



SQUAMOUS CELL CARCINOMA DEVELOPING IN A CASE OF LONG-STANDING DISSEMINATED DISCOID LUPUS ERYTHEMATOSUS – A CASE REPORT

Yogesh D¹, Romita Bachaspatimayum², Bijayanti Devi TH²

¹Department of Dermatology, Venereology and Leprology, Adichunchanagiri Institute of Medical Sciences, B.G.Nagara, Nagamangala taluk, Mandya district, India

²Department of Dermatology, Venereology and Leprology, Regional Institute of Medical Sciences, Imphal, Manipur, India.

Corresponding Author:- **Yogesh D**
E-mail: yogeshdevaraj86@gmail.com

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ABSTRACT

Discoid lupus erythematosus (DLE) is a chronic autoimmune mucocutaneous disease of unknown aetiology. Typically, central atrophy, small white keratinized plaques with elevated hyperpigmented borders, and telangiectasia are seen in these patients. Squamous cell carcinoma (SCC) developing in a lesion of DLE is very rare. We report a case of a 45 year old lady who developed SCC in a lesion of DLE after 10 years.

INTRODUCTION

Discoid lupus erythematosus (DLE) is a chronic autoimmune mucocutaneous disease of unknown aetiology. Typically, central atrophy, small white keratinized plaques with elevated hyperpigmented borders, and telangiectasia are seen in these patients. DLE is subdivided into a localized form in which lesions are confined to the face and neck or a disseminated form in which lesions also occur elsewhere on the body [1-3]. Oral cavity lesions are found in 20% of patients with DLE. Squamous cell carcinoma (SCC) developing in a lesion of DLE is very rare [2]. We report a case of a 45-year-old lady who developed SCC in a lesion of DLE after 10 years.

Case report

A 45-year-old female farmer with SCC over right arm was referred from the Department of Radiotherapy of our Institute as she was a diagnosed case of disseminated

DLE. She was on irregular treatment with hydroxychloroquine, topical steroid and sunscreen for the last 10 years. One year back, she developed an asymptomatic verrucous growth on her right arm over one of the skin lesions which gradually increased in size. There was history of bleeding from the lesion on and off for the last 3 months. Examination revealed multiple, well-defined discoid plaques with atrophic, depigmented centres and hyperpigmented borders over both forearms, arms, dorsa of hands and feet, legs, thighs, ears and back (Figure 1). Scalp showed diffuse alopecia and the nails showed melanonychia and longitudinal ridges. Right arm showed a non-tender ulcero-proliferative growth 8cm x 6cm in size with haemorrhagic verrucous surface (Figure 2), not fixed to underlying structures. There was no axillary lymphadenopathy. Systemic examination was normal. ANA was positive (49.1 I.U) with homogeneous pattern.



Anti-ds DNA was negative. Haematological examination and other routine investigations like liver and renal function tests and chest X-ray were within normal limits. Biopsy of lesional skin showed hyperkeratosis with keratotic plugging, hydropic degeneration of basal layer with pigment incontinence and small foci of extravasated RBCs and patchy predominantly lymphoid infiltrate in the upper dermis suggestive of discoid lupus erythematosus. Histopathological examination of incisional biopsy from the lesion on the right arm showed features of a well-differentiated SCC. The patient was treated with hydroxychloroquine 200mg b.d, and topical corticosteroid for DLE lesions, along with broad-spectrum sunscreen

lotion improvement at 3 weeks follow up (Figure 4). External beam radiation therapy (EBRT) of 39 Gy over 13 fractions with Cobalt 60 in antero-posterior and postero-anterior fields was given for SCC lesion at the Radiotherapy Department with significant improvement at 3 weeks and complete clinical resolution by 2 months (Figure 5). At 3 months follow up, she had a verrucous growth near the margin of the site of previous SCC (Figure 6). FNAC of the new lesion showed SCC in-situ, for which she received radiation therapy in the same dosage in the Department of Radiotherapy with complete clinical response at two weeks. However, the DLE lesions were improving till the last follow up.

