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CASE REPORT

ORAL DISCOID LUPUS ERYTHROMATOSUS: A CLINICAL ENIGMA

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ABSTRACT

Lupus erythematosus (LE) is an autoimmune disorder, in which the body's own immune system attacks its own tissues, especially components of the cellular nuclei. The lesions mostly occur in sun-exposed areas like face, neck, ears and upper trunk. The oral discoid lesions are characterized clinically by the presence of white papules, central erythema with a border zone of irradiating white striae and peripheral telengiectasia. It can occur anywhere in the oral cavity. Lupus erythromatosus causes desquamative lesions which are non plaque induced inflammatory gingival lesions. It is not a specific disease entity, but a gingival response associated with a variety of conditions. The most common clinical presentation is an erythromatosus, centrally atrophic plaque with surface telangiectasia and pigmented borders. This paper reports two rare cases of DLE involving the two different sites and was treated with combined medical and surgical therapy with splendid outcome.

INTRODUCTION

Lupus erythematosus is described as having two distinct forms by Bietti 1828 and Kaposi 1872.[1] Discoid lupus erythematosus (DLE) is an autoimmune disease characterized by well-defined inflammatory, scaly plaques on skin. Discoid lupus erythematosus has more specific histopathological features, but initially it can be confused with lichen planus as the lesion comprises of central erythematous mucosa with slightly elevated white border. This border is characterized by fine perpendicular white "paint-brush"-like lines and a diagnosis is best based on the combination of clinical and histopathological findings.[2,3] This paper reports two rare cases of DLE which was treated with combined management.

Case report 1

A 70 year-old, female patient reported to our department with a chief complaint of pain and swelling on lips since 6 months. Extra-oral examination revealed pallor of skin, nail and conjunctiva present with a diffuse swelling present on the lower lip. It is extending from vermillion border of lip till the mucogingival junction with areas of hyper and hypopigmentation present in the borders of the lesion. A multiple diffused white plaque are present on the vermillion border of lip which resembles white radiating lines with central atrophy (Figure 1a). Correlating history and clinical examination, this case was differential diagnosis as Lichen planus, Discoid lupus erythromatosus, and cheilitis granulomatosis of lower lip was made. The patient was subjected for medical evaluation and laboratory tests to rule out any other systemic reasons. The patient hematological examination which revealed, ESR was mildly elevated, Anti-Ro (SS-A) antibodies were negative and other values are within normal limits. The patient was subjected for further investigations, incisional biopsy and immnofluorescence was done. The H and E section showed parakeratinized stratified squamous epithelium overlying the connective tissue. The connective tissue showed dense inflammatory infiltrate beneath the basement membrane chiefly composed of lymphocytes (Figure 1c). Based on the clinical and histopathologic features, lesion was suggestive of discoid lupus erythematosus. The immnofluorescence showed sharply-demarcated, brightly fluorescent apple-green band at basement membrane zone (BMZ) with antisera against IgG and/or IgM along with C3, in a granular band-like or homogenous pattern. A final diagnosis of Discoid lupus erythematosus was made. Patient was advised treatment with the reference of general physician. Triamcinolone acetonide oral paste was advised to apply two to three times per day and was recalled for follow-up. Tab. Wysolone 20mg was given along with topical, once daily for 1 week and tapered the dosage in 1 month and follow up for a period of 1 year without any recurrence (Figure 1b).

