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CARDIO-PULMONARY PROFILE IN LIMITED AND DIFFUSE SYSTEMIC SCLEROSIS.

General Medicine

Junumani Baishya Post Graduate Trainee Department of Medicine, Assam Medical College and Hospital
Rebecca R Marak Assistant professor Department of Medicine, Assam Medical College and Hospital,
Sangita Deka* Post Graduate Trainee Department of Medicine, Assam Medical College and Hospital *Corresponding Author
Mriganka Shekhar Chaliha Associate Professor Department of Cardiology Assam Medical College and Hospital,
Pronami Borah Associate Professor Department of Radiodiagnosis Assam Medical College and Hospital.

ABSTRACT

Background: Scleroderma, systemic sclerosis (SSc), is a chronic multisystem autoimmune disease characterized by a vasculopathy, diffuse fibrosis of skin and numerous internal organs, and immune abnormalities. The clinical manifestations of this disease are extremely heterogeneous.1 Pulmonary and cardiac involvement of scleroderma are responsible for significant morbidity and mortality.2 This study aims to evaluate and compare different cardiac and pulmonary manifestations between limited and diffuse systemic sclerosis. **Materials And Methods:** 54 Systemic Sclerosis patients were studied at Assam Medical College and Hospital, from June 2020 to May 2021. **Results:** In our study among 54 patients, 46 were female (85.19%) and male (14.81%). Maximum (40.74%) patients were between the age group 41-50 years. 53.70% had diffuse and 46.30% had limited cutaneous systemic sclerosis. Dyspnoea on exertion (59.26%), cough (35.19%) and palpitation (20.37%). Dyspnea was significantly higher in diffuse SSc (75.86%) compared to limited SSc (40%). 72.22% of the studied population had ILD. Predominant HRCT findings were ground glass opacity (53.70%). 2D-ECHO abnormalities were observed in (62.96%) patients of diffuse SSc (72.41%); 58.62% of diffuse and 44% of limited cutaneous disease had abnormal ECG. PAH was observed in 28% in limited and 13.79% in diffuse Ssc. **Conclusion:** From this study we can conclude that though systemic manifestations and organ involvement was more common in diffuse SSc. Thorough evaluation in cases of SSc is essential for early diagnosis, treatment & better prognostic outcome.

KEYWORDS

Systemic Sclerosis, Pulmonary Hypertension, Cardiopulmonary Manifestation

BACKGROUND:

Systemic sclerosis is divided into two groups: Limited and diffuse cutaneous disease. The limited form (limited cutaneous systemic sclerosis) is characterized by thickening of the skin distal to the elbows and knees and is related to less severe visceral organ involvement. In addition to distal area involvement the diffuse form (diffuse cutaneous systemic sclerosis) involves skin proximal to the elbows and knees and is associated with more severe organ involvement and damage.

Evidence of pulmonary disease is found in over 80% of patients with systemic sclerosis.[3] Cardiac involvement is the end result of microvascular alterations, collagen overproduction by altered fibroblasts with extracellular matrix deposition and complex immune system dysregulation these leads to ischaemic, fibrotic and inflammatory lesions involving all cardiac structures, including the cardiac conduction system.[4] Pulmonary involvement is the leading cause of death in patients with systemic sclerosis. The specific mechanisms of disease progression remain not clear, however it is believed that inflammation and endothelial injury are common precursors. It might also manifest as interstitial lung disease or pulmonary artery hypertension. ILD and PAH are the most common cardiopulmonary findings in SSc.[5] During early stages simple bedside examination is not sensitive and early diagnosis is very important, therefore every patient with scleroderma need to be screened using objective testing including electrocardiography (ECG), ECHO, and pulmonary function test.

AIMS AND OBJECTIVES:

1) To evaluate different cardiopulmonary manifestations in diffuse and limited systemic sclerosis. 2) To evaluate cardiac manifestations by echocardiography and electrocardiogram in systemic sclerosis. 3) To correlate the findings in limited and diffuse systemic sclerosis.

METHODS:

A hospital based observational cross-sectional study was conducted at Assam Medical College and Hospital Dibrugarh enrolling 54 cases of systemic sclerosis irrespective of sex who fulfilled the American College of Rheumatology preliminary criteria (2013) for the diagnosis of SSc attending Rheumatology OPD and other outpatient departments or in various wards of department of Medicine at Assam Medical College and Hospital who fulfilled the inclusion criteria.

Ethical Clearance:
Ethics clearance was taken from the Institutional Ethics Committee (H) of Assam Medical College & Hospital, Dibrugarh.

Inclusion Criteria:
- SSc patients above 12 years.

