Case Report

Systemic sclerosis and tuberculosis – A case report

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Abstract

Scleroderma is a chronic systemic autoimmune disease of unknown origin characterized by excessive collagen deposition and autoantibodies. Patients with autoimmune diseases are known to develop infections like tuberculosis due to the disease per se and also secondary to immunosuppressive treatment. Mycobacterial infections are also known to induce the development of autoantibodies. We present a case of simultaneous occurrence of interstitial lung disease and pulmonary tuberculosis (BAL positive for AFB) in a patient with features of systemic sclerosis. Antibody profile was done were 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, bronchoscopy and immunological test helped in the diagnosis of the patient. We highlight the importance of suspicion, careful general examination, radiological assessment and screening tuberculosis patients for autoantibody profile in presence of a multisystem involvement.

Key words

Systemic sclerosis, Tuberculosis, ANA.

Introduction

Pulmonary involvement is common in patients with systemic sclerosis (SSc), and this leads to substantial morbidity and mortality. Disrupted immunity from the disease or associated medication may render such patients subject to tuberculosis infection. Infections are also known to induce the development of autoantibodies. A rare case of simultaneous diagnosis of scleroderma and pulmonary tuberculosis is being reported here.

Case report

A 45 year old female presented with complaints of breathlessness on exertion (MMRC grade III), cough with expectoration, fever, loss of appetite and weight, difficulty in swallowing and tightening of skin around the mouth and left knee joint pain for 2 months. There was no history of chest pain, haemoptysis, and wheeze. There was no occupational exposure to chemicals dust and smoke.

On examination, patient was thin built. Patient had features of salt and pepper appearance, fish mouth deformity, mask like face, pinched nose, facial melanosis, telangiectasia over the cheeks, sclerodactyly and resorption of digits diagnostic of scleroderma. There was no pallor, cyanosis, clubbing, lymphadenopathy or pedal oedema. Pulse was 110/min and BP: 100/60mmhg. Respiratory examination revealed bilateral end inspiratory crackles of velcro type. Other systems were normal.

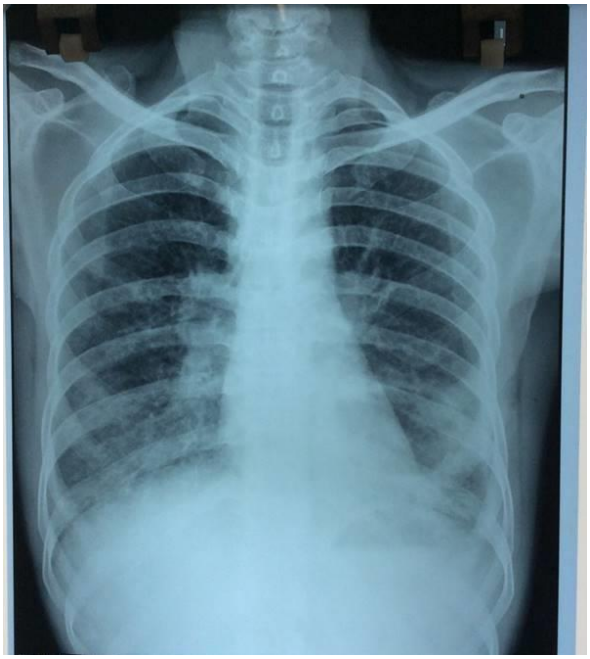
Provisional diagnosis of systemic sclerosis with interstitial lung disease was made. Due to financial constraints and she was fitting into clinical ACR criteria [1] for diagnosis ,ANA alone was sent as a part of autoimmune antibody profile which was positive, contributing towards the diagnosis.

Further serological investigations revealed anaemia with Hb of 9.8gm%, elevated leucocyte count with72% polymorphs and elevated ESR. Renal and liver functions were within normal limits.HIV serology was Non-reactive.

Chest X-ray (**Figure - 1**) revealed thick walled cavity in the left lower lobe and increased reticular opacities bilaterally. In view of thick walled cavity, secondary infections due to bacterial, fungal and tuberculous organisms were considered. Sputum was negative for AFB and gram stain, cultures revealed no growth. Mantoux was negative.

HRCT (**Figure - 2**) chest showed a large thick walled cavity with adjacent areas of consolidation in superior segment of left lower lobe, sub pleural reticulations with honey combing (**Figure - 2.1**) with areas of traction bronchiectasis in both lower lobes and linear branching opacities bilaterally with mediastinal lymphadenopathy. She had features of both progressive ILD and active pulmonary tuberculosis. Bronchoscopy was done which revealed stenosis of left lower lobe apical segment, opening of apical segment bronchus was covered with whitish plaque. BAL revealed AFB (**Figure - 3**) in acid fast staining. Patient was initiated on anti tuberculous drugs and treatment for ILD after 1 month of antituberculous treatment.

Figure – 1: Chest X-RAY showing thick walled cavity in the Left lower lobe and increased reticular opacities bilaterally.



Discussion

Systemic sclerosis is a multisystem disease involving skin, lungs, kidneys, heart, GI tract and skeletal muscles. Incidence is three times more in women than in men with the peak incidence between ages 20-60 [2]. Pulmonary involvement in the form of interstitial lung disease is common in patients with systemic sclerosis (SSc).

