

SKIMages

Lichen Sclerosus et Atrophicus

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A Black woman in her 20s presented to dermatology with a three-year history of a slowly expanding eruption beneath her left breast. The rash was occasionally pruritic but otherwise asymptomatic. Review of systems was notable for several months of vaginal irritation and pruritus, but no dysuria or dyspareunia.

On physical exam, there were wrinkled, shiny, atrophic thin plaques with peripheral hyperpigmentation and central mottled hypo- and hyperpigmentation involving the left breast, upper abdomen, and flank. On the left back there were clustered hyperpigmented papules. There was generalized edema and erythema of the

vulva, without erosions, dyspigmentation, or sclerosis. Rheumatologic workup, including antinuclear antibodies, rheumatoid factor, and anti-Ro and anti-La antibodies, was negative.

Biopsies were performed from a hypopigmented plaque on the flank and a hyperpigmented papule on the back. Both showed zones of faint homogenous hyalinization of the papillary dermis, dermal sclerosis with loss of CD34-positivity, lymphoplasmacytic periadnexal inflammation, and loss of fat consistent with a diagnosis of lichen sclerosus-morphea overlap.

The patient was initially treated with high-potency topical steroids, topical tacrolimus, and narrow-band ultraviolet B (UVB) phototherapy, but she had limited adherence and her rash continued to progress. She was started on hydroxychloroquine, and at 3-month follow up her pruritus had improved and the rash stabilized. Her vulvar symptoms were found to be due candida and fully resolved on fluconazole. There were no features concerning for genital lichen sclerosus.

Lichen sclerosus et atrophicus (LSeA), is a benign, chronic, lymphocyte-mediated inflammatory dermatosis affecting the dermis and the epidermis^{1,2}, predominantly in anogenital areas. Extragenital LSeA is rare, with a prevalence of up to 0.3%⁴. It affects all age groups, especially postmenopausal women and prepubertal girls (1). Inadequately treated genital LSeA can lead to atrophy, scarring, and physical dysfunction, with profound social, psychological, and sexual impacts³. Additionally, genital LSeA increases the risk of vulvar squamous cell carcinoma by 4-5%³, therefore, all patients with LSeA should be asked about genital symptoms and a genital examination is recommended.

Extragenital disease typically presents with clusters of white or erythematous papules coalescing into plaques, which later become porcelain-white and atrophic with a wrinkled, cigarette paper-like appearance². As seen in our patient, hyperpigmentation can be a predominant feature in individuals with darker skin. The back, upper torso, armpits, forearms, and inframammary skin are commonly involved¹. The pathogenesis of LSeA is thought to be due to the alteration of fibroblast function in the papillary dermis by inflammation, which results in fibrosis of the upper dermis³.

Diagnosis of LSeA is often made clinically or with biopsy, and while there is no cure, treatment can alleviate symptoms of discomfort such as itching, and reduce the risk of malignant transformation in genital LSeA. Therapeutic options for extragenital LSeA include topical and intralesional corticosteroids, topical tacrolimus, and phototherapy³. LSeA-morphea overlap should be managed similarly to morphea, with lower threshold to initiate immunosuppression.

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