831_EPAT CARE: PATIENT CASE DR. MOLINA
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CASE#1
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CASE OVERVIEW

A 61 year-old man was referred to the Unit of Interstitial Lung Diseases (ILD) for evaluation of radiological images obtained during an abdominal pre-surgical examination (cholelithiasis). Here we discuss some aspects of the subclinical idiopathic pulmonary fibrosis (IPF) diagnosis.
PATIENT ANAMNESIS

Patient history
- Ex-smoker (20 packs/year)
- Builder (no silica, beryllium or asbestos exposure)
- No pets, feather or fungi exposure
PATIENT ANAMNESIS

Physical examination

- No digital clubbing

- Respiratory symptoms:
  - Dyspnoea, only with big effort (running or climbing)
  - No cough
  - Bi-basal crackles
LUNG FUNCTION

Spirometry and pletysmography

The lung function tests at rest show a mild restrictive ventilatory pattern

<table>
<thead>
<tr>
<th>Value</th>
<th>Absolute</th>
<th>% of predicted</th>
</tr>
</thead>
<tbody>
<tr>
<td>FVC (L)</td>
<td>3.190</td>
<td>86.0</td>
</tr>
<tr>
<td>FEV$_1$ (L)</td>
<td>2.780</td>
<td>96.6</td>
</tr>
<tr>
<td>FEV$_1$/ FVC (%)</td>
<td></td>
<td>87.1</td>
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<table>
<thead>
<tr>
<th>Value</th>
<th>Absolute</th>
<th>% of predicted</th>
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<tbody>
<tr>
<td>TLC (L)</td>
<td>5.120</td>
<td>78.7</td>
</tr>
<tr>
<td>RV (L)</td>
<td>1.600</td>
<td>64.8</td>
</tr>
<tr>
<td>RV/TLC (%)</td>
<td></td>
<td>31.3</td>
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</table>
LUNG FUNCTION

Diffusion test

The study of diffusing capacity of the lung for carbon monoxide (DL$_{CO}$ or TL$_{CO}$) shows a mild decrease which normalizes with effective alveolar ventilation.

<table>
<thead>
<tr>
<th>Value</th>
<th>Absolute</th>
<th>% of predicted</th>
</tr>
</thead>
<tbody>
<tr>
<td>DL$_{CO}$ (mmol/min/mmHg)</td>
<td>18.4</td>
<td>74.3</td>
</tr>
<tr>
<td>DL$_{CO}$/VA (mmol/min/mmHg/L)</td>
<td>3.92</td>
<td>94.2</td>
</tr>
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</table>
Serial pulmonary function test results can be used to (select all that apply):

• Quantify disease severity*
• Diagnose IPF
• Monitor disease progression*
• Identify patients with increased risk of mortality*

(* correct answer)
Author’s solution:

The results of pulmonary function tests cannot be used to positively diagnose IPF. A restrictive effect seen in the tests can be an indication of IPF, but it can also be caused by many other ILDs.

LABORATORY

- LDH: normal
- ACE: normal
- Haematology: normal
- Metabolic assessment: normal
- Immunology: negative
- Serum precipitins: negative
- Biochemistry: hyperglycaemia
QUESTION 2

What is the next step which should be taken?

• BAL cell count
• Biopsy
• HRCT scan*
• Repeat lung function measurements after 6 months

(*correct answer)
The HRCT scan results can diagnose IPF. In contrast, biopsy is only indicated after an HRCT scan, if the scan yields inconclusive results. BAL cell count is not thought to be of great benefit for the detection of IPF, it can, however, be helpful in diagnosing certain forms of ILD. After diagnosis has been made, follow-up every 6 months with repeated lung function measurements can be useful to monitor disease progression.


IMAGING

- Chest radiography almost normal
- HRCT shows possible UIP
  - Sub-pleural reticular septa
  - Some traction bronchiectasis with bi-basal predominance
IMAGING

HRCT

Upper section

Sub-pleural reticular opacities
IMAGING
HRCT
Upper section

Sub-pleural reticular opacities
IMAGING

HRCT

Middle section

Traction bronchiectasis
IMAGING

HRCT

Middle section

Traction bronchiectasis

Sub-pleural reticular opacities
IMAGING

HRCT

Lower section

Traction bronchiectasis

Sub-pleural reticular opacities
IMAGING

HRCT

Lower section

Traction bronchiectasis

Sub-pleural reticular opacities
BRONCHOSCOPY

• Macroscopic assessment showed no airway abnormalities
• BAS:
  • Microbiology negative
  • No atypical cells
• BAL cell count (middle section):
  • Macrophages 84%
  • Neutrophils 4%
  • Lymphocytes 9%
  • Eosinophils 1%
VATS-guided pulmonary biopsies from the left lung (upper and lower sections) were performed.
PATHOLOGY

The biopsy shows a histological UIP pattern featuring:
- Fibroblastic foci
- Honeycombing
- Temporal heterogeneity
- Sub-pleural predominance
PATHOLOGY

Honeycombing
PATHOLOGY

Sub-pleural predominance

Sub-pleural predominance
Temporal heterogeneity

Temporal heterogeneity (areas of normal lung and areas of lung fibrosis within the same sample)
PATHOLOGY

Fibroblastic foci
QUESTION 3

What is the final diagnosis of this patient?

- Chronic Hypersensitivity Pneumonitis (chronic HP)
- Idiopathic Pulmonary Fibrosis (IPF)*
- Non-Specific Interstitial Pneumonia (NSIP)
- Asbestosis

*correct answer
Author’s solution:

HRCT shows a possible UIP pattern, while histology shows a definite UIP pattern (different from NSIP).

Other causes of UIP were excluded, including antigens (patient history and specific blood tests) as well as asbestos exposure.

The final diagnosis, therefore, is IPF.

FINAL DIAGNOSIS

1. HRCT shows a possible UIP pattern
2. Histology shows a definite UIP pattern
3. Other causes of UIP were excluded, including antigens (patient history and specific blood tests) as well as asbestos exposure.

→ The final diagnosis is IPF.
LEARNINGS FROM THE CASE

1. Evidence of subclinical IPF can be found by combining HRCT results and other clinical signs, even if the HRCT lacks characteristic honeycombing (possible UIP).

2. If IPF is suspected in a patient and the HRCT is not clear, lung biopsy can be helpful in order to obtain a definite diagnosis.

3. It is important to detect IPF as early as possible, in order to advise the patient on pharmacological and non-pharmacological treatment.