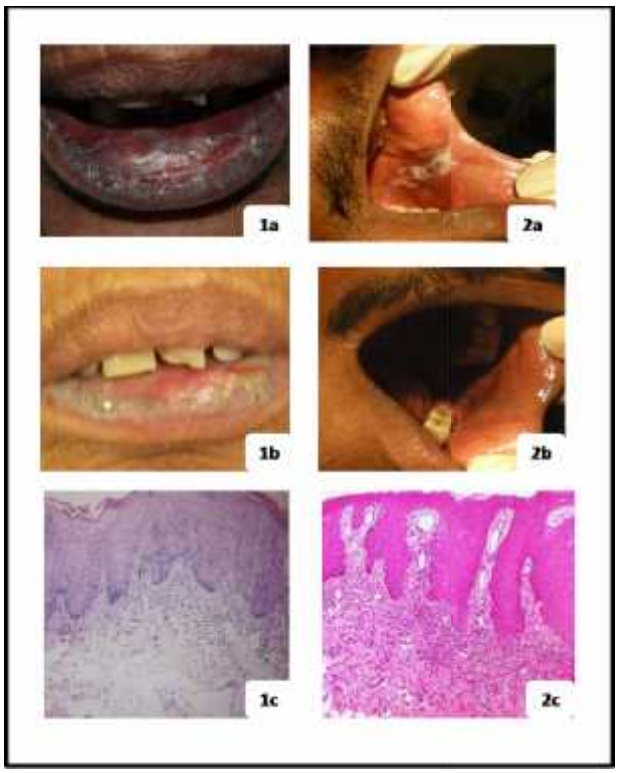

Figure 1a shows central atrophy with white striations at the periphery.

Figure 1b shows post treatment picture with remission of lesion.

Figure 1c Photomicrograph showing hyperorthokeratosis and acanthotic stratified squamous epithelium and connective tissue shows juxta epithelial cell-free zone, vasodilation and edema.

Figure 2a shows radiating lines giving a lacelike pattern resembling Wickham's striae with central atrophy and diffused areas of hyper pigmentation.

Figure 2b shows post treatment picture with complete remission of lesion.

Figure 2c shows parakeratinized stratified squamous epithelium, liquefaction degeneration of basal cell layer and patchy mononuclear infiltrate with inflammatory infiltrate beneath the basement membrane.

Case report 2

A 30 year-old, male patient reported to our department with a chief complaint of burning sensation since 6 months. The patient gives history of burning sensation which was gradual in onset, dull aching type and intermittent in nature. Intraoral examination revealed, a solitary, well defined white plaque present on the occlusal plane of left side of buccal mucosa with a radiating lines giving a lacelike pattern resembling Wickham's striae. It extends from commissure of lip till the distal aspect of 38. Diffused areas of hyper pigmentation present in and around the lesion. It is approximately 2x4cm in diameter, irregular borders and smooth and shiny surface. It is white in color in anteriorly and posteriorly reddish in color. On palpation, the lesion was soft in consistency without any evidence of bleeding (Figure 2a). Correlating history and clinical examination, this case was provisionally diagnosed as Lichen planus of left side of buccal mucosa.

The patient was subjected for routine blood examination, which was under normal limits. The biopsy was done on the left side of buccal mucosa and specimen was sent for histopathological examination. The H and E section showed parakeratinized stratified squamous epithelium overlying the connective tissue. The liquefaction degeneration of basal cell layer is seen with patchy mononuclear infiltrate. The connective tissue showed dense inflammatory infiltrate beneath the basement membrane chiefly composed of lymphocytes. The histopathological report gives suggestive of discoid lupus erythematosus (Figure 2c). The patient was subjected for further investigations, direct immnofluorescence was done, which revealed the presence of thickening of epithelial basement membrane. Correlating clinical and histopathological investigations, a final diagnosis of Discoid lupus erythematosus was made. The patient was advised Triamcinolone acetonide oral paste to apply two to three times per day and was recalled for follow-up for a period of 1 year (Figure 2b). There was no recurrence of the oral lesions seen.

