ORIGINAL RESEARCH PAPER

Volume-8 | Issue-6 | June-2019 | PRINT ISSN No. 2277 - 8179

INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

THE PROFILE OF PULMONARY INVOLVEMENT IN COLLAGEN VASCULAR DISEASES : A HOSPITAL BASED STUDY IN A TERTIARY CARE CENTRE

Pulmonary Medicine

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ABSTRACT

BACKGROUND: Collagen vascular diseases are a heterogeneous group of autoimmune disorders of unknown etiology. The pulmonary involvement in collagen vascular diseases varies according to the type of underlying disease.

OBJECTIVE: To study the clinical and radiological profile of pulmonary involvement in collagen vascular diseases.

MATERIALS AND METHODS: his is a cross sectional study which included all cases of collagen vascular disease attending the Pulmonary Medicine OPD with respiratory symptoms during thestudy period.

RESULTS : Dyspnoea is the predominant symptom in all types of collagen vascular diseases.(96.5%).The most common chest X ray finding is reticulonodular shadows (43.0%) . Interstitial(inter and intralobular)septal thickening is the commonest HRCT finding.(83.1 %).

CONCLUSIONS: Thisstudy shows that the pattern of pulmonary involvement in each type of collagen vascular disease is different. It is important to understand the differences in clinical and radiological features of each disease which greatly influences the treatment decision.

KEYWORDS

Collagen Vascular Diseases, Pulmonary Involvement, Clinical Profile, Radiological Profile

INTRODUCTION

Collagen vascular diseases are a heterogenous group of autoimmune disorders of unknown etiology. Many collagen vascular diseases involve the lungs directly or as a complication of treatment of the collagen vascular disease.

Collagen vascular diseasesinclude :SLE, Rheumatoid arthritis, Progressive systemic sclerosis / Scleroderma,Dermatomyositis& Polymyositis ,Ankylosing spondylitis ,Sjogrenssyndrome&MCTD .[12]

.ILD is the most common pulmonary complication of collagen vascular diseases & its incidence is increasing. Early detection of pulmonary involvement is very important for the initiation of targeted therapy.

Systemic sclerosis (SSc) is a connective tissue disorder of unknown etiology.Diffuse cutaneous SSc is associated with progressive skin induration.Patients who have limited cutaneous SSc coexisting with features of SLE, polymyositis, and rheumatoid arthritis may have mixed connective tissue disease (MCTD). [2]

Rheumatoid arthritis (RA) is a chronic inflammatory disease of unknown etiology marked by a symmetric, peripheral polyarthritis. Pleural disease is the most common thoracic manifestation and is seen much more frequently in men. [10,12] Interstitial lung disease,pleuritis, and occasionally obliterative bronchiolitis may be the first and only manifestation of the rheumatoid state in upto 20 percent of patients.[1]

Pleurisy and pleural effusion are the most common primary pulmonary complications of SLE, occurring in 50 to 80 percent of patients.[1]Acutelupus pneumonitis,community acquired pneumonias, diffuse alveolar haemorrhage and interstitial lung disease are the other pulmonary manifestations.

Polymyositis is a systemic autoimmune disorder characterized by an inflammatory myopathy. Aspiration pneumonia is a common pulmonary complication,occurring in 10 to 20 percent of patients .[1,21]Interstitial fibrosis occurs in 5%–30% of patients and appears as a fine reticular pattern that progresses to a coarse reticulonodular pattern and honeycombing [13].

Sj¨ogren's syndrome refers to a triad of xerophthalmia, xerostomia,andpolyarthritis.Obliterativebronchiolitis,constrictivebronchiolitis,ILD and bronchiolectasis have been reported.[14]

In a study of chest radiographic findings in 2,080 patients with ankylosing spondylitis, 26 (1.2%) had fibrosis in the upper lobes .[15]The most common abnormalities are peripheral interstitial lung disease, bronchiectasis, paraseptal emphysema, and apical fibrosis.[16]

METHOD

OBJECTIVE:

To study the clinical and radiological profile of pulmonary involvement in patients with collagen vascular diseases such as Systemic Lupus Erythematosus, Rheumatoid arthritis, Progressive systemic sclerosis / Scleroderma, Dermatomyositis & Polymyositis, Ankylosing spondylitis, Sjogrens syndrome, Mixed Connective Tissue Disease,attending the Pulmonary Medicine OPD, Medical college, Thiruvananthapuram during the time period from February 2012 to November,2012.

