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ORIGINAL RESEARCH

EARLY DETECTION OF CARDIO-PULMONARY MANIFESTATIONS IN SYSTEMIC SCLEROSIS: A CROSS-SECTIONAL STUDY

Dr. M. Babu[1], Dr. N.K. Shiva[2], Dr. N. Karthick[2], Dr. S. Dinesh[2]

[1]Senior Assistant Professor, Department of General Medicine, Coimbatore Medical College and Hospital, Coimbatore, Tamil Nadu.
[2]Senior Resident, Department of General Medicine, Coimbatore Medical College and Hospital, Coimbatore, Tamil Nadu.

Corresponding Author: Dr. **S. Dinesh**, Senior Resident, Department of General Medicine, Coimbatore Medical College and Hospital, Coimbatore, Tamil Nadu.

ABSTRACT

Background: Systemic sclerosis (SSc) is a complex autoimmune disease that significantly impacts cardio-pulmonary health, leading to increased morbidity and mortality. Early detection of cardio-pulmonary manifestations is crucial for improving patient outcomes.

Methods: This cross-sectional study was conducted at Coimbatore Medical College Hospital over one year (July 2021 - June 2022). Thirty systemic sclerosis patients were evaluated using ECG, chest X-ray, echocardiogram, HRCT, and PFT. Inclusion criteria included diagnosed SSc patients above 18 years. Exclusion criteria involved secondary immunodeficiencies, diabetes, cancer, corticosteroid or cytotoxic drug use, pregnancy, lactation, psychiatric conditions, and non-consent.

Results: Among the participants, 83% were female, and the majority were aged 51-60 years. Pulmonary hypertension was detected in 40% of patients, ILD in 60%, and 10% showed diastolic dysfunction. Ground-glass opacities were the most common chest X-ray finding (46.7%).

Conclusion: Early detection of cardio-pulmonary complications in systemic sclerosis through comprehensive screening significantly enhances management and patient outcomes.

Keywords: systemic sclerosis, early detection, cardio-pulmonary manifestations, pulmonary hypertension, interstitial lung disease, echocardiography

INTRODUCTION

Systemic sclerosis (SSc), also known as scleroderma, is a complex autoimmune disease characterized by widespread microvascular damage and fibrosis of the skin and internal organs[1]. This progressive disorder poses significant challenges due to its heterogeneity and the involvement of multiple organ systems, leading to considerable morbidity and mortality. Among the various complications of systemic sclerosis, cardio-pulmonary manifestations are particularly devastating, often determining the overall prognosis and quality of life of affected individuals[2].

Cardio-pulmonary involvement in systemic sclerosis includes a spectrum of conditions such as pulmonary arterial hypertension (PAH), interstitial lung disease (ILD), and cardiac complications like myocardial fibrosis, pericarditis, and arrhythmias. These manifestations are not only common but also represent the leading causes of death in SSc patients. Early detection and timely intervention are crucial in managing these conditions, thereby reducing the associated morbidity and mortality and improving patients' quality of life[3].

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Early detection of cardio-pulmonary manifestations in systemic sclerosis is paramount. The insidious onset and progressive nature of these complications often mean that by the time clinical symptoms become apparent, significant and sometimes irreversible damage may have already occurred[4]. For instance, pulmonary arterial hypertension, a condition characterized by high blood pressure in the arteries of the lungs, often presents late in the disease course when therapeutic options are limited and less effective. Similarly, interstitial lung disease, which involves inflammation and scarring of lung tissue, can progress silently, leading to substantial loss of lung function before being diagnosed[4].

Advances in diagnostic techniques have enabled earlier identification of these complications. High-resolution computed tomography (HRCT) of the chest, echocardiography, pulmonary function tests (PFTs), and biomarkers such as N-terminal pro-brain natriuretic peptide (NT-proBNP) and cardiac magnetic resonance imaging (MRI) are invaluable tools in the early detection of pulmonary and cardiac involvement. Utilizing these modalities can significantly alter the disease trajectory by facilitating timely and appropriate therapeutic interventions[5].

