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Title: Subtypes of pulmonary emphysema on HRCT affect prognosis on combined pulmonary fibrosis and emphysema in idiopathic pulmonary fibrosis

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Body: Background: The disease concept, combined pulmonary fibrosis and emphysema (CPFE), has been proposed as fibrotic lung disorder in recent years, although its diagnostic criteria and precise prognosis have not been established yet. On the other hand, it is known that prognosis of idiopathic pulmonary fibrosis (IPF) is very poor. In this study, we investigated the prognosis of IPF providing the characteristics of CPFE. Methods: Seventy-two patients with IPF who visited to our hospitals from 2007 to 2012 were investigated retrospectively. We diagnosed the patients as IPF according to the ATS/ERS-2002 statement. In the 72 patients, 66 were with findings of obvious honeycombing on HRCT. IPF with pulmonary emphysema (PE) on HRCT were divided into 3 groups by subtypes of PE as follows: IPF with paraseptal (p-PE) or centrilobular (c-PE) emphysema at the initial state, or, furthermore, mixed type of paraseptal and centrilobular emphysemas (m-PE) recognized at the final state. Prognosis of these groups was compared to that of IPF without PE (w/o-PE). Results: IPF with PE (n=28) had significantly worse prognosis compared to w/o-PE (n=38). The p-PE (n=16) showed worse prognosis than c-PE (n=12) at the initial state. During observation period, some c-PE patients resulted in m-PE (n=6). They showed poor prognosis similar to p-PE and were characterized by lower %DLco than c-PE patients who remained the initial state. Conclusions: IPF patients with p-PE or m-PE showed significantly worse survival than those without PE. In IPF patients with c-PE in HRCT, decline of %DLco suggests possibility of change in the subtype of emphysema to m-PE and poor prognosis.