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Idiopathic Pulmonary Fibrosis

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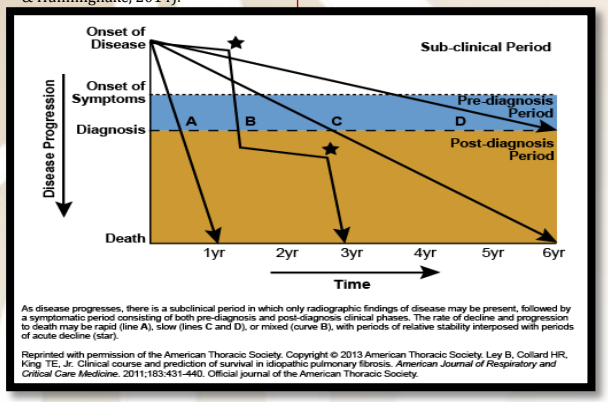
Introduction

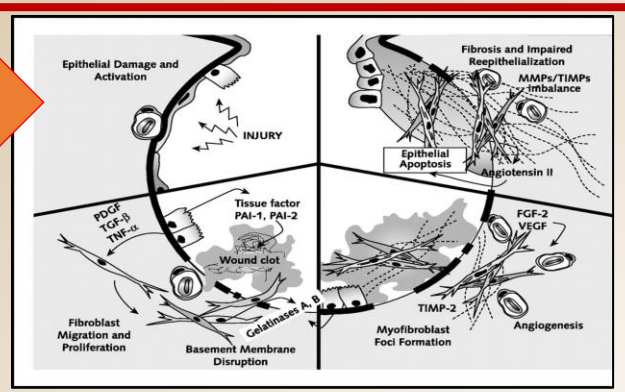
Idiopathic pulmonary fibrosis (IPF) is a progressive, irreversible lung disease, characterized by chronic inflammation and fibroproliferation of the parenchymal cells of the lung that lead to chronic respiratory failure and ultimately death (Kotsianidis et al., 2009). IPF is more prevalent in men than in 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 persons in the United States alone (Ding et al., 2011). Most of IPF cases are considered to be unpredictable and sporadic in nature, however approximately 15-20% of cases have a family history of IPF and linked to autosomal dominance disorder (Tsang, Wyatt, Ting, & Beattie, 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 quality of life. Though efforts are being made to decrease morbidity and mortality of this disease IPF remains to have a poor prognosis because it responds very little to medications and other treatments. Median survival after diagnosis is two to three years (Putman, Rosas, & Hunninghake, 2014).



American Thoracic Society (2013). Schematic Drawing of the Potential Clinical Course of Patients with Idiopathic Pulmonary Fibrosis [Figure 2]. Retrieved from http://lmt.projectsinknowledge.com/Activity/index.cfm?showfile=b&jn=2162&sj=2162.11&sc=2162.11.2

Pathophysiology

Extensive research on the development and underlying etiology of IPF has been studied over the years. Though the pathophysiology of IPF is complex and not entirely understood, Hauber & Blaukovitsch (2010) simply stated that IPF is an autoimmune disease that results in a fibrotic lung from underlying epithelial injury of unknown cause and abnormal healing of the alveolar-capillary barrier basement membrane due to dysregulated tissue repair of epithelial and endothelial cells (p. 159).
Taille et al. (2010) presented 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 on an observed IPF lung; organized with activated T lymphocytes and mature dendritic cells suggesting significant antigen activity 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 fibrogenic mediators, such as IL-10, transforming growth factor β-1, or tumor necrosis factor-α. Third, CD 4 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 the alveolar epithelial cell as a possible target of autoimmunity in IPF (p. 759).



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Signs & Symptoms

Subjective:
- worsening dyspnea
- dry cough
- chest pain
- fatigue
- low-grade fever
- weight loss

Objective:
- bilateral inspiratory crackles
- nailbed clubbing
- increased pulmonic second heart sound
- right ventricular lift
- tricuspid regurgitation
- Cyanosis (late clinical manifestation) (Ryu et al., 2014).

