Interstitial Lung Disease with Non-Specific Interstitial Pneumonia pattern in Systemic Sclerosis: A case report

[a]G. N. Srivastava, [b]Mrityunjay Sharma

[a]Professor, Department of TB and Respiratory Diseases, Institute of Medical Sciences, Sir Sunderlal Hospital, Banaras Hindu University, Varanasi, India

[b]JuniorResident, Department of TB and Respiratory Diseases, Institute of Medical Sciences, Sir Sunderlal Hospital, Banaras Hindu University, Varanasi India

Abstract

Systemic sclerosis is an autoimmune disorder with unknown etiology. Depending upon the symptoms and signs it has limited or diffuse variants. Diffuse systemic sclerosis extensively involves the skin of the extremities and face along with the visceral organ system while the limited form, also known as CREST variant is characterized by calcinosis, Raynaud phenomenon, esophageal dysmotility, sclerodactyly, and telangiectasia. Here we report a case of Interstitial Lung Disease with Non-Specific Interstitial Pneumonia pattern in Systemic Sclerosis presented in our hospital with complaints of multiple pitted scars, bluish discoloration of extremities on cold exposure, loss of wrinkles over face and breathlessness on exertion. On examination there was tight and shiny skin with loss of hair, pitted scars and breathlessness on exertion evaluated by six minute walk test. Bilateral crepitation were present on auscultation. On investigation anti RO-52 was found positive. PFT was suggestive of restrictive disorder and radiological features were suggestive of interstitial lung disease with non-specific interstitial pneumonia (NSIP) pattern.

KEYWORDS: Systemic Sclerosis, Interstitial Lung Disease

Introduction:

Systemic Sclerosis or scleroderma, an autoimmune, inflammatory-fibrotic disease with unknown etiology is characterized by multi organ system involvement. It involves skin and several visceral organs including the lungs, GIT, heart and kidneys[1].

Systemic sclerosis has two subtypes: diffuse and limited[2].Diffuse systemic sclerosis, extensively involves the skin of the extremities and face along with the visceral organ system.The limited form is also known as CREST variant (calcinosis, Raynaud phenomenon, esophageal dysmotility, sclerodactyly, and telangiectasia). In limited form visceral organ involvement tends to show insidious progression.

Involvement of lungs is common in systemic sclerosis. Postmortem series indicates a 70% to 100% incidence[6]. It usually results into interstitial lung disease (ILD) and pulmonary arterial hypertension (PAH). Around 90% of patients with ILD in systemic sclerosis have interstitial changes on high resolution computed tomography scan[8], and between 40-75% of patients show restrictive pattern on pulmonary function testing[9,

[3].Several reports suggest an increased incidence of lung neoplasms in patients with scleroderma[4,5.]

The symptoms in interstitial lung disease predominantly include dyspnea on exertion and cough. Bibasilar crackles are heard on auscultation. Radiographic features are typically bibasilar interstitial infiltrates, reticular pattern, traction bronchiectasis, honeycombing and loss of lung volume. Patients with diffuse systemic sclerosis or Scl-70 (anti-topoisomerase) antibodies are at higher risk for the development of ILD than the patients with limited systemic sclerosis or anti-centromere antibodies.

2013 ACR/EULAR CLASSIFICATION CRITERIA FOR SCLERODERMA[7]
These criteria were developed by the American College of Rheumatology (ACR) and the European League Against Rheumatism (EULAR).

Table No 1: The ACR-EULAR Criteria for the classification of Systemic Sclerosis

Items	Sub items	Score
Skin thickening of the fingers of both hands extending proximal to the metacarpophalangeal joints (sufficient criterion) Skin thickening of the fingers (only count the highest score)	 Puffy fingers Sclerodactyly of the fingers (distal to MCP but proximal to the PIPs)	9 2 4
Fingertip lesions (only count the highest score)	Digital Tip Ulcers Finger Tip Pitting Scars	2 3
Telangiectasia		2
Abnormal nail fold capillaries		2
Pulmonary arterial hypertension and/or Interstitial lung Disease (Maximum score is 2)	PAH ILD	2
Raynaud's phenomenon		3
Scleroderma related antibodies (any of anti-centromere, anti-topoisomerasel, anti-RNA polymerase III) (Maximum score is 3)	Anti-centromere Anti-topoisomerasel Anti-RNA polymerase III	3

Patients with a total score of ≥ 9 are classified as having definite scleroderma.
Skin thickening of the fingers extending proximal to the metacarpophalangeal joints is sufficient for the diagnosis.

