Original Article

DISSEMINATED DLE PROGRESSING TO SLE- A CASE REPORT

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Abstract

Lupus erythematosus is an autoimmune disease with a clinical spectrum ranging from mild cutaneous lesions to life-threatening diseases like nephritis, serositis and CNS involvement. James N. Gilliam developed a classification for LE-specific lesions which was divided into Acute cutaneous lupus erythematosus (ACLE), Subacute cutaneous lupus erythematosus (SCLE), chronic cutaneous lupus erythematosus (CCLE). The most common subset of chronic cutaneous lupus erythematosus is DLE. Most patients with DLE do not have significant systemic disease. The risk of developing SLE is 20% in disseminated DLE versus 5% in the localized form. Here we report a 41-year-old female patient who was a known case of DLE presentedwith history of new skin and mucosal lesions, fever, and joint pain. On further investigation, it was found that the patient has progressed to SLE. Careful history-taking , clinical examination and complete investigations should be carried in DLE patients to rule out progression to SLE.

Keywords: DLE, Lupus nephritis, SLE.

Introduction

Lupus erythematosus is a multisystem disorder that predominantly affects the skin. There are several types of cutaneous lupus. The most common types are acute cutaneous lupus (ACLE), subacute cutaneous lupus (SCLE), and chronic cutaneous lupus erythematosus which is also known as discoid lupus erythematosus.These patients may or may not report photosensitivity, but lesions are frequently photo distributed and tend to have secondary atrophy or scarring.Other forms of chronic cutaneous lupus erythematosus include hypertrophic lupus erythematosus, tumid lupus erythematosus, lupus erythematosus panniculitis (LEP or lupus profundus), chilblain LE, oral DLE, as well as DLE lesions on the palms and/or sole.There are haematological and serological changes in approximately half of patients, and these changes, with other evidence, suggest an autoimmune aetiology.The incidence of cutaneous lupus was 4/100 000, with 80% being DLE.Females are more commonly affected males,20% disseminated DLE can be converted to –SLE , 5% localised DLE – can be converted to SLE

Case report

A 41-year-old who was a known case of Discoid lupus erythematosus for 2 years presented in the dermatology opd with multipleskin lesions over the face, bilateral arms and upper back,multiple raw areas in the oral cavity for 5 days associated with fever,joint pain,fatigue,abdominal pain. On examination multiple well-defined annular hypopigmented plaque with surrounding hyperpigmentation and erythema with central atrophy was seen over the

bilateral chest,upper back,bilateral arm forearm,bilateral ear and scalp. She also had multiple hyperpigmented violaceous plaques present over the face,chest ,upper back and scalp(figure1, 2,3). Oral cavity examination showed multiple erosions over the palate and tip of the tongue(figure 4). Scalp examination showed scarring alopecia over occipital region.Nail and genital mucosa were normal.Dermascopic examination of the lesions in the arm showed hyperpigmentation at the periphery and central depigmentation associated with dilated blood vessels.Baseline investigation showed proteinuria and ultrasound abdomen showed hepatomegaly with a slight alteration of LFT.A punch biopsy of size 3mm was taken from the upper back and the diagnosis was confirmedby histopathology -HPE of the upper back whichshowed: hyperkeratosis with follicular plugging, thinning and flattening of the stratum malpighii, hydropic degeneration of basal cells, dyskeratosis, and squamatization of basilar keratinocyte,thickening and tortuosity of basement membrane correlating with deposition of immune reactants , interstitial mucin deposition edema, vasodilation, slight extravasation of erythrocytes which conformed the presence of DLE .Her anti-dsDNA was positive and serum complement c3 c4 levels were profoundly decreased .All the clinical and laboratory work up confirmed the progression of Disseminated Lupus Erythematosus to Systemic Lupus erythematosus. The patient was admitted and treated with systemic steroids and hydroxychloroquine after ophthalmological screening. Nephrologist opinion obtained for lupus nephritis management. Cutaneous and mucosal lesions started healing and patient is in regular followup.

FIGURE 1Multiple well defined hyperpigmented plaque present over the forehead cheeks and peri oral area.

