Hyperkeratotic Nummular Plaques on the Upper Trunk

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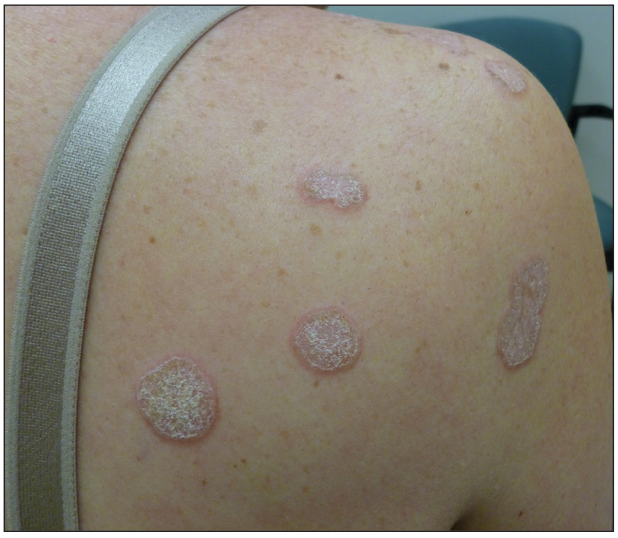

A 48-year-old woman with a history of type 2 diabetes mellitus and nonalcoholic steatohepatitis presented with papules and plaques on the upper trunk, proximal extremities, and volar wrists. Clear fluid–filled bullae occasionally developed within the plaques and subsequently ruptured and healed. Aside from intermittent lesion tenderness and irritation with the formation and rupture of the bullae, the patient's plaques were asymptomatic, and she specifically denied pruritus. A review of systems revealed a history of genital irritation evaluated by a gynecologist; nystatin–triamcinolone cream 0.1% applied as needed provided relief. The patient denied any recent flares or any new or changing oral mucosa findings or symptoms, preceding medications, or family history of similar lesions. Physical examination revealed well-demarcated, round, pink plaques with keratotic scale scattered across the upper trunk and central chest. The bilateral volar wrists were surfaced by well-circumscribed, thin, pink to violaceous, hyperkeratotic papules.

WHAT'S YOUR **DIAGNOSIS?**

a. extragenital lichen sclerosus et atrophicus
b. lichen planus
c. lichen simplex chronicus
d. nummular eczema
e. subacute cutaneous lupus erythematosus

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THE **DIAGNOSIS:** Extragenital Lichen Sclerosus Et Atrophicus

Histopathologic evaluation revealed hyperkeratosis, follicular plugging, epidermal atrophy, and homogenization of papillary dermal collagen with an underlying lymphocytic infiltrate (Figure 1). Direct immunofluorescence of a plaque with a superimposed bulla was negative for deposition of C3, IgG, IgA, IgM, or fibrinogen. Accordingly, clinicopathologic correlation supported a diagnosis of extragenital lichen sclerosus et atrophicus (LSA). Of note, the patient's history of genital irritation was due to genital LSA that preceded the extragenital manifestations.

Lichen sclerosus et atrophicus is an inflammatory dermatosis that typically presents as atrophic white papules of the anogenital area that coalesce into pruritic plaques; the exact etiology remains to be elucidated, yet various circulating autoantibodies have been identified, suggesting a role for autoimmunity.[1,2] Lichen sclerosus et atrophicus is more common in women than in men, with a bimodal peak in the age of onset affecting postmenopausal and prepubertal populations.[1] In women, affected areas include the labia minora and majora, clitoris, perineum, and perianal skin; LSA spares the mucosal surfaces of the vagina and cervix.[2] In men, uncircumscribed genital skin more commonly is affected. Involvement is localized to the foreskin and glans with occasional urethral involvement.[2]

In contrast, extragenital LSA tends to present as asymptomatic papules and plaques that develop atrophy with time, involving the back, shoulders, neck, chest, thighs, axillae, and flexural wrists[2,3]; an erythematous rim often is present,[4] and hyperkeratosis with follicular plugging may be prominent.[5] Our patient's case emphasizes the predilection of plaques for the chest and intermammary skin (Figure 2A). Approximately 15% of LSA cases have extragenital involvement, and extragenital-limited disease accounts for roughly 5% of cases.[6,7] Unlike genital

