A rare presentation of verrucous/hypertrophic lupus erythematosus: A variant of cutaneous LE

Sir,

Cutaneous lupus erythematosus (LE) is seen commonly as classical discoid lesion (DLE) or papulosquamous/annular lesions of subacute cutaneous LE.[1] Hypertrophic or verrucous variant of chronic cutaneous LE is a rare clinical variety that may be misdiagnosed for various other closely mimicking conditions, e.g. hypertrophic lichen planus or keratoacanthoma.[2]

A 32-year-old male patient presented with complaints of small, round, well-defined, non itchy, whitish, scaly skin lesions on the right ear [Figure 1] and lower lip accompanied with thick, vegetative, whitish skin lesions over both elbow and forearm since 1 year [Figure 2]. The lesions were slowly progressive and were not associated with local or systemic complaints. There was no involvement of other body parts. Examination revealed single, round, well-defined, atrophic scaly plaque of about 1-cm diameter with hyperpigmented border on helix of the right ear and similar lesion on the lower lip. Large, hypertrophic, verrucous, whitish plaques of irregular dimensions (approximately 5 × 5 cm) were present on both elbows (three on right and two on left). Routine serum biochemistry along with urinalysis was within normal reference range. Antibody to native double-stranded DNA was found to be within the normal range (14.2 U; normal range 0-30 U).

Radiography of both elbow joints were done to rule out bony origin and secondary calcification, which came out to be normal. Histopathology from both lesions at ear and elbow revealed classical epidermal and dermal changes of interface dermatitis seen in DLE, i.e. follicular plugging and liquefactive degeneration of basal cell layer with perivascular and periadnexal lymphocytic infiltrate [Figures 3-c]. Special stains performed did not show infection either with mycobacterium or fungi on Fite and Periodic acid Schiff stains, respectively.

On clinicopathological correlation, patient was diagnosed as having hypertrophic LE.

The patient is being treated with tab hydroxychloroquine 200 mg twice daily for 5 months and tab Acitretin 25 mg once at night time for the last 3 months and is showing regression of verrucous lesions and symptomatic relief on monthly follow-up [Figure 4].

Hypertrophic LE or verrucous DLE was first described by Behçet's in 1940 as lupus erythematosus hypertrophicus et profundus.[3] This entity is characterized clinically by hypertrophic verrucous or vegetative plaques with indurated borders and minimal scaling frequently reported on extensors of forearms, face, and upper part of trunk. In contrast, classical lesions of DLE present with well-circumscribed plaques having atrophy, follicular plugging, and adherent scale.[2]

Systemic involvement may occur in 5-10% cases of DLE with lesions confined to head and neck. This proportion may be higher in cases with widespread cutaneous lesions involving trunk and extremities.[4] On the contrary, cases with long-standing systemic lupus erythematosus developing erythematous, indurated, hypertrophic LE lesions have also been reported.[5] No such involvement was seen in our case.

In a retrospective study of 14 cases of hypertrophic DLE carried out by Daldon *et al*.,[6] all hypertrophic lesions occurred on sun-exposed sites and were accompanied in all patients with classic discoid lesions. Nine cases had involvement of face. Our patient also showed similar involvement. The patient is being followed up for development of squamous cell carcinoma.[7,8]

Histopathologically, our case showed all classical features of interface dermatitis including follicular plugging, liquefactive

