

**A CASE OF SCLERODERMA MISDIAGNOSED AS PULMONARY TUBERCULOSIS**Dilbag Singh<sup>\*1</sup>, Amritpal Kaur<sup>1</sup>, Naveen Pandhi<sup>2</sup>, N. C. Kaja<sup>3</sup>, Mukul Sharma<sup>4</sup>, Srijna<sup>1</sup><sup>1</sup>Junior Resident, Department of Pulmonary Medicine, Government Medical College, Amritsar Punjab, India.<sup>2</sup>Professor and Head, Department of Pulmonary Medicine, Government Medical College, Amritsar.<sup>3</sup>Professor, Department of Pulmonary Medicine, Government Medical College, Amritsar Punjab, India.<sup>4</sup>General physician. Gurdaspur, Punjab, India.**\*Corresponding Author: Dilbag Singh**

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**ABSTRACT**

Systemic sclerosis (SSc) is a systemic autoimmune disease that is characterised by endothelial dysfunction resulting in a small-vessel vasculopathy, fibroblast dysfunction with resultant excessive collagen production and fibrosis, and immunological abnormalities. We present a case of scleroderma misdiagnosed as a case of pulmonary tuberculosis. Antibody profile was done and was found to be positive for ANA. She also had most of the classical features of scleroderma on clinical examination thus confirming the diagnosis. A detailed history, clinical examination, radiological examination, and immunological test helped in the diagnosis of the patient. We highlight the importance of suspicion, careful general examination, radiological assessment to confirm the diagnosis of scleroderma misdiagnosed as a case of pulmonary tuberculosis.

**KEYWORDS:** Scleroderma, Tuberculosis, Anti Nuclear Antibodies, Anti Topoisomerase antibodies.**INTRODUCTION**

Pulmonary involvement is common in patients with systemic sclerosis (SSc), and this leads to substantial morbidity and mortality. Multiple factors can cause pulmonary (lung) involvement in systemic scleroderma. Build-up of collagen thickens lung tissue and causes fibrosis or scarring, making the transport of oxygen into the bloodstream more difficult. Symptoms of lung involvement include shortness of breath, a decreased tolerance for exercise and a persistent cough. While virtually any organ system may be involved in the disease process, fibrotic and vascular pulmonary manifestations of SSc, including interstitial lung disease (ILD) and pulmonary hypertension (PH), are the leading cause of death. A combination of clinical, radiological and immunological criteria is used to diagnose scleroderma.<sup>[1]</sup> Patients with autoimmune diseases are known to develop infections like tuberculosis either due to the disease activity or secondary to the immunosuppressive therapy. Tuberculosis per se is known to induce the development of autoantibodies which in turn stimulate the manifestation of autoimmune diseases

**CASE REPORT**

A 45 year old female presented to Chest and Tb hospital Amritsar with complaints of breathlessness on exertion, cough with expectoration, fever, loss of appetite and weight loss for past six months. Other complaints were difficulty in swallowing and tightening of skin around

the mouth, bluish discoloration of fingers on exposure to cold with a history of chest pain, distension of abdomen and swelling over both feet. She had been evaluated in the peripheral hospital and was started on treatment for TB under a national program for the past 2 months but with no improvement. Thereafter, she presented to the hospital with worsening of breathlessness since the past 2 weeks.

On examination, patient was thin built. Patient had features of salt and pepper appearance, fish mouth deformity, pinched nose, facial melanosis, telangiectasia over the cheeks, sclerodactyly and resorption of digits diagnostic of scleroderma. There was pallor, cyanosis of fingers of hand, clubbing, and bilateral pitting pedal oedema. Pulse was 120/min and BP; 90/70mmhg. Respiratory examination revealed bilateral end inspiratory crackles with decrease intensity of breath sound at right and left infrascapular area with muffling of heart sounds with a pericardial rub. Other systems were normal.

Provisional diagnosis of systemic sclerosis with interstitial lung disease was made. ANA was sent as a part of autoimmune antibody profile which was positive, with a value of 144.8 AU/MI, contributing towards the diagnosis, then anti topoisomerases antibodies was sent which also came out to be positive. Further serological investigations revealed anaemia with Hb of 8.1gm%, elevated leucocyte count with 72% polymorphs and

elevated ESR. Renal and liver functions were within normal limits. HIV serology, HCV and HBS were Non-reactive. ECG shows low voltage complex and t wave inversion in chest leads, followed by echocardiography showing mild to moderate pericardial effusion present posterolaterally more on laterally (8mm). Mantoux test was negative, sputum for AFB and CBNAAT came out to be negative. Bronchoscopy was performed, and on

cartridge based nucleic acid amplification test (CBNAAT) of bronchial aspirate, *Mycobacterium tuberculosis* was not detected further ruling out the diagnosis of TB.

**ChestX-ray** (Figure - 1) revealed bilateral heterogeneous opacities involving middle and lower zone of right lung and paracardiac of left lung with cardiomegaly.



**HRCT**(Figure -1)



High-resolution computed tomography of the patient with showing basilar predominate reticulation and ground-glass opacities with an absence of significant honeycombing in a pattern consistent with nonspecific

interstitial pneumonia. The patient also has an air–fluid level in the oesophagus consistent with scleroderma-associated oesophageal dysfunction.

#### HRCT(Figure 2)



High-resolution computed tomography of the patient showing peripheral and basilar predominate reticulation and honeycombing with an absence of significant ground-glass opacities in a pattern consistent with usual interstitial pneumonia. The patient also has an air-filled oesophagus consistent with scleroderma-associated oesophageal dysfunction.

