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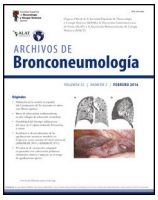

Editorial

Idiopathic Pulmonary Fibrosis? A Form of Presentation of Other Types of Pulmonary Fibrosis☆

Fibrosis pulmonar ¿idiopática?: una forma de presentación de otras fibrosis pulmonares

Since it was first described in the year 2000, idiopathic pulmonary fibrosis (IPF) has been considered a distinct clinical entity.[1] However, a very recently published comment has suggested that IPF is not a nosological entity with its own characteristics, but rather an advanced stage of other interstitial lung diseases.[2] Specifically, the usual interstitial pneumonia (UIP) radiological pattern of IPF can also be seen in silicosis, asbestosis, connective tissue diseases, and sarcoidosis, but most particularly, as a result of hypersensitivity pneumonitis, as reported in 2013.[3]

The idea that IPF is an advanced form of other pulmonary fibroses is not new: this concept was proposed in both Archivos de Bronconeumologia in 2016,[4] and Clinical Medicine in 2017.[5]

It now seems clear that the initial description of IPF as a distinct entity[1] is controversial, to say the least, since claims for its individual nature were based solely on the presence of certain radiological and histological characteristics known as the UIP pattern; furthermore, in many cases there are discrepancies in these characteristics between the 2 diagnostic procedures.[6]

Since IPF was described in 2000, the infrequent finding of patients with a typical UIP pattern, together with variability in the radiological manifestations needed to accept a diagnosis of IPF, meant that experts were forced to expand and divide the high-resolution computed tomography (HRCT) criteria into several diagnostic categories: *definitive, possible, and inconsistent UIP pattern.* Similarly, histological criteria were also divided into *definitive, possible, probable and non-UIP pattern.*[7]

Eight years later, as difficulties in the diagnosis of IPF persisted, the 2018 international guidelines[8] proceeded to split the HRCT diagnostic categories again into: *usual interstitial pneumonia (UIP), probable UIP, indeterminate for UIP*, while histological findings were divided into: *UIP, probable UIP and indeterminate for UIP.*

As can be seen, pulmonologists had to make numerous diagnostic adaptations and speculations, and still do, in order to fit patients into the nosological concept of IPF. Furthermore, as we have already mentioned,[6] the diagnostic concordance between the characteristics of the HRCT and biopsy findings, even in expert centers, is only moderate.[9] It is logical to assume that diagnostic difficulties will be even greater in less experienced centers.

Our proposal for the study of all patients with pulmonary fibrosis who do not have a definitive diagnosis is to approach the problem from a nosological and etiological point of view. In other words, we should try to identify the underlying disease and, at the same time, try to find the cause of it. If we identify the cause, we can recommend mitigation tactics and also prescribe a more targeted treatment for the specific disease that we have diagnosed.

To this end, we proposed a 10-point protocol for the study of the entities and causes of pulmonary fibrosis, which we have already published[5] and which consists of: 1) an exhaustive and repetitive history of exposures, performed by an expert in interstitial diseases; 2) pulmonary auscultation for velcro crackles and also for end-inspiratory wheezes (chirping rales); 3) chest HRCT; 4) blood tests with determination of specific IgG against the most common fungi and bird serum antigens most frequently associated with human contact, and a battery of anti-tissue autoantibodies, and determination of angiotensin-converting enzyme, rheumatoid factor, and anti-citrullinated peptide antibodies; 5) bronchoscopy with bronchoalveolar lavage cell count for lymphocytosis and histological study of cryobiopsy; 6) if a particular material is suspected to be causative, it will be cultured for fungi; 7) evaluate the possibility of the cause being smoking, in accordance with the established criteria[10]; 8) specific inhalation test if a known antigen is suspected to be the causative agent; 9) surgical lung biopsy will only be performed if still considered necessary to confirm the diagnosis after performing the above actions; and 10) clinical follow-up, with evaluation of the data provided by the patient and the results of other biopsies, or, possibly, study of the surgical specimen.

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