

Porcelain White, Crinkled, Violaceous Patches on the Inner Thighs

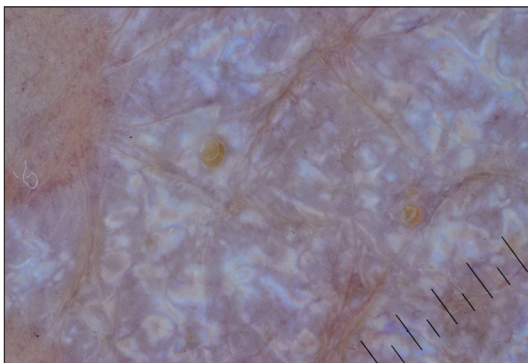
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A 50-year-old woman presented with multiple pruritic lesions on the right inner thigh of 2 years' duration. Physical examination revealed porcelain white, crinkled, violaceous patches extending from the right inner thigh to the inguinal fold (top). Dermoscopic examination revealed follicular plugs, white structureless areas, white lines, and a rainbow pattern arranged over white polygonal clods on polarized mode (bottom).

WHAT'S YOUR DIAGNOSIS?

- extragenital lichen sclerosus et atrophicus
- lichen planus
- lichen simplex chronicus
- morphea
- vitiligo



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THE DIAGNOSIS: Extragenital Lichen Sclerosus et Atrophicus

A punch biopsy of the lesion revealed epidermal hyperkeratosis, atrophy, follicular plugs with basal vacuolar degeneration, and homogenous dense fibrosis in the papillary dermis with a dense lymphocytic infiltrate beneath the fibrosis (Figure 1). Dermoscopic examination was remarkable for a distinctive rainbow pattern. Clinical, histopathologic, and dermoscopic findings led to the diagnosis of extragenital lichen sclerosus et atrophicus (LSEA). A potent corticosteroid cream was prescribed twice daily for 2 months, after which the lesions completely resolved. At 2-year follow-up, a relapse was not observed (Figure 2).

Lichen sclerosus et atrophicus is an inflammatory dermatosis that clinically presents as atrophic or hypertrophic plaques that may show pigmentation changes with anogenital and extragenital involvement. It is common among females and predominantly occurs in prepubescent girls and postmenopausal women. The exact etiology is unclear; however, it is hypothesized to occur secondary to autoimmunity with an underlying genetic predisposition. Local trauma, hormonal influences, and infections are other suspected etiologic factors. Genital lesions often lead to itching, pain, and dyspareunia, whereas extragenital lesions predominantly are asymptomatic. When symptomatic, itching usually is the main concern. Unlike genital LSEA, extragenital lesions are not associated with squamous cell carcinoma development. Reported dermoscopic features of LSEA are white structureless areas with scaling, comedolike openings, follicular plugs, white shiny streaks, blue-gray peppering, pigment network, and red-purple globules.¹ In our case, the dermoscopic finding of a rainbow pattern in LSEA is rare.² Although the mechanism behind this appearance unclear, it can be the result of the birefringence effect—local variations in refractive index—influenced by the direction of structures within the dermis such as collagen. In this case, there was diffuse and dense homogenous fibrosis in the superficial dermis that corresponded to dermoscopic white polygonal clods.

Extragenital LSEA commonly is located on the neck, shoulders, wrists, and upper trunk and manifests clinically as whitish papules coalescing into scarlike plaques. Of all patients who have LSEA, 20% have extragenital lesions, and most of these lesions are seen in patients who also have genital LSEA. Approximately 6% of all LSEA patients have extragenital LSEA without genital involvement.³

For experienced dermatologists, clinical symptoms and lesion characteristics usually are sufficient for diagnosis; however, a differential diagnosis of atypical lesions and isolated extragenital presentations such as

