

A STUDY OF PLEURO-PULMONARY MANIFESTATIONS IN SYSTEMIC SCLEROSIS - HOSPITAL BASED OBSERVATIONAL STUDY

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ABSTRACT

BACKGROUND

Systemic sclerosis (SSC) is a chronic multisystem disorder of unknown aetiology characterized clinically by thickening of the skin caused by accumulation of connective tissue and by involvement of visceral organs, including the gastrointestinal tract, lungs, heart and kidneys. The annual incidence of systemic sclerosis occurs at a rate of 2 per million population per year. The prevalence of systemic sclerosis is 50 per 1,00,000 population.

The objectives of the study were- to detect pleuropulmonary involvement in systemic sclerosis patients by clinical examination and investigations including x-ray chest, high resolution computerized tomography, pulmonary function test, electrocardiograph, and echocardiogram with Doppler study of pulmonary arterial pressure and to analyse the incidence of various types of pulmonary manifestations.

MATERIALS AND METHODS

This is a hospital based prospective observational study conducted in Thanjavur Medical College. About 22 patients who were diagnosed as systemic sclerosis were enrolled in the study between January 2017 to December 2017. The pleuro-pulmonary manifestations are analysed both clinically and radiologically and results were studied in detail.

RESULTS

The incidence of pulmonary involvement in SSC was significantly high in study and the pattern of involvement were interstitial lung disease being the most common followed by parenchymal lung lesion and pleural involvement as pleural thickening and effusion. The most common symptoms were unproductive cough and non-specific chest pain and effort dyspnoea. Most of the above symptoms were found in combination in the same patients. Chest x-rays, PFT, HRCT Procedures were used, and HRCT happens to be having the highest sensitivity rate even more than the PFT among the 3 diagnostic methods.

CONCLUSION

The incidence of Pulmonary involvement in SSC was significantly high in study and the pattern of involvement were interstitial lung disease being the most common followed by parenchymal lung lesion and pleural involvement as pleural thickening and effusion. The most common symptoms were unproductive cough and chest pain which were not specific and effort dyspnoea. Most of the above Symptoms were found in combination in the same patients. Chest x-rays, PFT, HRCT Procedures were used, and HRCT happens to be having the highest sensitivity rate even more than the PFT among the 3 diagnostic methods.

To conclude, the pleuropulmonary involvement was significantly high and almost all the findings concur with that as found in the literature and still more facts can come to light with the use of advanced measures like immunological study.

KEYWORDS

Systemic sclerosis, pleuro-pulmonary manifestations.

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BACKGROUND

Pulmonary Involvement in Systemic Sclerosis

The most common initial symptoms is shortness of breath on exertion, later a non-productive cough can be

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problematic. Chest pain is generally not caused by scleroderma lung disease and if present can usually be explained by another process such as musculoskeletal pain, reflux oesophagitis, pleurisy and pericarditis.^{1,2} Both interstitial fibrosis and pulmonary arterial vascular disease are present in the lung of patients. Approximately 80% of patients will have abnormal function measured by specific and sensitive testing. Majority of lung disease will be clinically silent.³ Pulmonary interstitial fibrosis is preceded by a neutrophilic alveolitis that is best detected by high - resolution computed tomography and or bronchoalveolar lavage. A chest radiograph is relatively insensitive, but

fibrosis appears as reticulonodular changes in the lower lobes of lung parenchyma. When active alveolitis is present demonstrated by a 'ground glass' appearance of the lung in high resolution CT the patient will have higher level of dyspnoea.^{4,5} Pulmonary hypertension is detected by measuring pulmonary artery pressure with 2D-echocardiography.^{6,7} The onset of pulmonary fibrosis is

within first 3 years of disease in most patients. The greatest loss of lung volume occurs within first 2 years.

Exertional dyspnoea is the most common symptom. A subset of patients develops progressive pulmonary fibrosis and proceed to have severe restrictive lung disease. Pulmonary related deaths occurred with greater frequency in the second 5 years from disease onset.⁸

Items	Sub items	Weight/score
Skin thickening of the fingers of both hands extending proximal to metacarpophalangeal joints (sufficient criterion)	-	9
Skin thickening of the fingers (only count the higher score)	Puffy fingers	2
	Sclerodactyly of the fingers (distal to MCP but proximal to PIP)	4
Fingertip lesions (only count the higher score)	Digital tip ulcers	2
	Finger pitting scars	3
Telangiectasia	-	2
Abnormal nail fold Capillaries	-	2
Pulmonary arterial hypertension and/or Interstitial lung disease	Pulmonary arterial hypertension	2
	Interstitial lung disease	2
Raynaud's phenomenon	-	3
Scleroderma related Auto-Antibodies	Anti-Centromere	3
	Anti-Topoisomerase	
	Anti-RNA Polymerase III	
ACR/EULAR Criteria for Diagnosing Systemic Sclerosis (2013)⁹		

Patients with total score of more than 9 are classified as having definite Systemic Sclerosis.

