PULMONARY FIBROSIS

Clinical Manifestations of Autoimmune Lung Disease
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Idiopathic pulmonary fibrosis can occur as the sole disease or as an accompanying disorder in patients with SLE and other autoimmune conditions.

What is Pulmonary Fibrosis?

Pulmonary fibrosis or interstitial lung disease is a condition of lung tissue scarring, which affects more than 5 million people worldwide. Pulmonary fibrosis can occur as a primary autoimmune condition, and it can occur in conjunction with a number of autoimmune inflammatory conditions such as sarcoidosis, scleroderma, Wegener’s granulomatosis, rheumatoid arthritis, systemic lupus erythematosus (SLE), mixed connective tissue disease, and hypersensitivity pneumonitis after exposure to inhaled organic dusts or occupational chemicals.

Pulmonary fibrosis can also develop in conditions referred to by the collective term pneumoconiosis. These disorders occur as a result of inhaling dust contaminated with bacterial, fungal, viral, or animal products or certain gases, and exposure to ionizing radiation such as radiation therapy used to treat chest tumors.

Environmental Triggers

Certain drugs, such as the cytotoxic agents bleomycin, busulfan, and methotrexate; the antibiotics nitrofurantoin and sulfasalazine; the anti-arrhythmics amiodarone and tocainide; anti-inflammatory medications such as gold and penicillamine; and illicit drugs such as crack cocaine, methamphetamine and heroin can cause a drug-induced form of pulmonary fibrosis.

Disease Process

Pulmonary fibrosis is an inflammatory disease of the lower respiratory tract that damages the lung tissue and the epithelial cells of the alveolar air sacs through a process characterized by diffuse interstitial fibrosis with mild inflammation. In fibrosis, normal tissue becomes scarred by persistent inflammation, causing the tissue to become wood-like or hardened. Damaged, the lung tissue in pulmonary fibrosis can’t provide sufficient oxygen for the body’s needs.

Some conditions of pulmonary fibrosis respond well to treatment while others are progressive and unresponsive to treatment. Women are more likely to develop pulmonary fibrosis associated with connective tissue diseases, whereas men are more likely to develop pulmonary fibrosis associated with rheumatoid arthritis and occupational exposures, for instance exposures to silica dust.
Because pulmonary fibrosis can occur as a gradual process, patients with condition related to occupational exposure are typically older than 50 years, whereas patients with pulmonary exposure associated with rheumatological conditions are usually affected at an earlier age. The incidence of pulmonary fibrosis appears to be on the rise, with twice as many people diagnosed in the last decade compared to previous years. Median survival at diagnosis is 3 to 5 years.

**Symptoms**

Symptoms of pulmonary fibrosis include dry cough, wheezing, exercise intolerance, and shortness of breath. In advanced disease, normal activities such as climbing stairs become impossible. Lack of oxygen (hypoxia) causes central cyanosis, reduced lung volumes, clubbing of the fingertips and a susceptibility to infection. In certain conditions such as sarcoidosis, patients may also have enlarged lymph nodes. In rheumatological conditions, patients may have skin changes and joint inflammation.

**Diagnosis**

Diagnosis is made by blood oxygen capacity and respiratory function tests, imaging tests, and biopsy studies. In tissue studies performed on biopsy specimens, honeycombing is indicative of end-stage disease.

**Treatment**

Corticosteroids such as prednisone are used to reduce inflammation. Recent studies suggest that treatments targeting caveolin-1, a natural substance considered to be a protective inhibitor of pulmonary fibrosis, could be a promising therapy as are treatments targeting osteopontin.

In advanced disease, supplemental oxygen is used to compensate for the pulmonary deficit. Lung transplantation is used in selected patients.

Lifestyle changes include smoking cessation and avoidance of inhalants and medications that may contribute to toxicity. Natural treatments include the herb Centella Asiatica, which is used to heal damaged capillaries and aloe vera oil and D-Mannose, which are used to rebuild tissue and fight infection. Antioxidants, anti-inflammatory herbs, interferon-gamma-1b, and N-acetylcysteine (NAC) are used to reduce inflammation and oxidative stress. Sodium perborate is used to help with systemic healing and gingko biloba is used to increase circulation.

**Resources:**


*Cure Idiopathic Pulmonary Fibrosis*, Earth Clinic, folk remedies and holistic cures, accessed March 10, 2007.
Study Offers Clues to Origins of Autoimmune Disease, Media Contact, National Jewish Medical and Research Center, Denver, CO, Oct 2, 2005.

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