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Bullous hemorrhagic lichen sclerosus of the breast: a report of two cases and review of the literature

Michael P Ryan[1] BS, Seena Monjazeb[2] MD, Lindy S Ross[2] MD, Kathleen T Kroger[2] MD

Affiliations: [1]School of Medicine, University of Texas Medical Branch, Galveston, Texas, USA, [2]Department of Dermatology, University of Texas Medical Branch, Galveston, Texas, USA

Corresponding Author: Kathleen T. Kroger MD, University of Texas Medical Branch, 301 University Boulevard, 5.120 McCullough Building, Galveston, TX 77555-0583, Tel: 409-772-5047, Fax: 409-747-0333, Email: kakroger@utmb.edu

Abstract

Lichen sclerosus (LS) is a chronic dermatologic condition characterized by atrophic porcelain-appearing plaques that can cause intense discomfort and eventually lead to destruction of local architecture. Lichen sclerosus most commonly arises in the anogenital area, but up to 20% of cases can present on extragenital skin. Rarely, LS can also present with hemorrhage and bullae; the mechanism by which this occurs is unknown. We report two cases of bullous hemorrhagic LS of the breast arising in 66-year-old and 77-year-old women. Bullous hemorrhagic LS of the breast is exceedingly rare, with few reported cases, and presents a diagnostic challenge to clinicians.

Keywords: lichen sclerosus, bullous, hemorrhagic, breast, extragenital, treatment, variant, dermatology

Introduction

Lichen sclerosus (LS) is a chronic dermatologic condition characterized by atrophic, white-to-porcelain appearing plaques that can cause intense pruritis, discomfort, and soreness [1-3]. These plaques are most commonly located in the anogenital region in up to 85-98% of cases and affect women 6-10 times more frequently than men [2, 3]. There is a bimodal age distribution in women with LS commonly affecting prepubescent children and postmenopausal women. The exact epidemiology of LS is unknown, but studies have estimated that one in 300-1000 referrals to dermatology clinics are for evaluation or management of LS [2]. The pathophysiology of the disease is also incompletely understood, but it is believed to have underlying genetic and autoimmune components [4]. Early in the disease, LS has a short inflammatory phase that eventually leads to atrophy and scarring. This can lead to dysuria, dyspareunia, pain with defecation, and loss of architecture depending on the location and extent of involvement. Lichen sclerosus induced by trauma to the area (Koebner phenomenon) has also been noted [2, 3]. Patients with anogenital LS are at increased risk for malignancy. In women the risk of developing squamous cell carcinoma (SCC) of the vulva is around 4-5% and in men there is likely a link between LS and penile SCC. However, the exact frequency is difficult to establish [2]. Extragenital LS is less common, presenting in 15-20% of LS cases, but does not appear to have the same association with development of malignancy [2, 3].

Case Synopsis

Case 1

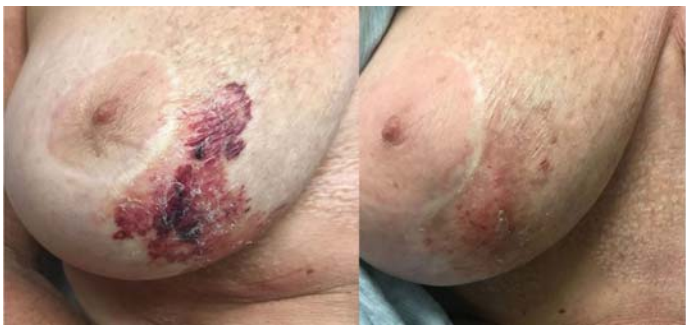

Figure 1. *Bullous hemorrhagic lichen sclerosus.* ***A)*** *Scaly violaceous plaque located on the right inferomedial aspect of the breast at time of presentation.* ***B)*** *Residual and resolving areas of erythema two months after starting twice daily clobetasol 0.05% ointment.*

A 66-year-old woman with a several-year history of biopsy-proven LS of her vulva, abdomen, and breast presented to the dermatology clinic as a new patient for evaluation of a new hemorrhagic lesion on her right breast (**Figure 1A)**. The plaque was pruritic and would often bleed. The patient had no history of breast cancer but did have bilateral breast reduction surgery years prior. Physical examination revealed a violaceous scaly plaque on her inferomedial right breast. A biopsy of the plaque was performed in-office and was consistent with the diagnosis of LS (**Figure 2**). Clobetasol 0.05% ointment was prescribed for twice-daily use and marked improvement was seen at her two-month follow-up appointment (**Figure 1B)**.

Case 2

A 77-year-old woman with an 11-month history of biopsy-proven LS presented to the dermatology clinic for evaluation of a tender bleeding "sore" on her left breast that had been present for two months. She had previously been diagnosed with biopsy proven LS of the trunk (**Figure 3**), vulva, extremities, and breast (**Figure 4A)**, which had been treated with betamethasone 0.05% cream. The physical examination revealed hemorrhagic tense bullae on the inferior aspect of the left breast (**Figure 4B)**, which was diagnosed as bullous hemorrhagic lichen sclerosus based on the clinical history and presentation. The patient was advised to start clobetasol 0.05% ointment twice daily and the plaque showed rapid improvement after three weeks (**Figure 4C**, **D)**.