FIGURE-2 Hyperpigmented plaque with central hypopigmentation seen in the ear

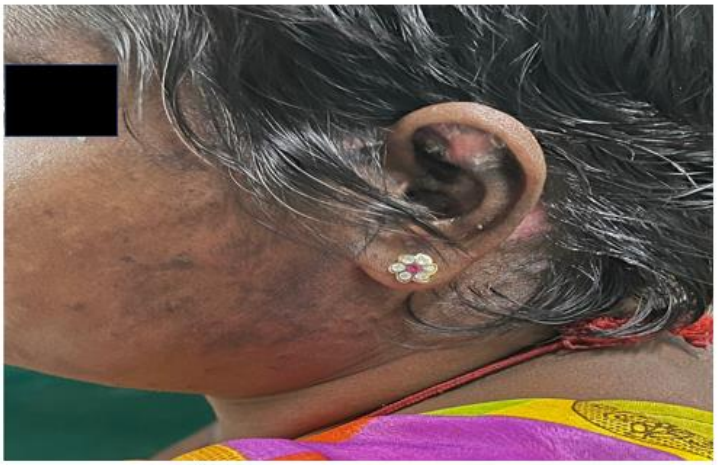


FIGURE 3 Hyperpigmented plaque in the upper back

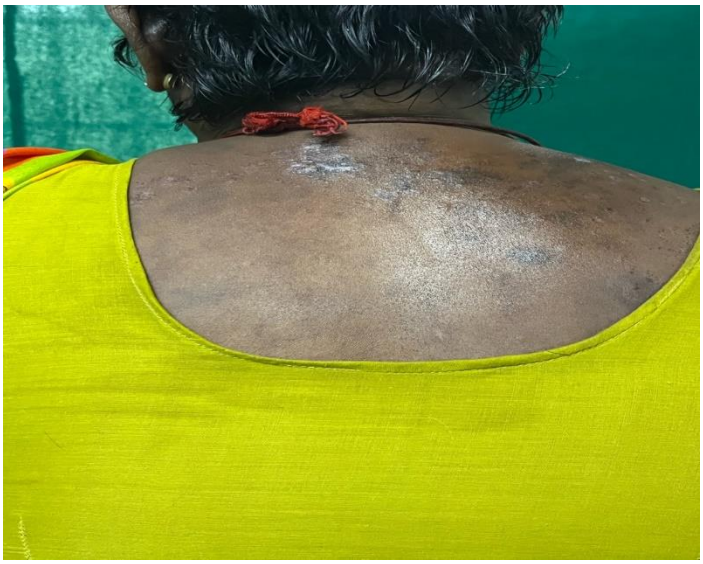


FIGURE 4Multiple well defined Hypopigmented plaques with surrounding Hyperpigmentation present in the upper limbs