Case report

A 30 years old lady presented with the complaints of multiple pitted scars below elbows and bluish discoloration of fingers and toes on exposure to cold for 4 years. Gradually she developed loss of wrinkling over face and difficulty in movements of fingers due to thickening and tightness of skin. She developed dry cough and shortness of breath on exertion also for last one year which was insidious in onset, persistent and gradually progressive.

On examination-

Patient had masked facies with decreased opening of mouth (figure 1). Skin of hands forearms, legs and feet appeared shiny and tight with loss of hair (figure 2a, b& 4). Pitted scars were present over finger tips and forearms (figure 2a& b). She had exertional dyspnea of mMRC grade 3. Bilateral crepitations were present on auscultation.

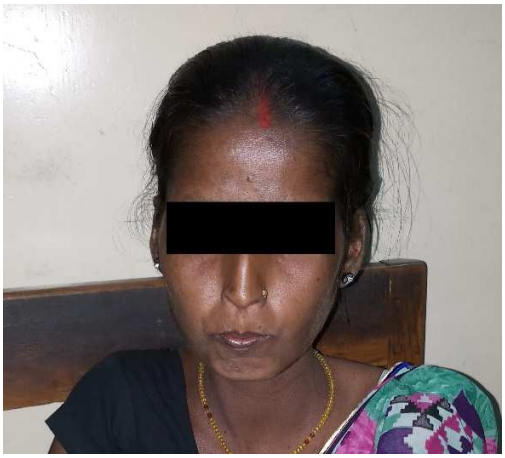

Figure 1: Face of the patient

Figure 2a & b:Upper extremities

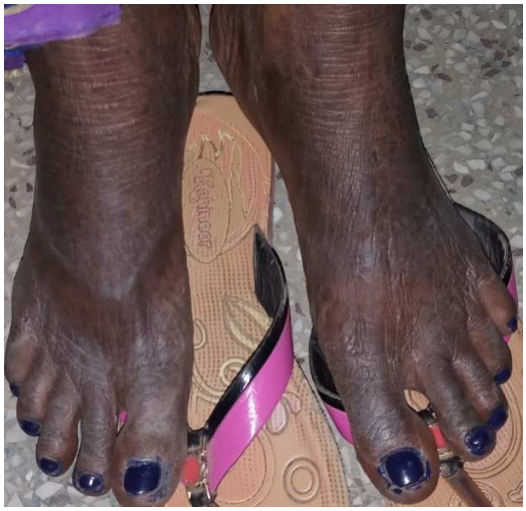

Figure 4: Lower extremities

Laboratory investigations:

Routine	
Hemoglobin	9.3 gm/dl
Total Leucocyte Counts	11800/µl
Differential Leucocyte Counts	N-69.2, L-21.4, M-4.9, E-4.2
Platelets	473000/µl
Creatinine	0.4 mg/dl
UREA	37 mg/dl
Na	141 mmol/L
K	4.2 mmol/L
Cl	108 mmol/L
SGPT/SGOT	36/40
Total Bilirubin/Direct Bilirubin	0.8/0.3
Alkaline Phosphatase	48 U/L
Anti RO-52	Positive
ANA	Negative
U1RNP,	Negative
SS-A	Negative
SS-B	Negative
SCL-70	Negative
PM	Negative
SCL	Negative
JO-1	Negative
CENP-B	Negative
PCNA	Negative
dsDNA	Negative

RNP	Negative
SM	Negative

Six minute walk test- 9% fall in spo2 was noted.

PFT was suggestive of restrictive disorder.

Chest x-ray showed diffuse heterogeneous opacities predominantly in middle and lower zones of bilateral lung fields with irregular diaphragmatic pleural borders.

