SJÖGREN'S SYNDROME: CLINICAL, CYTOLOGICAL, HISTOLOGICAL AND COLPOSCOPIC ASPECTS IN WOMEN

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Summary: Sjögren's syndrome, characterized by a progressive alteration of the exocrine glands which ultimately results in their atrophy, has the highest incidence among females.

The main etiopathogenetic mechanism is autoimmunological. The symptomatology is made up of symptoms depending on an altered glandular secretion and mucosal dryness. Dyspaurenia and pruritus, due to vaginal and vulvar dryness, are quite common symptoms.

Few studies have considered the clinical and histological consequences of Sjögren's syndrome in the external female genitalia. In the present study 26 women (mean age 46 years) affected by Sjögren's syndrome were examined by means of a series of tests including gynecological examination exam, colposcopic inspection and cervical biopsy.

All the patients showed a particular drynof a series of tests including gynecological examination failed to evidence malign cells and, in 15% of the subjects, showed an estrogenic insufficiency in various degrees.

The colposcopic inspection revealed dystrophic processes resulting in the atrophy of the cervicovaginal mucosa in 50% of the cases.

The histological findings of the cervical biopsies evidenced the presence of a chronic cervicitis in 10% of the cases.

Key words: Sjögren's syndrome; colposcopy; genital tract.

INTRODUCTION

Sjögren's syndrome was systematically described for the first time in 1933 by the Swedish opthalmologist who gave it its name ([1]).

The major clinical manifestations (xerostomia, xeropthalmia and keratoconjunctivitis sicca) are attributable to decreased lacrimal and salivary secretions secondary to an inflammatory infiltrate causing organic and functional glandular damage.

Sjögren's syndrome may occur alone (idiopathic Sjögren's syndrome) or in association with systemic or localized diseases of autoimmune nature ([2]). In fact, it has been described in 30% of the cases of rheumatoid arthritis, in 50% of the cases of systemic lupus erythematosus, in 40% of the cases of scleroderma and in a variable percentage of the cases of mixed cryoglobulinemia, chronic active hepatitis, primary biliary cirrosis and autoimmune thyroiditis ([3]). The basic histopathologic lesion is a lymphocytic infiltrate in the salivary and lacrimal glands causing atrophy of the secretory structures followed by fibrosis ([2]).

In the idiopathic type of Sjögren's syndrome many exocrine glands are compromised ([3]): nasal, laryngeal, bronchial, gastric, cutaneous, pancreatic and sweat glands all show dystrophic and functional lesion in various areas or an increased likelihood of infection (table 1). The diagnostic criteria in Sjögren's syndrome are described in table 2.

The presence in these patients, who are almost all women, of symptoms localized to the external genitalia, was first noted by Bloch and Buchanan ([7]) in their classic work in 1965 where they reported, in a large percentage of cases, dyspareunia, vulvar pruritus and vaginal dystrophy in most cases.

The gynecological examination showed vaginal erythema and irritation which the authors hypothized could be due to decreased function of the vulvar and vaginal

Table 1. – *Exocrine glands which can be affected in Sjögren's syndrome.*

Localization	*Clinical effects*
Lacrimal	keratoconjunctivitis sicca
Salivary	xerostomia, loss of teeth
Cutaneous	cutaneous dryness
Pharynx and oesophagus	dysphagia
Stomach	atrophic gastritis
Nasal cavity	rhinitis sicca
Respiratory tract	dry cough, infections
Pancreas	chronic pancreatitis
External genitalia	dyspareunia and pruritus

Table 2. – *Primary Sjögren's syndrome: diagnostic criteria.*

1) Presence of xerostomia and xerophtalmia.
2) Presence of keratoconjunctivitis sicca evidenced by Schirmer's test (4) and/or rose Bengal staining (4).
3) Presence of salivary compromission detected by radiological (sialography (5)) and/or histological (labial biopsy (6)) exams.
4) Absence of indications and symptoms of other connectivitis.

lubrification glands caused by Sjögren's syndrome.

No further systematic study has been performed to clarify this issue even though there are many reports of this type of gynecological problem associated with Sjögren's syndrome in Literature.

The purpose of this study is to evaluate the genital symptoms and signs seen in association with Sjögren's syndrome.

MATERIAL AND METHODS

26 patients affected by Sjögren's syndrome were divided into two groups. The first group (Group 1) was formed by 9 menopausal women of mean age 55.5 yr., the second (Group 2) by 17 women in the fertile period of life, mean age 38.8 yr.

Accurate histories were taken from both groups of patients along with: gynecological visit, cytological examination of the vagina and of the cervix to evidence eventual cellular atypia and to assess hormonal status; colposcopic inspection and cervical biopsy were also performed for a histopathological assessment of the cervix.

RESULTS

Table 3 shows symptoms and signs localized in the genital tract of the patients examined.

