Dermatology Practical & Conceptual

Extragenital Lichen Sclerosus et Atrophicus-Morphea Overlap Masquerading as Lupus Vulgaris: Histopathology to the Rescue

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Introduction

Lichen sclerosus et atrophicus (LSA) presents as atrophic plaques in genital region usually but can present at extragenital locations such as neck, shoulder, trunk and proximal extremities [1,2]. Morphea is characterized by skin thickening and increased collagen within the indurated lesion [3]. Although both these pathologies have been described as distinct clinical and histopathological entities, there are cases and retrospective studies which suggest that both the diseases could be a part of the spectrum of localized sclerosing disorders [1,2].

Case Presentation

A 56-year-old male presented with an asymptomatic reddish plaque over neck, gradually increasing in size for 8 months. He gave history of taking anti-tuberculous drugs for inguinal tuberculous lymphadenitis twenty years ago. Physical examination revealed an erythematous plaque with central atrophy and purpuric borders, approximately 4 x 3 cm in size, with an advancing elevated edge and an atrophic regressing edge (Figure 1A) with no regional lymphadenopathy. Mantoux skin test was positive (15 mm). However, interferon gamma release assay (IGRA) and chest X-ray were normal. Dermoscopy revealed follicular plugging, white structureless areas, yellowish to orange globules, network of linear vessels within white streaks along with few unfocussed violaceus vessels (Figure 1, B and C)]. A provisional diagnosis of plaque type lupus vulgaris (LV) was given and a skin biopsy was done for histopathology and CBNAAT (Cartridge based Nucleic Acid Amplification Test). Histopathology from the advancing edge of the plaque revealed follicular plugging, pan dermal sclerosis with thickening and homogenization of dermal collagen with replacement of subcutaneous fat below eccrine apparatus by thickened collagen. There was sparse interstitial lymphocytic infiltrate between the collagen fibres (Figure 1, D-F). Ziehl Neelson staining of

