Lichen Sclerosus: An Atypical Presentation

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We describe the case of a 66-year-old Hispanic man with an atypical presentation of lichen sclerosus (LS). The unusual presentation included bilateral axilla involvement (not previously reported to our knowledge), scrotal involvement (not common in men, despite common vulvar involvement in women), and an uncommonly thick plaque on his back.

Lichen sclerosus (LS) is a chronic inflammatory dermatosis of uncertain etiology. A case report by Hallopeau in 1887 usually is considered the first clinical description of LS.[1] Since 1887, many synonyms have been used in the medical literature, the most common being lichen sclerosus et atrophicus, balanitis xerotica obliterans (involvement of the glans penis), and kraurosis vulvae (vulvar involvement).[2] We report the case of a patient with an atypical presentation of LS in both axillae, on the scrotum, and as a thick plaque on the back.

Case Report

A 66-year-old Hispanic man was referred to our institution for an enlarging pruritic plaque on his back of one year's duration that was currently being treated with a topical antifungal cream, clotrimazole. Physical examination revealed a thick, erythematous, scaly plaque on the patient's mid back (Figure 1). Further examination revealed areas of cigarette paper atrophy overlying violaceous and hyperpigmented, scaly, coalescing, annular patches. Both axillae (Figure 2), the lower abdomen, and the genital area were involved. Noteworthy was the involvement of the scrotum and penile shaft. No other cutaneous findings were noted.

Results of a histologic examination of punch biopsies of the back and lower abdomen revealed hyperkeratosis, follicular plugging, thinning of the epidermis, and vacuolar alteration of the basal layer. A broad zone of subepidermal edema, with homogenization of collagen and diffuse perivascular infiltrate of lymphocytes in the mid dermis also were noted (Figure 3). Results from spirochetal silver stains and lyme serologies were normal.

The patient initially was started on topical triamcinolone 0.1% cream. However, after biopsy results were known, he was switched to clobetasol 0.05% cream. After the patient showed marked improvement, we used both triamcinolone (for weekdays) and clobetasol (for weekends).

Comment

LS is a chronic inflammatory dermatosis. The pathogenesis is unclear. Among the suggested causes are infectious agents (eg, *Borrelia*), Köbner phenomenon (eg, posttraumatic appearance in burn scars, surgical scars, cases of sexual abuse), autoimmune disorders (eg, vitiligo, thyroid disease, pernicious anemia, diabetes mellitus, alopecia areata), genetic predisposition, and decreased hormone levels.[2-7] Because of the unknown pathogenesis, biopsy confirmation of a suspected area is the only consistent workup recommended.

There is an up to 10:1 ratio in female-to-male predominance of LS. Population frequency and true incidence are unknown, though 10% to 15% of the reported cases occur in children. No racial predilection has been established. A distinct bimodal distribution (prepubertal and middle age/menopausal) of occurrences has been noted in both sexes.[2]

Classically, the lesions in LS present as discrete and confluent white papules and plaques, with surrounding mild erythema that coalesce into atrophic patches and show a smooth, slightly wrinkled texture.[2,3] Also, it has been reported that the lesions can follow Blaschko lines.[8] Lesions are commonly classified as extragenital, genital, or a combination. Extragenital lesions may be asymptomatic and occur anywhere on the body, most often on the neck, shoulders, or upper portion of the trunk. An English literature search of the Ovid database using the terms

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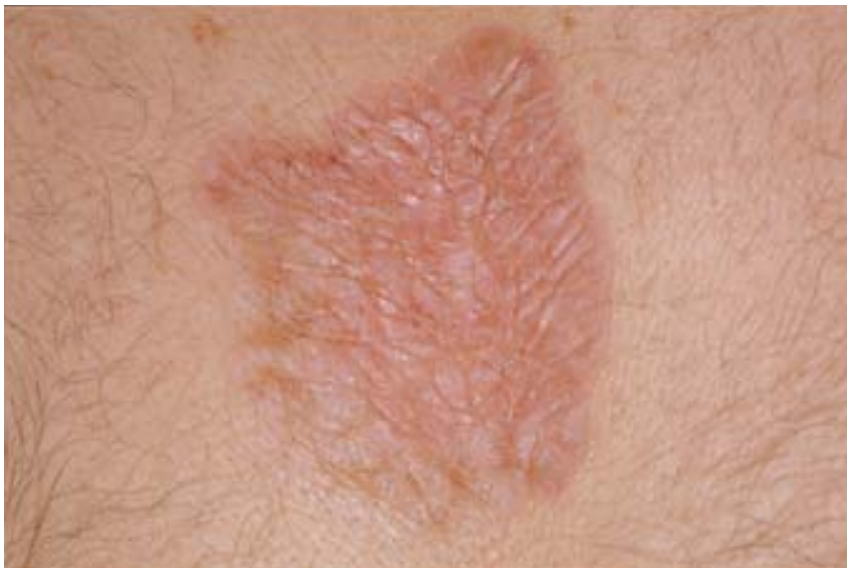

