Improving the Radiologic Diagnosis of UIP

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IPF References
Arakawa H and Horima K. Honeycomb lung: history and current concepts. AJR 2011

Usual Interstitial Pneumonitis

§ Most common idiopathic ILD
§ Idiopathic UIP
  • IPF
§ Secondary UIP
  • CVD
  • Asbestosis

UIP

Temporal Heterogeneity, Fibroblastic Foci

§ Heterogeneous (variegated) pattern of fibrosis-inflammation-normal lung
§ Subpleural and perilobular
§ Fibroblastic foci
§ Variable “honeycombing”
  • macroscopic: visible on CT
  • microscopic: occult to CT

CT in IPF

§ Diagnosis
§ Temporal changes and complications
§ Pitfalls and atypical features
**Honeycombing**

*Fleischner Society Glossary, 2008*

§ Pathology
- “destroyed and fibrotic lung tissue containing numerous cystic airspaces with thick fibrous walls, representing the late stage of various lung diseases, with complete loss of acinar architecture. The cysts range in size from a few mm to several cm in diameter, have variable wall thickness, and are lined by metaplastic bronchiolar epithelium.”

**End-stage Fibrosis**

*Honeycombing*

Many causes: could be UIP/IPF

**“Honeycombing”**

“The cysts range in size from a few mm to several cm in diameter, have variable wall thickness,...”

*Yale Rosen Pulmonary Pathology*

§ Radiology
- “clustered cystic air spaces, typically of comparable diameters on the order of 3-10 mm, but occasionally as large as 2.5 cm. Honeycombing is usually subpleural and is characterized by well-defined walls.”
Thick walled cystic spaces arranged in concentric layers with shared walls associated with other findings of fibrosis:

- Traction bronchiectasis, intralobular lines, reticulation, lobular distortion

Must be distinguished from traction bronchiolectasis, bronchiectasis, paraseptal emphysema, other cystic lung disease, etc.
Why is the Definition Important?

2011 ATS/ERS/JRS/ALAT Statement
Requirements for IPF Diagnosis

§ Exclusion of other known ILD causes (CVD, domestic or environmental exposures, drug toxicity)
§ Combinations of specific HRCT and surgical lung biopsy patterns in patients undergoing biopsy
§ UIP pattern on HRCT in patients without biopsy

2011 ATS/ERS/JRS/ALAT Statement
HRCT Criteria for UIP Pattern

§ Subpleural, basal predominance
§ Reticular abnormality
§ Honeycombing with or without traction bronchiectasis
§ Absence of features inconsistent with UIP pattern (including air trapping, cysts, others)

UIP With Honeycombing
But is “honeycombing” what makes this UIP?

UIP Without Honeycombing
But is this therefore not UIP?

UIP Diagnosis Without Honeycombing
Gruden JF et al, AJR 2013

§ Lower and peripheral zone predominant
§ SOME upper lobe reticulation (peripheral)
§ Non-segmental (crosses fissures)
§ Traction bronchiectasis
§ HETEROGENEOUS

Interobserver agreement for these findings is much better than for honeycombing
§ 53 patients 2009-2011 with specific CT findings
  • Peripheral reticulation that is non-segmental
  • Lower lung predominance with SOME upper lobe involvement
  • Traction bronchiectasis with no honeycombing
  • Heterogeneous appearance

§ Excluded patients with CVD, prior biopsy, occupational or other exposure history, or those with follow-up less than 6 months
§ Final group of 38 patients

§ Average age 80 years (61-93)
§ Follow-up 6-104 months (mean 38, median 37)
§ All had clinical diagnosis IPF, 9 asymptomatic
§ 34 died (most respiratory failure) or progressed
§ 4 stable have relatively short follow-up
§ None had diagnosis reversed or improved
51 F cough, dyspnea, asthma
Multiple prior episodes of pneumonia

OLB and IPF

§ OLB not always required
§ OLB and CT should reconcile to the diagnosis
§ Cases where the OLB shows UIP and the CT is discordant need further clarification

Summary

§ Honeycombing is currently required for CT diagnosis, but the term is inconsistently defined and applied and observers vary
§ UIP has a typical CT appearance even without definite honeycombing if strict criteria applied
§ There are limitations to the dx even at OLB

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§ Reticular abnormality
§ Honeycombing with or without traction bronchiectasis
§ Absence of features inconsistent with UIP pattern (including air trapping, cysts, others)

Proposed Modification

HRCT Criteria for UIP Pattern

§ Subpleural, basal predominance with some upper lobe involvement
§ Reticular abnormality that is nonsegmental
§ Traction bronchiectasis with or without honeycombing
§ Heterogeneous appearance
§ Absence of features inconsistent with UIP

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