ORIGINAL ARTICLE

Prevalence of Cardiac Manifestations in Connective Tissue Disorders (Systemic Lupus Erythematosus, Rheumatoid Arthritis, Systemic Sclerosis)

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ABSTRACT

Introduction: Connective tissue diseases comprises several immunologic systemic disorders. The cardiological manifestations frequently occur in these patients. So the identification of the cardiological manifestations is a major concern.

Aim: To find out the prevalence of different cardiac manifestations in most common connective tissue disorders (systemic lupus erythematosus, Rheumatoid arthritis, Systemic sclerosis).

Methodology: A Hospital based cross-sectional, observational study conducted on 63 diagnosed cases of connective tissue diseases, of which 42 are SLE, 13 RA and 8 SSc. All these patients underwent clinical evaluation followed by ECG and 2D Echocardiography.

Results: Mean age of presentation was 49 and female: male ratio was 28:5. ECG changes were present in 23.8% patients (38.5% in RA, 21.4% in SLE, 12.5% in SSc). 2D Echocardiography showed non rheumatic valvular heart disease in 54% of total patients (52.4% of SLE, 46.2% of RA and 75% of SSc patients). Left ventricular diastolic dysfunction was present in 25.4% of total patients (31% of SLE). Pericardial involvement in term of pericardial effusion was seen in 23.8% of total patients (30.8% in RA, 23.8% in SLE, 12.5% in SSc). Pulmonary artery hypertension was found in 11.1% patients (23.1% in RA, 7.1% in SLE). Mean LVEF in respective connective tissue disease is 60% in RA, 59% in SLE, 64% in SSc.

Conclusion: Regarding cardiological system, most of the patients in our study developed non rheumatic valvular heart disease. Pericardial involvement and pulmonary artery hypertension occurs in patients with connective tissue diseases but are not very common. Valvular involvement and left ventricular diastolic dysfunction are dominant cardiological manifestations in our study.

Key word: SLE, SSc, RA, Pulmonary Hypertension.

INTRODUCTION

The term "connective tissue disease" (CTD) is defined as a heterogeneous group of disorders, some hereditary, others acquired, characterized by abnormal structure or function of one or more of the components of connective tissue, i.e., collagen, elastin, (and cells) or the mucopolysaccharides. But most Commonly this term is limited to a group of autoimmune disorders that are classified among the systemic rheumatic diseases and include rheumatoid arthritis (RA), systemic lupus erythematosus (SLE), systemic sclerosis (SSc), polymyositis-dermatomyositis (PM-DM), primary Sjögren's syndrome (SS), primary antiphospholipid syndrome (APS), and mixed connective tissue disease (MCTD).[1] Among the above mentioned CTDs RA, SLE and SSc are more frequently found all over the world as well as in India. The cardiological manifestations are common in patients with connective tissue disorders (CTDs). All the elements of the cardiological system are more or less affected. This includes the endocardium, myocardium, pericardiam, pulmonary vasculature and the conduction system of the heart. The frequency and type of cardiac involvement in connective tissue disorders varies based on the underlying disease. Although most of the cardiac complications appear after some time of diagnosis of CTDs, in some situations, cardiac involvement may be there preceding the more typical manifestations of the underlying disease.

Cardiac manifestations of SLE are diverse and can involve any aspect of the heart.[1,2] Pericarditis will develop in as many as 54% of patients with SLE.[3] Pericardial effusions, if present, are typically small and do not cause hemodynamic problems.[4] Myocarditis is the most characteristic feature of myocardial in-

volvement in SLEseen in as many as 8% cases that can progress to ventricular dysfunction, dilated CM and heart failure.[5] Lupus associated sterile endocarditis, also known as Libman-Sacks endocarditics is found in 11% -74% cases.[6]

The most common cardiac involvement in RA is pericarditis, occurring in 30-50% of patients, mainly in patients with severely destructive and nodular RA.[7] RA-associated cardiomyopathy may be the result of focal non-specific, diffuse necrotizing or granulomatous myocarditis which may be found in 3–30% of RA patients.[7] ventricular tachyarrhythmias were seen as well as QT-dispersion and corrected QT-dispersion intervals were significantly longer in RA patients.[9] The most prevalent valve disease in RA is mitral valve insufficiency, varying from 30-80% followed by aortic valve insufficiency varying from 9-33%.[8]

Cardiac involvement is found in most patients with systemic sclerosis. A 'mosaic', 'patchy' distribution of myocardial fibrosis is a pathognomonic feature of the disease.[10] Arrhythmias and conduction abnormalities are also seen as a result of conduction system fibrosis.[11] Among the conduction defects, Left bundle branch block and right bundle branch block with left anterior fascicular block were associated with abnormal left ventricular diastolic function. Supraventricular arrhythmias are more common in SSc, occurring in nearly 66% of the cases.[12] Pericardial abnormalities may manifest as fibrinous or fibrous pericarditis, with pericardial effusions detected in up to 41% of patients, but rarely causing tamponade.[13]

Hence, the present study was undertaken to evaluate the various cardiac manifestations in various CTDs.