The morbidity and mortality associated with cardio-pulmonary manifestations in systemic sclerosis are significant. Studies have shown that patients with PAH have a markedly reduced survival rate compared to those without. Similarly, the presence of ILD is a major determinant of long-term outcomes, with progressive fibrosis leading to respiratory failure. Cardiac involvement further exacerbates the prognosis, as myocardial fibrosis and arrhythmias can precipitate heart failure and sudden cardiac death[6].

These complications not only shorten lifespan but also severely impair the quality of life. Patients often experience debilitating symptoms such as severe shortness of breath, fatigue, chest pain, and palpitations, which limit their daily activities and overall functional capacity. The psychological burden of living with a chronic and progressive illness also contributes to decreased quality of life, emphasizing the need for comprehensive and multidisciplinary care approaches[7].

Given the profound impact of cardio-pulmonary manifestations on patients with systemic sclerosis, this study seeks to identify these complications at a stage where interventions can be most effective. Early identification and treatment of PAH, ILD, and cardiac involvement can slow disease progression, reduce hospitalizations, and improve survival rates. By mitigating the severity and frequency of cardio-pulmonary symptoms, the study aims to enhance the overall quality of life for patients.

MATERIALS & METHOD

Study Design: This research was conducted as a cross-sectional study. The design was chosen to evaluate the prevalence and early detection of cardio-pulmonary manifestations in systemic sclerosis patients at a specific point in time.

Study Place: The study was carried out at Coimbatore Medical College Hospital, a tertiary care center that provides comprehensive medical services, including specialized rheumatology care.

Study Time: The data collection took place over one year, from July 2021 to June 2022. This timeframe was selected to ensure a sufficient sample size and to account for any seasonal variations that might influence cardio-pulmonary symptoms.

Study Participants: Inclusion criteria were patients diagnosed with systemic sclerosis, both males and females, and patients aged above 18 years. Exclusion criteria were patients with secondary immunodeficiency states, such as HIV, individuals with diabetes mellitus, cancer patients, patients on corticosteroids or cytotoxic drugs, pregnant or lactating women, patient's incapable of giving consent, including psychiatric patients, and patients who were not willing to participate or refused to consent.

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Sample Size: The sample size for this study was determined to be 30 patients. This number was considered adequate to detect significant trends and patterns in the early detection of cardio-pulmonary manifestations in systemic sclerosis within the study period.

Sampling Technique: Patients attending the Rheumatology outpatient department and those admitted to the medical ward at Coimbatore Medical College Hospital during the study period were screened for eligibility. A purposive sampling technique was employed to select participants who met the inclusion criteria.

Study Methodology: Upon obtaining informed consent, patients were enrolled in the study. Baseline characteristics, detailed medical history, and physical examination findings were recorded for each participant. The study excluded minors, pregnant women, mentally ill, and non-volunteering patients, as well as those with conditions listed in the exclusion criteria.

Study Tools: The following diagnostic tools and investigations were utilized to assess cardio-pulmonary involvement:

1. Chest X-Ray: Used to detect any structural abnormalities in the lungs.
2. Electrocardiogram (ECG): Conducted to identify any cardiac arrhythmias or abnormalities in heart function.
3. Echocardiogram: Utilized to assess cardiac structure and function, including the detection of myocardial fibrosis and pericardial effusion.
4. High-Resolution Computed Tomography (HRCT) of the Chest: Performed to identify interstitial lung disease and evaluate the extent of lung involvement.
5. Pulmonary Function Test (PFT): Conducted to measure lung volumes and capacities, aiding in the detection of restrictive lung disease and pulmonary hypertension.

Statistical Analysis: Data were analyzed using appropriate statistical methods to identify the prevalence and early signs of cardio-pulmonary manifestations in systemic sclerosis patients. Descriptive statistics, such as numbers and frequencies, were used to summarize patient characteristics and clinical findings.

Ethical Issues: The study was conducted in compliance with ethical standards and principles outlined in the Declaration of Helsinki. Approval was obtained from the Institutional Ethics Committee of Coimbatore Medical College Hospital prior to the commencement of the study. Informed consent was obtained from all participants, ensuring that they were fully aware of the study's objectives, procedures, potential risks, and benefits. Confidentiality of patient information was maintained throughout the study, and data were anonymized to protect the privacy of participants.

