ORIGINAL RESEARCH ARTICLE
pISSN 0976 3325 | eISSN 2229 6816
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www.njcmindia.org

A study of Pulmonary Manifestations in Collagen Vascular Diseases

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Financial Support: None declared
Conflict of Interest: None declared

How to cite this article:
Vadgama P, Garg P. A study of Pulmonary Manifestations in Collagen Vascular Diseases. Natl J Community Med 2018; 9 (12): 889-892

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Date of Submission: 30-01-18
Date of Acceptance: 27-11-18
Date of Publication: 31-12-18

ABSTRACT

Introduction: Collagen vascular diseases are group of diseases with specific characteristics. Pulmonary manifestations are commonly present in collagen Vascular Diseases. The research was conducted to study pulmonary manifestations of collagen vascular diseases clinically; to study the demographic incidence in patients with pulmonary manifestations in collagen vascular diseases; to study radiological manifestations in terms of Chest X-Ray & HRCT Thorax; and also to study pulmonary functions tests in collagen vascular diseases.

Material & methods: This is a retrospective study on 50 patients of already diagnosed patient collagen vascular diseases.

Results: Pulmonary manifestations are present in 40% of patients had scleroderma (SC), 30% of cases of Systemic Lupus Erythematosis (SLE), 20% of patients of Rheumatoid Arthritis while 10% of Patients with Mixed Connective Tissue Disorders. Restrictive pattern is common in all types of collagen vascular diseases on pulmonary function testing. 24% of patients of collagen vascular disease had pulmonary arterial hypertension. On HRCT reticulonodular pattern is more common.

Conclusion: Pulmonary manifestations are commonly present in collagen vascular diseases. Broad choice of investigations should be done to prevent early mortality & morbidity.

Keywords: Collagen vascular diseases, HRCT, Pulmonary Arterial Hypertension

INTRODUCTION:

Collagen vascular diseases are a group of diseases with specific autoimmune characteristics that affects several organs including lungs.[1,2] Collagen Vascular diseases involves whole of the connective tissue system. They originate from degeneration of collagen from ground substance, muscle, fibrin & other plasma proteins. Almost all the collagen disorders can either controlled or symptomatically relieved. They require persistent & lifelong monitoring as they are progressive or can undergo remissions. The collagen disorders have varied systemic symptoms. Pulmonary abnormalities in patients with Collagen Vascular Diseases may not be due to underlying disease but may be because of its treatment[1,2 3]. HRCT (High Resolution Computed Tomography) thorax is the diagnostic tool for evaluating the histopathology type & lung abnormalities of collagen vascular diseases[3,4,5,6]. Interstitial lung diseases & pulmonary arterial hypertension are main causes of mortality & morbidity among patients with collagen vascular diseases[2].

In present study patients who were already having collagen vascular diseases and attended chest/ medicine OPD for their complaints were examined clinically & needful investigations were carried out

for the same. A variety of pulmonary manifestations were found in those patients. The analysis was done to gain general knowledge to understand the disease presentations for better treatment.

MATERIAL & METHODS

This is a retrospective study of total 50 Patients. Patients of diagnosed collagen vascular diseases that had respiratory symptoms came to pulmonary medicine OPD or Medicine OPD were taken in this study. Detailed analysis of their respiratory symptoms, High Resolution Computed Tomography (HRCT) reports, pulmonary functions tests (PFT) & 2-D Echo was taken into account.

All cases with confirmed diagnosis of collagen vascular diseases and were having respiratory symptoms were included in the study. Subjects who do not have respiratory symptoms were excluded.

RESULTS

Present study is a retrospective analysis of 50 patients of collagen vascular diseases. In present study 40% of patients had scleroderma (SC), 30% of cases had Systemic Lupus Erythematosis (SLE), 20% of patients had Rheumatoid Arthritis while 10% of Patients had Mixed Connective Tissue Disorders (Table 1).

