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Extragenital Lichen Sclerosis et Atrophicus

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Lichen sclerosus et atrophicus (LSA) is an uncommon chronic inflammatory dermatosis involving mainly the anogenital area in postmenopausal woman. Extragenital LSA is rare as an isolated form and is most commonly located on the neck, upper arms and flexor surfaces of the wrist. Only 6% of women and men with extragenital LSA do not have genital lesions.

This rare condition is cited in most of the cases as lichen sclerosis without the atrophic portion and is relevant to findings that lichen sclerosus can result in hypertrophic rather than atrophic epithelium. We report three patients – 2 men and 1 woman with solely extragenital involvement with multiple asymptomatic hyper and hypopigmented atrophic lesions on the trunk and discuss the histopathological and dermatoscopic data to achieve a proper diagnosis.

Key words: Lichen sclerosus et atrophicus, extragenital sites, unusual form, hypertrophic epithelium, histology

Introduction

Lichen sclerosus et atrophicus (LSA) is an acquired, inflammatory skin disease of unknown etiology. It usually involves the anogenital area – vulva, perineum and perianal skin with itching, soreness and sexual dysfunction. It could be also asymptomatic. The primary lesions are flat ivory-colored spots, which conflate into thin atrophic lesions or hyperkeratotic plagues. Women are more affected than men with a reported female: male ratio 10:1 [9]. This statistic might be influenced by male circumcision. Postmenopausal women are at higher risk to develop the disease, followed by girls between the age of 8 and 13 years, men and children. The course of LSA is usually chronic and is associated with an increased risk of squamous cell carcinoma of the affected area. In children the signs and symptoms may improve at puberty, but they need monitoring for disease activity.

LSA is most commonly localized only on the anogenital area with 85% to 98% of the cases. However, in some patients both genital and extragenital involvement is reported up to 15% [11].

Extragenital LSA is rare as an isolated form and the most often localization is on the neck, upper arms and flexor surfaces of the wrist. Only 6% of women and men with extragenital LSA do not have genital lesions.

We report three patients with solely extragenital form of LSA as two of them are men. One of the men has an uncommon presentation, which includes lesions on the scalp.

Case report

The first case is a 71-year old healthy man. He noticed white spots on the skin of his back three years ago. He had neither itching nor discomfort but because of the coalescence of the spots and thickening of the involved area decided to go to a dermatologist. On the physical examination we saw one large whitish-colored irregular plaque and numerous atrophic lesions around it with a diameter 2-3 mm up to 1 cm on the back. Linear and nummular violaceous brownish plaques with different sizes and slightly atrophic center were localized laterally on both sites on the truncus and both thighs symmetrically. A biopsy was taken from the lesion of the back showing hyperkeratosis and partial atrophy of the epidermis, oedema and hyalinization of the upper dermis, dilated vessels and slightly thickened hyalinized collagen in the lower dermis (**Fig. 1**).

The second case is a 52-year old woman. She suffered from mild itching on the back and several thick and dense plague on the back, which appeared six years ago. The treatment is with topical corticosteroids and emollients with no improvement. A month

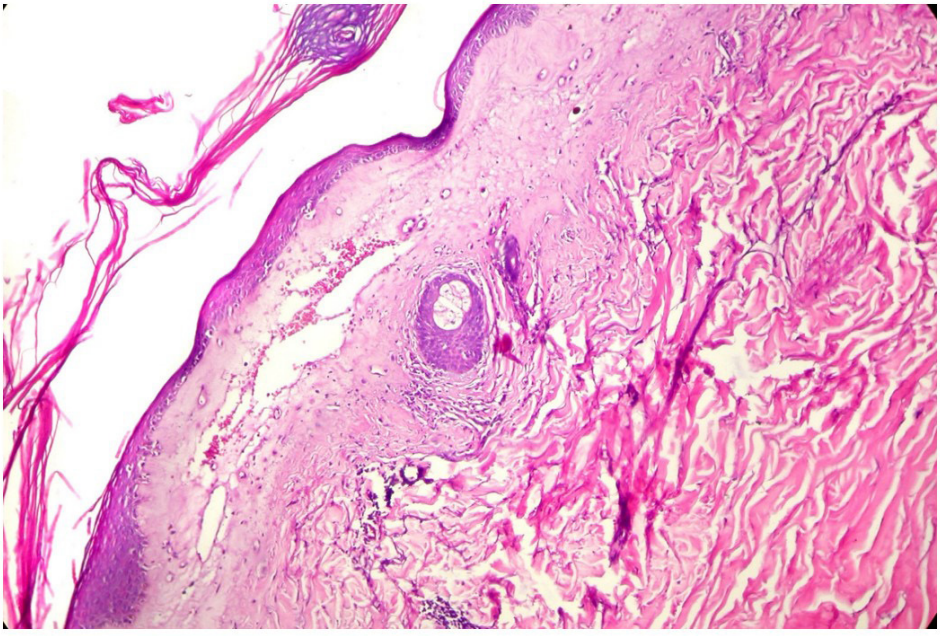

Fig. 1. Hyperkeratosis and partial atrophy of the epidermis, oedema and hyalinization of the upper dermis, dilated vessels and slightly thickened hyalinized collagen in the lower dermis (H&E, × 200).

