Nasal and paranasal involvement in primary Sjögren's syndrome*

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Summary

Background: The aim of this study is to investigate nasal and paranasal signs and symptoms of the primary Sjögren's syndrome patients and compare them with healthy controls.

Methodology: Seventy-seven (7 M, 70 F) primary Sjögren's syndome patients and 77 healthy controls were included in the study. Anterior rhinoscopy, nasal endoscopy, 5 component smell discrimination test, nasal clearance analysis with saccharin test and electrorhinomanometer were performed.

Results: Nasal crusting was present in 31 and 24 individuals in patient and control groups, respectively. Sinusitis was present in 2 and 1 individuals in patient and control groups, respectively. Nasal polyposis was present in 7 and 1 individuals in patient and control groups, respectively. These differences were not statistically different.

Conclusion: Although there were some findings in a few patients, nasal findings were insignificant and mild even in patients with severe oral or ocular findings. Rhinomanometry, nasal clearance determination or smell discrimination tests have very little value in the diagnosis or management of primary Sjögren's syndrome. Nasal polyposis was higher in the patient group, though it did not reach a significant level. Nasal glandular involvement is mild and insignificant in primary Sjögren's syndrome.

Key words: Sjögren's syndrome, nose, paranasal sinus

Introduction

Sjögren's syndrome is an autoimmune disease which affects exocrine glands and causes dryness of mucosa, skin and conjunctiva [(1)]. The prevelance of the disease is 1-3%, however, most patients live without the diagnosis due to a slow progression of the inflammation. The syndrome is named as primary Sjögren's syndrome if another rheumatological disease is absent. It is systemic with the involvement of the lungs, liver, kidneys, vessels and blood, yet it manifests mainly with opthalmological and otolaryngological symptoms. Oral cavity, ear and salivary gland involvement of the syndrome were investigated by many studies and reports previously. However, there is very little information about the nasal and paranasal involvement of the syndrome. Since nasal mucosa is comprised of exocrine glands, Sjögren's syndrome might present itself with nasal symptoms as well.

The aim of this study is to investigate nasal and paranasal signs and symptoms of the primary Sjögren's syndrome patients and compare them with healthy controls.

Materials and methods

Study population

The study was conducted in Ege University Medical School Hospital between 2001 and 2002. Seventy-seven (7 M, 70 F) primary Sjögren's syndome patients diagnosed and followed-up in the Rheumatology Department were included in the study. The patients were diagnosed according to the criteria of the European Study Group on Diagnostic Criteria for Sjögren's Syndrome [2]. Seventy-seven (7 M, 70 F) healthy individuals were included in the study as controls. The control group was chosen from patient companions on a voluntary basis. Patients or healthy controls receiving medical therapy were excluded from the study as this could affect nasal symptoms or signs. The mean age was 48.4 years (23-70) and 46.1 years (22-72) in the patient and control groups, respectively. Consent was obtained from all individuals included in the study. Clinical experiments in the study conform the principals outlined by the Declaration of Helsinki.

Patient analyses

Detailed systemic and otolaryngological physical examinations were performed on all patients and healthy controls. Nasal symptoms questionnaires were applied to all subjects. It questioned symptoms including nazal dryness, nasal crusting, nasal obstruction, epistaxis and allergic rhinitis. Allergy positivity was determined by patient history.

Anterior rhinoscopy, nasal endoscopy, a previously validated 5 component smell discrimination test, nasal clearance analysis with saccharin test and electrorhinomanometer were performed to all individuals. If at least one component was abnormal, the smell discrimination test was regarded as abnormal. Active anterior rhinomanometry was established in constant 150 pascal as described by European Rhinomanometry Standardization Comittee [3]. Nasal resistance was calculated as dividing the pressure difference by air volume (R = P/V). Radiological work-up was performed in case of any chronic nasal symptoms. Sinus waters radiography was taken from 12 patients and coronal paranasal computed tomography (CT) scan was taken from 39 patients.

Statistical analysis

Statistical Package for Social Sciences (SPSS 12.0) software was used in statistical analysis. Both groups were compared with Chi-square test, Fishers Exact test and t-test.

Results

Allergy was present in 15 (19.5%) and 14 (18.2%) individuals in patient and control groups, respectively ($p > 0.05$). Epistaxis was present in 3 (3.8%) and 4 (5.2%) individuals in patient and control groups, respectively ($p > 0.05$). Nasal crusting was present in 31 (40.3%) and 24 (31.2%) individuals in patient and control groups, respectively ($p > 0.05$). Sinusitis was present in 2 (2.6%) and 1 (1.3%) individuals in patient and control groups, respectively ($p > 0.05$). Nasal polyposis was present in 7 (9.1%) and 1 (1.3%) individuals in patient and control groups, respectively. A difference in nasal polyposis was present between the two groups. However, it could not reach a statistically significant level ($p > 0.05$).