#### DISCUSSION

In scleroderma, the two most common types of direct pulmonary involvement are ILD and PH, which together account for 60% of SSc-related deaths.<sup>[2]</sup> While certain pulmonary manifestations may occur more commonly in a subset of SSc (i.e. ILD is more common in dcSSc while PH is more common in lcSSc)<sup>[3]</sup>, all of the known pulmonary manifestations reported have been described in each of the subsets of disease.<sup>[4]</sup> Pulmonary disease can even occur in SSc with no skin involvement (an entity known as scleroderma sine scleroderma).<sup>[5]</sup> These patients can be misclassified as having idiopathic ILD and the presence of telangiectasias, Raynaud's phenomena, reflux or pericardial effusions; a nucleolar-antinuclear antibody test should alert the clinician to the possibility of scleroderma sine scleroderma.<sup>[6, 7]</sup> Patients with autoimmune diseases are known to develop infections like tuberculosis either due to the disease activity or secondary to the immunosuppressive therapy. Tuberculosis per se is known to induce the development

of autoantibodies which in turn stimulate the manifestation of autoimmune diseases.<sup>[8]</sup> Sreeram V Ramagopalan, et al. analyzed a database of statistical records of patients in England (1999 to 2011), and found a significant association of tuberculosis and autoimmune diseases. High levels of risk for tuberculosis was found in diseases like Addison's disease, SLE, polymyositis and patients with scleroderma had a relative risk of 6.1 (95% CI 4.4 to 8.2).<sup>[9]</sup> Shachor et al, suggests that a diffusely damaged lung increases the susceptibility to tuberculosis or activation of dormant tuberculosis.<sup>[10]</sup> Bhatia, et al.<sup>[11]</sup> had reported a case of tuberculosis presenting as bilateral pneumothoraces who was later found to have features of scleroderma. Another study done by Subramanian, et al. highlights the association of tuberculosis and autoimmune diseases.<sup>[12]</sup> In the present case, though initially due to similar complaints of cough with expectoration, fever, loss of weight and loss of appetite a false diagnosis was made and patient was started on TB treatment from peripheral hospital, but in view of progressive symptoms, negative bacteriological studies for TB, poor response to treatment, and predominant positive immunological tests, we were prompted to investigate for other causes such as interstitial lung disease (primary and secondary)

**CONCLUSION**

Tuberculosis is endemic in India and physicians play a vital role in its diagnosis and control. This case is reported to highlight the importance to differentiate between tuberculosis and immune mediated diseases mimicking as pulmonary tuberculosis. Detailed physical examination, high clinical suspicion and also to create the awareness regarding recognition of connective disorders can help to diagnose rare diseases with misleading clinicoradiological presentation of pulmonary tuberculosis as seen in the present case of scleroderma.

**REFERENCES**

1. Gabrielli A, Avvedimento EV, Krieg T. Scleroderma. *N Engl J Med.*, 2009; 360: 1989–2003. CrossRefPubMedGoogle Scholar
2. Steen VD, Medsger TA. Changes in causes of death in systemic sclerosis, 1972–2002. *Ann Rheum Dis.*, 2007; 66: 940–944. Abstract/FREE Full TextGoogle Scholar
3. Morelli S, Barbieri C, Sgreccia A, et al. Relationship between cutaneous and pulmonary involvement in systemic sclerosis. *J Rheumatol.*, 1997; 24: 81–85. PubMedGoogle Scholar
4. Highland KB, Garin MC, Brown KK. The spectrum of scleroderma lung disease. *Semin Respir Crit Care Med* 2007; 28: 418–429. CrossRefPubMedGoogle Scholar
5. Toya SP, Tzelepis GE. The many faces of scleroderma sine scleroderma: a literature review focusing on cardiopulmonary complications. *Rheumatol Int.*, 2009; 29: 861–868. CrossRefPubMedGoogle Scholar
6. Fischer A, Meehan RT, Feghali-Bostwick CA, et al. Unique characteristics of systemic sclerosis sine scleroderma-associated interstitial lung disease. *Chest*, 2006; 130: 976–981. CrossRefPubMedGoogle Scholar
7. Fischer A, Pfalzgraf FJ, Feghali-Bostwick CA, et al. Anti-th/to-positivity in a cohort of patients with idiopathic pulmonary fibrosis. *J Rheumatol.*, 2006; 33: 1600–1605. Abstract/FREE Full TextGoogle Scholar
8. Pradhan V, Patwardhan M, Athavale A, Taushid S, Ghosh K. Mycobacterium tuberculosis triggers autoimmunity. *Indian Journal of Tuberculosis*, 2012; 59(1): 49-51.
9. Ramagopalan SV, Goldacre R, Skingsley A, Conlon C, Goldacre MJ. Associations between selected immune mediated diseases and tuberculosis: Record linkage studies. *BMC Medicine*, 2013; 97(11): 1741-7015.
10. Y Shachor, D Schindler, ASiegal, DLieberman, Y Mikulski, I Bruderman. Increased incidence of pulmonary tuberculosis in Chronic interstitial lung disease. *Thorax*, 1989; 44: 151-1530.
11. Bhatia JL, Kallan BM, Kanwar MS. Recurrent alternate bilateral spontaneous pneumothoraces in scleroderma. *Indian Journal of Tuberculosis*, 1980; 27(1): 17-19.
12. Subramanian S, et al. Co-existence of scleroderma and Tuberculosis. *Sch J Med Case Rep.*, 2015; 3(1): 22-24.