Exclusion Criteria:
- Patients with other collagen vascular disease, mixed connective tissue disorder, overlap syndrome
- Previous history of Bronchial asthma/ Chronic Obstructive Airway Disease.
- Patients with respiratory infections, including pulmonary tuberculosis.
- Patients who are smokers.
- Patients with coronary artery disease, rheumatic heart disease or congenital heart disease.
- People who do not give consent.

a) Socio-demographic data including the age, gender, and duration of the disease were recorded.
b) History and detailed examination pertaining to the disease were recorded
c) Presenting cutaneous features (Raynaud"s phenomenon, skin sclerosis etc) and duration of the same were recorded.
d) Systemic manifestations (gastrointestinal, respiratory, cardiac, renal, musculoskeletal) were recorded.
e) Hemoglobin, White blood cell count, Differential count, ESR, Blood Urea, Serum Creatinine, Blood sugar, ECG, Chest X ray were done for all patients.
f) The antibody profile of the patients which include ANA, anti ds DNA, Scl 70 antibody and anticentromere antibody was also done.
g) All patients were subjected to spirometric evaluation, High Resolution Computerized Tomography (HRCT), Electrocardiogram and Echoardiography.
h) All the results were tabulated and statistical analysis were done. The statistical significance was fixed at 5% level (p value <0.05).

RESULTS:

1. In the study out of 54 patients, 85.19% were females and 14.81% were males respectively.

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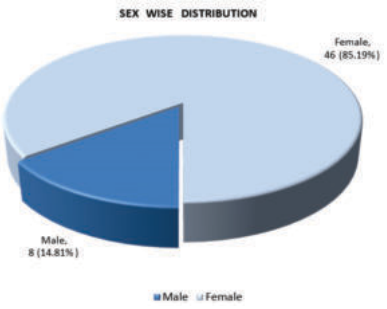
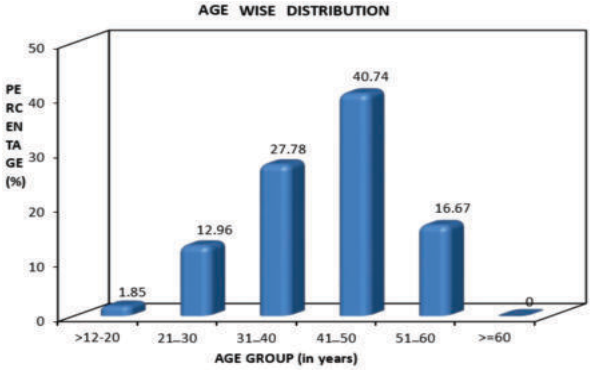
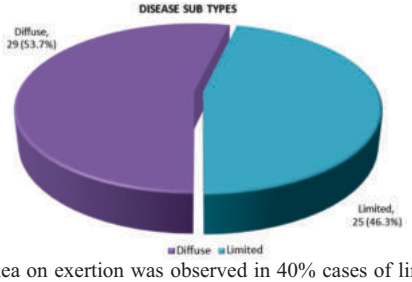


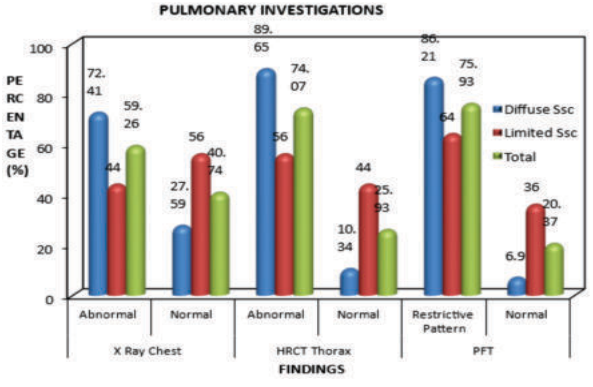
2. In the study maximum patients 40.74% were between 41-50 years of age. The mean age was 41.74 ± 10.17 years



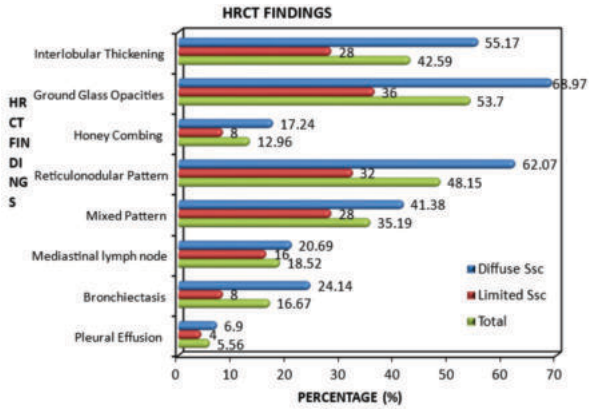
3. In the study maximum (29) were diffuse systemic sclerosis patients 53.70% of the study population and 46.30% had limited cutaneous systemic sclerosis.



