The Role of Extracellular Matrix Quality in Pulmonary Fibrosis

This review discusses the role of extracellular matrix (ECM) quality in the pathogenesis of pulmonary fibrosis (PF). In PF, the highly ordered structure of collagens and elastin within the ECM of the lung is severely disrupted and lacks its original tissue quality. Discussions about the ECM have focused on the role of protein quantity in relation to the progression of PF, while the importance of lung ECM quality, defined by the levels of ECM protein modifications and by the protein distribution in lung tissue, has not been properly addressed. The quality and function of proteins may be altered by different post-translational modifications (PTMs), such as crosslinking, proteolytic cleavage, citrullination, misfolding and glycosylation. This paper is the first to review key data from the literature related to the lung ECM at the molecular level, relate these to changes observed at a macroscopic level and evaluate which PTMs most likely contribute to PF. This paper also reviews the role of novel neo-epitope-specific biomarkers in the early diagnosis and prognosis of fibrotic disorders. We discuss and argue that the altered quality of the individual ECM proteins contributes to the progression of PF and may also lead to the increased quantity of lung proteins. Thus, both quantity and quality appear to be of utmost importance.

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