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Case Report

Atypical Ground Glass Opacification in Systemic Sclerosis Related Interstitial Lung Disease: A Case Report

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ABSTRACT

Ground glass opacification is generally indicative of active alveolitis and this finding on high resolution C.T scan of chest can identify scleroderma patients with early interstitial lung disease that may be amenable to treatment with aggressive immunosuppression in an attempt to prevent or slow progression to pulmonary fibrosis and hence to end stage lung disease. Further it can be used to assess the response to treatment and may also predict prognosis in such patients. We report a 30 year old female who had clinical, immunological and histological features of systemic sclerosis. Evaluation for progressive dyspnoea and non-productive cough revealed features of early interstitial lung disease i.e. restrictive physiology on spirometry and bibasal subpleural ground glass opacity in high resolution C.T. scan of chest. Echocardiography did not reveal elevated pulmonary arterial pressures and esophagus was normal on endoscopic examination.

Key Words: Ground glass opacification, alveolitis, systemic sclerosis.

INTRODUCTION

Scleroderma (SSc) is an uncommon systemic collagen vascular disease. [1] During the course of systemic sclerosis, pulmonary involvement can be observed in most patients. ILD can be demonstrated in virtually all patients, and it is observed in both limited and diffuse systemic sclerosis. [2] Moreover, pulmonary disease is the leading cause of death in patients with systemic sclerosis. [3] Interstitial lung disease as a first manifestation of disease in systemic sclerosis, however, is extremely rare, and was previously reported to be associated with the presence of autoantibodies against topoisomerase I (anti-Scl-70 antibodies). [4,5] In most cases of systemic sclerosis, however, lung involvement is observed relatively late in the course of the disease. [5,6] Early recognition of ILD is mandatory since symptoms will occur relatively late in the course of the disease. At that stage the patients may suffer from already severely impaired lung function due to irreversible interstitial fibrosis.

CASE REPORT

A 30 year old female was investigated for complaints of gradually progressive exertional breathlessness and non-productive cough of six months duration. She also had on and off joint pains and had begun experiencing pain and bluish discolouration of finger tips on exposure to cold water. It was associated with progressive tightening of skin over hands, face and feet. General physical examination revealed diffuse alopecia, tight shining skin over hands, forearms, feet, face and chest. She also had hyperpigmentation of face and digital pitted scars on index and middle fingers.

Cardiopulmonary examination revealed bibasal end-inspiratory crepitations, however there were no signs of pulmonary arterial hypertension (PAH).

Complete blood count and biochemical investigations were normal. She had positive tests for anti-nuclear antibody and anti-topoisomerase1 antibody. Rheumatoid factor, anti-centromere antibody as well as anti U3RNP antibody tests were negative. Her 24 hour urinary protein was normal as was esophago-gastroscopy. Skin biopsy was consistent with early progressive systemic sclerosis.

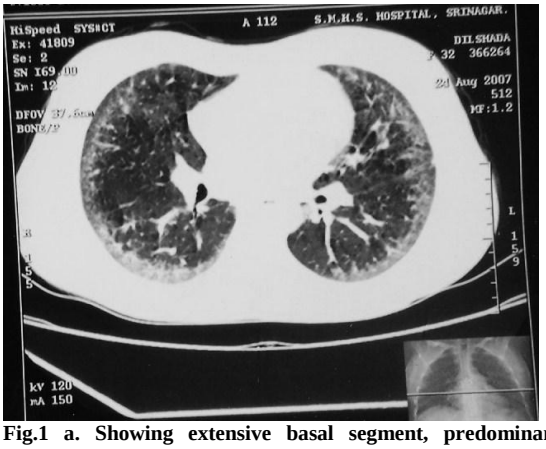

Fig.1 a. Showing extensive basal segment, predominantly subpleural ground glass opacification (GGO) in both lung fields.

