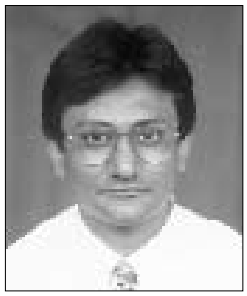

# Interstitial Lung Diseases - Imaging

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## ABSTRACT

Interstitial lung diseases (ILDs) are commonly seen in practice and encompass a large number of conditions ranging from idiopathic interstitial pneumonias to occupational lung diseases and drug reactions.

Radiology is one of the cornerstones of management. High-resolution CT (HRCT) has changed the way we approach ILDs. The diagnosis, classification and prognosis can all be assessed using HRCTs. Plain radiographs also help, especially in the initial assessment of ILDs.

## DEFINITION

The interstitium is defined as a continuum of loose connective tissue throughout the lung consisting of three subdivisions:

a. Axial (bronchovascular), surrounding the bronchovascular bundles from the hila to the secondary pulmonary lobules
b. Parenchymal (acinar), which is situated between the alveolar and capillary membranes, and
c. Subpleural, which is situated between the pleura and the lung parenchyma and is continuous with the interlobular septa and perivenous interstitial space

Interstitial diseases are diseases which affect the interstitium and the alveolar spaces.

## ETIOLOGY

There are over 250 diseases which are known to affect the interstitium. These include infections such as miliary tuberculosis, granulomatous disorders such as sarcoidosis, neoplasms such as lymphangitis carcinomatosis and lymphoma, other systemic diseases such as rheumatoid arthritis and scleroderma, many of the pneumoconiosis and toxic conditions related to drugs or radiation. In such a situation, it becomes sometimes very difficult to sift through the chaff.

However, the following nine conditions account for over 90% of ILDs seen in practice in the Western countries. With the addition of two more, to account for Indian conditions, we have just eleven conditions accounting for the majority of ILDs. The conditions are:

1. Idiopathic interstitial pneumonia and its subtypes, including cryptogenic organizing pneumonia
2. Sarcoidosis
3. Hypersensitivity pneumonitis
4. Eosinophilic granuloma
5. Asbestosis
6. Silicosis

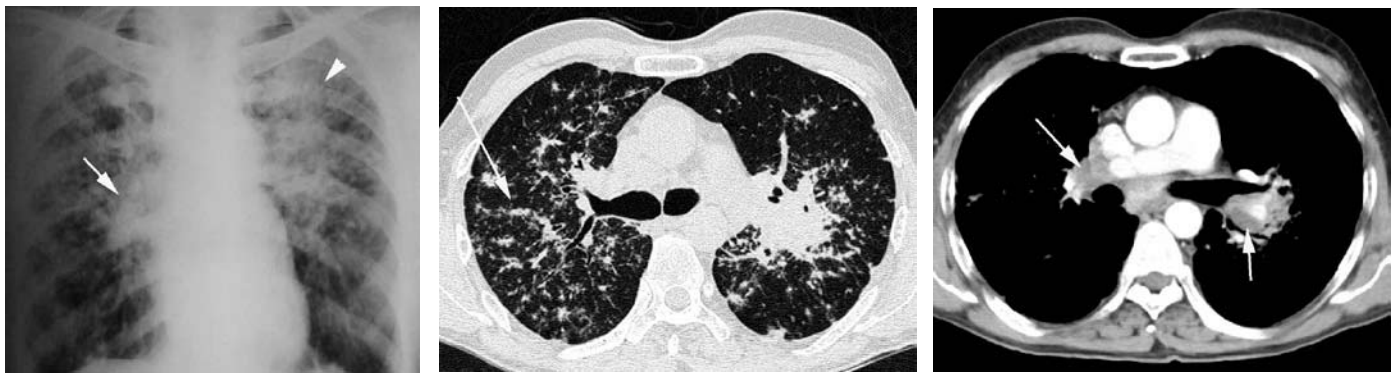

**Fig. 1 (A-C) :** Sarcoidosis. Plain radiograph (A) shows a reticulo-nodular pattern of disease (arrowhead) with an upper zone distribution pattern, with bilateral hilar lymphadenopathy (arrow). HRCT (B) shows a peribronchovascular pattern of nodules (arrow). Contrast-enhanced CT (C) shows bilateral hilar adenopathy (arrow).

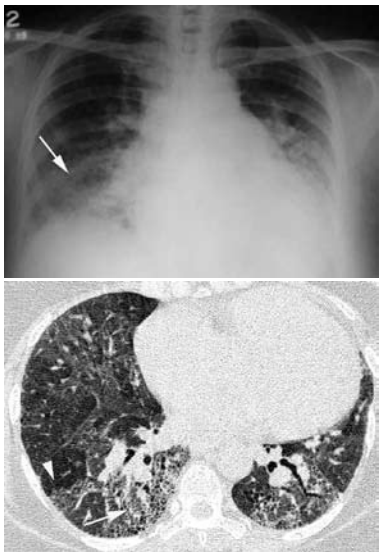

**Fig. 2 (A,B) :** Usual interstitial pneumonia. Plain radiograph (A) shows a reticular pattern of disease (arrow) with lower zone predominance and low volume lungs. HRCT (B) shows honeycombing (arrow) with intralobular interstitial thickening (arrowhead).

