Squamous cell carcinoma over lower limb in an Indian patient with disseminated discoid lupus erythematosus

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Abstract

Disseminated discoid lupus erythematosus (DLE) is a subtype of DLE when the typical lesions are found below the neck. Squamous cell carcinoma being a very rare complication of discoid lupus erythematosus in India. It is usually seen over the scalp, lips, cheek or ears. Case report: A 54 year old woman presented 2 years back with multiple scaly lesions with dark colored borders over the lower limbs. She gave history of such lesions since last 12 years. She was then diagnosed with DLE, she now presented with asymptomatic growths over the right leg since last 1 year. On examination, there were multiple vegetative plaques over the right lower limb which had central ulceration. They were non tender and with no local rise of temperature. The inguinal and femoral groups of lymph nodes were not enlarged. Histopathological examination revealed hyperkeratosis, papillomatosis, keratin pearls and malignant atypical cells invading dermis arranged in cords and sheets, the features were consistent with that of a well differentiated SCC. We evaluated our case of disseminated DLE for any progression to systemic lupus erythematosus with no systemic involvement. She was referred to an onco-surgeon for excision.

Key words

Squamous cell carcinoma, disseminated, discoid lupus erythematosus.

Introduction

Discoid lupus erythematosus (DLE) is a chronic, benign disorder of the skin and mucous membranes. It is a prototype of chronic cutaneous lupus erythematosus (CCLE). It typically presents as white or pinkish keratinized scaly plaques which heal with central atrophy, scarring and pigmented changes. The plaques have elevated and hyperpigmented borders. 20% patients have lesions in the oral cavity and can encroach even the upper lip. Histopathologically DLE shows follicular plugging, vacuolar degeneration of basal cell layer of epidermis along with patchy dermal lymphocytic infiltrate.

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Case report

A 54-year-old female patient came 2 years back with multiple scaly lesions with dark colored circumference over the lower limbs. She gave
history of such lesions since last 12 years. Cutaneous examination showed erythematous scaly lesions with central hypo and depigmentation along with atrophy, the outer borders of the lesions were hyperpigmented. They were present bilaterally from knees to the foot region in symmetrical fashion, sparing the scalp, face, upper limbs and the trunk (Figure 1 & 2). Her investigations included complete haemogram, LFTs, RFTs etc. which were found to be within normal limits. Anti-nuclear antibodies (ANA) were found to be elevated (1:80 dilution). She underwent skin biopsy and histopathology revealed hyperkeratosis with follicular plugging, thinning and flattening of the stratum malphighi, hydropic degeneration of basal cells, dyskeratosis and squamatization of basilar keratinocytes in the epidermis. Dermis had predominant lymphocytic infiltrate arranged along the dermal-epidermal junction and around hair follicles (Figure 3). Clinically and histopathologically, a diagnosis of disseminated DLE was made. She was given hydroxychloroquine (after ophthalmological examination), topical corticosteroids and sunscreen agents. She now presented to us with asymptomatic growths over the right leg since last 1 year. On examination, there were multiple vegetative plaques over the right lower limb which had central ulceration. They were not tender and with no local rise of temperature. The inguinal and femoral groups of lymph nodes were not enlarged. (Figure 4 & 5) Systemic examination was normal. Hematological investigations revealed a raised ESR. Excisional biopsy from the fungating growth from the lower limb was done and sent for histopathological examination. It revealed hyperkeratosis, papillomatosis, keratin pearls and malignant atypical cells invading dermis arranged in cords and sheets, (Figure 6). The features were consistent with that of a well differentiated SCC. Patient was counseled to
restart the treatment for DLE and was referred to an onco-surgeon for aggressive surgical excision of the carcinoma.

Discussion

Characteristic lesions of DLE occur in widespread pattern over trunk and limbs in the disseminated variety. It tends to be persistent and resistant to therapy. It is more common in females. There are various complications associated with DLE like scarring alopecia, hyper and hypopigmentation, systemic features like arthritis, pleuritis, pericarditis or even psychosis. SCC is one of the dreadful complications of DLE. Though its prevalence is very low in our country but there have been studies from other parts of world showing prevalence of up to 3.3%-5.26% cases of CCLE. Precipitating factors for SCC include age more than 40 years, male sex, sun/ultraviolet ray exposure, skin pigmentation and chronic inflammatory processes. Though DLE is a disease of mostly females but malignant transformation is seen more in males. There is an inverse relation between skin pigmentation and development of SCC because of the protective effect of melanin. The interval between development of DLE and SCC has varied from 4 to 20 years, but earlier onset has also been reported. The scalp is the most common site affected, followed by the lips, cheek, ears, trunk and even groin. In contrast, our case has developed SCC over the lower limb. Exposure to chemicals such as coal tar, soot, arsenic and a variety of oils have been implicated in its pathogenesis along with actinic rays. Due to mutation in p53 tumor suppressor gene, UV-radiation-induced DNA damage occurs which eventually leads to SCC. Some authors have postulated it to be an end-stage complication of a wide array of inflammatory skin conditions. The long-term prognosis of a complicated case of DLE is quite variable. SCCs are usually locally aggressive but are low-grade carcinoma with frequent recurrences. Simpson et al. documented an association and likely aggressiveness of SCC in patients infected with Human Papilloma Virus. Cutaneous SCC is more common to occur in individuals with skin type I, II and III. We present a rare report of a tumor arising over the lower limb in India.

We even worked on our case to look for any progression to systemic lupus erythematosus (SLE) as disseminated DLE has more chances; our patient did not have any features of SLE, despite having disseminated DLE for 12 years.

Conclusion

India is a country where we usually don’t see many cases of disseminated DLE due to darker skin type and its location in the temperate region, though few cases have been reported. Occurrence of a rare complication like SCC over lower limb is even rarer in India.

References