Table 3. – *Incidence of symptoms and clinical features in the 26 female patients with Sjögren's syndrome.*

	Group 1 Menopausal age (9 subjects) No. %	Group 2 Fertile age (17 subjects) No. %
Vaginal dryness	9 (100)	9 (52.9)
Dyspareunia	6 (66.6)	12 (70.6)
Vaginal pruritus and/or burning	5 (55.5)	6 (35.3)
Erythematous vulvitis	6 (66.6)	6 (35.3)
Uterine fibroma	2 (22.3)	4 (23.5)
Ovarian cysts	–	1 (5.9)
Negative internal genital finding	7 (77.7)	12 (70.6)

Table 4 shows the results of the cytological endocrine assessment, colposcopic and histological findings respectively of the first and second group.

In the menopausal group, vaginal dryness was present in 100% of the 9 women, dyspareunia in 66.6%, pruritus and or burning in 55.5%; 66.6% of these women had a clinical picture of erythematous vulvitis. As far as the internal reproductive organs were concerned, 77.7% were normal and 22.3% had uterine leiomyomas.

Of the 17 women of reproductive age, vaginal dryness was seen in 9 subjects (52.9%), dyspareunia in 12 cases (70.6%), pruritus and/or burning in 6 cases (35.3%). Erythematous vulvitis was present in 6 women (35.3%). 12 women

Table 4. – *Cytological, colposcopic and histological findings in the 26 female patients with Sjögren syndrome.*

	Group 1 No.	%	Group 2 No.	%
Cytological endocrine findings				
Normal estrogenic effect	2	(22.3)	13	(76.5)
Elevated estrogenic effect	3	(33.3)	–	
Mild estrogenic insufficiency	–		1	(5.9)
Moderate estrogenic insufficiency	4	(44.5)	–	
Severe estrogenic insufficiency	–		–	
Luteal insufficiency	–		–	
Normal luteinic effect	–		3	(17.6)
Colposcopic findings				
Normal cervix	2	(22.3)	4	(23.5)
Dystrophic conditions	5	(55.4)	2	(11.8)
Inflammatory conditions	2	(22.3)	5	(29.4)
Ectopy	–		7	(41.2)
Normal Transformation Zone	–		3	(17.6)
Atypical Transformation Zone	–		1	(5.9)
Histological findings				
Normal cervical tissue	6	(66.7)	9	(52.9)
Non specific chronic cervicitis	3	(33.3)	8	(47.1)

(70.6%) did not present any pathology of the internal reproductive organs, 4 patients (23.5%) were affected by uterine leiomyomas and in 1 woman (5.9%) a left ovarian cyst was found.

Results of the cytological examination or Pap test failed to evidence displastic or malign cells in any subjects studied.

Table 4 demonstrates that in the first group 4 women had moderate estrogenic insufficiency, 2 women had normal estrogen effects and 3 women had increased estrogen effects. Colposcopic findings reported are atrophic conditions in 5 cases, inflammatory processes in 2 cases and a normal cervix was seen in 2 cases. Histological evaluation evidenced 3 cases of chronic non-specific cervicitis and in the remaining 6 cases no abnormality was found. Table 4 also reports the results of these tests in the subjects of group 2.

Normal cytologic estrogen effects were seen in 13 women (76.5%), 1 case (5.9%) of mild estrogenic insufficiency, and 3 cases (17.6%) of normal luteinic effect were seen.

Colposcopically, a normal cervix was seen in 4 cases (23.5%), dystrophic conditions in 2 cases (11.8%), inflammatory conditions in 5 cases (29.4%), ectopy in 7 cases (41.2%), NTZ (Normal Transformation Zone) was seen in 3 cases (17.6%) and ATZ (Atypical Transformation Zone) was seen in 1 case (5.9%).

Histologically, a report of normality was found in 9 cases (52.9%) and non-specific chronic cervicitis was found in 8 patients (47.9%).

CONCLUSIONS

The resu.lts of this study indicate that the subjects with idiopathic Sjögren's syndrome present a characteristic gynecological clinical picture characterized by vaginal dryness, dispareunia, vulvar pruritus and/or burning that cannot be attributed only to the vulvo-cervico-vaginal dystrophic processes normally seen in post-menopausal women, as these conditions are also present in a high percentage of women of reproductive age with normal hormonal findings.

The pathologic changes are localized mainly to the vulva and are seen as a non specific inflammation most often in the form of an erythematosus voulvitis, which was seen in a high percentage of subjects (42.1% of all subjects examined).

Etiopathogenetic interpretation of the vulvar findings is difficult but they are most likely due to the same mechanisms responsible for the mucous alterations seen in other areas of the body.

The results of colposcopic examination indicate that there are no alterations distinctive enough to be considered indicative of idiopathic Sjögren's syndrome. However, interesting results were obtained by means of histological investigation which revealed chronic non specific cervicitis in 11 cases (42.2%) whereas colposcopic investigation revealed acute or chronic inflammation in only 7 cases (26.9%).

Is also of note that many patients with the above mentioned symptomatology had been treated in the past with topical anti inflammatory agents and/or with topical estrogens without symptomatic relief.

We want to continue our study with the specific intent of verifying whether or not the genital symptomatology as described above is responsive to the specific treatment of Sjögren's syndrome.

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