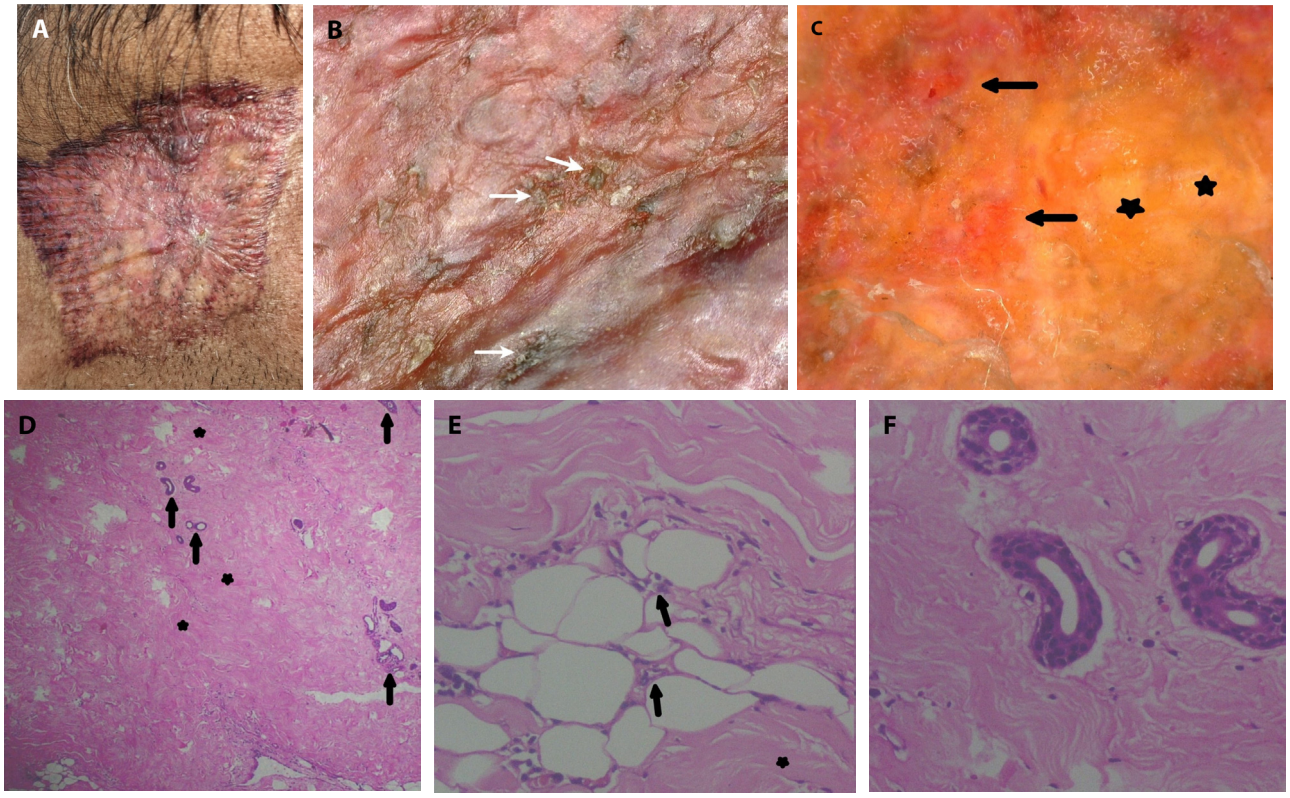

Figure 1. Cutaneous examination and dermoscopic and histopathological findings (H&E stain). (A) Erythematous plaque with central orange yellow hue, advancing edge at one border and regressing atrophic edge at the other end. (B) Dermoscopy showing follicular plugs (black arrow) with yellowish brown crust, 100x without polarization. (C) Yellowish orange globules (black star) with few unfocused vessels (black arrow) at 100x magnification with polarization. (D) Pandermal sclerosis with thickened collagen (black star) and scarce appendages pulled up in the mid dermis (arrow), 2x magnification, H&E stain. (E) Subcutaneous fat below eccrine apparatus replaced by thickened collagen (black star) with lymphocytic interstitial infiltrate (black arrow) 40x magnification, H&E stain. (F) Pulled-up eccrine ducts, 100x magnification, H&E stain.

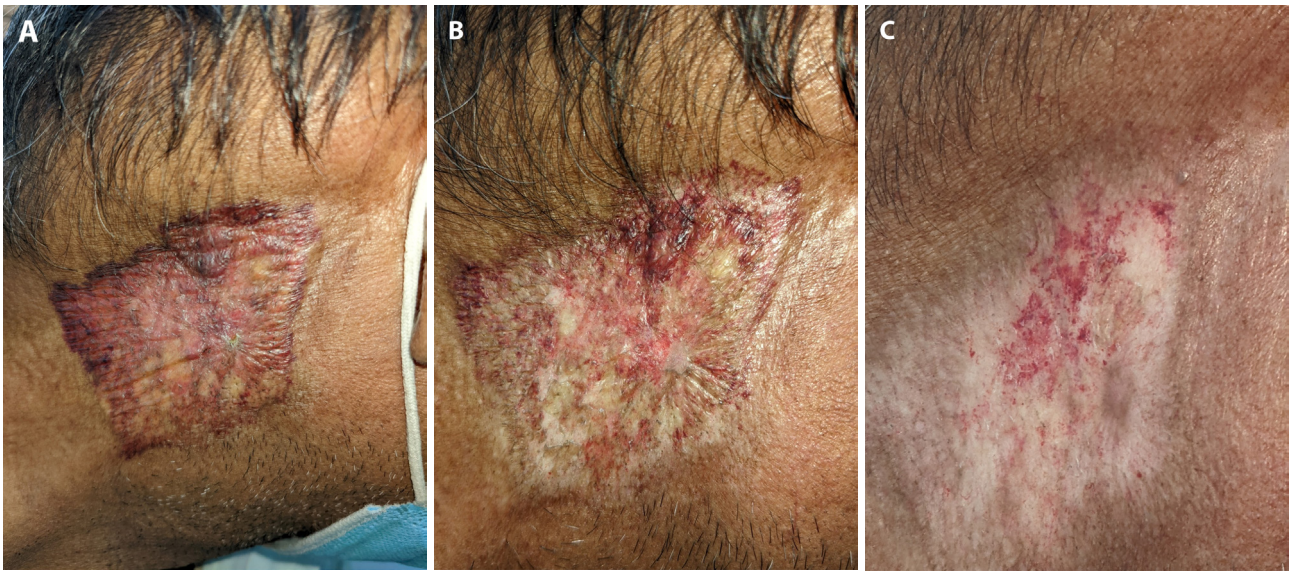

Figure 2. Results after topical calcitriol 0.0003% ointment application. (A) Pretreatment. (B) After 2 weeks of treatment. (C) After 4 weeks of treatment.

histopathological section was negative for acid fast bacilli. Skin biopsy CBNAAT was negative for mycobacterium tuberculosis. A final diagnosis extragenital LSA-morphea overlap was given and the patient was started on calcitriol 0.0003% ointment. There was decrease in the size of plaque within 4 weeks of commencing treatment (Figure 2, A-C).

Conclusions

LSA presents with white atrophic plaques and papules, most commonly on the vulvar and perianal area but may be seen anywhere, with trunk and extremities being the most common extragenital sites.

Dermoscopy of LSA shows porcelain white structureless areas, comedo like openings and a network of dotted blood vessels [1] .On histopathology, LSA presents with follicular plugging, lichenoid papillary dermal infiltrate and homogenization of papillary dermis whereas morphea presents with increased basal layer melanization, reticular dermal sclerosis, thickened collagen bundles, interstitial lymphocytic infiltrate and decreased appendages [1].

LV on dermoscopy presents with follicular plugging, hemorrhagic crusts, yellowish orange background suggestive of granuloma, linear vessels arranged along the white streaks, and white structureless areas. On histopathology, tuberculoid granulomas in reticular dermis with absent or scanty caseation are seen along with hyperkeratosis, atrophy and fibrosis.

Findings in support of LV included past history of TB lymphadenitis with positive Mantoux test, classical morphology of the lesion with an advancing edge, patulous follicles, orange red background with white streaks and structureless areas

However, the presence of pan dermal sclerosis, diminished appendages, replacement of subcutaneous fat beneath eccrine glands by thickened dermal collagen were against LV.

This case posed a diagnostic challenge due to its disguise as cutaneous tuberculosis which emphasizes on the importance of clinico-dermoscopic-histopathological correlation.

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