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Original Research Article

The Clinical Importance of HRCT Chest Anomalies in Scleroderma Patients: A Hospital-based Observational Study

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Abstract:

Aim: The aim of the present study was to discuss the significance of various pulmonary and extrapulmonary abnormalities that may be identified on high-resolution computed tomography (HRCT) chest of systemic sclerosis (SSc) patients.

Methods: The present study was a cross-sectional, observational including 50 patients in the Department of Radio-Diagnosis for one year . Patients with clinical diagnosis of SSc having pulmonary involvement were included in the study. Pregnant females and patients having active chest infection or history of pulmonary tuberculosis were excluded.

Results: Age ranged from 13 to 61 years (mean 35.5±10.2 years) and most of the patients belonged to 20-30 age group followed by 31-40 age group. All the patients had clinical complaints of skin thickening and tightness, barring a few. Mean duration of skin manifestations was 5.4±5.8 years. Other than skin thickening, dyspnoea and dry cough were the frequently associated symptoms. 36% patients had duration of <6 months. 56% had 61%-80% FEV. Chest radiography was not found to be a sensitive modality in evaluating ILD, particularly in the early stages of the disease. Among them, 56% had right lung involvement and 24% had left lung involvement. Most common chest radiograph finding was fine or coarse reticular opacity. Variable degree of lung volume loss was observed on radiography.

Conclusion: Systemic sclerosis commonly occurs in middle aged females; the presenting complaint being diffuse skin thickening over the limbs and face and dyspnoea on exertion with or without dry cough as the primary respiratory symptom. PFT reveals restrictive pattern. The limitations of chest radiography may overcome by HRCT chest. Non-specific interstitial pneumonia pattern is the most common ILD found in SSc.

Keywords: Ground-glass opacity; HRCT Chest; lung fibrosis; pulmonary hypertension, systemic sclerosis

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Introduction

Systemic sclerosis (SSc) is a chronic multisystem autoimmune disorder that involves the skin and several internal organs. Depending on the extent of cutaneous involvement, the disease is classified into diff use cutaneous (dcSSc) and localized cutaneous (LcSSc) forms. Among the potential organ involvements, the lung has a very important bearing on the prognosis of these patients. Pulmonary involvement and related complications is the largest driver of morbidity and mortality

in SSc.[1] Interstitial lung disease (ILD) and pulmonary hypertension (PH) are the two most important complications which are also the leading cause of morbidity and mortality. Over 90% of SSc patients have evidence of ILD at autopsy and 40% of SSc patients demonstrate abnormal pulmonary function tests.[2] HRCT chest is now a well-established and reliable imaging tool to detect and characterize ILD in SSc.[3,4] Classically, the disease initially affects the sub pleural, posterior, and dependant aspects of the lungs. The pattern of ILD in SSc is similar to that seen in the idiopathic nonspecific interstitial pneumonitis (NSIP).[5,6]

Patients with SSc can be classified by the extent of skin involvement: in patients with limited cutaneous SSc (lcSSc), the affected skin is restricted to the hands, forearms, feet, and face, while in patients with diffuse cutaneous SSc (dcSSc), the affected skin extends proximal to the elbows, and may involve the trunk.[7] The lung is frequently involved in SSc, with interstitial lung disease (ILD) a common manifestation.[8,9] Indeed, ILD is included in the American College of Rheumatology (ACR)/ European League Against Rheumatism Collaborative Initiative (EULAR) joint classification criteria to identify SSc in individuals who do not have skin thickening of the fingers extending proximal to the metacarpophalangeal joints.[10] ILD associated with SSc (SSc-ILD) is usually detected during the evaluation of a patient suspected or known to have SSc, but may be the initial presentation of the disease in some patients.[11]

Interstitial lung disease in systemic sclerosis (SSc) patients is common in both forms of the disease (diffuse cutaneous and limited cutaneous). The disease is diagnosed at an advanced stage most of the time and this is probably due to the insidious onset of the disease accompanied by subtle clinical symptoms; at that point, the lung has become extensively fibrosed.[12] High-resolution computed tomography (HRCT) plays a major part in diagnosing interstitial lung disease in SSc patients. Its benefit lies more in assessing the extent of the disease.[13]

The aim of the present study was to discuss the significance of various pulmonary and extrapulmonary abnormalities that may be identified on high-resolution computed tomography (HRCT) chest of systemic sclerosis (SSc) patients.