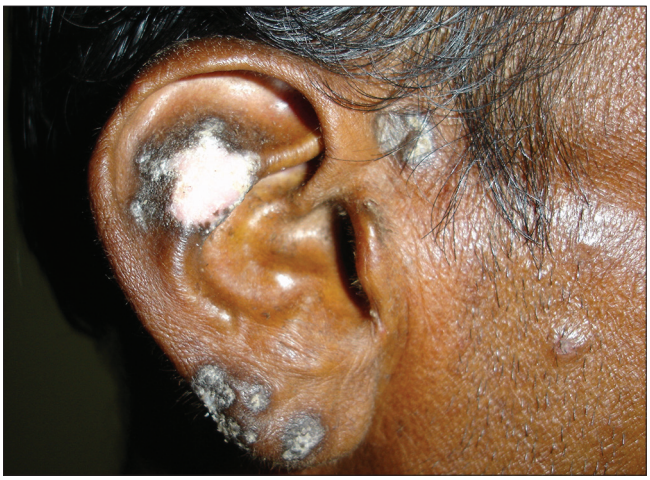

Figure 1: Classical DLE lesion on right ear

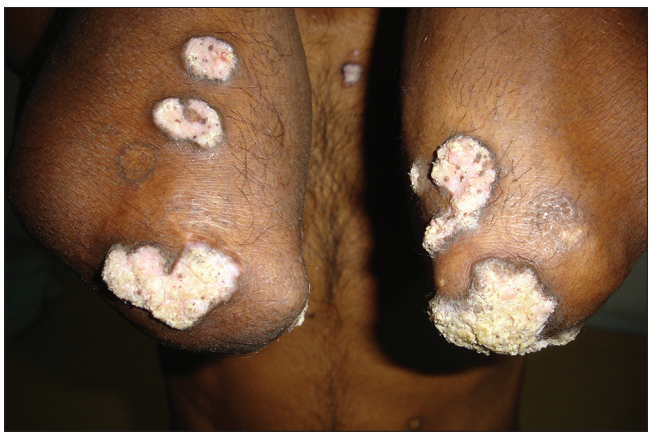

Figure 2: DLE lesions on both elbows

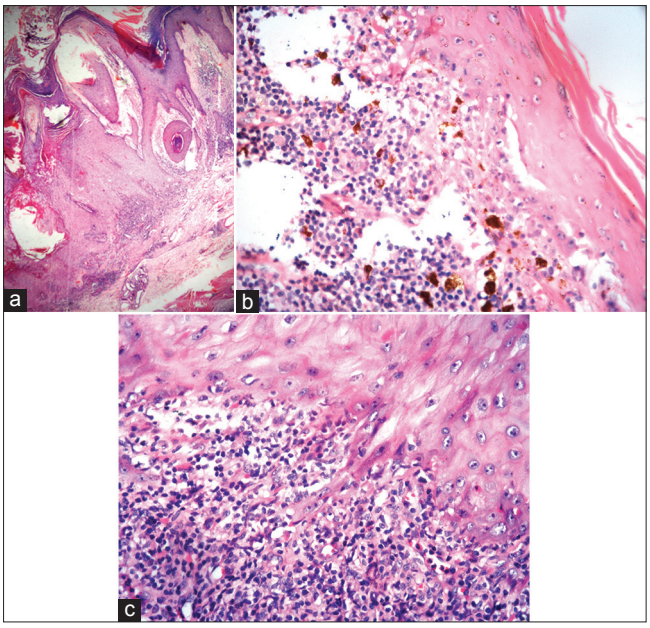

Figure 3: (a) Histopathology of verrucous lesion on lesion showing irregular epidermal hyperplasia with hyperkeratosis, acanthosis, and band-like infiltrate at dermoepidermal interface. (b) Histopathology of elbow lesion (high-power view) showing marked epidermal hyperplasia, follicular plugging, and basal cell degeneration. (c) Histopathology of elbow lesion (×100) showing dermoepidermal interface showing basal cell degeneration and band-like lymphohistiocytic infiltrate with pigmentary incontinence

degeneration of the basal layer, and perivascular lymphocytic infiltrate Biopsy specimen from the verrucous plaque (elbow lesion) showed marked epidermal hyperplasia in contrast to classical histology of DLE, which shows epidermal atrophy.

Apart from hydroxychloroquine, intralesional triamcinolone,[2] isotretinoin,[9] thalidomide,[10,11] and acitretin[12] are other options available for treatment of hypertrophic LE.

The importance of this case is diagnostic dilemma posed by this rare clinical entity, which may be easily missed and misdiagnosed as hypertrophic lichen planus or tuberous xanthomas. This case is being reported to create awareness about the probable clinical presentations of the rare variants of cutaneous lupus and to account them into consideration while approaching clinical diagnosis. Such discrepancies can be minimized by clinician's vigilance and appropriate histopathological correlation.

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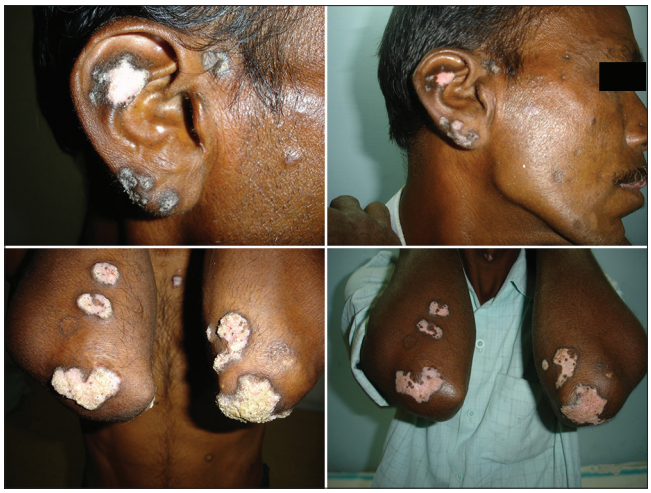

Figure 4: Comparison between pretreatment and post-treatment ear and elbow lesions after 6 months of treatment

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