



## Patient with IPF and no honeycombing on HRCT

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# CASE OVERVIEW

A 76-year-old male patient presented with progressive exertional dyspnoea refractory to current formoterol/beclomethasone medication. Auscultation revealed velcro-like crackles with basal predominance and diminished breath sounds. Features on high-resolution CT (HRCT) scans showed a possible UIP pattern.

Despite absence of honeycombing – which is a pre-requisite for the diagnosis of a definite UIP pattern – considering of all available data in the presence of a possible UIP pattern can lead to a “working diagnosis” of IPF in clinical practice without resorting to surgical lung biopsy.

Raghu G et al. *Am J Respir Crit Care Med* 2011;183:788–824.

# MEDICAL HISTORY

- Male, 76 years old
- Symptoms: progressive dyspnoea on exertion during the last 2 years which was aggravated in the past 6 months and was refractory to formoterol/beclomethasone
- Smoking status: ex-smoker, quit smoking 26 years ago, 50 pack-years
- Occupation: retired textile industry worker
- Comorbidities: diabetes, hypertension, benign prostatic hyperplasia
- Current medication: formoterol/beclomethasone, glimepiride, ramipril, dutasteride

# MEDICAL HISTORY AND TESTS

## Physical examination

- Lung auscultation: Velcro-like crackles with basal predominance mainly regarding the left lung. Diminished breath sounds in the lower right zone.
- No digital clubbing
- No leg oedema
- No arthralgia
- SpO<sub>2</sub>: 98% (on ambient air)
- Heart rate: 60 bpm

# LUNG FUNCTION TESTS

Parameter	Value
FVC	66% predicted
FEV <sub>1</sub> /FVC	78%
TLC	65% predicted
DL <sub>CO</sub>	47% predicted

Conclusion: Lung function testing reveals a restrictive pattern

# LABORATORY

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

# IMAGING

## HRCT

- Irregular reticular pattern with subpleural distribution and lower zone predominance
- Traction bronchiectasis/bronchiolectasis
- Asymmetric distribution with greater involvement of the left lung
- Absence of features listed as inconsistent with UIP pattern

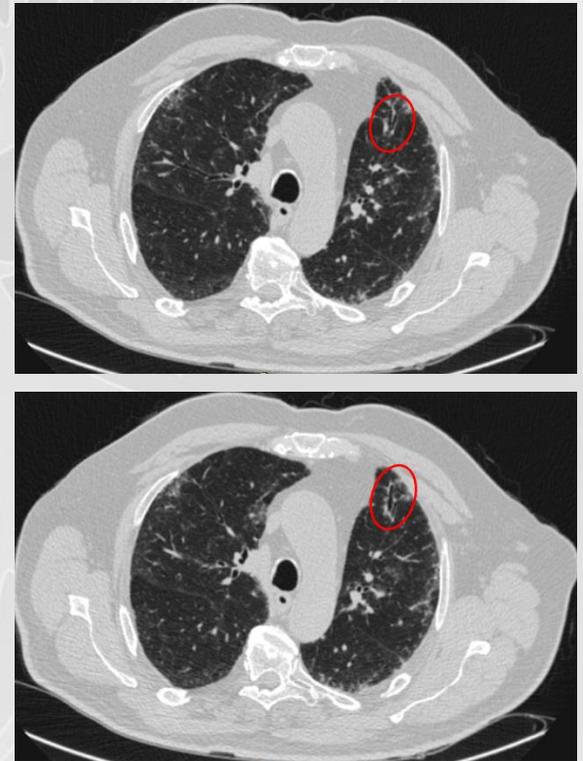
**Conclusion:** Possible UIP pattern

- Slice thickness: 0.9 mm

# QUESTION 1

What radiologic finding in these images points to the diagnosis of traction bronchiectasis?

- A. Signet ring sign
- B. Visible airway within 1 cm from the pleura
- C. Loss of tapering



# ANSWER 1

Author's solution: Correct answer: C

In the circles, a bronchus is identified that is larger than its parental and has a stable diameter while progressing peripherally.