Prognosis is worse with patients with¹⁰

- (1) DLCO is less than 40% of predicted value
- (2) Increased percentage of neutrophils or eosinophils on Broncho alveolar lavage cell differential count.

The serum markers that are increased in systemic sclerosis with pulmonary fibrosis are:¹¹

- (1) Monocyte chemoattractant protein - 1
- (2) Macrophage inflammatory protein - 1
- (3) Connective tissue growth factor
- (4) Interleukin - 6 receptor level
- (5) Surfactant - D
- (6) Earliest changes in open lung biopsies from patients who have systemic sclerosis with interstitial lung disease are patchy lymphocyte and plasma cell infiltration of the alveolar walls.
- (7) Lung biopsies also show evidence of endothelial and epithelial injury plus interstitial oedema and excess collagen deposition.
- (8) Pulmonary fibrosis in systemic sclerosis is associated with increased deposition of type I and type III collagen.

Although chest radiography has limited value in detecting early stages of disease, conventional chest radiography remains an invaluable aid in documenting the presence of ILD. Currently HRCT is the imaging method of choice is evaluating patients with known or suspected chronic or acute infiltrative lung disease. It is also indicated for the evaluation of small airway diseases such as

bronchiectasis and bronchiolitis. PFT are used to aid diagnosis, assess functional impairment and monitor treatment of progression of disease.

MATERIALS AND METHODS

A total of 22 patients who were provisionally diagnosed as systemic sclerosis (SSC) in Thanjavur Medical College Hospital from January 2017 to December 2017 were admitted and evaluated in department of general medicine, TMCH, Thanjavur for this study. This is a prospective observational study.

Inclusion Criteria

All patients who satisfied the criteria for the classification of Systemic sclerosis by American College of rheumatology were included in this study.

Exclusion Criteria

Patients who give history of chronic obstructive pulmonary disease, nicotine abuse and industrial exposure were excluded from this study.

Evaluation of Patients

Symptoms related to respiratory system such as cough with or without expectoration, haemoptysis, wheeze, fever, pleuritic chest pain and breathlessness, pedal edema were recorded.

Investigation

The Blood investigations included were, complete Blood count Renal Biochemistry, metabolic parameter and immunological test like ANA, estimation by indirect

immunofluorescence assay using Hep - 2 cells substrate. Special Blood test like Antitopoisomerase I was also done. X-ray Chest, ECG and Echocardiogram were taken for all the patients.

All the 22 Patients were subjected to pulmonary function tests and HRCT Scan. PFT was done to make out whether patients had restrictive or obstructive pattern. HRCT was done to identify and assess pleural and parenchymal involvement. Sputum culture for bacteria and sputum for AFB were done in relevant cases. Skin biopsy was carried out for 10 patients who were willing for the procedure.

