Idiopathic pulmonary fibrosis (IPF) in upper Egypt, a single center study

M. Alaa Rashad a,*, Ahmed K. Ibrahim b

a Department of Chest Diseases and Tuberculosis, Faculty of Medicine, Assiut University, Assiut, Egypt
b Department of Public Health, Faculty of Medicine, Assiut University, Assiut, Egypt

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Abstract  Aim of work: Idiopathic pulmonary fibrosis (IPF) is the second most common cause of admission to Chest Department at Assiut University hospital, the pattern of presentation, method for diagnosis and Co-morbidities need further studies.

Aim: To explore demographic data and pattern of presentation of patients with IPF in upper Egypt and to study the difference from international data.

Methods: A total of 568 patients with final diagnosis of IPF were studied, retrospective study was done using the available hospital database for all patients admitted at Assiut University hospital (Tertiary hospital for all upper Egypt Governorates) during the period from 2007 to 2012. Patients with incomplete data were excluded from study, all patients underwent chest X-ray, high resolution computed tomography, spirometry, arterial blood gases, in addition to routine laboratory investigation. Patients with clinical or laboratory evidence of collagen vascular disease or extrinsic allergic alveolitis were excluded from current study.

Results: The current study included 568 patients, 191 males and 377 females, mean age 44 ± 12 years. In all cases diagnosis was made according to clinical, spirometry and HRCT chest, no one was subjected to thoracoscopic or bronchoscopic lung biopsy. Most cases were house wife or farmer, 76% of cases were non-smokers, 17% ex-smokers and 7% current smokers, 86% of cases have restrictive spirometry. Usual interstitial pneumonia was the most common high resolution chest computed tomography pattern (51%) (Fig. 5). 39% of cases have at least one Co-morbid disease such as systemic hypertension, ischemic heart disease, pulmonary embolism and/or diabetes mellitus.

There was a significant difference between male and female patients with IPF with regard to smoking status (P value < 0.005) and HRCT pattern (P < 0.01).

Conclusion: IPF in upper Egypt has a different age and sex distribution compared to international data. Domestic air pollution, indoor exposures or other environmental factors may account for this difference. Lack of local resources for lung biopsy and lack of national guidelines for IPF
Introduction

Idiopathic pulmonary fibrosis (IPF) is a chronic progressive disease with unclear etiology and pathophysiology [1,2]. Its prognosis is poor; survival time after initial diagnosis is only 2.5–3.0 yr. There is currently no effective therapy known to improve survival [3,4].

Numerous studies on the etiology of IPF have identified many possible inciting factors including saw dust, metal particles, smoking, gastroesophageal reflux, and viruses [1]. Since IPF is more prevalent in the aged population, it has been hypothesized that the development and progression of IPF may be affected by age-related diseases such as diabetes mellitus (DM), metabolic syndrome, obesity, and cardiovascular disease [5–8].

Criteria for the diagnosis of IPF were set forth in an international consensus statement formulated by members of the American Thoracic Society (ATS) and the European Respiratory Society (ERS) [9]. In the absence of biopsy evidence of usual interstitial pneumonia, a constellation of typical clinical findings may be used to support the diagnosis of IPF. However, the extent to which these diagnostic guidelines are followed in actual clinical practice is unknown [9].

Median survival among persons with IPF is believed to be from 3 to 5 yr. Respiratory failure is the most frequent cause of death, and has been reported to account for over 80% [10,11–15].

Surprisingly, little is known about the epidemiology of IPF in the United States. Initial estimates of its prevalence, based largely on case series from pulmonary clinics and tertiary-care hospitals, ranged from 3 to 6 cases per 100,000 persons [16].

Substantially higher rates of prevalence (20 per 100,000 among men, 13 per 100,000 among women) and incidence (11 and 7 per 100,000, respectively) were reported in a study based on the adult population of Bernalillo County, New Mexico [17]. It is unclear, however, whether these rates are generalizable to the present-day United States as a whole, because they are based on data more than 15 yr old and come from an area of the United States known to attract persons with chronic lung diseases [18].

Although more recent estimates of disease prevalence (range, 1–24 cases per 100,000) are available from several European studies, their generalizability to the United States is not clear [19–23].

Methods

A single center retrospective hospital-based study was carried out in chest department, Assiut University hospital, Assiut Egypt (Tertiary hospital for all upper Egypt Governorates) between January 2007 and December 2012.

