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Title: Combined pulmonary fibrosis and emphysema: A distinct entity?

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Body: Introduction: Although combined pulmonary fibrosis and emphysema (CPFE) syndrome has been proposed as a distinct entity, it is controversial. Objectives: To investigate whether the combined pulmonary fibrosis and emphysema has different prognosis and prognostic factors from pure idiopathic pulmonary fibrosis. Methods: Clinical data and high resolution computed tomography images of 300 patients with idiopathic pulmonary fibrosis were retrospectively reviewed. The extent of emphysema and fibrosis were scored and the patients with moderate to severe emphysema (\geq grade 2) were categorized as combined disease group. Results: Seventy five (25.0%) patients had combined disease and the survival period was not significantly different from isolated idiopathic pulmonary fibrosis (n=200, p=0.190). In both groups, survival period was significantly correlated with fibrosis score, but not with emphysema score. In isolated idiopathic pulmonary fibrosis group and also all patient group, forced vital capacity and fibrosis score were predictors for mortality on multivariate analysis. In combined disease group, pulmonary hypertension was more frequent and an independent prognostic factor. However, in both groups, diffusion capacity for carbon monoxide was a sole determinant of pulmonary hypertension, suggesting the combined effect of both fibrosis and emphysema on the development of pulmonary hypertension. Conclusions: The combined pulmonary fibrosis and emphysema seems to be co-morbidity with similar survival period rather than a distinct disease entity.