Caplan's Syndrome

Description

This is pulmonary fibrosis, usually in coal miners who have rheumatoid arthritis (RA). The syndrome is named after Dr Tony Caplan who was a doctor on the pneumoconiosis board in Cardiff. There are a few earlier papers on the subject, calling it Caplan's syndrome. It seems to have been discovered in the 1950s.

Epidemiology

It was said to affect 1 in 100,000 people but the incidence is falling as the coal mining industry has been in decline. The prevalence is so low that much of the literature is case reports rather than trials. PubMed lists no more than 20 papers in the past 10 years.

Risk factors

Rheumatoid arthritis (RA) is a systemic disease and not just a disease of joints. The combination of RA and exposure to coal dust produces the condition. It develops especially in miners working in anthracite coal mines and in persons exposed to silica and asbestos. There is probably also a genetic predisposition and smoking is thought to be an aggravating factor.

Presentation

Symptoms

There is cough and shortness of breath. In addition there are the features of rheumatoid arthritis (RA) with painful joints and morning stiffness.

Signs

There are features of RA, including tender swollen metacarpophalangeal (MCP) joints and rheumatoid nodules. The nodules may pre-date the appearance of RA by several years. Examination of the chest may show diffuse rales that do not disappear on coughing or taking a deep breath.

Differential diagnosis

The combination of rheumatoid arthritis (RA) and exposure to coal dust are essential for the diagnosis but silicosis and asbestosis must be considered. The X-ray appearance can resemble tuberculosis (TB).

In RA, lung disease can develop even in the absence of dust exposure. It includes interstitial fibrosis, pleural effusion, pulmonary nodules, pulmonary arteritis and pulmonary hypertension.

Investigations

CXR shows multiple, round, well-defined nodules, usually 0.5-2.0 cm in diameter, which may cavitate and resemble tuberculosis (TB). CT scanning gives a better picture of cavitation.

Spirometry may reveal a mixed restrictive and obstructive ventilatory defect with a loss of lung volume. There may also be irreversible airflow limitation and a reduced gas transfer factor.

Rheumatoid factor, antinuclear antibodies, and non-organ specific antibodies may be present in the blood. ESR or PV and CRP will be elevated. X-rays of affected joints will show the features of rheumatoid arthritis (RA) with bone erosions.

Associated diseases
Tuberculosis (TB) must be sought and treated if found.

Management

Nondrug
- Exposure to coal dust must cease.
- Physical treatment should proceed as for rheumatoid arthritis (RA).
- Smoking should cease.

Drugs
After exclusion of TB, steroids are used. Treatment of the RA will include disease-modifying anti-rheumatic drugs (DMARDs) at an early stage.

Complications
Tuberculosis (TB) may co-exist. There can be complications from steroids and other forms of treatment, whether non-steroidal anti-inflammatory drugs (NSAIDs) or DMARDs.

Prognosis
This is as for RA. Severe respiratory disability is uncommon but massive pulmonary fibrosis can progress at times. Spontaneous remission of the lung disease can occur.

Prevention
People with rheumatoid arthritis (RA) must not be exposed to additional risk factors for lung disease.

Further reading & references

- Dhingra A et al, Coal Worker's Pneumoconiosis, Medscape, Jul 2010

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Author: Dr Richard Draper
Peer Reviewer: Dr John Cox

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