Case Report / Olgu Sunumu

Co-Existence of Oral Lichen Planus and Primary Sjogren's Syndrome

Oral Liken Planus ve Primer Sjögren Sendromu Birlikteliği

Tayfun Güngör[1], Muhammed Şahin[1], Seda Doğan[2], Ayşe Gökçe Yıldız[3], Adem Küçük[4], Salim Güngör[3]

[1]*Necmettin Erbakan University, Meram Faculty of Medicine, Department of Physical Therapy and Rehabilitation, Konya, Turkey*
[2]*Necmettin Erbakan University, Meram Faculty of Medicine, Department of Dermatology, Konya, Turkey*
[3]*Necmettin Erbakan University, Meram Faculty of Medicine, Department of Pathology, Konya, Turkey*
[4]*Necmettin Erbakan University, Meram Faculty of Medicine, Department of Internal Medicine, Rheumatology Division, Konya, Turkey*

ABSTRACT

Sjogren's syndrome is a chronic autoimmune disease associated with the lymphocytic infiltration of exocrine glands such as the salivary and lacrimal glands. Lichen planus is a chronic inflammatory disease which may affect the skin, nails, scalp and mucous membranes. Both diseases show similarities in many aspects such as oral mucosal involvement, etiopathogenesis and relation with immune diseases. And the studies report the concurrence of these two diseases.We diagnosed a 55-year-old female patient, who had had mouth ulcers for a year, with oral lichen planus and Sjogren's syndrome. We aim to draw attention to co-existence of these two diseases which have very limited data in the literature.

Keywords: Oral lichen planus, Sjogren's syndrome, autoimmunity

ÖZET

Sjögren sendromu, tükrük ve gözyaşı gibi ekzokrin bezlerin lenfositik infiltrasyonu ile karakterize kronik otoimmün bir hastalıktır. Liken planus ise deri, tırnak, skalp ve mukoz membranları etkileyebilen kronik inflamatuvar bir hastalıktır. Her iki hastalık mukozal tutulum, etyopatogenez ve otoimmün hastalıklarla ilişki gibi bir çok yönden benzerlikler gösterir. Çalışmalarda bu iki hastalığın birlikteliğinden bahsedilmektedir. Biz bir senedir ağız yaraları olan 55 yaşında kadın hastaya oral liken planus ve Sjögren sendromu tanısı koyduk. Literatürde birlikteliğine dair sınırlı veri bulunun bu iki hastalığa dikkat çekmek istiyoruz.

Anahtar sözcükler: Oral liken planus, Sjögren sendromu, otoimmünite

Corresponding Author
Yazışma Adresi

Tayfun Güngör

Necmettin Erbakan Üniversitesi, Meram Tıp Fakültesi, Fiziksel Tıp ve Rehabilitasyon Anabilim Dalı, Konya, Turkey

E-mail: drtgungor@hotmail.com

Received/Geliş Tarihi: 03.03.2014
Accepted/Kabul Tarihi: 25.03.2015

Introduction

Sjogren's syndrome (SS) is a chronic autoimmune disease associated with the lymphocytic infiltration of exocrine glands such as the salivary and lacrimal glands (1). SS may be primary or secondary to other autoimmune diseases such as rheumatoid arthritis. The frequency of the disease varies between % 0,5- 2,7 according to the different classification criteria. The estimated prevalence of the primary SS is about % 1-1,5. It is seen especially 40-50 years old women and the female:male ratio is 9:1 (2,3).

Symptoms and findings such as impaired oral hygiene associated with dry mouth, difficulty in eating dry foods, parchment-like tongue, and dry and cracked lips are observed, and these pathologies may result in candidiasis and dental cavities (4).

Furthermore, oral lesions of autoimmune etiology (OLAIE) are usually ignored in SS patients. Likar-Manookin et all reported that OLAIE such as lichen planus (LP) and ulcerative stomatitis are not uncommon among SS patients (5). One of these lesions, LP, is a chronic

inflammatory disease which may affect the skin, nails, scalp and mucous membranes (6). LP is characterised by isolated or groups of papulas which have special locations and these papulas have different colours and distrubition(7). The prevalence of the disease is less than % 1. The incidence of the disease may vary according to the regions such as % 0,78 in Sweden, % 0,44 in USA, % 0,14 in Palestine. In a study it is reported % 0,68 of the patients who admitted to the dermatology outpatient clinic is diagnosed as LP(8).

