Interstitial Lung Disease:
A Practical Approach to CT Diagnosis

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Disclosures

- Consultant for Riverain Medical
- Minor stockholder in Hologic, Inc.
- Consultant for GE Healthcare
- Research Support from Philips Healthcare
- License and royalty fees from University of Chicago (UC Tech)

Credits

Radiology Assistant
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Interstitial Lung Disease

- CT Technique for ILD
- CT patterns in ILD
- Classification of Interstitial pneumonias
- Integrating imaging findings with patient history
Interstitial Lung Disease

- **Scan Protocol**
  - Mediastinum Axial 3/3 mm
  - Lungs Axial 3/3mm
  - **Lungs Axial 1/1mm**
  - Coronal 3/3mm
  - Sagittal 3/3mm
  - MIP Axial, 10/5mm
  - MINIP Coronal 7/7mm
  - Source Images Axial 0.6mm
  - Expiration Scan (Low Dose)
    - Axial 3/3mm
    - Coronal 3/3mm
  - Inspiration Axial Prone 1/10mm
Utility of thin sections

3mm

1mm
Utility of thin sections
Interstitial Lung Disease

- Scan Protocol
  - Mediastinum Axial 3/3 mm
  - Lungs Axial 3/3mm
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- Expiration Scan (Low Dose)
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- Inspiration Axial Prone 1/10mm
Utility of Expiration Scan: HP with Air Trapping
Evaluating Expiratory Effort

Inspiration

Expiration
Utility of Prone Series

Supine 1mm

Prone 1mm
MINIP – Minimum Intensity Projection

Axial Slab

Coronal MINIP – Mosaic perfusion
Value of MINIPs for Honeycombing & Traction Bronchiectasis
CT Features of ILD

- Ground-glass opacity
- Reticular Pattern
- Honeycomb pattern
- Traction Bronchiectasis
- Architectural Distortion
- Nodular pattern
- Airspace opacity / Consolidation
- Bronchial Wall Thickening
- Mosaic perfusion

Fleischner Society: Glossary of Terms for Thoracic Imaging
Ground-Glass Opacity

Features:
- Hazy lung opacity
- Preservation of the bronchial and vascular margins
- Less opaque than consolidation

Caused by:
- Partial filling of airspaces
- Interstitial thickening
- Partial collapse of alveoli
31 y/o male with increasing dyspnea and fever

- Infections
- Edema
- Drug related
- Hemorrhage
31 y/o male with increasing dyspnea and fever

Pneumocystis Pneumonia
Classification of ILD by Predominant Pattern

Predominantly Ground-glass Opacity (GGO)

- Pneumocystis or viral pneumonia
- Acute Drug/Hypersensitivity Reactions
- Pulmonary Edema
- Hemorrhage
- Acute Aspiration
- AIP, NSIP, DIP
- Mucinous adenocarcinoma
Drug Toxicity

Imaging Findings:
- Pulmonary Edema
- Diffuse Alveolar Damage (DAD)
- Pulmonary hemorrhage
- Organizing Pneumonia
- Eosinophilic Pneumonia
- UIP/NSIP
GGO appearance is caused by density averaging
Pseudo Ground Glass Opacity

GGO appearance is caused by density averaging
Reticular Pattern

Features:
• Netlike pattern, linear opacities

Cause:
• Infiltration of the interstitial framework of the secondary pulmonary lobule
Classification of ILD by Predominant Pattern

Predominantly Linear or Reticular Pattern

- Edema
- Lymhangitic Mets
- UIP (Usual Interstitial Pneumonia)
- NSIP (Non-specific Interstitial Pneumonia)
- HP (Hypersensitivity Pneumonia)
- Asbestosis
Acute Pulmonary Edema

- Smooth thickening of interlobular septa
- Ground glass and airspace opacity
- Pleural effusions
- Thickened axial interstitium
- Irregular thickening of interlobular septae
Lymphangitic Metastases

- Thickened axial interstitium
- Irregular thickening of interlobular septae
Honeycomb Pattern

Features:

- Clustered cystic airspaces ~2-10mm
- Subpleural
- Well-defined walls

Indicates:

