Case study

SQUAMOUS CELL CARCINOMA COMPLICATING PLAQUE OF LUPUS ERYTHEMATOSUS - A CASE SERIES

Abstract:

Lupus Erythematosus is a group of autoimmune disorders with a wide spectrum of clinical presentations ranging from cutaneous involvement to widespread systemic involvement. Squamous cell carcinomas formation in cutaneous lesions of LE is rare with greater chances of metastases. Here, we report two cases, one of Discoid Lupus Erythematosus and other of Systemic Lupus Erythematosus complicated by development of squamous cell carcinoma over cutaneous lesions.

Keywords:

Discoid Lupus Erythematosus, Squamous Cell Carcinoma, Systemic Lupus Erythematosus

Introduction:

Systemic lupus erythematosus (SLE) is a systemic disease which causes multisystem organ inflammation which includes skin, joints and vasculature, kidneys, lungs, heart and central nervous system. Cutaneous Lupus Erythematosus (CLE) is a chronic, relapsing autoimmune condition encompassing a wide range of dermatologic manifestations. Skin

involvement in CLE patients can be divided into two categories based on histology: Lupus Erythematosus- (LE-) specific and LE-nonspecific skin lesions. The presence of LE-specific lesions is necessary to confirm the diagnosis of CLE. LE-specific skin lesions are divided into several subtypes based on clinical characteristics: Acute CLE (ACLE) Subacute CLE (SCLE) and Chronic CLE (CCLE) with several variants including Discoid LE (DLE) presenting as a localized or generalized form, LE Profundus (LEP) (Also called lupus panniculitis or subcutaneous LE) Hypertrophic LE (HLE) Chilblain LE (CHLE) and Lupus Erythematosus Tumidus (LET).[1] Squamous cell carcinoma is a malignant tumour arising from epidermal keratinocytes or its appendages. It has risk factors as, HPV infection, UV radiation, immunosupression, chronic infections and inflammatory conditions, fair skin, age over 50, premalignant conditions like actinic keratosis. Squamous cell carcinomas formation in cutaneous lesions of LE is rare with greater chances of metastases.[2]

Presentation of Case:

A 35-year-old female, a known case of systemic lupus erythematosus since 8 years who is on irregular treatment, came with complaints of exophytic growth over the lower lip since last three months. It later progressed in size gradually to size of 2 X 3 cm approximately. Since one week, burning sensation over the lesions was present. There was development of ulcers over preexisting lesions over her back and legs. There was history of scaling and crusting over the lesions. Patient had photosensitivity and arthalgia of small joints of her hands, wrists and shoulders. She is also a known case of seizure disorder and is on regular treatment for the same on the advice of neurologist. On inspection, multiple hypopigmented discoid scaly lesions over face, scalp, posterior neck and extremities were seen. Few of the lesions were erythematous and ulcerated with crust formation. Scarring alopecia of the scalp was seen. Patient had an exophytic mass of size 2x2 cm over her lower

lip with crust formation. It was non-tender and firm on palpation. A biopsy was taken from the lesion which showed proliferative epidermal mass, keratin pearls and features of atypicality yielding a diagnosis of squamous cell carcinoma. Patient was then referred to general surgery and wide local surgical excision was performed. Hydroxychloroquine and systemic prednisolone with other supportive medications were given as a treatment of SLE.

Another case of 50yr old female came with chief complaints of hyperpigmented elevated lesions with central depigmentation over right shoulder and lips since 1.5 years followed by development of a verrucous plaque over right shoulder since 2 months for which she was on treatment. History of photosensitivity was present. On examination, verrucous growth was seen over the depigmented scaly plaque over the right shoulder. Similar depigmented lesion seen over the lip. There is no tenderness, no ulceration. A biopsy was performed from the verrucous lesion which showed features of atypicality and keratin pearls. Hence, a diagnosis of Squamous cell carcinoma was made. Wide local surgical excision was done by the general surgeon. Patient was started on hydroxychloroquine as a treatment for lupus erythematosus.

Discussion :

Lupus erythematosus is a multisystem auto-immune disorder which prominently affects the skin. It mainly occurs due to production of autoantibodies against own body tissue components which later leads to formation of antigen-antibody complexes which cause inflammation and injury to the tissue. Nucleic acid and their associated protein constituent of cell nuclei are targetted by autoantibodies, called as anti-nuclear antibodies (ANA). Other autoantibodies like anti-dsDNA, anti-Sm, anti-la, anti-rocan also be detected. There are few factors which prompts the onset and progression of the disease which include ultraviolet radiation, medications, EBV infection, emotional stress, estrogen levels.[3] Development of malignancy like Squamous cell carcinoma, Basal cell carcinoma, malignant fibrous histiocytoma and atypical fibroxanthoma have been

reported. In Indian population, development of SCC over DLE in the is found to be 0.98% to 3.4% with a male-to-female ratio of 1.6:1.[4] In SCC development, preference for lips might be due to reasons like, it being a common site of DLE, constant sun exposure, tobacco use, and irritation from food.[5] These factors may accelerate carcinogenic process in turn leading to shorter time for development of carcinoma after DLE diagnosis.[6] Fitzpatrick skin type I,II and 3 are more prone for Squamous cell carcinoma.[7] Prognosis of the actual LE disease is considered to be poor when development of carcinoma is seen.[8] Long term follow up for such patients is warranted as there are high chances of recurrences.[9] The interval between development of DLE and SCC has varied from 4 to 20 years.[10]