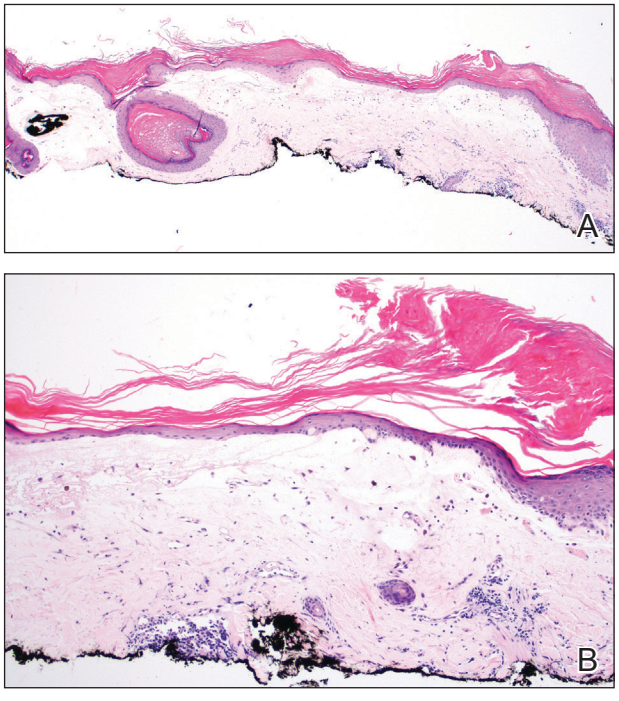

FIGURE 1. A, Hyperkeratosis with prominent follicular plugging and epidermal atrophy with flattening of rete ridges overlying homogenized papillary dermal collagen and a transected lymphocytic infiltrate (H&E, original magnification ×40). B, Associated effacement of the basal layer and scattered Civatte bodies (H&E, original magnification ×100).

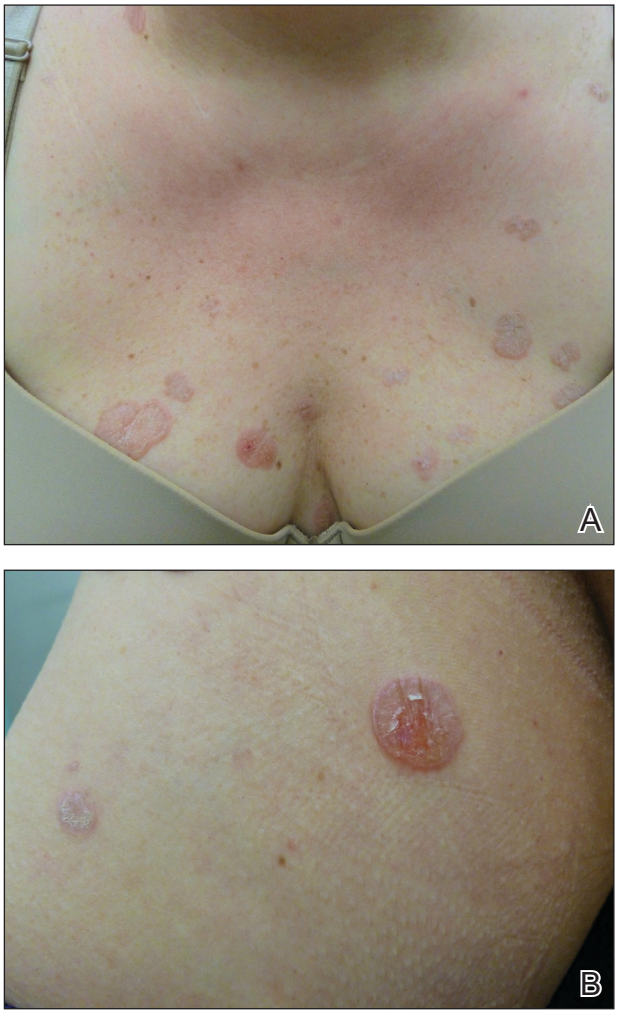

FIGURE 2. A, Additional well-demarcated, hyperkeratotic, pink plaques scattered across the sternal chest, bilateral breasts, and intermammary skin. B, A peeling collarette of fine scale at the site of a recently ruptured bullae centered over a proximal anteromedial thigh plaque.

LSA, extragenital disease has not been associated with an increased risk for squamous cell carcinoma.[1] Bullae formation within plaques of genital or extragenital LSA has been reported[3,8] and is exemplified in our patient (Figure 2B). Intralesional bullae formation likely is due to a combination of internal and external factors, mainly the inability to withstand shear forces due to an atrophic epidermis with basal vacuolar injury overlying an edematous papillary dermis with altered collagen.[8] Dermatoscopic findings may aid in recognizing extragenital LSA[9,10]; our patient's plaques demonstrated the characteristic findings of comedolike openings, structureless white areas, and pink borders (Figure 3).

The clinical differential diagnosis for well-demarcated, pink, scaly plaques is broad. Nummular eczema usually presents as coin-shaped eczematous plaques on the dorsal aspects of the hands or lower extremities, and histology shows epidermal spongiosis.[11] Nummular eczema may be considered due to the striking round morphology of various plaques, yet our patient's presentation was better served by a consideration of several papulosquamous disorders.