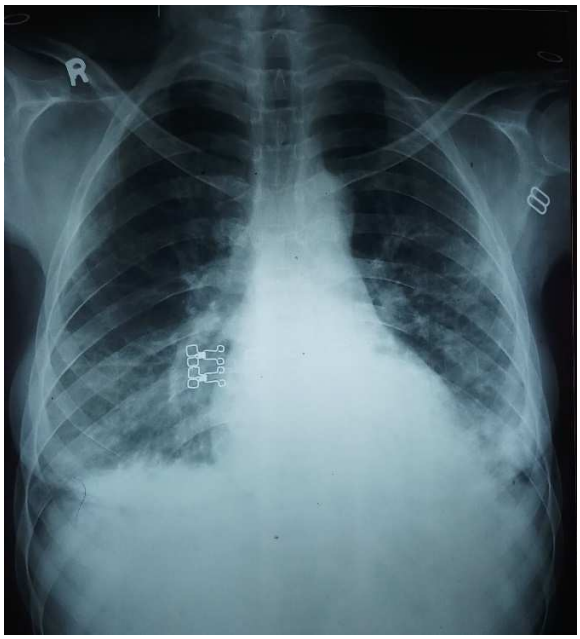

Figure 5: Chest X-Ray

High resolution computed tomography (HRCT) thorax showed fibro bronchiectatic changes along with areas of intralobular septal thickening scattered in bilateral lung fields, predominantly in lower lobes with sub-pleural sparing which favored interstitial lung disease with non-specific interstitial pneumonia(NSIP) pattern.

