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CASE REPORT

Extragenital bullous lichen sclerosus: A case report and literature review

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ABSTRACT

Lichen sclerosus (LS) is a chronic dermatosis characterized by atrophic white papules or plaques, most commonly occurring on the anogenital skin. Blisters have been rarely described developing in the background of extragenital LS. A 74-year-old woman with a 4-year history of sclerotic patches on the trunk showed a flaccid bulla on the lower back for 3 months. The histopathological finding of the skin biopsy was consistent with the diagnosis of bullous LS. In this paper, we present this uncommon case, review the literature on extragenital bullous LS, discuss the pathogenesis, and provide some treatment options for the case.

Introduction

Lichen sclerosus (LS) is a chronic inflammatory disorder, with white porcelain-like sclerotic lesions. The etiology of this chronic process is unknown. It mainly affects the genital area in postmenopausal women, but is rarely reported occurring exclusively on the extragenital area. Bullous LS is an unusual form of the disease, with hemorrhagic bulla in the genital and/or extragenital areas.[1,2] The clinical features and long-term outcomes of patients with extragenital bullous LS have been poorly described. We introduce a case with bullous LS exclusively found in the extragenital area, review similar cases reported in the literature, and discuss the clinical features, pathophysiology, and treatment options in an effort to raise awareness of this rare clinical presentation.

Case report

A 74-year-old woman presented with a 4-year history of asymptomatic whitish patches on her abdomen and back. There was no other lesion in the genital area. She came to our clinic because a blister occurred on her back 3 months earlier and did not subside. Her medical and family history was unremarkable. On examination, some sclerotic depigmented or hypopigmented patches were found on her abdomen (Figure 1A). There was a large 6 × 4 cm, ivory-colored sclerotic plaque with a tense bulla present on the lower back (Figure 1B and C). 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 titer of 1:40. Serology for *Borrelia* was negative. A biopsy specimen showed marked edema in the dermal papillae with bulla formation, atrophic epidermis featured by flattening of the rete ridges, mild orthohyperkeratosis, and homogenization of the collagen in the reticular dermis. Mild perivascular infiltration of lymphocytes was focally seen (Figure 2). Direct immunofluorescence was negative.

The patient was diagnosed with extragenital bullous LS. Treatment with clobetasol dipropionate ointment 0.05% (GlaxoSmith Kline, Barnard castle, UK) and oral prednisolone (China Chemical & Pharmaceutical Co. Ltd., Hsinchu, Taiwan) 10 mg twice daily for 4 weeks was not effective. Then, intralesional injection with triamcinolone (Yung Shin Pharmaceutical Co. Ltd., Taichung, Taiwan) 10 mg/mL was performed every 2 weeks for three sessions. At the 5-month return visit, the blister resolved and an atrophic whitish plaque remained. The plaque has remained stable for 1 year and no other new lesions have appeared.

Conflicts of interest: The authors declare that they have no financial or non-financial conflicts of interest related to the subject matter or materials discussed in this article.

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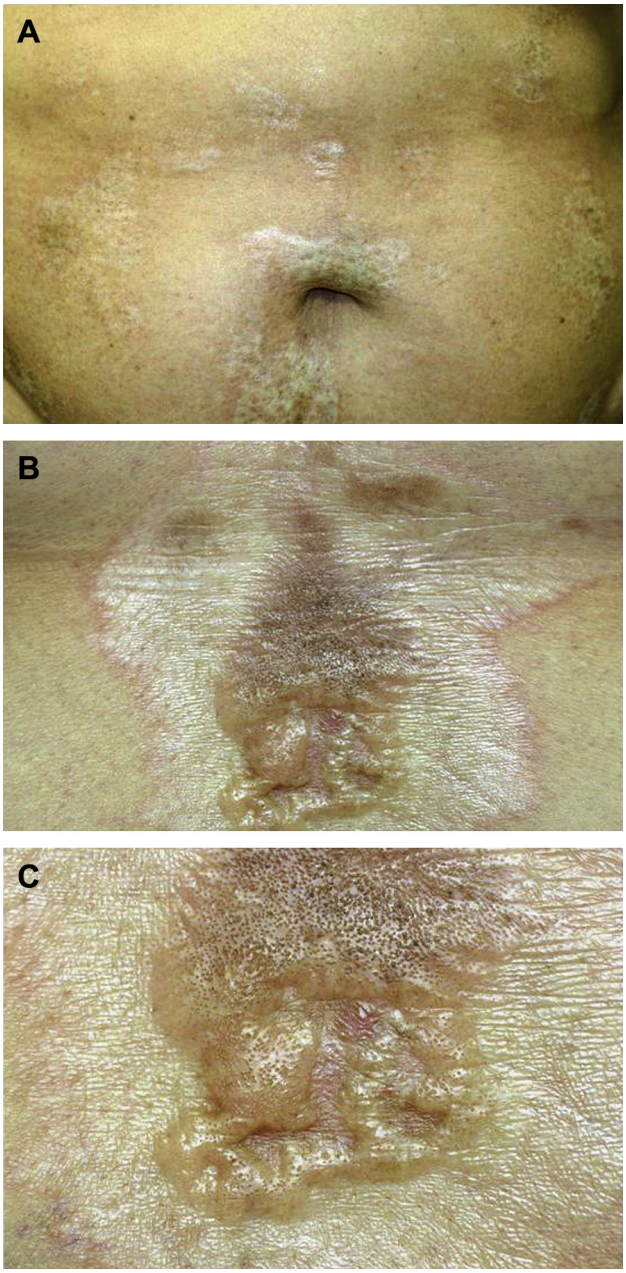

Figure 1 (A) The patient initially presented with sclerotic patches on her abdomen. (B) A tense blister developed in the lesion on her back. (C) A closer view of the blister demonstrated an irregular configuration.

