Clinical evaluation of interstitial lung disease

Michael Kreuter
Center for interstitial and rare lung diseases
Thoraxklinik
University of Heidelberg
Röntgenstr. 1
69126 Heidelberg
Germany
kreuter@uni-heidelberg.de

AIMS
To discuss the right approach and meaningful diagnosis of patients with interstitial lung disease, including potential pitfalls and challenges.

SUMMARY
Interstitial lung diseases (ILDs) are a heterogeneous group of disorders including known causes (e.g. drug related or connective tissue related ILDs), idiopathic interstitial pneumonias (IIPs), granulomatous disorders (e.g. sarcoidosis) and other causes, mainly rare lung diseases (e.g. LAM, LHC). Recent years have shown that a precise diagnosis is most essential, as the correct diagnosis determines treatment and prognosis in these patients. In this context, the integration of clinical, radiological and histological data into a multidisciplinary team (MDT, consisting of clinicians, pathologists and radiologists) approach, has dramatically improved ILD diagnosis by adjusting interobserver variation and diagnostic confidence and by this has become standard of care in ILDs [1, 2].

Efforts must be undertaken to improve early diagnosis of ILD, e.g. in training young medical students in auscultation and ascertain medical history. Especially the correct identification of velcro crackles which should then lead to a subsequent further diagnostic procedure is most helpful [3]. Recent guidelines have dealt with a structured diagnostic approach [4, 5, 6]. The correct diagnostic procedures compromise the collection of several clinical data. These include medical history at primary presentation, exposures (including drugs, occupational and environmental exposures, smoking status), associated diseases, i.e. comorbidities which may influence symptoms and perhaps prognosis, pulmonary function testing and laboratory findings including specialised laboratory examinations e.g. auto-antibodies, precipitating IgGs and biomarkers. However, negative findings, e.g. normal values in pulmonary function testing do not exclude an interstitial lung disease. Also positive autoantibodies or precipitating IgGs have to be placed into the clinical context as they might be false positive. The role of radiological and pathological diagnosis as well as the role of bronchoscopy including bronchoalveolar lavage, cryobiopsy and surgical lung biopsy will be discussed elsewhere. In order to diagnose an idiopathic interstitial pneumonia all clinical, radiological and pathological data have to exclude known causes of interstitial lung diseases such as drug or connective tissue disease.

The clinical, radiologic and perhaps pathological findings have finally to be discussed in a multidisciplinary team in order to come to a potential definite diagnosis and to establish a correct therapy.
REFERENCES

   Recent review on advances in the clinical evaluation of pulmonary fibrosis
   Trial on the effect of multidisciplinary team discussions in order to improve ILD diagnosis
   Discussing the importance of correct auscultation leading to suspicion on ILDs
   First international classification of IIPs including a standardized approach diagnosing ILDs
   International guideline on diagnosing and managing IPF
   2nd international classification of IIPs

EVALUATION

1. Which sentence is correct?
   a. Diagnosing ILD must not be multidisciplinary
   b. Multidisciplinary diagnosis might influence survival positively
   c. A standardized questionnaire is never helpful
   d. The multidisciplinary team must include a thoracic surgeon

   Answer B

2. Which sentence is incorrect?
   a. A normal pulmonary function test (PFT) does not exclude interstitial lung disease
   b. PFTs might be obstructive
   c. PFTs might reveal restrictive lung disease
   d. Diffusion capacity is never influenced by comorbidities

   Answer D

3. Which sentence is correct?
   a. Auto-antibodies are not helpful
   b. Functional assessment can only be done by 6-minutes walking distance
   c. ILDs can be separated into (1) known causes (2) idiopathic IIPs (3) granulomateous ILDs (4) others
   d. Idiopathic pleuroparenchymal fibroelastosis is not a new rare IIP

   Answer C