CASE REPORT

Discoid lupus erythematosus at the vermilion border of the lip: A rare clinical presentation

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Abstract

Discoid lupus erythematosus (DLE) is a chronic disease which affects the dermatome which causes scarring, hair loss, and hyperpigmentation changes in the skin. It has an effect on one’s quality of life, due to its chronic course. Prognosis can be improved if treatment is initiated as early as possible. The condition can be diagnosed clinically, and histopathology may be required to confirm. DLE lesions turning into squamous cell carcinoma has been reported sparsely and due to inadequate data following the reporting of such lesion, this topic deserves attention. Here is an unusual case of DLE which was seen in a young male patient at the vermilion border of the lip.

Keywords:
Cutaneous lupus erythematosus, discoid lupus erythematosus, human lymphocyte antigen, immunoglobulin G, systemic lupus erythematosus

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Introduction

Lupus erythematosus is a condition which is immunologically mediated and is one of the most common “collagen vascular” or connective tissue diseases.[1] The lupus erythematosus is described as having two distinct forms: (1) On the skin or mucosa, chronic discoid type lesions are seen. (2) When almost any organ of the body is involved, it is called the systemic type. Lesions in the oral cavity may be present in each of these two types of lupus erythematosus. Histopathological features discoid lupus erythematosus (DLE) are more specific, but overlapping features may be confused with lichen planus, and a diagnosis is best based on the clinic-histopathologic findings. Oral discoid lesions are seen in 20% of patients. Without the involvement of skin lesions or before the skin lesions develop oral lesions can be seen. Labial mucosa are the most common affected that to on the vermilion border and buccal mucosa. The occurrence of white papules, central erythema, a border zone of irradiating white striae and peripheral telangiectasia are pathognomic.[1,2]

Case Report

A 19-year-old male patient, farmer by profession came with a chief complaint of painful ulcerations on his lower lip since 6 months [Figure 1]. The pain was insidious in onset, localized throbbing to pricking type of pain which is of moderate intensity aggravates on taking food or trauma to that region. Pain preceded ulcers which were initially small in size and gradually progressed to the present size with the incidence of white serous discharge from the ulcers. Bilateral lymph nodes were non palpable. The systemic evaluation was conducted, and no other lesions were evident. The patient had a class three facial profile with a protuberant lower lip. Two solitary ulcers, roughly oval in shape were present on either side of the lip measuring 1 cm x 1 cm in size with regular borders with the thin marginal zone of erythema. There was a presence of radiating striae from the zone of erythema [Figure 2]. Considering the clinical presentation, a provisional diagnosis of mixed red and white lesions involving lower labial mucosa was given. Differential diagnoses considered were DLE, actinic keratosis, and erythema multiforme minor. The lesion was treated with corticosteroid therapy systemically and topically for 21 days [Figure 3]. The patient was asked to cover the mouth with a soft cloth while he is out in the fields. Based on the nature of the lesion, its progress, its clinical presentation and the response to the treatment given reaffirmed our diagnosis of a DLE.
Discussion

