Disseminated extragenital bullous lichen sclerosus

Tu-Van T. Trinh, Karina Parr, David F. Butler

Department of Dermatology, Scott and White Memorial Hospital and Clinic; Scott, Sherwood and Brindley Foundation, and The Texas A&M University System Health Science Center College of Medicine, Temple, Texas, United States of America

ABSTRACT

Lichen sclerosus commonly affects the genitalia of post-menopausal women. We describe a woman with painful, disseminated, bullous, extragenital lichen sclerosus that responded to oral acitretin and topical calcitriol and triamcinolone.

Key words: Acitretin, bullous, calcitriol, disseminated, lichen sclerosis et atrophicus, non-genital, painful

INTRODUCTION

Lichen sclerosus (LS) is a chronic inflammatory dermatosis of unknown etiology that presents with severe pruritus, epidermal atrophy and dermal sclerosis affecting predominately the anogenital area of post-menopausal females. The extragenital areas most commonly involved are neck, shoulders, trunk, and proximal extremities and are generally asymptomatic. We describe a case of symptomatic disseminated extragenital lichen sclerosus with the development of hemorrhagic bullae.

CASE REPORT

A 61-year-old Caucasian female was seen with a several year history of large pruritic plaques on the trunk and proximal extremities that proved to be lichen sclerosus et atrophicus on biopsy. Over the preceding months many of the truncal lesions developed painful bullae. Multiple therapies including ultra-potent topical steroids, topical vitamin D derivative (Dovonex), narrowband ultraviolet (UV) B, and hydroxychloroquine had failed to alleviate her symptoms or halt the progression of her dermatitis in the past.

Physical examination revealed several hypopigmented atrophic plaques, ranging in size from 5 cm to 10 cm, consistent with LS lesions that were widely distributed over the trunk, arms, forearms, and medial thighs. Hemorrhagic bullae were present within pre-existing atrophic lesions involving her abdomen, left flank, bilateral medial arms and antecubital fossae [Figure 1]. No oral or genital lesions were noted. The remainder of the physical examination was unremarkable.

Biopsies of the bullous lesions on the left flank were obtained for routine histological examination and immunofluorescence staining. Hematoxylin and eosin staining demonstrated features consistent with bullous LS including thinning of the epidermis with compact hyperkeratosis, degeneration of the basal layer, edema with the early stages of a subepidermal blister, and a band of homogenized collagen in the dermis [Figure 2]. Direct immunofluorescence staining was negative.

The patient was treated with topical calcitriol 3 mcg/g ointment and topical triamcinolone 0.1% cream daily in combination with oral acitretin 25 mg daily and responded with dramatic improvement of her lesions and level of pain. Future therapy considerations include high intensity ultraviolet A1 and immunosuppressive therapy with prednisone or methotrexate.

COMMENT

LS was first described clinically by Hallopeau in 1887 and histopathologically by Darier in 1892.[1] LS is a chronic inflammatory dermatosis presenting with sclerosis, atrophy, and pruritus affecting predominately the anogenital region. Both sexes are affected with a predilection to females and a peak incidence during the fifth and sixth decades.[2] Recent studies indicate a higher prevalence of LS in patients with morphea.[3] The etiology of LS is unknown, although there

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Address for correspondence:
Dr. Karina Parr,
Scott and White Northside Clinic,
409 West Adams Street,
Temple, Texas 76508,
United States of America.
E-mail: kparr@sw.org

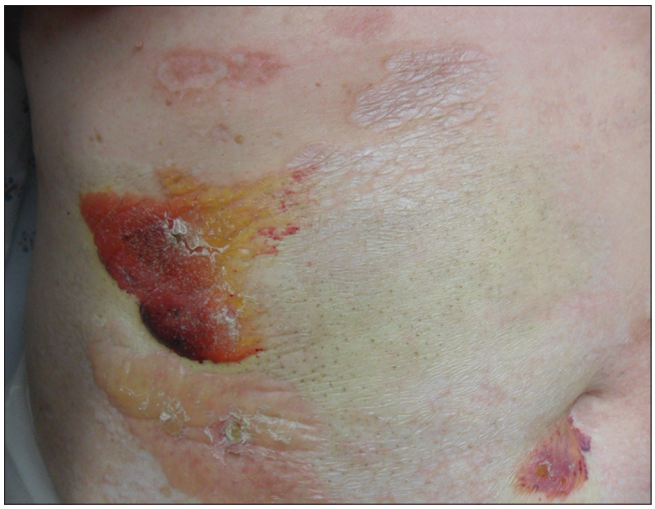

Figure 1: Hemorrhagic bullae within pre-existing atrophic lesions involving her abdomen

is an association with autoimmune diseases including thyroid disorders, vitiligo, alopecia areata, and type I diabetes.[4] Several other factors including genetic susceptibility, low levels of androgens, chronic infections, and trauma have been implicated as pathogenic factors.[4-6]

