Mycosis Fungoides with Photosensitivity Mimicking Chronic Actinic Dermatitis

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Sir,

Mycosis fungoides (MF) represents malignant transformation of CD4 T cells in the skin. MF often presents as randomly distributed patches, plaques, and later tumors; hence, it is often difficult to distinguish it clinically from a myriad of benign dermatoses.[1] Chronic actinic dermatitis (CAD) presents as persistent or recurring dermatitis affecting photo exposed areas with abnormal photosensitivity. There are occasional reports of lymphoma developing in long-standing cases of CAD particularly the actinic reticuloid (AR) variant.[2] However, lesions of MF developing in a photo-distributed pattern resembling CAD have been reported very sporadically in the preceding literature.[3] We report a case of MF associated with photosensitivity resembling chronic actinic dermatitis.

A 50-year-old lady presented with complaints of asymptomatic gradually progressive red raised lesions over face and upper limbs of 5 years duration with intermittent exacerbation on exposure to sunlight. Patient had been treated in the past with multiple courses of oral and topical steroids, sunscreens, and tablet hydroxychloroquine with suboptimal response. Present examination revealed multiple discrete hyperpigmented to violaceous papules and plaques over forehead, dorsum of both hands, and forearms in a photo-distributed pattern [Figures 1 and 2]. Differential diagnoses of CAD, polymorphous light eruption, and photoallergic contact dermatitis were considered clinically. Patch test with Indian standard series and photo patch test were negative compared to control. Skin biopsy from a representative lesion was performed, which showed dense dermal lymphoid infiltrate in the background of acanthosis, dermal fibroplasia, and focal epidermotropism [Figure 3a and 3b]. The infiltrate was composed of atypical lymphoid cells showing nuclear membrane irregularity and moderate nuclear pleomorphism [Figure 3c]. Pautrier's microabscess in the epidermis by these atypical lymphocytes was noted without spongiosis [Figure 4a and b]. On immunohistochemistry, these atypical cells showed positivity for CD3 and CD4 immunostain, thus confirming T-cell origin [Figure 5a and b]. These cells further showed partial loss of CD7. CD4: CD8 ratio was 4:1. Routine haematological investigations including peripheral blood smear and biochemistry was normal and whole-body PET scan did not reveal any subclinical metastasis. The patient was diagnosed as a case of mycosis fungoides (Clinical stage 1a) and managed with PUVA phototherapy with topical steroid application.

MF is a great imitator with several atypical morphological patterns. However, photosensitivity is rarely observed in MF. In fact, only six isolated reports of MF with photosensitivity were found in the preceding literature.[3] CAD is a common photo dermatosis that presents as a persistent dermatitis predominantly affecting photo exposed areas with abnormal photosensitivity to predominantly the ultraviolet B spectrum of wavelength. Its presentation varies from the most severe pseudo lymphomatous AR to milder variants like photosensitive eczema and persistent light reactors.[4] While our patient lacked the persistent infiltrated skin lesions of the pseudo lymphomatous variant AR; with the coexisting photosensitivity, the

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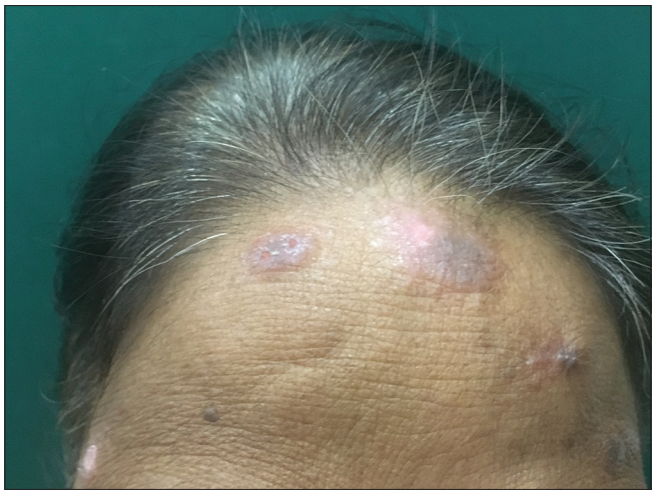

