MEDICINE

432 Team

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Extra: Interstitial Lung Diseases and Lung Cancer

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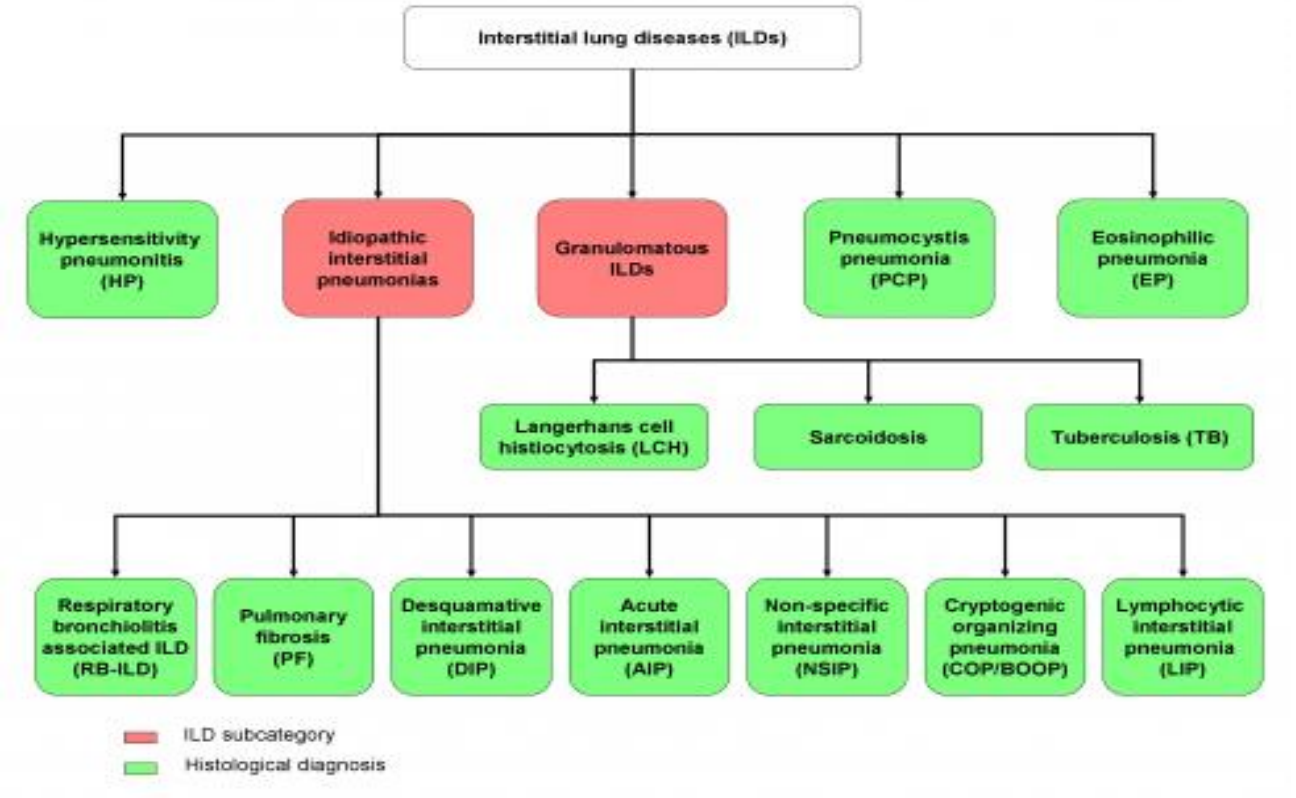
COLOR GUIDE: • Females' Notes • Males' Notes • Important • Additional

Objectives

Not Given :P

Interstitial Lung Diseases:

Diffuse parenchymal lung disorders (DPLD, also referred to as interstitial lung diseases) are a heterogeneous group of disorders accounting for about 15% of respiratory clinical practice. There is diffuse lung injury and inflammation that can progress to lung fibrosis. The classification is shown in the table.