Figure – 2: Large thick walled cavity associated with consolidation in superior segment of left lower lobe.

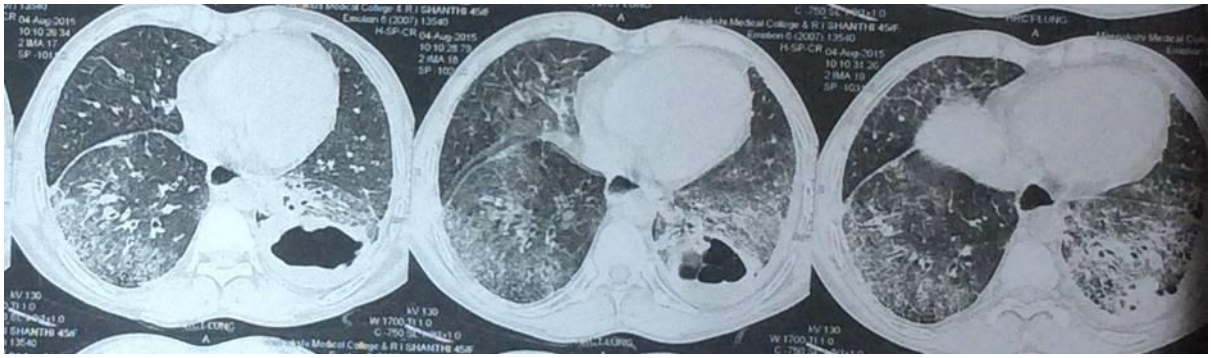


Figure - 2.1: Sub pleural reticulations with Honey combing.

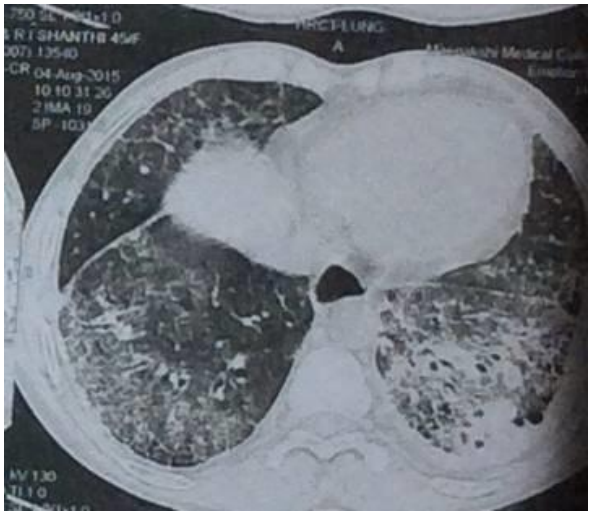
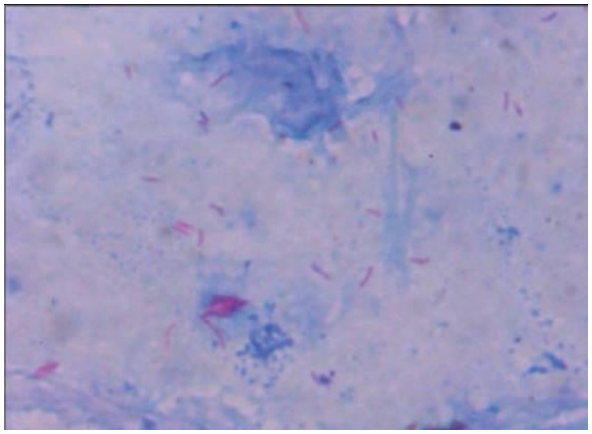


Figure – 3: BAL showing AFB.



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 [3].

Sreeram V Ramagopalan, et al. [4] 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).

He also looked at the incidence of immune diseases after 5 years of first occurrence of tuberculosis and found significant increase in Addison's disease, Sjogrens and SLE. The possible mechanism for this association was tuberculosis being infective trigger acting by molecular mimicry, bystander activation or acting as an adjuvant.

Pradhan et al did a screening of autoantibodies in tuberculosis endemic areas and concluded a possible role of mycobacterial infection triggering autoimmunity. Importance of screening all tuberculosis patients for autoantibody profile and follow up for autoimmune related symptoms was highlighted [2].

Shachor et al, suggests that a diffusely damaged lung increases the susceptibility to tuberculosis or activation of dormant tuberculosis [5].

In a study done in Taiwan, 838 patients with systemic sclerosis were followed up for a period of four years for the occurrence of tuberculosis infection and they found that the incidence of tuberculosis infection was 4.5 times higher than the controls [6]. Bhatia, et al. [7] 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 [8].

Our patient was diagnosed to have interstitial lung disease and pulmonary tuberculosis concurrently and also had characteristic features of systemic sclerosis.

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 of association between tuberculosis and immune mediated diseases, detailed physical examination and high clinical suspicion and also to create the awareness regarding recognition of co-existence of connective disorders and tuberculosis. Further studies are needed to look into the autoimmune phenomenon and triggering mechanisms linked with Mycobacterium tuberculosis infections.

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