STUDY DESIGN: Cross sectional study

STUDY POPULATION : All diagnosed cases of collagen vascular disease attending the Pulmonary Medicine OPD , Medical college , Thiruvananthapuram .

STUDY SETTING : Department of Pulmonary Medicine, Medical college, Thiruvananthapuram

STUDY PERIOD : February 2012 to November 2012

INCLUSION CRITERIA : All cases of collagen vascular diseases of age group above 12 yearsattending the Pulmonary Medicine OPD ,Medical College, Thiruvananthapuram .

EXCLUSION CRITERIA : Collagen vascular disease patients who are not willing to participate in the study.

METHODOLOGY : All cases of collagen vascular disease attending the Pulmonary Medicine OPD during the time period February 2012 to November 2012 with respiratory symptoms were involved in the study. Diagnosis of collagen vascular diseases was based on the diagnostic criteria..The following tests were done for the diagnosis of pulmonary manifestations in all cases taken up for the study:Chest radiograph,spirometry,electrocardiogram, echocardiography,HRCT Chest. The clinical and radiological profile of these patients was studied which included Age, Sex, symptoms, duration of symptoms, Clinical examination,Chest X Ray findings,Spirometry,ECG,HRCT chest findings ,Echocardiography- presence of pulmonary hypertension

RESULTS

A total of 86 patients with collagen vascular diseases were enrolled in this study.Systemic sclerosis constituted 47.7% of patients ;Rheumatoid arthritis34.9%;MCTD 8.1%;SLE 5.8%;PM/DM 2.3% and Sjogrens syndrome 1.3%.These patients were divided into 3

groups namely Rheumatoid arthritis,Systemic sclerosis and Others and further analysis done.

GENERAL CHARACTERISTICS

MEAN AGE OF THE PATIENTS IN EACH GROUP:

In Rheumatoid arthritis mean age of patients is 54.27.In Systemic sclerosis it is 38.49 and in Others 33.2.There is a significant difference (P-<0.001,F-22.487)

MEAN AGE OF ONSET OF COLLAGEN VASCULAR DISEASE :

For Rheumatoid arthritis 50.40. For Systemic sclerosis 35.89.For Others 31.45.There is a significant difference(P<0.001,F-18.923)

SEX DISTRIBUTION OF PATIENTS IN EACH GROUP:

Majority of patients were females(86%) .73.3% of patients with Rheumatoid arthritis,902% of those with Systemic sclerosis and all patients in Others group were females.

DISTRIBUTION OF SYMPTOMS :

In Rheumatoid arthritis group all patients had cough and dyspnea,53% had expectoration,13.3% chestpain and 3.3% had hemoptysis.In systemic sclerosis group 95.1% complained of dyspnea, 51.2% of cough,29.3% expectorationand 9.8% chestpain.In others 93.3% had dyspnea and 40% had cough, 26.7% had chestpain.

DISTRIBUTION OF PHYSICAL SIGNS :

In Rheumatoid arthritis group 3.3% had pallor, 53.3% had clubbing, 13.3% had pedal edema,90% had crackles and 20% had rhonchi on auscultation. In systemic sclerosis group 2.4% had pallor,24.4% had clubbing,17.1% had pedal edema,87.8% had crackles on auscultation. In others group 13…3% had pallor,20% had clubbing,13.3% had pedal edema,73.3% had crackles and 6.7% had rhonchi.

CHEST X-RAY FINDINGS

Table 1

CHEST X-RAY FINDINGS	DIAGNOSIS						
	RA		SS		OTHERS		P Value
	NO	%	No	%	NO	%	
RETICULAR	4	13.7	17	41.5	3	20	
NODULAR	0	0	0	0	0	0	
RETICULO NODULAR	14	46.7	15	36.6	8	53.3	0.471
CYSTIC SPACES	5	16.7	0	0	1	6.7	0.025
PLEURAL EFFUSION	1	3.3	1	2.4	2	13.3	0.088
PNEUMOTHORAX	0	0	0	0	1	6.7	
CONSOLIDATION	2	6.7	0	0	2	13.3	0.09

DISTRIBUTION OF LESIONS IN CHEST X-RAY

In rheumatoid arthritis group 16.7% was normal,6.7% had unilateral involvement and 76.7% had bilateral involvement. In systemic sclerosis group 22% was normal, 4.9% had unilateral involvement and 73.2% had bilateral involvement.In others group 13.3% had unilateral and 86.7 % had bilateral changes.