Figure 1. Thick, erythematous, scaly plaque on the patient's mid back.

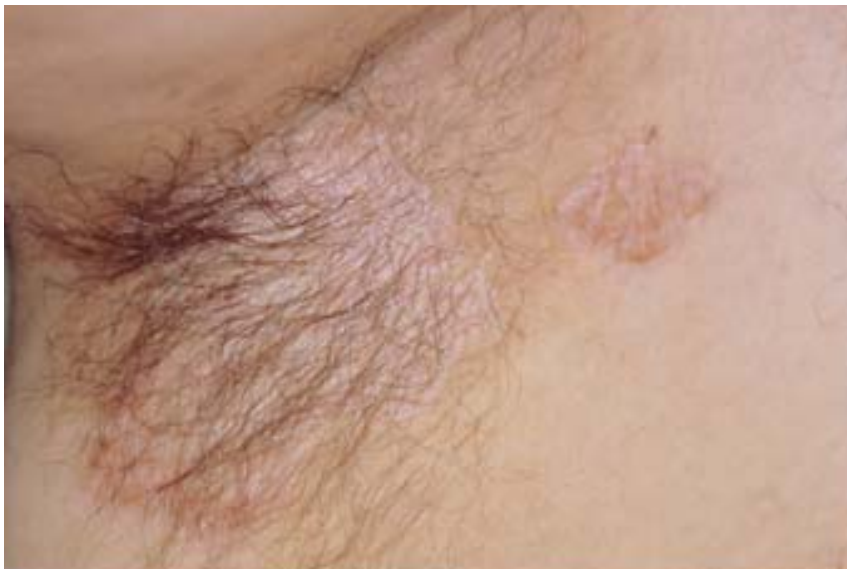

Figure 2. Annular atrophic patch on the axilla.

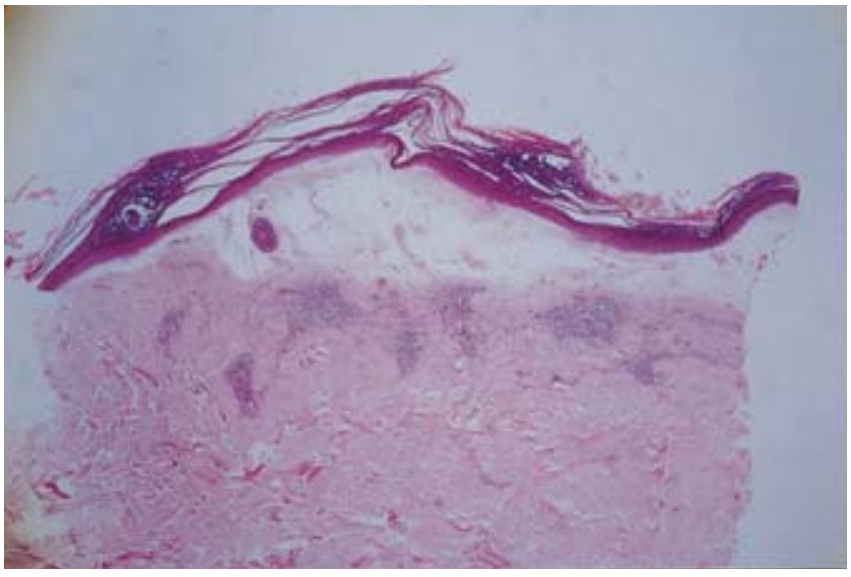

Figure 3. Histologic view of lichen sclerosus (H&E, original magnification ×100).

