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Hemorrhagic Bullous Lichen Sclerosus: A Case Report

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Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
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Conflict of interest: None declared

Patient: Female, 63-year-old
Final Diagnosis: Lichen sclerosus
Symptoms: Itching skin
Medication: —
Clinical Procedure: Biopsy
Specialty: Dermatology

Objective: Rare disease

Background: Lichen sclerosus (LS) is a chronic autoimmune dermatosis characterized by white, sclerotic, atrophic plaques. Classic LS commonly occurs in the anogenital region, while extragenital lichen sclerosis typically occurs on the trunk and proximal extremities. Bullous lichen sclerosus is a rare variant that can occur in both genital and extragenital LS. Flaccid bullae can form, which may become hemorrhagic and produce a characteristic appearance clinically.

Case Report: In this report, we describe the case of a 63-year-old female patient who presented for evaluation of a rapidly growing, erythematous, scaly growth on her back/shoulder that was biopsied and found to be hemorrhagic bullous LS. We will discuss the clinical and histologic features of this case as well as treatment of bullous LS, which in this case was a topical high-potency corticosteroid.

Conclusions: Bullous LS has been poorly studied due to the rarity of the condition, with limited investigation of the clinical and histopathologic characteristics of bullous LS and the available treatment options. Although rare, extragenital LS with hemorrhagic bullous features is an important variant of LS that should be considered to ensure appropriate diagnosis and treatment.

MeSH Keywords: Administration, Topical • Blister • Lichen Sclerosus et Atrophicus • Skin Abnormalities

Full-text PDF: https://www.amjcaserep.com/abstract/index/idArt/919353

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Background

Lichen sclerosus (LS) is a chronic autoimmune disorder of unknown etiology characterized by white, sclerotic, atrophic plaques. Classic LS commonly occurs in the anogenital region and can be asymptomatic or present with dryness, pruritis, and pain. Extragenital lichen sclerosis may also occur, commonly on the back, chest, abdomen, and proximal extremities. Extragenital lesions may present as bullous lichen sclerosus, a rare variant of LS. Bullous LS has been poorly studied due to the rarity of the condition, with limited investigation of the clinical and histopathologic characteristics of bullous LS and the available treatment options. Apart from a few case reports, there is limited published information regarding this entity and its treatment. We will discuss the clinical and histologic features of bullous LS in this case as well as treatment of bullous LS.

Case Report

We describe the case of a 63-year-old female patient who presented for evaluation of a rapidly growing, erythematous, minimally symptomatic, scaly growth on her back/shoulder that erupted 6 months ago and had rapidly grown since. The patient noted occasional pruritis over the site of the lesion but was otherwise asymptomatic. The patient reported no previous trauma to the site and no personal or family history of skin cancer. Her medical history was significant for hypothyroidism that was well-controlled with levothyroxine.

On physical exam, the patient had an erythematous, scaly, well-demarcated, 4.3×5.1 cm plaque on the right upper back and shoulder (Figure 1). No other lesions were noted on exam. Due to concern for malignancy, a shave biopsy of this thin plaque was performed. The biopsy revealed compact hyperkeratosis, effacement of underlying rete ridge architecture, epidermal atrophy, and numerous extravasated erythrocytes with superficial perivascular lymphocytic inflammation. A subepidermal cleft was noted in the central portion of the lesion with partial homogenization of the underlying dermal collagen (Figure 2). Hemorrhagic bullous lichen sclerosus was diagnosed based on clinical and histological presentation.

Treatment was initiated with betamethasone 0.05% ointment twice daily for 8 weeks. At the follow-up visit, the lesion had markedly improved, with partial resolution of the hemorrhagic blister (Figure 3). Treatment was continued for an additional 4 months with betamethasone twice daily, with application for 3 weeks followed by a 1-week break. Follow-up at 6 months showed almost complete resolution, with an atrophic white plaque replacing the hemorrhagic blister (Figure 4). At final follow-up at 15 months, the lesion was completely resolved, with only an atrophic white patch remaining with re-pigmentation at the edges of the lesion (Figure 5).