ago she noticed new plagues on the skin of her left inguinal fold and internal part of the left thigh. The histopathological findings from the new lesion showed hyperkeratosis, flattening of the dermoepidermal junction, oedema in the upper dermis and occasional perivascular infiltrates (**Fig. 2**). Fragmented white yellow structure – expression of atrophy, keratotic plugs and superficial desquamation were seen on dermatoscopy.

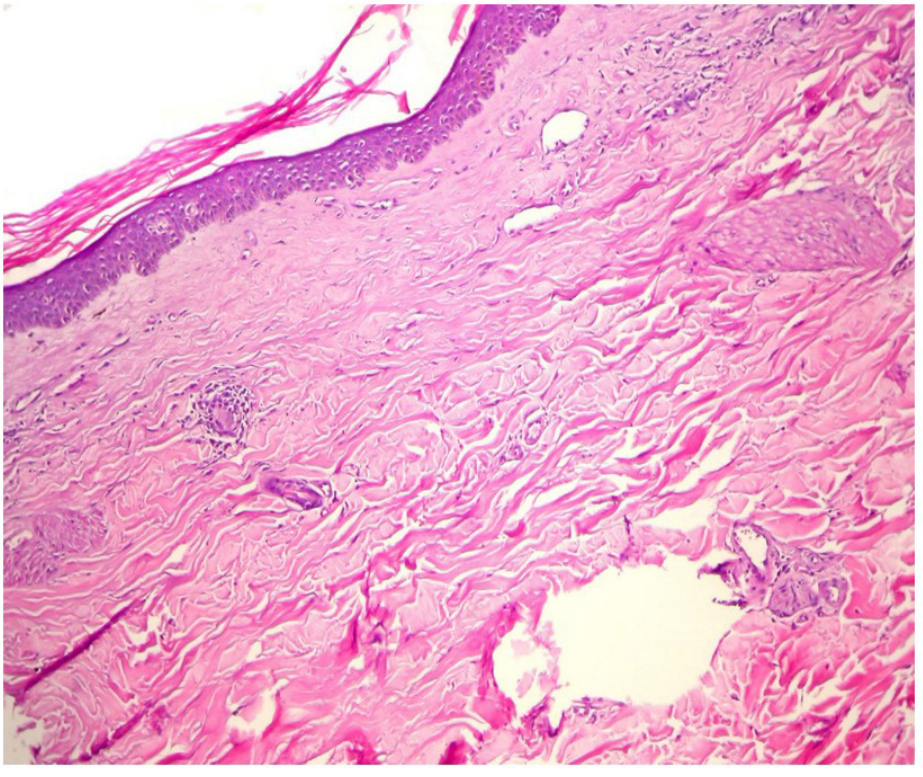

Fig. 2. Hyperkeratosis, flattening of the dermoepidermal junction, oedema in the upper dermis and occasional perivascular infiltrates (H&E, × 200).

Third case is a 36-year old man who suffered from itchy papules on the scalp and body from one year. The lesions on the scalp are perifollicular and polygonal papules, which merged and formed atrophic areas. The histopathology from here showed atrophy of the epidermis, not well defined upper dermis and dermal inflammatory infiltrates. The biopsy from the lesion of the lateral part of the truncus defined mild hyperkeratosis and partial mild acanthosis, oedema, subepidermal slight hyalization and rare perivascular infiltrates (**Fig. 3**).

The patients are diagnosed with extragenital LSA based on the clinical and histopathological findings.

