

Co-Existence of Scleroderma and Tuberculosis

Subramanian S*, Ragulan. R, Natraj. M, Meenakshi. N, Viswambhar. V

Department of Respiratory Medicine, Chettinad Hospital and Research institute, Padur, Kancheepuram District, Chennai-603103, Tamilnadu, India

*Corresponding Author:

Name: Dr. Subramanian

Email: drssmani@gmail.com

Abstract: Scleroderma is a chronic systemic autoimmune disease characterized by fibrosis, vascular alterations, and autoantibodies. This causes widespread symmetrical thickening of the skin followed by atrophy and pigmentation. Patients with immune-mediated diseases are known to have an increased risk of Tuberculosis. Mycobacterial infections are also known to induce the development of autoantibodies. We report a case presented with clinical features of pulmonary tuberculosis with general examination and radiological findings raising the suspicion of a co-existent connective tissue disorder. Antibody profile done and was found to be positive for ANA and anti-Scl-70 thus confirming the diagnosis of scleroderma coexisting with Pulmonary Tuberculosis. Hence here by we highlight the importance of suspicion, careful general examination, radiological assessment and screening tuberculosis patients for autoantibody profile in presence of a multisystem involvement.

Keywords: Scleroderma, Pulmonary Tuberculosis, Autoantibodies.

INTRODUCTION

About one-third of population in the world is infected with *Mycobacterium tuberculosis*. Infections are known to induce development of autoantibodies [1]. Studies suggest the possible autoimmune phenomenon linked with Tuberculosis. Infections triggering autoimmunity is linked to two mechanisms: 1) Up regulation of co-stimulators on antigen presenting cells which present the self-antigen to the T cells and 2) Molecular mimicry. i.e. microorganisms express antigens having same amino acid sequence as self-antigen resulting in activation of self-reactive T cells [1]. Case reports of simultaneous diagnosis of scleroderma and pulmonary tuberculosis is rare and is being reported here.

CASE REPORT

A 75 year old female, non-smoker presented with complaints of dry cough, loss of appetite and loss of weight for 1 month duration. There was no history of dyspnea, wheeze, chest pain and haemoptysis. There was no occupational exposure to chemicals, dust and smoke. On examination, patient was thin built with pallor. Patient had thickening of skin in lower palpable fissure, salt and pepper appearance over scalp, diffuse joint tenderness-bilateral metacarpophalangeal joints and knee joints. No icterus, clubbing, lymphadenopathy or pedal edema. Pulse was 84/mt. and B.P. 128/82mm of Hg. Respiratory examination revealed bilateral fine mid to late inspiratory crackles in infrascapular and infra axillary region. Other systems were normal. Total

leucocyte count was 7,900/cu mm with polymorphs 71 %, Lymphocytes 15%, monocytes 12%, Eosinophils 2 %, Hemoglobin-8.8 gm% and E.S.R.-68 mm in 1 hour. Renal and liver functions were within normal limits. HIV serology was non-reactive. Sputum smear examination was negative for AFB. Mantoux was positive (18 mm). Chest skiagram e.g. (Fig. 1) revealed heterogeneous opacity with air bronchogram in right upper lobe, pleural based opacity in right middle lobe, Reticularity in right lower lobe. CT Thorax revealed apical pleural thickening, cavity lesion with surrounding consolidatory changes in right upper lobe posterior segment (Fig. 2), interlobular and intralobular septal thickening noted in right posterior basal segments (Fig. 3). Coincidental finding of patulous esophagus was also noted. No evidence of pleural effusion or mediastinal lymphadenopathy. Bronchoscopy revealed narrowed right main bronchus and secretions in right upper lobe. Bronchial washings revealed numerous AFB in acid fast staining (Fig. 4). Based on the above general examination findings, radiological findings of interlobular and intralobular septal thickening and patulous esophagus, coexistent connective tissue disorder was suspected and autoimmune antibody profile done and was found to be positive for ANA and anti-Scl-70 thus confirming the diagnosis of scleroderma.



