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Original Research Article

Diffuse Parenchymal Lung Diseases - Aetiology and Clinical Profile: A Prospective Study

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Abstract:

Background: A complex combination of diffuse parenchymal lung conditions, interstitial lung disease is characterized by restrictive physiology, decreased gas perfusion, inflammation of the lung parenchyma, and fibrosis. The pulmonary interstitium, which is made up of the connective tissue space between the alveolar epithelial cells and the nearby capillary endothelial cells, is primarily responsible for the pathophysiology of interstitial lung disease.

Methods: The study was conducted from July 2022 to December 2022 in the department of Respiratory Medicine at Varun Arjun Medical College, Banthra, and Shahjahanpur. Total 60 patients, 36 male and 24 female included in this study. The male female ratio 1.5:1.

Results: Exertional dyapoea was the most common presenting symptom; second most common symptom was nonproductive cough. COPD was the commonest comorbid illness, second commonest comorbid condition absorbed was mellitus. Out of the 36 men, 25 were Smokers (42%) of the 19 Patients with IPF 16 weresmokers. Average smoking index was found to be 180. 21.58% patients had upper zone predominance in chest x- ray. 4 out of 60 patients had mid zone perihilar distribution of lesion. 29 out of 60 patients had lower zone predominance. Spirometry showed restrictive in 86 % of Patients.

Conclusion: The profile of ILDs with their demographic, clinical and outcome data were analyzed and compared with other regional and global studies. The results recognized certain similarities and differences compared to other reports, formulating a distinctive study among others. Idiopathic interstitial pneumonias were the commonest type of ILD in studied sample, followed closely by secondary ILDs.

Keywords: Diffuse Parenchymal Lung Disease, Idiopathic Pulmonary Fibrosis, and connective tissue – interstitial lung disease.

Introduction

Diffuse parenchymal lung diseases (DPLD) comprises of a wide spectrum of lung disease characterized by extensive parenchymal infiltrates. By radiological definition, the term diffuse means widely distributed not necessarily involving the entire lung but bilateral infiltrates. Lung parenchyma is the tissue distal to the terminal bronchioles.

The incidence of DPLD ranges from 3 to 26 lakhs per year. The clinical presentation can be acute or chronic. In acute settings aetiological diagnosis is a clinical puzzle and the clinician is forced to give empirical therapy under high index of suspicion. In such situations aetiological diagnosis can be often lifesaving.

This study is an attempt to identify the aetiologies of DPLD by clinical and radiological means supported by some relevant noninvasive tests obviating the need for potentially risky procedures like lung biopsy.

Since Hamman Rich fist described progressive pulmonary fibrosis, there has been variable degree of lung fibrosis that is commonly referred to as ILD. DPLD will be the more appropriate terminology since the disease often involves the whole lung parenchyma including the airways and blood vessels as well.

DPLD is a syndrome with the following common features i.e. exertional dyspnea, bilateral diffuse CXR infiltrates, nonproductive cough restrictive ventilatory defect, low diffusion capacity (DLCO) and variable degree of fibrosis and inflammation in the lung histopathology. ILD includes a heteroge-

neous group of disorders among which IPF is the prototype.

Causes

1. Occupational and environmental- asbestos, silica, beryllium etc
2. Organic dusts, microorganisms like fungus, actinomyces, animal protein
3. Collagen diseases
4. Inherited disorders like tuberous sclerosis, neurofibromatosis etc
5. Infections : Vasculitis, granulamatoses, Sturge Strauss, PAN, Wegeners Granulomatosis
6. Toxic fumes like vapours, chlorine, fluorine etc
7. Drugs: cytotoxins penicillamine gold etc
8. Poisons like paraquat, toxic oil syndrome, radiation
9. Unknown aetiology like cryptogenic fibrosising alveolitis, sarcoidosis, langerhans cell histiocytosis, amyloidosis

Idiopathic pulmonary fibrosis

IPF is an inflammatory lung disease of unknown origin. This disease typically affects people above 60 yrs; symptoms worsen over a period of months to years leading to end stage lung fibrosis and death. This carries 30% 5yr mortality. There are two histological types:-1.Usual interstitial pneumonia (UIP) 2.Desquamating interstitial pneumonia (DIP)

Investigations

- Chest X-ray patterns-Reticular, Nodular, Reticulonodular, Ground glass.
- HRCT Early stages-Ground glass, honey combing predominantly basilar and sub pleural (late stage).
- SPIROMETRY- Restrictive pattern.
- DLCO-Most sensitive test to diagnose early ILD.
- 6 Minute walk test-A fall of 4% of SpO2 from base line is usual.
- OTHER TESTS-The above investigations are common to all DPLDS. Tests other than these are clinically directed and depend on which disease is under consideration. Open lung biopsy is the gold standard for diagnosis of IPF.

