Interstitial lung disease

Interstitial lung disease (ILD) is a general term for a number of diseases characterised by progressive pulmonary fibrosis or "scarring" of the lungs. The term fibrosis implies formation of abnormal connective tissue within the lung parenchyma. These diseases are less commonly seen than airways diseases such as asthma and chronic obstructive pulmonary disease (COPD). However like COPD, they produce progressive debilitating breathlessness for the patient.

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The main area of the lung affected by interstitial lung disease (ILD) is the interstitium — the area between the alveolar epithelium and capillary endothelium. Other areas involved include the tissues that make up the framework of the lungs, airways, vasculature and alveolar air spaces. The deposition of abnormal connective tissue causes the lung to become stiff. This may also be referred to as "restriction" and will produce a restrictive pattern on spirometry. Impairment of the transfer of oxygen from the air to the blood across the alveolar membrane ("gas transfer") also occurs due to the deposition of connective tissue. In early disease, this is most troublesome during exercise.

The incidence and severity of fibrotic lung disease increase with age. Although the exact mechanisms of these increases are not clear,[1] prolonged exposure to reactive oxygen species contribute to ageing and that causes oxidant stress that is thought to be involved in the fibrotic response.

Different studies have confirmed that idiopathic lung fibrosis (IPF) is predominantly a disease of older patients, with a mean age of presentation of 60–65 years.[2] The incidence and prevalence of the disease are not precisely known.

IPF-related mortality seems to be higher in the elderly, although studies on this subject are rare. In one study,[3] the median survival time was significantly shorter in older patients (60–70 years of age, 27·2 months; 70 years of age, 14·6 months) compared with younger patients (50 years of age, 116·4 months; 50–60 years of age, 62·8 months).

Causes

A range of occupations and illnesses can cause ILD (Box 1), but there can also be no known

Box 1: Causes

Connective tissue disorder (eg, rheumatoid arthritis)

Drugs: more than 200 have been implicated (eg, chemotherapy, amiodarone, and antibiotics)[4]

Radiotherapy

Inorganic dust disease (eg, pneumoconioses such as asbestosis or silicosis)

Organic dust disease (eg, extrinsic allergic alveolitis)

Others sarcoidosis or Langerhans cell histiocytosis

Vasculitides.

cause. ILD of unknown cause is usually termed idiopathic pulmonary fibrosis, the most common of which is called usual interstitial pneumonia (UIP).[5]

The most aggressive type of ILD is idiopathic pulmonary fibrosis (IPF) and usually affects people aged 60 years and over. IPF is particularly challenging in elderly patients (>80 years),[4] whose tolerance to drugs is lower compared with younger patients.

Older people have been reported to be more susceptible to vasculitis related lung disease.[6,2] Large case series have revealed that advancing age is a risk factor for mortality in ANCA-associated vasculitis, independent of renal involvement.[2,7,8]

A study has reported that pulmonary involvement is significantly more common and more severe in patients aged over 65 years. The prevalence of interstitial fibrosis, as assessed by chest radiography or high-resolution computed tomography (HRCT), is also significantly higher in this age group.[9]

Diagnosis

ILD usually presents in mid-to-late life and is equally prevalent in men and women.[4] Breathlessness on effort is typically the presenting symptom. This usually becomes progressively worse and ultimately the patient may become breathless at rest. Another, less common symptom is a dry non-productive cough, and wheezing is not prominent.

The patient's history, including their occupation current medications, and previous illnesses, can provide clues to the cause of the ILD. In elderly patients, signs and symptoms of ILD can be easily mistaken for manifestations (or worsening) of other conditions more common in the elderly. For instance, in patients with concomitant chronic obstructive pulmonary disease (COPD), the appearance of radiographic nodules may be mistaken for a neoplasm or infection. Or in those with known heart failure, new dyspnoea and pulmonary infiltrates could be erroneously attributed to the heart failure.

Investigations

In spirometry, ILD causes a restrictive pattern that should be contrasted with the results seen in airways disease (ie obstruction to airflow). For detailed measurement of lung function, the patient should be referred to a specialist clinic. However, spirometry can clearly distinguish between two patterns: obstruction (airways disease) and restriction (ILD).

