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Clinicoradiological Profile of Patients with Connective Tissue related Diffuse Parenchymal Lung Disease.

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ABSTRACT

Background: Diffuse Parenchymal Lung Disease (DPLD) is one of the most serious pulmonary complications associated with connective tissue diseases (CTDs), resulting in significant morbidity and mortality. Although the various CTDs associated with DPLD often are considered together because of their shared autoimmune nature, there are substantial differences in the clinical presentations and management of DPLD in each specific CTD. **Methods:** An observational cross-sectional study was carried out in a single tertiary care center in northern India which included 50 consecutive subjects (age >12 years) of DPLD and amongst them patients having connective tissue diseases associated DPLD (CTD)-DPLD were studied.In this study, the clinical, radiological data of the subjects was collected. **Results & Conclusion:** Out of 50 patients, 13 patients turned out to be CTD-DPLD. The most common of these was Rheumatoid arthritis (53.8%) followed by Systemic sclerosis (38.4%) and sjogresn syndrome (7.7%). The average age in CTD-DPLD came out to be 47.8 years as opposed to 49.04 in DPLD patients. The average age in RA was 58.6 years, Systemic sclerosis was 40 years and Sjogrens syndrome was 45 years. Majority (92.3%) were females.

Keywords: Lung disease Connective Tissue.

INTRODUCTION

Diffuse parenchymal lung disease (DPLD) refers to a large group of acute and chronic pulmonary diseases characterized by damage to the lung parenchyma, with varying patterns of fibrosis and/or inflammation.[1,2] These diseases also frequently affect the airspaces, peripheral airways, vasculature, and corresponding epithelial and endothelial surfaces even though the pulmonary interstitium (i.e. The space between the epithelial and basement membranes) is the primary site of the parenchymal damage establishing an accurate diagnosis of DPLD can be challenging for clinicians as these comprises of a heterogeneous group of more than one hundred distinct lung disorders that tend to be grouped together because they share clinical, radiographic, and pathologic features.[3,4]

Connective tissue diseases (CTDs) cause a myriad of pulmonary complications, including bronchiolitis and bronchiectasis, pleuritis, and pulmonary hypertension. Interstitial lung disease (DPLD) is a common and serious form of pulmonary involvement characterized by various patterns of inflammation and fibrosis on high-resolution CT (HRCT) scan and in lung biopsy specimen. Advances in the description of radiologic patterns and pathologic findings used in the idiopathic interstitial pneumonias are now being applied to patients with CTD.[5-8] The lung is a common site of complications of systemic connective tissue disease (CTD), and lung involvement can present in several ways. Although it is generally thought that interstitial lung disease develops later on in CTD it is often the initial presentation ("lung dominant" CTD). DPLD can be present in most types of CTD, including rheumatoid arthritis, scleroderma, systemic lupus erythematosus, polymyositis or dermatomyositis, Sjögren's syndrome, and mixed connective tissue disease.[9-13]

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MATERIALS AND METHODS

This study was carried out in the department of chest and tuberculosis, Government Medical College, Amritsar. This was an observational cross-sectional study in which 50 patients who had diffuse parenchymal lung diseases and came to outpatient department or were admitted in wards were studied and out of these patients having CTD-DPLD were

further study to see their clinical radiological profiles.

This study was conducted after approval from the institutional ethics committee, Govt. Medical College, Amritsar. Each patient was considered for the study after taking an informed consent.

Inclusion criteria

1. Patients with age 12 years and above, either sex having chief complaints of dyspnea & cough and bilateral diffuse shadows on chest radiography.

Exclusion criteria

1. Patients not consenting for the study.
2. Diffuse parenchymal lung diseases cases diagnosed to have tuberculosis.
3. Patients with cardiac disorders.
4. Patients with very severe hypoxemia.

A pre-structured proforma was filled in those cases which were included in the study. Their personal data including age, sex, occupation, address, a detailed occupational history were sought out. Their symptoms, past and personal history with special focus on smoking were noted down. This was followed by their physical examination, investigations including Chest x-rays, SpO2 with pulse oximetery. Spirometeric analysis,6 min walk test were done. The data was documented, tabulated and analyzed by using unpaired t-test and Chi-square test wherever applicable.