Lichen planus (LP) presents as intensely pruritic, violaceous, polygonal, flat-topped papules with overlying reticular white lines, or Wickham striae, that favor the flexural wrists, lower back, and lower extremities. Lichen planus also may have oral and genital mucosal involvement. Similar to LSA, LP is more common in women and preferentially affects the postmenopausal population.[12] Additionally, hypertrophic LP may obscure Wickham striae and mimic extragenital LSA; distinguishing features of hypertrophic LP are intense pruritus and a predilection for the shins. Histology is defined by orthohyperkeratosis, hypergranulosis, sawtooth acanthosis, and vacuolar degeneration of the basal layer with Civatte bodies or dyskeratotic basal keratinocytes overlying a characteristic bandlike infiltrate of lymphocytes.[12]

Lichen simplex chronicus (LSC) is characterized by intense pruritus and presents as hyperkeratotic plaques with a predilection for accessible regions such as the posterior neck and extremities.[13] The striking annular demarcation of this case makes LSC unlikely. Comparable to LSA and LP, LSC also may present with both genital and extragenital findings. Histology of LSC is characterized by irregular acanthosis or thickening of the epidermis with vertical streaking of collagen and vascular bundles of the papillary dermis.[13]

Subacute cutaneous lupus erythematosus (SCLE) is important to consider for a new papulosquamous eruption with a predilection for the sun-exposed skin of a middle-aged woman. The presence of papules on the volar wrist and history of genital irritation, however, make this entity less likely. Similar to LSA, histologic examination of SCLE reveals epidermal atrophy, basal layer degeneration, and papillary dermal edema with lymphocytic inflammation. However, SCLE lacks the band of inflammation underlying pale homogenized papillary dermal collagen, the most distinguishing feature of LSA; instead, SCLE shows superficial and deep perivascular and periadnexal lymphocytes and mucin in the dermis.[14]

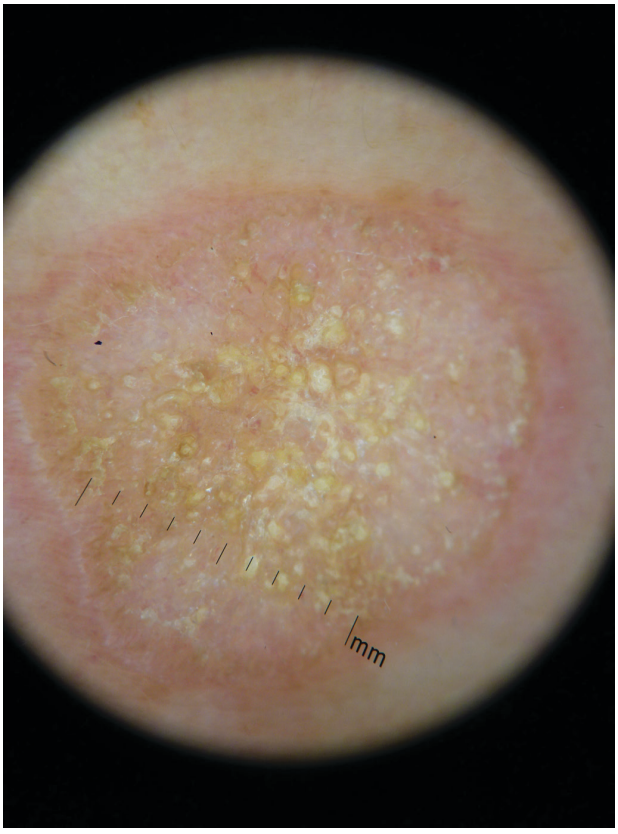

FIGURE 3. Dermoscopy showed a prominent yellow-white comedolike opening, white patches, and a pink border that correlated to the histologic findings of follicular plugging, epidermal atrophy, and bandlike lymphocytic inflammation, respectively (original magnification ×10).

Lichen sclerosus et atrophicus may be chronic and progressive in nature or cycle through remissions and relapses.[2] Treatment is not curative, and management is directed to alleviating symptoms and preventing the progression of disease. First-line management of extragenital LSA is potent topical steroids.[1] Adjuvant topical calcineurin inhibitors may be used as steroid-sparing agents.[2] Phototherapy is a second-line therapy and even narrowband UVB phototherapy has demonstrated efficacy in managing extragenital LSA.[15,16] Our patient was started on mometasone ointment and calcipotriene cream with slight improvement after a 6-month trial. Ongoing management is focused on optimizing application of topical therapies.

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