INTRODUCTION - IPF

Idiopathic pulmonary fibrosis (IPF) is a disease that has an aggressive course and is usually fatal an average of 3 to 6 years after the onset of symptoms. Although the mechanisms underlying pulmonary fibrosis are not clearly understood, current evidence suggests that it is characterized by an excessive accumulation of extracellular matrix (ECM) remodeling of the lung architecture (fig. 1). The ultimate effector cell in pulmonary fibrosis is the myofibroblast characterized by the presence of alpha-smooth muscle actin (α-SMA).

EMT

The epithelial-mesenchymal transition (EMT) is one of the main processes involved in the development of fibrosis in IPF. In this process fully differentiated epithelial cells undergo transition to a mesenchymal phenotype giving rise to fibroblasts and myofibroblasts. The transforming growth factor (TGF-β) induces EMT in alveolar epithelial cells.

Epigenetic regulators and miRNAs

Most of these processes mainly affect suppressor factors or inducers of myofibroblasts. Among these are DNMT1, HDCA4 and miRNAs.

TREATMENTS

At the moment, the available treatments can reduce the symptoms but not the progression of the disease. Some research treatments are follows

CONCLUSIONS

- It is possible that IPF develops as a consequence of abnormalities occurring in multiple biological pathways that affect inflammation and wound repair.
- Epigenetic studies IPF are relatively new and are providing data of great interest.
- Investigate markers improve early detection could benefit the efficiency of treatments.

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