Primary Sjögren syndrome and development of another connective tissue disease during follow-up.

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INTRODUCTION: Primary Sjögren's Syndrome (pSS) is a chronic autoimmune disease characterized by lymphocytic infiltration of the exocrine glands. It is the connective tissue disease that is most frequently associated with other autoimmune disorders and approximately 30% of patients with SS have another associated autoimmune condition. To date, there are no studies that evaluate the frequency in which patients with initial diagnosis of pSS develop another autoimmune connective tissue disease in their evolution.

OBJECTIVES: Estimate the prevalence and incidence of development of another autoimmune rheumatic disease during the follow-up of patients with SSp using the GESSAR database (Sjögren Syndrome Study Group of the Argentine Society of Rheumatology). Detail the connective tissue diseases that occurred most frequently in these patients. Describe the clinical, serological and salivary gland histology characteristics of these patients.

MATERIALS AND METHODS: Observational study, retrospective cohort. We analyzed the data of patients diagnosed with pSS (American-European criteria 2002 / ACR-EULAR 2016), included in the GESSAR database. It was considered the development of another connective tissue autoimmune disease in those cases in which the diagnosis was added during follow-up.
STATISTIC ANALYSIS: For descriptive statistics, continuous variables were expressed as mean and standard deviation (SD) or median and interquartile range (IQR), according to distribution. Categorical variables were expressed in percentages. Continuous variables were compared with t-test or Wilcoxon rank-sum test. Categorical variables with chi-square or Fisher's exact test.

RESULTS: 681 patients were included, Table 1 details the general characteristics of the population. The prevalence of overlap of another autoimmune disease during follow-up was 4.4% (95% CI: 3.1-5.7%), 30 patients developed another connective tissue disease (CTD) (FIGURE 1). For the survival analysis, 670 patients provided information. The incidence of development of another CTD during follow-up was 0.91 per 100 patients per year (95% CI 0.58-1.24). The median follow-up was 4 years (IQR: 3-9). The differences between the general pablation and the patients who presented development of another connective tissue disease are expressed in table 2. Table 3 shows the clinical and laboratory characteristics presented by the patients prior to the development of the CTD.

TABLE 1: general characteristics

	N= 681
Female sex n (%)	511 (94,8)
Age, mean (± SD)	54 (±14)
Age to Dx. mean (± SD)	50 (± 13)
Time of follow -up, mean (± SD)	4.7 (± 4.9)
Xerophthalmia n(%)	618/659 (93.78)
Xerostomía n(%)	581/672 (86.46)
Schimer test n(%)	454/499 (90.98)
Positive Sialometry n(%)	213/258 (82.56)
RF + n(%)	303/589 (51.34)
Ro + n(%)	467/627 (74.48)
La + n(%)	262/608 (43.09)
Positive biopsy n(%)	345/400 (86.25)
Parotid swelling n (%)	186 (27.31)
Neuropathy n(%)	67 (9.84)
Arthralgias n(%)	436 (64.02)
Arthritis n(%)	191 (28.05)
Raynaud n(%)	101 (14.83)

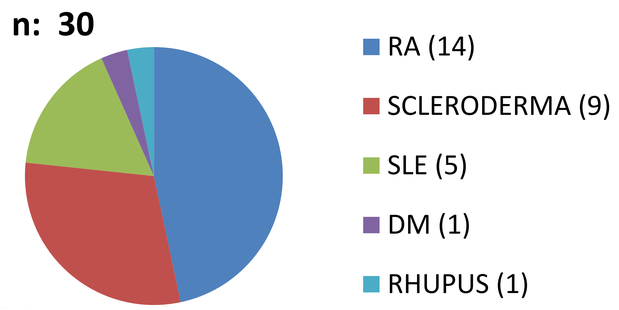

FIGURE 1:
RA: rheumatoid arthritis, LES systemic lupus erythematosus, DM: dermatomyositis.:

	pSS (n=651)	pSS + other CTD (n=30)	*p*
Female sex (%)	93.33	96.10	*0.34*
Mean age (± SD)	54 (±14)	53 (± 14)	*0.83*
Mean age to Dx (± SD)	50 (± 13)	48 (± 13)	*0.63*
Ro + (%)	74.12	82.14	*0.50*
La + (%)	43.55	33.33	*0.29*
RF + (%)	50.80	62.96	*0.21*
Positive biopsy (%)	86.53	78.57	*0.42*

TABLE 2 - Ssp; primary sjögren sindrome, CTD: connective tissue disease, RF: rheumatoid factor

DISEASE	PREVIOUS FEATURES
RA	78% arthralgia and arthritis 85.7% FR + 50% antiCCP dosage that resulted +
SCLERODERMIA	44% Raynaud 22.2% centromeric ANA 11% HTAP 33.3% GERD
LES	100% arthralgia 80% arthritis 100% ANA and RO +
RHUPUS	100% ANA + /100% RO + /100% Arthritis
DM	100% myalgia and myositis

TABLE 3: Previous features

CONCLUSIONS: The results showed a low percentage of development of another autoimmune rheumatic disease during follow-up. We consider it important to pay special attention to the presence of certain clinical and serological manifestations suggestive of this evolution.