Figure 1: Erythematous to violaceous papules and plaques over forehead

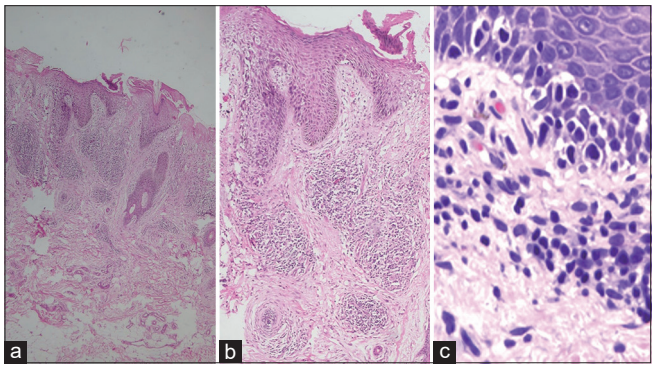

Figure 3: Dense dermal lymphoid infiltrate in the background of acanthosis, dermal fibroplasia and focal epidermotropism (a (H and E stain, × 10), and b (H and E stain, ×40)), which was composed of atypical lymphoid cells (3c (H and E stain ×100))

lesions were consistent with the clinical description of CAD.

Histologically, it may be difficult to differentiate between MF and pseudo lymphomatous variants of CAD, as epidermotropism and atypical lymphocytes can be seen in both, particularly in AR.[5] The presence of Pautrier's microabscesses, disproportionate epidermotropism, larger epidermal lymphocytes than dermal lymphocytes, alignment of lymphocytes along the basal layer of the epidermis, and the presence of intraepidermal cerebriform atypical lymphocytes are the most important features in distinguishing MF from CAD and other inflammatory dermatoses.[6] On the contrary, histological features favoring a diagnosis of CAD over MF include epidermal spongiosis and exocytosis, with the dermis containing large activated lymphocytes with eosinophils, plasma cells, and prominently increased multinucleated dermal dendrocytes.[7] Additionally, IHC can further differentiate that these two entities as a predominance of CD4 cells is seen in the skin biopsies of MF as opposed to predominantly CD8 lymphocytes in CAD.[5] Though CD8+ phenotype has been reported in less than 5% cases of MF, the clinical course

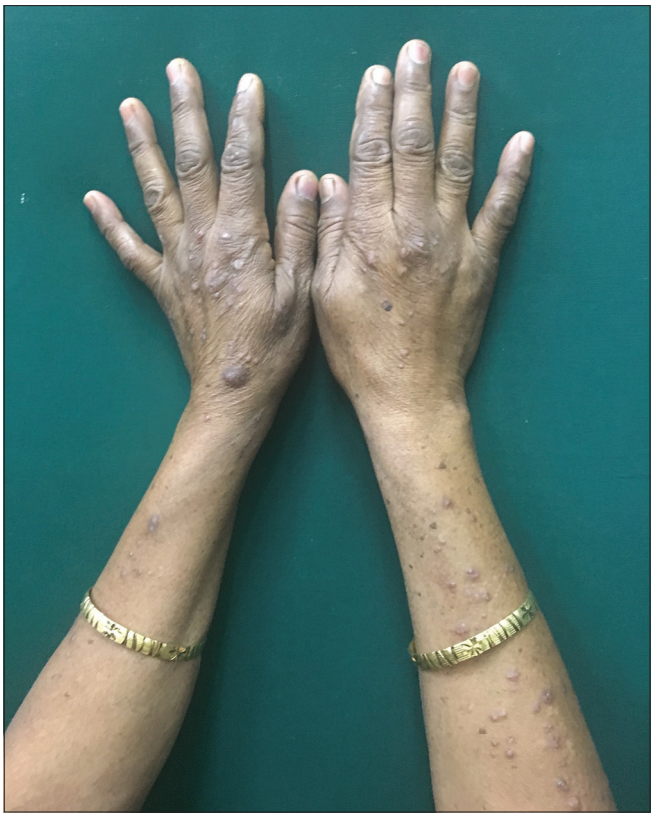

Figure 2: Discrete to confluent hyper pigmented papules and plaques over dorsa of both upper extremities

