

Lichen Sclerosus et Atrophicus with Scalp and Oral Mucosa involvement

SCALP VE ORAL MUKOZA TUTULUMU OLAN LİCHEN SCLEROSUS ET ATROPHİCUS

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SUMMARY

Lichen sclerosus et atrophicus is a benign but chronic disease characterized by ivory or white, shiny round macules or papules. In this report a 52-year-old woman who was diagnosed as lichen sclerosus et atrophicus with scalp and oral mucosa involvement is described. As these unusual locations are seen fairly rare, our case is remarkable.

Key Word: Lichen sclerosus et atrophicus

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ÖZET

Lichen sclerosus et atrophicus fildişi veya beyaz, parlak yuvarlak makül veya papüllerle karakterize benign fakat kronik bir hastalıktır. Bu yazıda saçlı deri ve oral mukoza tutulumu ile birlikte lichen sclerosus et atrophicus tanısı konan 52 yaşında bir kadın hasta tarif edilmektedir. Bu alışılmışın dışında lokalizasyonlar oldukça nadir görüldüğü için olgumuz dikkat çekicidir.

Anahtar Kelime: Lichen sclerosus et atrophicus

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Also known as lichen albus, Csillag's disease and white spot disease, lichen sclerosus et atrophicus (LSA) is a benign chronic disease with unknown etiology (1,2). It is considerably more common in women than in men and it appears to be much more frequent in whites, especially in prepubertal girls and pre- and postmenopausal women (3). When it is seen in boys it may be a common and distinctive cause of phimosis (4). The genitals are the usual site of involvement in both sexes. In this case the disorder is termed kraurosis vulvae in women and balanitis xerotica obliterans in men. Skin lesions unrelated to the genital site may appear, particularly on the trunk and neck (5). Some unusual locations including periorbital region, scalp and oral mucosa have been reported (2,5,6). The clinical course of LSA is rather variable with the possibility of spontaneous resolution generally at puberty (2).

CASE REPORT

52-year-old woman, had genital pruritus, alopecia, an extensive eruption as well-delimited white spots

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and grayish white plaques in the mouth which had first appeared 20,10,4 and 2 years ago respectively. Despite her symptomatology, her only complaint was the severe pruritus in all lesions and alopecia. She had 4 gravida, 4 para and a history of ligamentopexy operation (Modified Gillian Suspension Procedure) 23 years ago. She reported to be in menopause for the last 10 years. Dermatological examination revealed generalized eruption of white, shiny, round, small atrophic macules and papules showing a tendency to coalesce, to form plaques on the trunk and scalp with scarring alopecia (Fig. 1, 2). In oral mucosa there existed grayish white plaques on the tongue and a reticulate appearance particularly on the buccal mucosa, buccal sulcus and on the corners of the mouth (Fig.3). In gynecological examination vulvae, vagina, collum, fornix and uterus were found to be atrophic. The dimensions of uterus was measured as 2x3x4 cm. by pelvic ultrasound. A hormonal evaluation revealed high values of testosterone at 158 ng/dl (normal 30 to 120 ng/dl). Results of routine laboratory studies and other hormonal values were either within normal limits or negative. The diagnosis arrived after the histopathologic examination of the biopsy specimens taken from scalp, oral mucosa and trunk lesions, was LSA (Fig.4a, b.).



Fig. 1. Scalp involvement with scarring alopecia In our LSA patient

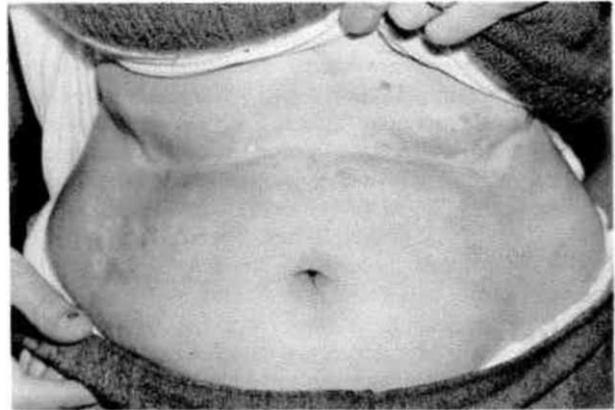


Fig. 2. Trunk lesions which consist of white, shiny, round atrophic macules and papules forming plaques

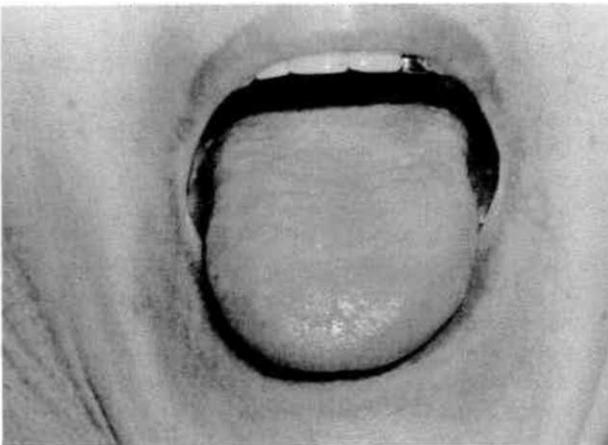


Fig. 3. Grayish white papules and plaques on the tongue in the same patient.