RESULTS

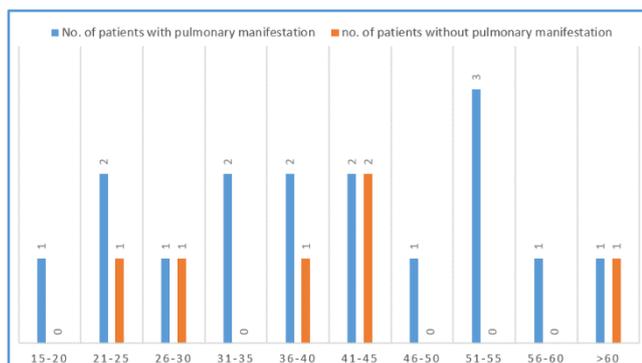


Figure 1. Correlation of Age and Pulmonary Manifestations

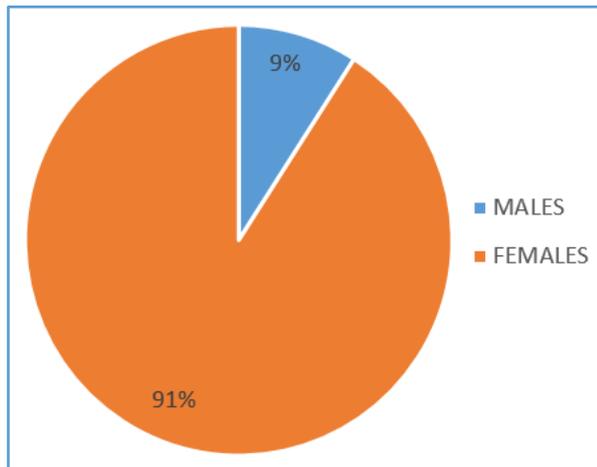


Figure 2. Sex Distribution

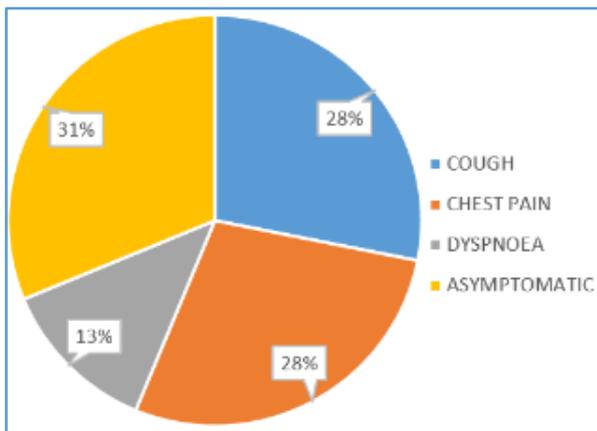


Figure 3. Respiratory Symptoms

There was overlap of symptoms and they were found in combination.

Symptomatic patients	:	12(54%)
Asymptomatic Patients	:	10(46%)

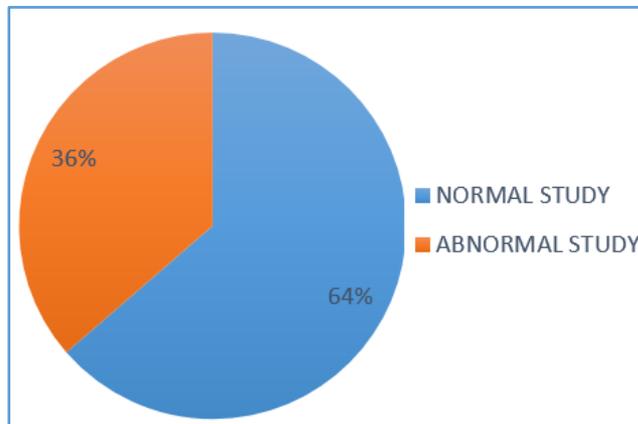


Figure 4. Incidence of X-Ray Abnormalities

Abnormal x-ray findings are-

Patchy infiltration	:	2
Apical infiltration	:	1
Homogenous Opacity	:	5

X ray is 36% sensitive in detecting SSc

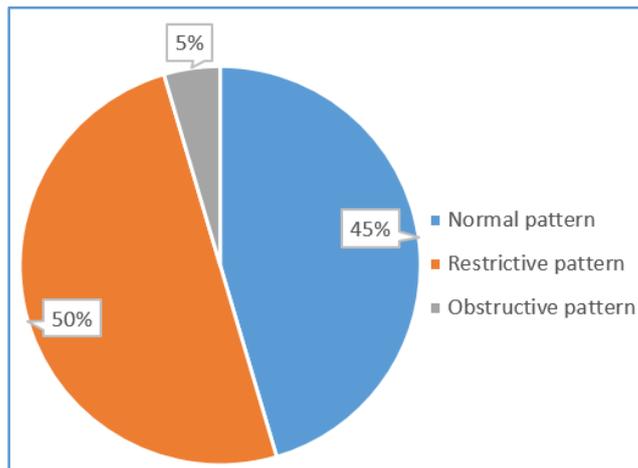


Figure 5. Pulmonary Function Test Results

Among 12 symptomatic patients, 10 were PFT positive and hence PFT is 83% sensitive in detecting SSc.

