Current concepts of the management of pulmonary fibrosis

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The last century experienced remarkable advances in the classification, diagnosis, and our understanding of the pathogenesis of the interstitial lung diseases. Technological advances, particularly physiological testing, lung imaging studies, bronchoalveolar lavage, surgical lung biopsy and histopathological assessment improved our understanding of these entities. This presentation will highlight some of the advances in the management of patients with pulmonary fibrosis.

Distinguishing usual interstitial pneumonia (UIP) from the other histopathological subgroups of idiopathic interstitial pneumonia (IIP) identified important therapeutic and prognostic implications. The historical belief that UIP represents the final common pathway for all forms of lung injury appears incorrect. Instead, UIP seems to be a distinct pathophysiologic entity characterized by minimal inflammation and chronic fibroproliferation due to abnormal parenchymal wound healing. In addition, the recognition and separation of nonspecific interstitial pneumonia (NSIP) from UIP has had major impact on our understanding of the IIPs.

The clinical course of idiopathic pulmonary fibrosis (IPF) is variable; however, the long-term survival is distinctly poor. Recent data from large clinical trials suggest that a large proportion of patients with mild-to-moderate IPF remain stable for prolonged periods while others experience an accelerated phase with a rapid decline and subsequent death. It is increasingly apparent that "acute exacerbations" or an "accelerated phase of rapid clinical decline" characterizes the clinical course of IPF and portends a poor prognosis. Histological examination of the lung has shown a pattern of acute lung injury (diffuse alveolar damage) on the background of UIP. Better understanding and management of these episodes appear critical to reducing the death rate in IPF. A number of features measured at the time of diagnosis or during the clinical course have been identified to be predictors of worse survival in IPF.

There has been a lack of large, randomized clinical trials to guide treatment for these diseases. Conventional management of IPF has been based on the concept that suppressing inflammation would prevent progression to fibrosis. Untreated patients invariably progress. Corticosteroids with or without azathioprine or cyclophosphamide show limited efficacy and frequent adverse effects in patients with IPF. Currently, there is no good evidence to support the routine use of any specific therapy in the management of IPF. The results of recent clinical trials will be discussed.

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