ZONES INVOLVED IN EACH GROUP

In Rheumatoid arthritis 26.7% had involvement of all zones,23.3% MZ andLZinvolvement ,23.3% had LZ involvement,6.7% had UZ and MZ involvement, 3.3% had MZ involvement In Systemic sclerosis 58.5% had LZ involvement ,22% had no involvement,12.2% had MZ and LZ involvementIn Others group 46.7% had LZ involvement ,33.3% had involvement of all zones , 20% had MZ and LZ involvement.

HRCTFINDINGS

In Rheumatoid arthritis interstitial (inter and intralobular) septal thickening was the predominant finding (66.7%)followed by honey combing (46.7%),bronchiectasis(40%),GGO(36.7%),Emphysema (10%),consolidation (6.7%),Nodular (3.3%) ,mediastinal LNE (2.67%),mass (1.33%).In systemic sclerosis the most common finding was interstitial septal thickening (95.1%) followed by GGO (46.3%),honey combing (46.3%),bronchiectasis (3.9%),emphysema (4.9%).In others interstitial septal thickening was seen in 83.3%,GGO in 41.7%,honey combing (33.3%),bronchiectasis in 8.3%,emphysema (8.3%),consolidation (8.3%)

DISTRIBUTION OF HRCT FINDINGS

LOBEWISE DISTRIBUTION : In Rheumatoid arthritis majority (36.7%)had lower lobe involvement,26.7% had involvement of all lobes,13.3% had upper and lower lobe and 10% had middle and lower lobe involvement. In systemic sclerosis majority of patients (75.6 %) had lower lobe involvement. In Others also lower lobe was the most involved (41.7%).

HRCT PATTERN

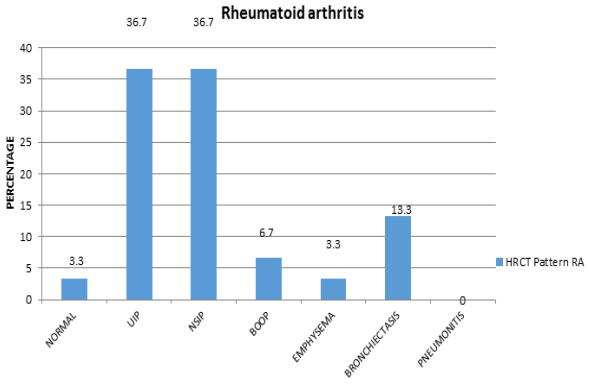

Figure 1

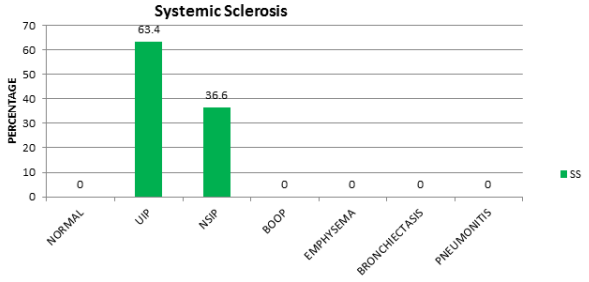

Figure 2

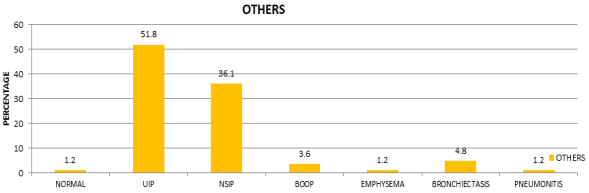

Figure3

PULMONARY HYPERTENSION

In Rheumatoid arthritis 3.3%, in Systemic sclerosis 22% ,in Others 16.7% had pulmonary hypertension (χ2- 4.911 ,df-2 ,P 0.086)

DISCUSSION

The study is a cross sectional study. A total of 86 patients with collagen vascular diseases were enrolled in this study. The ,patients were divided into three groups. Systemic sclerosis and Rheumatoid arthritis were the two predominant groups out of the three groups. Others comprised of only a small number each of the other collagen vascular diseases.