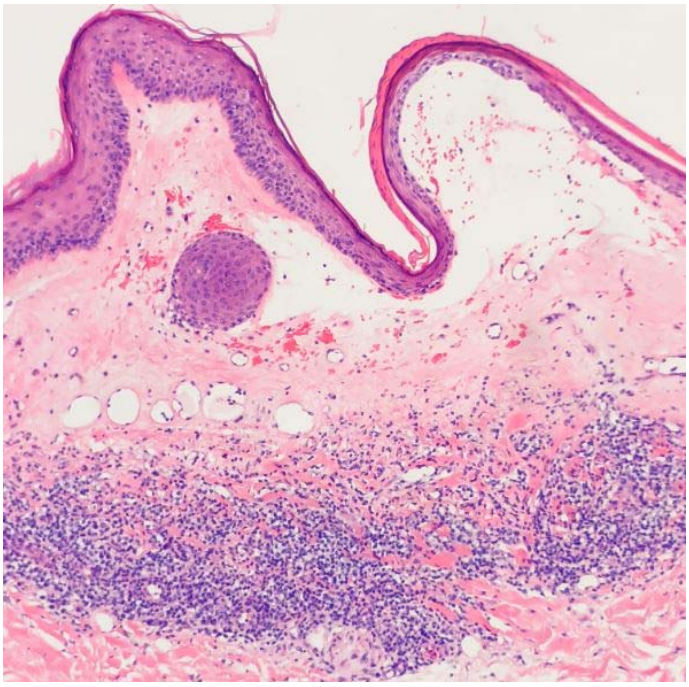

Figure 2. *H&E stain revealing epidermal atrophy with a wide band of hyalinization as well as a dense lymphohistiocytic infiltrate in the superficial dermis. A subepidermal blister with extravasated erythrocytes is also present, 200×.*

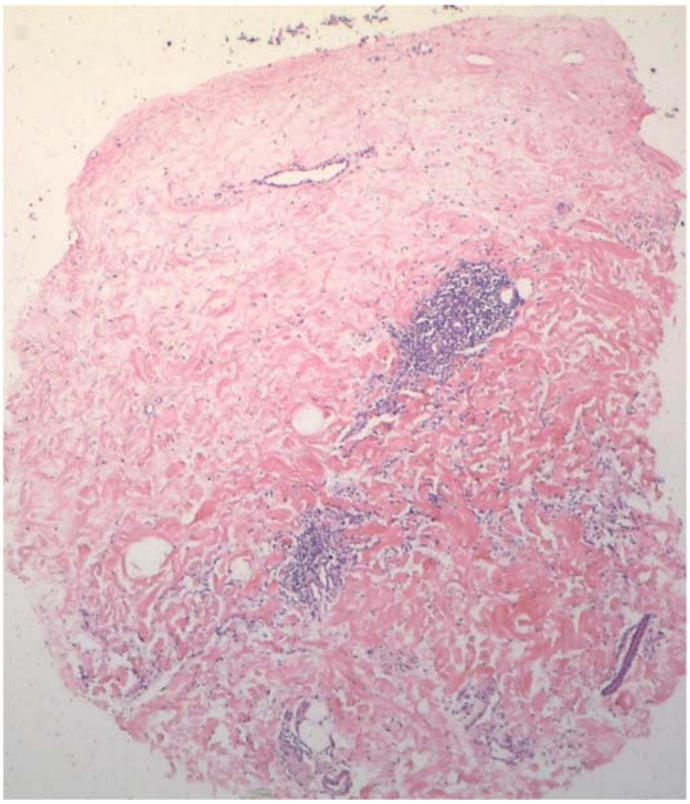

Figure 3. *H&E stain from original biopsy of clinically non-bullous lichen sclerosus of the chest. The epidermis is missing with sclerosis of the upper-mid dermis and associated perivascular lymphocytic inflammation, 40×.*

Case Discussion

Lichen sclerosus classically presents in the anogenital area. However, extragenital lichen sclerosus is present in 15-20% of cases and has many of the same features as classic anogenital LS. It often presents on the trunk, back, axillae, head, neck, shoulders, wrist, inner thighs, and inframammary area. Extragenital lesions tend not to be as symptomatic or pruritic and there is no association with malignancy [2, 3]. The treatment of choice for both anogenital and extragenital LS is high potency