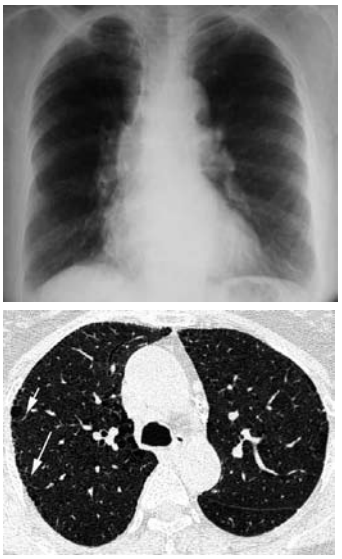

**Fig. 3 (A,B) :** Idiopathic interstitial pneumonia. Plain radiograph (A) is normal. HRCT (B) shows subtle honeycombing and intralobular interstitial thickening (arrows).

7. Lymphangitis carcinomatosis
8. Pulmonary edema
9. Drug-induced
10. Miliary tuberculosis
11. Tropical eosinophilia

## CLINICAL PRESENTATION

Radiologists see patients of ILD in two clinical settings:

a. To rule out the presence of ILD in a patient with a known predisposing factor or occupational disorder
b. To rule out / confirm and to characterize ILD in a patient with breathlessness and restrictive lung disease on PFTs

## RADIOLOGY OF ILD

The modalities involved are

I. Plain radiographs
II. CT
III. MR

### Plain Radiographs

On plain radiographs, two aspects need to be looked at:

A. The recognition of ILD
B. Characterization of ILD

***Recognition of ILD***

This is based on the following parameters

1. The presence of an "interstitial" pattern
2. Repeated "expiratory" films
3. Experience and expertise

Often, the only way interstitial disease is diagnosed is because of the experience of the observer

An interstitial pattern consists of one of the following appearances

a. Reticular
b. Reticulo-nodular
c. Nodular

***Characterization of ILD***

This is based on the following parameters

a. Pattern of interstitial disease
b. Distribution of ILD
c. Associated features
   i. Lung volume
   ii. Lymph nodes
   iii. Pleural effusion
   iv. Pericardial effusion
   v. Esophageal dilatation
   vi. Pneumothorax
   vii. Arthritis

*a. Pattern of interstitial disease*

This can be either "reticular", "nodular" or "reticulo-nodular" (Fig. 1A). Nodular diseases include miliary tuberculosis, silicosis, miliary metastases and sarcoidosis. The rest of the diseases can have either a reticulo-nodular or a reticular appearance.

*b. Distribution of disease*

This helps in the following way

*i. Central v/s peripheral*

Centrally distributed diseases (along the bronchovascular space), are usually pulmonary edema, sarcoidosis or lymphangitis carcinomatosis (Fig. 1A). The rest of the diseases are peripherally distributed.

*ii. Upper zone v/s mid-lower zone*

If the disease is in the upper zones, the differentials are restricted to tuberculosis, silicosis, eosinophilic granuloma, end-stage hypersensitivity pneumonitis and

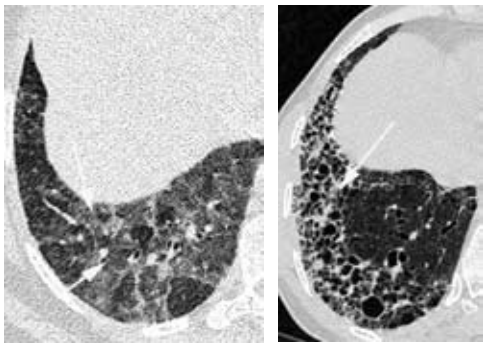

**Fig. 4 (A,B):** Prognosis. HRCT (A) shows ground-glass attenuation (arrow) suggesting alveolitis. This usually carries a good prognosis and suggests that there will be good response to treatment. HRCT (B) shows extensive honeycombing (arrow) suggesting an end-stage lung, which is unlikely to respond to treatment and carries a poor prognosis.

ankylosing spondylitis (Fig. 1A). The rest of the ILDs have mid and lower zone presentations (Fig. 2A).

*c. Lung volumes*

Increased or normal lung volumes (Fig. 1A) in the presence of significant disease are seen in sarcoidosis, eosinophilic granuloma and lymphangioleiomyomatosi. The rest of the ILDs, show reduced lung volumes (Fig. 2A).

