Multi-disciplinary Approach to Diffuse Lung Disease: The Imager’s Perspective
Role of HRCT

- Diagnosis
- Fibrosis vs. inflammation
- Next step in management
- Response to treatment

Confidence in diagnosis

- Definitive HRCT pattern
- HRCT + clinical: diagnostic
- Nonspecific HRCT pattern
Clinical Context

- Bird exposure -> hypersensitivity pneumonitis
- Smoker -> respiratory bronchiolitis
- Connective tissue disease -> follicular bronchiolitis
- Iron welder -> siderosis
- Acute symptoms -> viral infection, HP
End stage IPF

Markedly reduced TLC and DLCO

End stage constrictive bronchiolitis

Inspiration

Expiration

Markedly reduced FEV1
HRCT may show reduced sensitivity for:

- Small airways diseases
  - Constrictive broncholitis
  - Hypersensitivity pneumonitis
  - Asthma
- Emphysema
- Pulmonary hypertension

NSIP + pulmonary hypertension

Markedly reduced DLCO
Fibrosis vs. Inflammation

| No GGO - fibrosis | GGO - inflammation | GGO - fibrosis |

HRCT guides further work-up

| Bronchoscopy | Sputum | VATS |

11/10/2014
HRCT: follow-up after tx

Clinical/PFT deterioration

6 months later  Initial
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<th>Idiopathic Clinical Syndrome</th>
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### Radiology <-> Pathology

- 1. Microscopic honeycombing
- 2. Collagenous fibrosis
- 3. Fibroblastic foci
- 4. Normal lung
**Usual interstitial pneumonia (HRCT)**

*Raghu et al. Am J Respir Crit Care Med 2011; 183: 788*

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**Definite UIP: IPF**
UIP: non-idiopathic causes

- Asbestosis
- Drug
- Rheumatoid

Inconsistent with UIP

Final diagnosis: IPF
What % of patients have IPF?
*Raghu et al. Am J Respir Crit Care Med 2011; 183: 788*

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Atypical appearances of IPF
*Sverzellati et al. Radiology. 2010; 254: 957*

- All biopsy proven UIP
- HRCT probability of IPF
  - High: 27% of cases
  - Intermediate: 11% of cases
  - Low: 62% of cases
- Favored diagnosis with low probability HRCT
  - NSIP: 53%
  - Nonspecific: 24%
  - Chronic HP: 12%
  - Sarcoidosis: 9%
Familial Interstitial Lung Disease

Lee et al. Chest 2012; 142: 1577

- Genetic mutation (e.g. telomerase) or idiopathic
- 2-20% cases of IPF
- Earlier age of onset (<50 years old)
- Pathology
  - Unclassifiable fibrosis: 60%
  - UIP: 40%
- Radiology
  - Definite/possible UIP (22%)
  - Honeycombing (32%)
**UIP (HRCT)**

*Raghu et al. Am J Respir Crit Care Med 2011; 183: 788*

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**What diseases/patterns may mimic IPF on HRCT?**

- Nonspecific interstitial pneumonia (NSIP)
- Desquamative interstitial pneumonia
- Hypersensitivity pneumonitis
What diseases/patterns may mimic IPF on HRCT?

- Nonspecific interstitial pneumonia (NSIP)
- Desquamative interstitial pneumonia
- Hypersensitivity pneumonitis

Findings inconsistent with UIP

- 1. Ground glass opacity
- 2. Mosaic perfusion/air trapping (≥3 lobes)
- 3. Profuse micronodules
- 4. Discrete cysts
- 5. Consolidation
- 6. Mid-upper lung predominance
- 7. Peribronchovascular predominance
Can we distinguish UIP and NSIP?

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<td>Usually fibrotic</td>
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Sensitivity for UIP: 45%
Specificity for UIP: 96%

UIP vs. NSIP

Potential UIP pattern

Fell et al. Am J Resp Crit Care Med 2010; 181: 832

- Age >50 + fibrosis (+/- HC)
- Interstitial score: 0.6-1.0
- Specificity for UIP: 61-100%

UIP vs. NSIP

Ground glass opacity

Elliot et al. JCAT 2005; 29: 339

- Sensitivity for NSIP: 96%
- Specificity for NSIP: 42%
Sensitivity for NSIP: 64%
Specificity for NSIP: 93%

Silva et al. Radiology 2008; 246: 288

### UIP vs. NSIP

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Progression from NSIP to UIP

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- Nonspecific interstitial pneumonia (NSIP)
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Desquamative interstitial pneumonia
Idiopathic pulmonary fibrosis

Desquamative interstitial pneumonia
Findings favoring DIP over UIP

- Atypical morphology of “honeycombing”
  - Irregular shape (not round)
  - Thin walled
  - Resembles emphysema in upper lobes
- Lack of traction bronchiectasis
- Significant ground glass opacity

What diseases/patterns may mimic IPF on HRCT?

- Nonspecific interstitial pneumonia (NSIP)
- Desquamative interstitial pneumonia
- Hypersensitivity pneumonitis

>50% have no exposure history
Findings favoring HP over UIP

- Mosaic perfusion (inspiration)
- Air trapping (expiration)
- Distribution
  - Axial: central or diffuse
  - Craniocaudal: mid-upper lung

Unknown Case #1
Headcheese

- Head cheese is in fact not a cheese, but rather a terrine made of meat taken from the head of a calf or pig (sometimes a sheep or cow) that would not otherwise be considered appealing.
Ground glass opacity

- Changes below resolution of CT
- Processes
  - Alveolar
  - Interstitial
- Very nonspecific
- Broad differential

Mosaic perfusion

- Geographic decreased lung density
- Reflects differences in blood flow
- Causes
  - Bronchiolar disease
  - Vascular disease
Mosaic perfusion (lucent lung abnormal)

- Very geographic
- Air trapping
- Vessel size discrepancy

Headcheese sign

- Two components
  - Infiltrative (ground glass)
  - Obstructive (mosaic perfusion)
- Both in significant amounts
- Diagnosis usually HP
Hypersensitivity Pneumonitis

*Headcheese courtesy of Oscar Meyer

Headcheese: when to consider an alternative diagnosis

- Smoking → Respiratory bronchiolitis/DIP
- Acute symptoms only → Viral infection
- Connective tissue disease or immune suppression → Follicular bronchiolitis/LIP
- Two separate processes → Edema and asthma
Hypersensitivity pneumonitis

Headcheese  Centrilobular GGO nodules  Fibrosis + mosaic perf.