Patient with IPF and concomitant emphysema

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CASE OVERVIEW

A 74-year-old male patient presented with progressive exertional dyspnoea for 5 years, non-productive cough and bilateral velcro-like crackles with basal predominance on auscultation. Functional tests showed an obstructive as well as a restrictive pattern. HRCT revealed a definite UIP pattern with concomitant paraseptal emphysema.

This diagnostic case is an example of normal FVC values not excluding a diagnosis of pulmonary fibrosis. The presence of velcro crackles is a specific indicator of an underlying fibrotic disease and should prompt for further investigation with HRCT.
Basic data

• Male, 74 years old
• Symptoms: progressive dyspnoea on exertion during the last 5 years, non-productive cough
• Smoking status: ex-smoker, quit smoking 15 years ago, 50 pack-years
• Occupation: retired farmer
• Comorbidities: GERD, COPD
• Current medication: indacaterol, tiotropium, esomeprazole
Physical examination

Lung auscultation: Bilateral velcro-like crackles with basal predominance

- Digital clubbing was observed
- No leg oedema
- No arthralgia
- SpO$_2$: 97% (on ambient air)
- Heart rate: 65 bpm
LABORATORY

- Normal CBC, biochemistry
- Negative serology (ANA, RF, anti-CCP, ENA panel)
LUNG FUNCTION

Conclusion:
Mixed PFTs pattern (both obstructive and restrictive pattern) with severe reduction in $DL_{CO}$ is noted.
QUESTION 1

What of the following should prompt for an HRCT?

A. Presence of obstructive functional pattern
B. Presence of reduced DL$_{CO}$
C. Presence of velcro crackles*
D. Presence of digital clubbing*

Correct answer: C and D
Author’s solution

- The presence of the characteristic velcro crackles should always raise suspicion for an underlying fibrotic lung disease\(^1\).
- The presence of digital clubbing (although a characteristic of fibrotic lung disease) in a smoker imperatively renders the diagnostic assessment for an underlying malignancy\(^2-3\).

Short summary of the findings

- Paraseptal emphysema with upper lobe predominance
- Honeycombing with bibasilar, peripheral distribution, mainly in the posterior and lateral segments
- No inconsistent features with a UIP pattern
- Definite UIP pattern with concomitant paraseptal emphysema

- Slice thickness: 1.0 mm
QUESTION 2

What pattern is shown in this image?

A. Honeycombing
B. Paraseptal emphysema*
C. Pneumothorax
D. Bronchiectasis

Correct answer: B
In this image, paraseptal emphysema is identified. It must be emphasised that the differential diagnosis between paraseptal emphysema and honeycombing can be extremely difficult.

Findings that favour the diagnosis of paraseptal emphysema are\textsuperscript{1,2}:

1. Upper zone distribution
2. No discernible walls. Usually, the subpleural lucencies are margined by interlobular septa and form a single layer. The absence of clearly defined walls is mainly seen in relation to the adjacent normal lung parenchyma
3. Presence of centrilobular emphysema

References:

QUESTION 3

What pattern is shown in this image?

A. Honeycombing*
B. Paraseptal emphysema
C. Pneumothorax
D. Bronchiectasis

Correct answer: A
In this image honeycombing is identified. Note that the cystic air spaces:

- are clustered
- share clearly definable walls
- occur in several layers; the first layer is in immediate contact with the pleura

In early honeycombing a few, isolated, single-layered cystic air spaces can be seen.

References:
The identification of honeycombing on HRCT leads to the diagnosis of definite UIP pattern.

A. True
B. False*

Correct answer: B
Honeycombing is a pattern observed on HRCT. By itself, it cannot support a radiological diagnosis of a definite UIP pattern. Still, honeycombing is critical for making a definite diagnosis, for it must be present in combination with a specific distribution pattern, i.e. subpleural and bibasilar, and other HRCT features:

- Subpleural, basal predominance
- Reticular abnormality
- Absence of features listed as inconsistent with UIP pattern

Reference:
The most important take home messages of the case are:

1) A normal FVC does not exclude the diagnosis of pulmonary fibrosis.
2) The presence of velcro crackles is highly suspicious of an underlying fibrotic disease and should prompt for further investigation with HRCT.
3) The presence of digital clubbing in a smoker imperatively renders the diagnostic assessment of an underlying malignancy.