



Editorial

Idiopathic Pulmonary Fibrosis: Importance of Accurate Diagnosis and Treatment[☆]

Fibrosis pulmonar idiopática: importancia de un diagnóstico preciso y tratamiento

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In the last decade, Idiopathic Pulmonary Fibrosis (IPF), once a “mixed bag” where various interstitial diseases were included, has become a much better defined entity. This follows the expert statements on IPF, published in 2000¹ and 2011,² which have established the diagnosis of this disease more precisely. In fact, in the 21st century, Non-Specific Interstitial Pneumonia (NSIP), Large Cell Interstitial Pneumonia (due to exposure to heavy metals), smoking-induced Pulmonary Fibrosis, and Chronic Hypersensitivity Pneumonitis (CHP), which present with clinical symptoms of IPF and a histological pattern of Usual Interstitial Pneumonia (UIP), are already found separate from the so-called Idiopathic Pulmonary Fibroses (PF) or individualized within them.

However, in my opinion, the recommendations for the study of this entity described in the last international expert consensus on IPF in 2011² still do not place sufficient emphasis on recommending that the eventual cause of the PF be exhaustively sought; only if no etiology is found should we then call it idiopathic (IPF). Indeed, although it is recommended that an initial search be made for a possible cause of the symptoms, it is not stressed that this search must be painstakingly and repetitively continued at subsequent visits. The international consensus does not recommend the systematic determination of specific IgGs either, which is rightly recommended in Spanish Guidelines on the diagnosis and treatment of IPF³ published in this issue of *Archivos de Bronconeumología*, and which has led to this Editorial. Moreover, due to their sensitivity and specificity, it would even be advisable to perform lymphocyte stimulation tests⁴ against antigens suspected of being the source of the fibrosis, in an attempt to systematically find a cause of CHP.

Likewise, neither of the guidelines remark on the frequent etiological role of smoking in (Idiopathic) Pulmonary Fibrosis. In the 2011 international guidelines, the role of smoking is mentioned in the section on Risk Factors,² and in the Spanish guidelines, in the section on Etiology and Risk Factors.³ It is obvious that in many patients with concomitant emphysema (paraseptal), some of whom also show a predominance of macrophages (pigmented) and

an increase in the number of these cells on bronchoalveolar lavage (BAL), there is no other environmental exposure or potential cause of fibrosis other than smoking.

Moreover, the presentation of some CHPs (occasionally also with a histological pattern of UIP) in the form of IPF is well known.⁵ This finding should lead to an attempt to systematically discard environmental/occupational causes of HP. This systematic study should comprise the abovementioned examinations: thorough, repeated anamnesis and immunological study and, in the case of wanting to confirm suspected CHP, a specific bronchial provocation test against the antigen suspected of causing the symptoms should also be indicated, providing of course that the patient's lung function allows (forced vital capacity (FVC)>50% and carbon monoxide diffusion capacity (DLCO)>40%).⁶

The general diagnostic criterion for IPF recommended in both the international² and Spanish guidelines,³ based only on high resolution computed tomography (HRCT) typical of IPF, may also be debatable. It appears rather unreliable to diagnose IPF by typical HRCT if, as the Spanish guidelines state, “A UIP pattern can also be identified [on HRCT] in chronic hypersensitivity pneumonitis, asbestosis and some connective tissue diseases”. It is also known that the agreement between experts in the interpretation of HRCT in IPF is only 0.40 (Kappa index).⁷ In our public healthcare system, in which the patient generally has great confidence in their doctor and who in the interest of an accurate diagnosis consents to undergoing invasive procedures, the recommendation of basing the diagnosis on the clinical, radiological and pathological triad seems much more reliable when making a firm definitive diagnosis.

It is also relevant to highlight the role of BAL here, which, although not sufficiently recommended in the guidelines,^{2,3} helps in many cases to point us toward the entity underlying the fibrosis symptoms. Our opinion is that BAL should be performed in all cases, especially when bronchofibroscopy could become an even more essential examination with the recent introduction of cryobiopsy, thus avoiding surgical biopsy in a great many patients.

The importance of being strict and accurate when making a diagnosis of IPF will lead, on one side, to being able to prescribe the best treatment in each case and, on the other, to avoid attributing to genetics familial cases in which the cause was exposure to the same or different antigens that eventually cause CHP/fibrosis.

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Genetic studies should be initiated following confirmation, after several visits, that this is indeed a patient with IPF and not a possible secondary PF. Finally, diagnostic accuracy will result, in cases with a specific etiology, in being able to promote the prevention and early treatment of the initial causes of this serious disease called PF.

We believe that, for now, this strict diagnostic behavior is the best therapeutic weapon, since ensuring the type of PF (which could be a result of CHP, secondary to another entity or ultimately, classified as idiopathic) will help to be able to avoid the cause in some patients, to use corticosteroids in many cases and, in others, to avoid them. In patients in whom it is not possible to identify the entity that precedes the fibrosis or the cause, the diagnosis will remain as IPF; in these cases, we should prescribe anti-fibrotic drugs which, like pirfenidone, have demonstrated a favorable effect in slowing disease progression.^{8–10} Later, when the pharmaceutical development of other drugs that are currently under study has been completed, if their effectiveness is confirmed, we will add them to pirfenidone in an attempt to slow the process, which is what we pulmonologists dedicated to the care of these patients have wished for years.

Conflict of Interests

Dr. Morell belongs to the Advisory Board of InterMune.

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