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Extragenital Bullous Lichen Sclerosus et Atrophicus

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Abstract

Observation: Lichen sclerosus et atrophicus is a chronic, inflammatory dermatosis that is characterized by pruritic, white, atrophic plaques. It classically affects the anogenital region of postmenopausal women. Extragenital involvement also may occur with several reported morphologic variants. Extragenital bullous lichen sclerosus et atrophicus is a rare variant, which presents as flaccid bullae. It favors the trunk and proximal aspects of the extremities. We describe a 58-year-old woman with a six month history of extragenital lichen sclerosus et atrophicus, who developed a bullous eruption within a pre-existing patch of lichen sclerosis on the lower back.

Introduction

Lichen sclerosus et atrophicus (LSA) is a rare, chronic inflammatory disease of unknown etiology. It characterized by white porcelain-like sclerotic lesions. LSA mainly affects the genital area in postmenopausal women. It is rarely reported occurring exclusively on the extragenital area [**1**]. Bullous LSA is an un usual form of the disease, with hemorrh agic/nonhemorrhagic bulla in the genital and/or extragenital areas. We introduce a case of extragenital bullous LSA in a women.

Case Report

A 58-year-old woman presented with a six month history of asymptomatic whitish patches on her abdomen and back. There was no lesion in the genital area. She came to our clinic because a blister occurred on her lower back 3 weeks earlier. Her medical and family history was unremarkable. On examination, some sclerotic depigmented or hypopigmented patches were found on her abdomen (**Figure 1**). There were 6x4 cm and 2x3 cm ivory-colored sclerotic plaques with tense bullaes present on the lower back (**Figure 2**). She denied either itching or rubbing the affected areas. Laboratory findings including a complete blood count, urinalysis and liver function tests were normal. Serologic analysis was notable for a positive antinuclear antibody with a low titer. Serology for Borrelia was negative. A biopsy specimen showed mild orthohyperkeratosis, atrophic epidermis featured by flattening of the rete ridges, marked edema in the dermal papillae with bulla formation and homogenization of the collagen in the reticular dermis. Mild perivascular infiltration of lymphocytes was seen (**Figure 3**). The patient was diagnosed with

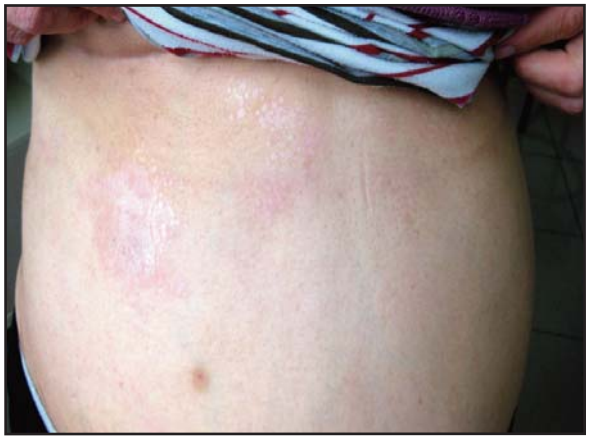

Figure 1. The patient presented with sclerotic patches on her abdomen

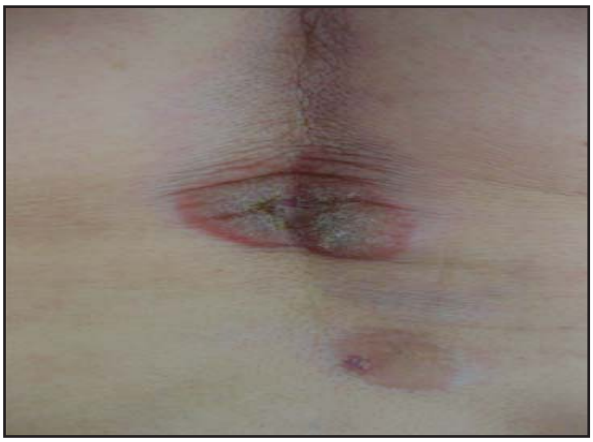

Figure 2. Tense blisters developed in the sclerotic patches on her lower back

extragenital bullous LSA. Treatment with betamethasone 17 valerate ointment 1% was performed. A bullous lesion improved with topical treatment, but the patient did not come to other appointments.

