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Psoriasiform Mycosis Fungoides Involving the Face- A Rare Case Report

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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Case Report

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ABSTRACT

Mycosis fungoides is represented as the most common epidermotropic cutaneous T-cell lymphoma, which is mainly characterized by the proliferation of atypical cells within the epidermis. We report a rare presentation of mycosis fungoides in a 60-year-old male presenting with chronic psoriasiform plaque involving the face. Punch biopsy of the lesion from the forehead was taken for routine histological examination and immunohistochemical stains. Results of biopsy and immunohistochemical findings were consistent with mycosis fungoides and diagnosed as psoriasiform presentation of mycosis fungoides involving the face.

Keywords: Mycosis fungoides; persistent facial plaque; epidermotropism; atypical lymphocytes.

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1. INTRODUCTION

Mycosis fungoides is a primary cutaneous T-cell lymphoma that accounts for 50% of all cutaneous lymphoma. It primarily affects the skin and can present with various progressive lesions such as patches, plaques, tumors, or erythroderma [1,2,3]. It is also called as Alibert-Bazin syndrome or granuloma fungoides [1].
It mainly affects the male population with onset between 45 to 60 years of age-group. MF usually manifests with diffuse, scaly erythematous patches and plaques which can progress into cutaneous tumors, generalized erythroderma, or lymph node involvement [2].

The cutaneous manifestations are mostly seen over the covered areas such as the trunk and body folds. Patch or plaque stage of lesions involving the sun-exposed parts like face and scalp is a very rare presentation in mycosis fungoides. The tumor stage of MF is the presenting sign in only 10% of cases mainly involving the face and may lead to leonine facies [2].

2. CASE REPORT

A 60-year-old male patient came to dermatology OPD with chief complaints of persistent raised thickened pigmented lesion over the face for the past 4 years. The Patient was normal before 4 years followed by which he developed pruritic flat pigmented lesions initially over the forehead and then the lesions were gradually progressed to elevated lesions involving other sites of the face with increased itching.

The patient also had a history of recurrent episodes of severe itching for the past 4 years which were not relieved by oral anti-histamines. No history of similar lesions in the family members and no history of any drug intake. No history of any mucosal involvement.

On cutaneous examination of lesions, the patient has single, non-tender, ill-defined, mild scaly, diffuse infiltrated hyper-pigmented plaque was seen over the forehead, and multiple infiltrated hyper-pigmented patches involving the nose and sides of the face (Fig 1 & Fig 2A, B, C). No scalp and oral involvement. No nail changes on examination. Lymph nodes are not palpable and systemic examination was normal.

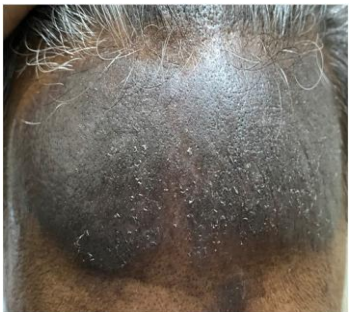

Fig. 1. Hyper-pigmented diffuse plaque over the forehead

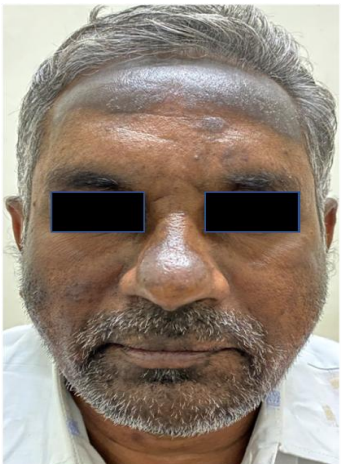

Fig. 2A. Hyperpigmented diffuse plaque over the forehead and multiple hyperpigmented patches over the nose and sides of the face