DISCUSSION

Discoid lupus erythematosus is an immunologically mediated condition and is one of the most common of the so called "collagen vascular" or connective tissue diseases. It occurs by the age of 20 to 50 years. In present cases 1st, it occurs in 70 years, which is very rare and in present case 2nd, it occur in 30years of age which is consistent with the literature. Marked gender predilection in females, male female ratio is 8:1. But in cases 1st, it was female, which is consistent with literature and case 2nd it was male, which is very rare. It is caused by inter play of genetics, hormonal, and environmental. DLE is essentially a cutaneous disease with negligible propensity for systemic organ involvement. Its less frequent presentations include tumid, warty, chillblain, telangiectatic rosaceous, ulcerative, linear, bullous and acneiform. These lesions may appear similar to oral lichen planus. [4,5,6] Characteristic feature include itching or burning sensation. In present cases, burning sensation was present, which is consistent with literature. Incidence of oral lesions of DLE is about 20 to 50%. Present with or without skin lesion. The margins of the lesions are not sharply demarcated, but frequently show the formation of narrow zone of keratinization, hyperemia with edema, superficial, painful ulceration may occur with crusting, but no actual scale formation as is seen on the skin. Early classic DLE lesions typically evolve into sharply demarcated, coin shaped (i.e., discoid) erythematous plaques covered by a prominent, adherent scale that extends into the orifices of dilated hair follicles. The lesions typically expand with erythema and hyperpigmentation at the periphery leaving hallmark atrophic central scarring, telangiectasia, and hypopigmentation.[7] In present cases there was no systemic involvement and any skin lesions, which was consistent with the literature. The case 1st showed the lesion which was a white plaque type with areas of hyperpigmentation and central atrophy, while in 2nd case, the lesion was similar as lichen planus with white radiating lines and Wickham's striae, which is consistent with literature given. The diagnosis of discoid lupus is generally made based on clinical features.

Histology may be required to confirm the diagnosis; it is that of a lichenoid tissue reaction with changes at the dermo-epidermal junction that include thickening of the basement membrane (best demonstrated by periodic acid-Schiff staining) and vacuolar degeneration of the basal cells along with perivascular and peri-appendage inflammatory cell infiltration of a variable degree in the reticular dermis. Hyperkeratosis is more evident and follicular plugging may be seen in more mature lesions. Regarding the histological criteria of oral discoid lupus erythematosus, hyperortho and/or parakeratosis (hyperparakeratosis), liquefaction degeneration of the basal layer, focal or perivascular infiltrates of lymphocytes.[8,9] but

present cases, The H and E section showed parakeratinized stratified squamous epithelium overlying the connective tissue. The connective tissue showed dense inflammatory infiltrate beneath the basement membrane chiefly composed of lymphocytes, which was consistent with literature. DLE is less severe course than SLE and has a better prognosis. It is important for dentist to recognize DLE because it is a potentially scarring disease.

The treatment of DLE would in most instances be initiated at a dermatology department, but before instituting treatment for discoid lupus patients should be assessed for systemic involvement. This should include a full history and physical examination, full blood count, erythrocyte sedimentation, rate, midstream urine, and antinuclear antibody.

In present cases, the values were under normal limits. Topical steroids are the mainstay of treatment of DLE. Patients usually start with a potent topical steroid applied twice a day, then switch to a lower potency steroid as soon as possible. The minimal use of steroids reduces the recognized side effects like atrophy, telengiectasia, striae, and purpura. Intralesional steroids are particularly useful to treat chronic lesions, hyperkeratotic lesions, and those that do not respond adequately to topical steroids. Oral steroids may be required for the control of systemic lupus but are not generally beneficial in DLE. Treatment with antimalarials drugs constitutes first line systemic therapy for DLE. Therapy with antimalarials, either used singly or in combination is usually effective. The 3 commonly used preparations include chloroquine, hydroxychloroquine, and mepacrine. Potentially more toxic therapeutic medication needs to be used in the management of many cases of DLE; however, topical tacrolimus ointment has been found recently to be useful in the management of DLE.[10] In present cases, half of the lesion was removed during incision biopsy and other half was treated by Triamcinolone acetonide oral paste two to three times daily for 4 weeks and patient was recalled and reviewed for 1year.

CONCLUSION

Oral lesions may be the first clinical manifestation of DLE, so proper diagnosis is required for early detection and to establish better treatment planning, this promotes resolution of established lesions and prevents scarring of skin lesions and also, alleviates discomfort of the patients.

It is especially important for the dentist to recognize not only that some dermatoses exhibit concomitant lesions of the oral mucous membranes, but also that manifestation of some diseases may be preceded by oral lesions.

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