- Late stage fibrosis
- Destroyed fibrotic lung tissue with numerous cystic airspaces with thick fibrous walls
Mild Subpleural Microcystic Honeycomb Pattern
Traction Bronchiectasis

- Irregular bronchial dilatation caused by surrounding retractile pulmonary fibrosis
50 y/o female with DOE and hypoxia
50 y/o female with DOE and hypoxia
50 y/o female with DOE and hypoxia
Follow up CXR after antibiotic RX
Emphysema with Bacterial Pneumonia
Airspace Opacity/Consolidation

- Air space filling process
- Obscures vascular structures
- Air bronchograms
Classification of ILD by Predominant Pattern

**Consolidation**

**Acute**
- Infection, edema, hemorrhage

**Chronic**
- COP (Cryptogenic Organizing Pneumonia)
- Chronic infections
- Chronic Eosinophilic Pneumonia
- Lipoid Pneumonia
- Mucinous adenocarcinoma “BAC”
- Lymphoma
78 y/o female w cough and low grade fever: organizing pneumonia
Cryptogenic Organizing Pneumonia (COP)

Clinical Features

- Mean age 55 years
- Nonsmokers > smokers 2:1
- Dyspnea, cough, fever
- Respond ++ to steroid rx
- Good prognosis

CT

- Multifocal
- Lower lobes more frequently involved
- Opacities vary from ground glass to consolidation
Cryptogenic Organizing Pneumonia (COP)

Clinical Features

- Mean age 55 years
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CT

- Multifocal
- Lower lobes more frequently involved
- Opacities vary from ground glass to consolidation
  - Subpleural arcuate opacities
  - Atoll sign
Classification of ILD by Predominant Pattern

Predominantly Nodular Pattern

- Hematogenous infection (TB etc)
- Metastases
- HP
- RBILD
- LCH
- Sarcoid
- Lymphangitic mets
Classification of ILD by Predominant Pattern

Predominantly Nodular Pattern

- Sharply defined
  - Metastases
  - Granulomas (Mycobacterial, fungal, sarcoid, pneumoconiosis)

- Poorly defined
  - ± Centrilobular (HP, RBILD, LIP, LCH, “BAC”)
Nodular Pattern
Sharply defined, Random, Uniform
Nodular Pattern
Miliary MTB
Nodular Pattern: Non-Uniform
Nodular Pattern: Non-Uniform

Metastatic Lung Cancer
Diffuse small sharp solid nodules

- Metastases
  - Thyroid, lung, GI etc
- Granulomas
  - Miliary infection: TB, Histo etc
  - Sarcoid
  - Silicosis etc
Perilymphatic/ Peribronchovascular Distribution

Sarcoidosis
Centrilobular Nodules

- Inflammation of the terminal bronchiole and lobular pulmonary artery
- Small round opacity in the center of a secondary pulmonary lobule
- **Hypersensitivity Pneumonitis**

  - **Acute**: Air-space opacities
  - **Subacute**: Centrilobular nodular opacities, ground glass opacity
  - **Chronic**: Fibrosis, honeycombing, air-trapping
- Hypersensitivity Pneumonitis

Acute: Air-space opacities
Subacute: Centrilobular nodular opacities, ground glass opacity
Chronic: Fibrosis, honeycombing, air-trapping
Centrilobular Nodules

Respiratory Bronchiolitis ILD

Tobacco smoke produces mild immunosuppression. Smokers get RB/RBILD, but not get HP
Aspiration Bronchiolitis

- Centrilobular nodules
- Tree-in-bud opacities
- Airspace opacities
Classification of ILD by Predominant Pattern

Cystic Patterns

- Emphysema
- LAM (Lymphangioleiomyomatosis)
- LCH (Langerhans Cell Histiocytosis)
- Honeycomb pattern in UIP, Sarcoid etc.
- Chronic PJP
- Lymphocytic Interstitial pneumonia (LIP)
Emphysema

- Centrilobular
- Paraseptal
- Panlobular
Centrilobular Emphysema - Coronal MINIP
Upper lobe emphysema, basilar fibrosis
Combined Pulmonary Fibrosis and Emphysema (CPFE)