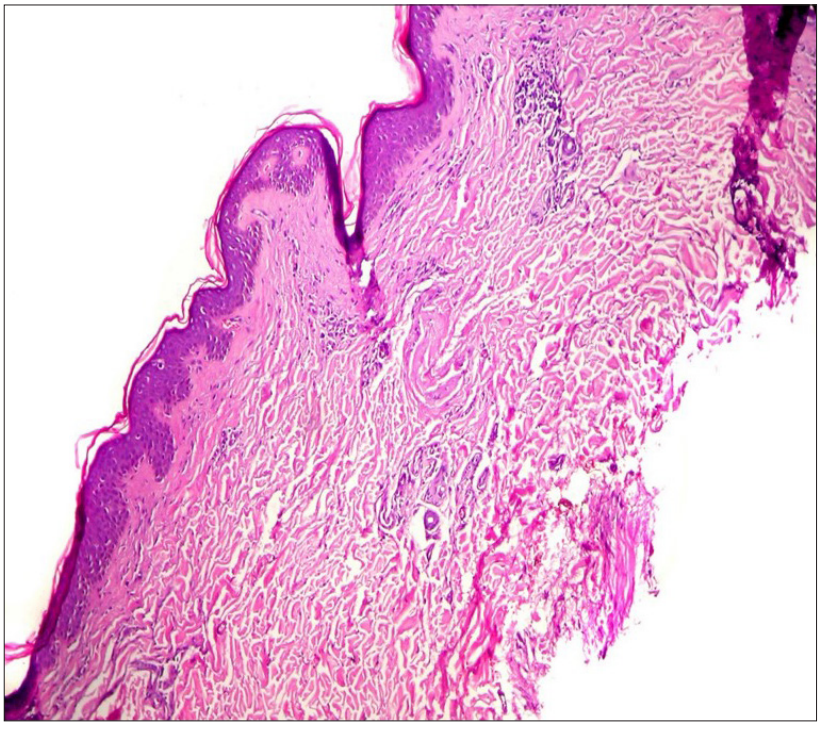

Fig.3. Mild hyperkeratosis and partial mild acanthosis, oedema, subepidermal slight hyalization and rare perivascular infiltrates (H&E, x 200).

Disscusion

LSA was first described by Hallopeau in 1987 [6] and Darier reported the histological changes in 1892 [4]. They considered the disorder to be a type of lichen planus. Others thought that it was related to circumscribed scleroderma. Now, LSA is known as a separate entity because of the distinct clinical sights and pathological changes.

The cause of LSA is unknown, but most studies suggest that it is multifactorial. A genetic predisposition, based on familial clustering was observed [13]. The association of specific human leukocyte antigen (HLA) type - HLA II DQ7 and other autoimmune diseases suggests that LSA is an autoimmune process [5]. Autoimmune diseases are observed in up to 33% of women, particularly thyroid disease, alopecia areata, vitiligo, diabetes mellitus, rheumatoid arthritis, pernicious anemia, lupus erythematosus and morphea [7, 8]. Immunoreactivity to extracellular matrix protein 1 has been demonstrated in up to 74% of cases [10].

LSA may occur in skin, which is already scarred or damaged – so called Koebner phenomenon. From this point of view trauma, injury and scarring are suggested as possible triggers of symptoms in genetically predisposed people. Extragenital LSH involvement commonly develops in preexisting scars and damaged areas [11]. Lesions preferentially occurring on left side of body in most of the reported cases and has been

attributed to stronger cell-mediated immune hypersensitivity in the left side of the body in healthy young subjects. It is speculated that the cellular immune responsiveness might influence the confinement of the Blaschko-linear LSA to the left side of the body [3]. This is in contrast to our first case where lesions were on the back and on the both sides of the body and on the third case – lateral sides of the truncus and scalp. These two patients are man and late onset in the first one, which is not common for this diagnosis. The skin involvement in our second case, which is a woman was on the left side – left inguinal fold and internal part of the left thigh.

Extragenital lesions occur most often on the chest, upper back, and breasts. Cases of linear LSA have been reported along the lines of Blashko [2]. Most often hypopigmented atrophic patches are observed, which are typically asymptomatic. However, progressive disease may cause discomfort and pruritus.

Histopathlogically, these lesions were likely to show concominant changes of lichen simplex chronicus or dermal eosinophils in one series [1]. The striking histological features in LSA is a band of hyalinization of the dermal collagen bellow the epidermis. The hyalinized tissue appears formless, oedematous and may contain sparse cells or has dilated cappilaries. The epidermis shows variable thickening, hyperkeratosis and follicular plugging. Later, the epidermis could become thinner. Perivascular round cell infiltration is also present. In older lesions the lymphocythic infiltrations are more scanty and focal. Extragenital LSA showed decreased expression of the proliferation marker Ki-67 and p53 in comparison to genital LSA, which may explain in part the lack of reported malignant transformation in the extragenital subtype [12].

Conclusions

We present three patients with the rare extragenital form of LSA. The diagnosis is clinically and histopathology confirmed. All of them are followed to detect any atypical or malignant changes and no transformation of the lesions happens. Two of them are men which is not common according to the statistic of mainly women affected. All of them have disseminate lesions and one of them has scalp involvement, which is unusual localization. Histopathological finding varies depending on the evolutionary stage and localization of the investigated lesions. There are scarce perivascular infiltrates unlike those found in LSA with genital lesions. Extragenital LSA in most of the cases has no atrophy and may lead to hypertrophic epithelium.

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