Mutilating Squamous Cell Carcinoma: A Rare Complication of an Early Plaque of Discoid Lupus Erythematosus S Kaur, P Sahu, S Dayal, VK Jain, N Jindal, V Jairath

ABSTRACT

Cutaneous Squamous cell carcinoma (SCC) is the second most common form of nonmelanotic skin cancer after basal cell carcinoma (BCC). It is a well-known complication of DLE and usually favours the depigmented lesions located over sun-exposed areas. The clinical course of this complication may be aggressive, with a high risk of mortality. Literature describes numerous case reports of SCC complicating the lesions of DLE but mutilating SCC is rarely mentioned. Here in, we report a rare case of mutilating squamous cell carcinoma arising in an early (2 years duration) lesion of DLE.

Keywords: Discoid lupus erythematosus, mutilation, squamous cell carcinoma

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INTRODUCTION

DLE is an autoimmune cutaneous disorder, characterized by one or more asymptomatic, sharply demarcated, erythematous plaques with adherent scales which heal with scarring, atrophy and pigmentary changes (1). Due to evolving treatment options and growing awareness of patients, complications of DLE like SCC and mutilating changes are seldom encountered (2). There are several case reports of SCC arising in DLE plaques, however mutilating SCC is barely reported in the literature. Thus, we are reporting a rare presentation of mutilating SCC arising in an early lesion of DLE in a 70 year old female.

CASE REPORT

A 70 year old female presented to the outpatient department of dermatology with multiple asymptomatic, erythematous plaques over the sun exposed and covered area since 6 months. The lesions aggravates upon exposure to sun. There was no history of fever, joint pain or Raynaud's phenomenon. Physical examination revealed multiple, well defined erythematous to depigmented scaly, hypertrophic plaques, varying in size from 0.5 cm to 7cm surrounded by a rim of hyperpigmentation over the scalp, face, hands and extensor surface of bilateral forearm and lower limbs [Figure1a]. Oral cavity was spared. Routine haematological and biochemical investigations were within normal limits. The antinuclear antibody and rheumatoid factor were also negative. The skin biopsy was suggestive of DLE. After fundus examination, she was started on tablet hydroxychloroquine 200 mg twice a day and topical steroids. General measures like regular use of sunscreen were also advised.

She was on regular follow up for the next one year and lesions were gradually improving. But unfortunately, patient lost her follow up and presented after eight months with a large mass over the nose. On examination, there was a well-defined erythematous

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ulcerative fungating mass of 6×8 cm in size over the nose. The margins were rolled out and the surface was vertucous, on palpation it bled on touch. The mass roofed whole of the nose, extending to the right cheek and bilateral inner canthus [Figure1b]. Bilateral submandibular lymphadenopathy of size varying from 2×1 cm to 1.5×2 cm was noted. On palpation lymph nodes were firm, mobile and non- tender. Systemic examination was normal.

A complete haemogram revealed haemoglobin of 8.4 gm% and a raised ESR 60 mm/hr. Biochemical investigations and chest X ray were normal. Skin biopsy from the mass over the nose showed irregular nest of malignant squamous cell with eosinophilic cytoplasm and multiple small and large keratin pearls in the dermis. The stroma consists of loose connective tissue and dense infiltrate of lymphocytes [Figure 2]. These histopathological features were consistent with the diagnosis of well differentiated squamous cell carcinoma. Based on the clinical features and histopathological findings she was diagnosed as a case of disseminated DLE with mutilating SCC. Dermatological part of treatment was continued and she was referred to oncosurgery department for further evaluation and possible reconstructive surgery.

DISCUSSION

DLE is a chronic form of cutaneous lupus erythematosus, manifested clinically by well defined, discoid erythematous plaques covered by a prominent adherent scales, which tend to heal with atrophy and scarring. It is subdivided into a localised form, confined to the head and neck and a disseminated form in which lesions also occur both above and below the neck.(1)

SCC is a rare, but well-recognized, long-term complication of chronic DLE. According to Millard et al the incidence of SCC arising in lesions of DLE was 3.3%, scalp being the most common site followed by the lips (3). It usually develops in a depigmented chronic discoid lesions that are located over sun-exposed areas. The time period between the occurrence of SCC in DLE plaque varies from 7 to 41 years, but earlier onset of one year has been reported by Parikh et al (4, 5). Precipitating factors of SCC are (a) age more than 40 years, (b) male sex with early onset DLE, (c) exposure to ultraviolet rays, (d) chronic inflammatory processes with scarring and (e) patients resistant to treatment. (6) In the present case, the SCC developed in a DLE plaque after 2 years which is quite early. The early onset of this malignancy may be attributed to the age (i.e. 70 years), photo-exposed area (i.e. nose) and lack of treatment (due to lost follow).

Cutaneous SCC, the second most common form of non-melanotic skin cancer, is an indolent tumour with low metastatic potential and excellent prognosis.(7) However, SCC developing on chronically diseased skin has a substantial risk of recurrence, metastases and death as compared to those developing on non-diseased skin.(4)

Literature has described near about 53 cases of DLE complicated by SCC, and in most, the lesions were on sun exposed area (4). Factors that might help in preventing the complications of DLE plaque are a) protecting the affected area with high sun protection factor (SPF) sunscreen, b) examination of any new change in the morphology of DLE plaques, and c) biopsy of a suspicious lesion for early diagnosis and timely recognition of malignancy and consequently preventing mutilation.

To the best of our knowledge this is the first case of early mutilating SCC developing in the lesion of DLE. This case has been presented to emphasize that mutilating malignant transformation can develop in an early lesion of DLE .In our case patient lost her follow up for merely eight months , occurrence of SCC with mutilating changes in such a short period is unexplainable. However, this can be explained on the basis of literature mentioning the aggressive nature of SCC on a diseased skin. Hence, a high index of suspicion is warranted in cases of DLE even if the lesions are of recent onset, especially with risk factors like age, lesions over sun- exposed area and chronic scarring.

CONCLUSION

Initiation of treatment on time, regular visit of the patient and keen eye to look for any suspicious change in the morphology are the only possible steps which collectively can prevent complications.

There are few factors that make this case interesting. Firstly, malignant changes in a 2 yr old DLE plaque, which is quite early according to the literature. Secondly, mutilating SCC as a complication of DLE is a rare presentation, third, occurring over the nose, considered to be the most common site of DLE.

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Fig.1 (a): Multiple erythematous, scaly depigmented plaques surrounded by rim of hyperpigmention over legs and dorsum of hands.



Fig. 1 (b): Mutilating mass of SCC over the nose, with erythematous, depigmented plaques of DLE involving face and dorsum of the hands.



Fig. 2: H&E: 40x; Photomicrograph of a well differentiated squamous cell carcinoma of the nose-showing malignant squamous epithelial cells and multiple foci of keratin pearls.