Extragential bullous and haemorrhagic lichen sclerosus: Case report

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ABSTRACT

Lichen sclerosus is a chronic, inflammatory dermatitis of unknown etiology affecting any site but mainly the anogenital area. It most commonly occurs in women, but also in men and children. Extragential forms of the disease and hemorrhagic transformation or blister formation are really uncommon. Three cases of extragential bullous and haemorrhagic lichen sclerosus receiving different therapeutic choices were presented respectively.

Keywords: Blister, hemorrhage, lichen sclerosus.

INTRODUCTION

Lichen sclerosus (LS) was described for the first time in 1887. It is a chronically relapsing disease with a potential for atrophy, destructive scarring, functional impairment and malignant evolution. It mainly affects the anogenital area and extragenital forms of the disease are uncommon, especially hemorrhagic transformation or blister formation. It is hardly cured, but can be controlled by adequate treatment. With early treatment, long-term sequelae such as destruction of anatomic structures and progression to squamous cell carcinoma (SCC) may be prevented (Fistarol and Itin, 2013). Here, we reported three patients of extragential bullous and haemorrhagic LS receiving different therapeutic choices respectively.

CASE REPORT

Case 1

A 58-year-old man presented with minimally pruritic white colored lesion on the left upper back for almost ten years. Though several kinds of ointments were prescribed to him for usage but this was to no avail as the lesion gradually increased in size to 2.5 cm × 2.2 cm and there was associated mild stinging sensation and minimal watery discharge from the surface since half a year ago. There was no history suggestive of involvement of oral or genital mucosa. Dermatological examination revealed atrophic plaque with superficial wrinkled appearance and follicular plugging on the surface (Figure 1A).

Under the atrophic epidermis, there was a collection of fluid which was confirmed after puncturing the lesion with needle. Although surgery is not the first-line therapeutic choice, the patient insisted to receive a surgery to excise the lesion completely since this issue has been on for almost ten years. Histological examination revealed the presence of an atrophic epidermis with hydropic degeneration of basal cell layer forming a subepidermal split with pronounced dermal edema and homogenization of collagen in upper dermis (Figure 1B) and relatively dense lymphocytic perianillageal infiltrate in the mid dermis. The final histopathologic findings allowed us to establish the diagnosis of bullous variant of LS. After surgery he applied topical tacrolimus immediately and remained asymptomatic with no signs of recurrence at the 2-years follow up.

Case 2

A 38-year-old woman consulted the dermatology clinic for a pruritic lesion on her lower back that had been present for six years. The lesion, which had always been whitish, became hemorrhagic and more raised in the previous 2
Figure 1: (A) Atrophic plaque with blister on the left upper back. (B) Keratotic follicular plug and subepidermal blister formation (hematoxylin-eosin stain, original magnification × 100). (C) Atrophic plaque with a hemorrhagic blister. (D) Subepidermal blister with marked edema of the papillary dermis and extensive areas of hemorrhage (hematoxylin-eosin stain, original magnification × 100). (E) White plaque with a hemorrhagic blister. (F) Keratotic follicular plug and subepidermal blister with hemorrhage (hematoxylin-eosin stain, original magnification × 100).

months. The patient reported that there had been no previous trauma. On physical examination the patient had a white plaque with a hemorrhagic blister and local erosion on the surface (Figure 1C). There were no other lesions in the genital area or in other sites. A biopsy was done and histological examination revealed a subepidermal blister with marked edema of the papillary dermis, homogenization of collagen, atrophy of the epidermis, and extensive areas of hemorrhage (Figure 1D). The diagnosis was extragential haemorrhagic LS.

Concerning the long-term safety of corticosteroid, topical corticosteroid and tacrolimus 0.1% ointment were both administered on this patient at the same time. The symptom relieved her a lot quickly and she was still in the follow-up.

Case 3

A 29-year-old man was presented with minimally pruritic
white plaque on the left neck for 2 years. The central region of the lesion raised and became hemorrhagic spontaneously 1 month ago. Physical examination revealed a white plaque with a hemorrhagic blister and local erosion on the central part of the surface (Figure 1E). Histological examination revealed hyperkeratosis, epidermal atrophy with flattening of the rete ridges, vacuolar interface changes, subepidermal blister with hemorrhage and homogenization of collagen (Figure 1F).

Based on the clinical manifestation and histological features, the diagnosis was extragential haemorrhagic LS. The hemorrhagic part of the lesion had an excellent respond to topical corticosteroid. Considering wider involved area, tacrolimus 0.1% ointment was added to reduce the adverse effects of long-term with corticosteroid subsequently.

**DISCUSSION**

LS mainly affect the genital region of pre-pubertal and post-menopausal females (Heymann, 2007). Extragential involvement is associated with classic genital LS in 15 to 20% of cases and only 2.5% cases show exclusive involvement of extragenital site (Ballester et al., 2009). The most common locations for extragenital LS are the buttocks, thighs, breasts, submammary area, neck, back and chest, shoulders, axillae and wrists (Fistarol and Itin, 2013).

The classic form of LS is characterized by pruritic flat-topped white papules that coalesce to form white plaques with epidermal atrophy and a ‘cigarette-paper’ appearance. Occasionally, hemorrhagic transformation or blister formation occurs due to basal cell degeneration resulting in dermal-epidermal separation and marked edema in upper dermis. Extragential LS is much less symptomatic than genital LS. This may result from the occlusion and maceration of genital area, which play a role in causing pruritus, soreness and pain (Fistarol and Itin, 2013).

Histologically, well-developed lesion of LS is characterized by orthokeratosis, keratotic follicular plugs, atrophy of stratum corneum with flattening or loss of rete ridges. The basal layer shows liquefactive degeneration. Beneath the epidermis there is a broad zone of pronounced edema and homogenization of the collagen. Within this zone collagen fibers are swollen, homogenous and contain very few nuclei. The blood and lymphatic vessels are dilated and there may be areas of hemorrhage. Edema in the upper part of the dermis may be enough to result in clinically apparent vesiculation (Khatu and Vasani, 2013) and the fragility of the flattened epidermal-dermal interface may give rise to bullous and hemorrhagic lesions in extragenital LS (Fistarol and Itin, 2013) just like in the three reported cases.

Topical corticosteroids remain the first-line treatment for LS, but there are no published randomized controlled trials or case series reporting the use of topical corticosteroids for extragenital LS. Topical calcineurin inhibitors (TCIs) are an alternative treatment option and should not be applied to malignant or to potentially malignant skin lesions and that treatment should be short term, although only three cases were observed in genital LS treated with TCIs in the worldwide reports collated by the US FDA (Fistarol and Itin, 2013). The safety evaluation of TCIs used for extragenital LS is still lacking. The patient has to be informed about this circumstance prior to the prescription. From our treatment experience, TCIs are effective and safe in extragenital LS. Surgery should be limited to patients with associated malignancy or to patients who need correction of functionally restricting, scarring processes and not be performed until the disease activity has ceased. In the first case, the patient received the complete excision owing to his insistence and the small size and inactivity of the lesion.

**REFERENCES**


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