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Authors
Arnold, Nichelle
Manway, Mitch
Stephenson, Sean
et al.

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Extragenital bullous lichen sclerosus on the anterior lower extremities: report of a case and literature review

Nichelle Arnold¹, Mitch Manway², Sean Stephenson³, Howard Lipkin³


Corresponding Author: Nichelle Arnold, Beaumont Hospital Farmington Hills, 28050 Grand River Ave, Farmington Hills, MI 48336, Email: arnoldnichelle@gmail.com

Abstract

Lichen sclerosus (LS) is a benign, chronic, inflammatory skin disease with a predilection for the anogenital region in women. Although males can also be affected, the ratio of female to male incidence has been reported to be as high as 6-10:1 and possesses a bimodal age distribution of pre-pubertal girls and postmenopausal women [1, 2]. Affected skin usually demonstrates polygonal papules that coalesce into porcelain white plaques and can be associated with edema, telangiectasias, and comedo-like plug formation [3]. Lichen sclerosus can be debilitating for some patients causing significant pruritus, pain, dysuria, and dyspareunia [4]. Rarely, lichen sclerosus appears in various extragenital areas, although most cases are relatively asymptomatic [3]. Even more uncommonly, as displayed in this case report of a 69-year-old woman, LS can present extragenitally with a bullous or hemorrhagic appearance [5].

Keywords: bullous; lichen sclerosis; lichen sclerosis et atrophicus; extragenital lichen sclerosis

Case Synopsis

The patient is a 69-year-old woman who presented with a chief complaint of a skin rash located on her left and right pretibial regions. The lesions had been present for one year and associated with bleeding, blistering, itching, and pain. She stated that the spots come and go but never completely resolve. She had not used any topical or oral treatments. She denied a history of skin lesions on her legs or elsewhere on her body prior to this episode, including her genital area. The patient was taking many medications, none of which were started around the onset of the rash. She was otherwise well, denying any recent illnesses or significant changes in her health status. Review of symptoms was negative for oral pain or lesions, vaginal pain, dyspareunia, vaginal pruritus, genital lesions, fever, chills, joint pain, muscle pain, nausea, vomiting, or headaches.

Past medical history included: diabetes (type 2), hypercholesterolemia, hypertension, and chronic obstructive pulmonary disease. Her daily medications included: fluticasone propionate, aspirin, metformin, esomeprazole, sertraline, simvastatin, fluticasone propionate inhaled, insulin glargine, and tiotropium inhaled. She has no known drug, environmental or food allergies. The patient is a former smoker but denied any current tobacco use. Surgical history and family history are non-contributory.

Physical exam revealed scattered erythematous papules, plaques, vesicles, and erosions on the anterior tibia bilaterally (Figure 1). Lesions did not appear lichenified and are in various stages ranging from 3mm papules to 1cm erosions with serosanguinous crusting. The surrounding skin was hypopigmented and atrophic. Vesicles were 3mm-5mm and tense; Nikolsky sign was negative. Oral examination did not reveal ulceration, erosion, or lacy white patches. Examination of the labia and perineum was negative for signs of disease including: ulceration, erosion, lichenification, or white plaques. At the time of initial examination two 3 mm punch biopsies were completed on the left anterior lower
leg for hemolysin and eosin (H&E) as well as direct immunofluorescence (DIF). Laboratory evaluation including a complete blood count, metabolic panel, and anti-nuclear antibody test revealed no abnormalities.

Figure 1. Clinical image of right and left anterior lower legs demonstrating hemorrhagic bulla, atrophic epidermis, and erythematous plaques.

Figure 2. H&E histology demonstrating epidermal atrophy, follicular plugging, and papillary dermal edema A) 4x, B) 20x.
Dermatology Online Journal  ||  Photo Vignette

The differential diagnosis included bullous pemphigoid, pemphigus vulgaris, bullous arthropod, bullous lichen sclerosis, and bullous lichen planus.

Pathology: The biopsy for H&E demonstrated hyperkeratosis with follicular plugging and atrophy of the epidermis (Figure 2). There was separation between the epidermis and dermis exhibiting gossamer strands. Focal homogenization of superficial collagen bundles was present, along with a superficial perivascular predominantly lymphocytic inflammatory cell infiltrate. DIF was negative. These results were consistent with a diagnosis of bullous lichen sclerosis.

With the diagnosis of bullous lichen sclerosis, treatment was initiated with clobetasol propionate 0.05% ointment under occlusion twice daily. Upon follow-up, she had no new lesions and reported decreased pain and pruritus.

Case Discussion

The exact pathogenesis of lichen sclerosus is not clear, but there is evidence to support multiple etiologies. Associations include genetic, autoimmune, infectious, chronic irritation/trauma, as well as hormonal influences [2]. There are some who believe that the skin disease morphea, which possesses many similar clinical and histological features, is not actually a separate entity but is a presentation along the same disease spectrum [6]. Extragénital disease most commonly appears on the thighs, buttocks, breasts, back, chest, axillae, shoulders, and wrists. However, many unique presentations have been reported, including cases of infraorbital and intraoral LS [1, 2, 7]. Although the clinical presentation is somewhat variable, LS usually begins as slightly elevated polygonal bluish-white papules, which coalesce over time into erythematous plaques with an increasingly atrophic and wrinkled appearance. Advanced features include follicular plugging, telangiectasia, and bullous and hemorrhagic lesions, which are caused by the fragility of a flattened epidermal-dermal interface [2, 4]. Histologic features include epidermal atrophy, luminal hyperkeratosis, follicular plugging, edematous homogenized superficial dermis, dilated blood vessels, loss of rete ridges, hydropic degeneration of the basal layer, and a dermal lymphocytic infiltrate that is sometimes referred to as band-like [2, 8, 9]. Rarely, as in our patient, a subepidermal blister can be present. That being said, histologic characteristics are not diagnostic alone and a combination of clinical and pathologic information must be considered for proper diagnosis. Dermoscopic evaluation may reveal scales, keratotic plugs, erosions, and chrysalis structures [10]. On the other hand, immunohistochemical studies have shown decreased immunoreactivity to Ki-67 and p53 [11]. The differential diagnosis of extragenital bullous LS includes bullous morphea, bullous lichen planus, cicatricial bullous pemphigoid, bullous scleroderma, and circumscribed lymphangioma [12].

Some reports have shown an increase in risk of squamous cell carcinoma development in anogenital forms of LS, but no strong association has been shown with extragenital cases [13]. Treatment is aimed at relieving symptoms of dryness and pruritus and improving appearance. Unfortunately, there are very few well-designed randomized clinical trials and therefore, treatment options are based upon limited observations [4]. Ultra-potent topical corticosteroids such as clobetasol propionate 0.05% cream are first line and will be effective in most patients, but not all [14]. Length of treatment with this method has varied from 12-24 weeks [15]. Should this treatment prove ineffective or if long-term corticosteroid use is undesirable owing to potential adverse effects, other potential treatment modalities include injection of corticosteroids, topical calcineurin inhibitors such as pimecrolimus or tacrolimus, vitamin derivatives such as vitamin D and systemic retinoids, and UV phototherapy [4, 5, 14]. Use of hormone therapies such as topical testosterone or progesterone have not shown good efficacy and should be avoided [16]. Regrettably, there is no permanent cure for lichen sclerosus and relapses may transpire [4]. Therefore, we will continue to follow our patient with regular examinations to ensure that her lesions remain under control and that she has no signs of malignancy. Although rare, this case is a reminder that lichen sclerosus should remain on the differential diagnosis list for a patient who presents with a blistering dermatosis.

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References