Oral lichen planus (OLP), a subtype of LP, involves the tongue, lips and hard palate, particularly the buccal mucosa (9). It has various clinical types such as bullous, atrophic, reticular, papullar, plaque-like and erosive (10). The mucosal involvement is seen 2/3 of the LP patients. Oral and genital regions are the most common localized for mucosal involvement. Mucosal involvement may also be the first and only sign of the disease in the % 15-25 of LP patients. The frequency of the oral mucosal involvement is about %15-35 and it increases up to % 50-65 with the cutanous involvement (7). OLP is diagnosed by the histopathological examination of the characteristic lesions (11).

In this case, we are reporting a patient whom we diagnosed with OLP(histopathologically reticular patern and sporadic erosions) and SS.

Case Report

A 55-year-old female patient applied to our outpatient clinic with complaints of dry mouth for 2 years, gritty sensation of the eyes, and oral ulcers which had been increasing for the last 1 year. The patient also showed signs of Raynaud's phenomenon. The patient had no family history, and did not smoke or consume alcohol. The patient's physical examination did not reveal any findings of arthritis or rash. However, she had bilateral symmetrical reticular web structure with sporadic erosions in the oral mucosa and upper and lower gingival, and extensions to the buccal mucosa over the erosions (Figure 1). No pathology was found in the examination of the genital mucosa. Her laboratory test results were as follows: erythrocyte sedimentation rate: 15 mm/hour, CRP: 1.83 mg/L, RF: <20 IU/mL, ANA: Homogenous 1/160, Schirmer test 2/4. Because of these results, a salivary gland biopsy requesting an ENA profile was performed on the patient (Figure 2). Mononuclear cell infiltration around the ductal structures was observed, and the histopathological appearance was consistent with Sjogren's syndrome (Grade II according to Masson and Chisholm classification). Anti SS-A was positive, but anti-SS-B was negative in the ENA profile. Additionally, Anti-dsDNA was negative. Sporadic parakeratosis was presented on the epidermis and was becoming more visible on the granular layer, and mononuclear inflammatory cell infiltration under the epithelium on the epidermis was observed in the biopsy of the patient's oral lesion (Figure 3). Histopathological appearance was consistent with lichen planus. Also the patient was diagnosed with Sjogren's syndrome according to American-European classification criteria.

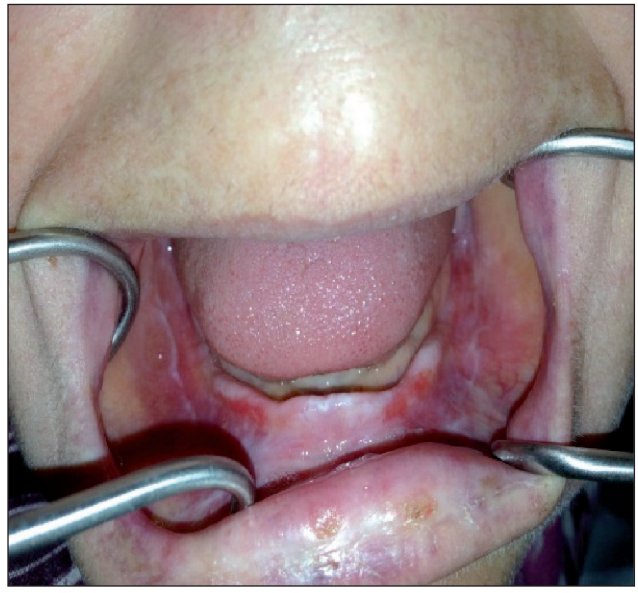

Figure 1. Lichen planus: Sporadic erosions in the gingiva, bilateral symmetric reticular web structure with extensions to the buccal mucosa.

