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Malar Mimicry: Discoid Lupus Erythematosus And The Butterfly Deception

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Abstract:
Introduction: *Discoid lupus erythematosus (DLE) is a chronic autoimmune skin condition characterized by distinctive, scaly, erythematous plaques, primarily affecting sun-exposed areas. While typically presenting with well-defined lesions, DLE can sometimes mimic other dermatoses, posing diagnostic challenges.*
This case report describes a cutaneous lupus(CLE) exhibiting a butterfly-shaped distribution on the face with a malar rash of systemic lupus erythematosus (SLE) but a scale representing discoid lupus erythematosus leading to a diagnostic dilemma.
Case presentation: *A 22-year-old male presented with discoid lupus erythematosus (DLE) exhibiting an unusual butterfly-shaped rash on the face, mimicking the malar rash commonly associated with systemic lupus erythematosus (SLE). This atypical presentation posed a significant diagnostic challenge, highlighting the importance of considering DLE in the differential diagnosis of facial rashes, even when the clinical picture suggests SLE.*
Discussion: *The case is of particular interest to the readers because it underscores the variable and sometimes deceptive nature of DLE, particularly in younger patients. The case emphasizes the crucial role of histopathological examination and direct immunofluorescence (DIF) in accurately diagnosing DLE and differentiating it from other conditions, such as SLE and the overlaps. Furthermore, it highlights the importance of a high index of suspicion for DLE, even in the absence of systemic symptoms or positive serological markers.*
Key Word: *Butterfly rash, Malar rash, Lupus, Discoid*

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I. Introduction

The autoimmune disease lupus erythematosus (LE) is a fairly common occurrence that manifests in two broad forms: systemic (SLE), which is abrupt in onset and involves several organ systems and, discoid (DLE), which is chronic and confined to the skin. Numerous cellular antigens in blood and tissue are targeted by antibodies, and serum antigen-antibody complexes drive the systemic form of disease (SLE), resulting in multi-organ involvement. DLE involves both humoral and cell-mediated immune response (1,2).

Subacute cutaneous lupus erythematosus (SCLE), is a third common form of the cutaneous lupus erythematosus (CLE), with DLE being the most common subtype, often affecting middle-aged women who exhibit lesions on the skin and the vermillion border of the lip. Classic DLE lesions are characterized by erythematous discoid-raised plaques with an adherent scale, which leads to atrophy and scarring. The disc-shaped lesions expand perpendicularly and heal in the centre with subsequent scarring due to loss of pigment. Mucous membrane involvement, if seen, exhibits white radiating striae often affecting the oral mucosa (2-5).

SLE causes modest skin involvement, butterfly rash, and organ system involvement, and presents as fever, weight loss, and malaise. It then spreads systemically to the joints, liver, kidneys, and lungs. It seldom develops from DLE. However, 20% cases of SLE have shown DLE as a manifestation and in 25% SLE, development of discoid lesions occur during a period of disease course. DLE frequently involves the sun-exposed areas of the face in a scattered lesional pattern (5,6).

This case describes an unusually presenting cutaneous lupus erythematous with a butterfly dispersion of the lesion, highlighting the diagnostic challenges and the significance of thorough clinical evaluation and histological confirmation.

II. Case Report

A male patient, age 22, approached our hospital after experiencing a chronic face rash for six months. There were no similar lesions found on the ears, arms, legs, or back. However, the under-eye region was partly affected by the rash. He did not report any systemic symptoms or any noteworthy medical history. Upon

examination, the patient had a distinct, erythematous rash that was butterfly-shaped and spread across the cheeks and bridge of nose, bypassing the nasolabial folds. The edges of the scaly lesions were somewhat elevated. Other sun-exposed areas showed no signs of well-defined DLE lesions or acute inflammation. There were no oral cavity lesions and no mucosal involvement. SLE, rosacea, seborrheic dermatitis, and contact dermatitis were among the differential diagnoses**.**

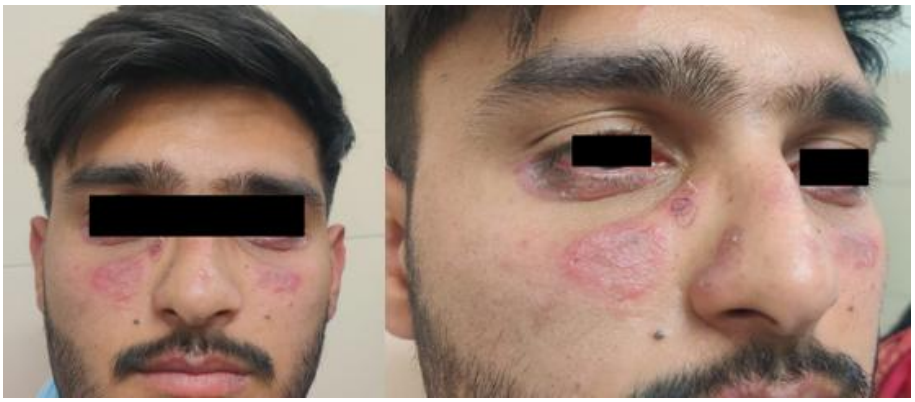

Fig.1 A&B: Patient Exhibiting An Erythematous Rash On Malar, Bridge Of Nose And Ocular Region.

