Idiopathic Pulmonary Fibrosis

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Introduction
Idiopathic pulmonary fibrosis (IPF) is a progressive, irreversible lung disease characterized by chronic inflammation and fibro-proliferation of the parenchymal cells of the lung that leads to chronic respiratory failure and ultimately death (Kearns, 2009). IPF is more prevalent in men than women, and risk for disease increases after age 60. IPF is the most common form of idiopathic interstitial pneumonia and it affects over 100,000 people in the United States alone (Ding et al., 2014). Most IPF cases are considered to be unexplained and sporadic in nature, however approximately 15-20% of cases have a family history of IPF and linked to antinuclear ribonucleoprotein (anti-RNP) autoantibodies (Tyring, Yung, & Toon; 2012). IPF is a debilitating disease with minimal treatment options and current research is being done to determine treatments that will optimize patient's lung capacity and improve treatments that will optimize patient’s quality of life. Although idiopathic pulmonary fibrosis affect most specific target of autoimmunity in IPF (p. 759).

Pathophysiology
Extensive research on the development and underlying etiology of IPF has been studied over the years. Although, the pathophysiology of IPF is complex and not entirely understood. Bajwa, Goyal & Blackstock (2010) simply stated that IPF is an autoimmune disease that results from a fibrotic lung from long standing epithelial injury of unknown cause and abnormal healing of the alveolocapillary barrier basement membrane due to dysregulated tissue repair of epithelial and endothelial cells (p. 159). Taille et al. (2010) present data that strongly suggests that IPF is an autoimmune disease. First piece of data that strongly suggests that IPF is autoimmune is the presence of B cell aggregates in an observed IPF lung; second, circulating CD4 T cells from the patient with IPF exhibits immune activation, CD4 T cells help produce cytokines and also immune activation. CD4+CD25+FOXP3+regulatory T cells in the lung parenchyma. Second, circulating CD4 T cells from the patient with IPF exhibits immune activation. CD4 T cells help produce cytokines and also interfere with fibrogenenic mediators, such as IL-10, transforming growth factor β-1, tumor necrosis factor-α. Third, CD4 + T cells purified from lymph nodes from patients with IPF proliferate when cultured with autologous lung tissue protein extracts. The responsible antigen(s) is still unidentified but several studies point to epithelial cell injury. Thus, the existence of IPF is an autoimmune disease. First piece of data that strongly suggests that IPF is autoimmune is the presence of B cell aggregates in an observed IPF lung; second, circulating CD4 T cells from the patient with IPF exhibits immune activation. CD4 T cells help produce cytokines and also interfere with fibrogenenic mediators, such as IL-10, transforming growth factor β-1, tumor necrosis factor-α. Third, CD4 + T cells purified from lymph nodes from patients with IPF proliferate when cultured with autologous lung tissue protein extracts. The responsible antigen(s) is still unidentified but several studies point to epithelial cell injury. Thus, the existence of IPF is an autoimmune disease. First piece of data that strongly suggests that IPF is autoimmune is the presence of B cell aggregates in an observed IPF lung; second, circulating CD4 T cells from the patient with IPF exhibits immune activation. CD4 T cells help produce cytokines and also interfere with fibrogenenic mediators, such as IL-10, transforming growth factor β-1, tumor necrosis factor-α. Third, CD4 + T cells purified from lymph nodes from patients with IPF proliferate when cultured with autologous lung tissue protein extracts. The responsible antigen(s) is still unidentified but several studies point to epithelial cell injury. Thus, the existence of IPF is an autoimmune disease.