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Original Research Article

TO STUDY THE FREQUENCY OF OCCURRENCE OF PULMONARY INVOLVEMENT IN PROGRESSIVE SYSTEMIC SCLEROSIS (PSS)

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Abstract

Background: Systemic sclerosis is a generalized disorder of connective tissue affecting skin and internal organs. Lung involvement accounts for significant morbidity and is a leading cause of mortality in patients. This study intends to study the frequency of occurrence of pulmonary involvement in progressive systemic sclerosis (PSS) and to describe the clinical and radiological picture of pulmonary involvement in PSS. **Materials and Methods:** This was a descriptive cross‐sectional study. A detailed history, modified Rodnan score, clinical examination, routine investigation, antinuclear antibody, immuno biot, chest X‐ray (CXR), pulmonary function test (PFT), and 6 min walk test (6MWT) were performed on all patients. High resolution computed tomography was done on those who consented. **Result:** Hundredsubjects with PSS were included in the study; 90 were females and 10 were males. Common presenting complaints were skin thickening in 99% and Raynaud's phenomenon in 97%. Skin thickening of digits beyond metacarpo phalangeal was seen in 99%, face and neck in 93%, and hands in 93%. Chest wall thickening was seen in 50 subjects (40%). 90 (90%) of the studied subjects had pulmonary involvement, longer duration of disease was significantly associated with pulmonary involvement ($P < 0.05$). Dyspnea, cough, bilateral crepitations, CXR, Borg score, and Rodnan score was found to be significantly associated with severe pulmonary involvement ($P < 0.05$). **Conclusion:** The prevalence of pulmonary involvement in this cohort study was 90%. Almost 1/3rd of patients, that is 30 (30%) were detected to have pulmonary involvement despite being asymptomatic for respiratory complaints, hence early screening and evaluation is recommended. PFT and 6MWT are noninvasive, cost‐effective, and easily available screening tests which can be used in resource‐limited settings.

INTRODUCTION

Systemic sclerosis is a systemic autoimmune disease that is characterized by endothelial dysfunction resulting in a small‐vessel vasculopathy, fibroblast dysfunction with resultant excessive collagen production and fibrosis, and immunological abnormalities.[1] Systemic sclerosis is an acquired, sporadic disease with worldwide distribution, with peak age incidence of 30–50 years, preferentially affecting women (M: F = 1:3–4). Lung involvement accounts for significant morbidity and is a leading cause of mortality in patients with systemic sclerosis.[2,3] The exact prevalence of interstitial lung disease (ILD) in systemic sclerosis (SSc) is difficult to estimate because the patient is clinically asymptomatic early in the course.[4] Earlier studies have reported 74–100% prevalence of ILD in SSc patients at autopsy.[5,6] Ninety‐one percent of patients had interstitial abnormalities on high resolution computed tomography (HRCT).[6,7] This study intends to study the frequency of occurrence of pulmonary involvement in SSc and to describe the clinical and radiological picture of pulmonary involvement in SSc.[8] Statistical analysis also made to study the predictors of severity of pulmonary involvement in SSc.

MATERIALS AND METHODS

This was a descriptive cross‐sectional study. SSc patients attending Immunology/Pulmonary medicine outpatient departments of a tertiary care hospital in DMCH, Darbhanga, Bihar from March 2022 to June 2023. The Institution Review Board of the hospital approved the study. The patients aged more than 18 years with SSc satisfying the American Rheumatism Association criteria who consented for the study were included. Patients with other collagen vascular disease/mixed connective tissue disorder, and

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overlap syndromes were excluded. After obtaining an informed consent, basic demographic details were collected, detailed history, and clinical examination was noted. Modified Rodnan skin score was calculated for all the patients.[1] Antinuclear antibody was determined by indirect immunofluorescence in all the patients and its various patterns were noted. Immunoblot was used to determine antigenic specificities by immunodiffusion kits.[3,4] Pulmonary function test (PFT) and 6 min walk test (6MWT) were performed according to American thoracic society guidelines.[5] Restrictive lung disease was diagnosed if the percentage predicted forced vital capacity (FVC) was <80%.[9] Obstructive lung disease was diagnosed if the forced expiratory volume at 1 s/FVC was <70%. The percent of predicted DLCO was obtained using the single breath technique. Abnormal diffusing capacity was defined by a percent predicted DLCO <80%. All patients underwent a chest radiograph, and HRCT was performed when, indicated.