Chest radiograph (CXR) typically shows changes of bilateral, mostly peripheral, shadowing. In advanced disease, the lungs look much smaller than normal and "honeycombing" may be seen.

HRCT can be used to confirm the presence of ILD and can be used to identify the precise type of disease. However, the "gold standard" test for ILD is lung biopsy, which is used for the histological examination.

Management

The prognosis depends on a number of factors. If the disease is due to an identifiable external factor, such as a drug, environmental allergen or an occupation, removal of the factor can prevent further damage and in some cases complete recovery follows. Conversely, in diseases of unknown cause, especially UIP, the progress is inexorable and can be worse than some forms of cancer, with typical survival of less than five years.

Death is usually due to respiratory failure. Supportive care is key for those with progressive disease. The use of home oxygen is important if the patient is limited by breathlessness and is in respiratory failure. Other measures include stopping smoking, pulmonary rehabilitation and regular vaccination programmes.

Opioids such as oral morphine are used as palliative measures with severe breathlessness. Unfortunately there are no effective treatments for some forms of pulmonary fibrosis, although steroids and immunosuppressive drugs help some patients.

Drug treatments

- Corticosteroids are valuable for some diseases, such as reactions to environmental allergens and sarcoidosis. However, their role in many diseases, especially UIP, is questionable. Biopsies and CT scanning can be used to guide the likelihood of a response to steroid therapy
- Cytotoxic drugs (for example, azathioprine and cyclophosphamide) have been used in conjunction with steroids as "steroid sparing" agents. There is limited evidence to support their use. Older patients receiving cytotoxic agents, such as cyclophosphamide, are particularly prone to the development of malignancies

and cytopenia, which, in turn, place them at risk of infection or bleeding[5]
- A range of drugs affecting various aspects of the pathological process underlying ILD have been evaluated. In particular, agents that might affect the fibrotic process are under extensive investigation.

Non-drug measures are an integral part of management of non-malignant respiratory disease. The care of ILD patients should include support, symptom control and communication. Primary care nurses who have regular contact with the patient and who have built up a relationship with the patient and family are best placed to ensure access to a package of care that meets their changing needs and condition.

Symptom management

Low doses of opiates can be effective for relief of breathlessness, and benzodiazepines and other anxiolytics have a place in managing breathlessness in patients with associated anxiety and panic attacks.

Ambulatory oxygen therapy may improve exercise performance. Patients with ILD will often ultimately need continuous high-flow oxygen therapy.

Cough may be troublesome. Medication can include conventional antitussive drugs such as codeine linctus and oral opiates.

For depression and anxiety, which are often associated with dyspnoea, antidepressants, counselling, relaxation and complementary therapies may be helpful.

Muscle weakness and fatigue accompany the immobility associated with ILD, and patients may benefit from pulmonary rehabilitation.

End-of-life decisions

Advanced directives/living wills and preferences and priorities of care allow patients to maintain a degree of control by allowing them to express their wishes about treatment and care before the disease progresses. Given the relatively slow progression of many forms of ILD, these issues can be introduced and discussed during patient follow-up.

Sources of support

An aspect of supportive care is referral to the appropriate resources and multidisciplinary working, to ensure that patients have the appropriate level of care to meet their needs and that of their families as their condition changes.

- Respiratory specialist nurse practitioners provide specialist advice and support to patients, carers and other healthcare professionals
- Social services provide homecare to support daily activities of living, respite care, day care and nursing/residential care
- Occupational therapists are helpful for the independent living aids, adaptations to home and lifestyle adjustment advice
- Physiotherapists/chest physiotherapists can help patients with breathing recovery strategies, breathing exercises and mobility maintenance
- The British Lung Foundation (BLF) provides information/leaflets on specific respiratory conditions, a patient advice line (08458 50 50 20) and a useful website (www.lunguk.org).

Conclusion

Patients with suspected with pulmonary fibrosis in primary care need early urgent referral for specialist review. Diagnosis is made with a high-resolution CT scan, but often no underlying cause is identified. Treatments such as steroids may provide some relief of symptoms, but have a number of side effects. The mainstay of treatment is supportive care, and palliative care planning is key in patients who are rapidly declining.

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