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Case Report

Lichen sclerosus et atrophicus and morphea occurring over different sites in a young male: an interesting association

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ABSTRACT

Lichen sclerosus et atrophicus (LSA) is a chronic inflammatory disorder primarily affecting the female genitalia, presenting as white, atrophic skin plaques; extra-genital LSA is less common, occurring in about 15% of cases. Morphea, or localized scleroderma, is a rare connective tissue disease that typically presents with sclerotic skin plaques. We report a 15-year male patient with dark coloured itchy lesions over lower back since childhood and left knee since 6 months. Various investigations, including hemogram, serum biochemistry, thyroid profile, lipid profile, viral markers, and histopathological examination, were conducted. Patient was treated with potent topical corticosteroid, antihistamines and methotrexate 10 mg weekly once for 6 weeks with moderate improvement. Common immune-pathogenic mechanisms and trigger factors such as genetic predisposition and trauma may be responsible for development of 2 different immune mediated dermatoses in the same patient.

Keywords: Lichen sclerosis et atrophicus, Morphoea

INTRODUCTION

Lichen sclerosus et atrophicus (LSA) is a chronic inflammatory disorder primarily affecting the female genitalia, presenting as white, atrophic skin plaques; extra-genital LSA is less common, occurring in about 15% of cases.[1] Morphea or localized scleroderma, is a rare connective tissue disease that typically presents with sclerotic skin plaques. The coexistence of morphea and LSA in the same patient suggests that these lesions represent a spectrum which may reflect similar etiologic events or closely related pathologic processes in these two diseases.[2]

Although an autoimmune hypothesis has been proposed for LSA and morphea, the pathogenesis of LSA is still under discussion. We report a young male who was diagnosed to have LSA and morphea concomitantly at different body sites.

CASE REPORT

A 15-year male patient presented with dark coloured itchy lesions over lower back since childhood. It started as a dark papule which increased in size over the years to form a plaque with wrinkled texture. The lesion was not associated with any itching, pain or erosions. He also developed thick black lesions over knees since 6 months. It started as an elevated lesion with mild itching over left knee later involving the right knee. No history of trauma, vaccination or insect bites was elicited. There was no history of atopy, bronchial asthma or tuberculosis. No significant family and personal history was present. General examination was done. The patient was moderately built and nourished. There was no pallor, icterus, cyanosis, clubbing, lymphadenopathy and pedal edema. On thorough systemic examination, cardiovascular, central nervous, gastro-intestinal and respiratory systems were normal.

On cutaneous examination, two well marginated closely arranged shiny atrophic plaques with dyspigmentation and prominent follicular openings were seen over left paravertebral region (Figures 1a and b). On the left knee, a well-marginated hyperpigmented scaly indurated atrophic plaque was seen (Figure 2). Similar smaller lesions were present over surrounding skin and right knee. Involvement of genital skin was absent. Based on clinical features, differential diagnoses considered for trunk lesion were - LSA, discoid lupus erythematosus and atrophoderma; differentials considered for knee lesion were morphea, discoid eczema and follicular LP.

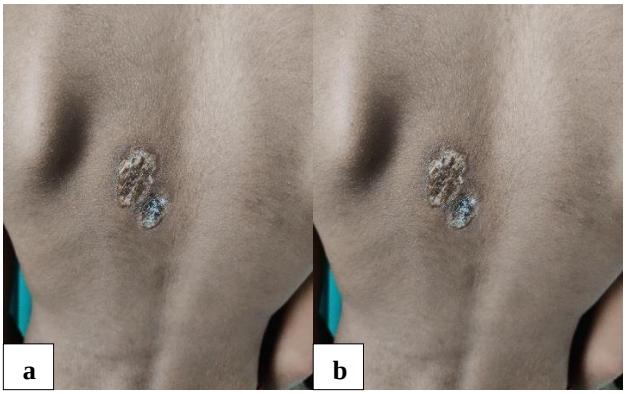

Figure 1 (a and b): Two well marginated closely arranged shiny atrophic plaques with dyspigmentation and prominent follicular openings were seen over left paravertebral region.

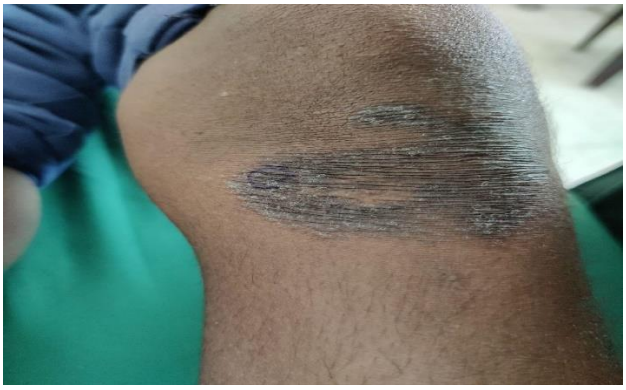

Figure 2: Well-marginated hyperpigmented scaly indurated atrophic plaque over left knee.

The patient was thoroughly evaluated. Complete hemogram, serum biochemistry, viral screening, X-ray, ultrasound abdomen, and electrocardiograph were found to be within normal limits. A 4 mm punch biopsy from edge of the lesion from the back was done. Histopathology showed atrophic epidermis, sub-epidermal band of sclerosis and dense inflammatory infiltrate in mid and lower dermis suggestive of LSA (Figure 3). A 4 mm punch from the knee lesion was also taken. Histopathology from the knee lesion revealed thin epidermis, closely placed collagen bundles in dermis and loss of adnexal structures suggestive of morphea (Figure 4). Based on clinical features and histopathological examination (HPE) findings, back lesion was diagnosed as LSA whereas knee lesions were diagnosed as morphea. Patient was treated with potent topical corticosteroid, antihistamines and methotrexate 10 mg weekly once for 12 weeks along with daily folic acid 5 mg resulting in satisfactory improvement in skin lesions over lower extremity. No significant improvement was noticed over lesions on the back. No adverse effects were seen with the prescribed drugs.

