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Extragenital lichen sclerosus et atrophicus-morphea overlap as an initial presentation of genital lichen sclerosus

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Abstract

Lichen sclerosus et atrophicus (LSA) is a chronic inflammatory disorder, most often characterized by atrophic skin plaques located on female genitalia. Infrequently, LSA may present extragenitally; however, much is unknown about the temporal relationship between genital and extragenital LSA. Morphea, also known as localized scleroderma, is a rare inflammatory skin condition characterized by sclerotic plaques. Investigators debate whether LSA and morphea exist on the same spectrum of disease, with LSA representing a superficial variant of morphea involving genitalia, or if they are distinct but coincidental entities. Although researchers have described LSA and morphea occurring in different locations on the same patient, few reports describe LSA and morphea occurring in the same lesion and in the inguinal folds. Herein, we report a case of a 62-year-old woman with extragenital LSA-morphea overlap in the inguinal folds, who three months later developed genital LSA. Extragenital LSA-morphea in the same plaque, with no signs of genital lesions on initial exam, with later development of genital LSA, is especially uncommon. The temporal progression of extragenital LSA-morphea overlap to genital LSA over a three-month period is an important contribution to the literature, as the temporal relationship between extragenital and genital LSA is not previously discussed.

Keywords: extragenital, lichen sclerosus et atrophicus, morphea, overlap, temporal

Introduction

Lichen sclerosus et atrophicus (LSA) is a chronic inflammatory disorder, most often characterized by white, atrophic skin plaques involving female genitalia. Extragenital LSA is uncommon, affecting 15% of LSA patients [1]. Morphea, or localized scleroderma, is a rare inflammatory skin condition that presents with sclerotic plaques. Much is unknown about the temporal relationship between genital and extragenital LSA and the relationship between LSA and morphea is controversial. Herein, we present a 62-year-old woman with extragenital LSA-morphea overlap who later developed genital LSA.

Case Synopsis

A 62-year-old woman presented to Columbia University Irving Medical Center with three months of asymptomatic white plaques on her back and lower extremities. She had no prior dermatologic history and attempted no therapies for her plaques. The patient had a full-body physical exam, including mucosal surfaces. Her examination was notable for round white shiny atrophic papules coalescing into plaques with firm erythematous borders on her back and lower extremities, particularly in her inguinal folds (**Figure 1A**, **B**). Of note, the patient's genital examination was unremarkable. The differential diagnosis was broad, including extragenital LSA, morphea, atrophoderma, vitiligo, and cutaneous T-cell lymphoma.

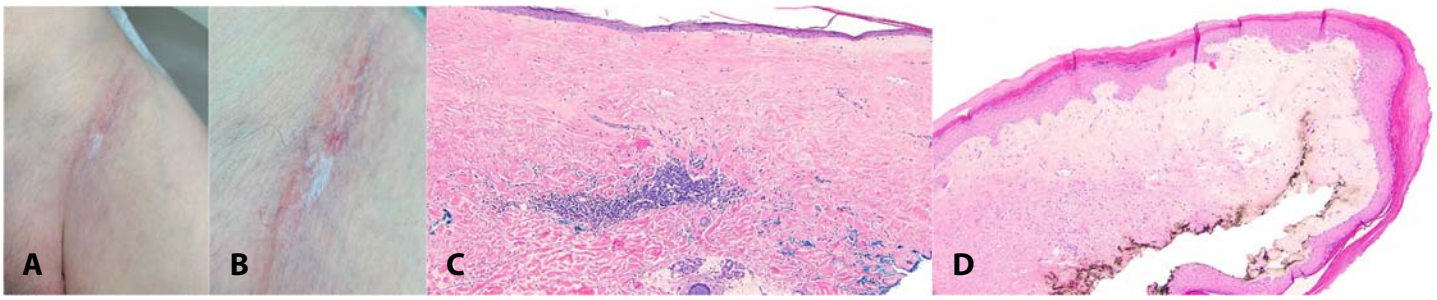

Figure 1. ***A, B)*** *Clinical image: white shiny atrophic plaques with erythematous borders at the left inguinal fold.* ***C)*** *Thigh, H&E histopathology. Flattened epidermis, edema and pallor within the upper dermis, mononuclear cell infiltrate in mid and deep dermis. Thickening of collagen in mid and deep dermis with compression of adnexal structures. Findings are consistent with lichen sclerosus et atrophicus-morphea, 100×.* ***D)*** *Vulva, H&E histopathology. Atrophic epidermis, edema and pallor of the upper dermis, infiltrate of lymphocytes in mid-dermis. Findings are consistent with lichen sclerosus et atrophicus, 100×.*

Skin biopsy of the thigh plaque demonstrated hyalinized connective tissue, entrapment of adnexal structures, thickened collagen bundles deep in the dermis, patchy mononuclear cell infiltrate, edema and pallor of the upper dermis, and flattened epidermis with overlying hyperkeratosis (**Figure 1C**). A diagnosis of LSA-morphea was rendered. The patient was prescribed topical triamcinolone 0.1% ointment alternating with pimecrolimus 1% ointment with minimal improvement.