Indications for lung biopsy in DPLD

This includes relatively young age, h/o fever, family h/o ILD, pneumothorax, symptoms and signs of peripheral vasculitis, atypical radiographic features and rapidly progressive disease.

Materials and Methods

Total 60 patients were studied. The study was conducted from July 2022 to December 2022 in the department of Respiratory Medicine at Varun Arjun Medical College, Banthra, and Shahjahanpur.

Inclusion criteria

All patients of age above 18yrs with CXR and HRCT findings consistent with DPLD attending the department of respiratory medicine were include in the study.

Exclusion criteria

- Patients with cardiogenic pulmonary edema.
- Corona infected patients.

Investigations

- CXR, HRCT DLCO, oximetry done in all patients studied.
- Blood tests like ANA, anti DSDNA, ANCA. etc done according to clinical indications.
- Sputum examination in relevant cases.
- Fiber optic bronchoscopy -Done in 2 patient, one had pulmonary alveolar proteinosis, second patient had sarcoidosis.

Lung biopsy

Two patients had undergone lung biopsy. One had alveolar protenosis (transbronchial biopsy) and the other one was diagnosed as IPF (open lung biopsy).

Observations

- Total no. of patients= 60.
- Male= 36.
- Females =24.
- Male Female ratio 1.5:1.

Clinical Profile

Exertional dyapoea was the most common presenting symptom, second most common symptom was nonproductive cough.

Table 1:

Symptoms	Number	Percentage
Dyspnoea	47	78.33%
Cough	13	21.67%
Chest Pain	14	36.0%
Fever	8	13.33%
Joint Symptoms	16	26.67%
Body ache	6	10.0%

Table 2: Clinical signs

Category	Number	Percentage
Clubbing	24	40.0%
Cyanosis	7	11.62%
Bibasal crackles	52	87.0%

Co-morbidity: COPD was the commonest comorbid illness, second commonest comorbid condition absorbed was mellitus.

Table 3:

Disease	Number	Percentage
COPD	7	11.67%
CAD	3	5.0%
Hypertension	4	6.67%
Diabetes	6	9.96%
HIV	4	6.67%

Smoking Status: Out of the 36 men, 25 were Smokers (42%) of the 19 Patients with IPF 16 weresmokers. Average smoking index was found to be 180.

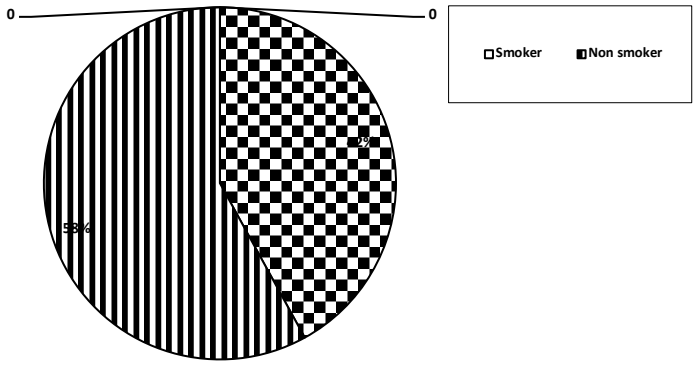

Figure 1:

Chest X - Ray

Upper zone predominance

- 21.58% Patients had upper zone predominance in chest x- ray
- Sarcoidosis 3
- Silicosis 2
- Extrinsic Allergic Alveolitis 7
- Berylliosis 1

Mid zone predominance

- 4 out of 60 patients had mid zone perihilar distribution of lesion
- Pulmonary alveolar proteinosis 1
- Pneumocystis Carinii pneumonia 3

Lower zone predominance

- 29 out of 60 patients had lower zone predominance
- IPF 19
- RA 5
- SLE 2
- Scleroderma 1
- Polymyositis 1
- Drug induced 1

Non – specific Pattern

- Observed in 23.4% of Patients
- TB 7
- Dengue fever 1
- Laptospirosis 2
- Malignancy 2

Spirometry Pattern: Spirometry showed restrictive in 86 % of Patients.