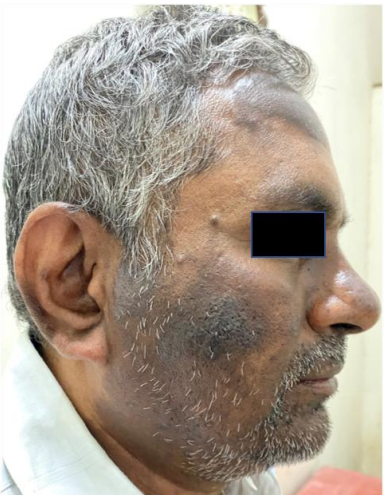

Fig. 2B. Multiple hyperpigmented patches and plaque over right side of face

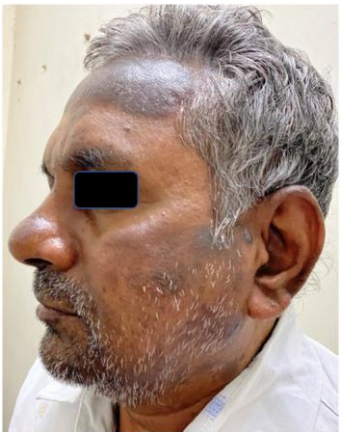

Fig. 2C. Multiple hyperpigmented plaque and patch over the left side of the face

A provisional diagnosis of psoriasis Vulgaris was made and started on topical tacrolimus 0.1% and topical corticosteroids on alternate days. But the lesions were persistent and did not respond to any topical treatments.

Dermoscopy of facial plaque showed multiple white rosettes, which is mainly suggestive of mycosis fungoides.

Hence, a 3.5mm punch biopsy was taken from the plaque over the forehead. On histopathological examination, it showed epidermotropism of atypical lymphocytes with pautrier's microabscess and some cells showed cerebriform nuclei and clear cytoplasm (Fig3A&B).

Immunohistochemistry showed CD3, CD4, CD7,and CD8 positive (Fig 4). Hence, the findings of biopsy and immunohistochemistry were consistent with mycosis fungoides.

Hence, a final diagnosis of psoriasiform mycosis fungoides was made. We started the patient on systemic methotrexate 7.5mg/ week and PUVA therapy twice weekly with topical corticosteroids and topical tacrolimus 0.1 %.

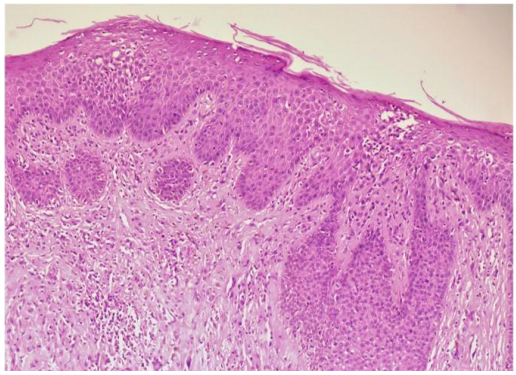

Fig. 3A. HPE: on scanner view showing epidermotropism and pautrier's microabscesses with atypical lymphocytes

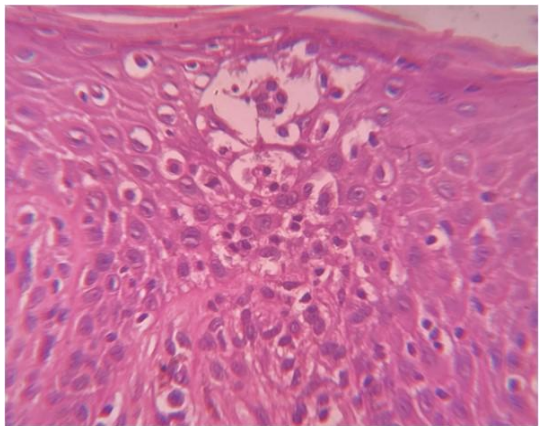

Fig. 3B. HPE: on high power view showing epidermotropism & pautrier's microabscesses with atypical lymphocytes