I. Granulomatous lung disease:

A granuloma is a mass or nodule composed of chronically inflamed tissue formed by the response of the mononuclear phagocyte system (macrophage/histiocyte) to an insoluble or slowly soluble antigen or irritant. If the foreign substance is inert (e.g. an inhaled dust), the phagocytes turn over slowly; if the substance is toxic or reproducing, the cells turn over faster, producing a granuloma. A granuloma is characterized by epithelioid multinucleate giant cells, as seen in tuberculosis. Granulomas are also seen in other infections, including fungal and helminthic, in sarcoidosis, and in hypersensitivity pneumonitis, and can also be due to foreign bodies (e.g. talc).

A. Sarcoidosis

Sarcoidosis is a multisystem granulomatous disorder, commonly affecting young adults and usually presenting with bilateral hilar lymphadenopathy, pulmonary infiltration and skin or eye lesions. Beryllium poisoning can produce a clinical and histological picture identical to sarcoidosis, though contact with this element is now strictly controlled.

Epidemiology and aetiology:

Sarcoidosis is a common disease of unknown aetiology that is often detected by routine chest X-ray. There is great geographical variation. The prevalence in the UK is approximately 19/100,000. Sarcoidosis is common in the USA but is uncommon in Japan.

Clinical features:

Sarcoidosis can affect many different organs of the body. The most common presentation is with respiratory symptoms or abnormalities found on chest X-ray (50%). Less common presentations include fatigue or weight loss (5%), peripheral lymphadenopathy (5%) and fever (4%). Neurological presentations are rare but well recognized and can mimic a variety of conditions. Chest X-ray may be normal in up to 20% of non-respiratory cases, though pulmonary lesions may be detected later. There are four stages of pulmonary involvement based on radiological stage of the disease, which is helpful in prognosis:

- ***Stage I:*** bilateral hilar lymphadenopathy (BHL) alone
- ***Stage II:*** BHL with pulmonary infiltrates
- ***Stage III:*** pulmonary infiltrates without BHL
- ***Stage IV:*** fibrosis.

Investigations:

- ✓ ***Imaging***. Chest X-ray
- ✓ High-resolution CT is useful for assessment of diffuse lung parenchymal involvement.
- ✓ ***Full blood count***. There may be a mild normochromic, normocytic anaemia with raised ESR.
- ✓ ***Serum biochemistry***. Serum calcium is often raised and there is hypergammaglobulinaemia.
- ✓ ***Transbronchial biopsy*** is the most useful investigation, with positive results in 90% of cases of pulmonary sarcoidosis with or without X-ray evidence of lung parenchymal involvement.

- ✓ ***Serum angiotensin-converting enzyme (ACE) level*** is raised by two standard deviations above the normal mean value in over 75% of patients with.

Treatment:

Persisting infiltration visible on the chest X-ray with normal lung function tests should be monitored carefully. Patients with abnormal lung function tests are unlikely to improve without corticosteroid treatment. If the disease is not improving spontaneously 6 months after diagnosis, treatment should be started with prednisolone 30 mg for 6 weeks, reducing to alternate-day treatment with prednisolone 15 mg for 6–12 months.

B. Granulomatous lung disease with vasculitis:

The classification of pulmonary vasculitis and granulomatous disorders is unsatisfactory. In broad terms there are two main groups: the respiratory manifestations of systemic diseases, and disorders associated with the presence of anti-neutrophil cytoplasmic antibodies (ANCAs). The most common types:

- ➢ Wegener's granulomatosis (granulomatosis with polyangiitis)
- ➢ Churg–Strauss syndrome

II. Idiopathic interstitial pneumonia (IIP):

(IIP is characterized by diffuse inflammation and fibrosis in the lung parenchyma)

A.Idiopathic pulmonary fibrosis (IPF)

This is also known as *usual interstitial pneumonia* (UIP) and was previously known as cryptogenic fibrosing alveolitis (CFA). It is relatively rare with a prevalence of about 20/100 000 population, mean onset is in the late 60s and it is more common in males. The cause is unknown but possible contributory factors include cigarette smoking, chronic aspiration, antidepressants, wood and metal dusts and infections, e.g. Epstein–Barr virus.

Clinical features:

The main features are progressive breathlessness, a non-productive cough and cyanosis, which eventually lead to respiratory failure, pulmonary hypertension

and cor pulmonale. Fine bilateral end-inspiratory crackles are heard on auscultation and gross finger clubbing occurs in two-thirds of cases.

