

INTERSTITIAL LUNG DISEASE

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Abstract: ILD is a group of diseases affect interstitium of the lung. Repeated insult to the lung cause the interstitium to be damaged. Similarly occupational history of great concern in this regard. The major presentation is chronic cough and history of progressive dyspnea. It is diagnosed by HRCT chest. Honey comb shadows mostly in lower lungs are found on HRCT. Scan chest. Treatment is according to the cause, including steroids, Immunosuppressants.

Key words: Interstitium, Fibrosis, Honey comb pattern, HRCT

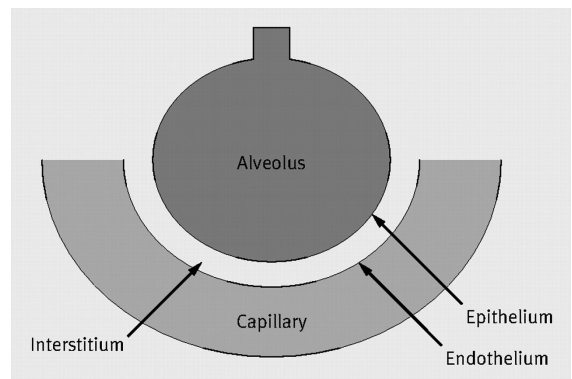
Definition:

Interstitial lung disease[ILD] is a group of diseases that affects interstitium of the lung, it is also known by the term diffused parenchymal lung disease [DPLD].

Interstitium is a lace like tissue network around air sacs of lungs and provides support to the lungs, small blood vessels runs through this network exchange of gases takes through this interstitium between air sacs and blood vessels.

Interstitium is not visible on X-Rays or CT scan.

So any pathology or injury of lungs affecting interstitium will be labelled under ILD.



Interstitial tissue of lungs is present between alveoli and pulmonary blood vessels.

Pathogenesis:

Repeated injuries due to exogenous or endogenous stimuli causes the lungs to generate enough amount of tissue to repair its damaged parts, but some time the repair process does not stop causing the tissue around air sacs and blood vessels to become thickened, scarred & fibrosed. Thus this makes exchange of gases between air-sacs and blood vessels difficult.

Clinical Presentation:

There is a progressive history of dyspnoea,

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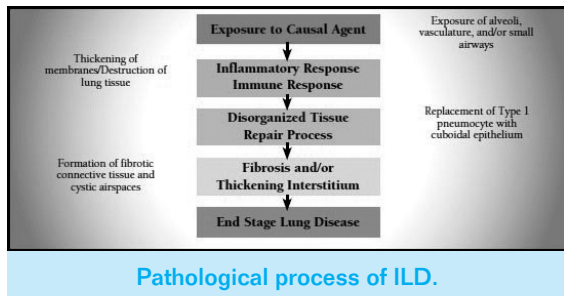
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and cough which is usually dry and non-productive, there is finger clubbing and in advanced stages central cyanosis, and on auscultation there is bilateral basal fine late inspiratory crackles.

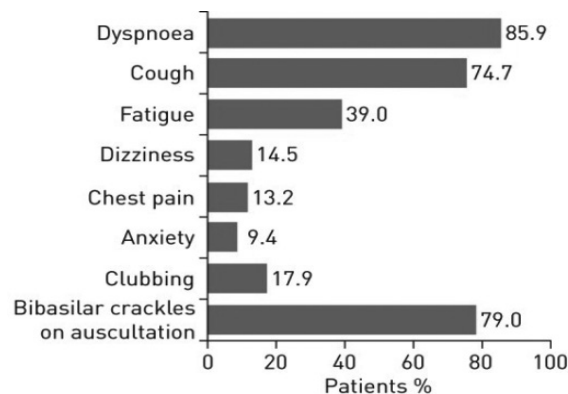
COMPLICATIONS

Long standing disease can cause other systemic complications.



1) GERD:

[Gastro-esophageal reflux disease] Recent studies suggest that GERD is associated with rapid progression of Idiopathic pulmonary fibrosis which is a specific form of ILD.



2) Pulmonary hypertension:

Pulmonary hypertension develops due to chronic ILD, this might lead to dysfunction of right sided heart (Cor-Pulmonale).

3) Respiratory Failure:

It develops in end stage ILD due to hypoxemia.

INVESTIGATIONS

Complete Blood Count (CBC):

Eosinophills are usually raised.