Discussion

Cases of bullous LS have been uncommonly reported in English literature, occurring in genital and/or nongenital forms. After reviewing the cases reported in published studies, bullous LS was found exclusively at the extragenital site in only 13 cases (Table 1).[1–13] Fourteen cases with extragenital bullous LS including ours were investigated. Among them, seven were females, four were males, and three were not available. The age distribution was wide, from 14 years to 84 years, but most of the patients were elderly (average 60 years). Five of the cases were with generalized distribution and the rest were locally involved. The most common site for the localized lesions was the back, followed by abdomen, palms, wrists, buttocks, the pretibial area, and the posterior neck.

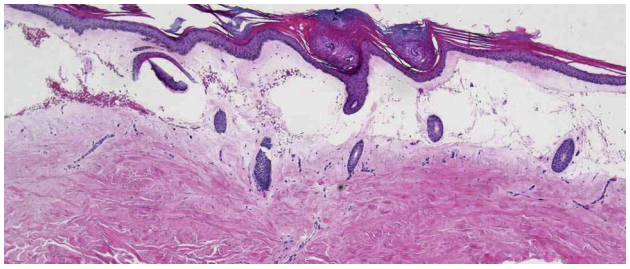

Figure 2 Skin biopsy of the lesion on the woman's back revealed hyperkeratosis, atrophy of the stratum malpighii, pronounced edema with bulla formation in the papillary dermis, sclerotic dermis, and mild inflammatory cell infiltrate. (Hematoxylin and eosin, original magnification 4×.)

The extragenital LS was asymptomatic, often not itchy[2]; patients usually paid attention to the disease when blister(s) formed.

The underlying cause of LS is obscure, and many factors may be involved, such as genetic susceptibility and a link with autoimmune mechanisms.[2,10] An infectious etiology has been suggested in bullous LS, with some reports proposing a connection with *Borrelia* infection and hepatitis C.[8,14] However, we could not find any evidence of this in our patient. Furthermore, although our case had a positive antinuclear antibody with a titer of 1:40, titers that are less than 1:160 are usually not considered as clinically significant or associated with autoimmune disorders.

Another postulation regarding the bullous formation of LS is thought to result from lymphoid edema or trauma. Marked edema with dilated vessels and hemorrhagic areas induces blister formation. Minor trauma with mechanical stress easily causes bullae

Table 1 List of the 14 published cases of bullous lichen sclerosus (LS) exclusively in the extragenital area.

Case	First author (year of publication)	Age at diagnosis (y)	Sex	Location of lesion	Treatment
1	Gottschalk HR (1947)[3]	NA	NA	Extensive	NA
2	Di Silverio A (1975)[4]	NA	NA	Extensive	ACTH
3	Tudino ME (1984)[5]	54	F	Palms and wrists	NA
4	Klein LE (1984)[6]	NA	NA	Pretibial	Tangential excision
5	Marren P (1992)[1]	82	F	Generalized and scalp	NA
6	Wakelin SH (1994)[7]	55	F	Generalized	Hydroxychloroquine
7	Boulinguez S (1997)[8]	59	F	Buttocks	NA
8	Hallel-Halevy D (1998)[9]	47	M	Lower back	Topical corticosteroids
9	Gomez-Calcerrada MR (1999)[10]	61	F	Lumbar, abdominal area, scalp	Antiseptics, analgesics, topical corticosteroids, and oral antihistamines
10	Madan V (2008)[11]	84	M	Generalized and scalp	Topical corticosteroids and doxycycline
11	Ballester I (2009)[2]	80	M	Back	Topical clobetasol propionate cream
12	Kimura A (2011)[12]	44	M	Upper back	Topical clobetasol propionate cream
13	Shiver M (2012)[13]	14	F	Posterior neck	Topical clobetasol propionate cream, followed by pimecrolimus cream
14	Present case (2013)	74	F	Back	Intralesional injection with triamcinolone 10 mg/mL

ACTH = adrenocorticotropic hormone; F = female; M = male; NA = not available.

because of epidermal atrophy in extragenital LS.[12] This may explain why our case had many sclerotic patches on the trunk, but only showed a blister on the back, where the possibility of being rubbed by tight clothes was high.

Interestingly, there were some published cases on bullous morphea, which can be confused clinically with LS because both diseases may cause bullae in the sclerodermatous plaques.[15] Furthermore, patients with long-standing LS that subsequently developed new onset of bullous pemphigoid and pemphigus vulgaris at affected sites were reported in the literature.[16] Therefore, a skin biopsy is needed to accurately differentiate the disease.

An increased risk of squamous cell carcinoma may exist in genital LS, but no increased risk of malignant transformation has been found in extragenital LS.[2] There is no definitive treatment for LS. The use of potent corticosteroid has proved to be an effective therapeutic option. However, extragenital lesions are not as responsive as genital diseases to topical corticosteroid therapy. Topical clobetasol propionate cream resolves the problems of blistering but has little effect on the underlying atrophic plaque.[13] Our patient had a poor response to topical and oral corticosteroids but had better resolution of the blister after intralesional corticosteroid injection.

In conclusion, we would like to emphasize the possibility of bullous formation in patients with chronic extragenital LS. The fair response to local triamcinolone injection provides an alternative choice of therapy. By acknowledging this phenomenon, the clinician can provide better management to the patient.

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