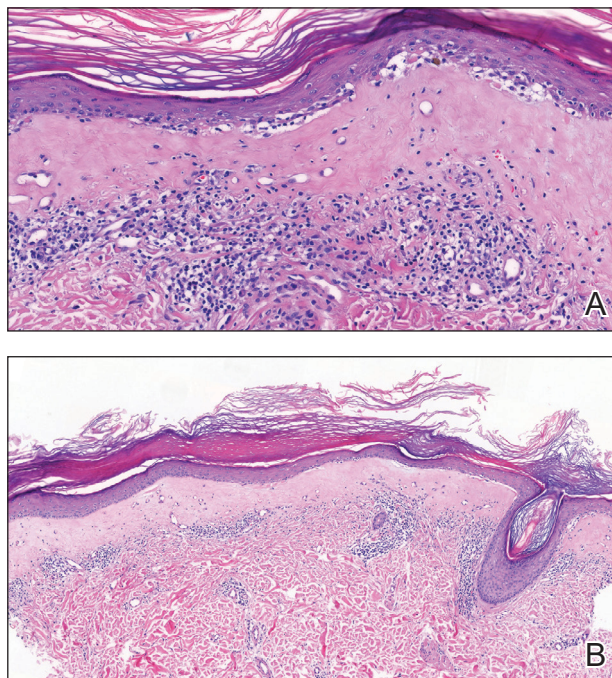


FIGURE 1. A and B, Histopathology revealed epidermal hyperkeratosis, atrophy, follicular plugs with basal vacuolar degeneration, and homogenous dense fibrosis in the papillary dermis with a dense lymphocytic infiltrate beneath the fibrosis (H&E, original magnifications $\times 7.3$ and $\times 21.9$, respectively).

morphea, lichen simplex chronicus, lichen planus, and vitiligo requires the correlation of clinical findings with histopathology and dermoscopy. Morphea, known as localized scleroderma, is an idiopathic inflammatory skin disease with sclerotic changes. It manifests as inflammatory plaques that vary in color from red to purple. If there is moderate sclerosis in the center of this plaque, the color progressively fades to white, leaving a purplish ring around the edges. Dermoscopic features of morphea are reported as areas of erythema; red-focused vessels of linear, irregular, or dotted morphology; white fibrotic beams; and pigmentary structures.² Lichen simplex chronicus is characterized by single or multiple dry and patchy skin lesions that are intensely pruritic. It commonly occurs on the neck, scalp, extremities, genital areas, and buttocks. Scratching the lesions leads to scarring, thickening of the skin, and increased frequency of itching. Histopathology of lichen simplex chronicus most frequently demonstrates a thickening of the epidermis and papillary dermis, irregularly elongated rete ridges, and fibroplasia with stellate or multinucleated fibroblasts completed by perivascular lymphocytic inflammation.⁴



FIGURE 2. At 2-year follow-up, there was no relapse in lichen sclerosus et atrophicus lesions, and only mild atrophy and depigmentation remained after 2 months of topical steroid use.

Lichen planus presents with pruritic, polygonal, purple papules and/or plaques that can present in a variety of clinical forms, including atrophic and hypertrophic lichen planus.⁵ Lichen planus was an unlikely diagnosis for our patient due to the presence of patchy scarlike lesions and dermoscopic features that are well described in patients with LSEA. Lichen sclerosus et atrophicus presents with hypopigmented and/or hyperpigmented patches

and plaques, distinguishing itself from vitiligo, which has flat lesions.

Topical steroids are the first-line therapeutic agents in the treatment of LSEA.⁶ Despite frequent use in this setting, common side effects such as localized scarring and atrophic degenerations have led to debate about their use. In our patient, the lesions resolved almost completely in 2 months, and no relapse was observed in the following 2 years. In the setting of topical steroid resistance, topical calcineurin inhibitors, UVA/UVB phototherapy, and topical tacrolimus can be used for treatment.⁶

The diagnosis of isolated extragenital LSEA may be a clinical challenge and generally requires further workup. When evaluating extragenital lesions, dermatologists should keep in mind extragenital LSEA as a differential diagnosis in the presence of a dermoscopic rainbow pattern arranged over white polygonal clods.

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