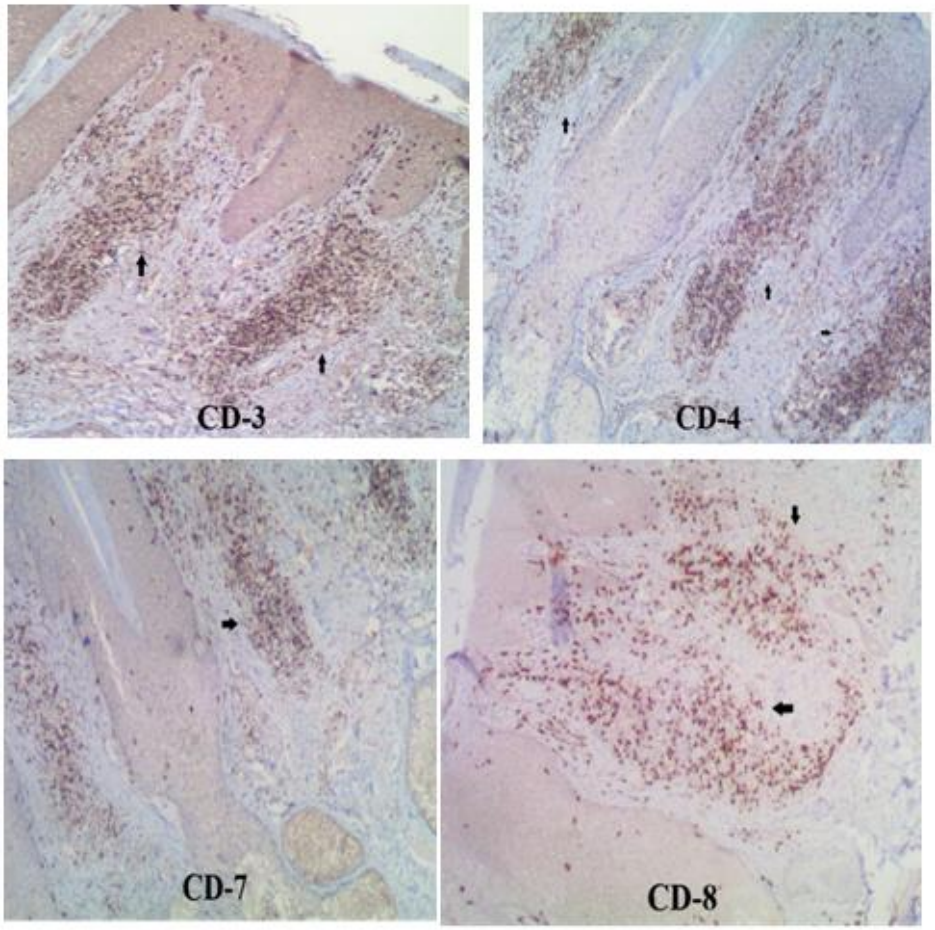

Fig. 4. IHC CD3 , CD4 , CD7 ,CD8 Positivity

3. DISCUSSION

Mycosis fungoides (MF) is majorly considered as rare, non-Hodgkin's lymphoma, which involves sites other than lymph nodes with clonal, malignant T-lymphocytes with an incidence of 0.36 / 1,00,000 persons per year [1]. MF can be classified according to its clinical presentation as patch stage, plaque stage, tumor, and erythrodermic stage.

MF classically manifests with indolent cutaneous eruptions with Patch or plaque stage lesions mainly present over the covered parts of the body (trunk, breast, buttocks, medial thighs). But, the occurrence of plaque or patch stage of lesions over the face is a very rare presentation in mycosis fungoides. Patch and plaque stage lesions can present with hypopigmentation, hyperpigmentation, atrophy, or petechiae[1].

In addition to various clinical presentations of MF, it can be associated with severe pruritus or it could be asymptomatic. Patch stage of mycosis fungoides can progress to an infiltrated plaque with generalized distribution and the minority of cases may progress to the exophytic tumor[2]. Lymphadenopathy and visceral dissemination are usually presented as late occurrences [2].

Diagnosis of lesions during the early stages of MF is mainly difficult because it can be misdiagnosed as chronic contact dermatitis and chronic plaque-type psoriasis [4-6]. Diagnosis is mainly based on clinical aspects and it is always confirmed by skin biopsy and immunohistochemistry [7-8].

Histopathological features of mycosis fungoides mainly include epidermotropism (migration of atypical lymphocytes from the dermis into the epidermis within the basal cell layer), atypical dermal lymphocytes with cerebriform nuclei, pautrier's microabscesses, and grandiosity sign [2]. Classically, MF shows a CD2+, CD3+, CD4+, CD8–, CD30–, CD45RO+ immunophenotype [2].