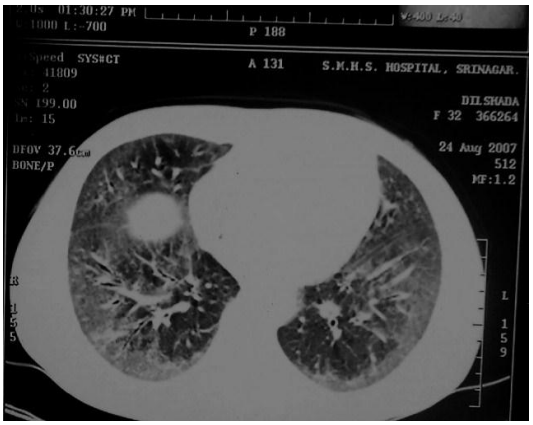

Fig. 1b

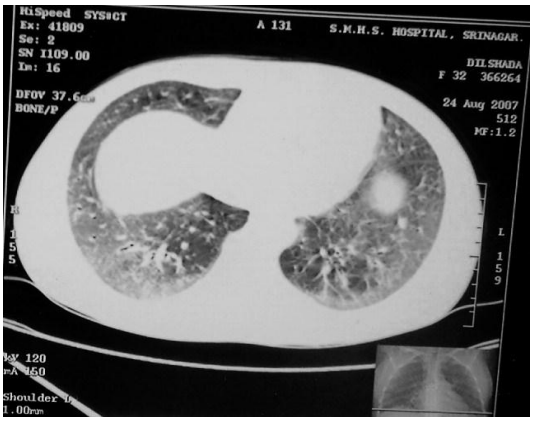

Fig. 1c

Spirometry revealed restrictive physiology with FVC equal to 66% of predicted for her age and sex. Chest X-ray showed bilateral ground glass haziness and high resolution C.T. scan chest revealed extensive basal segment, predominantly subpleural ground glass opacification (GGO) in both lung fields i.e. Figs: 1a, 1b and 1c. There was no honeycombing, intra- or interlobular septal thickening. Pulmonary arterial pressure (RVSP) was within normal limits on transthoracic echocardiography.

Patient was started on immunosuppressive agents that included Azathioprine (2.5 mg/kg) and prednisolone (1 mg/ kg) and was doing well on last out-patient follow-up.

DISCUSSION

Systemic sclerosis (SSc) is a clinically heterogeneous generalized disorder which affects the connective tissue of the skin and internal organs such as gastrointestinal tract, lungs, heart and kidneys. It is characterized by alterations of the microvasculature, disturbances of the immune system and by massive deposition of collagen. [7] Diffuse and limited forms of the disease exist, depending on the extent of cutaneous involvement, with a different clinical course and prognosis for each.

Pulmonary disease is found in over 80 percent of patients with systemic sclerosis (SSc). Pulmonary involvement is second in frequency only to esophageal involvement as a visceral complication of SSc and has surpassed renal involvement as the most common cause of death. [8]

Pulmonary fibrosis is the most common radiographic finding, being present in 20% to 65% of patients. The fibrosis usually has a basilar predominance, initially as a fine reticular pattern that progresses to coarse reticulation and honeycombing.[9] Non-Specific Interstitial Pneumonitis (NSIP) being much more common than Usual Interstitial Pneumonia UIP.[10,11] However, pulmonary hypertension is also common, either as an isolated finding or in association with lung fibrosis. Pulmonary hypertension is particularly common in patients with limited scleroderma (CREST syndrome). [12]

HR-CT is a widely accepted diagnostic tool to detect interstitial lung disease, and has been proven to be highly superior to the chest X-ray. [13] Because computed tomography has improved the accuracy of non-invasive testing of idiopathic pulmonary fibrosis (or UIP), presently the main indication of lung biopsy is to assess the relative degrees of cellularity and fibrosis. If HRCT could predict the pathological appearances, the need for lung biopsy would be further reduced.

Two distinct patterns of disease are seen in tomograms: ground glass increase in attenuation of lung parenchyma and a reticular pattern. Ground glass opacification (GGO) is defined as increase in attenuation of lung parenchyma without obscuration of vascular markings on CT images. The GGO is associated with a cellular histological appearance of that area of lung where as a predominantly reticular pattern is found in patients whose subsequent lung biopsy confirms fibrosis. There is evidence that a predominant GGO pattern is more likely to represent active inflammatory disease and to respond to appropriate immunosuppressive therapy. It is still unproven whether GGO pattern proceeds reticular or honeycomb pattern, although this seems likely. Correlation between pathology and HRCT shows that most, but not all, patients have NSIP; [12] the degree of cellular versus fibrotic NSIP determines the subsequent clinical course.[14]

Our patient had history of Raynaud's phenomenon and typical signs of systemic sclerosis who underwent evaluation for exertional breathlessness and cough which confirmed interstitial lung disease and she had good early response to immunosuppressive therapy. Thus, HRCT done early in the course of systemic sclerosis can pick up patients with active alveolitis who might benefit from immunosuppressive therapy. It can also be used for follow up and prognostication.

Because of its ability to differentiate between cellular and fibrotic disease with reasonable accuracy, HRCT scanning can be used to predict response to treatment and is significantly more accurate than chest radiography in this respect. In treated patients with idiopathic pulmonary fibrosis (UIP) improvement in lung function in those with a predominantly GGO pattern is

significantly better than in those with a reticular pattern or a mixture of GGO and reticular patterns. This improved response rate is matched by improved survival, the predictive value of the HRCT scan being independent of lung function or duration of breathlessness. The extent of fibrosis on HRCT scanning shows 80% sensitivity in predicting survival.

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