lichen sclerosus and *axilla* failed to reveal previous reports involving the axillae. Female genital lesions typically present with pruritus, bleeding, or pain and occur mainly on the labia majora but in time can involve the labia minora and introitus. Male genital lesions usually present with pruritus or phimosis and involve the glans penis and prepuce. Much less common is involvement of the penile shaft, and scrotal involvement, as seen in our patient, is quite rare.[2]

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reasons, isotretinoin is not indicated for ongoing long-term treatment. Therefore, if a patient still has severe draining lesions after completing treatment with isotretinoin, management is a difficult problem.

Acitretin, the primary metabolite of etretinate, is used to treat severe psoriasis and disorders of keratinization. A second-generation retinoic acid derivative, acitretin exerts its pharmacological effect through inhibition of epidermal growth and differentiation.[6] Because acitretin is indicated for ongoing treatment, it may be useful for the management of nodulocystic acne and hidradenitis suppurativa that are not adequately suppressed by isotretinoin.

Case Report

A 41-year-old man presented with severe nodulocystic facial acne and hidradenitis suppurativa on the inguinal folds. The conditions were not controlled with various oral antibiotics prescribed by previous physicians. The patient was treated with 2 full courses of isotretinoin. The first course of treatment consisted of 4 months of isotretinoin at 1.4 mg/kg per day, followed by $4^1/_2$ months at 2 mg/kg per day. Although some draining cysts on the patient's face and groin improved throughout treatment, they were persistent. A second course of isotretinoin at 2 mg/kg per day for 6 months yielded similar results. During treatment with isotretinoin, the patient's triglyceride levels were greater than 350 mg/dL, despite taking simvastatin 40 mg by mouth at bedtime. Adding dapsone 100 mg daily to the patient's treatment with isotretinoin did not improve his response. A pulse therapy trial of isotretinoin for one week per month also was ineffective. Between courses of isotretinoin, the patient's condition, including his triglyceride level, returned to baseline with numerous draining facial and inguinal cysts.

Because of the inability of isotretinoin to achieve long-term remission of the patient's condition, acitretin was considered as a possible maintenance drug. After 2 months of acitretin 50 mg by mouth daily, the patient's hidradenitis was completely controlled, and his acne improved to only a few inflamed nondraining facial cysts. The patient's triglyceride level remained within normal limits at 174 mg/dL. Based on the patient's weight, 85 kg, the dose was increased to 75 mg daily (a dose tolerated in premarketing trials) to see if further improvement of acne could be achieved. After one month on this dosage, the patient was completely free of inflammatory lesions on the face and groin. After 4 months at this dosage, however, alopecia and unacceptable joint pain developed. After one month off acitretin, the patient's side effects resolved. Treatment was resumed at a dose of 50 mg daily, with results similar to when the patient was previously on this dosage, with no resumption of joint pain. Because this improvement was satisfactory to the patient, it was decided he would remain at this dosage. After 5 months of therapy, improvement continued to be satisfactory.

Comment

Both nodulocystic acne and hidradenitis suppurativa have been widely managed with short courses of isotretinoin.[1-4] However, it is an unacceptable maintenance drug for patients who are unable to obtain long-term remission of these conditions. Concerns have been expressed regarding adverse events and long-term use of isotretinoin. Acitretin has reportedly had success in the long-term outcomes in patients treated for skin disorders such as the pustular or erythrodermic types of psoriasis.[7,8] Systemic retinoid therapy side effects such as conjunctivitis; hair loss; dry skin; and elevated levels of triglycerides, aspartate aminotransferase, alanine aminotransferase, and lactate dehydrogenase can generally be effectively managed through dosing adjustments, lipid therapy, and careful patient monitoring.[7,8] Acitretin was used in our patient for ongoing treatment of recalcitrant nodulocystic acne and hidradenitis suppurativa. The patient was almost completely improved after 5 months of acitretin therapy. Acitretin also was effectively used for the patient's ongoing maintenance. Acitretin may be a promising treatment for severe nodulocystic acne and hidradenitis suppurativa, which requires long-term suppression when isotretinoin fails to give long-term remission.

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