- Almost all male
- Predominant paraseptal emphysema
- PFTs (Lung Volumes, FEV₁) less abnormal than expected
- DLCO very abnormal
- High incidence of PAH
- Prognosis intermediate between COPD and UIP

Middle aged woman with SOB

- Thin-walled cysts in otherwise normal lung
Lymphangioleiomyomatosis (LAM)

- Thin-walled cysts in otherwise normal lung
Lymphangioleiomyomatosis (LAM)

Women of child bearing age or older
Smooth muscle proliferation
Can present with hemoptysis, pneumothorax or chylous effusion
Pulmonary cysts with thin walls, + nodules
Related to TSC (Tuberous Sclerosis Complex)
+ Renal angiomyolipomas
Example #1: 48 y/o female smoker with PA hypertension
Case #2: 41 y/o smoker

- Irregularly shaped cysts
- Small nodules
- Upper lobe predominant
- Sparing of CP angles
Langerhans Cell Histiocytosis (LCH)

- Irregularly shaped cysts
- Small nodules
- Upper lobe predominant
- Sparing of CP angles
Smoking Related Lung Disease

- Respiratory Bronchiolitis (RB)
  - Usually asymptomatic – all smokers have it.

- RB-ILD
  - 5-10% of smokers have clinically significant lung disease in association with RB

- DIP
  - More diffuse and severe than RB-ILD

- LCH (Langerhans Cell Histiocytosis)
  - 90-100% are cigarette smokers
  - < 40 yrs old
  - Cysts, nodules
  - UL predominance, sparing of CP angles
50 y/o man with fever, SOB and hypoxia
Pneumocystis Jiroveci Pneumonia (PJP)

Clinical:
- AIDS: CD4 < 200
- Organ transplant
- Lymphoma / leukemia

CT:
- Hazy interstitial opacity (GGO)
- Usually diffuse
- Thin-walled cysts >UL ≤ 38%
Interstitial Pneumonias: Definition and Classification

- **Definition**
  - Idiopathic: Uncertain etiology
  - Interstitial Pneumonia: interstitial lung inflammation and fibrosis which also affects airspaces, peripheral airways, and vessels

- **Classification**
  - Standardized by American Thoracic Society and European Respiratory Society in 2001
  - Based on clinical information, radiology, and pathology.
<table>
<thead>
<tr>
<th>Histologic Pattern</th>
<th>Distribution</th>
<th>Typical CT Findings</th>
<th>Smoking Related?</th>
</tr>
</thead>
<tbody>
<tr>
<td>UIP</td>
<td>Peripheral, Subpleural, Apicobasal Gradient</td>
<td>Reticular, Honeycombing, Traction Bronchiectasis, Architectural Distortion</td>
<td>Usually</td>
</tr>
<tr>
<td>NSIP</td>
<td>Peripheral, Subpleural, Basal, Symmetric</td>
<td>Ground Glass, Reticular, Consolidation</td>
<td>NO</td>
</tr>
<tr>
<td>OP</td>
<td>Subpleural, Peribronchial</td>
<td>Patchy Consolidation and/or Nodules</td>
<td>NO</td>
</tr>
<tr>
<td>DAD</td>
<td>Diffuse</td>
<td>Consolidation, Ground Glass, Lobular Sparing, Traction Bronchiectasis</td>
<td>NO</td>
</tr>
<tr>
<td>RB</td>
<td>Diffuse, Upper lobe</td>
<td>Bronchial Wall Thickening, Centrilobular Nodules, Patchy Ground Glass</td>
<td>YES</td>
</tr>
<tr>
<td>DIP</td>
<td>Lower Zone, Peripheral</td>
<td>Ground Glass, Reticular Lines</td>
<td>YES</td>
</tr>
<tr>
<td>LIP</td>
<td>Diffuse</td>
<td>Centrilobular Nodules, Ground Glass, Septal and Bronchovascular Thickening, Thin-walled Cysts</td>
<td>NO</td>
</tr>
</tbody>
</table>
Common Causes of Fibrosing ILD