ESR:

ESR is raised

LDH:

LDH is raised in pneumonitis.

Pulmonary Function Tests:

PFT's show restrictive defect and reduced lung volumes and gas transfer.

Fibro-Optic Bronchoscopy:

Bronchoscopy & bronchoalveolar-lavage is done to confirm diffused alveolar hemorrhages in ILD.

Lung Biopsy:

It is done to confirm type of ILD like granulomatous disease like sarcoidosis and berylliosis.

Chest X-Ray:

It shows bi-basal retico-nodular opacities, and in some cases ground glass appearance in the lower lobes.

HRCT:

(High Resolution – Computed Tomography) HRCT shows honey-combing scarred tissue.

CAUSES

1) Inhaled Substances

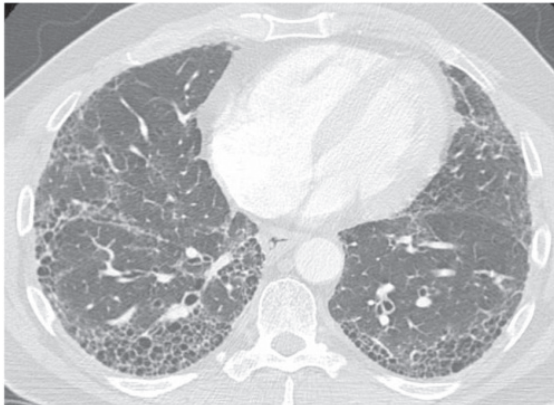
A) Inorganic:

- Silicosis: caused by inhalation of free or crystalline silica, and manifests after 10 to 20 years of continuous exposure.
- Asbestosis: caused by exposure to microscopic asbestos mineral fibers.
- Berylliosis : Caused by esposure to beryllium and its compound.

- Industrial Printing Chemicals: eg Black carbon(Coal), Iron oxide , Tin Oxide.

B) Organic:

- Hypersensitivity Pneumonitis: caused by immunological mechanism in reaction to organic antigens. examples are
- Farmer's Lung : Caused by mouldy hay or straw containigatigen like aspergillus fumigatus.
- Bird Fancier's Lung: caused by exposure to avium serum protiens in bird's excreta.
- Byssinosis: Caused by exposure to cotton, flax and hemp dust in textile industry.



HRCT-lung shows honey-combing appearance and peripheral reticulation

2) Drug Induced:

Most frequently caused by following drugs

- Bleomycin.
- Methotrexate.
- Amiodarone.
- Nitrofurantoin.

3) Connective-Tissue and Inflammatory Diseases affecting Lungs:

eg,

- Rheumatoid Arthritis.
- Systemic Sclerosis.

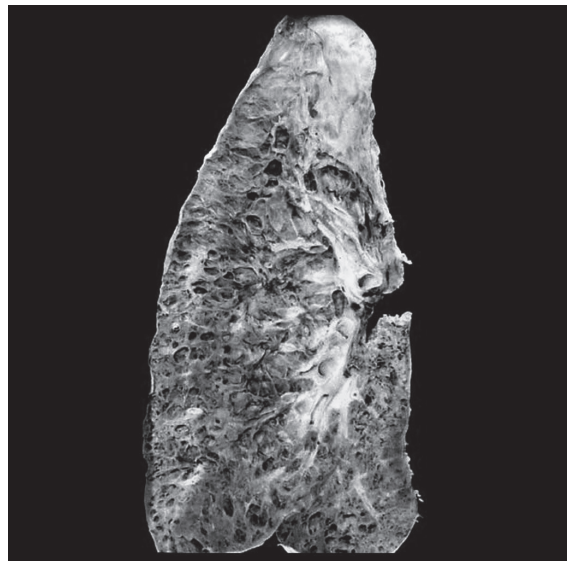
- SLE (Systemic Lupus Erythematosus).
- Polymyositis

4) Idiopathic:(due to unknown cause)

- Idiopathic Pulmonary fibrosis.(IPF)
- Non-specific interstitial pneumonia. (NSIP)

5) Grannulomatous Disease:

- Sarcoidosis: It is a granulomatous disorder marked by presence of non-ceasating granulomas.