The diagnosis of bronchiectasis is based on the following:

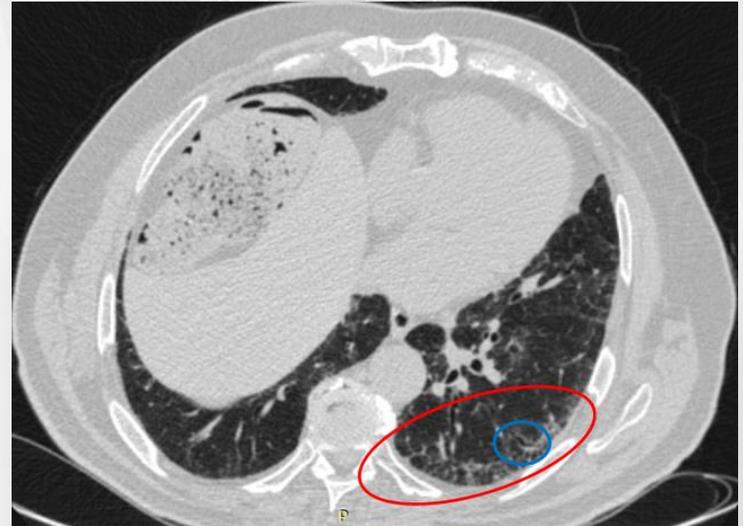
1. Loss of tapering
2. Signet ring sign (bronchial dilatation with respect to the accompanying pulmonary artery)
3. Visible airway within 1 cm of the pleura

Hansell DM et al. *Radiology* 2008;246:697-722.

## Question 2

What patterns are shown in this image?

- A. Honeycombing
- B. Paraseptal emphysema
- C. Bronchiectasis
- D. Irregular reticulation



## Answer 2

Author's solution: Correct answer: C, D

- In this image there is an irregular reticular pattern with bronchiectasis/bronchiolectasis (blue circle).
- The distribution is basal/subpleural.
- In the absence of features inconsistent with UIP pattern the radiologic diagnosis is that of possible UIP pattern.
- As an incidental finding, Morgagni hernia is recognised.
- Based on the current guidelines, in order to establish the diagnosis of IPF, a lung biopsy would be recommended. However, in clinical practice when alternative diagnoses are excluded, the presence of possible UIP pattern on HRCT (i.e. without the presence of honeycombing) is considered sufficient to form a “working diagnosis” and start treatment.

## Question 3

Which of the following parameters are useful clinical predictors of a diagnosis of IPF?

- A. Age
- B. Sex
- C. HRCT interstitial score
- D. Pulmonary function
- E. 6-minute Walk Distance
- F. Desaturation during 6-minute Walk Test

## Answer 3

Author's solution: Correct answer: A, C

- Age and extent of interstitial abnormalities on HRCT seem to be clinical predictors for an underlying UIP pathology pattern (OR: 1.11 and 17.20 respectively).
- This observation is of clinical value as it can support a “working diagnosis” of IPF in a patient with a compatible history and HRCT findings and obviate the need for lung biopsy.

1. Raghu G et al. *Am J Respir Crit Care Med* 2006;174:810–816.
2. Fell CD et al. *Am J Respir Crit Care Med* 2010;181:832-837.

# LEARNINGS FROM THE CASE

The most important take home messages of the case are:

- 1) Honeycombing is a pre-requisite for the diagnosis of a definite UIP pattern.
- 2) In clinical practice after examining all the available data (history, physical examination, serology) in order to exclude alternative diagnoses, the presence of a possible UIP pattern on HRCT can lead to a “working diagnosis” of IPF without resorting to lung biopsy.
- 3) Increased age favours the diagnosis of IPF in a patient with a possible UIP pattern.

**More patient cases can be found on [www.inIPF.com](http://www.inIPF.com)**