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Interstitial lung disease associated with idiopathic inflammatory myopathies: the importance of recognizing an early pulmonary phenotype

Enfermedad pulmonar intersticial asociada a miopatías inflamatorias idiopáticas: el valor de reconocer un fenotipo pulmonar temprano

Castro Zorrilla, Liliana[1, 2,3]

"In idiopathic inflammatory myopathies, the lung is not just another organ: it is the one that defines the clinical course."

Idiopathic inflammatory myopathies (IIM) comprise a heterogeneous group of autoimmune diseases in which pulmonary involvement has become a decisive prognostic factor. Interstitial lung disease (ILD) associated with IIM is one of the main determinants of morbidity and mortality[1,2]. In this context, early identification of clinical, immunologic, and radiologic risk patterns is essential to modify the natural course of the disease.

The multicenter study analyzed[3] provides significant data by characterizing a representative cohort of regional clinical practice, with a predominance of women, a high frequency of antisynthetase antibodies –particularly anti-Jo-1– and a dominant pattern of nonspecific interstitial pneumonia (NSIP). These findings are fully consistent with recent international series[4-6], underscoring that accurate identification of the antisynthetase phenotype is a cornerstone of diagnosis, prognosis, and therapeutic decision-making in this context.

Radiologic findings reinforce the importance of expert interpretation of high-resolution computed tomography. The predominance of the NSIP pattern, also observed in contemporary studies[5,7], requires active surveillance, given its potential reversibility when treated promptly. Multidisciplinary collaboration among pulmonology, rheumatology, and radiology is therefore essential to ensure early diagnosis and timely management.

From a functional standpoint, early reduction in diffusing capacity, exercise-induced desaturation, and restrictive ventilatory impairment represent sensitive markers of early pulmonary involvement[1,2]. Systematic assessment with DLCO and walk tests –simple, low-cost tools– plays a central role in detecting subclinical progression and adapting treatment in a timely manner.

Therapeutically, the favorable outcomes observed with mycophenolate and the use of rituximab in refractory cases align with contemporary evidence and recent multicenter studies.[6,8] Early implementation of intensive immunomodulatory therapy has shown benefit in preventing irreversible fibrotic progression and improving long-term survival.

A valuable contribution of the study is the comparison between patients with and without ILD. The higher prevalence of antisynthetase markers among those with ILD, and the greater muscular involvement and ANA positivity in those without pulmonary disease, reflect the phenotypic heterogeneity of IIM.[9] This variability reinforces the need to stratify patients according to distinct clinical and serologic profiles to guide personalized management strategies.

Taken together, this regional study reinforces a key clinical message: active screening for ILD in patients with IIM is essential. Early detection allows intervention before irreversible lung damage occurs, optimizing immunomodulatory therapy and improving long-term outcomes.

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[1]Specialist in Pulmonology and Clinical Immunology. [2]Associate Professor Faculty in Pulmonology. School of Medicine, University of Buenos Aires.[3] Former Head of the Department of Immunology, School of Medicine, University of Buenos Aires

Conflicts of interest

The author declares no conflicts of interest related to this publication.

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