A novel take on idiopathic pulmonary fibrosis disease progression: localised autoimmunity

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Localised autoimmune responses in the lungs might contribute to disease progression in IPF
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It is well established that several of the interstitial lung diseases (ILDs) are driven by autoimmunity, such as rheumatoid arthritis-associated ILD and systemic sclerosis-associated ILD. These are therefore considered to be autoimmune diseases and frequently treated with immunosuppressive drugs. Idiopathic pulmonary fibrosis (IPF) is not viewed as an autoimmune disorder, but nevertheless, autoantibodies routinely studied in clinical practice are found in IPF and other types of idiopathic interstitial pneumonias. Serological autoreactivities, such as anti-cyclic citrullinated peptide antibody (anti-CCP), antinuclear antibody, rheumatoid factor (RF) and anti-neutrophil cytoplasmic antibody, have been reported in IPF [1] similar to patients with a connective tissue disease. These are sometimes classified as interstitial pneumonia with autoimmune features (IPAF) [2], based on a combination of clinical factors consisting of pulmonary and extra-thoracic features, serological evidence of autoantibodies, and certain imaging patterns. A Korean retrospective cohort study (n=512) showed that the IPAF serological pattern was present in 27% of IPF patients and was an independent risk factor for 5-year mortality [3]. A Canadian registry recently reported 15% of IPF patients being seropositive for RF or anti-CCP [4], though the clinical relevance of these findings is yet to be determined. The indirect significance of these autoantibodies in exacerbating disease was corroborated when 24 IPF patients with acute exacerbations showed clinical improvement and less requirement for supplemental oxygen upon autoantibody reduction therapy [5].