5 老化関連疾患 3）IPF

Idiopathic pulmonary fibrosis

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Key Words
■ 加齢
■ 特発性肺線維症
■ 遺伝子異常
■ 国際ガイドライン

Summary

It has been revealed that accumulation of internal and external factors with aging also triggers in lung fibrosis. As external factors, repeated inhalation can cause chronic inflammation such as dust inhalation including smoking. Internal factors include decreased numbers of alveolar epithelial cells and increased apoptosis due to genetic abnormalities such as surfactant protein and telomerase and epigenetic changes known as DNA methylation, and cell senescence, oxidative stress and decreased autophagy also contributes to the progression of fibrosis.

A new clinical practice guidelines summary for clinicians regarding idiopathic pulmonary fibrosis (IPF) has been released that describes clinical manifestations and diagnostic interventions for the disease. According to the guidelines, a useful strategy for evaluating the severity of lung function impairment in monitoring IPF progression. The preferred diagnostic strategy for IPF includes chest high resolution computed tomography (HRCT). The new guideline contains information on 4 diagnostic categories on HRCT: Usual interstitial pneumonia (UIP) patterns, probable UIP pattern, indeterminate for UIP pattern, and alternative diagnosis. In addition, the new guideline describes 4 distinct histopathologic patterns for IPF on biopsies. In order to be diagnosed for IPF, other causes of interstitial lung disease (ILD), including domestic and occupational environmental exposures, connective tissue disease (CTD), drug toxicity, should be excluded. According to the guideline, initial examination should identify the underlying conditions of ILD. If specific diagnosis is not made, then a multidisciplinary discussion focused on clinical findings, HRCT features, and, as appropriate, lung biopsy may help ascertain or exclude the diagnosis of IPF.

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概念，定義

加齢に伴い増加してくる呼吸器疾患として肺がん，慢性閉塞性肺疾患（chronic obstructive pulmonary disease：COPD），肺炎などがその代表であるが，特発性肺線維症（idiopathic pulmonary fibrosis：IPF）