Three months later, the patient presented to her gynecologist with vulvar pruritus. Physical examination demonstrated hypopigmented plaques on her labia minora and vulvar biopsy was performed. Histopathology showed atrophic epidermis, edema and pallor of the upper dermis, and infiltrate of lymphocytes in mid-dermis, consistent with LSA (**Figure 1D**). The patient was prescribed clobetasol propionate 0.05% ointment alternating with tacrolimus 0.1% ointment and experienced moderate improvement in her symptoms.

Case Discussion

Few reports describe LSA and morphea occurring in the same lesion and in this anatomic location [2-4]. Cases of extragenital LSA-morphea overlap describe plaques on the shoulders, trunk, arms, legs, or submammary folds. They rarely present in the inguinal folds, as in this case. Extragenital LSA-morphea in the same plaque, with no signs of genital lesions on initial exam, with later development of genital LSA is especially uncommon. The temporal progression of extragenital LSA-morphea overlap to genital LSA over a three-month period is an important contribution to the literature, as the temporal relationship between extragenital and genital LSA is not previously discussed. One of the few cases of overlapping LSA and morphea in the same lesion is only extragenital, does not include any genital manifestations of LSA, nor does it discuss the temporal relationship between the two presentations [2].

Investigators debate whether LSA and morphea exist on the same spectrum of disease, with LSA representing a superficial variant of morphea involving genitalia, or if they are distinct but coincidental entities. Clinically, both LSA and morphea present with sclerotic and dyschromic plaques and, histopathologically, they have inflammatory dermal infiltrates. Specifically, LSA is associated with follicular plugging, epidermal atrophy, hyperkeratosis, and homogenized, edematous papillary-upper reticular dermis, whereas morphea is associated with homogenized, sclerotic collagen bundles in the reticular dermis and superficial subcutis, and trapped adnexa [3].

Genital and extragenital manifestations of LSA have been seen in patients with morphea (in distinct skin lesions) and an association has been proposed. Farrell et al. examined 9 patients with genital LSA and found that 7 had genital LSA and morphea [4]. Lutz et al. found that of 76 patients with morphea, 45% also had genital LSA [5]. Kreuter et al. examined genital and extragenital LSA in 472 morphea patients and found that 6% of patients with morphea had LSA, with 19 extragenital and 8 genital cases [6].

These authors concluded that LSA and morphea may share a common pathogenetic pathway. Although the etiology of both conditions remains unknown, associations have been found with certain HLA alleles, environmental exposures, infections (such as *Borrelia burgdorferi*), autoimmune disease, and trauma [7, 8]. In this case, the patient did not have a history consistent with these causes; however, further investigation into family history and HLA alleles would be necessary to confirm. Additionally, although these studies examined the presence of LSA and morphea in the same patient, they did not assess the presence of LSA-morphea in the same lesion, which may provide further insight into their coexistence and is a unique contribution of our case to the literature. They also did not assess the temporal association between genital LSA, extragenital LSA, and morphea.

Both LSA and morphea have few effective treatments including topical or intralesional corticosteroids, topical calcineurin inhibitors, phototherapy, and systemic therapies such as methotrexate, retinoids, and cyclosporine. There are few studied treatments for extragenital LSA, as most investigations have focused on genital LSA. Further research into treatment of co-existing extragenital LSA and morphea is needed.

Conclusion

Overall, we present a rare case of extragenital LSA-morphea preceding genital LSA. Initial extragenital LSA in the inguinal folds with an unremarkable initial genital exam, with later development of genital LSA is unique. Although there is great debate over classification of LSA and morphea, they ultimately have similar presentations, etiologies, and treatments. Complete examination, including inspection of genital mucosa, is prudent in patients with a diagnosis of extragenital LSA-morphea, as they may subsequently develop genital LSA which is a known risk factor for squamous cell carcinoma [9].

Potential conflicts of interest

Larisa Geskin has served as an investigator for and/or received research support from Helsinn Group, Johnson & Johnson, Mallinckrodt, Kyowa Kirin, Soligenix, Innate, Merck, BMS, and Stratpharma; on the speakers' bureau for Helsinn Group and Johnson & Johnson; and on the scientific advisory board for Helsinn Group, Johnson & Johnson, Mallinckrodt, Sanofi, Regeneron, and Kyowa Kirin. The remaining authors declare no relevant financial interests.

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