All patients with confirmed IPF admitted during the study period to Chest department at Assiut University, who agreed to participate in the research, were included in this study. The diagnosis of IPF was made based on the diagnostic criteria of American Thoracic Society and the European Respiratory Society by history taking, clinical examination, high-resolution computerized tomography (HRCT) of the chest and pulmonary function testing (PFT). None of the cases accepted to confirm the diagnosis by either thoracoscopic lung biopsy or transbronchial lung biopsy. The presence of typical clinical and HRCT features of IPF, when identified by expert clinicians and radiologists, is sufficiently characteristic to allow a confident diagnosis and eliminate the need for surgical lung biopsy. All cases had basal fine crackles in auscultation and predominantly peripheral, sub-pleural, bi-basal fine reticular shadows and/or honeycombing, occasionally with traction bronchiectasis on HRCT. All cases had also abnormal pulmonary function studies including evidence of restriction—reduced vital capacity with increased FEV1/FVC ratio. There was no evidence of either coexisting collagen-vascular disease or history of known occupational exposure to agents that might produce a clinical picture similar to that of IPF in any of the cases.
The study was approved by the Research Ethics Committee Assiut Faculty of Medicine, Assiut University.

**Results**

The current study included 568 patients, 191 males (34%) and 377 females (66%) (Fig. 1), mean age 44 ± 12 years. In all cases diagnosis was made according to clinical, spirometry and HRCT chest, no one was subjected to thoracoscopic or bronchoscopic lung biopsy. Most cases were housewife 53% or farmer 12% (Fig. 2), 76% of cases were non-smokers, 17% ex-smokers and 7% current smokers (Fig. 3), 86% of cases have restrictive spirometry, mixed 9% or normal 5% (Fig. 4). Usual interstitial pneumonia was the most common high resolution chest computed tomography pattern (51%), desquamative 20% and nonspecific 5% (Fig. 5). 39% of cases have at least one Co-morbid disease such as systemic hypertension, ischemic heart disease, pulmonary embolism and/or diabetes mellitus (Fig. 6).

There was a significant difference between male and female patients with IPF with regard to smoking status ($P$ value < 0.005) (Fig. 7) and HRCT pattern (Fig. 8) ($P$ < 0.01). Most females were non-smoker (97%).

**Discussion**

The current study included 568 IPF cases, showed that most cases diagnosed with IPF in upper Egypt are middle aged female housewife, epidemiology of IPF in the united states showed that IPF is more prevalent among aged males.

In a population-based study in Bernalillo County, New Mexico, Coultas et al. found a prevalence of ILD of 81 per 10\(^5\) in males and 67 per 10\(^5\) in females, and an incidence of 32 per 10\(^5\) per yr in males and of 26 per 10\(^5\) per yr in females [24].

In another study, farming and raising birds with the potential exposures to dusts of animal feeds, products and waste as well as pesticides were significant risk factors for the development of IPF among female workers. Also, the environmental exposure to domestic birds and cats was positively associated with IPF development in both genders. These findings were in accordance with results of current study.

Agricultural workers are exposed to very high levels of dust and aerosolized particulates from a variety of sources including feed grains, bedding, and livestock fecal material, [24] and tend to have a higher prevalence of lung fibrosis [25].

In Egypt, the poultry industry had expanded rapidly over the past 25 years to provide approximately 55% of the per capita animal protein consumption. Problems with raising birds in Egypt include widespread roof-top and back-yard raising bird, unhygienic local marketing and home slaughtering as well as the presence of approximately 40,000 poultry farms lacking biosecure and hygienic production systems and unprotected exposure to birds. 30, 31 these widespread unplanned and unprotected activities in raising birds and their environmental impacts help in magnifying the role of raising birds in IPF.
development. In Egypt, women were found to be more involved in raising birds than men and this may explain the elevated risk of IPF among women [24].

This study has several limitations. First, data from the medical records may be incomplete due to limited information. Second, this study was based on earlier, 2002 ATS-ERS guidelines for diagnosis of IPF and on retrospective study. Therefore, this study may have involved atypical IPF features on HRCT. Third, the interpretation of HRCT images may have differed in quality among radiologists. Further prospective studies are needed to confirm our results using the new IPF guidelines. Fourth, the IPF in patients was not confirmed by lung tissue biopsy.

Conflict of interest

We have no conflict of interest to declare.

References

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