Pneumoconiosis — Symptoms and Treatment

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Pneumoconiosis is an occupational lung disease that is related to coal dust exposure. Macrophages and fibroblasts become activated and focal lung fibrosis occurs. If left untreated, patients can progress to massive lung fibrosis which carries high mortality. History taking and chest x-rays are the mainstay diagnostic tests to confirm and stage pneumoconiosis. Current treatment of pneumoconiosis is mainly symptomatic. Lung transplantation might be an option for patients with advanced pneumoconiosis.

Overview of Pneumoconiosis

Pneumoconiosis can be classified into different types depending on the type of dust causing the disease. Examples are berylliosis, coal worker's pneumoconiosis (CWP), silicosis and asbestosis.
Pneumoconiosis is the **chronic accumulation of dust in the lung parenchyma** and the **inflammatory reaction** related to coal or other dust and minerals. The inflammatory or fibrotic response and the actual type of disease that will develop, depends on the type and amount of substance inhaled.

- **Building trades**
  - Asbestos – Asbestosis (identical to pulmonary fibrosis)
  - Pleural plaques, thickening and effusions
- **Mining**
  - Coal dust – COPD, micronodular infiltrates, progressive massive fibrosis
  - Silicosis – micronodular infiltrates, TB
- **Industrial exposure**
  - Metals – micronodular infiltrates, diffuse alveolitis
  - Beryllium – similar to sarcoid

Two distinct types of coal worker’s pneumoconiosis exist – **simple CWP** and **progressive CWP**. As the name implies, patients with simple CWP have limited lung disease that is not progressive while patients with progressive CWP have an ongoing disease process that is associated with **extensive lung fibrosis**.

### Epidemiology of Pneumoconiosis

Pneumoconiosis is an **occupational lung disease**, hence, it is known to be more common in people with certain professions such as **coal miners**. Therefore, most patients in the United States come from Pennsylvania, Maryland, Virginia and Kentucky where coal mining is common. Pneumoconiosis is a common condition among coal miners with an estimated prevalence of 30%, while the prevalence of progressive CWP can be as high as 2.5% in people exposed to coal dust.

Additionally, pneumoconiosis-related mortality has significantly decreased in the last decades due to several reasons. While the most important cause for the reduction of pneumoconiosis-related mortality can be traced to the decline of the coal-mining workforce, **early diagnosis** and **prompt treatment** have also been linked to less progression of the disease.

Pneumoconiosis is a chronic disease process, that takes years to happen. Therefore, the majority of the patients are usually **older than 50** years of age. Pneumoconiosis has been identified in both sexes and no racial differences have been noted.

### Pathophysiology of Pneumoconiosis

Different **stages** of pneumoconiosis can be identified; each has its own pathogenic features. The **earliest stage of coal dust accumulation in the lungs** is known as **anthracosis**. While anthracosis is common among **coal miners**, it has also been described in **tobacco smokers** without previous history of exposure to coal dust.

When the patient inhales coal dust, either due to smoking, or because of coal mining, carbon molecules reach the terminal bronchioles. Once there, the **alveolar macrophages** engulf the carbon molecules. While this process is usually enough to clear out coal dust, the system can sometimes become overwhelmed, such as occurs in coal miners. Once this happens, the coal dust-filled macrophages start accumulating in the alveoli and trigger an **inflammatory response**.
The first pathogenic step in pneumoconiosis is believed to be the activation of fibroblasts, which secrete reticulin. Reticulin is responsible for the entrapment of more macrophages in the alveoli. One important molecule in coal dust is silica, which is responsible for accelerated macrophage lysis and the recruitment of more fibroblasts. These recruited fibroblasts secrete collagen and start the process of fibrosis.

At first, distinct focal areas of macrophages that are filled with coal dust and activated fibroblasts form coal macules. These are pathognomonic histologic patterns of CWP. The next step in the pathogenesis of CWP is the accumulation of multiple coal macules together, the deposition of more collagen and interstitial lung fibrosis. The consequence of this pathology is the formation of focal emphysema which can be asymptomatic at first.

