INTRODUCTION
Cutaneous ulcers are challenging with respect to diagnosis and treatment because of heterogeneous etiopathogenesis. Cutaneous ulcers due to chronic cutaneous lupus erythematosus (CCLE) and lichen planus (LP) are infrequent.[1]

CCLE has many subtypes, mainly classic discoid lupus erythematosus (DLE), lupus hypertrophicus, lupus profundus, and chilblain lupus. Superimposed ulcerations can develop over cutaneous lesions of CCLE, particularly, chilblain lupus which may develop small overlying erosions and lupus profundus[2] (ulceration described in 28% cases of lupus profundus in one series).[3] However, ulcer as the only or predominant presentation of CCLE is rare.

Cutaneous ulcerated LP is an unusual variant, predominantly, involving plantar surface, usually associated with anonychia.[4] Ulcerated cutaneous LP of non-acral sites is even rarer.

CASE REPORT
Clinical details of all the cases are summarized in Table 1.

Ulcerted lupus erythematosus cases
Three cases of ulcerated CCLE included two women and one man, aged 22–41 years. Duration of illness ranged from 3 months to 10 years. All three patients presented with skin ulcers. Ulcers numbering two to multiple (>5), involved scalp, arms, abdomen, buttock, thigh, and leg (Figures 1a, b and 2a, d). Additional findings noted on cutaneous examination were macular...
pigmentation on the cheeks in Case 1 [Figure 1c] and atrophic scars in Case 3.

In all three cases, broad clinical differential diagnoses, particularly, mycobacterial and deep fungal infections were considered. CCLE was included in the clinical differential diagnosis in two cases.

Skin biopsies (number of biopsies: 1–3/patient) were performed in all three cases. Case 1 presented to us for a second opinion with a histopathological diagnosis of squamous cell carcinoma, reported elsewhere, which on review, was interpreted as pseudoepitheliomatosus hyperplasia. Subsequently, two more biopsies were performed, the one from the center of ulcer was non-contributory, and the one from the edge was diagnostic of lupus erythematosus (LE) [Figure 3]. The diagnosis was confirmed by a positive lupus band of immunoglobulin M and C3 on direct immunofluorescence (DIF) study and raised antinuclear antibodies (ANA) (133, normal level <20) and dsDNA levels (64.13, normal <35). In Case 2, the first biopsy was initially misinterpreted as hypertrophic LP. In view of patient developing typical DLE plaques over a period

### Table 1: Clinical details of cutaneous ulcerated chronic cutaneous lupus erythematosus and lichen planus cases

| Case | Age in years/sex | Site of ulcer | Number of lesions | Clinical description of ulcer | Nonulcerated lesions of DLE/LP | Clinical differential diagnosis |
|------|------------------|---------------|-------------------|-----------------------------|--------------------------------|--------------------------------|
| CCLE cases |
| Case 1 | 41/female | Buttock, leg, thigh, scalp, abdomen | Multiple | Superficial, irregularly shaped ulcers with hyperpigmented margins | Macular pigmentation on cheek | Atypical mycobacterial infection, DLE, leishmaniasis, PG |
| Case 2 | 32/female | Right leg, right arm | 2 | Hyperkeratotic plaque with central ulceration and hyperpigmented margins | Subsequent plaque DLE on elbow, knee, and lip | Lupus vulgaris, deep fungal infection, LSC, hypertrophic LP |
| Case 3 | 22/male | Back | 3-4 | 3-4 atrophic scars of size 2 cm×1-2 cm×3 cm, with central ulceration, violaceous margins, and mild surrounding erythema | None | Cutaneous tuberculosis, herpes zoster, DLE, sarcoidosis, deep fungal infection, paraspinal abscess |

| LP cases |
| Case 4 | 18/female | Left knee | 1 | Painful and itchy, 3 cm×3 cm ulcer with fleshy center and brown crusted margins | None | Lupus vulgaris, sarcoidosis, squamous cell carcinoma |
| Case 5 | 14/male | Shins | Multiple | Eroded papules | Violaceous plaque on knee, papules on legs, ankle and arms | LP, psoriasis, LSC, PG, fictitious dermatitis |
| Case 6 | 41/female | Right shin | 1 | Ulcerated plaque hyperpigmented edges | Multiple hyperpigmented, itchy papules and nodules over both legs below knee for over 16 years | LP, prurigo, vasculitis |

CCLE: Chronic cutaneous lupus erythematosus, DLE: Discoid lupus erythematosus, LP: Lichen planus, LSC: Lichen simplex chronicus, PG: Pyoderma gangrenosum

**Figure 1:** Case 1: (a and b) Irregular ulcers with violaceous borders on leg and buttock; (c) macular pigmentation on cheek; (d and e) after 6 months of treatment, scars of healed ulcers with pigmented rims, on leg and buttock
of 1 year and raised ANA levels, the second biopsy and review of the initial biopsy were done, and diagnosis was revised to LE [Figure 2b and c]. A diagnosis of LE was given in Case 3 based on the histopathological findings [Figure 2e and f] but, the patient was lost to follow-up, and no further confirmatory tests could be done.