Saccharin clearance time was 16.6 minutes (13-20 minutes) and 14.5 minutes (12-16 minutes) in patient and control groups respectively ($p > 0.05$). Smell discrimination test was abnormal in 2 (2.6%) and 1 (1.3%) in patient and control groups, respectively ($p > 0.05$). The difference between nasal resistance and nasal volume scores were statistically insignificant ($p > 0.05$). There were no statistically significant differences in rhinomanometry measurements ($p > 0.05$).

Thirty-nine patients underwent coronal paranasal CT scan. Seven patients had nasal polyposis, one with the sphenoid retention cyst, three with the maxillary retention cysts and three had concha bullosa in their middle turbinates on CT scan.

Discussion

Sjögren's syndrome is a chronic disease and relatively hard to diagnose. It's important to define and diagnose the nasal symptoms and signs of it. There are different studies that investigate the ear and the neck findings of the syndrome. Nevertheless, there is little information about the nasal and paranasal symptoms. Secondary Sjögren's syndrome patients were excluded to define the nasal symptoms more reliably.

Prevelance of Sjögren's syndrome in females is 9 times higher than males [1]. The female/male ratio was 10 in our study. Hormonal immuno-regulation was proposed as the major factor for female predominance. Disease prevelance is highest among post-menapausal women between the fifth and the seventh decades and this finding supports the hormonal hypothesis for gender predominance.

The major pathway in Sjögren's syndrome's pathophysiology is chronic immune system stimulation. The main histopathological finding in target tissue is lymphocyte infiltration. B cell activation is the most common finding of the disease. B and T cell infiltration are associated with exocrine gland damage. One may think that patients with this syndrome may suffer from nasal mucosal dryness, crusting and eventually epistaxis. Hochberg and Mahoney reported increased nasal crusting and epistaxis in Sjögren's syndrome patients [1,4]. However, in our study, nasal crusting as a sign or symptom and nasal dryness were not statistically significant between the disease and the control groups. It

was not prominent even in patients with severe oral and ocular findings.

Decreased exocrine gland secretion may cause dryness in nasal mucosa, increased nasal congestion as well as nasal resistance and may eventually lead to delayed nasal clearance. Nasal clearance in saccharin test was found 17 ± 3 minutes [5,6]. Mean nasal clearance was slightly increased in our patient group; yet it did not reach a statistically significant level. This interesting finding may be associated with the mild involvement of the nasal mucosa in Sjögren's syndrome.

Based on endoscopic and radiological findings, we diagnosed seven pansinusitis with nasal polyposis and two mild maxillary sinusitis in the patient group. There were some mild findings of mucosal disease such as retention cysts or few mild sinusitis patients. However, sinusitis and its association with the Sjögren's syndrome was not significant in our study. Septal perforation was previously reported to be significantly higher among Sjögren patients (13%) [7-9], but we did not diagnose any septal perforation in our patients. Only two patients described smell disorder. One had stage 4 nasal polyposis and the other had stage 2 nasal polyposis. Smell disorder was associated with nasal polyposis rather than mild mucosal involvement of the disease. Freeman reported frequent nasal symptoms in Sjögren's syndrome, and relatively less frequent nasal findings in rhinoscopy [10]. We believe that the difference between the previous studies and our study was primarily due to the patient selection criteria since we only included primary Sjögren's syndrome patients in this study. Association of nasal polyposis with primary Sjögren's syndrome was one of the new findings of this study. Larger series are warranted to enlighten this association. A nasal mucosa biopsy could be beneficial to clarify this association.

In conclusion, 77 patients with primary Sjögren's syndrome had none or mild nasal and paranasal signs and symptoms in our study. Although there were some findings in few patients, they were insignificant and mild even in patients with severe oral or ocular findings. Rhinomanometry, nasal clearance determination or smell discrimination tests have very little value in the diagnosis or management of the primary Sjögren's syndrome. Nasal polyposis was higher in the patient group, however, it did not reach a significant level. The diagnosis of the disease seems to be very hard based on nasal or paranasal symptoms and signs. We may conclude that nasal glandular involvement is mild and insignificant in primary Sjögren's syndrome.

Authorship contribution

RM: design, interpreting results
SG: writing of the manuscript
GO: patient selection
YK: patient selection
BK: design, patient selection

Conflict of interest

The authors state that there is no conflict of interest regarding this manuscript.

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