Table 4:

Pattern	Number	Percentage
Restrictive	33	86.80%
Mild	-	-
Moderate	13	39.4%
Severe	20	60.6%
mixed	8	13.1%

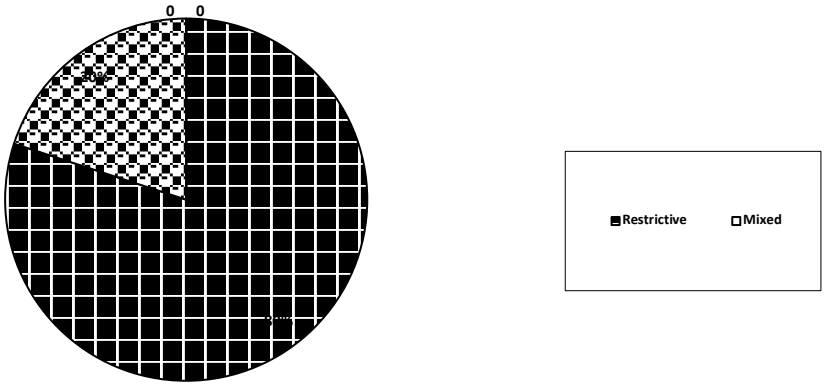

Figure 2:

Table 5: HRCT Pattern

Pattern	Number	Percentage
Ground Glass	5	11.36%
Reticulonodular	32	72.73%
Honey Combing	7	15.91%
Total (CT done)	44	100%

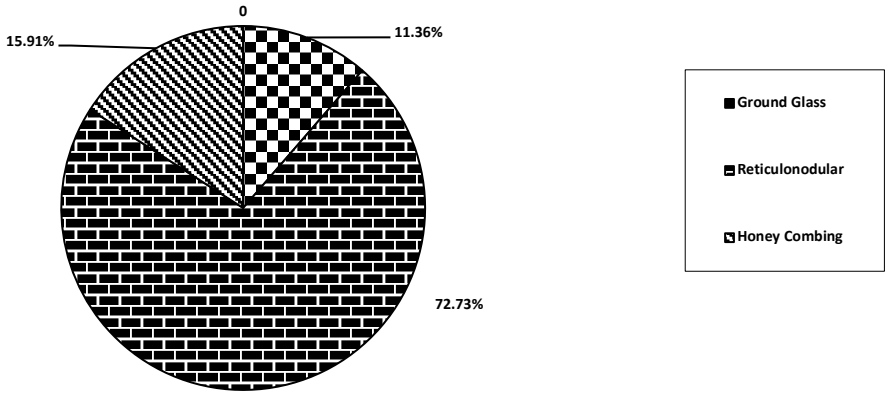

Figure 3:

Etiology: Exact aetiological diagnosis could be arrived at in the cases of 41 out of 60 (68.33%) patients. The remaining 31.67% was due to IPF.

Table 6:

Disease Entity	No. of cases	Percentage
IPF	19	31.67%
Infections	12	20%
CTD	9	14.99%
EAA	7	11.67%
Occupational LD	3	5%
Sarcoidosis	3	5%
BOOP	2	3.33%
Malignancy	2	3.31%
Others	3	5%
Total	60	100%

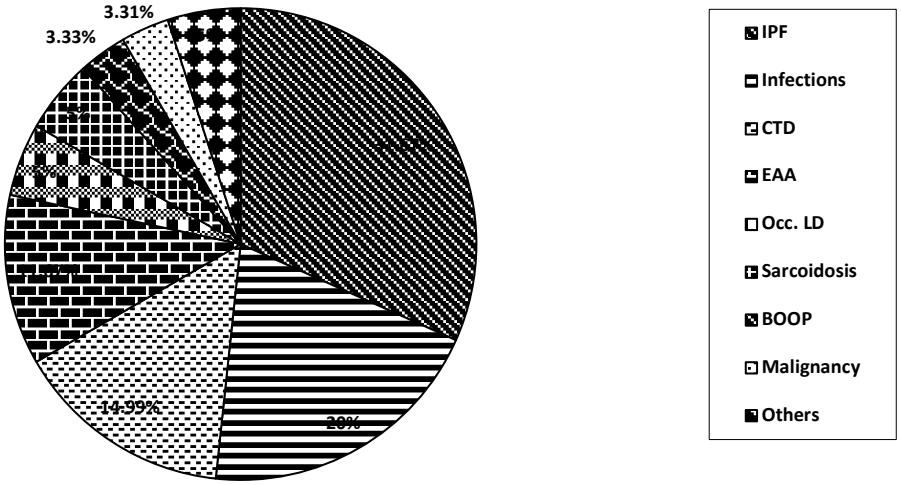

Figure 4:

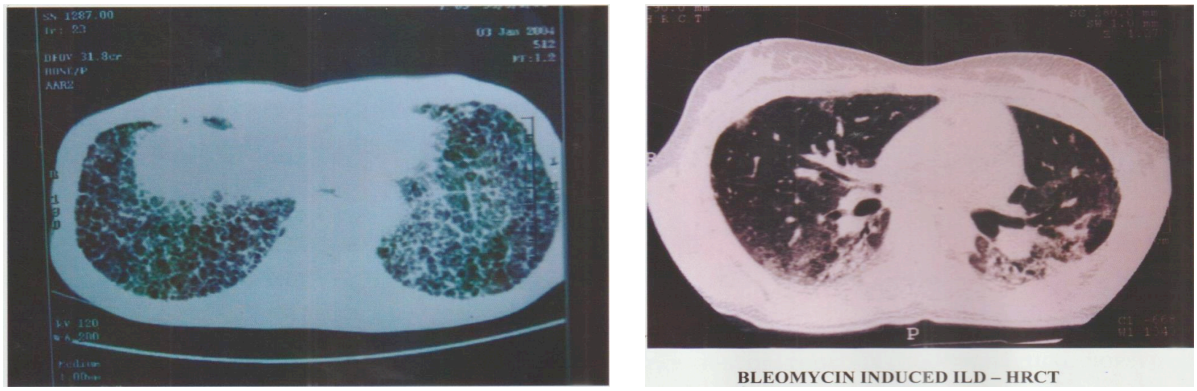

Figure 5: IPF - Honey Combing in HRCT

Discussion

Albeight a wide variety of aetiologies, DPLDS share some common clinical features. Almost all the patients had exertional dyspnea plus dry cough or both as presenting symptoms.3 Patients with IPF had sputum productions. But all of them were heavy smokers with age above 60 yrs. HRCT has revolutionized the aetiological diagnosis of DPLD. In experienced hands HRCT is like histopathology in this regard.

31 patients in the study were diagnosed as IPF the proportion of IPF in the study is comparable to the earlier study conducted by Gaenslor and Corrington (34%). Most common cause of DPLD differs in different parts of the globe. But the incidence of IPF is almost similar worldwide. The first published report of ILD in India came in the year 1997 (Jindel SK et al). According to ATS ERS criteria the minimum symptoms duration for diagnosis of IPF is 3months.Clubbing was reported in 79% IPF in this study. (As per earlier studies 25 to 30 %). Higher proportion of clubbing in our population might be attributable to higher smoking score and smoking related pulmonary diseases. The second most common aetiology in our study is infection study by Ganslor and Corrinmgton shows granulomatous disease as common second cause.

In many states of north India hypersensitive pneumonitis remains the leading cause. Many infections have ILD like presentation. Infections comprise 12 out of 60 cases here. 4 patients had pulmonary TB among which 2 were sputum AFB positive 3RD common aetiology was connective tissue disorders. Five out of nine had Rh. arthritis.

Two women were diagnosed as SLE, Sarcoidosis detected in 3women average age was 38 yrs. Lung biopsy was considered as the gold standard for aetiological diagnosis of DPLD. But lung biopsy is an invasive procedure and it carries an element of risk to the patient. The study here is an attempt to arrive at causal diagnosis in DPLD without lung biopsy.

Conclusion

- Aetiological diagnosis could be done in 69% of DPLDS
- 31% of cases are due to IPF
- Infections and connective tissue disorders are the 2nd and 3rd causes respectively

- The average age of IPF is 59yrs and mean duration of symptoms 9months Exertional dyspnea is the most common presenting symptom followed by dry cough
- HRCT is the most useful noninvasive tool for diagnosing DPLD
- Early diagnosis could be made in 11patients (6.6%) only
- Lung even though taken as gold standard is not a mandatory investigation for DPLD

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