The oral lesions of DLE as described by the WHO Collaborating Reference Centre for Oral Precancerous lesions defines it as "circumscribed, slightly elevated, white patches that may be surrounded by a (red) telangiectasia halo. A radiating pattern of thin white lines is seen commonly." DLE is a benign disorder of the skin, the most frequent involving the face, and characterized by well-defined red scaly patches of variable sizes, which heal with atrophy, scarring, and pigmentary changes. DLE can be classified into (1) localized form (2) generalized form.[1,2] Genetic deficiencies of the complement components including C2, C3, C4, and C5 as well as the C1 esterase inhibitor are associated with DLE. Significant increases of human lymphocyte antigen (HLA) B7, B8, DR3, and DQA0102 and a significant decrease in HLA A2 have been reported for patients with DLE and A*03, B*07, DRB1*15 haplotype was associated with DLE. It also occurs with increased frequency in female carriers of X linked chronic granulomatous disease.[3] Females below 40 years with HLAB8 have a high risk of converting to SLE. UV light could uncover an already established autoimmune state by triggering autoantigen release from an injured keratinocyte. Conceivably, UV light could also affect localization of pre-existing autoantibody containing immune complexes by altering endothelial permeability. UV light may additionally have an early critical role in HLA-DR molecules may result in the recognition of self-peptides that may have escaped thymic tolerance.[3-5] UV light may cause an overt release of immunological mediators such as interleukin-1 (IL-1), tumor necrosis factor-α, prostaglandin E, proteases, oxygen free radicals, and histamine in genetically susceptible individuals with LE. The aberrant expression of adhesion molecules such as intercellular adhesion molecule (ICAM-1) could play a role in the pathogenesis of LE photosensitivity. UV light may directly affect immunoregulatory cells such as cutaneous T-cells, which normally help suppress abnormal patterns of cutaneous inflammation.[5] DLE is seen in 20–40 years of age. Peak age of onset is 30 years in females and 40 years in males.[6-10] It is predominantly seen in females in the ratio 3:1 and 3:2 and more severe in blacks. Face, scalp, nose, ears, V area of the neck, and extensor aspect of the arms, any area of the face including the eyebrows, eyelids, nose, and lips can be affected.[6-10] Early lesions start as patches of erythema, occasionally with a urticarial component later they become papulosquamous and eventuate in elevated reddish edematous plaques covered with adherent graying scales. The lesions tend to enlarge peripherally and may coalesce to produce bizarre patterns.[10] Chronic lesions are well defined and circular, oval or irregular in shape. Often they have an elevated erythematous border. The center of the lesion is usually depressed. Dilated follicular openings plugged with horny epithelial plugs are seen. Large areas may be involved. Some of the patches may resolve, but residual scarring is more common. The scars are smooth, atrophic, flat, and white. Telangiectasia may be present at the edges of the scars so called telangiectatic lupus erythematosus is merely the morphological form in which telangiectasia predominates. Post inflammatory hyperpigmentation also may occur. Depigmentation may also be seen. Follicular involvement is a prominent feature. Keratotic plugs accumulate in dilated...
The lesions of the mucous membranes may be limited to these areas, but usually coexist with skin lesions of DLE. The mucous membrane lesions the more common occur during acute systemic episodes of lupus erythematosus. They typically consist of gingivitis, mucosal hemorrhage, erosion, and shallow ulcerations. Early lesions: - mucosal hemorrhage, erosion, superficial erythematous patches with dilated blood vessels on the borders. The center is depressed or superficially ulcerated. The chronic lesions - central atrophic areas with small white dots, surrounded by a keratinized border composed of radiating white striae.[10]

The oral lesions are prevalent in 7-52% of patients with SLE and studies have shown that up to 57% of mucosal lesions were painful and almost 82% of oral ulcers were painless.[11]

Associated features of DLE: Small telangiectasia on the face in 20%, bilateral enlargement of the parotids, livedo reticularis is on the legs, porphyria cutanea tarda, acute intermittent porphyria chronic lymphocytic leukemia, macroglobulinaemia, polychondritis, autoimmune thyroiditis, and carpal tunnel syndrome, PLE.[6]

Laboratory findings such as anemia, leukopenia, thrombocytopenia, raised erythrocyte sedimentation rate (ESR), raised serum globulin levels, elevation of gamma globulin, higher immunoglobulin G (IgG) levels - scarring, autoantibodies, T-cell counts are decreased, urine examination, and blood urea nitrogen has to be done to rule out SLE and to know patient’s renal function. Antinuclear antibodies are rarely present.[4]

DLE has negative response for LE cell inclusion test; usually patients with SLE typically develop LE cells. This cell, or phenomenon, consists of a rosette of neutrophils surrounding a pale nuclear mass apparently derived from a lymphocyte. Only rare occasions is the LE cell found in cases of DLE.[12]