Statistical analysis: Descriptive statistical analysis has been carried out results on continuous measurements are presented on (mean ± standard deviation), and results on categorical.

RESULTS

The baseline characteristics of the study subjects are tabulated in [Table 1]. Common presenting complaints and their duration such as skin thickening, Raynaud's phenomenon, dyspnea, heart burn, and cough are also described in [Table 1].

Skin changes namely, digital tip pitting was noted in 78 subjects, sclerodactyly in 65 patients, nail capillary loop abnormality in 57, nail fold infarct in 47, flexor deformity in 47, and digital gangrene in 24 subjects. In skin thickening, digit beyond metacarpo phalangeal was involved in 98 subjects, face and neck was involved in 93 subjects, and hands were involved in 93 subjects. Anterior chest wall thickening was seen in 45 subjects. Among systemic findings, 50 subjects had bilateral crepitations and 32 subjects had loud P2. Blood investigations showed Hb <10 g % in 17 subjects, peripheral smear performed in 41 subjects showed predominantly normocytic normochromic picture in 24 (57%). Erythrocyte sedimentation rate >30 mmHg was seen in 42 subjects, 5 subjects had creatinine >1.2 mg. Antinuclear antibody was present in 99 subjects. On analysis of the immunoblotting for antinuclear antibodies, anti‑Scl‑72 (84 subjects) was the most common followed by antiro‑54 in 27, and anti‑RNP in 22 subjects [Table 2]. Among radiological features, 50% subjects had reticulonodular shadows on CXR and 61% subjects had normal CXR. Among 100 subjects, 87 underwent HRCT, basal sub pleural honey combing was the predominant finding in 33.1% followed by fibrosing alveolitis seen in 18.5% of subjects. Pulmonary involvement was defined as either radiological involvement (CXR/HRCT) or by Spirometry.[8] Using this criteria, 91% of the studied subjects had pulmonary involvement. Among 90 subjects with pulmonary involvement, all of them had skin thickening and Raynaud's phenomenon. Among 90 subjects, 31 were asymptomatic for respiratory complaints, in whom 14 subjects had FVC <70%, 19 subjects had desaturation >4%, 7 had reticular shadows on CXR, and 24 had parenchymal involvement on HRCT. Either dyspnea or cough was not significantly associated with pulmonary involvement (P > 0.05).

Table 1: The baseline characteristics of the study subjects.

Characteristics	Mean±SD or median (range) or n (%)
n=100	
Male:Female	10:90
Age (years)	39.25±11.99
Skin thickening/Raynaud phenomenon (months)	
Dyspnea (months)	24.00 (1.00-180.00)
GERD (months)	6.00 (1.00-120.00)
Skin (%)	12.00 (1.00-120.00)
Digit tip pitting	78 (78)
Sclerodactyly	65 (65)
Nail capillary loop abnormality	55 (5)
Digital gangrene	16 (16)
Skin thickening (%)	
Digit beyond MCP	99 (99)
Hand	93 (93)
Face and neck	93 (93)
Forearm	78 (78)
Anterior chest wall	45 (45)

Table 2: Investigation

Characteristics Number (percentage) or Mean±SD	
ANA	
Positive (%)	98 (98)
Immunoline	
	Anti‑Scl‑70 (%) 84 (84)
	Anti‑RNP (%) 21 (21)

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Anti‐ centromere B antibody (%)	5 (5)
CXR	
Reticulo nodular shadows (%)	45 (45)
HRCT	
Total	88
Involved (%)	86 (97.7)
FVC (L/min)	1.81±0.63
FEV1/FVC (percentage of predicted)	89.81±8.52