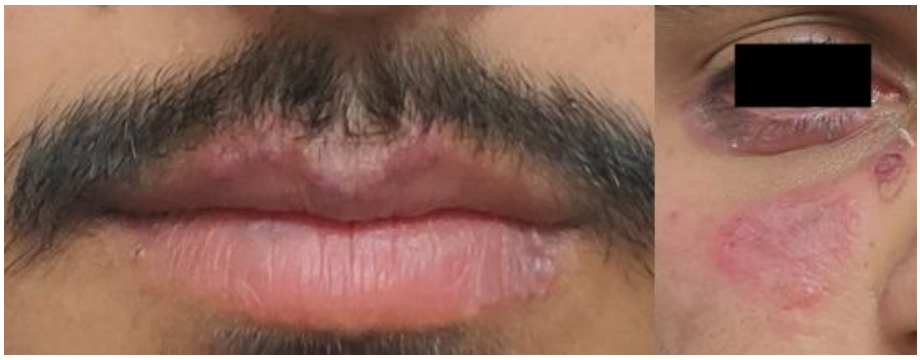

Fig. 2: Disc Shaped Lesions On The Vermillion Border.
Fig. 3: Biopsy Site With Markings.

Investigations:

Complete blood count and other routine blood investigations were within normal limits. A skin biopsy was performed from the affected facial skin. Histopathological examination revealed features consistent with DLE, including interface dermatitis, lymphocytic infiltration around hair follicles and blood vessels, thickening of the basement membrane, and increased dermal mucin.

Direct immunofluorescence (DIF) on the skin biopsy showed a granular deposition of IgG and C3 at the dermo-epidermal junction, further supporting the diagnosis of DLE. Antinuclear antibody (ANA) testing was negative. Lupus band test (LBT) presented complement components and immunoglobulins only on the involved lesional skin whereas normal skin did not exhibit the same.

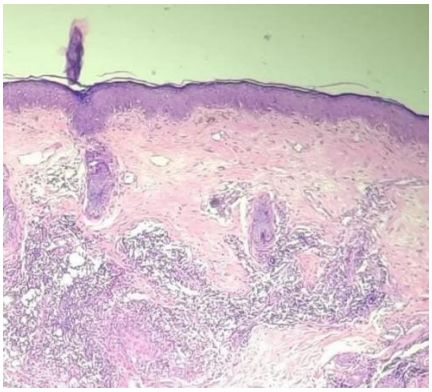

Fig 4. Histological Picture (10X) Showing Dense Inflammatory Infiltrate At Deeper Layer Of Dermis With Follicular Plugging.

Treatment and Outcome:

The patient was managed primarily by strong topical corticosteroids, such as clobetasol propionate. Strict sun protection measures, such as wearing broad-spectrum sunscreen with a high SPF, were also recommended. The erythema and scaling improved somewhat over the course of the following few months. The chronic nature of DLE and the significance of consistent sun protection and routine follow-up were discussed with the patient. The face involvement and possibility for long-term control made hydroxychloroquine a viable second-line treatment.

III. Discussion

Lupus erythematosus (LE) is a multi-organ disorder that predominantly affects the skin and has several subtypes with discoid Lupus erythematosus (DLE) being the most prevalent. DLE further has two main subtypes, localized; with lesions on the face, scalp, and ears, seen in 80% of cases and less often seen is a disseminated form which affects the extensor arms and hands that sometimes progresses to SLE. An overlap can be seen from 1% to 5% cases, where an existing DLE progresses to SLE. Likewise, SLE may progress to DLE in 25% of cases (4,6-8).

This case illustrates a diagnostic challenge posed by DLE mimicking the malar rash of SLE. The butterfly distribution of the rash raised initial concerns for SLE. The patient presented with photosensitivity, which is seen in all types of CLE; the incidence varies from 27-100% in SCLE and 25-90% in DLE (8). However, the absence of systemic symptoms, negative ANA, and the characteristic histopathological findings lay in the favor of DLE. DLE primarily involves the head and neck region, most commonly affecting the scalp and ears, but the present case exhibited a malar distribution. Thus the case highlights the importance of considering DLE in the differential diagnosis of facial rashes, even when the clinical presentation is suggestive of SLE.

Differentiating DLE from SLE with facial involvement can be challenging. Key distinguishing features often include the presence of thicker, scaly lesions in DLE, the potential for scarring, and the absence of systemic manifestations in purely cutaneous DLE. The other possibly close diagnosis could be Subacute lupus erythematosus (SCLE) but was ruled out by their annular and papulosquamous lesional type. However, overlap can occur, and a skin biopsy is often essential for a definitive diagnosis.

The involvement of the superficial dermis with inflammatory infiltrate represents the systemic and subacute forms, whereas DLE has a deeper involvement to the reticular dermis with a denser inflammatory infiltrate and thickened basement membrane with follicular plugging (9).

The other differentials for the present case included, overlapping DLE and SLE; DLE progressing to SLE; SLE later developed DLE (10). These were finally went unsupported by the reports of the histopathological, DIF and lupus band test (LBT).The negative ANA further reinforced that the diagnosis of DLE. Presentation of components of the complement system and immunoglobulins on the lesioned skin, is a feature consistent with DLE.

IV. Conclusion

This case report emphasizes the importance of recognizing the variable clinical presentations of DLE. A butterfly rash distribution on the face can mimic SLE. A high index of suspicion, thorough clinical examination, and histopathological examination are essential for accurate diagnosis and appropriate management. This case also highlights the utility of DIF in confirming the diagnosis of DLE, especially in cases with atypical clinical features. Clinicians should be aware of the diverse manifestations of DLE to avoid misdiagnosis and ensure timely intervention.

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