Table 1 Incidence of etiology of Collagen Vascular Diseases

Collagen Vascular Diseases	Cases (%)
Scleroderma	20 (40)
Systemic Lupus Erythematosus	15 (30)
Rheumatoid Arthritis	10 (20)
Mixed Connective Tissue Disorders	5 (10)
Dermatomyositis/Polymyositis	-
Sjogrens Syndrome	-
Ankylosing Spondylitis	-

Table 2: Demographic distribution of Collagen Vascular Diseases

Age Group	SC (n=20)	SLE (n=15)	RA (n=10)	MCTD (n=5)	Total (n=50)
0-10	-	-	-	-	0
11-20	1 (5%)	4 (2.66%)	1 (10%)	-	6 (12%)
21-30	7 (35%)	7 (46.66%)	1 (10%)	-	15 (30%)
31-40	8 (40%)	4 (26.66%)	2 (20%)	2 (40%)	16 (32%)
41-50	4 (20%)	-	5 (50%)	2 (40%)	11 (22%)
51-60	-	-	1 (10%)	1 (20%)	2 (4%)
>60	-	-	-	-	-
Mean Age	32.65±6.81	28.33±7.41	37.1±7.99	43.2±7.72	

Table 3: Respiratory signs & symptoms

Symptoms & Signs	SC (n=20)	SLE (n=15)	RA (n=10)	MCTD (n=5)	Total (n=50)
Breathlessness on Exertion	15 (75)	3 (20)	5 (50)	2 (40)	25 (50)
Dry Cough	7 (35)	3 (20)	1 (10)	2 (40)	11 (22)
Cough With Expectoration	3 (15)	2 (13.33)	6 (60)	3 (60)	12 (24)
Fatigue	12 (60)	7 (46.66)	2 (20)	4 (80)	25 (50)
Fever	-	2 (13.33)	4 (40)	2 (40)	8 (16)
Chest Pain	2 (10)	4 (26.66)	2 (20)	1 (20)	9 (18)
Haemoptysis	-	1 (6.66)	2 (20)	1 (20)	4 (8)
Clubbing	5 (25)	-	4 (40)	2 (40)	11 (22)
Crackles	10 (50)	-	3 (30)	3 (60)	16 (32)

Table 4: HRCT Manifestations in Collagen Vascular Diseases

Type of Opacity on HRCT Thorax	SC (n=20)	SLE (n=15)	RA (n=10)	MCTD (n=5)	Total (n=50)
Reticular	2 (10)	5 (33.33)	1 (10)	1 (20)	9 (18)
Nodular	4 (20)	1 (6.66)	-	-	5 (10)
Reticulonodular	10 (50)	1 (6.66)	3 (30)	2 (40)	16 (32)
Pleural effusion	-	6 (40)	4 (40)	1 (20)	11 (22)
Honey combing	4 (20)	2 (13.33)	2 (20)	1 (20)	9 (18)

Table 5: Pulmonary Arterial Hypertension in Collagen Vascular Diseases

	SC (n=20)	SLE (n=15)	RA (n=10)	MCTD (n=5)	Total (n=50)
Normal	13 (65)	13 (86.66)	8 (80)	1 (20)	38 (76)
Mild PAH (RVSP 30-50)	1 (5)	1 (6.66)	-	-	2 (4)
Moderate PAH (RVSP 51-75)	3 (15)	1 (6.66)	1 (10)	1 (20)	6 (12)
Severe PAH (RVSP ≥75)	3 (15)	-	1 (10)	3 (60)	7 (14)
Total Patients of PAH	7 (35)	2 (13.33)	2 (20)	4 (80)	12 (24)

Table: 6 Pulmonary Function Test in Collagen Vascular Diseases

Abnormality	SC (n=20)	SLE (n=15)	RA (n=10)	MCTD (n=5)	Total (n=50)
Obstructive Pattern	1 (5)		2 (20)	1 (20)	4 (8)
Restrictive Pattern	15 (75)	12 (80)	6 (60)	3 (60)	36 (72)
Combined Pattern	2 (10)	2 (13.33)	1 (10)	1 (20)	6 (12)
Small Airways Disease	2 (10)	-	1 (10)	-	3 (6)

The demographic characteristics of patients are given in Table 2. Mean age of patients in SC was 32.65±6.81, in SLE was 28.33%±7.40, in RA & MCTD was 37.1±7.99 & 43.2±7.72 respectively. Most of the patients were in age group of 31-40 years (32%). In present study female preponderance was there. 80% of total patients were female.

