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

Nichelle Arnold DO, Mitch Manway DO, Sean Stephenson DO, Howard Lipkin DO
Department of Dermatology  ■  Beaumont Hospital, Farmington Hills, MI

INTRODUCTION

Lichen sclerosis (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 sclerosis can be debilitating for some patients causing significant pruritus, pain, dysuria, and dyspareunia[4]. Rarely, lichen sclerosis appears in various extragenital areas, however most cases are relatively asymptomatic [5]. Even more uncommon, as displayed in this case report of a 69 year old female, LS can present extragenitally with a bullous or hemorrhagic appearance [5].

CASE REPORT

The patient is a 69 year old female who presents with a chief complaint of a skin rash located on her left and right pretibial regions. The lesions have been present for one year and are associated with bleeding, blistering, itching, and pain. She states that the spots come and go but never completely resolve. She has not used any topical or oral treatments. She denies a history of skin lesions on her legs or elsewhere on her body prior to this episode. The patient is on many medications, none of which were started around the onset of the rash. She is otherwise well, denying any recent illnesses or significant changes in her health status.

Review of symptoms is negative for fever, chills, joint pain, muscle pain, nausea, vomiting or headaches.

Past medical history includes: diabetes type 2, hypercholesterolemia, hypertension, and chronic obstructive pulmonary disease. Her daily medications include: flavocapone proprionate, Aspirin, Metformin, Nexium, sotalolin, simvasstain, Flovent disco, Lantus, and Spirliva. She has no known drug, environmental or food allergies. The patient is a former smoker but denies any current tobacco use. Surgical history and family history is non-contributory.

Physical exam reveals scattered erythematous papules, plaques, vesicles, and erosions on the anterior tibia bilaterally (figures 1 and 2). Lesions do not appear lichenified and are in various stages ranging from 3mm papules to 1cm erosions with serosanguinous crust. The surrounding skin is hypopigmented and atrophic. Vesicles are 3mm – 5mm, taut, and Nicksky’s sign is negative. At the time of initial examination two 3mm 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.

Differential diagnosis includes:

• bullous pemphigoid
• pemphigus vulgaris
• bullous arthropod
• bullous lichen sclerosis
• bullous lichen planus

Pathology: The biopsy for H&E demonstrated hyperkeratosis with follicular plugging and atrophy of the epidermis (figures 3 and 4). There is separation between the epidermis and dermis exhibiting follicular plugging, and papillary dermal edema.

DISCUSSION

The exact pathogenesis of lichen sclerosis is not clear, but there is evidence to support multiple etiologies. Associations include genetic, autoimmune, infectious, chronic inflammatory, and hormonal influences [5].

Extragenital disease most commonly appears on the thighs, buttocks, breasts, back, chest, axillae, shoulders, and elbows. However, cases of extragenital LS have been reported, including cases of infantile and infantile extragenital LS [1,2,7]. Although the clinical presentation is somewhat variable, LS usually begins as slightly elevated polygonal whitish papules which coalesce overtime into erythematous plaques with increasing atrophy and telangiectatic appearance. Advanced features include follicular plugging, telangiectasia formation, as well as 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, exudates, homogenized superficial dermis, dilated blood vessels, loss of rete ridges, hypogonic degeneration of the basal layer, and a dermal lymphocytic infiltrate that is sometimes referred to as band-like [2,4]. Rarely, as our case, a subepidermal blister can be present. That being said, histologic characteristics are not diagnostic alone and a combination of clinical and pathological information must be considered for proper diagnosis. Dermoscopic evaluation may reveal follicular plugging, keratin plugs, erosions, and chrysalis structures [16]. On the other hand, immunohistochemical studies have shown decreased immunoreactivity to Ki-67 and p53 [11]. The differential of extragenital 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, pruritus, and improvement in 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]. Lenght of treatment with this method has varied from 12-24 weeks [15]. Should this treatment prove ineffective or if long-term use is undesirable to stave potential adverse effects, other treatment modalities include injection of corticosteroids, topical calcium inhibitors such as pimecrolimus or tacrolimus, vitamin derivates 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 sclerosis and relapse may transpire [4].

CONCLUSION

In conclusion, we will continue to follow our patient with regular examinations to ensure that her lesions are remaining under control and that she has no signs of malignancy. Although rare, this case is a reminder that lichen sclerosis should remain on the differential for a patient presenting with a blistering dermatitis.

REFERENCES