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TO STUDY CLINICORADIOLOGICAL PROFILE OF IDIOPATHIC PULMONARY FIBROSIS (IPF) PATIENTS

Respiratory Medicine

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ABSTRACT

Background- The term "interstitial lung disease" (ILD), or diffuse parenchymal lung disease (DPLD), represents a group of about 200 distinct disorders. Idiopathic pulmonary fibrosis (IPF) is one of the most severe forms of IIP. This study aims to observe clinicoradiological profile of idiopathic pulmonary fibrosis patients. **Methods-** A cross-sectional observational study (inpatients and outpatients) visiting the Department of Respiratory Medicine, Gandhi Medical College, Bhopal for the period of 18 months from November 2019 to April 2021, diagnosed to have Idiopathic Pulmonary Fibrosis (IPF) by HRCT who meet the inclusion and exclusion criteria. **Result-** The incidence of IPF is higher in males as compared to females. It appears mainly between the fifth and seventh decades of life ,the mean age of IPF was 58.86 ± 11.93, and the maximum number of patients came under the age of 51–60 years .In this study, breathlessness and cough were the predominant symptoms in definitive UIP. The severity of breathlessness is higher in definitive UIP. In our study every patient with IPF, we found to have an abnormal chest xray. Hence, chest xray should be the first investigation in any suspected case of IPF. The commonest chest radiographic abnormality in patients with IPF is peripheral reticular opacity, most marked at the bases. In this study, the most common HRCT Chest findings in IPF patients were reticulations, subpleural and basal involvement, and fibrosis. **Conclusion-** In this study, the most common pattern of idiopathic pulmonary fibrosis was definitive UIP and most common age group was 51–60 years old and IPF reveals a male predominance. Clubbing was found in 39.4% of patient's maximum with definitive UIP (28.8%). On auscultation, the most common findings are fine crackles and the most common findings on HRCT are reticulations So, the final diagnosis of IPF is a multidisciplinary approach. Further study is needed with a larger sample size to understand the disease in detail.

KEYWORDS

Pulmonary Fibrosis, Clinicoradiological, Clubbing

INTRODUCTION-

The term "interstitial lung disease" (ILD), or diffuse parenchymal lung disease (DPLD), represents a group of about 200 distinct disorders that encompass clinical disorders that involve the alveolar structures, pulmonary interstitium, and small airways. [1]

Idiopathic pulmonary fibrosis (IPF) is one of the most severe forms of IIP. Idiopathic pulmonary fibrosis, also known as cryptogenic fibrosing alveolitis, is a chronic, progressive fibrosis characterized by an irreversible decline in lung function, progressive respiratory failure, and high mortality. It is characterized by progressive worsening of dyspnea and lung function and is associated with a poor prognosis. [2]

The incidence of IPF is about 10.7 per 100,000 people for males and 7.4 per 100,000 people for females. The prevalence of IPF is slightly higher at 20.2 males per 100,000 and 13.2 females per 100,000. IPF is not associated with any particular race, ethnic group, or environment. Cigarette smoking is strongly associated with IPF and most accepted risk factor for IPF, and it increases the risk of IPF by about 2 times. The incidence of IPF is higher in males as compared to females. It appears mainly between the fifth and seventh decades of life, and about 33% of patients' age is more than 60 years.

The mean age at the time of presentation was 66 years old. IPF can occur in young people and rarely in children. Familial IPF accounts for 0.5 to 2% of all cases of IPF. [3]

Patients with IPF can present with dry cough, progressive dyspnea, and bibasilar late inspiratory fine crepitation (Velcro crepts) on auscultation, which is characteristic of idiopathic pulmonary fibrosis. The patient may also have clubbing, but it is not necessary in all cases of IPF.

A typical interstitial pneumonia pattern on high-resolution CT is distinguished by reticular opacities, which are frequently associated with traction bronchiectasis, and little or no ground-glass opacifications. Honeycombing, which appears as subpleural, clustered cystic airspaces with well-defined walls (typically 3–10 mm in diameter), is common and essential for making a diagnosis. [4]

AIMS & OBJECTIVES -

To study clinicoradiological profiles of IPF patients.

MATERIALAND METHODS-

A cross-sectional observational study (inpatients and outpatients) visiting the Department of Respiratory Medicine, Gandhi Medical College, Bhopal for the period of 18 months from November 2019 to April 2021, diagnosed to have Idiopathic Pulmonary Fibrosis (IPF) by HRCT who meet the inclusion and exclusion criteria.sample size was 104 . Institute's Ethics Committee (EC) clearance was obtained and a written informed consent was obtained from all patients.

