LYMPHOMA ARISING AT THE TIME OF DIAGNOSIS OF PRIMARY SJÖGREN SYNDROME: A HIGHLY-ACTIVE SYSTEMIC SUBSET OF THE DISEASE


Background: By January 2019, The Big Data Sjögren Project included 11,420 consecutive patients with primary Sjögren syndrome (SjS) in whom a lymphoproliferative disease is diagnosed concomitantly. Among the 2,696 patients recruited from 24 countries of the five continents, 312 patients (11.6%) were diagnosed with lymphoma and primary SjS syndrome (SjSs) in whom a lymphoproliferative disease is diagnosed concomitantly.

Objectives: By January 2019, The Big Data Sjögren Project included 11,420 consecutive patients with primary SjS syndrome. The objective of the study was to describe the frequency and characteristics of lymphomas in patients with primary SjS syndrome (SjSs) recruited from 24 countries of the five continents.

Methods: In the present study, we describe the frequency and characteristics of lymphomas in patients with primary SjS syndrome (SjSs) recruited from 24 countries of the five continents.

Results: 117 (1%) patients were diagnosed with lymphoma and primary SjS syndrome. Age-gender adjusted multivariate analysis identified the following features associated with lymphoma (OR: C19%; male gender (4.61; 2.88-7.18), White ethnicity (3.51; 1.78-7.91), abnormal oral tests (3.4; 1.38-10.88), positive biopsy (3.2; 1.3-10.17), positive RF (2.7; 1.48-3.53), hypocomplementemia (3.39; 1.4-7.89), peripheral nerve (2.56; 1.4-4.41) and hematological (2.49; 1.4-3.75) ESSDAI domains were associated with lymphoma (frequencies summarized in the Figure).

Conclusion: Patients diagnosed concomitantly with primary SjS and lymphoma have a very specific, highly-active phenotype (men, White, severe oral involvement, cryoglobulinemic-related immunological markers, and high systemic activity).

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