Granular pattern or shaggy pattern of IgG, IgM, IgA, and C3 in the basement membrane or in the dermal epidermal junction positive in active lesions at least 6-week-old granular deposits of IgG (>IgM) at the dermoeidermal junction.[13] This is known as lupus band test (LBT) and is positive in 90% of active lesions not recently treated with topical corticosteroids but negative in burnt out or scarred lesions and in the normal skin both sun exposed and non-exposed. SLE in contrast, has positive LBT in lesional as well as both normal (70-80%) and non-exposed (50%) skin.[14]

The differential diagnosis for oral lesions includes lichen planus and leukoplakia.[14] The classic oral discoid lesion has four outstanding features according to Schiodt which can be differentiated from other lesions they are: [15]

1. A central atrophic area with

2. Small white dots and a slightly elevated border zone of

3. Irradiating white striae and

4. Telangiectasia.

The untreated skin lesions tend to be persistent. Usually heals with scarring. <5% of the cases may convert into SLE, relationship between DLE and SLE 5% of the patients presenting with classic DLE lesions subsequently develop unequivocal evidence of SLE.[6,8,13]

The patients with generalized DLE have somewhat higher risk for progressing to SLE, and a higher risk for developing more severe manifestations of SLE than patients with localized DLE. The following have been suggested as risk factors for the development of SLE than DLE: Diffuse non scarring alopecia, generalized phenomenon, sub-acute, or acute cutaneous lupus erythematous skin lesions. LE non-specific skin lesions such as vasculitis, unexplained anemia, marked leukopenia, false positive tests for syphilis, persistently positive high titer anti-nuclear antibody assay, anti-single stranded DNA antibody, hypergammaglobulinemia, elevated ESR (especially >50 m/h), positive sun protected, non-lesional lupus band test, and elevated levels of soluble IL-2 receptor.[4,4]

Squamous cell and less commonly basal cell carcinomas occasionally occur in scars of DLE, particularly on the scalp, ears, lips, and nose.[16,17]

Excessive exposure to sunlight, UV light, and heat are initiating or aggravating factors as seen in our case. Wear a broad-brimmed hat. Avoid short sleeved shirts and shorts. Sunscreen cream or lotion to applied regularly topical therapy includes: (1) 0.025%, flucinolone cream; (2) 0.1%, betamethasone 17-valerate cream; (3) 0.1%, triamcinolone acetonide cream; (4) intralesional corticosteroid injections - triamcinolone acetonide 5 – 10 mg/ml at 6 weekly interval are helpful in resistant cases and interferon-α.[18,19]

Oral therapy includes:

1. Oral prednisolone 0.5 mg/kg rapidly tapered over 6 weeks
2. Hydroxychloroquine, initially 200 mg twice daily, reducing to 200 mg/day after response
3. Chloroquine sulfate 200 mg twice daily
4. Tab auromycin 6-9 mg/day
5. Isoretinoin 20-80 mg/day
6. Dapsone 100 mg/day
7. Oral thalidomide has been tried for cases not responding to steroids and antimalarial drugs.

When all of the above have failed in patients with the severe and persistent disease, then the following drugs can be given.

1) Pulsed methylprednisolone 500-1000 mg/day for 2-3 days
2) Cyclophosphamide 50-200 mg/day
3) Intravenous pulses of above drugs at 10 mg/kg, at 3-4 weekly intervals.

Antimalarials have many side effects. So, pre-treatment screening has to be done. Hydroxychloroquine is preferred than chloroquine as it has less side effects than the latter. Pre-treatment screening when using antimalarials.[20,21]
Conclusion
Thus, with this overview of DLE and its causes we can attribute our case which has rarer clinical presentation of occurring at a younger age. The lesion on the vermilion border brings us to suspicion of other red and white lesions. Lesions on the lips should be attended promptly as it has the highest level of malignant transformation.

References