Table 3: Predictors of severity of pulmonary involvement in systemic sclerosis

Severe pulmonary involvement P		
No (55) Yes (20)		
Age	37.78±12.	19 41.65±11.27 0.219
Skin thickening	18.00 (0.00-120.00)	27.00 (6.00-180.00) 0.127
Raynaud's phenomenon 0.187	24.00 (1.00-120.00)	27.00 (3.00-120.00)
Digital tip pitting 0.430	47 (85.45)	19 (95.00)
Nail fold infarct 0.076	23 (41.82)	13 (65.00)
Sclerodactyly 0.801	34 (61.82)	13 (65.00)
Gangrene 0.693	6 (10.91)	6 (10.91)
Anterior chest wall		
Thickening		
0.147	20 (36.36)	11 (55.00)
Bilateral crepts 0.031	23 (41.82)	14 (70.00)
Borg scale 002	1.41±1.26	2.45±1.28 0.
Rodnan score 0.031		25.84±7.07 29.90±7.13

DISCUSSION

In earlier studies, it has been shown that organ damage occurs early in SSc. Renal crisis occurs within the first 4 years of disease and pulmonary fibrosis occurs in the first 2 years of disease, even though patients are often asymptomatic.[2-10] As the patients with organ damage have poor prognosis, all the patients should be carefully evaluated and followed‐ up in the initial 3 years for organ involvement.[9,2] This facilitates the early identification and initiation of appropriate therapy The present study was designed to evaluate the prevalence, clinical features, and predictors of pulmonary involvement in SSc. Majority, 90/100 (90%), of study subjects were females. The common presenting complaints were Raynaud's phenomenon, skin thickening, joint pain, dyspnea, and heart burn. The median duration of disease was 2 years (1 month – 13 years), increased disease duration was significantly associated with pulmonary involvement (P = 0.04). Almost all the patients had skin involvement, but its extent and severity was not significantly associated with pulmonary involvement. Four percent had creatinine >1.2 indicating the incidence of renal involvement was low compared to western studies which showed incidence of 19%. Loud P2 was seen in 28% of subjects and ECHO showed pulmonary artery systolic pressure >25 mmHg in 48% of subjects, incidence was higher compared to the western and other Indian studies. One third of patients were asymptomatic, FVC was <70% predicted in 13.3%, desaturation >4% was seen in 18.8% and 26.6% had pulmonary involvement on HRCT. HRCT was very sensitive in detecting pulmonary involvement even though patients were asymptomatic for respiratory complaints. Hence we should consider PFT, 6MWT, and HRCT in all patients to facilitate early detection. Various factors predicting the severity of pulmonary involvement was studied which showed respiratory symptoms, Rodnan score, Borg score, and CXR were significantly associated with severe organ damage. 6MWT is a simple, easy, and cost‐ effective test, and can be easily performed on all patients. Although PFT and HRCT were not done in all patients, it together increased the early detection. Recent update also recommends follow‐ up of PFT and 6MWT every 6 months for monitoring disease progression, so that appropriate treatment can start early.[10] This study examined only predictors of lung involvement at one point, further follow‐ up regarding disease progression, management, and response would help to plan better management protocol.

CONCLUSION

The study was conducted on 100 patients with SSc. Skin thickening and Raynaud's phenomenon was observed in all study subjects. Twenty‐ nine subjects were asymptomatic for respiratory complaints. Hypothyroidism as a co‐ existing illness was noted in 14 (46%) of all subjects. The prevalence of pulmonary involvement in this study was 90%. Duration of disease and abnormal CXR were significantly associated to be predictors of pulmonary involvement. 26.6% of them had severe pulmonary involvement. Respiratory symptoms, Rodnan score, bilateral crepts, and abnormal CXR were a predictor of severe pulmonary involvement. Almost 1/3rd of patients was detected to have pulmonary involvement despite being asymptomatic for respiratory complaints, hence early screening and evaluation is recommended. PFT and 6MWT are noninvasive, cost effective, easily available screening tests which can be used in resource limited settings.

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