|| ISSN(online): 2589-8698 || ISSN(print): 2589-868X ||
International Journal of Medical and Biomedical Studies
Available Online at www.ijmbs.info
NLM (National Library of Medicine ID: 101738825)
Index Copernicus Value 2019: 79.34
Original Research Article
Volume 3, Issue 3; March: 2019; Page No.257-262

A Radiographic Assessment Significance of Various Pulmonary and Extrapulmonary Abnormalities Chest in Scleroderma Lung

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Article Info: Received 12 February 2019; Accepted 21 March 2019

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Conflict of interest: No conflict of interest.

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 100 patients in the Department of Radiology 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.

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. 35% patients had duration of <6 months. 55% 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, 55% had right lung involvement and 45% had left lung involvement. No significant upper and middle zone involvement was seen. Most common chest radiograph finding was fine or coarse reticular opacity.

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.

Introduction

Chronic multisystem autoimmune condition systemic sclerosis (SSc) affects the skin and multiple organs. The condition is characterized as diffuse cutaneous (dcSSc) or localized cutaneous (LcSSc) based on cutaneous involvement. Lung involvement affects these individuals' prognoses. Pulmonary involvement and consequences cause most SSc morbidity and death.[1] Interstitial lung disease (ILD) and pulmonary hypertension (PH) are the most serious consequences and main causes of death. Over 90% of SSc patients exhibit ILD at autopsy, and 40% have abnormal pulmonary function tests.[2] HRCT chest is a trusted imaging method for SSc ILD detection and characterisation.[3,4] The illness often starts in the subpleural, posterior, and dependent lungs. SSc has ILD comparable to idiopathic nonspecific interstitial pneumonitis.[5,6]

SSc patients can be classified by skin involvement: limited cutaneous SSc (lcSSc) affects the hands, forearms, feet, and face, while diffuse cutaneous SSc (dcSSc) affects the elbows and perhaps the trunk.[7] SSc often causes interstitial lung disease (ILD).[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] Sometimes SSc-ILD is the first sign of the illness, although it is frequently found following a patient's examination.[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 100 patients in the Department of Radiology, Gouri Devi Institute of Medical Sciences and Hospital, Durgapur , West Bengal, 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%.

RESULTS

Table 1: Baseline profile and pulmonary function data

Age Group (years)	Number of Patients (%)
10-20	10 (10)
21-30	35 (35)
31-40	30 (30)
41-50	10 (10)
51-60	10 (10)
61-70	5 (5)
IIP Pattern	
NSIP	65 (65)

UIP	25 (25)
Organising pneumonia	3 (3)
No abnormality	7 (7)
Duration of cutaneous manifestation	
<5	72 (72)
6–10	20 (20)
>10	8 (8)
Duration of respiratory symptoms	
No complaints	5 (5)
<6 months	35 (35)
6–12 months	34 (34)
>12 months	26 (26)
Forced vital capacity	
>80%	15 (15)
61%–80%	55 (55)
41%–60%	30 (30)
<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. 35% patients had duration of <6 months. 55% had 61%-80% FEV.

Table 2: Pulmonary function tests and radiological imaging findings

FEV_1/ FVC	**Number of Patients (%)**
70%–85%	5 (5)
86%–100%	55 (55)
>100%	40 (40)
Lung zone involved	
Right	55 (55)
Left	45 (45)
Predominant chest radiograph finding	
Reticular	45 (45)
Reticulo-nodular	0
No abnormality detected	55 (55)
HRCT chest findings	
Only GGO	10 (10)
Mixed GGO + Reticular inter-lobular septal thickening	65 (65)
Only reticular fibrosis	15 (15)
Honey-combing	20 (20)
Traction bronchiectasis	40 (40)
HRCT chest score	
0–5	40 (40)
6–10	45 (45)

11–15	15 (15)
(Mean HRCT score) FVC% predicted	
>80% (4)	15 (15)
60%–80% (5.5)	50 (50)
<60% (8.8)	28 (28)
Could not perform (11.6)	7 (7)

Chest radiography was not found to be a sensitive modality in evaluating ILD, particularly in the early stages of the disease. Among them, 55% had right lung involvement and 45% 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. 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. 35% patients had duration of <6 months. 55% 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, 55% had right lung involvement and 45% 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. 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

In middle-aged women, systemic sclerosis causes extensive skin thickening on the limbs and face and exercise-induced dyspnoea with or without dry cough. Restricted PFT. HRCT overcomes chest radiography's drawbacks. Non-specific interstitial pneumonia dominates SSc ILD. HRCT is accurate for ILD diagnosis, especially in early SSc. Semi-quantitative HRCT grading determines sickness severity and extent. Highly associated with PFT. HRCT and clinical symptoms may treat and diagnose early pulmonary disease.