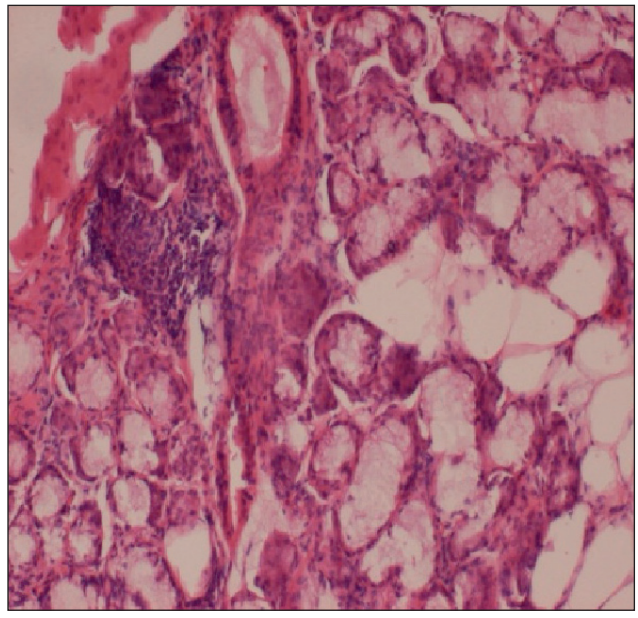

Figure 2. Salivary gland biopsy: Lymphocyte infiltration around the ductal structures, focal lymphoid infiltration in the upper left corner.

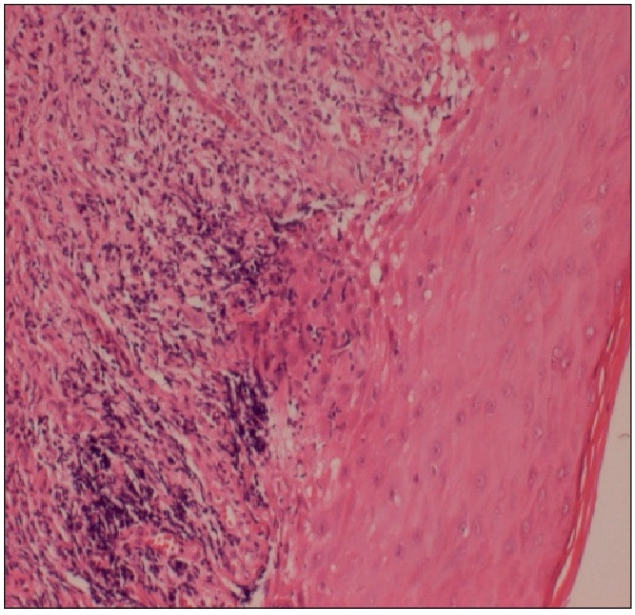

Figure 3. Biopsy from the lesion in the oral mucosa: Band-shaped lymphocytic infiltration in the dermoepidermal junction.

Discussion

SS and LP are chronic inflammatory diseases and similar in many aspects. The etiology of both diseases are unknown, but they are thought to be autoimmune conditions. SS and LP are less common in childhood and more common in women over 40 years of age (9, 12-14). In SS, the risk of lymphoproliferative tumors increases with environmental factors, cytokine stimulation, viral infections and genetic events (15). The risk of oral cancer also increases in addition to NHL (16). Particularly the erosive and atrophic variants of OLP (the prevalence is % 0.2 - 4) have tumor potential (9, 14, 17).

While SS is a rheumatic disease which involves many tissues and organs such as the mucosal membrane, skin, lungs and kidneys, and particularly the exocrine glands, LP is similar to SS with its involvement of tissues such as the mucosal membranes, skin and nails (9, 12).

Since the protective effect of saliva is eliminated in SS patients, these patients may experience a modified sense of taste, difficulty in speaking and swallowing, loss of teeth, and may develop oral infections. Nonspecific oral lesions are also common in SS (5).