For diagnosis of LE, basic laboratory investigations like CBC,ESR with ANA profile are done. A skin biopsy can yield the diagnosis with peculiar histopathological features like atrophy of epidermis hyperkeratosis, follicular plugging, interface dermatitis with a patchy lymphocytic infiltrate in dermis. Direct immunofluoroscence test from the normal sun protected skin shows presence of Lupus band which is a band of deposits of immunoglobulins along dermo-epidermal junction. Histopathology of squamous cell carcinoma reveals infiltration of epidermis into the dermis,features of atypicality like individual cell keratinization,loss of intercellular bridges,hyperchromasia and hyperplasia of the nuclei,increased mitotic figures, great variation in size and shape of the cells, moderate lymphoid infiltrates , keratin pearls.

Treatment depends on individual and on the severity of the disease. Photoprotection should be achieved by suitable means.Most important drugs used for treating LE include, NSAIDs, Hydroxychloroquine, corticosteroids, belimumab and other immunosupressive drugs like azathioprine, cyclophosphamide, methotrexate and mycophenolate mofetil. Topically corticosteroids and Calcineurin inhibitors like tacrolimus can be tried. For treatment of SCC, complete surgical excision and radiotherapy remains the mainstay.

Conclusion:

Development of malignancy over discoid lesions of LE is rare. However, close follow up of lesion with supervision for any atypical changes is important. These SCC as well as the disease LE as such both are more severe when associated with each other in comparison to conventional ones. Thus, here dermatologists play important role in suspecting SCC in early stage. In turn, early treatment can limit the spread in early stages itself.

Legends to figures:

Figure 1A - Case 1: Clinical photograph showing hypopigmented discoid scaly lesions on posterior neck and back.

Figure 1B - Case 1: Clinical photograph showing verrucous nodole over hypopigmented discoid scaly lesion on lower lip.

Figure 2A – Case 1: Histopathological picture showing infiltrative epidermal masses

Figure 2B – Case 1: Histopathological picture showing keratin pearl.

Figure 3 – Case 2: Clinical photographs showing hypopigmented discoid scaly lesions on lips and shoulder with verrucous lesion on shoulder.

Figure 4A – Case 2: Histopathological picture showing interface dermatitis.

Figure 4B – Case 2: Histopathological picture showing individual cell keratinisation.

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Figures:

Figure 1A - Case 1: Clinical photograph showing hypopigmented discoid scaly lesions on posterior neck and back.

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Figure 2A – Case 1: Histopathological picture showing infiltrative epidermal masses

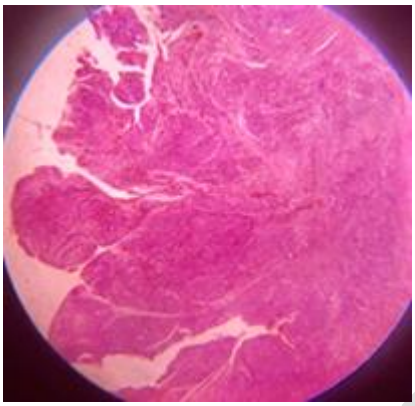

Figure 2B – Case 1: Histopathological picture showing keratin pearl.

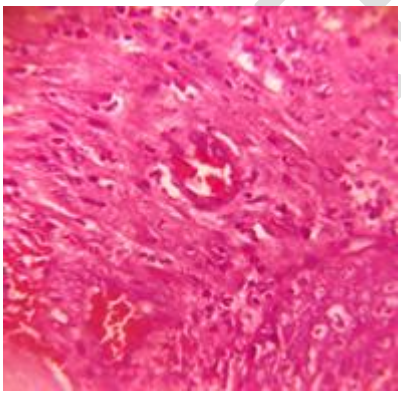

Figure 3 – Case 2: Clinical photographs showing hypopigmented discoid scaly lesions on lips and shoulder with verrucous lesion on shoulder.

Figure 4A – Case 2: Histopathological picture showing interface dermatitis.

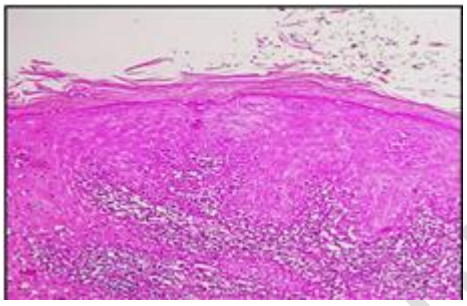

Figure 4B – Case 2: Histopathological picture showing individual cell keratinisation.