METHODOLOGY

The present study is a hospital based cross sectional, observational study conducted at Medical College And Hospital, Kolkata from April 2018 to March 2019. Total 63 patients, who were diagnosed to be suffering from any of the following three connective tissue diseases – SLE, RA and SSc , are selected from patients attending Medicine and Rheumatology outpatient department. Cases were diagnosed by relevant clinical and biochemical parameters of SLE, RA and SSc and were included in the study. Patients suffering from ischemic heart disease, hypertension, congenital autoimmune CTDs, cardiac involvement due to other disease and patients with any severe chronic co-morbidities are excluded from the study. After meeting the inclusion and exclusion criteria, and consents being taken from the patients, ECG and 2D echocardiography were done for the diagnosis of the cardiological manifestations in these cases. Presence of valvular involvement, left ventricular diastolic dysfunction (LVDD), left ventricular ejection fraction(LVFE%), Pericardial involvement and Pulmonary arterial hypertension(PAH) were looked for during 2D echocardiography.

All patients were observed over a period of one year for clinical and cardiological evaluation.

RESULTS

Basic characteristics of the patients are given in table 1 and 2. Out of all the patients, majority of patients are having SLE (42 in number, 66.7%), followed by patients having RA (13 in number, 20.7%) and then SSc (8 in number, 12.6%). In our study patients age ranged from 15 to 50 years, which were divided into four groups. In our study most of the patients were in the age group of 35y to 44y, around 42.8% patients belonged to this age group followed by 17 patients (26.9%) in age group of 15y to 24y, mean age being 49 years. We analyzed the sex distribution of our study group where we observed that majority of the patients belong to female (total 53 patients, 84%), where as male patients are only of 15%. We also analyzed the sex distribution of patients according to different diseases and that 88% of all SLE patients, 77% of all RA patients and 75% of all SSc patients were female and rest were male. It was observed that connective tissue diseases are more common in females than males.

Table 1: Demographic characteristics (sex) of the patients

Disease	Total patients (%)	Male	Female
RA	13 (20.7)	3	10
SLE	42 (66.7)	5	37
SSc	8 (12.6)	2	6
Total	63	10	53

SLE- Systemic Lupus Erythematosus; RA- Rheumatoid Arthritis; SSc- Systemic Sclerosis

Table 2: Age distribution of the patients studied

Age in Years	Total patients	SLE	RA	SSc
15-24	17	10	5	2
25-34	10	6	4	0
35-44	27	19	3	5
>45	9	7	1	1
Total	63	42	13	8

SLE- Systemic Lupus Erythematosus; RA- Rheumatoid Arthritis; SSc- Systemic Sclerosis

Table 3: ECG changes in patients studied

ECG Changes	RA	SLE	SSC	Total
Absent	8	33	7	48
Present	5	9	1	15
Total	13	42	8	63

SLE- Systemic Lupus Erythematosus; RA- Rheumatoid Arthritis; SSc- Systemic Sclerosis

Table 4: distribution of non-rheumatic valvular heart disease

Nonrheumatic Valvular Disease	RA	SLE	SSC	Total
Absent	7	20	2	29
Present	6	22	6	34
Total	13	42	8	63

SLE- Systemic Lupus Erythematosus; RA- Rheumatoid Arthritis; SSc- Systemic Sclerosis

Table 5: Distribution of LV morphology changes in different CTDs

Lv Morphology	RA	SLE	SSC	Total
No Rwma	10	29	8	47
LVDD	3	13	0	16
Total	13	42	8	63

SLE- Systemic Lupus Erythematosus; RA- Rheumatoid Arthritis; SSc- Systemic Sclerosis; RWMA- Regional Wall Motion Abnormality; LVDD- Left Ventricular Diatolic Dysfunction

Table 6: Distributon of pericardial involvement in different CTDs

Pericardial Involvement	RA	SLE	SSC	Total
No Effusion	9	32	7	48
Pericardial Effusion	4	10	1	15
Total	13	42	8	50

SLE- Systemic Lupus Erythematosus; RA- Rheumatoid Arthritis; SSc- Systemic Sclerosis

