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Clinical profile of idiopathic pulmonary fibrosis: A cross sectional study

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Abstract
Background: Idiopathic Pulmonary Fibrosis (IPF) is a chronic progressive disease that has a highly variable clinical course. IPF constitutes a subgroup of diffuse parenchymal lung diseases with distinct clinical, radiological and histopathological features. This study was undertaken to study the various signs and symptoms in the patients diagnosed with IPF on the basis of HRCT.
Objective: To study the clinical profile of patients with IPF.
Material and Method: This study was done on 60 patients who were diagnosed as IPF on the basis of HRCT chest findings. Each subject underwent complete physical examination and relevant investigations were done.
Result: Amongst 60 subjects, 16 were females and 44 were males. Most common age group was more than 60 years. Exertional breath lessness and dry cough were the most common symptoms. 15 patients were smokers and 25 had gastro esophageal reflux. 21-30 months is the most common duration of symptoms. Crackles are the commonest finding on physical examination.
Conclusion: Our study highlights the distribution of various signs and symptoms among the patients with IPF, which can help us in understanding the disease in a better manner which can contribute in its better management.

Keywords: idiopathic pulmonary fibrosis, HRCT, diffuse parenchymal lung disease

Introduction

The diffuse parenchymal lung diseases are a heterogenous group of inflammatory processes affecting the alveolar wall. About 15% of all cases in respiratory medicine comprises of diffuse parenchymal lung diseases.

The idiopathic interstitial pneumonias are a subset of diffuse parenchymal lung diseases of unknown aetiology characterized by expansion of the interstitial compartment with an infiltrate of inflammatory cells. In the updated classification, the major entities have been preserved and grouped into (a) "chronic fibrosing IIPs" (idiopathic pulmonary fibrosis [IPF] and idiopathic nonspecific interstitial pneumonia [NSIP]), *(b)* "smoking-related IIPs" (respiratory bronchiolitis–associated interstitial lung disease [RB-ILD] and desquamative interstitial pneumonia), and *(c)* "acute or subacute IIPs" (cryptogenic organizing pneumonia and acute interstitial pneumonia). In addition, idiopathic pleuroparenchymal fibroelastosis has been newly included and classified together with lymphoid interstitial pneumonia in a fourth group, "rare IIPs". In the updated classification, it has again been acknowledged that in a subset of cases, a final diagnosis cannot always be achieved despite extensive multidisciplinary discussion, and such cases are considered "unclassifiable" [1].

IPF is defined as a specific form of chronic, progressive fibrosing interstitial pneumonia of unknown cause, occurring primarily in older adults, limited to the lungs, and associated with the histopathologic and/or radiologic pattern of UIP [2, 3, 4]. The definition of IPF requires the exclusion of other forms of interstitial pneumonia including other idiopathic interstitial pneumonias and ILD associated with environmental exposure, medication, or systemic disease [2, 3].

IPF should be kept in mind in all patients presenting with unexplained chronic exertional breathlessness, cough, bibasilar inspiratory crackles and clubbing [5-7]. Incidence increases with age with sixth and seventh decades being most common [7-10]. More commonly reported in men and in cigarette smokers [5-8].

The incidence of IPF was estimated at 10.7 cases per 100,000 per year for men and 7.4 cases per 100,000 per year for women in a population-based study from the county of Bernalillo, New Mexico [11]. Prevalence estimates for IPF have varied from 2 to 29 cases per 100,000 in the general population [8].

Although idiopathic pulmonary fibrosis is, by definition, a disease of unknown etiology, a number of potential risk factors have been described. Cigarette smoking for example, is strongly associated with IPF. Also, environmental exposures like metal and wood dusts [12-14], stone cutting/polishing, microbial agents like Epstein Barr Virus [15, 16] and Hepatitis C [13, 17, 18], gastro esophageal reflux are found to be closely associated with IPF. Although accounting for less than 5% of total patients with IPF, familial forms of IPF (i.e., those affecting two or more members of the same primary biological family) have been reported [19-22].

HRCT is an essential component of the diagnostic pathway in IPF. UIP (Usual Interstitial Pneumonia) pattern in HRCT is characterized by the presence of reticular opacities, with

subpleural and basal predominance, often associated with traction bronchiectasis [23, 24]. Honeycombing is common, and is critical for making a definite diagnosis.