Fig 1. Erythematous scaly plaques with peripheral hyperpigmentation over dorsa of hands



Fig 2. Reddish-brown verrucous growth with haemorrhagic areas adjoining hypopigmented atrophic, scaly, plaques



Fig 3. Histopathological examination of specimen from the verrucous growth showing features of well-differentiated Squamous cell carcinoma (H and E, x100)

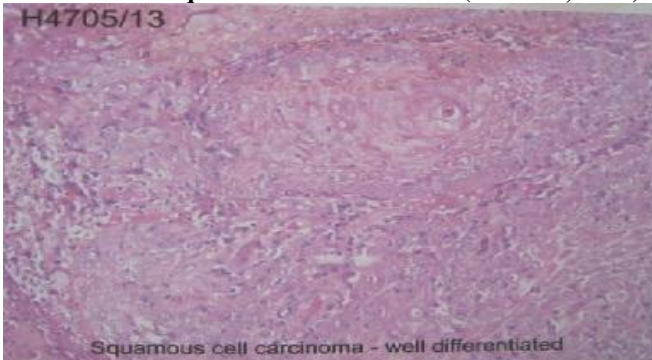


Fig 4. Reduction of erythema and scaling at 3 weeks



Fig 5. Complete clinical resolution of SCC at 2 months

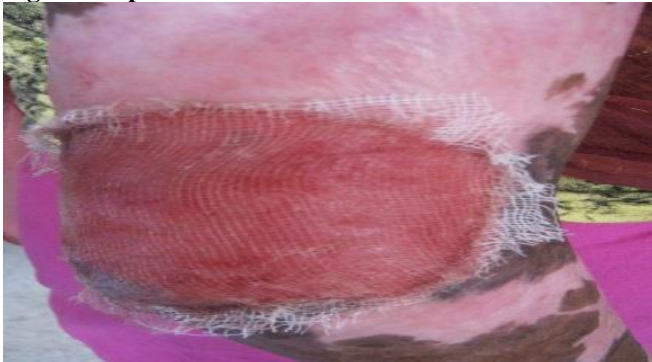


Fig 6. New verrucous growth at 3 months



DISCUSSION

SCC is a rare complication of long-standing DLE. The incidence of SCC developing in DLE varies from 3.3% to 3.4% [1]. SCC usually arises from skin damaged by actinic rays. Exposure to chemicals like coal tar, soot and arsenic are also implicated in its pathogenesis. It can also occur in scars following inflammatory or degenerative processes [5]. Due to mutation in p53 tumour suppressor gene, there may be defect in apoptosis of keratinocytes that have sustained UV-radiation-induced DNA damage which ultimately leads to SCC [6]. DLE-related SCCs have been observed to be more aggressive than conventional SCCs [1]. The recurrence, metastasis, and mortality rates were 10% to 20% higher than that of non-DLE-related SCCs [4]. When such lesions are encountered differentiation must be made between hypertrophic DLE and SCC by histopathological examination.

Reports of neoplastic change in DLE range from SCC and basal cell carcinoma to malignant fibrous histiocytoma to fibroxanthoma [7]. The interval between development of DLE and SCC varies from 4-20 years across different studies [8], which is in concordance with our case where SCC developed after 10 years of

appearance of DLE lesions. Precipitating factors for SCC are age more than 40 years, male sex, sun/ultraviolet ray exposure, skin pigmentation and chronic inflammatory processes. There is an inverse relation between skin pigmentation and development of SCC because of the protective effect of melanin [6]. In our case, it could be due to prolonged sun exposure as the patient was a farmer and also due to the fact that she was on irregular treatment.

The long-term prognosis of such cases is varied. SCC arising in DLE is regarded as a locally aggressive but low grade carcinoma. One study reported local recurrences in about 20% and metastasis in 30% cases [9]. Recurrence was also noted in our case. Death has also been reported from multiple metastases [10].

CONCLUSION

There should be a high index of suspicion for non-healing lesions or ulcers on discoid lesions of DLE and biopsy of these lesions should be done for early diagnosis and management of carcinoma and such cases have to be followed-up life-long to detect any metastases or recurrence.

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