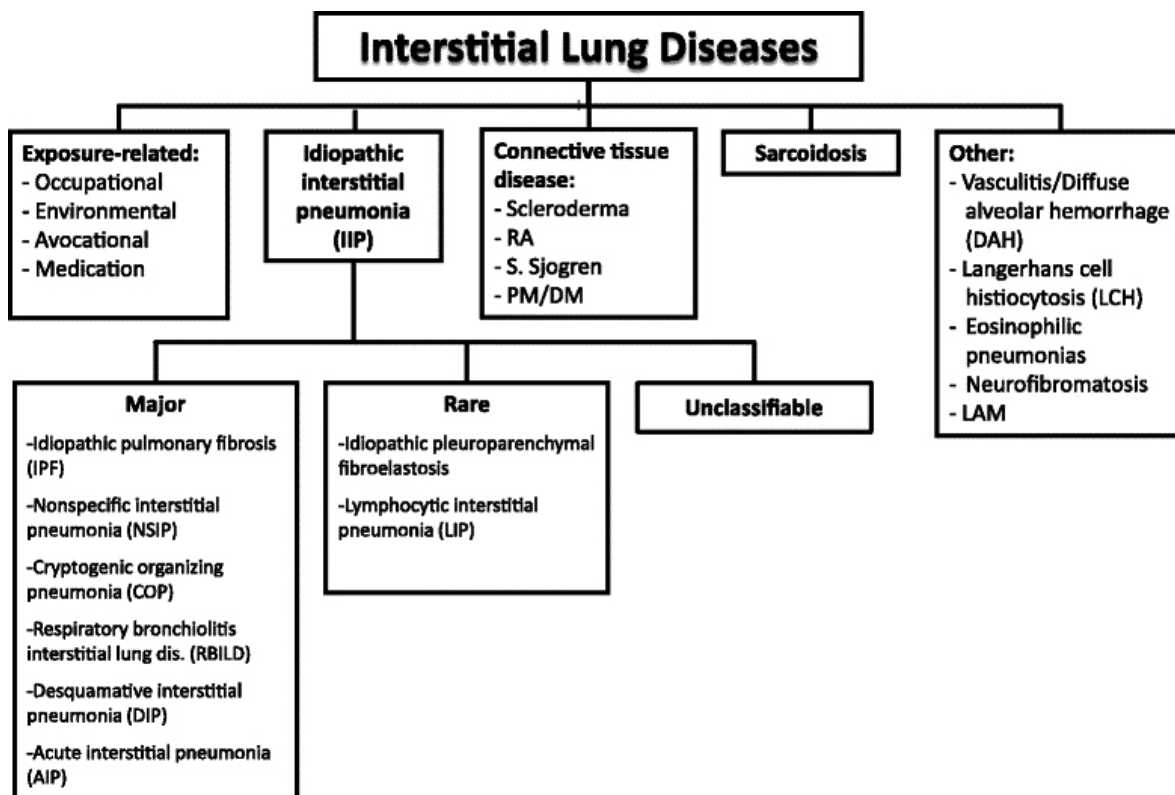


Cross-Section of lung with ILD showing peripheral honeycomb like scar tissue

Differential Diagnosis:

Conditions which mimic the sign and symptoms of interstitial lung disease are as follows

- Viral Pneumonia.
- T.B (Tuberculosis pulmonary).
- Bronchioalveolar carcinoma.
- Pulmonary edema.
- Aspiration Pneumonitis.



Above table shows ILD's grouped according to different causes.

MANAGEMENT

ILD is not a single disease, but encompasses many different pathological diseases, so treatment differs for every cause.

1) Eliminating exposure to causative agent:

- If a specific occupational exposure cause like coal or silica or asbestos then person should avoid that environment, and if possible change profession to get rid of external stimuli causing harm to the lungs.
- If any of the drug is causative agent it should be discontinued.

2) Oral Antibiotics and Corticosteroids:

- Idiopathic and connective tissue based

causes should be treated with oral corticosteroids e.g Prednisolone which is given as 0.5mg per body weight. Corticosteroids reduces the inflammation of lung causing scarring.

- Anti-Biotic therapy with Azithromycin should be started in patients with interstitial pneumonias.

3) Immuno-suppressants:

- Immunosuppression therapy with Azathioprine (2-3mg/kg) is given in patients which are highly symptomatic.

4) Oxygen Therapy:

- Patients with low level of Oxygen in blood (hypoxemia) should be given supplemental oxygen.

5) Lung Transplant:

- Lung-Transplantation should be considered in patients with progressive ILD causing severe lung impairment despite continuous therapy.

Reference:

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