Management of MF with topical therapies like topical potent-corticosteroids, topical cytotoxic agents like nitrogen mustard, topical retinoids like bexarotene, and PUVA therapy has been shown to improve the lesions during the early stage of MF. Systemic methotrexate and other targeted therapy like monoclonal antibodies, oral bexarotene, recombinant interferon-alpha therapy used in complicated stages of MF with systemic features [7-12].

Systemic methotrexate which was used to be given during the advanced stages of MF was given to our patient because of poor response with topical glucocorticoids alone.

4. CONCLUSION

Mycosis fungoides being considered as major dermatological masquerader with a wide spectrum of clinical manifestations mainly involving the covered parts of the body and involvement of the face is a very rare presentation mainly during the early stages of patch or plaque lesions. Hence, we hereby report a rare psoriasiform presentation of mycosis fungoides involving face disguised as chronic dermatitis and the importance of skin biopsy and immunohistochemistry in such cases.

CONSENT

As per international standard or university standard, patient's written consent has been collected and preserved by the author(s).

ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

REFERENCES

1. Mycosis Fungoides: Case Report and Literature Review: Akinbami et al. Mycosis Fungoides: Case Report and Literature Review. Clinical Medicine Insights: Case Reports 2014;7:95–98. DOI: 10.4137/CCRep.S15724.
2. A Rare Presentation of Mycosis Fungoides Mimicking Psoriasis Vulgaris S. Doukaki M. Aricò M.R. Bongiorno Department of Dermatology, University of Palermo, Palermo, Italy. Case Rep Dermatol. 2009;1:60–65. DOI: 10.1159/000249148.
3. The different faces of mycosis fungoides: Results of a single-center study Polytimi Sidiropoulou, Vasiliki Nikolaou, Leonidas Marinos, Dimitra Voudouri, Elena Komini, Afroditi Economidi, Dimitris Rigopoulos, Alexander Stratigos. The International Society of Dermatology; 2019. DOI: 10.1111/ijd.14735
4. Papathemeli D, Georgiou E, Koletsa T, et al. Mycosis fungoides in patients with psoriasis: an ongoing issue. Eur J Dermatol 2018;28:235–236.
5. Nikolaou V, Marinos L, Moustou E, et al. Psoriasis in patients with mycosis fungoides: a clinicopathological study of 25 patients. J Eur Acad Dermatol Venereol 2017;31:1848–1852.
6. Scarisbrick JJ, Quaglino P, Prince HM, et al. The PROCLIPI international registry of early-stage Mycosis Fungoides identifies substantial diagnostic delay in most patients. Br J Dermatol. 2019;181:350–357.
7. Willemze R. Mycosis fungoides variants-clinicopathologic features, differential diagnosis, and treatment. Semin Cutan Med Surg 2018;37:11–17.
8. Pope E, Weitzman S, Ngan B, et al. Mycosis fungoides in the pediatric population: report from an international childhood registry of cutaneous lymphoma. J Cutan Med Surg. 2010;14(1):1–6.
9. Marschalko M, Eros N, Kontar O, et al. Folliculotropic mycosis fungoides: clinicopathological analysis of 17 patients. J EurAcad Dermatol Venereol. 2015;29 (5):964-972.
10. Alberti-Violetti S, Talpur R, Schlichte M, et al. Advanced-stage mycosis fungoides and S_ezary syndrome: survival and response to treatment. Clin Lymphoma Myeloma Leuk. 2015; 15(6):e105-e112
11. Leonine facies (LF) and mycosis fungoides (MF): A single-center study and systematic review of the literature. Danielle Nicole Brown, BA, Iris Wieser, MD, Casey Wang, MD, Boutnaina S. Dabaja, MD, and Madeleine Duvic, MD. Houston, Texas,

and Salzburg, Austria. J Am Acad Dermatol. 2015;73:976-86.

12. Lim HLJ, Tan EST, Tee SI, et al. Epidemiology and prognostic factors for mycosis fungoides and Sezary syndrome in a multi-ethnic Asian cohort: a 12-year review. J Eur Acad Dermatol Venereol. 2019;33:1513–1521.

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