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PERSPECTIVE

Pneumoconiosis

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Introduction

The pneumoconioses are parenchymal lung diseases that arise from inhalation of (usually) inorganic dusts at work. Some such dusts are biologically inert but visible on a chest X-ray or CT scan; thus, while they are radiologically alarming they do not give rise to either clinical disease or deficits in pulmonary function. Others – notably asbestos and crystalline silica – are fibrogenic so that the damage they cause is through the fibrosis induced by the inhaled dust rather than the dust itself. Classically these give rise to characteristic radiological patterns and restrictive deficits in lung function with reductions in diffusion capacity; importantly, they may progress long after exposure to the causative mineral has finished.

In the UK and similar countries asbestosis is the commonest form of pneumoconiosis but in less developed parts of the world asbestosis is less frequent than silicosis; these two types are discussed in detail below. Other, rarer types of pneumoconiosis include stannosis (from tin fume), siderosis (iron), berylliosis (beryllium), hard metal disease (cobalt) and coal worker's pneumoconiosis.

Asbestosis

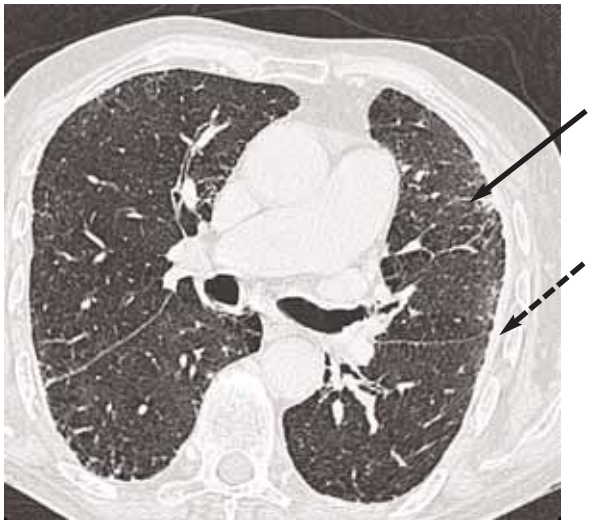
Clinical scenario

A man of 78 reports gradually worsening breathlessness; he has no relevant medical history of note and has never been a regular smoker. His spirometry reveals that both his FEV_1 and FVC are about 50% of their predicted values; the machine interprets this as a 'restrictive' picture. In his 20's-30's he spent about 15 years working in the boiler rooms of a power station.

A chest X-ray reveals several pleural plaques, some calcified, but no other abnormalities.

Because of his symptoms and pulmonary function abnormality (neither of which could be explained by pleural plaques alone – see Box 1) he was referred for further investigation. A thoracic CT scan identified a limited degree of bilateral, lower zone fibrosis. The combination of these findings with his history of occupational exposure was considered sufficient for a diagnosis of asbestosis.

Figure 1. Asbestosis; the HRCT scan shows the typical picture of subpleural fibrosis (solid arrow); in addition there is diffuse, left-sided pleural thickening (broken arrow), characteristic too of heavy asbestos exposure



How is the diagnosis made?

Asbestosis is the 'pneumoconiosis' that arises from exposure to asbestos in the workplace.[1] The diagnosis is made when, on the background of heavy occupational exposure to any type of asbestos, there is radiological evidence of pulmonary fibrosis (see Box 2). The diagnosis is not necessarily straightforward and is often a matter of judgement[2] after taking the following into account:

- The radiological features of asbestosis are non-specific and closely resemble the 'usual interstitial pneumonia' that is characteristic of 'idiopathic' pulmonary fibrosis (IPF). Changes are far more readily seen on CT scan (Figure 1) than on a conventional chest X-ray.[3] In fact, the increasing use of thoracic CT in both respiratory and cardiac care is probably responsible for some of the apparent increase in the incidence of asbestosis. Other radiological features of asbestos exposure such as pleural thickening or plaques are often seen and can be used to help make the distinction between asbestosis and IPF.