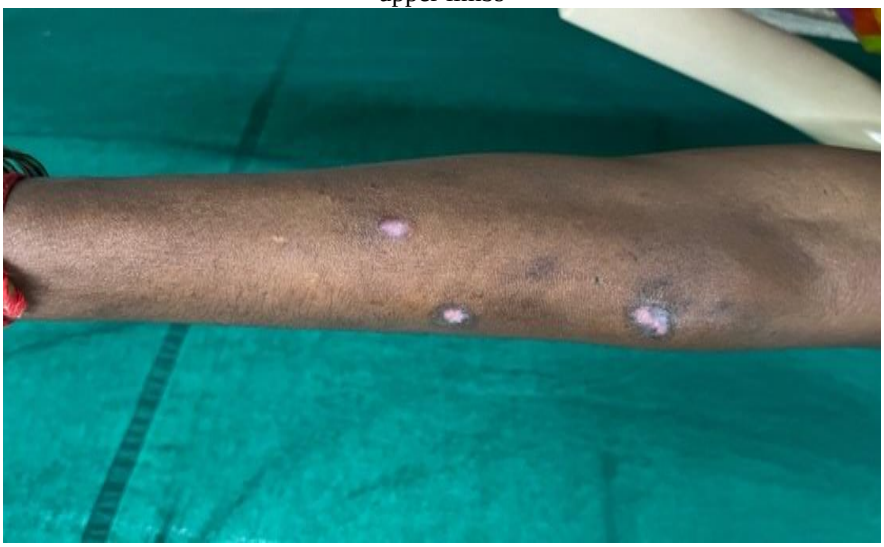


FIGURE 5erythema and erosions present over the tongue

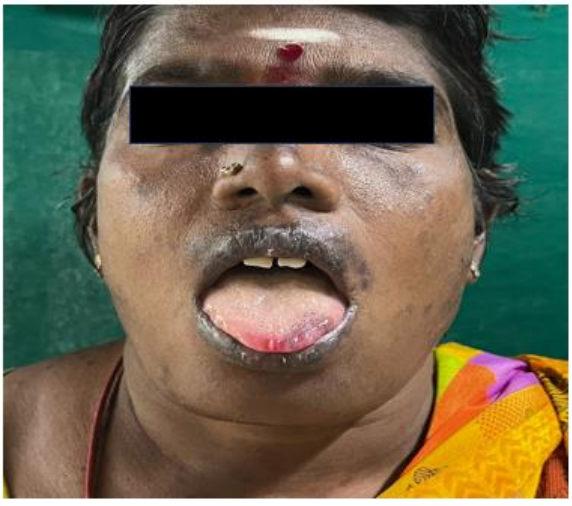


FIGURE 6 Dermascopic revealed telengiectasia, follicular red spots

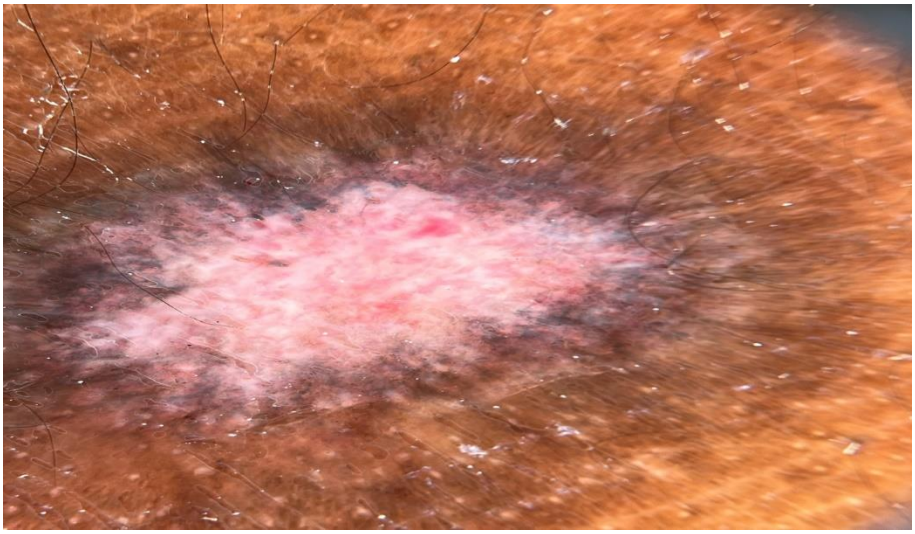
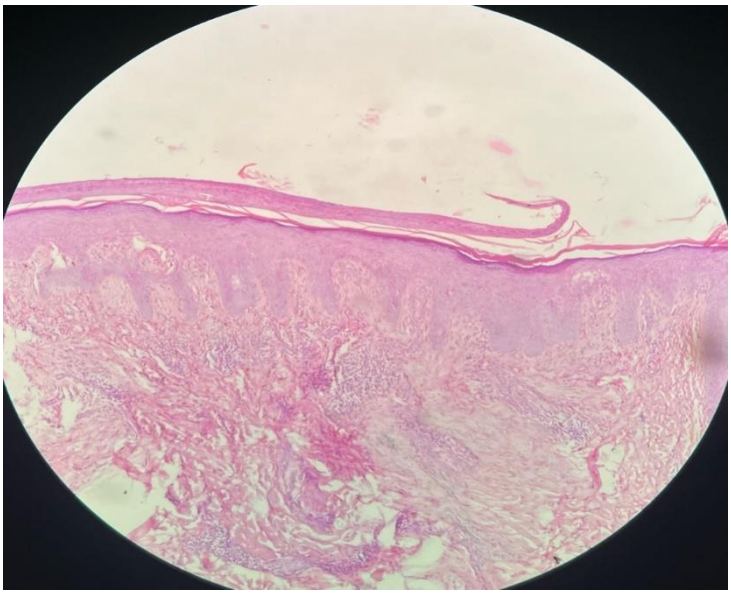


FIGURE 7H and E showing surface hyperkeratosis,follicular hyperkeratosis,perifollicular lymphoid infilterate