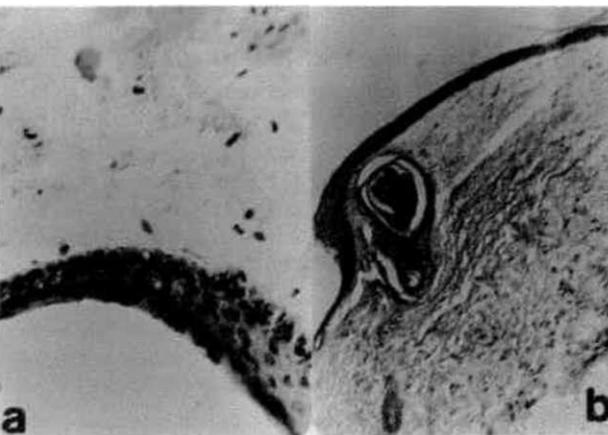


Fig. 4 a. Epithelial atrophy, hydrophic degeneration of the basal layer, edema and the loss of dermal infiltration in the oral mucosa of our patient (H Ex140). b. Epithelial atrophy, follicular plugging and the inflammatory infiltrate in the dermis of the trunk lesion (H Ex140).

Following the diagnosis of the disorder, etretinate (Tigason, La Roche) 0.6 mg/kg/day was given for 3

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months. As a response to the treatment the severe pruritus disappeared and trunk lesions showed a mild improvement.

DISCUSSION

The cause of LSA has not been elucidated, however it is undoubtedly multifactorial. The genetic aspects usually between mother and daughter and the relationship between HLA types of HLA B40, HLA W31 has been demonstrated by various researches (2,7,8). Patients with LSA was shown to have an increased incidence of organ specific antibodies, and these patients and their relatives have a significantly higher incidence of associated autoimmune diseases including pernicious anemia, hypothyroidism, thyrotoxicosis, alopecia areata, diabetes mellitus and vitiligo than normal controls (9,10). We were unable though to show any organ disturbance or a suggestion of genetic transmission in our patient.

Friedrich et al, showed that in patients with untreated vulvar LSA serum levels of dihydrotestosterone, free testosterone and androstenedione were below than normal values for their age and suggested that abnormal 5 α reductase activity might be responsible for this disease (11). High serum testosterone level in our patient did not converge with the results of Friedrich and limited the use of topical testosterone therapy as mentioned in previous reports (1,11,12).

There are also some reports which have linked LSA to infection with the spirochete, *Borrelia burgdorferi* (1,2,12).

The skin lesions of our patient have been found typical of LSA which is characterized by ivory or white shiny round macules or papules with the individual lesions generally becoming aggregated to form plaques. In these lesions atrophy is a hallmark and telangiectasias are occasionally prominent (2,10,12). Some unusual locations such as scalp, oral mucosa, periorbital region, palm and soles have been reported (2,5,6,12,13,14).

Among the unusual presentations of LSA oral mucosa lesions have been described in the cheek, palate, tonsillar pillars, tongue and lips as whitish plaques and these lesions are hardly distinguishable from the lesions of lichen planus (5,12,15,16). Oral LSA is often asymptomatic, although in one instance it was at least partially responsible for mouth pain that limited mouth opening (15). In our patient asymptomatic oral mucosa lesions converges with the infrequent reports of oral LSA clinically. Although it has been suggested that oral lesions appear before skin lesions (5), in our case the patient first presented skin lesions.

Scalp lesions have been reported to be pruritic, hypopigmented atrophic areas which result in scarring alopecia (2, 17), as was the case.

In LSA there is a particular predilection for involvement of the genitalia. When this is the case long-term observations and recurrent histologic examinations should be carried on although the risk of vulvar malignancies is uncertain (2,7). It is generally known that non genital lesions do not undergo malignant change but occasionally regress spontaneously. However whether oral lesions behave like mucosal lesions of the genitalia have not been documented (15).

In cutaneous lesions of LSA the histologic features are hyperkeratosis with follicular plugging (the latter is absent in genital disease), atrophy of the stratum malpighii with hydropic degeneration of basal cells, pronounced edema and homogenization of the collagen in the upper dermis and an inflammatory infiltrate in the middermis (18). The histologic features observed in oral LSA are quite similar to those for the skin with the exception of hyperkeratosis which may not appear in the oral lesions (5). The histopathologic findings of oral mucosa and trunk lesions in our case, were found to be in accordance with these as shown in Fig 4a and 4b.

The choice of treatment is still controversial and the various recommended treatment modalities include vulvectomy, topical or intradermal injection of corticosteroids, topical testosterone, progesterone and estrogen, cryosurgery, laser therapy and tretinate therapy particularly in pre- or postmenopausal women with moderate forms of vulvar dystrophy (1,2,11,12,19).

The disappearance of severe pruritus during tretinate therapy in our patient converges with the results of previous studies investigating the efficacy of this agent in LSA (19,20). Dryness of mucous membranes was the only side effect in a daily dose of 0.6 mg/kg. Since our protocol was only an experimental one, we administered the drug in a relatively low dose and for only 3 months. Even with this conservative approach the patient experienced a dramatic relief in her pruritus complaint.

A review of the literature reveals that the appearance of LSA in the oral mucosa and scalp is rare (2,5). Our case is remarkable for the presence of generally distributed LSA lesions and atrophic genitalia with histopathologically proven oral mucosa and scalp involvement.

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