*d. Lymphadenopathy*

The presence of lymphadenopathy restricts the diagnosis to sarcoidosis (Fig. 1A), silicosis, lymphoma or berylliosis. Tuberculosis and metastases are the other differentials

*e. Other features*

i. Pleural effusion - RA, lymphoma
ii. Pericardial effusion - lymphoma, SLE
iii. Esophageal dilatation - scleroderma
iv. Pneumothorax - eosinophilic granuloma, lymphangioleiomyomatosis
v. Arthritis - RA, scleroderma

## CT

CT (high resolution CT- HRCT) is an invaluable modality for the diagnosis of interstitial lung disease (ILD).[1-4]

### HRCT

It is a technique which utilizes a special bone algorithm which enhances edges. Other parameters include the use of thin sections and a reduced field of view. Special lung windows are used for optimal detection of pathology.

HRCT answers the following questions:

1. Is there ILD?
2. If there is ILD, then what is it?
3. Is there acute or active disease?
4. Where should the biopsy be performed?
5. Has there been change?

***1. Is there ILD?***

HRCT is an extremely sensitive technique for confirming the presence or absence of ILD in patients with pre-disposing diseases or symptoms / signs (Fig. 3). In a small percentage of patients of emphysema which presents with restrictive lung disease on PFTs, HRCT diagnoses the emphysema as well.

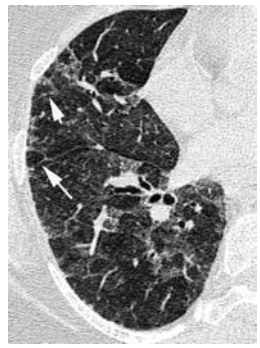

**Fig. 5 :** Non-specific interstitial pneumonia. HRCT shows septal thickening (arrows).

***2. If there is ILD, what is it?***

Using various criteria and signs (discussed below), HRCT allows some characterization of the disease process, often narrowing down the differential diagnosis and often allowing a precise diagnosis to be made (Fig. 1, 2).

Studies[3,4] have conclusively shown that in idiopathic interstitial pneumonias and in cystic lung diseases, HRCT has over 70% accuracy for making a diagnosis. It is able to differentiate between various types of idiopathic interstitial pneumonias as well.

***3. Is there acute or active disease?***

On the basis of the presence of ground-glass attenuation which suggests alveolitis, HRCT allows this distinction to be made - this has an important bearing on therapy and the expected response to drugs (Fig. 4).

***4. Where should the biopsy be performed?***

HRCT reliably allows accurate localization of the segment where the biopsy has to be performed.

***5. Has there been a change?***

Follow-up HRCTs allow monitoring of disease progress on treatment

### *HRCT Anatomy*

HRCT allows the visualization of the secondary pulmonary lobule which is the smallest unit which can be visualized with this technique

### *Secondary Pulmonary Lobule*

This is an anatomic unit consisting of 2-3 terminal bronchioles and their respiratory bronchioles, alveolar ducts and alveoli. It measures 1 - 2.5 cm in diameter and is supplied by a small bronchiole and its accompanying arteriole. The lobule is marginated by interlobular septae on all sides and in the periphery by the subpleural interstitium at one edge. The interlobular septae contain lymphatics and venules.

### *HRCT Signs*

The following signs aid in the diagnosis of ILDs

1. Septal thickening
2. Nodules
3. Cysts

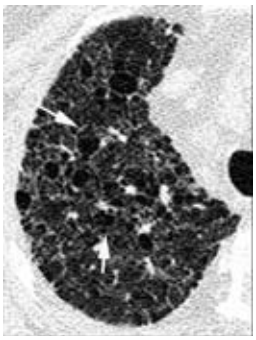

**Fig. 6 :** Langerhan's cell histiocytosis. HRCT shows extensive cystic change (arrows).

4. Ground-glass attenuation

*1. Septal thickening*

Thickening of the interlobular septae (Fig. 5) results in septal lines. This thickening could be due to fluid or cells and occurs in

i. Usual interstitial pneumonia
ii. Asbestosis
iii. Lymphangitis
iv. Pulmonary edema
v. Alveolar proteinosis

*2. Nodules*

Nodules (Fig. 1B) may be randomly distributed as in miliary tuberculosis or along the bronchovascular bundles as in sarcoidosis

They occur in the following conditions

i. Metastases
ii. Sarcoidosis
iii. Miliary tuberculosis
iv. Lymphangitis
v. Silicosis
vi. Hypersensitivity pneumonitis
vii. Eosinophilic granuloma

*3. Cysts*

Cysts (Fig. 4B, 6) occur as part of end-stage disease due to intra-alveolar cystic degeneration and traction bronchiectasis. However cystic disease per se is not very common and occurs in

i. Eosinophilic granuloma, and
ii. Lymphangioleiomyomatosis

*4. Ground-glass attenuation*

This is defined as focal increased opacity (Fig. 4A) within the lung parenchyma, but without obscuration of the vessels. It occurs due to alveolar filling with fluid, cells, pus or blood.

Commonly, ground-glass attenuation occurs in most ILDs and when it occurs, usually signifies the presence of acute disease / alveolitis.

In the various ILDs, different patterns of involvement are seen along with specific modes of distribution as described above. Central v/s axial patterns of distribution are better appreciated on HRCT than on plain radiographs.

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