It has been reported that autoimmune diseases are observed in 33% of SS patients. These comorbid conditions include primary biliary cirrhosis, hypothyroidism, Graves disease, discoid lupus, celiac disease and scleroderma (18). OLP is associated with many factors such as stress, chronic liver diseases, anxiety, diabetes, infections, hypertension and autoimmune disorders which include systemic lupus erythematosus (SLE), rheumatoid arthritis (RA) and SS (19, 20). This means that both SS and LP are associated with autoimmune disorders, and both have immune dysfunction. T-lymphocyte infiltration is a common immunological factor in SS and LP (5). Sistig S et al. (21) have obtained data on this relationship in their study with respect to the role of serum immunoglobulin levels in the pathogenesis of a group of oral diseases including OLP and SS. Previous studies have also reported that immunoglobulin levels increase in both OLP and SS (22, 23). Additionally, while cytokines play an important role in the pathogenesis of SS (24), they are also associated with pathophysiology of LP (25). This data gives us the idea that both diseases are also related in pathological manner.

OLP is diagnosed by histopathological examination. The characteristics of the lesions are : hyperkeratosis without parakeratosis, focal increase in the granular cell layer, irregular acanthosis which leads to a "saw blade" appearance, liquefaction degeneration in the basal cell layer and band-shaped lymphocytic infiltration in the dermoepidermal junction (11).

Our case had been experiencing mucocutaneous symptoms for 2 years. She also had oral lesions, which had been increasing for 1 year. She had bilateral symmetric reticular web structure with sporadic erosions in the oral mucosa and gingiva and extensions to the buccal mucosa over the erosions. The patient had ANA, Schirmer test, SS-A positivity, and the biopsy of her minor salivary gland was compatible for SS. The histopathological appearance of the biopsy obtained from the patient's oral lesion was consistent with lichen planus. The patient was diagnosed with Sjogren's syndrome and lichen planus. OLP is usually diagnosed with clinical and histological assessments (26). Another point of interest in this case was that the patient was SS-B negative. In their prevalence study, Likar-Manookin K et al. (5) found the SS-B antibodies to be significantly negative in patients diagnosed with SS and having oral lesions, and this situation was considered a potential risk factor.

In our case, systemic prednisolone 40 mg/day and also diclofenac sodium + triamcinolone acetonide gel 3x1, benzydamine hydrochloride + chlorhexidine gluconate oral rinse (3x1 as local treatment) was administered for treatment of OLP. The patient's complaints regressed in the follow-ups, and her steroid dose was gradually reduced. Additionally, hydroxychloroquine 200 mg tablets 2x1 were administered for SS. The patient responded positively to the treatment. Her oral lesions clearly improved.

In summary, the coexistence of SS and OLAIE has been drawing more attention lately. Since the mucocutaneous symptoms emerged earlier in our case we suppose that SS might have triggered the development of LP.

Another possibility is the hypothesis that the same immunological, genetic and/or environmental factor might have played a role in the development of the mentioned diseases because both diseases have similar features in etiopathogenesis. Therefore, we wanted to draw attention to the coexistence of OLAIE and SS.

