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Original Research

Assessment of risk factors and clinical profile of cases of Idiopathic pulmonary fibrosis

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ABSTRACT

Background: Idiopathic pulmonary fibrosis (IPF) is a fatal, chronic fibrosing interstitial pneumonia, of unknown aetiology. The present study assessed risk factors and clinical profile of cases of IPF. **Materials & Methods:** The present study was conducted on 48 cases of IPF of both genders. Onset and duration of symptoms, past and personal history to look for any risk factor, family history was recorded. Six minute walk test was performed to see exercise tolerance. Arterial blood gas was done to test hypoxia at rest. **Results:** Out of 48 patients, males were 28 and females were 20. Common risk factors were smoking seen in 21, positive family history was seen in 10 and GERD in 14 patients. Common symptoms in patients were cough in 45, chest pain in 42, fever in 12, hemoptysis in 7 and weight loss in 5 cases. The difference was significant ($P < 0.05$). **Conclusion:** Authors found that common risk factors in cases of IPF were GERD, positive family history and smoking and symptoms were cough, breathlessness and fever.

Key words: Cough, Fever, Idiopathic pulmonary fibrosis.

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INTRODUCTION

Idiopathic pulmonary fibrosis (IPF) is a fatal, chronic fibrosing interstitial pneumonia, of unknown aetiology, which affects primarily adults older than 50. Although there is a great variability in the occurrence of IPF, possibly due to geographic and demographic differences, the most reliable data estimate a prevalence ranging approximately 13–20 per 100,000 inhabitants in women and men, respectively. The IPF mean survival ranges between 2 and 4 years from diagnosis for patients not receiving anti-fibrotic treatment.¹

The American Thoracic Society and European Respiratory Society (ATS/ERS), in collaboration with the American College of Chest Physicians (ACCP), published an international consensus statement in 2000 on the diagnosis and management of IPF.² Importantly, the statement recognized IPF as a distinct clinical entity associated with the histologic appearance of usual

interstitial pneumonia (UIP), and provided specific recommendations for clinicians regarding its diagnosis and management. Since the publication of the 2000 ATS/ERS statement, studies have used the ATS/ERS statement recommendations to further our understanding of the clinical manifestations and course of IPF.³

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, stone cutting/polishing, microbial agents like Epstein Barr Virus and Hepatitis C, gastro esophageal reflux are found to be closely associated with IPF.⁴ The present study assessed risk factors and clinical profile of cases of IPF.

MATERIALS & METHODS

The present study was conducted in the department of Chest & TB. It comprised of 48 cases of IPF of both genders. All were informed regarding the study and written consent was obtained. Ethical approval for the study was taken from institutional ethical committee. Data such as name, age, gender etc. was recorded in case history proforma. The diagnosis was based on the ATS/ERS criteria for a diagnosis of IPF in the absence of open lung biopsy, exclusion of other known causes for interstitial lung disease; abnormal pulmonary

function testing results, HRCT findings of bibasilar reticular abnormalities with minimal ground-glass opacities; and a clinical history consistent with the diagnosis.

Onset and duration of symptoms, past and personal history to look for any risk factor, family history was recorded. Six minute walk test was performed to see exercise tolerance. Arterial blood gas was done to test hypoxia at rest. Patients were submitted to pulmonary function tests. Results were subjected to statistics. P vale <0.05 was considered significant.

RESULTS

Table I Distribution of patients

Total- 48		
Gender	Male	Female
Number	28	20

Table I shows that out of 48 patients, males were 28 and females were 20.