RESULTS

The patients diagnosed as CTD-DPLD based on clinical, radiological and PFT findings attending OPD and/or admitted in Chest and TB hospital, Amritsar were included in the study. They were studied according to their demographic features, clinical characteristics, and Radiological findings and the following observations were made which have been depicted in tabular form.

Out of 50 patients, 13 patients turned out to be CTD-DPLD. The most common of these was Rheumatoid arthritis (53.8%) followed by Systemic sclerosis (38.4%) and sjogresn syndrome (7.7%). These patients were further studied based on their clinical and radiological profiles and the results are as follows: the average age in CTD-DPLD came out to be 47.8 years as opposed to 49.04 in DPLD patients.The average age in RA was 58.6 years, Systemic sclerosis was 40 years and Sjogrens syndrome was 45 years. 92.3% were females.

Table 1: History of patients

Sex	
Male	1(7%)
Female	12(93%)
Occupation	
Labourer	1(7%)
Housewife	12(93%)
Age (years)	
Rheumatoid arthritis	58.6
Systemic sclerosis	40
Sjogren's syndrome	45

Table 2: Clinicoradiological profile

Signs and symptoms at presentation	
breathlessnes	13(100%)
cough	6(46.15%)
joint pains	6(46.15%)
skin thickening	5(38.5%%)
Spirometeric abnormality	
Restrictive	8(61.5%)
Obstructive	nil
Normal	5 (38.5%)
chest X-ray feature	
Reticular pattern	56%
Reticulonodular	22%
Nodular pattern	12%
Consolidation	10%
HRCT Pattern	
Honeycombing	50%
Ground Glass Opacity	30%
Septal Thickening	46%
Traction Bronchiectasis	56%
Consolidation	10%
Nodules	22%
Upper/Middle Lobe Predominant	8%
Lower Lobe Predominant	64%
Diffuse	28%

Table 3: Aetiological diagnosis

Aetiological diagnosis	
Rheumatoid arthritis	7(53.8%)
Systemic sclerosis	5 (38.5%)
Sjogren's syndrome	1(7%)

DISCUSSION

Despite similarities in clinical and pathologic presentation, the prognosis and treatment of CTD associated DPLD (CTD-DPLD) can differ greatly .Diffuse parenchymal lung diseases are heterogeneous group of diseases involving lung interstitium. They have features in common like similarities of symptoms, comparable radiographic appearances, consistent alterations in the pulmonary physiology and typical histological features. Reports from western literature show an increase in the prevalence and incidence of DPLD in recent decades. The present study was conducted in fifty cases of DPLD attending OPD and/or admitted in chest and TB hospital, Amritsar so as to to evaluate the clinical spectrum and radiological findings of DPLD which can aid in their diagnosis and early management.

Out of 50 patients, 13 patients turned out to be CTD-DPLD. The most common of these was Rheumatoid arthritis (53.8%) followed by Systemic sclerosis (38.4%) and sjogresn syndrome (7.7%). These patients were further studied based on their clinical and radiological profiles and the results are as follows: the average age in CTD-DPLD came out to be 47.8 years as opposed to 49.04 in DPLD patients. The average age in RA was 58.6 years, Systemic sclerosis was 40 years and Sjogrens syndrome was 45 years. Majority (92.3%) were females.

The common symptoms at presentation observed were breathlessness which was seen in all the

patients (100%), followed by cough (46.15%), (joint pains (46.15%), and thickening of skin in 38.5%.

Most common chest X-ray feature in our study group was reticular pattern which was present in 56%, followed by nodular pattern in 12% and consolidation in 10%. Reticulonodular pattern was seen in 22%.

The most common HRCT pattern in our study group was Traction Bronchiectasis seen in 56%, followed by Honeycombing seen in 50%, Septal Thickening in 46% and GGO 30%.

CONCLUSION

DPLD is now increasingly recognized as a frequent and serious complication of rheumatic diseases and CTDs. Early diagnosis of CTD-DPLD is of paramount importance to prevent/delay progression to irreversible damage especially as its a treatment responsive disease and most of these can be easily diagnosed on the basis of the clinical and the radiological features. Larger prospective epidemiological studies, increased education and awareness is required for a better understanding of their spectrum.

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