Discussion

Lupus Erythematosus is an autoimmune disorder.Itsaetiologyis multifactorial involving genetic,immunological and environmental factors. Apoptosis, necrosis, and chemokine production appear to mediate the recruitment and activation of autoimmune T cells and interferon-producing plasmacytoid dendritic cells, which subsequently release more effector chemokines, thus amplifying chemokine production and leukocyte recruitment. This drives a proinflammatory circle which leads to the development of the cutaneous LE phenotype. Ultraviolet (UV) exposure is the most accepted trigger factor for DLE. Several nuclear antigens are expressed on the surface of keratinocytes that undergo UV-induced apoptosis. Antinuclear antibodies that target these antigens are found in 20%–80% of CLE cases.The scarring seen in DLE is attributed to the dense infiltration of cytotoxic skin-homing T cells. Several HLA variants are associated with the development of LE. Recent studies have also focused on the genetics of interferon (IFN) signalling in CLE pathogenesis.Depending on the clincal and laboratory findings patient can be divided into cutaneous and/orsystemic lupus erythematous.Further James N.Gilliam divided cutaneous lupus erythematosus into acute, subacute and chronic lupus erythematosuswhich is also known as discoid lupus erythematosus. It is further classified as Localised, generalised /disseminated,hypertrophic

/verrucous, Lichenoid Lupus erythematosustelangiectatic, Hypertrophic, Linear Cutaneous lupus erythematosus,Rosaceous Lupus erythematosusAcneiform type,Pigmented macular type Dego's like lesions in Chronic cutaneous lupus erythematosus,Lupus erythematosus panniculitis /lupus profundus,Lupus erythematosus tumidus Chilblain,Mucosal Discoid lupus erythematosus,classically DLE presents as well defined erythematous plaques thick adherent scales,they may extend into dilated hair follicle causing follicular plugging,on the removal of the thick adherent scales results in tin tack /carpet tack sign.The usual sites of presentation arethe v region of the neck,face, lips, scalp, eyelids, and trunk. The distinction between localized DLE with involvement above the neck and disseminated DLE has prognostic implications since the risk of developing SLE is 20% in disseminated DLE versus 5% in the localized form.In patients of DLE scalp is affected more than 60% and it may be the only site involved in 10% of the patients,it can result in scarring alopecia.Dermoscopic features of DLE include perifollicular whitish halo, follicular keratotic plugs, and telangiectatic vessels. Laboratory tests are indicated for patients with disseminated DLE and those with systemic symptoms. Systemic symptoms like fever, joint pain and photosensitivity to be looked for in each follow-up visits to monitor the progression to SLE. ANA may be positive in low titer. Detection of anti-dsDNA could indicate the transition to SLE.Skin biopsy helps to establish the diagnosis of CLE.DIF test is a useful adjunct in cases where histopathologicfeatures include orthohyperkeratosis with keratotic follicular plugging and atrophy of the malpighian layer, (2) hydropic degenerationof basal cells, dyskeratosis and squamatization of basilar keratinocytes, (3) colloid bodies which are round-to-homogenous, eosinophilic structures, (4) thickened basement membrane and (5) lymphocytic infiltration which is seen at the dermo-epidermal junction (DEJ), and in a perivascular and periadnexal distribution. Interstitial mucin deposition is often present. The hair follicles may be involved. Lesional direct immunofluorescence (DIF) test is not mandatory to establish the diagnosis of classic DLE. However, it is a useful adjunct when histopathologic features are not typical. A positive DIF test is characterized by the deposition of two or more immunoreactants, IgG, IgM, and C3, and rarely IgA in a band along the DEJ of lesional skin in DLE- Seen as a linear granular-like deposits indicating positive lupus band test.

Conclusion

Progression of DLE to SLE though accounts for 5- 20%, strict monitoring of clinical symptoms and regular laboratory investigations to be done for early diagnosis of SLE in DLE patient. Earlier and aggressive treatment will prevent systemic damage in SLE. Photoprotection should be educated well to all DLE and SLE to prevent disease aggravation.

References

1. Vukicevic JS, Milobratovic DJ. Discoid lupus erythematosus of the eyelid. Indian J Dermatol Venereol Leprol 2010;76:418-420
2. Selva D, Chen CS, James CL, Huilgol SC. Discoid lupus erythematosus presenting as madarosis. Am J Ophthalmol 2003;136:545-6.
3. Goodfield M, Dutz J, McCourt C. Lupus Erythematosus. In: Griffiths C, Barker J, Bleiker T, Chalmers R, Creamer D, eds. Rook's Textbook of Dermatology.9th ed. West Sussex: Wiley Blackwell; 2016: 51.2
4. Achtman JC, Werth VP. Pathophysiology of cutaneous lupus erythematosus. Arthritis Res Ther. 2015;17(1):182. Published 2015 Aug 10.
5. Albrecht J, Taylor L, Berlin JA, et al. The CLASI (Cutaneous Lupus Erythematosus Disease Area and Severity Index): an outcome instrument for cutaneous lupus erythematosus. J Invest Dermatol.