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Original Research Paper

Pulmonary Medicine

A RARE CASE OF PLEURO-SCLERODERMA

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KEYWORDS :

INTRODUCTION:

Scleroderma or systemic sclerosis (SSc) is an inflammatory-fibrotic disease that results in deposition of excessive extracellular matrix in the skin and several visceral organs including the lungs, heart, kidneys, and gastrointestinal tract(1). Pulmonary complications of SSc remain one of the largest causes of morbidity and mortality in the disease. Interstitial lung disease (ILD) and pulmonary arterial hypertension (PAH) are the most common forms of lung manifestation associated with SSc(2).

Case Report

A 37 year old male who presented with lesions over face, abdomen and hands for the past 6 months to Department of Dermatology, was referred to us for the c/o dry cough and shortness of breath. He is a known case of chronic anemia. No other positive history was present.

On examination he was pale with thickening of facial skin, thinned out lips with difficulty in opening the mouth. He had multiple well demarcated shiny indurated hyperpigmented patches which were warm, non-pinchable and with loss to sensation. Vitals were, Spo_2 92% on RA at rest, BP 130/80 mm/hg, PR 102/min, RR 24/min and Temp 37.6 C.

On auscultation, breath sound was absent in left infra-scapular and infra-axillary areas. Fine Late Inspiratory Crepitations heard in the Right Basal Areas of Chest.

Lab investigation showed Normocytic normochromic anemia. ABG showed Type I Respiratory failure. Chest x-ray showed a homogenous opacity in left lower zone with blunting of costophrenic angle (fig 1). HRCT chest showed few linear subpleural parenchymal opacities in posterior and lateral basal segments of right lower lobe without honeycombing – suggestive of ILD (fig 2).

Small to moderate left sided pleural effusion causing passive atelectasis of underlying basal lung. PFT showed restrictive pattern. DLCO showed reduced diffusing capacity. ANA positive with speckled pattern. Biopsy of skin lesions was suggestive of scleroderma.

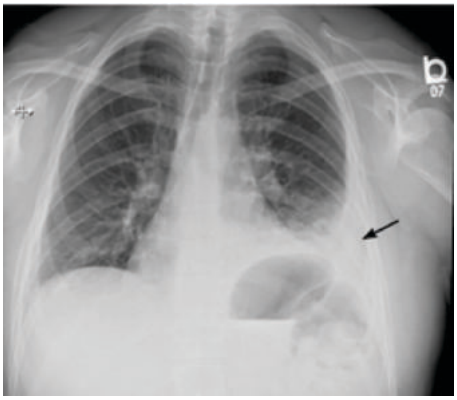

Fig1: Chest x-ray showed a homogenous opacity in left lower zone with blunting of costophrenic angle

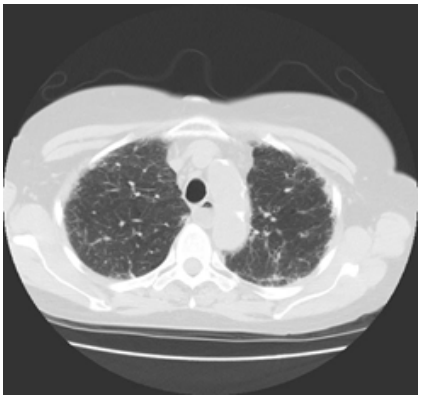

Fig 2: HRCT chest showed few linear subpleural parenchymal opacities right lower lobe without honeycombing

DISCUSSSION:

Rheumatologic disorder, such as Systemic Sclerosis (Scleroderma), may be associated with Interstitial Lung Disease(1). More than 80% of patients with systemic sclerosis have lung involvement but Pleural effusion is rare complications(1,2). Pleural effusion can be a complication for cardiac involvement if present but in this case, patient doesn't have any cardiac involvement to have pleural effusion(3). Pulmonary hypertension is commonly associated when skin involvement is present in systemic sclerosis(4). This case is a rare presentation where echo was normal but patient had pleural effusion on left side. ICD was done and 150ml of pleural fluid was drained and sent for analysis which was found to be exudative, ADA was 16, Gene-xpert MTB was not detected and cytology was normal.

CONCLUSION:

ILD and PAH are the leading causes of death in SSc patients(3). Due to other manifestations of their disease, as well as the asymptomatic nature of lung involvement in its early stages, pulmonary involvement often goes undiagnosed. Clinicians need to have a low threshold to evaluate for ILD and PAH in these patients. Once diagnosed, therapy or enrolment in a treatment trial is recommended. While lung transplant is an option in selected patients with advanced lung disease, other nonpharmacological treatment, interventions and goal-oriented measures may influence outcome for patients with pulmonary manifestations of Ssc.

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