Treatment details were available only for Case 1; the patient was initially started on hydroxychloroquine with oral and topical steroids, and later on, she was managed on azathioprine. Her lesions improved significantly over a period of few months, ulcers healed and were replaced by scars with hyperpigmented rim [Figure 1d and e].

**Ulcerated lichen planus cases**
Three cases of ulcerated LP included two women and one man in 18–41 years age group. Case 4 had a single lesion, a large ulcer on the shin, for 4 years [Figure 4a]. The ulcer healed leaving a thin central crust, after 3 months of oral and topical steroid therapy [Figure 4b]. Case 5, an adolescent, presented with 4–5 months history of multiple papules and plaques of LP showing koebnerization on the arms, legs, and ankles. Few ulcerated papules, present on the legs, were suspected to be secondary to mechanical/chemical manipulation [Figure 5a]. Skin lesions responded to oral hydroxyzine and topical steroid after 1 month of therapy. Case 6 had multiple papules and nodules on both legs for over 16 years, and one of the lesions on the shin had insidiously progressed to a large ulcer [Figure 5c]. The patient was started on oral Cyclosporine, and the ulcer healed over a period of few months.

In all three cases, varied clinical diagnoses were raised. In Cases 5 and 6, LP was considered as the most likely diagnosis because of the presence of accompanying typical LP lesions, whereas, in Case 4, LP was not suspected clinically.

In all three cases, two biopsies were performed one from the ulcer center and the other from either the ulcer edge (Cases 4 and 6) or non-ulcerated papular lesion (Cases 5). In all cases, biopsies from edge/non-ulcerated lesion yielded the diagnostic features of LP [Figures 4c, d and 5b, d] while biopsies from ulcer
center were non-diagnostic. DIF study was done in one patient (Case 4) which was negative for lupus band.

Clinical comparison of ulcerated chronic cutaneous lupus erythematosus and lichen planus

The mean age of CCLE patients was 31.6 years which was higher than the mean age of 24 years for LP patients. There was no difference in sex distribution of the cases in both categories. Interestingly, the site of ulceration in all three cases of LP was anterior leg whereas, in LE, the distribution of ulcer was more widespread. At initial presentation, none of the LE cases showed typical lesions of CCLE while two out of three cases of LP had typical lesions in addition to the ulcer. Morphology of cutaneous ulcer was similar in both groups except Case 5, who had eroded papules of LP.

Histopathological comparison of ulcerated chronic cutaneous lupus erythematosus and lichen planus

The comparative features are summarized in Table 2. Interface pathology was observed in both ulcerated LP and CCLE. In all but one case (Case 2 of CCLE), there was a dense lichenoid infiltrate in the papillary dermis. Similarly, capillary proliferation and mild papillary dermal edema were focally seen in both groups where biopsy site was the leg. Therefore, this finding is likely to be secondary to ulceration and/or site. Additional features such as plasma cells, mild deep dermal infiltrate, and mild basement membrane thickening on periodic acid–Schiff stain were variably seen in both groups.

Table 2: Comparison of histopathological findings in ulcerated lichen planus and chronic cutaneous lupus erythematosus

| Histopathological features                        | LP (total cases 3) | CCLE (total cases 3) |
|--------------------------------------------------|--------------------|----------------------|
| Interface pathology                               | 3                  | 3                    |
| Lichenoid infiltrate                              | 3                  | 2                    |
| Epidermal changes                                 |                    |                      |
| Saw-toothed hyperplasia                           | 3                  | None                 |
| Pseudoepitheliomatous hyperplasia                 | None               | 2                    |
| Atrophy                                           | None               | 1                    |
| Keratinocyte pallor                               | 3                  | None                 |
| Orthokeratotic hyperkeratosis                     | 3                  | None                 |
| Wedge shaped hypergranulosis                      | 3                  | None                 |
| Deep dermal infiltrate                            | 1 (mild)           | 3 (dense in 2, sparse in 1) |
| Increased dermal mucin (alcian blue stain)*       | None               | 2                    |
| Basement membrane thickening (PAS stain)          | 1 (mild)           | 3 (prominent in 2, mild in 1) |
| Capillary proliferation and edema in dermal papillae | 3                  | 2                    |

Table 2 continued...

