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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 lungs that lead to chronic respiratory failure and ultimately death (Kaplan 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 150,000 people in the United States alone (Boggs et al., 2013). Most of IPF cases are considered to be idiopathic and sporadic in nature, however approximately 15-20% of cases have a family history of IPF and linked to autosomal dominant disorder (Tsang, Wyatt, & Stacey, 2010). 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 be a poor prognosis because it represents a very little to no need for medications and other treatments.

Median survival after diagnosis is 5-10 years from diagnosis (Selman, King, & Parra, 2005). Idiopathic pulmonary fibrosis affect up to 150,000 people in the United States alone (Stanescu, 2014). Port Mortem Hallmark Sign: Honeycomb

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 & Blaak, 2009), it simply stated that IPF is an autoimmune disease that results in a fibrotic lung from long standing epithelial injury of unknown cause and abnormal healing of the air/sclerotic barrier basement membrane due to dysregulated tissue repair of epithelial and endothelial cells (p. 159).

Tseng 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 65% of cases with organized 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, and tumor necrosis factor-α. Third, CD4 T cells purified from lymph nodes from patients with IPF proliferate when cultured with autologous lung tissue in vitro. The antigen-specific (is) is still unknown but several studies point to lung epithelial cells as a possible target of autoimmunity in IPF (p. 739).

Signs & Symptoms

Subjective:
- worsening dyspnea
- dry cough
- fatigue
- palpable pleural friction 
- weight loss
- bilateral inspiratory cracks
- nafkle clothing
- paroxysmal nocturnal hemoptysis
- dyspnea on mild exercise
- clubbing
- Cyanosis
- cor pulmonale

Objective:
- Inspiratory crackles 
- clubbed fingers
- Kretschmers thoracic dystrophy
- use of accessory muscles of respiration
- Tachypnea
- flattening of the diaphragmatic silhouette

Diagnosis

Recently discovered biomarkers that can help assist in the diagnostic and prognostic identification of IPF are available through peripheral blood tests. These biomarkers include:
- matrix metalloproteinases (MMP-1 & MMP-7)
- Krupp von den Lenngen (KL-6) which is a glycoprotein that is expressed mainly on type II pneumocytes
- surfactant protein A
- C4C8 cells
- circulating myofibroblasts
- presence of oxidative stress

Some of these biomarkers can be useful in staging IPF but the more biomarkers detected indicates the amount of alveolar epithelial cell injury. Other laboratory tests:
- increased WBC
- C-reactive protein
- lactate dehydrogenase
- Alveolar hypoventilation and hypoxemia & anticipate any need for mechanical ventilation

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, and fibro-histiocytic foci, and hamorrhage with capillitium (Hauber, Gómez, Ouyang, & Joyce, 2013).

Signs & Symptoms


Nursing Implications

Idiopathic pulmonary fibrosis has a poor prognosis with median survival of 2-3 years thus as 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 and monitoring progression of disease, consultation to palliative care, assessment and follow-up of other comorbidities, and smoking cessation advice. Pharmacological interventions include steroids, immunosuppression, and n-acetylcysteine (Duck, 2014).

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Patients with IPF often develop pulmonary hypertension (PH) and right ventricular hypertrophy due to pulmonary vascular remodeling from hypoxic vasoconstriction thus echocardiogram is used to help determine the extent of IPF. Approximately 85% of patients with end-stage IPF have PH (Duck et al., 2014). 69% of IPF patients have PH. Some of these IPF patients also have pulmonary hypertension. Patients with pulmonary hypertension have a worse prognosis. The presence of PH is an important feature of IPF and is an indication for lung transplantation. Other hallmarks include lung cancer that can be difficult to diagnose due to the fibrotic changes in the lungs but typically appear as irregular nodules, also venous thromboembolism.

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 unravel its physiological characteristics, however effective therapy remains minimizable. Palliative care education should be provided to the patient and family, as supportive care is optimal for this therapy for patient.