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- The 'dose' of asbestos required to induce fibrosis is relatively high and usually acquired over at least several years of high exposure in the workplace; these doses are generally higher than those necessary to induce pleural plaques or mesothelioma. Common occupations associated with asbestosis include boiler lagging with asbestos (as is likely in this case), work with asbestos textiles, and the manufacture of asbestos-containing building materials. In the UK these are long-abandoned industries, but because the latency of asbestosis is also long, new cases continue to be identified; indeed the incidence of, and mortality from, asbestosis in older UK men is still rising.[4]
- The 'Helsinki criteria' suggest that a dose of at least 25 fibre/ml.years is necessary to cause asbestosis.[5] This cumulative exposure metric is similar to the 'pack years' used to measure cigarette smoking; thus, 10 years work in an average airborne concentration of 2.5 fibre/ml is equivalent to one year at 25 fibre/ml. This latter concentration is very high but may readily have been found, for example, in the asbestos textile industry in the 1950's and 1960's. In practice there is rarely any information available about exposure levels in individual cases and the criterion value is used only as a guide.
- Asbestos bodies are fibres, usually of 'blue' or 'brown' asbestos (so-called 'amphiboles') that have become coated in an iron-rich proteinaceous material and are readily visible on light microscopy. If a lung biopsy has been taken then the presence of asbestos bodies is helpful evidence of substantial asbestos exposure; their absence, however, does not rule out a diagnosis of asbestosis.

Box 1. Pleural plaques

Pleural plaques are essentially pathognomonic of asbestos exposure, usually acquired at work even if the patient is unaware of it. Plaques:

- can develop after quite low asbestos exposures, and
- are very common in older men from industrialised areas
- are benign and remain so
- are not responsible for any symptoms or significant loss in lung function
- should not be confused with diffuse pleural thickening
- do not themselves require follow-up
- can induce quite a lot of anxiety
- are not compensated in most countries (in Scotland claims for personal injury can be made)

The true issue for those with plaques is not the plaques *per se* but the fact that they reflect asbestos exposure with its attendant risks. The most important of these is mesothelioma. While plaques do not in themselves increase the risk of mesothelioma, the exposure that gave rise to them does; the size of the risk depends in part on the dose and age of exposure but will usually be around 5%. This is difficult information to impart and, arguably, is unhelpful anyway since there is currently no way of knowing which patients with plaques will develop mesothelioma and no evidence that routine monitoring results in a better prognosis of what is in essence an untreatable malignancy.

Box 2. 'Asbestos' and 'asbestosis'

Most patients (and some doctors) fail to distinguish the term 'asbestos' from 'asbestosis' and it is not uncommon for them to emerge from a clinical discussion about asbestos exposure – perhaps in relation to the finding of pleural plaques on a chest X-ray – with the impression that they have been told they have 'asbestosis'.

Managing asbestosis

The diagnosis of asbestosis is particularly frightening to most people. This is not necessarily warranted, since the prognosis of mild cases is good[6] and in almost all cases is better than the (dismal) prognosis of IPF. In limited disease an appropriate degree of reassurance can be very helpful.

Most patients with asbestosis will be followed-up in secondary care to monitor the radiological and functional progression of their disease; the gap between appointments may be long, for example 12 months, and an interval assessment of symptoms and spirometry can be helpful. There is no effective drug treatment for asbestosis and in most cases no pharmacological treatment is offered – in particular, neither bronchodilators nor corticosteroids (inhaled or oral) are helpful. Some patients may like to take n-acetyl cysteine which has been proven to be of some benefit in IPF and has at least the virtue of being harmless. Advanced cases may benefit from oxygen at home. Severe cases of asbestosis are sometimes treated with lung transplantation, but this is rare in the UK.

The course of the disease depends largely on its extent and on the quantity of asbestos retained in the lungs. In mild cases there is little if any evidence of progression for many years. In others with more advanced disease there is a steady decline towards respiratory failure. A precipitous deterioration, marked by increasing breathlessness and sometime cyanosis on exercise, is usually indicative of secondary pulmonary hypertension. This requires rapid assessment by a specialist service but successful treatment is rare.

