Summer 2015

Idiopathic Pulmonary Fibrosis: Understanding Has Led to Exciting Treatments

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Idiopathic Pulmonary Fibrosis: Understanding Has Led to Exciting Treatments

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Introduction

Pulmonary fibrosis can be described as an end-stage interstitial lung disease characterized by scarring of the lung tissue. It is a heterogeneous group of idiopathic interstitial pneumonias, of which idiopathic pulmonary fibrosis (IPF) is the most common form. The exact cause of IPF is unknown. When the disease cannot be determined, a diagnosis of idiopathic pulmonary fibrosis is made. The mean survival for patients with IPF is approximately 5-8 years from the time of diagnosis. IPF is found primarily in men over 50 years of age, with a higher incidence of cigarettes smokers compared to non-smokers. Environmental and occupational inhalation exposures (Lekes, 2011, p. 265), a genetic component (Barkauskas et al., 2014, p. 1402), and multiple cell and responses may be involved in the development of IPF (Coon, Wolff, & Cannon, 2014, p. 246).

Presentation of Case

A 76 year old man was admitted with shortness of breath and cough. He has a history of smoking until a few years ago and is now requiring increased amounts of home oxygen. He has a history of Pneumonia (Acute lower Respiratory disease) and Paroxysmal Nocturnal Dyspnea. Since he has no acute medical problems, he is being treated with diuretics and oxygen. He is being treated with prednisone and is doing well. He has a history of IPF for 2 years, which is characterized by thickening of the septal walls and subpleural honeycombings (Barkauskas, 2012, p. 256).

Signs and Symptoms of IPF

According to the National Lung Heart and Blood Institute (2011, p. 359), signs and symptoms of IPF can be divided into early and late symptoms. Early symptoms include: increased breathing rate, unintended weight loss, and secondary symptoms are: increased shortness of breath at rest. The other late symptoms are: decreased exercise capacity, unintentional weight loss, and secondary symptoms are: increased shortness of breath at rest. The other late symptoms are: decreased exercise capacity, unintentional weight loss, and secondary symptoms are: increased shortness of breath at rest.

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Underlying Pathophysiology of IPF

“Idiopathic pulmonary fibrosis (IPF) is a deadly lung disease with few therapeutic options. Apotasis of alveolar cells, followed by abnormal tissue repair characterized by hyperplastic epithelial cell formation, is a pathogenic process that contributes to the progression of pulmonary fibrosis. However the signaling pathways responsible for increased proliferation of epithelial cells is poorly understood” (Weng et al., 2014, p. 1402).

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Nurses with updated evidence based practice and information on IPF can be an invaluable source of education for patients and family.

Significance of Patho

Nursing Implications

- Nurses with updated evidence
- Practice and information on IPF can be an invaluable source of education for patients and family.

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References