High-resolution computed tomography (CT) has been shown to be superior to radiography in the assessment of the presence and extent of parenchymal, airway, and pleural abnormalities. [10]A survey combining the results of several studies estimates that the sensitivity of HRCT for ILD is 94% compared with 80% for chest radiography. [17]HRCT has a valuable role in confirming or refuting the presence of ILD, particularly given the low false positive rate for HRCT. [18]

Rheumatoid arthritis (RA) is the most common of the collagen vascular diseases to be associated with interstitial lung disease. Systemic sclerosis (SSc) is much less prevalent than rheumatoid arthritis but is more commonly complicated by interstitial lung disease . [3]

The histological entities of the ATS/ERS classification for Idiopathic Interstitial Pneumonias (IIPs) can all be associated with the collagen vascular diseases.[11]

In the only large study of lung histology in systemic sclerosis there was a high prevalence of Non Specific Interstitial Pneumonia (NSIP) (62/80, 75%) and a very low prevalence of Usual Interstitial Pneumonia (UIP) (6/80, 8%), [3,4] a finding that is broadly consistent with other smaller series. [5,6]Recent data suggest that NSIP and UIP subgroups make up similar proportions of patients with rheumatoid lung. [22] The spectrum of HRCT changes also suggests that NSIP and UIP-type patterns occur with similar frequency,[8,9]Bronchiolitis obliterans or obliterative bronchiolitis is a well-recognized cause of progressive and often severe obstructive lung disease in patients with rheumatoid arthritis.[7]With the possible exception of rheumatoid arthritis, NSIP is the predominant histological diagnosis in collagen vascular diseases .

In this study interstitial lung disease is the most common manifestation obtained in all the three groups studied. All the systemic sclerosis patients studied presented with interstitial lung disease . Usual Interstitial Pneumonia (UIP) (63.4%) is the predominant HRCT pattern obtained in systemic sclerosis patients with interstitial lung disease followed by Non Specific Interstitial Pneumonia (NSIP) (36.6%) which is not in agreement with the data available in literature. No other patterns were observed in systemic sclerosis in this study.

In Rheumatoid arthritis patients with interstitial lung disease both Usual Interstitial Pneumonia (UIP) (36.7%) and Non Specific Interstitial Pneumonia (NSIP) (36.7%) were equally present in HRCT and were the predominant findings in this group. Available data in radiology also supports this finding. Other patterns observed in Rheumatoid arthritis were Bronchiolitis obliterans Organising Pneumonia (BOOP) in 6.7% cases and bronchiectasis in 13.3% cases . In others also interstitial lung disease is the predominant manifestation in this study, with Usual Interstitial Pneumonia (UIP) being the commonest pattern (51.8%) and Non Specific Interstitial Pneumonia (NSIP) coming next (36.1%)

Outcome differs little between UIP and NSIP in SSc, even after adjustment for baseline disease severity.[4]Furthermore, UIP in SSc has a better outcome than IPF, perhaps reflecting a lower profusion of fibroblastic foci in UIP in CTD in general [23] or earlier diagnosis of the lung disease. As with SSc, however, UIP associated with RA appears to have a better outcome than the idiopathic form of the disease.

CONCLUSIONS

Majority of the patients studied were females. (86%)Dyspnoea is the predominant symptom in all types of collagen vascular diseases.(96.5%) .Clubbing is the most common finding on general examination (33.7%). Clubbing is more commonly seen in Rheumatoid arthritis compared to other collagen vascular diseases.(53.3%).The most common chest X ray finding isreticulonodular shadows (43.0%).

Interstitial thickening is the commonest HRCT finding.(83.1 %).The predominant HRCT pattern in Systemic sclerosis is UIP pattern (63.4%) Both UIP and NSIP patterns are equally seen in Rheumatoidarthritis(36.7%).Lower lobe is the most commonly involved lobe.(56.6%)

This study shows that the pattern of pulmonary involvement in each type of collagen vascular disease is different.It is found that a significant number of patients present with pulmonary symptoms prior to the onset of other systemic symptoms.Therefore it is important to understand the differences in clinical and radiological features of each disease which greatly influences the treatment decision.This signifies the role of pulmonologist in identifying more cases of collagen vascular diseases.

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