin-hardening or sclerodactyly, or those who only exhibited subtle respiratory symptoms. Early diagnosis would enable us to prevent progression to severe forms of pulmonary fibrosis.

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