Investigations:

- ✓ ***Chest X-ray***
- ✓ ***High-resolution CT***
- ✓ ***Full blood count***.
- ✓ ***Serum biochemistry***.
- ✓ ***Biopsy***
- ✓ ***Respiratory function tests***

Prognosis and treatment:

The median survival time for patients with IPF is approximately 5 years, although mortality is very high in the more acute forms. Treatment with prednisolone (30 mg daily) is usually prescribed for disabling disease although it produces little benefit.

B. Desquamative interstitial pneumonia (DIP)
C. Respiratory bronchiolitis interstitial lung disease (RBILD)
D. Acute interstitial pneumonia (AIP)
E. Nonspecific interstitial pneumonia (NSIP)

III. *Other types of diffuse lung disease*

Goodpasture's syndrome

This disease often starts with symptoms of an upper respiratory tract infection followed by cough and intermittent haemoptysis, tiredness and eventually anaemia, although massive bleeding may occur. The chest X-ray shows transient blotchy shadows that are due to intrapulmonary haemorrhage. These features usually precede the development of an acute glomerulonephritis by several weeks or months. The course of the disease is variable: some patients spontaneously improve while others proceed to renal failure.

TUMOURS OF THE RESPIRATORY TRACT:

I. Bronchial carcinoma

- Bronchial carcinoma is the most common malignant tumour worldwide, with around 1.4 million deaths annually.
- It is the third most common cause of death in the UK after ischaemic heart disease and cerebrovascular disease and is now the commonest cause of cancerrelated death in both men and women.
- Rates are declining in men but still increasing overall reflecting increasing incidence in women.
- Cigarette smoking (including passive smoke exposure) accounts for >90% of lung cancer. There remains a higher incidence of bronchial carcinoma in urban compared with rural areas, even when allowance is made for cigarette smoking.

II. Lung Cancer:

Pathophysiology:

Historically, lung cancers are broadly divided into small cell carcinoma and non-small cell carcinoma based upon the histological appearances of the cells seen within the tumour. This distinction is necessary with respect to the behaviour of the tumour, providing prognostic information and determining best treatment. Non-small cell carcinoma is further divided into a number of cell types (adenocarcinoma, squamous cell carcinoma, large cell carcinoma, large cell neuroendocrine).

Clinical features:

The presentation and clinical course vary between the different cell types (Table 15.26). Symptoms and signs may vary depending on the extent and site of disease. Common presenting features can be divided into those caused by direct/local tumour effects, metastatic spread and non-metastatic extrapulmonary features.

Table 15.26 Lung cancer cell types and clinical features

Cell type	Incidence in UK (%)	Features
Squamous cell carcinoma	35	Remains the most common cell type in Europe Arises from epithelial cells, associated with production of keratin Occasionally cavitates with central necrosis Causes obstructing lesions of bronchus with post-obstructive infection Local spread common, metastasizes relatively late
Adenocarcinoma	27–30	Likely to become the most common cell type in the UK in the near future (most common cell type in the USA) Increasing incidence over last 10 years possibly linked to low tar cigarettes Originate from mucus-secreting glandular cells Most common cell type in non-smokers Often causes peripheral lesions on chest X-ray/CT Subtypes include bronchoalveolar cell carcinoma (associated with copious mucus secretion, multifocal disease) Metastases common: pleura, lymph nodes, brain, bones, adrenal glands
Large cell carcinoma	10–15	Often poorly-differentiated Metastasize relatively early
Small cell carcinoma	20	Arise from neuroendocrine cells (APUD cells) Often secrete polypeptide hormones Often arise centrally and metastasize early

Local effects

- Cough
- Breathlessness
- Haemoptysis
- Chest pain
- Wheeze
- Hoarse voice

Treatment:

Treatment of lung cancer involves several different modalities and should be planned by a multidisciplinary team. Unfortunately, the majority of patients have incurable disease at presentation, or have significant co-morbidities which preclude radical treatment. The most important techniques are:

A. Surgery
B. Radiation therapy
C. Chemotherapy
D. Laser therapy, endobronchial irradiation and tracheobronchial stents

This is our last lecture in this semester, we apologize for being late and for the mistakes.

Good Luck 432

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