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PRIMARY SJÖGREN SYNDROME AT A UNIVERSITY HOSPITAL: A RETROSPECTIVE ANALYSIS

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BACKGROUND

Diagnosis of primary Sjögren's syndrome (pSS) is based on objective dry eye/mouth assessment, presence of antibodies (e.g., anti-SSA/SSB) and minor salivary gland biopsy (MSG). However, pSS extraglandular manifestations (EGM) have been increasingly recognized. EULAR Sjögren's syndrome disease activity index (ESSDAI) combines EGM useful for diagnosis and follow-up in pSS. This study aims to describe demographic, clinical, serological, MSG and EGM findings in 57 pSS patients regularly followed-up at a rheumatology outpatient clinic of a university hospital.

MATERIALS AND METHODS

A retrospective analysis of the medical records was conducted. Information regarding epidemiological, clinical and demographic data, serological results, ocular staining score (OSS), MSG findings and ESSDAI manifestations present at diagnosis were recorded. Descriptive and comparative analysis were performed. A p-value < 0.05 was significant.

RESULTS

Fifty-four (95%) patients were female. At diagnosis of pSS, mean age was 49.11 ($\pm$ 11.82 years). Median delay in diagnosis was 5 (0–46) years. Twenty (35%) patients had Raynaud's phenomenon (RP); 41 (72%) reported dry mouth and 49 (86%) had dry eye. Forty-seven (82.5%) patients present anti-SSA and 18 (31.6%) had anti-SSB; 33 (57.9%) had rheumatoid factor (RF) and 3 (5.3%) had anti-RNP positive. Ocular staining score $\geq$ 5 occurred in 28 (66%) patients. Thirty-four (59%) patients were submitted to MSG, 89.5% presenting FS $\geq$ 1. Median total ESSDAI at diagnosis was 2 (0–32). The FS median of MSG was 2 (0–7). Delay in diagnosis correlated negatively with FS (p = 0.028) and positively with the disease length (p = 0.001). Positive anti-SSA reduced diagnostic delay (p = 0.036). The score of ESSDAI respiratory assessment was lower in patients with FS $\geq$ 1; furthermore, hematological score was associated positively with RP.

CONCLUSION

Although the knowledge in pSS has increased exponentially in past decades, the delay in diagnosis is still expressive. Dry symptoms are prevalent but nonspecific. In agreement with a large cohort of pSS, RP was frequent in the studied sample. Anti-SSA is an accessible and useful test to confirm clinical suspicion of pSS and therefore reduce time for diagnosis. Otherwise, infrequent clinical presentation and long-term disease may be associated with diagnostic delay and with a worse performance of MSG due to glandular fibrosis.

REFERENCES

1. Retamozo S, Acar-Denizli N, Rasmussen A, Horváth IF, Baldini C, Priori R, et al. Systemic manifestations of primary Sjögren's syndrome out of the ESSDAI classification: prevalence and clinical relevance in a large international, multi-ethnic cohort of patients. *Clin Exp Rheumatol*. 2019;37Suppl118(3):97–106.
2. Seror R, Bowman SJ, Brito-Zeron P, Theander E, Bootsma H, Tzioufas A, et al. EULAR Sjögren's syndrome disease activity index (ESSDAI): a user guide. *RMD Open*. 2015;1:e000022. <https://doi.org/10.1136/rmdopen-2014-000022>
3. Shiboski CH, Shiboski SC, Seror R, Criswell LA, Labetoulle M, Lietman TM, et al. 2016 American College of Rheumatology/European League Against Rheumatism Classification Criteria for Primary Sjögren's Syndrome: A consensus and data-driven methodology involving three international patient cohorts. *Arthritis Rheumatol*. 2017;69(1):35–45. <https://doi.org/10.1002/art.39859>