Symptoms had been present for more than 9 months in most patients. The median survival from the time of diagnosis varies between 2.5 - 3.5 years. Lung physiology and radiological abnormalities rarely improve.

This study was undertaken to study the various signs, symptoms and risk factors in the patients diagnosed with IPF on the basis of HRCT.

Objective

To study the clinical Profile of Idiopathic Pulmonary Fibrosis in a tertiary care centre.

Materials and Methods

The study was carried out in Department of Pulmonary Medicine, Lilavati Hospital & Research Centre, a tertiary health care centre located in Bandra West, Mumbai, Maharashtra. This is a cross sectional observational study, conducted over a period of 1 year from 2015 to 2016. 60 cases of IPF diagnosed on the basis of UIP pattern on HRCT, as per ATS/ERS guidelines were included in the study after taking informed consent from each patient.

Inclusion criteria

1. Diagnosed cases of IPF on the basis of HRCT pattern, both newly and previously diagnosed.
2. Age above 45 years.

Exclusion criteria

1. Age ≤ 45 years.
2. Patient with diagnosis of chronic lung diseases like asthma, bronchiectasis, COPD.
3. Hemodyamically unstable patients, bed ridden/stroke patients who cannot perform 6 minute walk test.
4. Patients with history of chest trauma/lung contusion/rib fracture.

Methodology

Patients coming to Lilavati Hospital, both out patient and in patient, who were diagnosed as a case of IPF after thorough clinical examination and on the basis of HRCT chest, excluding all other causes of UIP pattern on HRCT, were included in the study. Detailed history of such patients was taken, including onset and duration of symptoms, past and personal history to look for any risk factor, family history to rule out familial association. Thorough physical examination was done to check for all the significant signs. Six minute walk test was performed to see exercise tolerance. Arterial blood gas was done to test hypoxia at rest.

Results

A total of 60 patients who were diagnosed as cases of idiopathic pulmonary fibrosis on the basis of HRCT chest findings were studied for their clinical presentation, risk factors, exercise tolerance and hypoxia. Amongst 60 patients, 16 were female and 44 were male (Figure 1), with a male: female ratio of 2.75:1. Minimum and maximum age of the subjects were 50 and 78 years respectively. Most common age group was 60-69 years (Figure 2), which had 40 subjects (66.67%) with a mean age of 63.12 years.

Amongst subjects, history of smoking was found in 15 subjects (25%) and in 25 subjects (41.67%), a history of gastroesophageal reflux was found (Table 1). Most of the subjects (n=39) gave history of duration of their symptoms of 21-30 months (65%), with a mean duration of 24.43 months (Table 2).

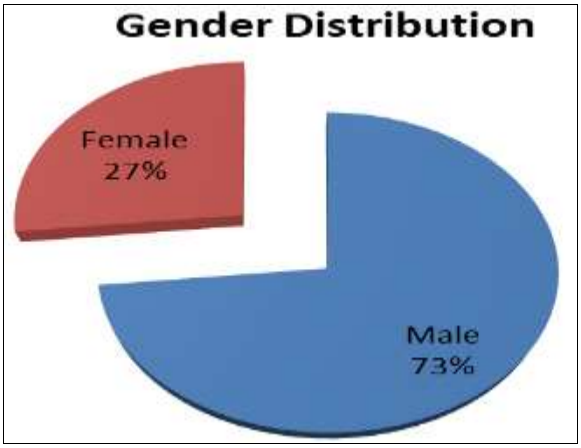

Fig 1: Distribution of gender

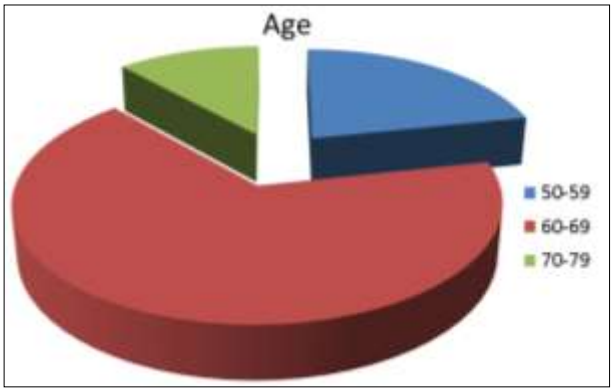

Fig 2: Distribution of Age

Table 1: Distribution of Risk Factors

Risk factors	No.	Percentage
Gerd	25	41.67%
Smoking	15	25%
Family H/O ILD	-	-

Table 2: Distribution of Duration of Symptoms

Duration of Symptoms (Months)	No.	Percentage
<10	01	01.67%
10-20	13	21.67%
21-30	39	65.00%
>30	07	11.67%
Total	60	100%