Discussion

Lichen sclerosus et atrophicus is a chronic, inflammatory disease that is classically characterized by pruritic, white, sclerotic, atrophic plaques. It most commonly affects the anogenital region. It is an uncommon disease with an estimated prevalence of 0.1 to 0.3% [**2**]. Both sexes are affected; however there is a bimodal distribution that occurrs prepubescently and postmenopausally [**3**]. 15 to 20% of all patients with LSA have extragenital involvement [**1**]. Extragenital LSA is generally asymptomatic and commonly involves the trunk and proximal extremities [**4**]. Bullous LSA, which is characterized by flaccid/tense bullae, may localized to both genital and extragenital. In this case, there is no pruritus on the lesions.

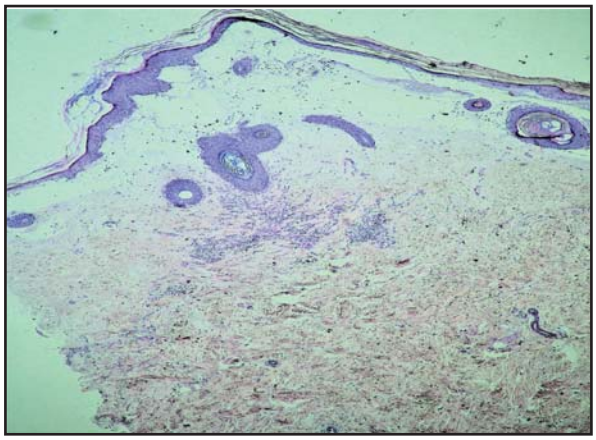

Figure 3. Skin biopsy of bullous lesion on the woman's lower back revealed mild orthohyperkeratosis, atrophic epidermis featured by flattening of the rete ridges, marked edema in the dermal papillae with bullae formation and homogenization of the collagen in the reticular dermis, mild perivascular infiltration of lymphocytes (HE, x40).

The etiology of LSA is stil unknown [**5**]. Several factors including genetic susceptibility, trauma, chronic infections and low levels of androgens have been implicated as pathogenic factors; however currently favored is an autoimmune processes. There is an association with autoimmune diseases including type I diabetes, vitiligo, thyroid disorders and alopecia areata [**6**]. In our case, there is only antinuclear antibody positivity with low titer. Several studies have shown an increase in circulating antibodies against extracellular matrix 1 protein and bullous pemphigoid antigens 180 and 230 [**7, 8**]. Theoretically, an autoimmune process could result in bullae formation in the bullous variant of LSA.

Characteristic histopathologic features of LSA include epidermal atrophy, follicular plugs and a band of edematous, homogenized collagen in the papillary dermis with varying degrees of lymphocytic infiltrate [**9**]. The histopathologic formation of subepidermal bullae has been attributed to two mechanisms. One is papillary dermal edema that leads to disruption of the supporting collagen fibers, and the second is a vacuolar interface dermatitis that causes basal layer degeneration and basement-membrane zone instability [**10**].

The treatment of extragenital bullous LSA is similar to that of genital LSA. However, extragenital LSA often is less responsive, so may present a therapeutic challenge [**11**]. The treatment of choices for extragenital LSA are the application of super-potent topical glucocorticoids with/ without topical calcineurin inhibitors, intralesional glucocorticoids and sistemik glucocorticoids for widespread or refractory cases [**11**]. For cases of extragenital LSA refractory to glucocorticoids, methotrexate or phototherapy (especially UVA1) may be considered [**12**].

In conclusion, we would like to emphasize the possibility of bullous formation in patients with extragenital LSA. By acknowledging this phenomenon, the clinician can provide better management to the patient.

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