RESULTS

The study included a total of 30 participants diagnosed with systemic sclerosis. The distribution of the participants by gender and age is presented in Table 1. Most of the participants were female (83%), with only 17% being male. The age distribution showed that the highest proportion of participants fell into the 51-60 years age group, accounting for 36.5% of the sample. The age groups 31-40 and 41-50 years comprised 26.8% and 20% of the participants, respectively. Participants under 30 years made up 13.4%, while only 3.3% were over 60 years old. Regarding the duration of the disease, 67% of the participants had been diagnosed with systemic sclerosis for 1-4 years, whereas the remaining 33% had been living with the disease for 5-8 years.

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Table 1: Characteristics of the study participants.

Variable		Number	Percentage
Gender	Male	5	17
	Female	25	83
Age group	<30	4	13.4
	31-40	8	26.8
	41-50	6	20
	51-60	11	36.5
	>60	1	3.3
Duration of disease	1-4 YRS	20	67
	5-8 YRS	10	33

The findings from the various diagnostic investigations conducted on the study participants are summarized in Table 2. The ECG results indicated that the majority of participants (90%) had normal findings. However, 6.7% of the participants showed left bundle branch block (LBBB), and 3.3% had supraventricular tachycardia (SVT). Chest X-ray findings revealed that 33.3% of the participants had normal results. Among those with abnormal findings, ground-glass opacity was the most common, observed in 46.7% of the participants. Additionally, 13.5% had pericardial effusion, and 6.5% exhibited a reticulonodular pattern.

Half of the participants (50%) had normal echocardiogram results. Of those with abnormalities, pulmonary hypertension was noted in 40% of the participants, while 10% had diastolic dysfunction. HRCT scans showed normal results in 40% of the participants. Among those with abnormal findings, usual interstitial pneumonia (UIP) was detected in 43.3%, and non-specific interstitial pneumonia (NSIP) was found in 16.7% of the participants. Pulmonary function tests revealed that 40% of the participants had normal forced vital capacity (FVC) and forced expiratory volume in one second (FEV1) values. However, 60% of the participants had reduced FVC with normal FEV1, indicating restrictive lung disease.

Table 2: Results of various investigations among the study participants.

Variable			Number	Percentage
ECG findings	Normal		27	90
	Abnormal	LBBB	2	6.7
		SVT	1	3.3
Chest X-ray findings	Normal		10	33.3
	Abnormal	Ground glass opacity	14	46.7
		Pericardial effusion	4	13.5
		Reticulonodal pattern	2	6.5
Echo findings	Normal		15	50
	Abnormal	Pulmonary hypertension	12	40
		Diastolic dysfunction	3	10
HRCT findings	Normal		12	40
	Abnormal	USIP	13	43.3
		NSIP	5	16.7
PFT findings		FVC-N/FEV1-N	12	40
		FVC<P value/FEV1 -N	18	60

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DISCUSSION

The present study aimed to identify and evaluate the early detection of cardio-pulmonary manifestations in patients with systemic sclerosis (SSc) attending Coimbatore Medical College Hospital. The study cohort consisted predominantly of female patients (83%), which aligns with the known higher prevalence of systemic sclerosis among women. This gender disparity is well-documented in literature, suggesting that hormonal and genetic factors may contribute to the higher susceptibility in females. The age distribution of participants showed that the majority were between 51-60 years old (36.5%), followed by the 31-40 age group (26.8%). These findings are consistent with the typical age of onset for systemic sclerosis, which often occurs in middle age[8]. Most participants had been living with systemic sclerosis for 1-4 years (67%). This duration is significant as it represents a critical window for early intervention. Early stages of the disease are crucial for implementing strategies to prevent or minimize severe organ involvement, particularly cardio-pulmonary complications that significantly impact morbidity and mortality[9].