REFERENCES

1. Ioannidis JP, Vlachoyiannopoulos PG, Haidich AB, Medsger Jr TA, Lucas M, Michet CJ, Kuwana M, Yasuoka H, Van Den Hoogen F, Te Boome L, Van Laar JM. Mortality in systemic sclerosis: an international meta-analysis of individual patient data. The American journal of medicine. 2005 Jan 1;118(1):2-10.
2. Varga J. Systemic sclerosis: an update. Bulletin of the NYU hospital for joint diseases. 2008 Sep 1;66(3):198-.
3. Goldin JG, Lynch DA, Strollo DC, Suh RD, Schraufnagel DE, Clements PJ. High resolution CT findings in scleroderma-related lung diseases: findings from Scleroderma Lung Study. Chest. 2008;134(2):358-67.
4. Kim EA, Johkoh T, Lee KS, Ichikado K, Koh EM, Kim TS, Kim EY. Interstitial pneumonia in progressive systemic sclerosis: serial high-resolution CT findings with functional correlation. Journal of computer assisted tomography. 2001 Sep 1;25(5):757-63.
5. Desai SR, Veeraraghavan S, Hansell DM, Nikolakopolou A, Goh NS, Nicholson AG, Colby TV, Denton CP, Black CM, Du Bois RM, Wells AU. CT features of lung disease in patients with systemic sclerosis: comparison with idiopathic pulmonary fibrosis and nonspecific interstitial pneumonia. Radiology. 2004 Aug;232(2):560-7.
6. Bouros D, Wells AU, Nicholson AG, Colby TV, Polychronopoulos V, Pantelidis P, Haslam PL, Vassilakis DA, Black CM, Du Bois RM. Histopathologic subsets of fibrosing alveolitis in patients with systemic sclerosis and their relationship to outcome. American journal of respiratory and critical care medicine. 2002 Jun 15;165(12):1581-6.
7. Wollheim FA. Classification of systemic sclerosis. Visions and reality. Rheumatology. 2005 Oct 1;44(10):1212-6.
8. Steele R, Hudson M, Lo E, Baron M, Canadian Scleroderma Research Group. Clinical decision rule to predict the presence of interstitial lung disease in systemic sclerosis. Arthritis care & research. 2012 Apr;64(4):519-24.
9. Vonk MC, Broers B, Heijdra YF, Ton E, Snijder R, Van Dijk AP, van Laar JM, Bootsma H, van Hal PT, van den Hoogen FH, van Daele PL. Systemic sclerosis and its pulmonary complications in The Netherlands: an epidemiological study. Annals of the rheumatic diseases. 2009 Jun 1;68(6):961-5.
10. Van Den Hoogen F, Khanna D, Fransen J, Johnson SR, Baron M, Tyndall A, Matucci-Cerinic M, Naden RP, Medsger Jr TA, Carreira PE, Riemekasten G. 2013 classification criteria for systemic sclerosis: an American College of Rheumatology/European League against Rheumatism collaborative initiative. Arthritis & Rheumatism. 2013 Nov;65(11):2737-47.
11. Fischer A, Brown KK. Interstitial lung disease in undifferentiated forms of connective tissue disease. Orphan Lung Diseases: A Clinical Guide to Rare Lung Disease. 2015:419-34.
12. Hoffmann-Vold AM, Aaløkken TM, Lund MB, Garen T, Midtvedt Ø, Brunborg C, Gran JT, Molberg Ø. Predictive value of serial high-resolution computed tomography analyses and concurrent lung function tests in systemic sclerosis. Arthritis & rheumatology. 2015 May;67(8):2205-12.
13. Wells AU. High-resolution computed tomography and scleroderma lung disease. Rheumatology. 2008 Oct 1;47(suppl_5):v59-61.
14. Ooi GC, Mok MY, Tsang KW, Wong Y, Khong PL, Fung PCW, et al. Interstitial lung disease in systemic sclerosis. Acta Radiol 2003;44:258–64.
15. Strollo D, Goldin J. Imaging lung disease in systemic sclerosis. Current rheumatology reports. 2010 Apr;12:156-61.
16. Pandey AK, Wilcox P, Julie O, Ellis J, Brown J, Leipsic J. Significance of various pulmonary and extrapulmonary abnormalities on HRCT of the chest in scleroderma lung. Indian journal of radiology and imaging. 2013 Oct;23(04):304-7.
17. Solomon JJ, Olson AL, Fischer A, Bull T, Brown KK, Raghu G. Scleroderma lung disease. Eur Resoir Rev 2013;22:6–19.
18. Peros-Golubicic T, Sharma OP. Clinical atlas of interstitial lung disease. Springer Science & Business Media; 2006 Nov 23.
19. Valeur NS, Stevens AM, Ferguson MR, Effmann EL, Iyer RS. Multimodality thoracic imaging of juvenile systemic sclerosis: emphasis on clinical correlation and high-resolution CT of pulmonary fibrosis. American Journal of Roentgenology. 2015 Feb;204(2):408-22.
20. Khanna D, Nagaraja V, Tseng CH, Abtin F, Suh R, Kim G, Wells A, Furst DE, Clements PJ, Roth MD, Tashkin DP. Predictors of lung function decline in scleroderma-related interstitial lung

disease based on high-resolution computed tomography: implications for cohort enrichment in systemic sclerosis–associated interstitial lung disease trials. Arthritis research & therapy. 2015 Dec;17:1-0.

21. Goh NS, Desai SR, Veeraraghavan S, Hansell DM, Copley SJ, Maher TM, Corte TJ, Sander CR, Ratoff J, Devaraj A, Bozovic G. Interstitial lung disease in systemic sclerosis: a simple staging system. American journal of respiratory and critical care medicine. 2008 Jun 1;177(11):1248-54.
22. Schurawitzki H, Stiglbauer R, Graninger W, Herold C, Pölzleitner D, Burghuber OC, Tscholakoff D. Interstitial lung disease in progressive systemic sclerosis: high-resolution CT versus radiography. Radiology. 1990 Sep;176(3):755-9.
23. Bastos AD, Corrêa RD, Ferreira GA. Tomography patterns of lung disease in systemic sclerosis. Radiologia Brasileira. 2016 Sep;49:316-21.
24. Shah RM, Jimenez S, Wechsler R. Significance of ground-glass opacity on HRCT in long-term follow-up of patients with systemic sclerosis. Journal of thoracic imaging. 2007 May 1;22(2):120-4.