Idiopathic Pulmonary Fibrosis in Soweto, South Africa: A Descriptive Study

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ABSTRACT

Background: Idiopathic Pulmonary Fibrosis (IPF) is a specific form of age-related fibroproliferative interstitial pneumonia that is chronic, progressive, and carries a poor prognosis, with median survival just 2.5-3.5 years from diagnosis. At present, there are very limited South African data available regarding the incidence, presentation, and risk factors in this population.

Objectives: This study sought to describe the cohort of patients that attended the respiratory outpatient services at a tertiary-level hospital in South Africa during the period 2007-2016. To the best of the authors' knowledge, this the first such descriptive study performed in Africa.

Methods: This was a retrospective, descriptive record review, that included patients ≥ 18 years of age who fulfilled 2011 ATS/ERS/JRS/ALAT diagnostic criteria for IPF.

Results: Data from 74 patients were used for analysis in this study, of which 60.8% were female. The mean age (SD) was 64.4 (10.9) years, and the majority (79.7%) were Black. Over half of the patients (40/74, 54.1%) were current or previous smokers, although there was no correlation between smoking history and age or baseline pulmonary function testing. All patients reported dyspnoea, which was mMRC grade 3 or 4 in 80% of patients. HRCT chest was reported as radiological UIP in 72 patients (97.3%), and three patients underwent lung biopsy, all of which showed a UIP pattern. Fifty-eight patients (78.4%) had spirometry results available, with median FVC 67.3% of predicted; this was significantly higher in females. Median TL_{CO} was 39% predicted. Twenty-five patients (33.8%) received corticosteroids, of whom five (6.8%) received the prednisone-azathioprine-NAC regime. Three patients (4.1%) received nintedanib; two of whom showed slowing of decline in lung function, although no significant symptomatic improvement was reported. Mean duration of follow up was 13.3 months, although 85.1% of patients were lost to follow up.

Conclusion: Despite a fairly small sample size and retrospective nature, this study contributes to the body of literature on IPF, and highlights the need for additional studies in developing countries, particularly in Africa. Of particular concern is the late presentation and advanced disease noted in this cohort of patients.