

Respiratory Compromise In Patients With Primary Sjögren Syndrome.

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Background/Purpose: Sjögren's syndrome (SS) is an autoimmune inflammatory disease that affects exocrine glands and may have extraglandular involvement. The respiratory compromise (RC) occurs in 19%–65% of the patients with SS and the most common type is nonspecific interstitial pneumonia (NSIP) (30–60%). Objectives: To determine the frequency, time of occurrence and type of RC in patients with primary SS. To establish the relationship between RC and antibody profile, complement levels, leukopenia, and other extraglandular manifestations. **Methods:** Observational, transversal and analytical study. We included patients who met ACR-EULAR criteria for primary Sjögren's syndrome, over 18 years old from the GESSAR (argentine SS study group) database from Jan/2011 to Jul/2011 inclusive. We evaluated demographic data, time of evolution, the presence of extraglandular manifestations and laboratory parameters. Categorical variables were compared with X2 test or Fisher test and continuous variables with the Mann-Whitney test. The p was considered significant when $p < 0.05$. We performed multivariate logistic regression analysis.

Results: Total N: 285. 95.9% female, age 55.8 \pm 14.8 y. Time of evolution of SS until the RC developed: 4 years (IQR 2–6 a). 75 patients (26.3%) had RC, of which the most frequent were xerotrachea (58.6%), recurrent infections (24%), NSIP (13.3%) and pulmonary fibrosis (12.5) (Box 1). RC was more common in patients with Raynaud syndrome (22.2% vs 12.3%, $p < 0.04$), fibromyalgia (26.8% vs 14.3%, $p = 0.01$) and tinnitus (5.9% vs 1.2%, $p = 0.050$). There were no significant differences in laboratory parameters. In the multivariate logistic regression analysis there was an independent association of with Raynaud's (OR 2.32, 95% CI 1.08 to 4.96, $P < 0.03$) and fibromyalgia (OR 2.50, 95% CI 1.21 to 5.13, $p < 0.01$) with RC.

Table 1. Distribution of the Types of Respiratory Compromise

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Xerotrachea n (%)	44 (58,6)
Recurrent infections n (%)	18 (24)
Nonspecific interstitial pneumonia (NSIP) n (%)	10 (13,3)
Pulmonary fibrosis (UIP) n (%)	9 (12,5)
Lymphocytic interstitial pneumonia n (%)	3 (4)
Pulmonary hipertensión n (%)	2 (2,6)
Pulmonary thromboembolism n (%)	2 (2,6)
Bronchiectasis n (%)	2 (2,6)
Bronchiolitis obliterans n (%)	2 (2,6)
Pleural effusion n (%)	1 (1,3)
Nonspecific interstitial pneumonia n (%)	1 (1,3)
Solitary nodule n (%)	1 (1,3)

Conclusion: A quarter of patients with SS had RC of which the most frequent were xerotrachea, recurrent infections, NSIP and pulmonary fibrosis. There was no association with laboratory parameters. RC was independently associated with the presence of lung disease raynaud and fibromyalgia. The authors declare no conflicts of interest.