Materials and Methods

The present study was a cross-sectional, observational including 50 patients in the Department of Radio-Diagnosis, Narayan Medical College and Hospital, Jamuhar, Sasaram, Bihar, India. Patients with clinical diagnosis of SSc having pulmonary involvement were included in the study. Pregnant females and patients having active chest infection or history of pulmonary tuberculosis were excluded. PFT was done for all the study patients. Other investigations done were chest radiography, as an initial imaging modality and relevant blood investigations as required.

High-resolution computed tomography chest, using Siemens SOMATOM™ definition flash 128-slice dual source CT scanner with appropriate protocols was performed in the supine position and with full inspiration, as end-inspiratory sections give fine details of lung parenchyma and allow reliable reconstruction of volumetric images. Thin axial sections were taken from the lung apex to lung base without intravenous contrast. Prone sections were obtained to distinguish gravity dependent changes, wherever indicated. Slice thickness of 5mm with 1mm reconstruction and 0.7mm increment was used. According to the semi- quantitative scoring method formulated by Ooi et al[14], all HRCT findings, like GGO, mixed ground-glass and reticular opacity, reticular fibrotic changes alone and honey-combing were assessed in each of the six lobes

(considering lingula as a separate lobe) of the lungs and expressed as percentage of each lobe affected as – 0 for 0%, 1 for 1%–25%, 2 for 26%–50%, 3 for 51%–75% and 4 for 76%–100% area involved.[14]

The extent of lobar involvement was calculated as percentage of each lobe affected for each HRCT abnormality. Total score was obtained by adding individual lobar scores for all the four HRCT abnormalities and correlated with the PFT parameters. Inflammatory index was derived using the score of GGO and mixed pattern and fibrotic index was found from the scores of reticular opacities only or honey- combing changes as described by Ooi et al.[14] All pulmonary function parameters were expressed as percentage predicted values, except FEV1/FVC ratio. Abnormal lung function was considered when predicted FVC and FEV1 value were <80% and FEV1/FVC was <75% or >85%.

Statistical Analysis

The data was entered in Microsoft Excel spreadsheet and analysis was done using Statistical Package for Social Sciences (SPSS). Categorical variables were presented in number and percentage and continuous variables were presented as mean ± standard deviation (SD) and median. Quantitative variables were compared using the student's t-test. A P value of <0.05 was considered statistically significant.

Results

Table 1: Baseline profile and pulmonary function data

Age Group (years)	**Number of Patients (%)**
10-20	5 (10)
21-30	18 (36)
31-40	16 (32)
41-50	5 (10)
51-60	4 (8)
61-70	2 (4)
IIP Pattern	
NSIP	32 (64)
UIP	12 (24)
Organising pneumonia	2 (4)
No abnormality	4 (8)
Duration of cutaneous manifestation	
<5	35 (70)
6–10	11 (22)
>10	4 (8)
Duration of respiratory symptoms	
No complaints	3 (6)
<6 months	18 (36)
6–12 months	17 (34)
>12 months	12 (24)
Forced vital capacity	
>80%	8 (16)
61%–80%	28 (56)
41%–60%	14 (28)
<40%	0

Age ranged from 13 to 61 years (mean 35.5±10.2 years) and most of the patients belonged to 20-30 age group followed by 31-40 age group. All the patients had clinical complaints of skin thickening and tightness, barring a few. Mean duration of skin manifestations was 5.4±5.8 years. Other than skin thickening, dyspnoea and dry cough were the frequently associated symptoms. 36% patients had duration of <6 months. 56% had 61%-80% FEV.

