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Recommended Citation
Rosselot, Stacey, "Idiopathic Pulmonary Fibrosis" (2014). Nursing Student Class Projects (Formerly MSN). 51.
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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 (Khanna et al., 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 persons in the United States alone (Hargreaves et al., 2013). 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 dominant disorder (Tang, Myntt, & Bourg; 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. Ethical concerns include being made to deceive morbidity and mortality of this disease. IPF remains a killer disease because it regards very little to modifications and other treatments. Median survival after diagnosis is two to three years (Bhatti, Bajwa, & Henshaw, 2014).

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 (Lai, Blaukovitsch & Kotsianidis, 2010) simply stated 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 initial and endoepithelial cells (p. 159). Tavolli et al. (2010) present data that strongly suggest that IPF is an autoimmune disease. First piece of data that strongly suggests that IPF is autoimmune in the presence of B-cell aggregates in an immune response pattern organized with activated T lymphocytes and mature dendritic cells which suggest 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 immune activation. T cells are also “target of autoimmunity in IPF (p. 759).”

Nursing Implications
Idiopathic pulmonary fibrosis has a poor prognosis with median survival of 2-3 years thus the nurse 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 and teaching to monitor progression of disease, consultation to palliative care, assessment and intervention of other comorbidities, and smoking cessation advice. Pharmacological interventions include steroid, immunosuppressant, and n-acetylcysteine (Duck, 2014).

References


Diagnosis
Recently discovered biomarkers that can help assist in the diagnostic and prognostic identification of IPF are available through peripheral blood tests. Some biomarkers include:

- matrix metalloproteinase (MMP-1 & MMP-7)
- KL-6 (Kreis von den Langen (KL-6) which is a glycoprotein that is expressed mainly on type II pneumocytes)
- surfactant protein A
- CEA
- circulating myofibroblasts
- pressure of oxidative stress

Some of these biomarkers can be useful in staging IPF however more biomarkers detected indicates the amount of alveolar epithelial cell injury. Other laboratory tests:

- increased WBC
- C-reactive protein
- lactate dehydrogenase

All of these biomarkers are hypercapnic and anticipate need for mechanical ventilation (Bluth, Gribar, Uman, Cary, & 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 reveal extensive “honeycombing” and resulting of lung biopsy shows typical usual interstitial pneumonia (UIP) pattern, diffuse alveolar damage with or without hyaline membrane, nonspecific fibrotic, and hemorrhage with capillaritis (Bhatti, Gribar, Oumay, Cary, & Bajwa, 2013). Patients with IPF often develop pulmonary hypertension (PH) and right ventricular hypertrophy due to pulmonary vascular remodeling from hypoxic vascular constriction thus echocardiogram should be used to help determine the extent of IPF. Approximately 85% of patients with end-stage IPF have PH (Duck et al., 2014). Patients with IPF with PH must be critically evaluated. Others include collapse that can be difficult to diagnose due to chronic changes in the lungs but typically appear as irregular nodules, also venous disease (Kotsianidis, Blaukovitsch, Nakou, & Leach, 2010). Idiopathic pulmonary fibrosis is 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 minimal. Palliative care education should be provided to the patient and family, as supportive care is the optimal therapy for this patient.

Signs & Symptoms

Subjective:

- worsening dyspnea
- dry cough
- fatigue
- chest pain
- weight loss

Objective:

- bilateral inspiratory cracks
- nailbed clubbing
- pleural or subcutaneous heart sound
- tricuspid regurgitation (post clinical manifestation) (Ryu et al., 2014).

Post Mortem

Honeycombing


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 minimal. Palliative care education should be provided to the patient and family, as supportive care is the optimal therapy for this patient.