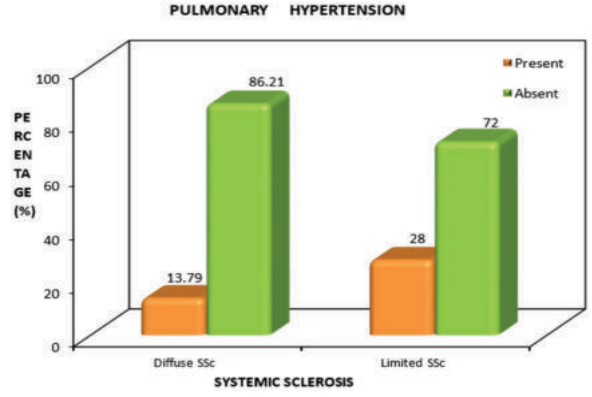
4. Dyspnea on exertion was observed in 40% cases of limited and 75.86% of diffuse systemic sclerosis and dyspnea was a significantly higher in diffuse SSc compared to limited SSc cases. Dry cough was observed in 35.19% cases, palpitation in 20.37% cases and chest pain in 18.52% case.
5. In the study 59.26% had abnormal chest x-ray, 74.07% showed abnormal HRCT findings and 75.93% had restrictive ventilatory abnormality on spirometry.
6. Restrictive pattern was found in 25 (86.21%) out of 29 patients with diffuse cutaneous systemic sclerosis compared to 16 (64%) out of 25 with limited cutaneous systemic sclerosis.



7. The commonest pulmonary manifestation found in both subset of systemic sclerosis was interstitial lung disease, detected in 72.22% of the study population.
8. The predominant patterns of involvement in HRCT included ground glass opacity in 53.70%, reticulonodular pattern in 48.15%, and inter lobar septal thickening in 42.59%.



9. 2D-ECHO abnormalities were observed in 34 (62.96%) patients; 21 (72.41%) in diffuse and 13 (52%) in limited cutaneous systemic sclerosis.
10. Electrocardiogram findings were abnormal in 28 (51.85%) patients. Right axis deviation and conduction block were found in 11 (20.37%), 3 (5.56%) patients had left axis deviation. 8 (14.81%) patients had non specific ST-T changes, 8 (14.81%) patients had sinus tachycardia. Arrythmia was detected in 3 (5.56%)
11. In the study 13.79% of dcSSc patients and 28% of lcSSc patients had Pulmonary hypertension.



DISCUSSION:

In the study majority of cases were female, which is similar to study done by Sharma VK et al.[6] In the study maximum patients (40.74%) were between 41-50 years of age, which was similar to the study done by Poormoghim et al.[7] In our study most of the patients belonged to the diffuse SSc group (53.70%), which is similar to those found in Indian study.[6] In the study, dyspnea on exertion was observed in (59.26%) of the study population, which was similar to the study by Owens et al. (60%).[8] In the study we observed chest X ray abnormality in 59.26%, This was comparable to that observed in earlier Indian studies by Sharma et al. (65.3%).[6] 74.07% shows abnormal HRCT findings in present study similar to a study by Seely et al. (73%).[9] Restrictive pattern on spirometry was significantly higher in diffuse than limited SSc patients; similar to Dilip Kumar SA et al.[10] In the study, (72.22%) commonest pulmonary manifestation found in both subset of systemic sclerosis in our present study was interstitial lung disease. Which was similar to the earlier Indian studies by Arakkal et al.[11] In the study, predominant patterns of involvement in HRCT included ground glass opacity in 53.7% and reticulonodular infiltrates in 48.15% which was similar to study done by Arakkal et al.[11] 2D-ECHO abnormalities were observed in 34 (62.96%) patients; 21 (72.41%) of diffuse and 13 (52%) of limited cutaneous systemic sclerosis patients which was similar to the study done by Arakkal et al.[11] In the study, Electrocardiogram findings were abnormal in 28 (51.85%) patients. It was similar to a study by Draeger et al.[12] in the study pulmonary artery hypertension was present in (20.37%). Which was similar to study conducted by A. Ghosh et al. where PAH was recorded in 24% of study population.[13]

Limitations:

1. Small sample size
2. Right heart catheterization to evaluate Pulmonary arterial hypertension, could not be done due to resource limitations.
3. Complete evaluation of arrhythmia with 24 hour holter

monitoring could not be done.

4. Further, since this was a cross sectional study we could not present any follow up data for these patients.

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