Table 3 depicts various respiratory manifestations of collagen vascular diseases. Overall, most common chest symptom in patients of collagen vascular diseases was breathlessness on exertion & fatigue which was 50% of all the symptoms. In Scleroderma breathlessness on exertion was present in 75% of cases, fatigue in 60% and dry cough was present in 35% of cases.

In SLE fatigue was the most common complaint in 46.66% of cases and chest pain was present in 26.66% of cases. 60% Cases with RA had cough with expectoration and 50% presented with breathlessness on exertion. In 80% MCTD presented with fatigue as main complaint followed by cough with expectoration (60%).

Table 4 shows various findings on HRCT Thorax. Reticulonodular pattern was observed in most of cases. 32% of all patients of collagen vascular diseases had reticulonodular pattern on HRCT Thorax. 50% patients of SC & 40% of patients of MCTD had reticular pattern. Pleural effusion was common finding in RA & SLE , 40% patients each of them had pleural effusion. Honeycombing was observed in almost all cases of collagen vascular diseases (SC ,RA, MCTD each 20% & SLE 13%). Pleural effusion was exudative in nature in all cases that had pleural effusion.

Pulmonary arterial hypertension was observed in 24% of patients. Pulmonary arterial hypertension was more common in scleroderma being 35% of patients with 15% each had moderate & severe variety. PAH is less common in SLE (14%) of patients (Table 5).

72% of all cases of collagen vascular diseases had restriction pattern on spirometry, out of them 38% of patients had severe restriction of pulmonary function test. Restrictive pattern was most commonly seen in scleroderma (75%) & SLE (80%). One patient of SLE could not perform PFT Test. 8% of patients had obstructive pattern & 6% had small airway disease while 12% had combined pattern on PFT. (Table 6)

DISCUSSION

In present study 40% of patients of collagen vascular diseases who presented with pulmonary manifestations had SC, 30% had SLE while 20% of patients had RA & 10% had MCTD. In study by Merhdad et al incidence of SLE & RA was 30.3%, SC was 32.15 while incidence of MCTD & polymyositis was 5.3% & 1.7% respectively[7].

Cough was also presenting symptoms in 22% of patients which is in accordance with study done by G.S. Gaude who reported dyspnoea & cough as most common presenting symptoms in collagen vascular diseases[8].

Participants show that in SC, RA & SLE reticulonodular pattern was common finding on HRCT Thorax. Similar findings were also reported by GS Gaude study in which 78 patients out of 195 had reticulonodular pattern was present on HRCT Thorax[5]. 40% of patients of RA had pleural effusion. Pleural effusion was also commonly observed in SLE (40%). Mayberry & colleagues founded that pleural involvement was most common in RA[9].

80% of patients of collagen vascular diseases had restrictive pattern on PFT. 20% of patients of RA had obstructive pattern. G C Kane et al reported 50% patients of diffuse systemic sclerosis had restrictive pattern of PFT[10]. In study group secondary PAH was more common (80%) of patients of MCTD, 35% of patients of Sc & 20% of patients of RA had PAH while 13.33% of SLE patients had PAH. In one study pulmonary arterial hypertension is more common in patients with progressive systemic sclerosis (10%-33%) & MCTD. It is 5% - 10% in SLE & rare in RA[11]. In study by Condliffe R et al prevalence of PAH was 10% in SLE patients[12].

CONCLUSION

Collagen vascular diseases presents with variety of pulmonary manifestations and share common manifestations with other chronic respiratory diseases. Great knowledge, high suspicion & broad choice of investigations are needed to diagnose and treat collagen vascular diseases.

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