Table 7: Distribution of pulmonary artery hypertension in CTDs

PAH	RA	SLE	SSc	Total
Absent	10	39	7	56
Present	3	3	1	7
Total	13	42	8	63

PAH: Pulmonary Artery Hypertension; SLE- Systemic Lupus Erythematosus; RA- Rheumatoid Arthritis; SSc- Systemic Sclerosis

23.8% of all the patients were having ECG changes. 38.5% of all RA patients had ECG changes, while 21.4% of all SLE patients and 12.5% of all SSc patients were having ECG changes (Table 3). It was seen that ECG changes are more common in RA patients than SLE and SSc patients. Studies show recent-onset SLE exhibited repolarization abnormalities, although severe abnormalities were rare.[14] Left ventricular hypertrophy, left axis deviation was also seen.[15] It is also found that arrhythmias and conduction defects are frequent manifestations of cardiac involvement in patients with SSc.[16] These abnormalities may be mild, but can also lead to a fatal outcome. Myocardial damage and fibrosis is main culprit for arrythmogenesis.

In our study, about 54% of patients have non rheumatic valvular heart disease. Among all, valvular involvement is more common in SSc 75%. In SLE and RA, there is more or less equal involvement with 52.4% in SLE and 46.2% in RA (Table 4). Studies show that non rheumatic heart disease is more common in RA(MR>AR).[17] It is found that non rheumatic heart disease (MR) is also more common in SLE but mild and asymptomatic.[18] Non rheumatic valvular heart disease is less common in SSc.[19]

Left ventricular diastolic dysfunction (LVDD) is less common in our study i.e 16(25.4%). Among all, LVDD not found in SSc and more common in SLE 31%(13 patients out of 42) in our study.(Table5) Studies show left ventricular morphology changes in youger patients in SLE.[20] and SSc does not seem to cause primary diastolic abnormalities.[21] Few study shows that LVDD is common in RA and is associated with duration and severity of the disease.[22]

Pericardial involvement in term of pericardial effusion is also not very common in our study i.e 23.8%(15 patients out of 63) but if present , it more commonly seen in RA patients i.e 30.8%. It is less common in SLE (23.8%) and SSc (12.5%). (Table 6) Study stated that and pericardial effusion may be the first presenting feature but taponade is rare in SSc. But if tamponade occurs, it indicates poor outcome as it is often associated with severe PAH.[23] Large pericardial effusion is rare in SLE.[24] Minimal pericardial effusion is common in RA.[25]

Pulmonary arterial hypertension (PAH) seen in 11.1% patients. It is more common in RA (23.1%) and less common in SLE (7.1%). PAH in RA can be attributed to interstitial lung disease, vascular disease and chronic thromboembolic disease[26]. PAH is 2nd most common in SLE after SSc[27] Pulmonary hypertension (PAH) is a frequent and severe complication of systemic sclerosis (prevalence 10%) [28].

DISCUSSION

A majority of the patients in the present study, 66.7% had SLE while RA contributed to 20.7% and SSc 12.6% of the cases.. Females are more commonly affected by the diseases than men. In our study female to male ratio was 28:5. Cardiological symptoms in patients ranged from asymptomatic to exertional dyspnea and palpitation.

In SLE, cardiac involvement was mostly in form of non rheumatic valvular heart disease, seen in 52.4% patients. LVDD was more common in SLE 31%(13 patients out of 42) in our study. Pericardial involvement in term of pericardial effusion was seen in 23.8% patients with SLE. No severe impairment of left ventricular ejection fraction was found. Pulmonary arterial hypertension (PAH) seen in 7.1% patients of in SLE. ECG changes were found in 21.4% SLE cases.

The most common cardiac involvement in RA In

our study, was non rheumatic valvular heart disease, which was found in 46.2% of RA cases. LVDD was seen in 23.6% cases of RA, whereas Pericardial effusion was discovered in 30.8% of RA patients. Our study shows no severe impairment of left ventricular ejection fraction in RA with mean LVEF being >55%. Pulmonary arterial hypertension (PAH) was seen in 23.1% of RA. ECG changes were found in 38.5% of RA patients.

In systemic sclerosis, commonest form of cardiac involvement in our study was non rheumatic valvular heart disease, seen in 75% of SSc patients (6 out of 8). LVDD was not found in SSc in our study and no impairment of LVEF was seen. Pericardial effusion was also not common in SSc , only 12.6% had mild pericardial effusion. Also only 12.5% SSc patients had pulmonary artery hypertension. ECG changes were seen in 12.5% patients.

CONCLUSION

In summary, the cardiac involvements are common in the CTDs included in our study, cardiac involvement may remain asymptomatic or manifest at some point in the course of the disease, where it can cause deterioration of quality of life as well as poor prognosis of disease. So, early recognition and introduction of the therapy will help in preventing the morbidity and mortality in such cases.

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