Diagnosis

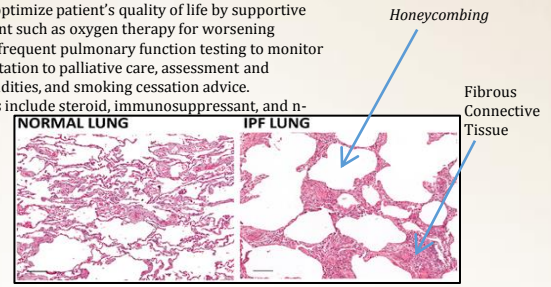
Recently discovered biomarkers that can help assist in the diagnostic and prognostic identification of IPF are available through peripheral blood smears. These biomarkers include:
- matrix metalloproteinases (MMP-1 & MMP-7)
- Krebs von den Lungen 6 (KL-6) which is a glycoprotein that is expressed mainly on type II pneumocytes
- surfactant protein A
- CD28 cells
- circulating myofibroblasts
- presence of oxidative stress
- (Torrens & Iwata, 2012).

Some of these biomarkers can be useful in staging IPF because the more biomarkers detected indicates the amount of alveolar epithelial cell injury.
Other laboratory tests:
- Increased WBC
- C-reactive protein
- lactate dehydrogenase.
- ABG- hypoxemia and hypercapnea & anticipate any need for mechanical ventilation (Bhatti, Girdhar, Usman, Cury, & Bajwa, 2013).

Idiopathic pulmonary fibrosis is a diagnosis of exclusion. In addition to the blood serum tests, a CT scan, bronchoalveolar lavage, and lung biopsy should be obtained. A hallmark sign of IPF on CT scan reveals extensive *"honeycombing"* and results of lung biopsy shows typical usual interstitial pneumonia (UIP) pattern, diffuse alveolar damage with or without hyaline membranes, numerous fibroblastic foci, and hemorrhage with capillaritis (Bhatti, Girdhar, Usman, Cury, & Bajwa, 2013).
Patients with IPF often develop pulmonary hypertension (PH) and right ventricular hypertrophy due to pulmonary vascular remodeling from hypoxic vasoconstriction thus echocardiogram should be used to help determine the extent of IPF. Approximately 85% of patients with end-stage IPF have PH (Ryu et al., 2014). Another common comorbidity of IPH is gastroesophageal reflux disease (GERD). Increased levels of pepsin are seen in the bronchoalveolar lavage fluid of IPF patients and suggest gastric aspiration. Others include lung cancer that can be difficult to diagnose due to fibrotic changes in the lungs but typically appear as irregular nodules, also venous thromboembolic disease due to immobilization (Ryu et al., 2014).

Nursing Implications

Idiopathic pulmonary fibrosis has a poor prognosis with median survival of 2-3 years thus as the nurse it is important to provide support, coping strategies, education regarding IPF, adjusting to a new lifestyle and offering pulmonary rehabilitation tailored to their needs and progression of disease (Duck, 2014). The nurse is also required to optimize patient's quality of life by supportive care and symptom management such as oxygen therapy for worsening hypoxia, treatment for cough, frequent pulmonary function testing to monitor progression of disease, consultation to palliative care, assessment and management of other comorbidities, and smoking cessation advice. Pharmacological interventions include steroid, immunosuppressant, and n-acetylcysteine (Duck, 2014).



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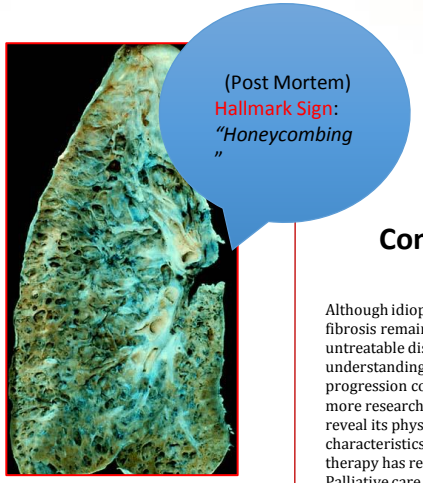

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Conclusion

Although idiopathic pulmonary fibrosis remains a debilitating and untreatable disease our understanding of the disease and its progression continues to advance as more research is being done to reveal its physiological characteristics, however effective therapy has remained indefinable. Palliative care education should be provided to the patient and family, as supportive care is the optimal therapy for this patient.

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