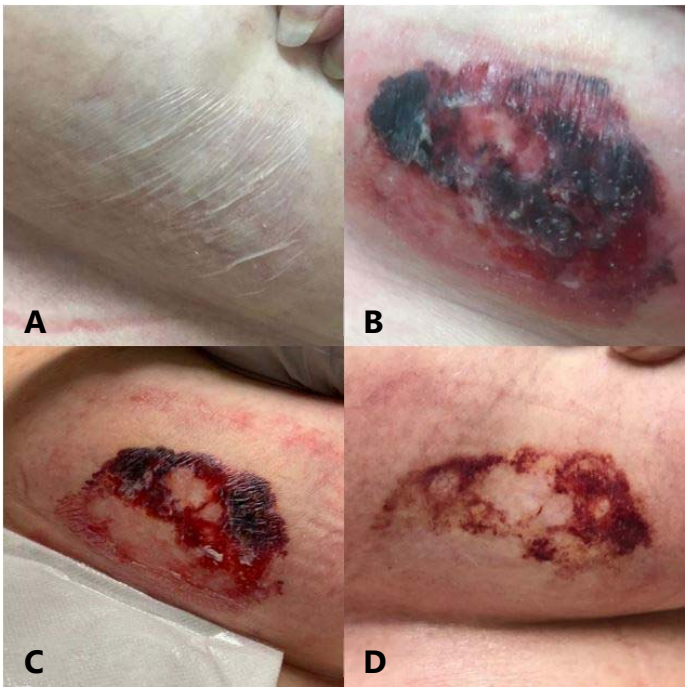

Figure 4. *Progression of lichen sclerosus.* ***A)*** *Previously diagnosed lichen sclerosus without hemorrhage.* ***B)*** *Bullous hemorrhagic LS at initial visit.* ***C)*** *Bullous hemorrhagic LS after one week of clobetasol 0.5% ointment.* ***D)*** *Few remaining areas of purpura after three weeks of clobetasol use.*

(Class I) topical corticosteroids [5]. Bullous hemorrhagic lichen sclerosus is a rare variant of LS in which flaccid bullae develop along with purpuric hemorrhages in the skin and these can be found in both anogenital or extragenital skin [6]. Despite the unique appearance, the prognosis and treatment for this variant of LS does not appear to be different from that of traditional LS. High potency topical corticosteroids have been used to successfully treat both anogenital and extragenital bullous hemorrhagic LS [6]. The treatment response to high potency topical steroids is highlighted in our reported cases as each showed rapid improvement with the use of clobetasol ointment.

Bullous hemorrhagic LS of the breast is exceedingly rare with very few reports in the literature (**Table 1**), [7-12]. Several of these cases were associated with previous radiation treatment for breast cancer and two of these cases arose within [9] and concurrently [12] with post-irradiation morphea. A separate series examining cases of post-irradiation morphea found histological evidence of concurrent LS and morphea in two of the five cases examined [13]. Several other unique cases of LS involving the breast have been reported. One unique case involved LS on both areolae of a 15-year-old boy, in which the left areola had a single small hemorrhagic vesicle but no other bullous or hemorrhagic involvement. That case could represent the earliest stage of, or a minor case of, bullous hemorrhagic LS [14]. Another entity, LS with telangiectasias but not hemorrhage or bullae, has also been reported. These cases of telangiectatic LS appeared after breast irradiation [15, 16].

The exact etiology of bullous hemorrhagic LS is unknown. One potential explanation for the observation that bullae and hemorrhage are so often found together has been proposed. Pronounced edema within the skin could leave the capillaries without surrounding collagen support, thus predisposing them to rupture from minimal trauma or damage, similar to what is seen in senile and corticosteroid induced purpura [17]. A potential explanation for the formation of bullae in LS arising on irradiated breasts is the alteration of lymphatic vessels by radiotherapy. The radiation and potentially also the surgical removal of lymph nodes could predispose the tissue to edema and bulla

Table 1. *Cases reported for bullous hemorrhagic lichen sclerosus.*

Year	Author & Reference	Age/Sex	Radiation	Comments
1979	Faergemann (7)	68 y F	No	Patient also had polymyalgia rheumatica and alopecia totalis
1993	Starzycki (8)	25 y F	No	LS located on both areolae
1994	Trattner et al. (9)	57 y F	Yes	LS arising on pre-existing area of morphea
2010	Vujovic (10)	77 y F	Yes	No pre-existing trauma, radiation, or cancer
2015	Quatrano et al. (11)	65 y F	No	History of breast cancer, but radiation not mentioned
2018	Petersen et al. (12)	57 y F	Yes	LS in superficial dermis, morphea in deep dermis on pathology

Reported cases of bullous hemorrhagic lichen sclerosus (LS) of the breast in the English literature found with the search terms "Bullous Hemorrhagic Lichen Sclerosus" and "Lichen Sclerosus Breast" in PubMed.

The authors declare no conflicts of interests

formation [13]. Neither of our reported cases had a history of breast cancer or radiation and the patient with bilateral breast reduction surgery only had unilateral bullous hemorrhagic involvement. Further exploration and research are necessary to determine the exact mechanisms by which hemorrhagic bullae arise in LS.

Conclusion

Lichen sclerosus is a chronic dermatologic condition that typically presents as atrophic porcelain plaques in the anogenital region of women. Lichen sclerosus can less commonly present in extragenital locations and only rarely presents with hemorrhage and bullae. The mechanism of hemorrhagic bullae formation is not understood but there may be an association with radiation therapy and post-irradiation morphea. Bullous hemorrhagic lichen sclerosus of the breast is exceedingly rare, with a markedly different clinical appearance than classic LS, making it a diagnostic challenge and an important entity for clinicians to recognize.

Potential conflicts of interest

The authors declare no conflicts of interests.

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