*Dermal mucin was assessed in reticular dermis away from granulation tissue or scar. CCLE: Chronic cutaneous lupus erythematosus, LP: Lichen planus, PAS: Periodic acid Schiff

Distinctive features seen only in CCLE cases were dense dermal perivascular/periadnexal infiltrate, increased dermal mucin, epidermal atrophy, or marked pseudoepitheliomatous hyperplasia with keratinocyte pallor and prominent basement membrane thickening. Features specific to LP cases were compact orthokeratotic hyperkeratosis, wedge-shaped hypergranulosis, and saw-toothed epidermal hyperplasia. Although the epidermal changes seem to be somewhat different in two diseases, it is difficult to entirely ascribe this feature to the disease per se as these might be related to the duration of the lesion and the site of the biopsy.

The statistical significance of the observed clinical and histopathological differences cannot be assessed because of small number of cases.
DISCUSSION

The clinical diagnosis of LP and CCLE is particularly challenging when cutaneous ulcer is the only manifestation, as exemplified in patients 1–4 of present series. Even in the absence of typical lesions, subtle clues may be observed in some cases. For example, in Case 1, macular pigmentation on the cheek (a feature sometimes seen in Indian patients),[9] and atrophic scars in Case 3 led to clinical suspicion of CCLE. The clinical differential diagnoses for cutaneous ulceration are wide and particularly in the Indian setting, infectious etiologies’ (mycobacterial and fungal) are prime considerations. The diagnosis relies primarily on histopathology and therefore appropriate site and adequate depth of biopsy are crucial for diagnosis. The edge of the ulcer should be biopsied instead of the center which is non-diagnostic.

Histopathologically, LP and CCLE may show overlapping features. In ulcerated lesions, the distinction becomes more difficult because of secondary changes. For instance, dermal mucin, an important clue to LE, is often present in granulation tissue and scars. Thus, mucin should be assessed in interstitial reticular dermis away from these areas. Other findings that are supportive of CCLE, such as deep dermal inflammation, plasma cells, papillary dermal edema, and telangiectasias, may also be seen as reactive changes along the ulcer margins. In dubious cases, clinico-pathological correlation and ancillary investigations, i.e., DIF and serology for (ANA and dsDNA) play an important role in making a definitive diagnosis.

Cutaneous ulcer is a rare morphological variant of CCLE. In a review by Pramatarov on clinical subtypes of CCLE, ulcerated form was not enlisted among more than twenty different subtypes of CCLE.[6] Ulcerated CCLE has rarely been documented in literature. In a study by Bajaj et al. comprising 110 patients of CCLE, ulcerative form constituted 7.3%.[7] A case of ulcerated CCLE involving ear lobe in the absence of other classic features i.e., atrophy or scarring, has been described.[8] Green and Pasternak have reported a case wherein multiple typical DLE lesions were present and one lesion on the hand progressed to hypertrophic plaque with extensive erosion.[9] Rai and Balachandran reported an Indian patient who presented with a large non-healing ulcer on the back along with smaller scaly depressed plaques of DLE over arms, thighs, and macular hyperpigmentation on the zygomatic area.[10] Macular hyperpigmentation as in this case and in one of our patients (Case 1) may be a manifestation of CCLE, particularly, in patients of Indian ethnicity.[5]

Erosive plantar LP was first described by Friedman in 1921.[11] Albeit, many times, it has been termed as erosive palmoplantar LP, the involvement of palms is very rare.[4] The erosive plantar LP is a crippling, cicatricial dermatosis manifesting as hypertrophic and bullous lesions on the plantar surface of foot and dorsum of toes, leading to superficial ulceration and onychia. The disease mainly affects adults and is more common in women than in men. It is usually accompanied by mucosal LP, papulo-squamous LP and cicatricial alopecia.[4,11] Erosive LP is known to be associated with various autoimmune diseases such as diabetes, chronic liver disease, primary biliary cirrhosis, Hashimoto thyroiditis, rheumatoid arthritis, Sjogren’s syndrome, abnormal autoantibody profile and Hepatitis C.[4,11,12]

Ulcerative cutaneous LP at non-acral sites is exceptionally rare. The Internet-based search yielded 15 cases,[13-27] summarized in Table 3. Surprisingly, two cases of severe generalized ulcerative LP, complicated by secondary infections leading to death, have been documented.[16,19] Pathergy was observed in one patient.[18] Similar to one of our cases (Case 5), in a report by Schepis et al.,[23] the lesions were suspicious for factitious dermatitis.

Associated autoimmune diseases and Hepatitis C have been documented in five of the 15 reported cases of ulcerated LP (four Hepatitis C and one Sjogren syndrome).[15,20,22,24,25] In the present series, there was no known history of autoimmune disease or Hepatitis C in any of the patients; however, patients were not specifically, investigated for this.