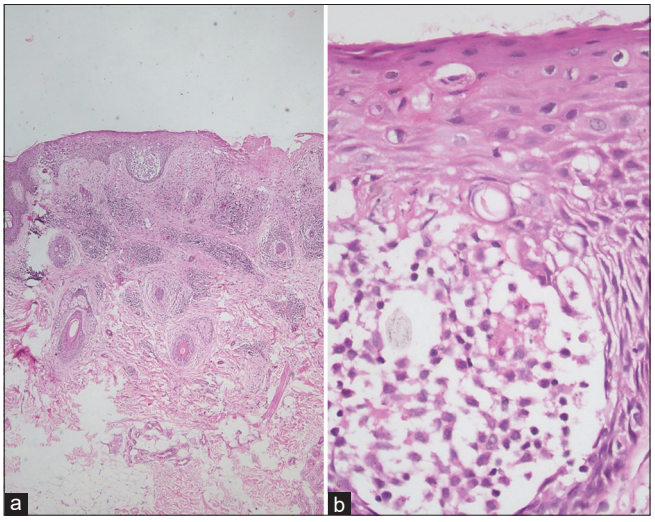

Figure 4: Pautrier's microabscess seen in the epidermis formed by these atypical lymphocytes without spongiosis (a (H and E stain, ×10), and b (H and E stain, ×100))

is more aggressive in this subset of patients.[8] In our case, the lymphocytic skin infiltrate consisted predominantly of CD4 T cells on IHC along with dense dermal lymphocytic infiltrate with focal epidermotropism on histopathology. T-cell receptor gene arrangement can be useful in differentiating neoplastic from reactive inflammatory lymphocytic infiltrates[9] but could not be done in our case due to nonavailability.

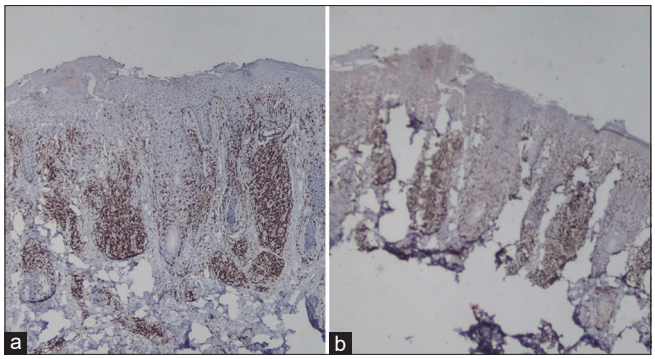

Figure 5: CD3 (a (IHC, × 100)) and CD4 positivity (b (IHC, × 100))

Though lymphomas have been occasionally reported in the chronic cases of CAD, long-term follow-up studies have shown no increased risk of lymphoma or other malignancies in cases of CAD.[10] Also, the frequency of positive patch and photo patch tests is high in CAD and a strong association between sensitivity to compositae plant species and CAD has been well documented.[11] Conversely, patch and photo patch tests are negative in MF as was observed in our case.

The management options for MF depend on the stage of the disease with skin-directed therapy with topical agents like steroids, nitrogen mustard, carmustine, retinoids, and phototherapy in early stages (stage Ia- IIa) and electron beam irradiation radiotherapy, immunotherapy, monoclonal antibodies, and oral retinoids in advanced stages of disease (IIB-IV).[12] Our patient had disease limited to the skin and hence was managed with topical steroids and phototherapy.

The case highlights a rare variant of MF associated with photosensitivity, with a total of only six cases in the literature prior to this report. The treating physician needs to have a high index of suspicion, keeping MF as a differential in any persistent, progressive, and treatment refractory "photosensitive" lesion, for early diagnosis.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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