- **UIP** (Usual Interstitial Pneumonia)
- **NSIP** (Non-Specific Interstitial Pneumonia)
- **HP** (Hypersensitivity Pneumonitis)
- Sarcoid
<table>
<thead>
<tr>
<th>Pathological Diagnosis</th>
<th>Etiology</th>
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</thead>
<tbody>
<tr>
<td>UIP</td>
<td>IPF (Idiopathic Pul. Fibrosis)</td>
</tr>
<tr>
<td></td>
<td>Collagen Vascular Disease</td>
</tr>
<tr>
<td></td>
<td>End stage HP</td>
</tr>
<tr>
<td></td>
<td>Asbestosis, etc</td>
</tr>
<tr>
<td>NSIP</td>
<td>Primary NSIP</td>
</tr>
<tr>
<td></td>
<td>Collagen Vascular Disease</td>
</tr>
<tr>
<td></td>
<td>Hypersensitivity (HP)</td>
</tr>
<tr>
<td></td>
<td>Drug reactions</td>
</tr>
</tbody>
</table>
UIP

Etiology
- IPF (Idiopathic Pul. Fibrosis)
- Collagen Vascular Disease
- End stage HP
- Asbestosis, etc

Histology
- Temporal heterogeneity
  - Different stages in the evolution of fibrosis
- Scattered fibroblastic foci
- Patchy lung involvement with subpleural and basal
- Honeycombing
UIP

- **CT Findings**
  - Apicobasal gradient
    - Most extensive in the most basal section
  - Subpleural reticular opacities
  - Honeycombing
  - Traction bronchiectasis
  - Heterogeneous
    - Fibrosis with areas of normal lung
  - Limited ground-glass opacity
UIP

Traction Bronchiectasis

Honeycombing
Idiopathic Pulmonary Fibrosis (IPF)

- Typical Features
  - Age > 50 years
  - Progressive dyspnea and nonproductive cough
    - Symptoms 3 months or greater
  - Fatigue and weight loss
  - Does not respond to high-dose corticosteroid therapy
  - Mean survival 2-4 years
  - Histological pattern is UIP
NSIP Pattern

- Patchy ground-glass attenuation
- Irregular, fine reticular opacities
  - May be associated with traction bronchiectasis
- Scattered micronodules
- Honeycombing inconspicuous or minimal
  - More likely in fibrosing subtype
- Consolidation is infrequent
Nonspecific Interstitial Pneumonia (NSIP)

- **Typical Features**
  - 40-50 years, decade younger than IPF
  - Worsening dyspnea over several months
    - Milder than IPF
  - Fatigue and weight loss
  - Treatment with corticosteroids and cyclosporin
  - Prognosis better than IPF
    - Correlates with the extent of fibrosis
  - Histological pattern is NSIP
Uncommon Interstitial Pneumonias

- **AIP (Acute IP):** Diffuse Alveolar Damage (DAD)
  - Only idiopathic interstitial pneumonia with acute onset of symptoms
  - Occurs in healthy individuals with prior illness suggestive of a viral upper respiratory infection
  - Severe dyspnea with need for mechanical ventilation in less than 1-2 weeks

- **DIP (Desquamative IP):**

- **LIP:** Lymphocytic Interstitial Pneumonia
Multidisciplinary Approach to ILD

- Clinical
  - Age, sex, Occupation
  - Clinical presentation (Acute or Chronic)
  - Immune status, drug exposures, RT

- Imaging
  - Lung volumes
  - Disease distribution
  - CT Pattern
  - Lymphadenopathy, effusions, CM, PAH
Clinical History

- Exposures
  - Tobacco:
    - + LCH, RBILD, DIP, UIP, Eosinophilic Pna
    - - HP
  - Drugs
    - Chemotherapy
    - Immunotherapy
    - Illicit drugs (Cocaine, Heroin etc)
  - Dust
    - Silica, Coal dust, Asbestos, Berylium
Summary

- Define key features including dominant pattern and distribution
- Consider clinical context, including age, gender, immune status, drug and occupational exposures
- Compare with previous scans and CXRs
- Multidisciplinary approach works best