ECG findings were normal in 90% of the patients, suggesting that gross cardiac electrical abnormalities are relatively uncommon in the early stages of systemic sclerosis. However, 6.7% of the participants exhibited left bundle branch block (LBBB), and 3.3% had supraventricular tachycardia (SVT). LBBB in systemic sclerosis patients can indicate underlying myocardial fibrosis or ischemic heart disease, while SVT could be related to autonomic dysfunction or myocardial involvement. These abnormalities, though not frequent, underscore the need for routine cardiac monitoring in systemic sclerosis patients[10].

Chest X-rays revealed that 33.3% of participants had normal results, while the majority exhibited various abnormalities. Ground-glass opacity was the most prevalent abnormality (46.7%), indicating the presence of interstitial lung disease (ILD). Pericardial effusion was observed in 13.5% of the participants, a complication that can range from asymptomatic to causing significant hemodynamic compromise. The presence of a reticulonodular pattern in 6.5% of participants further suggests fibrotic changes within the lung parenchyma, characteristic of advanced ILD[11].

Echocardiographic evaluation showed that 50% of the participants had normal findings, indicating that half of the patients did not have overt cardiac involvement detectable by echocardiography. However, a substantial proportion of patients (40%) had pulmonary hypertension (PH), which is a critical and often devastating complication of systemic sclerosis. Pulmonary hypertension results from vascular remodeling and increased pulmonary vascular resistance and is a major cause of morbidity and mortality in these patients[12]. Additionally, 10% of the participants had diastolic dysfunction, reflecting myocardial stiffness and impaired relaxation, likely due to myocardial fibrosis.

HRCT findings indicated that 40% of the participants had normal scans, while 43.3% had usual interstitial pneumonia (UIP) and 16.7% had non-specific interstitial pneumonia (NSIP). UIP is characterized by a more aggressive course and poorer prognosis compared to NSIP, which often has a better response to treatment. The high prevalence of UIP among the study participants highlights the aggressive nature of interstitial lung involvement in systemic sclerosis and underscores the necessity for early and precise imaging to guide management[13].

Pulmonary function tests showed that 60% of participants had reduced forced vital capacity (FVC) with normal forced expiratory volume in one second (FEV1), indicative of restrictive lung disease. This finding correlates with the high incidence of interstitial lung disease detected on HRCT. Reduced FVC is a hallmark of pulmonary fibrosis, which impairs lung compliance and gas exchange, leading to progressive dyspnea and decreased exercise tolerance[14].

The findings from this study have significant clinical implications. Early detection of cardio-pulmonary involvement in systemic sclerosis is crucial for improving patient outcomes. Routine

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screening using ECG, chest X-ray, echocardiography, HRCT, and PFTs should be an integral part of the management protocol for systemic sclerosis patients[15]. These investigations help in identifying subclinical disease, allowing for timely intervention and potentially altering the disease course.

Pulmonary Arterial Hypertension (PAH): Given the high prevalence of pulmonary hypertension found in this study, regular echocardiographic screening for PAH is recommended. Early treatment with vasodilators and other targeted therapies can significantly improve outcomes.

Interstitial Lung Disease (ILD): The detection of ILD, particularly UIP, necessitates early and aggressive management with immunosuppressive agents, antifibrotic drugs, and careful monitoring to slow disease progression and maintain lung function.

Cardiac Involvement: The presence of ECG abnormalities and echocardiographic evidence of diastolic dysfunction and pericardial effusion highlight the importance of cardiac evaluation in systemic sclerosis. Early identification of myocardial involvement can prompt the initiation of appropriate treatments to prevent further cardiac complications.

This study has several limitations. The sample size was relatively small, which may limit the generalizability of the findings. Additionally, the study was conducted at a single center, which may introduce selection bias. Larger, multicenter studies are needed to validate these findings and provide a more comprehensive understanding of cardio-pulmonary involvement in systemic sclerosis.

CONCLUSION

The study highlights the high prevalence of cardio-pulmonary manifestations in systemic sclerosis and the critical need for early detection and intervention. Early and aggressive management can improve survival rates and enhance the quality of life for patients with systemic sclerosis. By prioritizing early diagnosis and tailored therapeutic strategies, clinicians can better address the multifaceted challenges posed by this complex disease.

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