The various treatment options for ulcerated LP are systemic and topical or intralesional steroids, cyclosporin, oral and topical retinoids, thalidomide, tacrolimus, topical tetracycline, azathioprine, griseofulvin, dapsone, chloroquine, low molecular weight heparin, platelet derived growth factor, ultraviolet A phototherapy, and extracorporeal photochemotherapy.[4,11,12,17,28] Acral ulcerative LP is notoriously recalcitrant to treatment, at times requiring skin grafting.[4] However, in non-acral cutaneous ulcerative LP, as reviewed from the published reports and our experience from this series, the results seem to be promising with satisfactory response in most.

Hypertrophic ulcerated CCLE is managed with oral antimalarials (combining two antimalarials including
quinacrine with hydroxychloroquine or chloroquine), oral retinoids, topical steroids, and thalidomide.\(^9\)

Squamous cell carcinoma (SCC) is a documented risk in both DLE,\(^{28}\) and LP, particularly its hypertrophic and ulcerative variants.\(^{14,29}\) The development of squamous cell carcinoma on a preexisting lesion of DLE and LP might be the cause of ulceration. Therefore, biopsy in ulcerated lesions is of utmost importance. Any suspicious or clinically different looking area should be biopsied to exclude SCC. On the contrary, pseudoepitheliomatous hyperplasia and keratinocytic atypia seen in hypertrophic lesions of DLE may lead to misdiagnosis of SCC on histology,\(^{28}\) as also happened in Case 1 of this series.

**CONCLUSION**

Ulcerated LP and CCLE are uncommon causes of cutaneous ulcers. In the absence of typical skin lesions, LP/CCLE as the underlying cause of cutaneous ulcer may not be suspected clinically. Diagnosis is based on histopathology in conjunction with DIF and antinuclear antibodies.

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**Conflicts of interest**

There are no conflicts of interest.

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**Table 3: Nonacral cutaneous ulcerated lichen planus cases reported in literature**

| References       | Age in year/sex | Site                                                               | Number of lesions | Acral/mucosal ulceration | Nonulcerated LP |
|------------------|-----------------|--------------------------------------------------------------------|-------------------|--------------------------|-----------------|
| Altman and Perry | 55/female       | Anterolateral aspect of lower legs/ankles                         | Multiple          | Oral ulcers              | Reticulate LP of oral mucosa, acute generalized eruption of LP on trunk and extremities None |
| Higgins et al.   | 57/female       | Flexures: Both breasts, abdominal skin fold, inguinal area, natal cleft | Multiple          | None                     | Nail LP         |
| Brudy et al.     | 90/female       | Scalp                                                              | Multiple          | None                     | Cricatricial alopecia None |
| Matsuura et al.  | 67/male         | Generalized Flexures: Both breasts, inguinal region, perianal area, abdominal skin fold | Multiple          | None                     | None |
| Eisman and Orteu | 79/female       |                                                                   | Multiple          | Acral (hands and feet) and mucosal (oral and genital) ulcer | None |
| Henderson et al. | 50/female       | Pretibial area                                                    | 1                 | None                     | Hyperkeratotic lesions on limbs |
| Sheth et al.     | 64/male         |                                                                   | Multiple          | Oral mucosal erosions    | None |
| Neville et al.   | 50/male         | Scrotum                                                           | 1                 | Ulcer on palm and oral mucosal erosions | Nail (fingers and toes) LP, reticulate oral mucosal LP |
| Schepis et al.   | 58/male         | Inguinal region                                                   | 1                 | None                     | Reticulate LP of buccal mucosa, Papulo-plaque lesions on pretibial region and inguinal areas |
| Franchi et al.   | 76/female       | Forearms, legs, back, groin, axillae                              | Multiple          | None                     | Dark brown cutaneous lesions - postinflammatory hyperpigmentation or LPP |
| Schepis et al.   | 56/male         | Back, lower limbs and nostrils                                    | Multiple          | None                     | Nail LP, reticulate oral LP, cutaneous papules |
| Fisher et al.    | 42/male         | Lower legs and thighs                                             | Multiple          | Oral ulcers              | Hyperkeratotic LP on elbow, arms, trunk, and knees, reticulate oral LP |
| Binesh and Parichehr | 51/male | Scalp Bilateral upper limbs-forearm and dorsum of proximal hand | Multiple          | None                     | None |
| Patel et al.     | 45/female       |                                                                   | Multiple          | Oral erosions            | Reticulate oral LP |
| Mansouri et al.  | 77/female       | Shin                                                               | 1                 | None                     | None |

LPP: Lichen planus pigmentosus, LP: Lichen planus
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