Most common symptoms were cough (n=51) and breathlessness (n=54), overall, 90% subjects had complaints of breathlessness and 85% had complaints of cough (Table 3 and Figure 3).

Crackles were found in 93.34% patients (n=56), while rhonchi

were found in just 11.67% patients (n=7). Significant desaturation was seen in 65% patients (n=39), clubbing in 70% (n=42) and hypoxia in 60% (n=36) subjects. These characterstics are shown in table 4 and figure 4

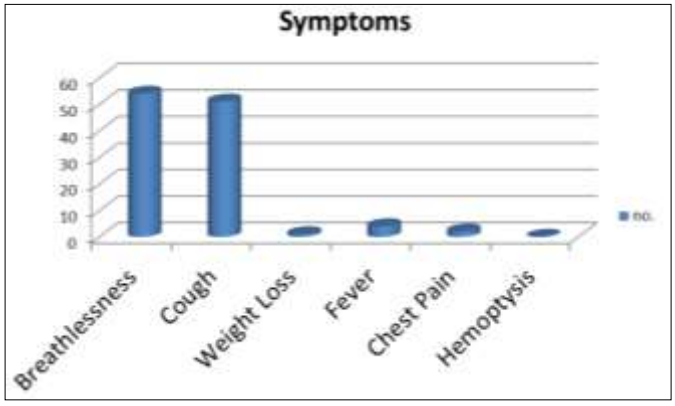

Fig 3: Distribution of symptoms

Table 3: Distribution of Symptoms

Symptoms	No.	Percentage
Breathlessness	54	90%
Cough	51	85%
Weight Loss	1	1.67%
Fever	4	6.67%
Chest Pain	2	3.33%
Hemoptysis	-	-

Table 4: Distribution of Signs

Signs	No.	Percentage
Crackles	56	93.34%
Rhonchi	7	11.67%
Desaturation on 6MWT	39	65%
Clubbing	42	70%
Hypoxia on ABG	36	60%

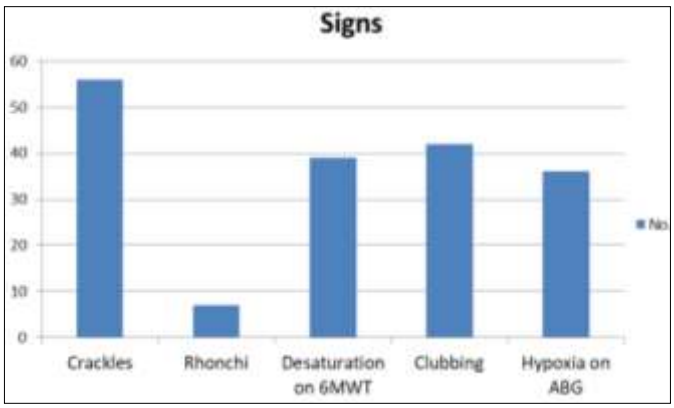

Fig 4: Distribution of signs

Discussion

Idiopathic pulmonary fibrosis is a progressive fibrosing inflammatory disease of the lungs of unknown etiology. It is approximately twice more common in males than females. Largest survey done reported the mean age in IPF patients being 67 years [26].

IPF is now a well known clinical entity rather than a diagnosis of exclusion. There is however very limited data on its clinical behavior from India. In a study done over 5 years period on patients with DPLD, Jindal Sk, *et al.* [27], IPF accounted for 46% of cases. In another Sharma SK, *et al.* [28], IPF was seen in 28.6% of 133 patients with DPLD.

Commonly seen in the middle aged patients, the incidence of IPF increases with advancing age. As per the reports from west, nearly 2/3rd patients are over 60 years at the time of diagnosis. In the present study, 47 patients (78.34%) were aged 60 years or more. Although 13 patients (21.67%) were <60 years of age.