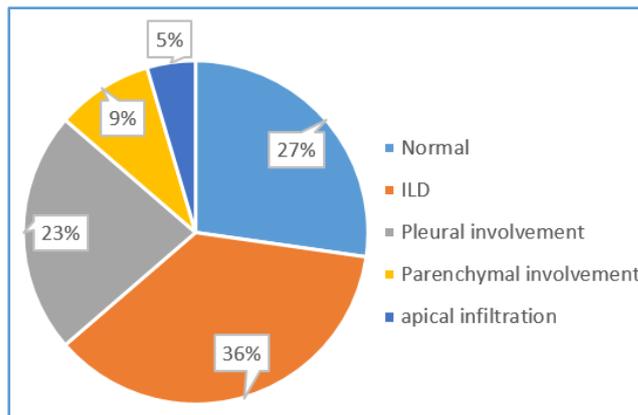


Figure 6. HRCT Scan Finding

Pleural involvement is defined as presence of pleural thickening with or without pleural effusion. ILD changes observed are reticulonodular and ground glass appearance. Parenchymal changes include consolidation with and without cavity.

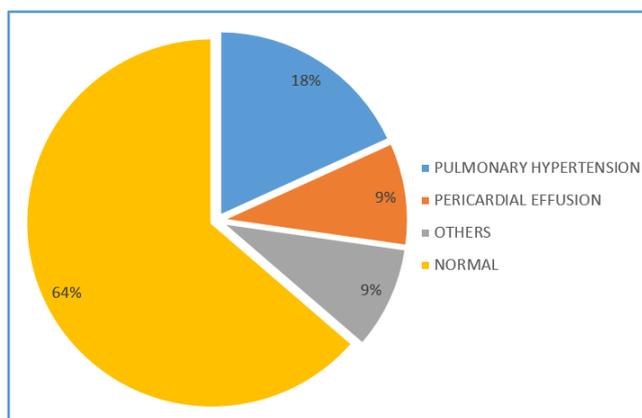


Figure 7. Echocardiogram Results

Respiratory Symptoms	No. of Patients with Pleuro-Pulmonary Involvement with HRCT	No. of Patients Without Pleuro-Pulmonary Involvement with HRCT
No Symptoms	6	4
With Symptoms	10	2

Table 1. Correlation of Respiratory Symptoms and HRCT Changes

X-ray Chest Result	No. of Patients with pleuro Pulmonary Involvement with HRCT	No. of Patients without Pleuro Pulmonary Involvement with HRCT
Normal Study	8	6
Abnormal Study	8	-

Table 2. Correlation of X-Ray Chest and HRCT Changes

DISCUSSION

In our study 22 patients admitted between January 2017 to December 2017 with Systemic Sclerosis were taken up for study regarding the pleuropulmonary involvement and all were found to be having diffuse Systemic Sclerosis. The age distribution was found between 15 years and 70 years and the maximum number of patients was in the age group between 41-45 years (4 number) (Fig. 1). Female: Male ratio was 10:1 in our study as compared to 7:1 cited in the literature (Fig. 2). Commonest Symptoms were unproductive cough, Chest pain attributable to respiratory, Gastro intestinal, cardiac causes and effort dyspnoea (Fig. 3). X-ray was able to pick up positive finding in 8 patients (36%) The percentage of Insensitivity is about 37% in the Symptomatic Patients in our Study (Fig. 4). So, X-ray Chest cannot be taken as a dependable investigation for SSC. PFT is abnormal among 12 patients in the study. Among that

12, 10 patients were symptomatic (Fig. 5), this study shows HRCT has the highest sensitivity rate compared with PFT and x-ray in diagnosing the pulmonary involvement in SSc (Fig -6). In Echocardiogram findings, pulmonary hypertension was present in 4 patients (18%), pericardial effusion in 2 patients (9%), and Normal echo study in 14 Patients (64%) (Fig. 7). HRCT also picked up 6 out of 10 asymptomatic cases (table. 1). X ray is abnormal in 8 patients, but HRCT detected abnormality in 8 patients in which Chest X ray was normal study (Table 2).

CONCLUSION

The incidence of Pulmonary involvement in SSC was significantly high in study and the pattern of involvement were interstitial lung disease being the most common followed by parenchymal lung lesion and pleural involvement as pleural thickening and effusion. The most common symptoms were unproductive cough and chest pain which were not specific and effort dyspnoea. Most of the above Symptoms were found in combination in the same patients. Chest x-rays, PFT, HRCT Procedures were used, and HRCT happens to be having the highest sensitivity rate even more than the PFT among the 3 diagnostic methods.

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