

PhotoDermDiagnosis

Erythematous plaque over face

Piyush Kumar*, Ashim Kumar Mondal*, Avijit Mondal*, Niharika Ranjan Lal*, R C Gharami*, Arunasis Maiti**

Department of Dermatology, Medical College, Kolkata. India

Department of Dermatology, Midnapore Medical College, Paschim Midnapore, India

A 25-year-old female presented with asymptomatic erythematous plaque over face for 1 year. The lesion had started as small erythematous papule and gradually increased in size to attain the present status (**Figure 1**). There was no history of insect bite or trauma prior to onset of lesion. There was no history of photosensitivity, fever, joint pains or any other systemic features. Patient had consulted some physician and had been applying steroids intermittently for last 5 months prior to presentation; however, there was not much improvement.

On examination, single erythematous plaque of size 3cm x 3 cm was found over right malar area. The surface was studded with many small papules and margin was hyperpigmented at places. The lesion was indurated; however, one part of lesion (near eye) was characterized by atrophy. There was no enlargement of cervical lymph nodes. Sensations over lesion were intact and no peripheral nerve trunks were thickened. Mucocutaneous and systemic



Figure 1

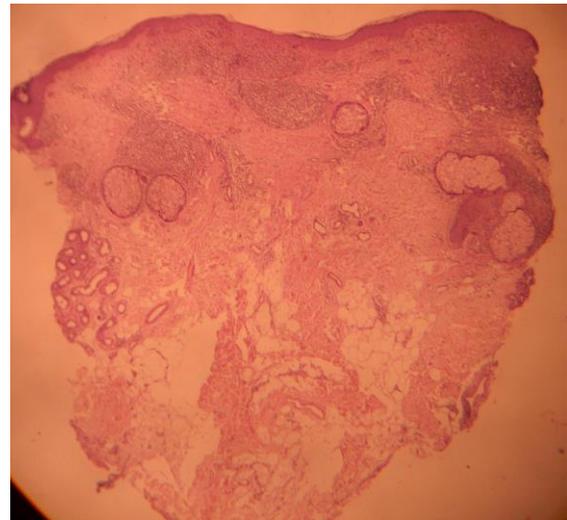


Figure 2

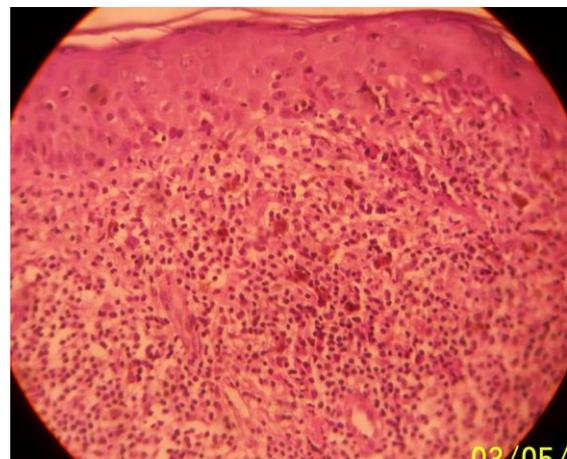


Figure 3

examination did not reveal any other abnormality. Routine blood examination, Mantoux test, and chest X-ray were within normal limits. Slit-skin smear was negative for acid-fast bacilli. Serology for antinuclear antibody was noncontributory. Histopathology sections are shown in **Figures 2** and **3**.

What is the diagnosis?

Diagnosis

Discoid lupus erythematosus (DLE)

