Damage Accrual In a Single Centre Cohort Of Patients With Primary Sjögren's Syndrome Followed Up For Over 10 Years

ACR Abstract: #505

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Description:
Background/Purpose: 1) To describe the progression and cumulative prevalence of damage accrued over the time in a single center cohort of patients with primary Sjögren's Syndrome (pSS) 2) to explore the influence of the patients' clinical and serologic profile on damage accrual.

Methods: Consecutive unselected pSS patients (AECG criteria) were enrolled in this study between September 2012 and April 2013. Demographic, clinical, serologic, histological and therapeutic data of the patients enrolled were collected. Damage scores were assessed at 1, 3, 5 and 10 years post-diagnosis of pSS using both the Sjogren's Syndrome Damage Index (SSDI) and the Sjögren's Syndrome Disease Damage Index (SSDDI). The category/item of damage was also noted. Friedman test and Wilcoxon signed-rank test were used to evaluate damage progression over the follow-up.

Results:

The study cohort consisted of 155 pSS patients (7M:148F; median age at diagnosis= 49 years, IQR 40-58 years; median follow-up=5 years, IQR 3-10 years). The total increase of patients with damage was 28% after 1 year, 44% after 3 years, 74% after 5 years and 83% at the end of ourstudy. Median SSDI and SSDDI total damage scores steadily increased over the 10 years from 0 (IQR 0-1) to 2 (IQR 1-4) and from 0 (IQR 0-1) to 2 (IQR 1-3), respectively, with a significant correlation between SSDI and SSDDI scores (p=0.000). The domains mainly contributing to the total damage were the oral and the ocular items. More specifically, over the follow-up, 77/155 (49.5%) patients presented teeth loss and/or caries and 53/155 (34%) showed salivary flow impairment. Persistent salivary gland swelling was detected in 22/155 patients (14%) and was associated with a lower age at pSS diagnosis (p=0.002), anti-Ro/SSA (p=0.03), cryoglobulinemia (p=0.05), low C4 (p=0.04), hypergammaglobulinemia (p=0.000) and lymphocytopenia (p=0.000). Ocular damage was observed in 35/155 patients (22.6%) with corneal ulcers in 17/155 (11%) and tear flow impairment in 31/155 subjects (20%). Systemic damage manifestations were observed in 21/155 patients (13.5%) and correlated with the ESSDAI scores at the baseline (p=0.000), low C4 levels (p=0.005) and lymphocytopenia (p=0.01). Lymphoproliferative disorders were detected in 7/155 patients (4.5%) and malignancy in 14/155 (9%) cases.

Conclusion:

this study demonstrated that the vast majority of pSS patients developed damage within 10 years. Damage accrued mostly in the oral and ocular domains, however systemic damage manifestations and malignancy might be observed in the 10-15% of the patients.