Symptoms in IPF include insidious onset of cough (dry) and exertional breathlessness. In previous studies, the incidence of cough has ranged from 26%-73% and that of breathlessness from 26% - 100% [27, 29, 30]. Clubbing may be seen n 25%-50% and crackles in more than 80% patients [31, 32]. Overall prevelance of signs and symptoms has been found to be almost similar in our study.

HRCT has almost obviated the need of lung biopsy in patients with IPF. Characterstic findings on HRCT are often sufficient to diagnose IPF.

Conclusion

1. There is no gender difference in clinical presentation of IPF as compared to the western world.
2. Most common age at presentation in our study is more than 60 years which is comparable to the previously done studies.
3. Most common symptom in the patients with IPF is cough and exertional breathlessness, and the most common finding on physical examination is crackles.
4. History of gastroesophageal reflux and smoking are fairly common in patients diagnosed with IPF.
5. Patients with IPF commonly ignore their symptoms of breathlessness and cough at initial stages or are commonly wrongly diagnosed, which is apparent in the duration of symptoms before being diagnosed in our study.
6. Every patient attending the respiratory clinic with unexplained dry cough and exertional breathlessness, having crackles on auscultation must be thoroughly examined for IPF.

References

1. Travis WD, Costabel U, Hansell DM, *et al*. An official American Thoracic Society/European Respiratory Society statement: update of the international multidisciplinary classification of the idiopathic interstitial pneumonias. Am J Respir Crit Care Med. 2013; 188(6):733-748.
2. American Thoracic Society; European Respiratory Society. Idiopathic pulmonary fibrosis: diagnosis and treatment: international consensus statement. Am J Respir Crit Care Med. 2000; 161:646-664.
3. American Thoracic Society, European Respiratory Society. American Thoracic Society/European Respiratory Society International Multidisciplinary Consensus Classification of the Idiopathic Interstitial Pneumonias. Am J Respir Crit Care Med. 2002; 165:277-304.
4. Visscher DW, Myers JL. Histologic spectrum of