Fig. 1: Chest X-ray revealing heterogeneous opacity with air bronchogram sign in right upper lobe, and reticularity in right lower lobe

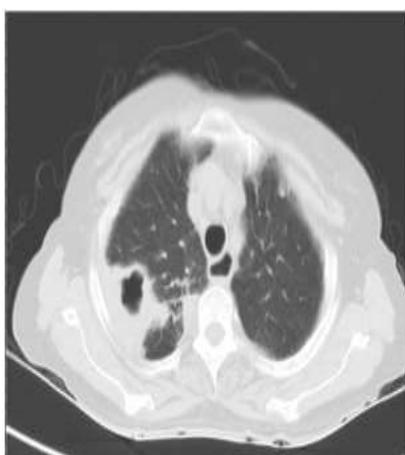


Fig. 2: Axial section of CT Thorax revealing cavity lesion with surrounding consolidatory changes in right upper lobe posterior segment and patulous oesophagus



Fig. 3: Axial section of CT Thorax revealing Inter and intralobular septal thickening in right posterior basal segment

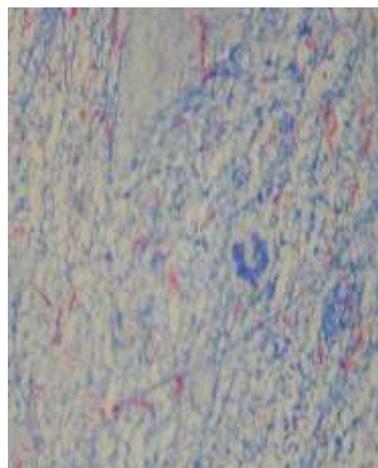


Fig. 4: Acid fast Staining of Bronchial washings revealing Acid Fast Bacilli

DISCUSSION

This patient presented primarily with clinical features suggestive of pulmonary Tuberculosis. Unusual general examination findings like thickening of skin in lower palpable fissure, salt and pepper appearance over scalp, diffuse joint tenderness with radiological finding of interlobular and intralobular septal thickening and patulous oesophagus helped us to suspect scleroderma.

In a similar previous case report by Agarwal *et al.* [2] in 1977 pulmonary tuberculosis was diagnosed in a known case of scleroderma who has taken systemic steroids for six months duration. In that case there was a possibility of Tuberculosis because of immunosuppression secondary to steroid therapy. J L Bhatia *et al.* [3] has reported a case of Tuberculosis whose past history revealed gradually progressive numbness, pain and cyanosis of fingers and toes every winter for 12 years with subsequent dyspnea, dysphagia and facial hyper pigmentation. The rare finding reported by him was occurrence of recurrent bilateral pneumothoraces in that patient. Sreeram V Ramagopalan *et al.* [4] analyzed a database of statistical records of England (1999 to 2011), and a similar database (the Oxford Record Linkage Study - ORLS) for southern England and concluded that patients with some immune-mediated diseases have an increased risk of TB and further studies of these associations may aid TB screening and its control and treatment policies. Shuo-Ming Ou *et al.* [5] conducted a nationwide study using the Taiwan Health Insurance Research Database during 2000–2006, where 838 patients of systemic sclerosis were identified and followed for emergence of TB infection and concluded that the incidence of TB was significantly higher among systemic sclerosis patients than in controls. In contrast to the above mentioned studies the point we would like to highlight in this case is the simultaneous diagnosis of Tuberculosis and Scleroderma. The diagnosis of scleroderma was feasible despite absent presenting

complaints, due to high degree of suspicion based on general physical examination and radiological findings.

CONCLUSION

Tuberculosis is endemic in India and physicians play a vital role in its control. This case is reported to highlight the association between Tuberculosis and immune-mediated diseases and also to create the awareness regarding recognition of pulmonary manifestations of co-existent connective tissue disorders and tuberculosis. Awareness of association between the above combination and high index of suspicion based on clinical and radiological findings will help in early diagnosis and management of immune mediated diseases there by reducing the morbidity of the illness.

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