METHODOLOGY-

We have a total of 250 patients with ILD, of which 104 were chosen for the study based on exclusion criteria and HRCT chest findings. Detailed demographical and clinical parameters including age, sex, height, weight, symptoms like dry cough and breathlessness, including mMRC grading, smoking history, history of ATT for similar illness, environmental, occupational, and, clubbing, end-inspiratory crackles, gastrointestinal reflux were assessed. Fever was defined as temperature more than 98.6 degree F (37.0 degree C). [5]Chest x-ray was performed in all cases and evaluated for parenchymal abnormality and HRCT of the thorax was evaluated for Reticulations, Honey combing, Fibrosis, subpleural and basal involvement, ground-glass opacities (GGO), Traction Bronchiectasis.

OBSERVATIONS AND RESULTS-

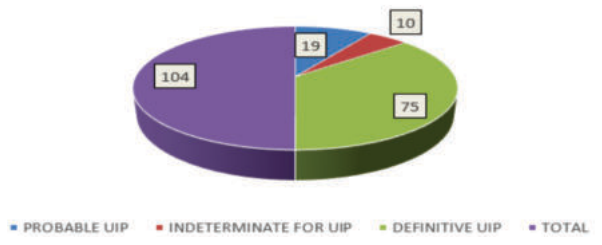
This study includes 104 participants, encountered during the 18 months of study (November 2019 to April 2021).The study was conducted in department of Respiratory Medicine Gandhi medical college Bhopal

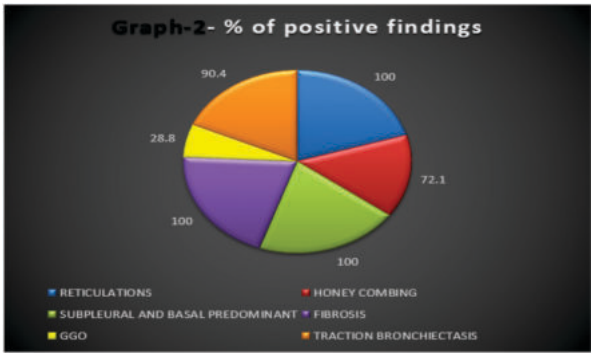
Table 1- Distribution of study participants on the basis of radiological diagnosis-

SL. NO.	DIAGNOSIS	FREQUENCY	PERCENTAGE
1	PROBABLE UIP	19	18.3
2	INDETERMINATE FOR UIP	10	9.6

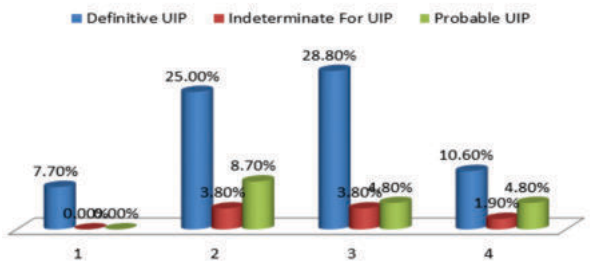
3	DEFINITIVE UIP	75	72.1
	TOTAL	104	100.0

Graph 1-Study participants on the basis of radiological diagnosis





Graph 3-Association Between mMRC Grading With Diagnosis



DISCUSSION-

Interstitial lung diseases (ILDs) are a group of parenchymal lung diseases that are heterogeneous. One of the most aggressive forms of IIP is idiopathic pulmonary fibrosis (IPF). The incidence of IPF is higher in males as compared to females. It appears mainly between the fifth and seventh decades of life, and about 33% of patients' age is more than 60 years. The average age at the time of presentation was 66 years old. IPF can present as dry cough, progressive dyspnea, and bibasilar late inspiratory fine crepitation (Velcro crepts) on auscultation, which is characteristic of idiopathic pulmonary fibrosis. The patient may also have clubbing, but it is not necessary in all cases of IPF.

AGE-

In our study, the mean age of IPF was 58.86 ± 11.93, and the maximum number of patients came under the age of 51–60 years, which is about 26.9% of all patients included in the study, followed by 61–70 years (26%). IPF is more frequent in people in their fifth and seventh decades of life, with two-thirds of all cases occurring in people over 60. The average age at presentation was 66. IPF is uncommon in those under the age of 40, and it rarely, if ever, affects children. IPF occurs in middle-aged and elderly adults (median age at diagnosis of 66 years, range 55–75 years). [6]

GENDER-

In our study,it was seen in 73 males out of a total of 104 patients, which was around 70.2% of all the study populations. A male preponderance was seen in IPF. Among 44 patients with Definitive IPF 32 (72.7%) were male. According to the ILD India Registry [7] IPF is more common in males. Among IPF alone, the male to female ratio is 2:1. It is in accordance with Kundu et al. [8] in which they found the male to female ratio was 4:3 and among western literature, a study done on the Danish population [9] found 77% of male IPF patients. However, Kumar et al. [10] found equal numbers of male and female patients in their study. Similarly, a study done by Sherbini et al. [11] found only 44% of males in their study.