A high proportion of those with asbestosis die from lung cancer, reflecting the synergistic effects of smoking and asbestos exposure. It is probably good practice to advise those with asbestosis who smoke to stop doing so, although the benefit in reducing the risk of lung cancer at that stage is uncertain. Around one in 10 patients will develop (and die from) mesothelioma – about the same proportion that will die from asbestosis itself.

Compensation for asbestosis

The provision of compensation for occupational lung diseases varies between different countries. In the UK, for example, patients with asbestosis can claim compensation through a number of routes:

1. Industrial Injuries Disablement Benefit is a statutory payment available to those who have developed a 'prescribed' disease (of which asbestosis is an important example) through their employment. Those who have been exposed through self-employment are ineligible. The process of claim – through the Benefits Agency – is deliberately simple and almost all claimants with asbestosis will be awarded compensation which at its lowest level is a little over £30 weekly, payable for life.

2. Alternatively, or in addition, patients may open a civil case against their employer, usually a previous employer given the long latency of the disease. This is best done through a trade's union or other specialist solicitor and almost always is on a no-win-no-fee basis. Where an employer (or their insurer) cannot be discovered, other avenues for compensation are available by statute. These claims can be quite complex and it is useful to seek advice from a suitably qualified lawyer or one of the many Asbestos Support groups in the UK.

Silicosis

Clinical scenario

A 35-year old stonemason was referred to hospital following routine health surveillance at his work. He reported no respiratory symptoms. Lung function tests had shown an FEV_1 of 5.0L (114% predicted) when he was aged 25 and, although remaining within normal limits, had fallen over the intervening 10 years to 3.3L (85% predicted). A chest X-ray showed a profusion of small nodules in the upper and mid zones consistent with silicosis (Figure 2). He had smoked 20 cigarettes per day from his late teens.

How is the diagnosis made?

Silicosis results from the accumulation of respirable particles of crystalline silica in the lung.[7] Crystalline silica is found in many types of stone, but sandstone is approximately 70% silica and therefore stonemasons – particularly if using angle grinders which generate large quantities of respirable dust – are at high risk of developing silicosis. Other groups at risk are quarry workers and tunnellers, foundry and pottery workers, and construction workers (such as paviers, who frequently cut or break stone, concrete or brick).

Classic silicosis is the most common presentation and typically follows 10-20 years of work during which time the individual often remains asymptomatic. The typical plain chest X-ray appearances are similar to those of coal worker's pneumoconiosis, with a profusion of small nodules in the upper and mid zones. Hilar and mediastinal lymph node enlargement may be present. Non-specific findings include a peripheral blood lymphopenia and raised serum angiotension converting enzyme (ACE), which together with the chest X-ray changes may give rise to confusion with sarcoidosis.

High resolution CT (HRCT) scan (Figure 3) provides greater diagnostic confidence, showing bilateral well-defined 3-5 mm nodules in a centrilobular and subpleural distribution with a posterior bias. In addition, HRCT facilitates the identification of progressive massive fibrosis (coalescence of nodules into irregular masses) and lymph node involvement. Lung biopsy may be considered if the diagnosis remains uncertain on clinical and radiological grounds; in silicosis it shows acellular whorls of hyaline collagen and, when viewed under polarised light, the presence of bi-refringent crystals confirming the presence of silica and other silicates.[8]

This case illustrates that substantial lung damage, including the development of progressive massive fibrosis, may occur without symptoms in young fit workers, and highlights two potential pitfalls in the interpretation of lung function tests. First, although the FEV_1 normally declines with age, it does not significantly do so until the late thirties and so any loss of lung function occurring before this should raise concern. Second, the predicted value represents the average value in healthy subjects of a given age, height and sex and it is possible to lose a significant amount of lung function yet remain within the 'normal' range.