In select patients, CWP can become progressive, as coal macules start recruiting more activated immune cells and extensive lung fibrosis ensues. It is believed that rheumatoid arthritis and the rheumatoid factor are two important factors for the progression of CWP. Pneumoconiosis, when associated with rheumatoid arthritis, is known to cause Caplan syndrome, which is characterized by progressive massive lung fibrosis.

Clinical Presentation of Pneumoconiosis

Pneumoconiosis like other forms of interstitial lung disease can be diagnosed with detailed history taking combined with advanced imaging modalities. Social history taking including occupational history is essential in the evaluation of the patient with suspected pneumoconiosis. Those who work in the coal mining industry are at an increased risk of pneumoconiosis. Additionally, the identification of the silica content amount of the coal that the patient was exposed to is essential to estimate the risk of progressive CWP.
Smoking history is also important, because coal miners who smoke are more likely to exhibit symptoms of CWP compared to non-smokers. Unfortunately, the majority of the patients with pneumoconiosis do not consider changing their career as a viable option.

Patients with simple CWP can be asymptomatic or can present with non-specific symptoms such as productive cough. Once the disease progresses, patients can develop dyspnea that is typically progressive. If left untreated, patients can develop signs of right-sided heart failure.

Pneumoconiosis can also be associated with the occlusion of small bronchioles, which predisposes the patient to infectious complications. Those with infectious disease can develop fever, night sweats and chest pain.

Diagnostic Workup for Pneumoconiosis

Laboratory investigations are not helpful in the confirmation of the diagnosis but can exclude infectious complications. Therefore, patients with pneumoconiosis who present with fever might be good candidates for a complete blood count and sputum culture to exclude possible infectious disease.

Chest X-ray provides valuable information in the diagnosis and staging of pneumoconiosis. Patients with simple CWP can present with lung nodules that are less than 1 cm in diameter. These nodules are usually in the upper lung lobes.
Patients who present with lung nodules that are larger than 1 cm in diameter are considered to have **progressive massive fibrosis**. These patients usually have **multiple small nodules and one large nodule** in addition to the simple CWP picture.

**Pulmonary function tests** usually are normal in people with simple CWP. Patients with complicated or progressive CWP can have **decreased ventilatory capacity** on pulmonary function testing.

### Treatment of Pneumoconiosis

Unfortunately, there is **no definitive treatment** for pneumoconiosis regardless of the stage of the disease. Current treatment approaches are mainly **symptomatic**. One of the main problems with pneumoconiosis is **hypoxia**, therefore, **oxygen therapy** is indicated in patients with moderate to severe hypoxemia.

Additionally, recent evidence has shown that **stopping exposure to coal dust** will not stop the progression of the disease once the patient develops progressive CWP. Therefore, replacing the patient in another line of work rather than coal-mining is questionable. On the other hand, patients with simple CWP can halt the disease progression by transferring to a job that has less exposure to coal dust.

Patients with **Caplan syndrome** can be hypoxemic. **Oxygen therapy** combined with **bronchodilators** is indicated in these patients. As Caplan syndrome is related to rheumatoid arthritis, **disease modifying antirheumatic drugs** are usually indicated, per current guidelines for the treatment of rheumatoid arthritis.

Patients with pneumoconiosis are at a significant risk of **influenza-related mortality** and immunization against influenza is recommended. Patients with unexplained weight loss, fever, night sweats and chronic cough might have a **superimposed mycobacterial infection** and specific treatment for mycobacterial infection should be started.

Patients with progressive massive lung fibrosis or complicated CWP might be possible candidates for bilateral or single **lung transplantation**. In one study, it was reported
that patients do relatively well after lung transplantation. These patients, however, had not reached the stage of cor-pulmonale or right-sided heart failure. Therefore, mortality remains high in patients with cor-pulmonale due to massive lung fibrosis.

References

Silicosis and coal workers’ pneumoconiosis via nih.gov
Coal Worker’s Pneumoconiosis via medscape.com

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