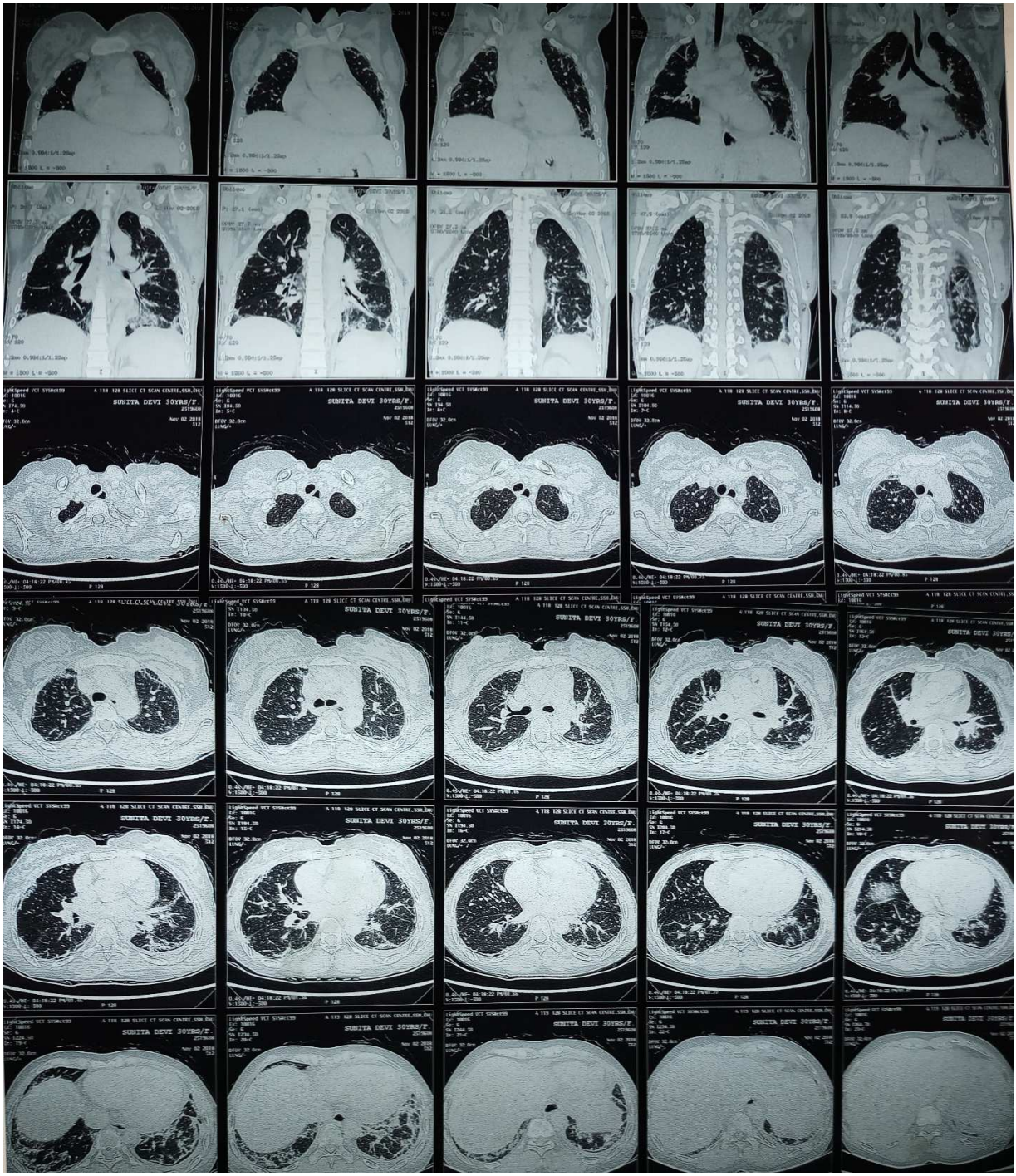

Figure 6: HRCT Thorax

On the basis of clinical, hematological and radiological grounds a diagnosis of Systemic sclerosis with interstitial lung disease of Non-specific interstitial pneumonia (NSIP) pattern was made.

Discussion

Systemic sclerosis, an autoimmune fibrotic disease has diffuse and localized variants. Diffuse variety involves multiple visceral organs like lungs, heart, kidneys and GIT. Lung involvement in systemic sclerosis can result in pulmonary fibrosis and secondary pulmonary hypertension. Patients present with exertional dyspnea, fatigue, and cough. Non-specific interstitial pneumonia pattern in interstitial lung diseases are characterized by honeycombing, tractional bronchiectasis, interseptalthickening, with lower lobes involvement predominantly. Immunosuppression is the mainstay of treatment in systemic sclerosis, with corticosteroids and cyclophosphamide being the agents of choice[10]. Mycophenolate mofetil and rituximab have also been reported to benefits patients with progressive disease in scleroderma[11].

References:

1. Gandage G., Aironi VD, Gauri Sawadkar. Radiological manifestations of scleroderma: a case report. Pravara Med Rev. 2011;3(2):40-2

2. Cleveland Clinic - Soumya Chatterjee. Systemic scleroderma, 2014. Available at:http://www.clevelandclinicmeded.com/medicalpubs/diseasemanagement/rheumatology/systemic-sclerosis/Default.htm.

3. Steen VD, Conte C, Owens GR, et al. Severe restrictive lung disease in systemic sclerosis. Arthritis Rheum. 1994; 37(9):1283–9. [PubMed: 7945490]

4. Colaci M, Giuggioli D, Sebastiani M, et al. Lung cancer in scleroderma: results from an Italian rheumatologic center and review of the literature. Autoimmun Rev. 2013;12:374–379.

5. Omair MA, Lee P. Development of adenocarcinoma of the lung in a transplanted lung of a scleroderma patient. ClinExpRheumatol. 2012;30:317–318.

6. Schurawitzki H, Stiglbauer R, Graninger W, et al. Interstitial lung disease in progressive systemic sclerosis: high-resolution CT versus radiography. Radiology. 1990;176:755–759.

7. van den Hoogen, Frank et al. “2013 classification criteria for systemic sclerosis: an American College of Rheumatology/European League against Rheumatism collaborative initiative” Arthritis and rheumatism vol. 65,11 (2013): 2737-47.

8. Schurawitzki H, Stiglbauer R, Graninger W, et al. Interstitial lung disease in progressive systemic sclerosis: high-resolution CT versus radiography. Radiology. 1990; 176(3):755–9. [PubMed: 2389033]

9. Steen VD, Owens GR, Fino GJ, et al. Pulmonary involvement in systemic sclerosis (scleroderma). Arthritis Rheum. 1985; 28(7):759–67. [PubMed: 4015723]

10. Strange C, Bolster MB, Roth MD, et al. Bronchoalveolar lavage and response to cyclophosphamide in scleroderma interstitial lung disease. Am J RespirCrit Care Med. 2008;177:91–98.

11. Zamora AC, Wolters PJ, Collard HR, et al. Use of mycophenolate mofetil to treat scleroderma-associated interstitial lung disease. Respir Med. 2008;102:150–155