Figure 2. Silicosis: the chest X-ray shows multiple small nodules in the upper zones with evidence of upper lobe fibrosis causing loss of volume in the upper lobes and elevation of the hila

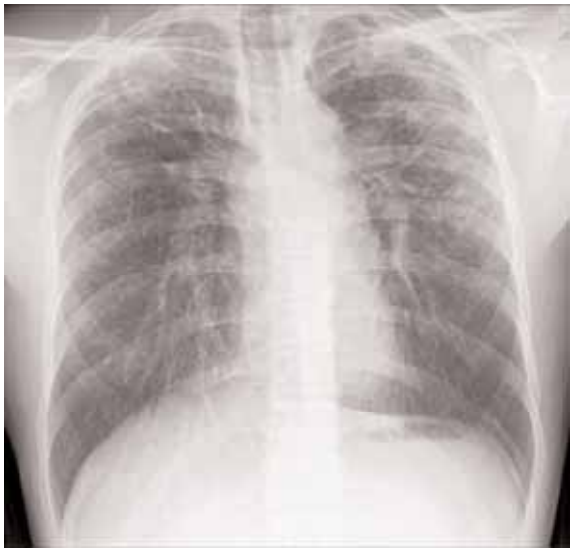
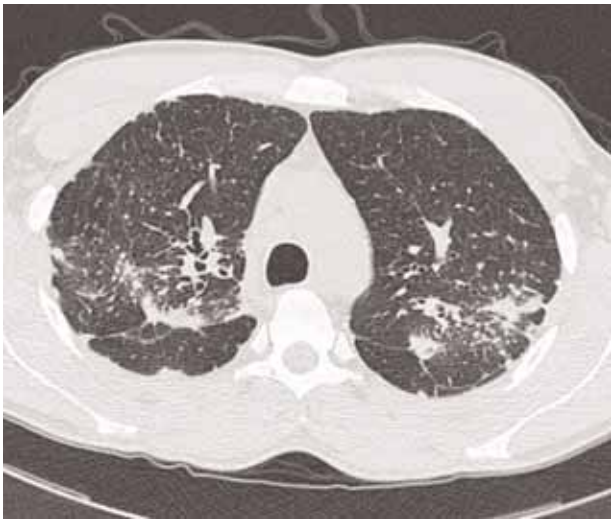


Figure 3. Silicosis; the HRCT shows multiple small pulmonary nodules with a posterior bias and confirms the presence of irregular conglomerate masses consistent with progressive massive fibrosis



Managing Silicosis

Silicosis is invariably progressive, even following complete cessation of exposure, and regular assessment should be undertaken including more detailed lung function testing and serial chest X-rays.[9] As the condition progresses, typical symptoms include cough and shortness of breath occurring on exertion.

There are no effective pharmacological treatments for silicosis. Smoking cessation is particularly important as silica is a carcinogen, and individuals with silicosis are at increased risk of lung cancer.[10,11] The development of silicosis also increases the risk of contracting pulmonary tuberculosis (TB), which is an important consideration in areas where TB is endemic, especially as the typical upper lobe chest X-ray changes of pulmonary TB may be masked by the presence of silicosis.[12] Less common but well recognised complications include the development of connective tissue diseases such as scleroderma and SLE and, rarely, glomerulonephritis. Advanced disease may be complicated by pulmonary hypertension, *cor pulmonale* and respiratory failure, and any evidence of these should trigger a pulmonary transplant assessment.

The patient should be encouraged to inform the company's occupational health team of his diagnosis. In the UK the employer is legally bound to notify the Health and Safety Executive through the Reporting of Injuries, Diseases and Dangerous Occurrences (RIDDOR) mechanism. The diagnosis of silicosis in a worker should trigger a thorough review of the occupational hygiene measures and supply of personal protective equipment in the factory.[13]

Compensation for silicosis

The diagnosis of silicosis invariably represents the end of a career. Companies are reluctant to re-employ stonemasons or others diagnosed with silicosis and doctors should be cautious in advising continued exposure to any form of dust. The implications of this, especially to younger workers, can be devastating, and finding alternative sufficiently remunerative work can be challenging. They should be encouraged to seek compensation from the Industrial Injuries Disablement Benefit scheme (silicosis is a prescribed disease) or the appropriate equivalent in other countries, and informed that they can seek legal advice with a view to pursuing a civil claim, as described above.

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Conflicts of interest The authors declare that they have no conflicts of interest in relation to this article.

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