idiopathic interstitial pneumonias. Proc Am Thorac Soc. 2006; 3:322-329.
5. Douglas WW, Ryu JH, Schroeder DR. Idiopathic pulmonary fibrosis: Impact of oxygen and colchicine, prednisone, or no therapy on survival. Am J Respir Crit Care Med. 2000; 161:1172-1178.
6. King TE Jr, Tooze JA, Schwarz MI, Brown KR, Cherniack RM. Predicting survival in idiopathic pulmonary fibrosis: scoring system and survival model. Am J Respir Crit Care Med 2001; 164:1171-1181.
7. Gribbin J, Hubbard RB, Le Jeune I, Smith CJ, West J, Tata LJ. Incidence and mortality of idiopathic pulmonary fibrosis and sarcoidosis in the UK. Thorax. 2006; 61:980-985.
8. Scott J, Johnston I, Britton J. What causes cryptogenic fibrosing alveolitis? A case-control study of environmental exposure to dust. BMJ. 1990; 301:1015-1017.
9. Mannino DM, Etzel RA, Parrish RG. Pulmonary fibrosis deaths in the United States, an analysis of multiple-cause mortality data. Am J Respir Crit Care Med 1996; 153:1548-1552-1979-1991.
10. Raghu G, Freudenberger TD, Yang S, Curtis JR, Spada C, Hayes J, *et al*. High prevalence of abnormal acid gastro-oesophageal reflux in idiopathic pulmonary fibrosis. Eur Respir J. 2006; 27:136-142.
11. Coultas DB, Zumwalt RE, Black WC, Sobonya RE. The epidemiology of interstitial lung diseases. Am J Respir Crit Care Med. 1994; 150:967-972.
12. Hubbard R, Lewis S, Richards K, Johnston I, Britton J. Occupational exposure to metal or wood dust and aetiology of cryptogenic fibrosing alveolitis. Lancet 1996; 347:284-289.
13. Miyake Y, Sasaki S, Yokoyama T, Chida K, Azuma A, Suda T, *et al*. Occupational and environmental factors and idiopathic pulmonary fibrosis in Japan. Ann Occup Hyg. 2005; 49:259-265.
14. Hubbard R, Cooper M, Antoniak M, Venn A, Khan S, Johnston I, *et al*. Risk of cryptogenic fibrosing alveolitis in metal workers. Lancet. 2000; 355:466-467.
15. Hubbard R, Cooper M, Antoniak M, Venn A, Khan S, Johnston I, *et al*. Risk of cryptogenic fibrosing alveolitis in metal workers. Lancet. 2000; 355:466-467.
16. Kuwano K, Nomoto Y, Kunitake R, Hagimoto N, Matsuba T, Nakanishi Y, Hara N. Detection of adenovirus E1A DNA in pulmonary fibrosis using nested polymerase chain reaction. Eur Respir J, 1997; 10:1445-1449.
17. Ueda T, Ohta K, Suzuki N, Yamaguchi M, Hirai K, Horiuchi T. *et al*. Idiopathic pulmonary fibrosis and high prevalence of serum antibodies to hepatitis C virus. Am Rev Respir Dis. 1992; 146:266-268.
18. Irving WL, Day S, Johnston ID. Idiopathic pulmonary fibrosis and hepatitis C virus infection. Am Rev Respir Dis. 1993; 148:1683-1684.
19. Bitterman PB, Rennard SI, Keogh BA, Wewers MD, Adelberg S, Crystal RG. Familial idiopathic pulmonary fibrosis: evidence of lung inflammation in unaffected family members. N Engl J Med. 1986; 314:1343-1347.
20. Marshall RP, Puddicombe A, Cookson WO, Laurent GJ. Adult familial cryptogenic fibrosing alveolitis in the UK. Thorax. 2000; 55:143-146.
21. Hodgson U, Laitinen T, Tukiainen P. Nationwide prevalence of sporadic and familial idiopathic pulmonary fibrosis: evidence of founder effect among multiplex families in Finland. Thorax. 2002; 57:338-342.
22. Allam JS, Limper AH. Idiopathic pulmonary fibrosis: is it a familial disease? Curr Opin Pulm Med. 2006; 12:312-317.
23. Nishimura K, Kitaichi M, Izumi T, Nagai S, Kanaoka M, Itoh H. Usual interstitial pneumonia: histologic correlation with high-resolution CT. Radiology. 1992; 182:337-342.
24. Johkoh T, Muller NL, Cartier Y, Kavanagh PV, Hartman TE, Akira M, *et al*. Idiopathic interstitial pneumonias: diagnostic accuracy of thin-section CT in 129 patients. Radiology. 1999; 211:555-560.
25. Selman M, King TE, Pardo A, Idopathic Pulmonary Fibrosis; Prevailing and evolving hypothesis about the pathogenesis and implications for therapy, Ann Intern Med. 2001; 134: 136-151.
26. Johnston IDA, Procott RJ, Ehalmes JE, Rudd RM, British Thoracic Society study of Cryptogenic Fibros-ing Alveolitus, Current presentation and initial manage-ment. Thorax. 1997; 52:38.
27. Jindal SK, Malik SK, Deodhar SD, Sharma BK. Fibrosing alveolitis: A report of 61 cases seen over the past five years. Indian J Chest Dis Allied Sci. 1979; 21:174-79.
28. Sharma SK, Pande JN, Guleria JS. Diffuse interestitial pulmonary fibrosis. Indian J Chest Dis Allied Sci. 1984; 26:214-19.
29. Louw SJ, Bateman ED, Benatar SR. Cryptogenic fibrosing alveolitis: Clinical spectrum and treatment. South Afr Med J. 1984; 65:195-200.
30. Smith C, Feldman C, Levy H, Kallenbach JM, Zwi S. Cryptogenic fibrosing alveolitis: A study of an indigenous African population. Respiration. 1990; 57:364-71.
31. Johnston ID, Prescott RJ, Chalmers JC, Rudd RM. Fibrosing Alveolitis Subcommittee of the Research Committee of the British Thoracic Society. British Thoracic Society study of cryptogenic fibrosing alveolitis: Current presentation and initial management. Thorax. 1997; 52:38-44.
32. American Thoracic Society and the European Respiratory Society. Idiopathic pulmonary fibrosis: Diagnosis and treatment. International consensus statement. Am J Respir Crit Care Med. 2000; 161:646-64.