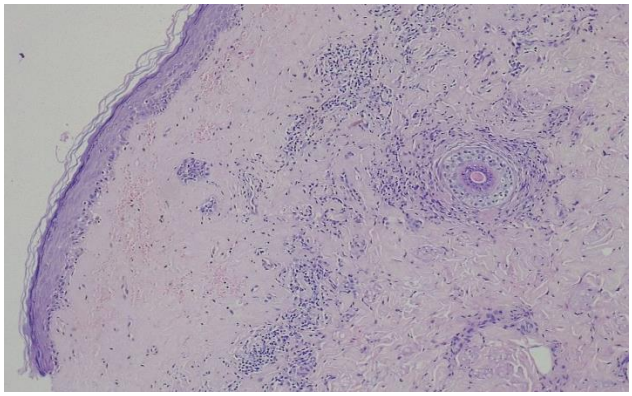

Figure 3: Atrophic epidermis, sub-epidermal band of sclerosis and dense inflammatory infiltrate in mid and lower dermis suggestive of LSA (H&E 10X).

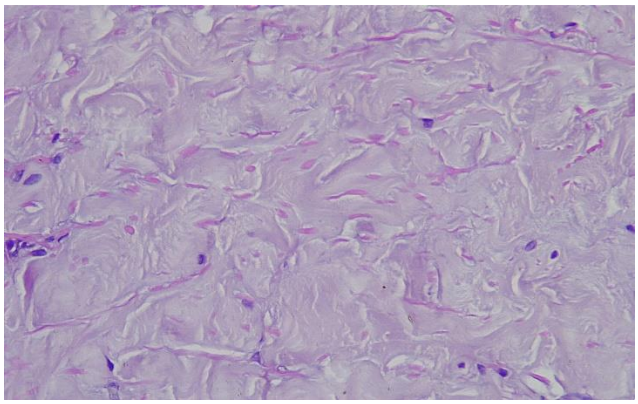

Figure 4: Closely placed collagen bundles in dermis and loss of adnexal structures suggestive of Morphea (H&E 40X).

DISCUSSION

LSc is a chronic inflammatory scarring dermatosis with an ano-genital predilection. Genital LSc (GLSc) is more common than extragenital or oral disease, but there may (rarely) be concomitant involvement of these sites. In adults, ano-genital LSc is said to be about 10 times more common in women than men.[3] Anogenital lichen sclerosus may present with pruritus, pain, dysuria or dyspareunia, but extragenital lesions are usually asymptomatic. Extragenital LSc presents with ivory white papules or plaques often with follicular delling. The disruption of ECM-1-mediated control of MMP-9 activity may be important in pathogenesis.[4] In the present case,

hypopigmented atrophic plaques with prominent follicular openings seen over back which were clinically in favour of LSc.

Morphoea is a group of diseases characterized by inflammation and increased collagen deposition, primarily affecting the skin and subcutaneous tissues.[5] It can involve the fat, fascia, muscle, bone, joints, and occasionally the mouth, eye, and brain. Extracutaneous manifestations, including musculoskeletal, neurological, ocular, oral, and dental complications, are observed in up to 75% of cases. It can be classified as limited, generalised plaque, pansclerotic, linear and mixed types. Limited morphea is further classified as circumscribed plaque, guttate and atrophoderma of Passini and Pierni. Triggers include borrelia, hepatitis C, varicella, herpes zoster; vaccinations (MMR, BCG, hepatitis B); radiation; and drugs.[6] In the present case, multiple plaques were present over knee region with atrophic surface which clinically suggested morphea. However, no history of any of the common trigger factors could be elicited.

Review of literature indicates that 5.7% of morphea cases may have concomitant LSA; association of both extra-genital and genital LSA with morphea, and co-localization of LSA like skin changes over morphea plaques is also reported.[7] Upregulated miR-155, TNF and IL-6 may represent further overlaps in pathogenesis of lichen sclerosus and morphoea enhancing Th1 immune response and regulating fibrotic tissue formation.[8]

Common treatments used in lichen sclerosus are topical clobetasol propionate 0.05%, tacrolimus or pimecrolimus, Ciclosporin and UVA.[9] The mechanism explaining action of potent topical corticosteroid is anti-inflammatory (suppression of pro-inflammatory cytokines**,** such as IL-1, IL-6, and TNF-α) and anti-fibrotic effects (inhibition of TGF- β) which prevents progression to more severe disease. Common treatments used in morphea are tacrolimus ointment 0.1%, imiquimod, topical/ intralesional corticosteroids, calcipotriol 0.005%, UVA-1, broad-band UVA, and narrowband UVB.[10] The mechanism explaining action of UVA1 is anti-fibrotic and anti-inflammatory effects.

In the present case, in view of limited extent of skin lesions, conservative treatment was administered. LSA was treated with topical corticosteroids and morphea was treated with MTX 10 mg weekly once which resulted in limited progression.

CONCLUSION

Our case highlights association of morphea with extra-genital LSA which is less commonly seen. Common immune-pathogenic mechanisms and trigger factors such as genetic predisposition and trauma may be responsible for development of 2 different immune mediated dermatoses in the same patient. In cases which present with overlapping clinical features of LSA and morphea, meticulous clinical examination and histological demonstration of typical changes is paramount to achieve diagnostic accuracy.

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