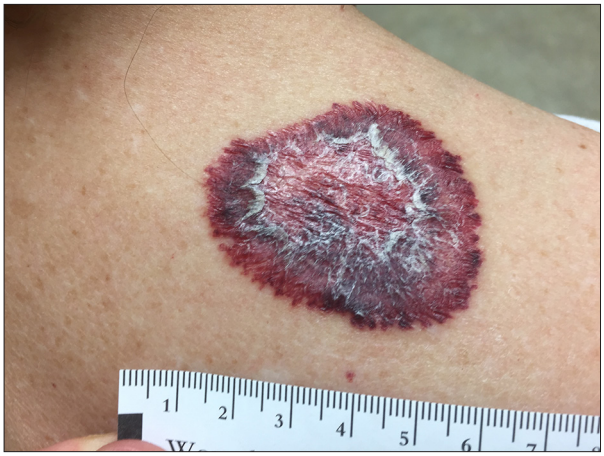

Figure 1. Hemorrhagic bullous LS on initial presentation.

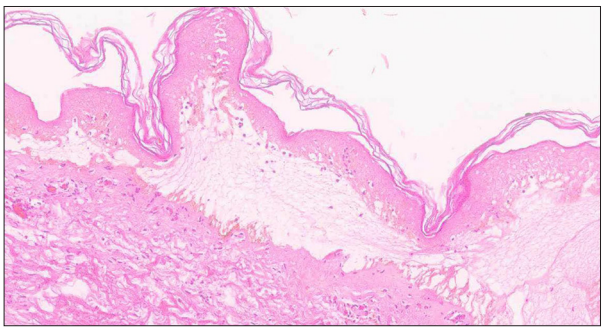

Figure 2. Biopsy of lesion taken on initial presentation demonstrating lichenoid dermatitis with subepidermal cleft and erythrocyte extravasation highly suggestive of hemorrhagic bullous LS (100× magnification).

Discussion

Lichen sclerosus was first clinically described in 1887 by Hallopeau, with the histopathological features characterized shortly thereafter by Darier [1]. Although numerous studies and guidelines have been published regarding anogenital LS, extragenital LS remains poorly studied, with few established guidelines for treatment. Although LS can occur at any age in both sexes, it predominantly occurs in postmenopausal females in the fifth and sixth decades of life. Most LS cases occur in the anogenital region (85–98%), while the remaining 15–20% occur in extragenital regions, primarily the back, chest, abdomen, and proximal extremities [2]. Unlike anogenital LS, extragenital LS does not have malignant potential. LS lesions may initially begin as polygonal, white, elevated papules that eventually progress to plaques that become atrophic and wrinkled [2]. While classic LS often presents with pain and severe pruritis, extragenital LS is often minimally symptomatic or asymptomatic.

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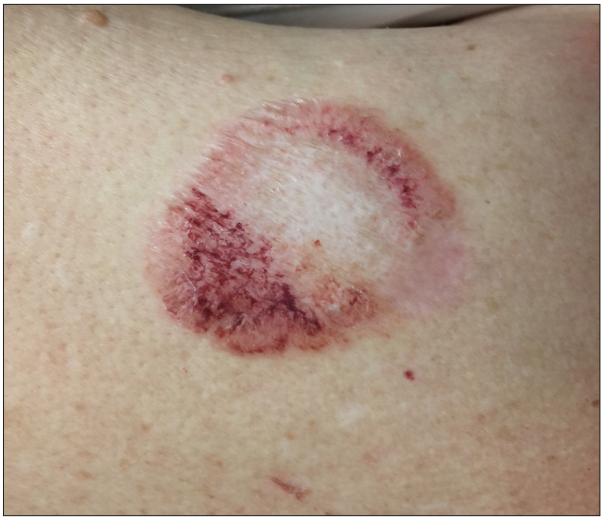

Figure 3. Hemorrhagic bullous LS after 8 weeks of treatment with topical betamethasone.

