

Background

The 2022 ATS/ERS/JRS/ALAT criteria for progressive pulmonary fibrosis (PPF) were developed for fibrotic interstitial lung diseases other than idiopathic pulmonary fibrosis (IPF). Whether this multidimensional framework has prognostic value in patients with treated IPF remains uncertain. We evaluated the prevalence of PPF-defined progression and its association with mortality in patients with IPF receiving antifibrotic therapy.

Methods

This prospective cohort study included patients with multidisciplinary discussion-confirmed IPF enrolled in the Registry of Interstitial Lung Disease (REGILD) between 2018 and 2022. Disease progression was assessed one year after initiation of antifibrotic therapy using the 2022 PPF criteria. The primary outcome was 3-year all-cause mortality. Cox regression and nested Cox model analyses were performed to identify prognostic factors and evaluate incremental prognostic value beyond the GAP score.

Results

Ninety patients were included, of whom 50 (55.6%) fulfilled PPF criteria after one year of antifibrotic therapy. During a median follow-up of 3.6 years, 28 patients (31.1%) died. Three-year mortality was significantly higher in patients with PPF-defined progression than in those with stable disease (46.0% vs. 12.5%, $p = 0.001$). In multivariable analysis, lower baseline body mass index (BMI) (HR 0.87, 95% CI 0.78–0.96, $p = 0.006$) and PPF-defined progression (HR 3.39, 95% CI 1.23–9.34, $p = 0.018$) were independently associated with mortality. Addition of PPF-defined progression to a model containing the GAP score significantly improved model fit ($\Delta\chi^2 = 5.610$, $p = 0.018$).

Conclusions

PPF-defined progression and lower baseline BMI were independently associated with mortality in patients with IPF receiving antifibrotic therapy. PPF-defined progression provided incremental prognostic information beyond the GAP score, suggesting that assessment of longitudinal disease trajectory may complement conventional baseline risk stratification in treated IPF.

