



Letters to the Editor

The Importance of Diagnosis in Interstitial Lung Disease***La importancia del diagnóstico en enfermedades intersticiales pulmonares**

To the Editor:

Guidelines on the diagnosis and treatment of idiopathic pulmonary fibrosis have recently been published,¹ following on from the ATS guidelines published in 2011.² Consensus and guidelines are always essential, particularly for diseases that are difficult to manage due to their rarity, difficult diagnosis or the lack of therapeutic resources.

One of the reasons for this “revived” interest in interstitial disease and its diagnosis is the development of new drugs for use in situations in which no effective long-term treatments were previously available. The same is true of lung cancer, biomarker determination and personalized or precision medicine. Nowadays, reaching a detailed and accurate diagnosis with the smallest amount of tissue using multidisciplinary techniques and in the shortest time possible is the mantra of Pathology Departments. No clinician would propose initiating treatment with new drugs designed for specific therapeutic targets or tumour types (adenocarcinoma or squamous cell carcinoma of the lung, for example) without a firm, well-corroborated diagnosis, along with all the radiological, extension, histological and molecular studies available for predicting response to treatment.

Diffuse interstitial lung diseases or ILD (in the plural), however, are very rare, and few hospitals have specialists (pulmonologists, radiologists, thoracic surgeons and pathologists) with sufficient experience to reach a definitive, isolated diagnosis of idiopathic pulmonary fibrosis (without the clinical, radiological and pathological triad mentioned by Dr Morell in his editorial³). This raises the possibility of all the information derived from the tissue studies being missed; in other words, patients may be diagnosed solely on the basis of high resolution computed tomography, and may be given treatment that is only indicated in truly idiopathic cases.

Tissue studies, meanwhile, can show the presence of a secondary pathology. The predominantly central distribution of fibroblastic nests in the lobule, the presence of microgranulomas, the detection of substances such as asbestos bodies, the use of immunohistochemistry to detect immunoglobulin subtypes, or the presence of follicular bronchiolitis, to name but a few, can be key factors in the diagnosis of extrinsic allergic alveolitis,

pneumoconiosis, IgG4-associated syndromes or rheumatoid arthritis. This is only the tip of the iceberg that symbolizes our profound ignorance of this type of disease, an ignorance that can only be lifted with study and research (for which paradoxically tissue is also required).

All the information derived from the study of these patients is essential, and we must have all the data at our disposal in order to determine and predict how the new drugs will work and to further knowledge of the disease. As Dr Morell so rightly points out in his editorial, there are numerous cases of secondary ILD (extrinsic allergic alveolitis, drug toxicities, autoimmune disease, etc.) that can manifest as standard interstitial pneumonia and that are radiologically indistinguishable from idiopathic pulmonary fibrosis. Are patients with secondary diseases going to respond to the new treatments in a similar way as idiopathic cases? Would it be acceptable for an oncologist to treat a patient with tyrosine kinase inhibitors for non-small cell lung cancer without making a more detailed diagnosis, using all the techniques available?

These are questions that we all, pulmonologists, radiologists, surgeons, pathologists, hospital managers, public policymakers and the pharmaceutical industry (in whose interest it is to ensure that its products perform as well as possible), need to take into consideration today.

Conflict of Interests

Javier Gómez Román declares that he has received funding from InterMune for giving lectures at educational events.

References

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