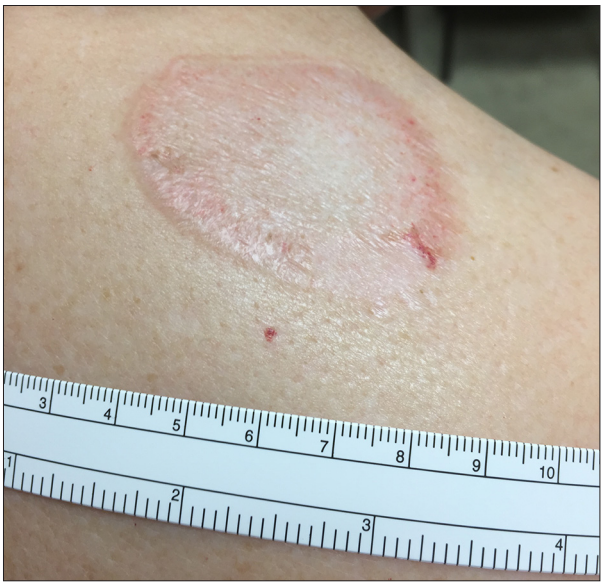

Figure 4. Hemorrhagic bullous LS after 6 months of treatment with topical betamethasone.

The underlying etiology and pathogenesis of LS has yet to be elucidated, although multiple mechanisms have been implicated. The prevailing theory based on current research is that autoimmunity in predisposed individuals leads to the development of both anogenital and extragenital LS [3]. A study of 350 women with LS found that 21.5% had at least 1 autoimmune disorder at the time of diagnosis, with the most prevalent disorder being autoimmune thyroiditis [4]. In the same study, 42% of patients with LS had autoimmune antibodies.

Specific antibodies that have emerged in the pathogenesis of LS are IgG1 and IgG2 autoantibodies against extracellular matrix protein 1 (ECM1), a membrane glycoprotein found in the basement membrane. These autoantibodies have been found in 75% of patient with LS. ECM1 interacts with perlecan, a major proteoglycan in the basement membrane. This interaction shows the important role of ECM1 in the binding of dermal collagens and elastic fibers [5]. These autoantibodies are thought to disrupt these interactions, resulting in the dermatopathological findings of LS. The flaccid bullae that develop in bullous LS may be due to an exaggerated response of this mechanism, with the disruption of the basal layer leading to instability of the basement membrane. Another proposed mechanism for the formation of bullae is the development of edema of the papillary dermis secondary to dermal lymphocytic infiltration and inflammation, which interferes with supporting collagen fibers and flattens the rete ridges [6]. This combination of basement membrane disruption and inflammation results in blister formation with minimal trauma.

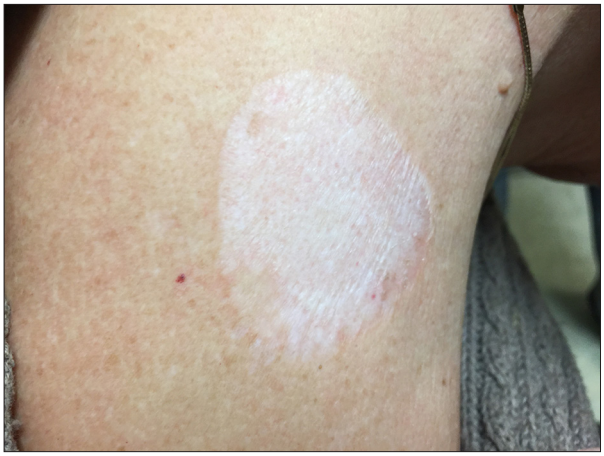

Figure 5. Hemorrhagic bullous LS after 15 months of treatment with topical betamethasone.

Due to the relative rarity of extragenital LS, there are no randomized, controlled studies for treatment of LS, and treatment recommendations are mainly based upon prior case reports. First-line treatment consists of topical corticosteroids, commonly clobetasol propionate 0.05%, a super-potent steroid, once or twice daily for 2 weeks [6]. Topical calcineurin inhibitors may be used for both initial treatment and maintenance therapy following initial treatment. A double-blind study comparing clobetasol and pimecrolimus in patients with vulvar LS found that both effectively decreased inflammation, but clobetasol was more effective than pimecrolimus [7].

Extragenital LS is typically less responsive to topical steroids and calcineurin inhibitors compared to anogenital LS, and additional therapies are often required. Phototherapy has been found to be effective in certain patients, with case reports specifically showing clinical improvement with narrowband UVB, UVA1, and psoralen-UVA (PUVA) [8].

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Conclusions

This case was a unique example of an almost completely hemorrhagic bullous lichen sclerosus lesion treated effectively with a high-potency topical corticosteroid. Although relatively rare, extragenital LS is an important variant of LS that should be considered to ensure appropriate diagnosis and treatment.

Conflict of interest

None.

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