References

1. Margaix-Munoz M, Bagan JV, Poveda R, et al. Sjogren's syndrome of the oral cavity. Review and update. Med Oral Patol Oral Cir Bucal 2009;14:325-30.
2. Aksu K. Sjögren sendromunda immunsupresif tedavi. RAED Dergisi 2009;1:35-41.
3. Karabulut G, Kitapçıoğlu G, Argın M, ve ark. Primer Sjögren sendromlu olgularda sakroiliit sıklığı nedir?. Ege Journal of Medicine 2011;50:43-7.
4. Yan Z, Young AL, Hua H, et al. Multiple oral candida infections in patients with Sjogren's syndrome -- prevalence and clinical and drug susceptibility profiles. J Rheumatol 2011;38:2428-31.
5. Likar-Manookin K, Stewart C, Al-Hashimi I, et al. Prevalence of oral lesions of autoimmune etiology in patients with primary Sjogren's syndrome. Oral Dis 2013;19:598-603.
6. Ghaleyani P, Sardari F, Akbari M. Salivary IgA and IgG in oral lichen planus and oral lichenoid reactions diseases. Adv Biomed Res 2012;1:73.
7. Ceyhan AM, Yüksel Babak P, Baysal Akkaya V, ve ark. Liken planusun farklı klinik yüzleri:olgu sunumu. SDÜ Tıp Fak Derg 2007;14:33-6.
8. Yanık ME, Aliağaoğlu C, Turan H, ve ark. Son 10 yılda kliniğimizde takip edilen liken planus olgularının retrospektif değerlendirilmesi. Tıp Araştırmaları Dergisi 2012;10:6-11.
9. Moger G, Thippanna CK, Kenchappa M, et al. Erosive oral lichen planus with cutaneous involvement in a 7-year-old girl: a rare case report. J Indian Soc Pedod Prev Dent 2013;31:197-200.
10. Lavanya N, Jayanthi P, Rao UK, et al. Oral lichen planus: An update on pathogenesis and treatment. J Oral Maxillofac Pathol 2011;15:127-32.
11. Bolognia JL, Jorizzo JL, Rapini PR. Lichen planus and lichenoid dermatosis. In: Tetsuo Shiohara, Yoko Kano, editors. Dermatology. Mosby: Elseiver third edition, 2008:183-203.
12. Tincani A, Andreoli L, Cavazzana I, et al. Novel aspects of Sjögren›s syndrome in 2012. BMC Med 2013;11:93.
13. Aminzadeh A, Jahanshahi G, Ahmadi M. A retrospective comparative study on clinico-pathologic features of oral lichen planus and oral lichenoid lesions. Dent Res J (Isfahan) 2013;10:168-72.
14. Gümrü B. A retrospective study of 370 patients with oral lichen planus in Turkey. Med Oral Patol Oral Cir Bucal 2013;18:427-32.
15. Dong L, Chen Y, Masaki Y, et al. Possible mechanisms of lymphoma development in Sjögren›s syndrome. Curr Immunol Rev 2013;9:13-22.
16. Zhang W, Feng S, Yan S, et al. Incidence of malignancy in primary Sjogren›s syndrome in a Chinese cohort. Rheumatology (Oxford) 2010;49:571-7.
17. García-García V, Bascones Martínez A, Martinelli-Kläy CP, et al. New perspectives on the dynamic behaviour of oral lichen planus. Eur J Dermatol 2012;22:172-7.
18. Lazarus MN, Isenberg DA. Development of additional autoimmune diseases in a population of patients with primary Sjögren›s syndrome. Ann Rheum Dis 2005;64:1062-4.
19. Ismail SB, Kumar SK, Zain RB. Oral lichen planus and lichenoid reactions: etiopathogenesis, diagnosis, management and malignant transformation. J Oral Sci 2007;49:89-106.
20. Scully C, Beyli M, Ferreiro MC, et al. Update on oral lichen planus: etiopathogenesis and management. Crit Rev Oral Biol Med 1998;9:86-122.
21. Sistig S, Vucićević-Boras V, Lukac J, et al. Salivary IgA and IgG subclasses in oral mucosal diseases. Oral Dis 2002;8:282-6.
22. Gandolfo S, Carrozzo M, Carbone M, et al. Humoral immunological parameters in Italian patients with oral lichen planus. Bull Group Int Rech Sci Stomatol Odontol 1994;37:71-7.
23. Talal N, Asofsky R, Lightbody P. Immunoglobulin synthesis by salivary gland lymphoid cells in Sjögren's syndrome. J Clin Invest 1970;49:49-54.
24. Huang YF, Cheng Q, Jiang CM, et al. The immune factors involved in the pathogenesis, diagnosis, and treatment of Sjogren›s syndrome. Clin Dev Immunol 2013;2013:1-6.
25. Chauhan I, Beena VT, Srinivas L, et al. Association of cytokine gene polymorphisms with oral lichen planus in Malayalam-speaking ethnicity from South India (Kerala). J Interferon Cytokine Res 2013;33:420-7.
26. Eisen D, Carrozzo M, Bagan Sebastian JV, et al. Number V Oral lichen planus: clinical features and management. Oral Dis 2005;11:338-49.