Discussion

Clinically, many differential diagnoses can be considered for an erythematous plaque over face. Some of them include Hansen's disease-tuberculoid pole, sarcoidosis, lupus vulgaris, chronic cutaneous lupus erythematosus (tumid LE and LE profundus), granuloma faciale and pseudolymphoma (cutaneous lymphoid hyperplasia or lymphocytoma cutis and Jessner's lymphocytic infiltration of skin). Many malignant conditions like T cell or B cell lymphoma as well as cutaneous metastasis may present in a similar way. Fortunately, many of these conditions have characteristic histopathology. Hence, histopathology from the lesion may be of great help in such a situation. In our patient, histopathology revealed patchy lymphocytic infiltration in dermis (**Figure 3**). Such a finding is seen in conditions classically described as 'five L's'. They are lupus erythematosus, lymphoma, lymphocytoma cutis, polymorphous light eruption (plaque type), and lymphocytic infiltration of the skin of Jessner.¹ In our patient, there was prominent basal layer degeneration (**Figure 3**). This makes the diagnosis of LE most likely, as other conditions among 'five L's' are not associated with basal layer degeneration. Follicular plugging, thinned epidermis, prominent basal layer degeneration and perivascular and periappendageal infiltration were in favour of DLE. However, clinically, lack of adherent scales, atrophy and dyschromia were against the diagnosis of DLE. Clinically, lesion was more of tumid LE than of DLE. On the other hand, from the histopathological point of view, findings were in favour of DLE, rather than tumid LE. This posed a diagnostic dilemma.

The most accepted classification of specific cutaneous manifestation of LE is by Gilliam. He has classified LE specific lesions into three categories (acute, subacute and chronic) based on clinical findings and histopathology. Chronic LE consists of mainly DLE, lupus tumidus/ tumid LE, lupus panniculitis/lupus profundus and chilblain LE among others.²

DLE represents one of the most common skin manifestations of lupus. Discoid lesions are found most often on the face, scalp and ears, but may be present in a widespread distribution. A classical DLE lesion is clinically characterized by well-demarcated, erythematous, slightly infiltrated, discoid plaque with adherent thick scales and follicular plugging.³ The active lesion displays surrounding erythema and the lesion heals with atrophy and dyspigmentation.⁴ Histological hallmarks include hyperkeratosis with follicular plugging of stratum corneum but thinning and flattening of stratum malpighii with hydropic degeneration of basal cells and lymphocytic infiltrate arranged along the dermal-epidermal junction, perivascular and periappendageal structure.¹

Tumid LE was first described by Erich Hoffman in 1909.⁵ However, comprehensive criteria for definition of this condition remained elusive until Alexiades-Armenakas *et al.*⁶ reported a series of 15 tumid LE patients. Clinically, tumid LE presents as indurated papules, plaques, and nodules without follicular plugs, scaling, atrophy, or ulceration of surface.^{2,6} The number of lesions varies from 1-4 and are present over sun-exposed parts, most commonly on face. Photosensitivity is present in majority of cases.⁶ Histologically, there is superficial and deep perivascular, periappendageal lymphocytic infiltrate with abundant mucin deposition. The changes at dermo-epidermal junction and in epidermis are absent or minimal.⁶ These histopathological findings

make tumid LE unique among cutaneous LE, as surface changes (which are so characteristic of LE) are absent or minimal. Identification of mucin deposition can be utilised to differentiate it from other conditions of 'five L's'. In doubtful cases, direct immunofluorescence (DIF) is a very helpful tool. Alexiades-Armenakas *et al.*⁶ reported positive DIF findings in 50% cases. The findings include linear deposition of IgG, IgA, C3 and granular deposition of IgM along basement membrane zone.⁶ One interesting observation in their article was presence of focal epidermal changes in 60% cases of patients with positive DIF findings.⁶ This limits the usefulness of DIF in diagnosis of tumid LE as positive DIF is expected in patients with focal epidermal changes and these changes, when present, themselves are sufficient to differentiate tumid LE from other 'five L's'.

Our patient was diagnosed as DLE based on histopathological findings i.e. follicular plugging, epidermal thinning, prominent basal layer degeneration and periappendageal lymphocytic infiltration. The atypical clinical presentation (lack of adherent scales, central atrophy and dyschromia) might be explained by intermittent application of steroids for 5 months prior to presentation. Moreover, presence of only one lesion is unusual for DLE. The patient had been under our follow up for around 1 year. Still, no other lesion had been observed. As there was only one lesion, we settled on intralesional triamcinolone acetonide (10mg/ml) along with

photoprotection. The intralesional injections were given at an interval of 3 weeks (total of 6 such) and patient has responded favourably to above said treatment regimen.

Acknowledgement

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