Genital LS leads to progressive atrophy and destructive scarring resulting in dryness, severe pruritus, pain, and often functional impairment (phimosis in men and stenosis of the vaginal introitus in women). It is estimated that 15-20% of cases of anogenital LS also involve the extragenital areas while exclusive involvement of extragenital area, as in our patient, accounts for only 2.5% of all cases of LS.[5,6] Extragenital LS is generally asymptomatic and preferentially involves the neck, shoulders, trunk, and proximal extremities.[2] Occasionally, extragenital LS may be distributed along Blaschko lines.[7]

Clinically, early lesions present as pearly interfollicular papules that progress into sclerotic, atrophic, ivory-white plaques. In advanced stages, follicular plugging and telangiectasias may be noted. Characteristic histopathological findings of LS include marked thinning of the epidermis with hyperkeratosis, follicular plugging, degeneration of the basal layer, a band of homogenized collagen in the papillary dermis above a varying degree of lymphocytic infiltrate.[8] Extensive vacuolar degeneration of the basal layer and edema may lead to fragility of the dermal-epidermal junction resulting in bulla formation.[9] Bullous LS may be localized or generalized and is often associated with hemorrhage, as seen in our patient's presentation.

Although there is no definitive or satisfactory treatment for LS, ultra-potent topical corticosteroids remains the treatment of choice.[10,11] Numerous therapeutic modalities including topical and systemic vitamin A and D analogs, topical androgens, topical calcineurin inhibitors, systemic corticosteroids,

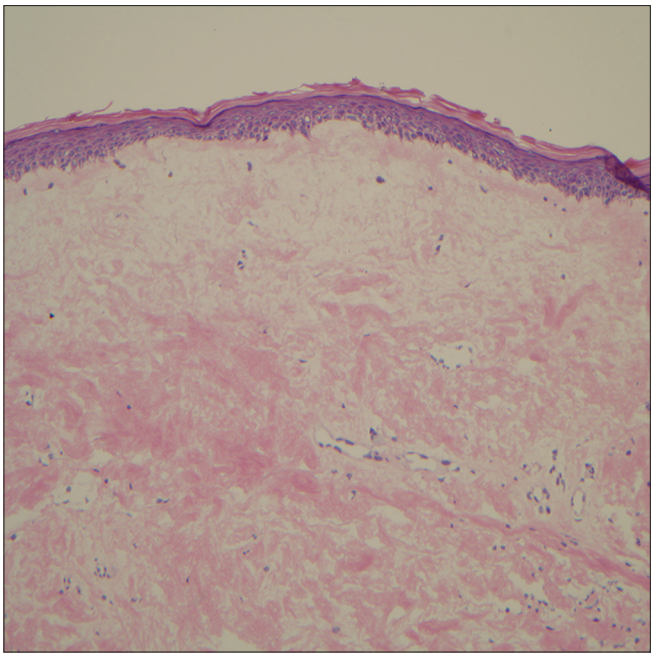

Figure 2: Punch biopsy of the left flank showing thinning of the epidermis with compact hyperkeratosis, degeneration of the basal layer, edema with the early stages of a subepidermal blister, and a band of homogenized collagen in the dermis (H and E, ×100)

systemic hydroxychloroquine, phototherapy, and surgery have been used with varying degrees of symptomatic relief.[10-13] Extragenital lesions are less responsive to conventional therapy.[11] Narrowband UVB has been reported to provide dramatic symptomatic relief in a case of extragenital LS recalcitrant to multiple topical modalities.[14] While several cases have demonstrated effectiveness of UV-A1 therapy and immunosuppressive therapy with oral prednisone and methotrexate in extragenital lesions.[15,16]

LS is a chronic condition that often progresses to include destructive scarring which results in functional impairment, reduced self-image, marked distress, and anxiety leading to a decreased quality of life. Our patient highlights the importance of considering bullous LS in any patient presenting with persistent pruritus and blister formation even in the absence of genital lesions. Moreover, histology and direct immunofluorescence is essential for the diagnosis when widespread bullae are present.

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