Table 2: Pulmonary function tests and radiological imaging findings

FEV_1/ FVC	**Number of Patients (%)**
70%–85%	3 (6)
86%–100%	28 (56)
>100%	19 (38)
Lung zone involved	
Right	28 (56)
Left	22 (44)
Predominant chest radiograph finding	
Reticular	22 (44)
Reticulo-nodular	0
No abnormality detected	28 (56)
HRCT chest findings	
Only GGO	5 (10)
Mixed GGO + Reticular inter-lobular septal thickening	33 (66)
Only reticular fibrosis	7 (14)
Honey-combing	11 (22)
Traction bronchiectasis	20 (40)
HRCT chest score	
0–5	19 (38)
6–10	23 (46)
11–15	8 (16)
(Mean HRCT score) FVC% predicted	
>80% (4)	7 (14)
60%–80% (5.5)	25 (50)
<60% (8.8)	14 (28)
Could not perform (11.6)	4 (8)

Chest radiography was not found to be a sensitive modality in evaluating ILD, particularly in the early stages of the disease. Among them, 56% had right lung involvement and 24% had left lung involvement. No significant upper and middle zone involvement was seen. Most common chest radiograph finding was fine or coarse reticular opacity. Variable degree of lung volume loss was observed on radiography.

Discussion

Systemic sclerosis (SSc) is a complex, chronic, multi- system autoimmune connective tissue disorder affecting around 15 people per 10 lakh population in the world. It has 3:1 female predilection and typically occurs in third to fifth decades of life.[15] The disease is characterised by vascular obliteration due to endothelial dysfunction, fibrosis of skin and internal

organs due to excessive collagen formation and immunologic abnormalities. Among internal organs, lungs, gastrointestinal tract and kidneys are frequently affected. More than 90% of the patients with SSc have evidence of interstitial lung disease (ILD) at autopsy and 40% of patients show abnormal pulmonary function tests (PFTs).[16] Pulmonary complications, such as ILD and pulmonary hypertension are the most common cardio-pulmonary findings in patients with SSc. These account for approximately 60% of SSc related deaths.[17] The disease is classified into two sub-types — diffuse scleroderma and limited cutaneous scleroderma. Diffuse scleroderma shows anti topoisomerase - I (scl-70) antibody. Limited scleroderma, also known as CREST syndrome is associated with anticentromere antibodies.[18]

Age ranged from 13 to 61 years (mean 35.5±10.2 years) and most of the patients belonged to 20-30 age group followed by 31-40 age group. Valeur et al[19] labelled juvenile SSc when the age of the patient was less than 16 years and they reported 3% to 10% of total cases were of juvenile SSc. All the patients had clinical complaints of skin thickening and tightness, barring a few. Mean duration of skin manifestations was 5.4±5.8 years. Other than skin thickening, dyspnoea and dry cough were the frequently associated symptoms. 36% patients had duration of <6 months. 56% had 61%-80% FEV. Khanna et al[20] also had similar findings of mean FVC% as 67.7%. In our study a restrictive pattern of ILD in SSc was observed in 90% of the cases, as there was a decrease in both % predicted FVC and FEV1 with an increase in the FEV1/ FVC ratio. Goh et al[21] observed a mean FVC of 77.6±18.6, FEV1 of 78.7±21.4; higher values than our study. The reason for this difference in observations might be due to more severe patients or less number of patients in our study. Schurawitzki et al[22] also noted that only 39% cases had findings on chest radiograph; most had interstitial opacification and 26% of cases had equivocal radiological findings, while Bastos et al[23] reported this in 25% to 53% cases.

Chest radiography was not found to be a sensitive modality in evaluating ILD, particularly in the early stages of the disease. Among them, 56% had right lung involvement and 24% had left lung involvement. Most common chest radiograph finding was fine or coarse reticular opacity. Variable degree of lung volume loss was observed on radiography. GGO as an abnormality was found in 10% of the cases. Shah et al[24] had observed GGO in 66% of cases and GGO alone in 7% of cases. Few patients with only GGO or associated with fine reticulation (17%) had better lung function results (predicted FVC >70%) but higher total HRCT score.

Conclusion

Systemic sclerosis commonly occurs in middle aged females; the presenting complaint being diffuse skin thickening over the limbs and face and dyspnoea on exertion with or without dry cough as the primary respiratory symptom. PFT reveals restrictive pattern. The limitations of chest radiography may overcome by HRCT chest. Non-specific interstitial pneumonia pattern is the most common ILD found in SSc. HRCT has a significantly high diagnostic accuracy in detecting the presence or absence of ILD, especially in the early stages of the SSc. HRCT semi-quantitative scoring system is valuable in assessing the disease severity and its extent. It correlates significantly with PFT. HRCT as an imaging modality along with clinical findings can directly help in the treatment and prognostic assessment in very early or sub-clinical pulmonary disease.

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