Symptoms At Presentation-

In this study, breathlessness and cough were the predominant symptoms in IPF(definitive UIP). Dry cough was found in 66 (63.35%), 19 (16.3%), and 9 (8.7%) of people with definitive UIP, probable UIP, and indeterminate UIP.

mMRC grade I was found only in definitive UIP cases, accounting for 8 (7.7 percent) of all definitive UIP cases. mMRC grade II was observed in 26 (25 percent), 9 (8.7 percent), and 4 (3.8 percent) for definitive UIP, 91 probable UIP, and indeterminate UIP, respectively. mMRC grade III was observed in 30 (28.8%), 5 (4.8%), and 4 (3.8%) cases of definitive UIP, probable UIP, and indeterminate UIP, respectively. For definitive UIP, probable UIP, and indeterminate UIP, mMRC grade IV were found in 11 (10.6 percent), 5 (4.8 percent), and 2 (1.9 percent), respectively. The total number of patients with definitive UIP, probable UIP, and indeterminate UIP was 75 (72.1%), 19 (18.3%), and 10 (9.6%), respectively.

It is in accordance with Kundu et al[8] in which they found 100% of patients with cough and breathlessness at presentation. Also, Maheshwari et al. [12] found breathlessness (98.6%) and dry cough (92.1%) as the most common presenting symptoms in IPF. In earlier reports, the incidence of cough has ranged from 26% to 73%. The prevalence of dyspnea ranged from 26% to 100%. [13,14]

The severity of breathlessness is higher in definitive UIP as compared to probable and indeterminate UIP. The higher the grade, the greater the chances of having a UIP pattern.

Chest X Ray-

The association between different types of IPF and abnormal parenchyma in chest x ray was found to be significant. It is most common in IPF. In our study every patient with IPF, we found to have an abnormal chest xray. Hence, chest xray should be the first investigation in any suspected case of IPF. The commonest chest radiographic abnormality in patients with IPF is peripheral reticular opacity, most marked at the bases, Chest radiographs may occasionally be normal in patients with IPF.[15] In this study abnormal lung parenchyma in Chest x ray was found in 104(100%) of patients. Which is similar to study done by Kumar et al. [10] in which they found reticulonodular shadow in 88.75% patients.

HRCT Chest-

In this study, the most common findings in IPF patients were reticulations, subpleural and basal involvement, and fibrosis, which were found in 104 patients (100%). Followed by traction bronchiectasis found in 94 (90.4%) patients, honey combing in 75 (72.1%) patients, and ground glass opacity in 30 (28.8%) patients, which was a statistically significant P value (<0.05)

This is in accordance with ATS guidelines [2], which state that the most common finding in IPF is reticulation, followed by honeycombing, fibrosis, and traction bronchiectasis. Also, Kumar et al. [10] found the most common HRCT findings to be interstitial infiltrates in 56 (75.67%) patients, honeycombing in 23 (31.08%) patients, fibrosis in 37 (50.0%) patients, and traction bronchiectasis in 9 (12.16%) patients.

CONCLUSIONS-

In this study, the most common pattern of idiopathic pulmonary fibrosis was definitive UIP, followed by probable UIP, and then indeterminate UIP. In our study, the most common age group was 51–60 years old, accounting for approximately 26.9 percent of all patients, followed by 61–70 years old (26 percent). People in their fifth and seventh decades of life are more likely to have IPF, and IPF reveals a male predominance. The majority of IPF patients presented with a dry cough and shortness of breath, and many were diagnosed with definitive UIP and had a history of smoking and GERD.

Clubbing was found in 39.4% of patient's maximum with definitive UIP (28.8%). On auscultation, the most common findings are fine crackles, which were found in about (98) 94.2% of patients. All the patients have parenchymal abnormalities on chest Xray and the most common findings on HRCT are reticulations, reticulations with basal

predominance, and fibrosis, which are found in 100% of patients with IPF.

So, the final diagnosis of IPF is a multidisciplinary approach. Further study is needed with a larger sample size to understand the disease in detail.

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