漫談間質性肺病

Spectrum of interstitial lung diseases

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Abstract

Interstitial lung diseases (ILDs) encompass various lung diseases composed of distinct degrees of inflammation and fibrosis. Some of them predispose to progressive fibrosis after the initial insult, and eventually, patients with widespread fibrosis will require advanced respiratory support. Due to the complexity of diseases and their comorbidities, ILD diagnosis and subsequent treatment are highly dependent on multidisciplinary discussion and cooperation, aiming to optimize patient care and outcome. In the past, interdisciplinary teamwork may best be done in an ILD referral center. However, during the COVID-19 pandemic, it is suggested that teleconferences may solve this issue efficiently for the priority to decrease person-to-person contact.

There is no consensus on the definition of progression to date. Most experts agree that, within 24 months of the observation period, a relative decline of forced vital capacity (FVC) ≥10%, or a relative decline of FVC ≥5% occurring with a relative reduction of diffusion capacity of the lungs for carbon monoxide (DLCO) ≥15%, worsening symptoms or radiographic extent justify progression. Idiopathic pulmonary fibrosis (IPF) is the prototype of progressive fibrosing ILD (PF-ILD). The antifibrotic drugs nintedanib and pirfenidone have been proven to slow down FVC decline in some IPF patients, although not the reversal of the fibrosis. Other entities of PF-ILD include idiopathic non-specific interstitial pneumonia (iNSIP), unclassifiable idiopathic interstitial pneumonias, rheumatoid arthritis-associated ILD (RA-ILD), systemic sclerosis-associated ILD (SSc-ILD), interstitial pneumonia with autoimmune features (IPAF), hypersensitivity pneumonitis (HP), sarcoidosis, and ILDs related to other occupational exposures. For the above diseases categories excluding occupational exposure-associated ILDs, immunomodulation therapies are the conventional mainstay treatment strategy, and a non-absolute selection criterion for PF-ILD. In the

context of PF-ILD, nintedanib has demonstrated its efficacy after failure to immunomodulation therapies.

Despite taking antifibrotic drugs, some PF-ILD patients ultimately progress to end-stage disease and suffer from refractory cough, dyspnea, and severe resting and exertional hypoxemia. Therefore it is obligatory to evaluate these patients for lung transplantation and give them palliative care, including opiates and oxygen therapy, even earlier before they enter the end-stage disease.