Table II Assessment of risk factors

Risk factors	Number	P value
Smoking	21	0.05
Family history	10	
GERD	14	

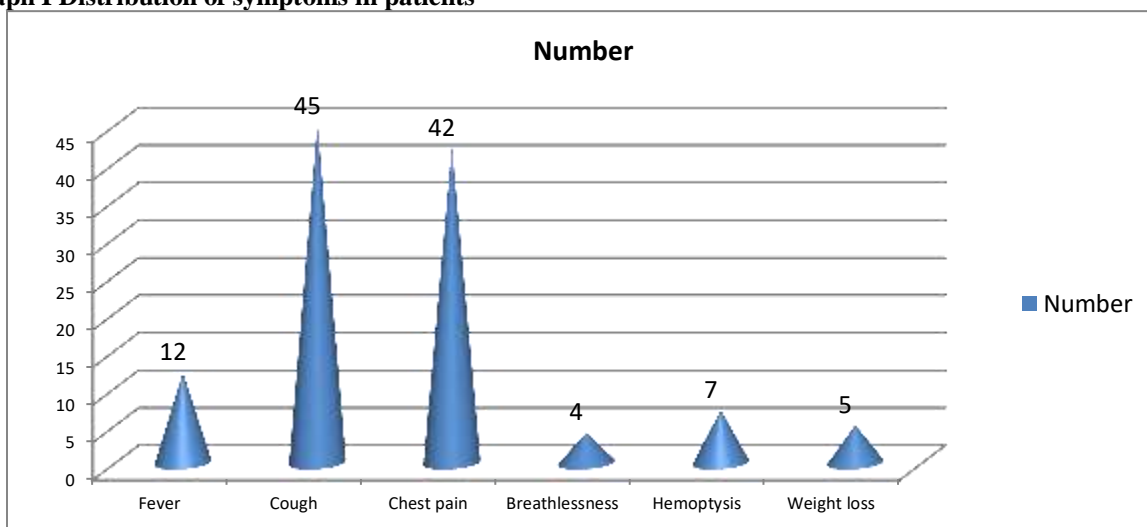
Table II shows that common risk factors were smoking seen in 21, positive family history was seen in 10 and GERD in 14 patients. The difference was significant (P< 0.05).

Table III Distribution of symptoms in patients

Symptoms	Number	P value
Fever	12	0.01
Cough	45	
Chest pain	42	
Breathlessness	4	
Hemoptysis	7	
Weight loss	5	

Table III, graph I shows that common symptoms in patients were cough in 45, chest pain in 42, fever in 12, hemoptysis in 7 and weight loss in 5 cases. The difference was significant (P< 0.05).

Graph I Distribution of symptoms in patients



DISCUSSION

Some factors have been identified to be associated with poorer prognosis and shorter survival time, such as older age, smoking status (smokers and ex-smokers), lower body mass index, more impaired pulmonary function (mainly on forced vital capacity, FVC, total lung capacity, TLC, and diffusing capacity for carbon monoxide, D_{LCO}), radiological findings (usual interstitial pneumonia, UIP), a pattern or greater extent of fibrosis, and the development of acute exacerbations or comorbidities, especially pulmonary hypertension and emphysema.^{5,6} There is evidence of high prevalence of up to 90% of classic gastroesophageal reflux disease (GERD) in IPF in a number of studies. Moreover, patients with scleroderma-associated lung fibrosis have significantly increased reflux episodes as compared with scleroderma patients without pulmonary fibrosis.⁷ The present study assessed risk factors and clinical profile of cases of IPF.

In this study, out of 48 patients, males were 28 and females were 20. The common risk factors were smoking seen in 21, positive family history was seen in 10 and GERD in 14 patients. Gupta et al⁸ conducted a study 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. Amongst 60 subjects, 16 were females and 44 were males. Most common age group was more than 60 years. Exertional breathlessness 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.

We found that common symptoms in patients were cough in 45, chest pain in 42, fever in 12, hemoptysis in 7 and weight loss in 5 cases. Fernández et al⁹ conducted a prospective, observational, multicentre and nationwide study that involved 608 IPF patients included in the SEPAR IPF Registry up to June 27th, 2017, and who received any treatment for their disease. IPF patients were predominantly males, ex-smokers, and aged in their 70s, similar to other registries. Upon inclusion, mean \pm SD predicted forced vital capacity was $77.6\% \pm 19.4$, diffusing capacity for carbon monoxide was $48.5\% \pm 17.7$, and the 6-min walk distance was $423.5\text{ m} \pm 110.4$. The diagnosis was mainly established on results from the high-resolution computed tomography in the proper clinical context (55.0% of patients), while 21.2% of patients required invasive procedures (surgical lung biopsy) for definitive diagnosis. Anti-fibrotic treatment was prescribed in 69.4% of cases, 51.5% pirofenidone and 17.9% nintedanib, overall with a good safety profile.

Anti-fibrotic treatments for IPF aim to slow down the disease progression and increase the survival time. To date, there are two effective disease-modifying therapies, pirofenidone and nintedanib. Besides the performance of clinical trials for investigating the efficacy and safety of novel drugs, observational studies from routine clinical practice are also required for understanding the natural course of the disease, and identifying differential patterns of diagnosis and treatments.¹⁰

CONCLUSION

Authors found that common risk factors in cases of IPF were GERD, positive family history and smoking and symptoms were cough, breathlessness and fever.

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