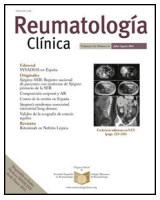
Reumatol Clin. 19 (2023) 407–408

Sociedad Española de Reumatología - Colegio Mexicano de Reumatología

Reumatología Clínica

www.reumatologiaclinica.org



Letter to the Editor

Does interstitial lung disease behave in the same way in all rheumatic diseases? A review of the literature

¿Se comporta la enfermedad pulmonar difusa igual en todas las enfermedades reumáticas? Revisión de la literatura

Dear Editor,

It is well known that there can be lung involvement in some connective tissue diseases (CTDs), and it may even be their first manifestation. For example, Mathai and Danoff described that the CTDs most frequently associated with interstitial lung disease (ILD) are systemic scleroderma and inflammatory myopathies, followed by rheumatoid arthritis (RA), and less frequently Sjögren's syndrome, or systemic lupus erythematosus (SLE).[1] In all of these diseases, the most frequently observed pattern is that of systemic scleroderma or systemic lupus erythematosus. Non-specific interstitial pneumonia (NSIP) is the most frequently observed pattern in all of these diseases, except RA, in which the pattern of usual interstitial pneumonia (UIP) prevails.

ILD is the most common pulmonary manifestation in RA, and its prevalence and incidence are unclear (1%–58%). In fact, the prevalence of subclinical ILD ranges from 19% to 57%, and that of clinically significant ILD is around 10% in men and 7% in women. Thus, the cumulative incidence of RA-ILD with clinical impact at 10 years is 5%, at 15 years 6.3%, and at 30 years 6.8%. UIP is the most frequent radiological pattern, which appears in around 50%–60% of cases, followed by the NSIP pattern, although other patterns such as organised pneumonia or desquamative interstitial pneumonia may also appear. In terms of mortality, ILD is the second leading cause of death in RA patients (after cardiovascular disease) and, in fact, women with RA-ILD are 3 times more likely to die than patients without RA.[2]

In systemic scleroderma, interstitial abnormalities are found on high-resolution computed tomography in up to 80% of cases, which rises to 90% in autopsies of these patients,[3] of these, about 40% develop clinical symptoms. Nihtyanova et al. found in an observational study that 47% of patients developed clinically significant ILD, most within 5 years of diagnosis, and none after 15 years. NSIP is the most frequent radiological pattern, which appears in 80%–90% of cases, the fibrotic type being more frequent than the cellular type, followed by the UIP pattern in less than 10% of patients. The course of this disease is highly variable; systemic scleroderma-associated ILD accounted for 35% of deaths in the EUSTAR cohort. Notably, the presence of fibrosis on high-resolution computed tomography and decreased FVC on respiratory function tests are independent predictors of mortality in this type of patient.[4]

The prevalence of ILD in inflammatory myopathies is estimated to be 20%–78%.[1] NSIP is the most frequent pattern, although it may exist with diffuse alveolar damage or organised pneumonia.

In Sjögren's syndrome, ILD is the most frequent and severe pulmonary complication, with a prevalence of 20%, and is the first manifestation in 10% of patients. NSIP was the most frequent radiological pattern, in 45% of patients, however, the lymphocytic interstitial pneumonia pattern is the most characteristic but rare. Data on the prognosis of ILD are limited, but 5-year survival rates of up to 89% have been estimated.[5]

In conclusion, it can be seen that the behaviour of the PDID associated with rheumatic diseases is very varied. In fact, Park et al. studied the evolution of these diseases. They observed that: a) patients with CTD with NSIP pattern had the same survival as patients with the same pattern but without CTD; b) patients with CTD and UIP pattern (except RA) had the same survival as patients with CTD and NSIP pattern, and c) patients with CTD with UIP pattern had longer survival than patients with idiopathic pulmonary fibrosis (IPF).[6]

Table 1 summarises the characteristics of CTD-associated ILD.

Table 1
Characteristics of CTD-associated ILD.

ILD	RA	SS	IM	Sjögren's
Prevalence, %	1–58	80–90	20–78	20
Radiological pattern	UIP > NSIP > OP, DIP	NSIP > UIP	NSIP	NSIP *LIP: characteristic
Prognosis	Second cause after cardiovascular diseases	35% mortality of SS	Survival at 5 years: Acute form: 52% Chronic form: 87%	Survival at 5 years of 89%

CTD: connective tissue disease; DIP: diffuse interstitial pneumonia; ILD: interstitial lung disease; IM: inflammatory myopathies; LIP: lymphocytic interstitial pneumonia; NSIP: non-specific interstitial pneumonia; OP: organised pneumonia; RA: rheumatoid arthritis; SS: systemic scleroderma; UIP: usual interstitial pneumonia.

References

1. Mathai SC, Danoff SK. Management of interstitial lung disease associated with connective tissue disease. BMJ. 2016;352:h6819.
2. Spagnolo P, Lee JS, Sverzellati N, Rossi G, Cottin V. The lung in rheumatoid arthritis: focus on interstitial lung disease. Arthritis Rheumatol. 2018;70:1544–54.
3. Perelas A, Silver RM, Arrossi AV, Highland KB. Systemic sclerosis-associated interstitial lung disease. Lancet Respir Med. 2020;8:304–20.
4. Cottin V, Brown KK. Interstitial lung disease associated with systemic sclerosis (SSc-ILD). Respir Res. 2019;20:13.
5. Luppi F, Sebastiani M, Silva M, Sverzellati N, Cavazza A, Salvarani C, et al. Interstitial lung disease in Sjögren's syndrome: a clinical review. Clin Exp Rheumatol. 2020;38 Suppl 126:291–300.
6. Park JH, Kim DS, Park IN, Jang SJ, Kitaichi M, Nicholson AG, et al. Prognosis of fibrotic interstitial pneumonia: idiopathic versus collagen vascular disease-related subtypes. Am J Respir Crit Care Med. 2007;175:705–11.

Diego Durán Barata,* Andrea Pérez Figuera,
Winnifer Briceño Franquiz, Beatriz Pintado Cort

Servicio de Neumología, Hospital Universitario Ramón y Cajal, Universidad Alcalá de Henares, Madrid, Spain

* Corresponding author.
E-mail address: iago_794@hotmail.com (D. Durán Barata).