Received: 26 June 2023 | Revised: 28 November 2023 | Accepted: 7 December 2023

DOI: 10.1002/ccr3.8347

CASE REPORT

Clinical Case Reports Open Access WILEY

Lichenoid mycosis fungoides: Report of a case with lichen planus-like histopathologic features

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Key Clinical Message

Mycosis fungoides is a diagnostic challenge. Herein, we report a case with marked lichenoid features in pathology assessments. After several biopsies and clinicopathologic correlation, the diagnosis of lichenoid mycosis fungoides was made.

Abstract

Mycosis fungoides (MF) is a great imitator and mimicks other dermatoses clinically and histopathologically. We report a 61-year-old patient with 5-year history of generalized violaceous patches and plaques. His biopsy revealed a marked lichenoid band-like infiltrate of inflammatory cells along the basal layer with basal layer vacuolar changes; the diagnosis of lichen planus was first made histopathologically. Several biopsy specimens, clinicopathologic correlation, and immunohistochemistry findings confirmed the diagnosis of lichenoid MF. Awareness of peculiar histopathologic findings of MF is essential to avoid a potential misdiagnosis. When in doubt, multiple biopsies with other diagnostic methods should be employed.

KEYWORDS

histopathology, lichen planus, lichenoid, mimics, mycosis fungoides

1 | INTRODUCTION

Mycosis fungoides (MF) is the most common form of cutaneous lymphoma of T-cell origin. Classical MF manifests with patch, plaque, tumor, and erythroderma. However, this entity can exhibit a diversity of clinical and histopathological presentations. Lesions could mimic benign inflammatory dermatoses, including but not limited to eczema, pigmented purpuric dermatosis, psoriasis, and lichen planus (LP). Besides, several peculiar new variants have been described to date; follicular, hypopigmented, and ichthyosiform MF, to name a few.[1–3] All these can pose a diagnostic pitfall to dermatologists. Herein, we report a case with a rare variant of lichenoid MF subjected to multiple histopathological assessments.

2 | CASE PRESENTATION

A 61-year-old Middle Eastern male presented in our dermatology clinic with a history of generalized lesions. Before that, he was otherwise healthy and took no medications. The lesions appeared in the past 5 years and progressed in size and number. On examination, multiple erythematous and violaceous patches and plaques were scattered all over his body. Some of these poikilodermic

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lesions were scaly, but none had signs of pruritus and were clinically compatible with classic MF. (Figure 1).

Since the onset, he underwent several histopathologic evaluations sent with a provisional diagnosis of MF. Accordingly, four punch biopsies (one at the initial presentation, one at the time of exacerbation during Narrow Band UVB (NBUVB) therapy, and two when he returned to the hospital after 25 months) were undertaken. The results were inconclusive; he had the histopathologic diagnosis of LP, MF, MF, and LP, respectively, throughout the years. All of his biopsies showed findings resembling those of LP. At the last session, the same histopathologist reexamined

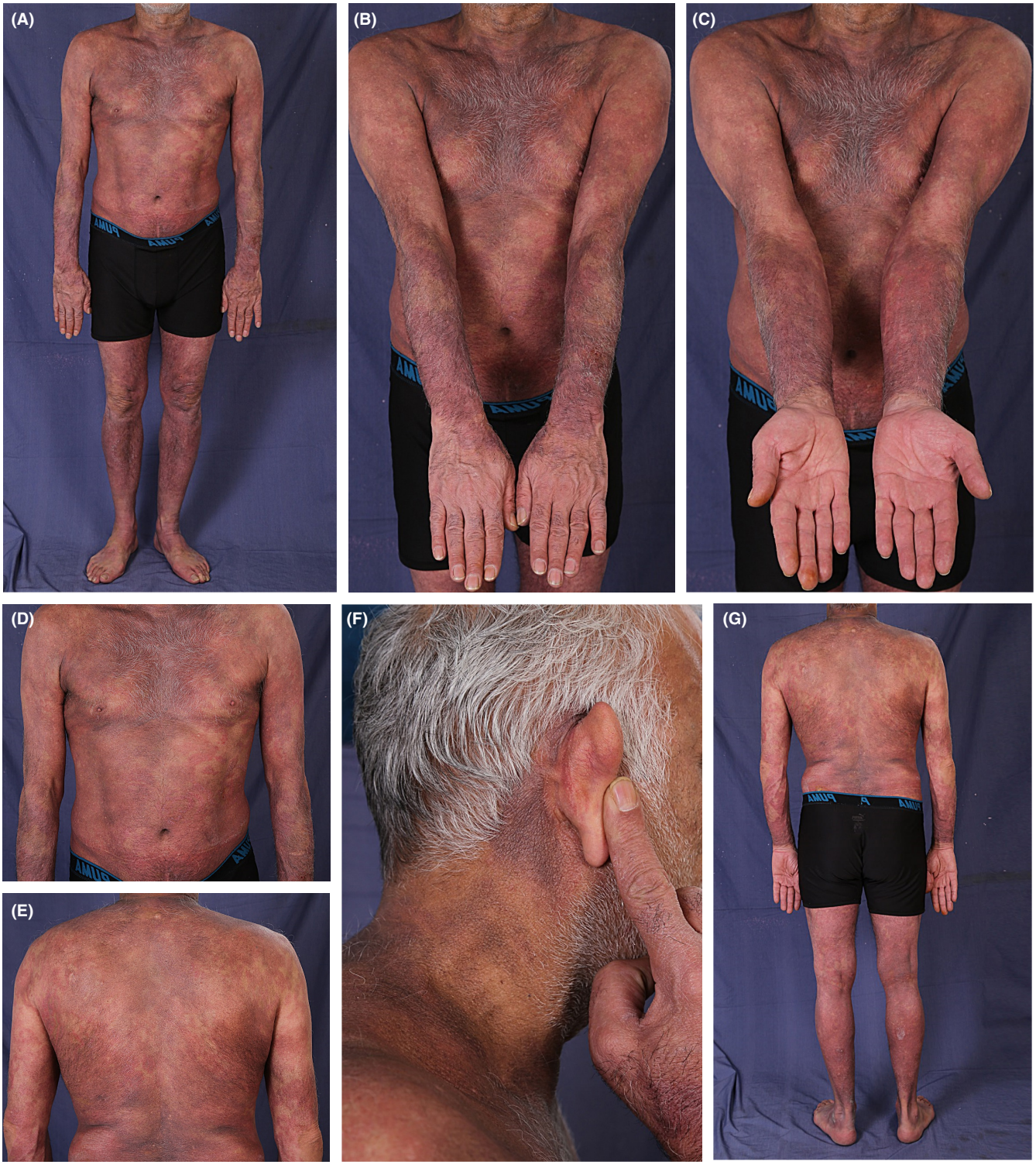

FIGURE 1 (A–G) The clinical presentation of a 61-year-old man with widespread erythematous to violaceous patches and plaques. Some lesions were poikilodermic, with the final diagnosis of lichenoid mycosis fungoides.

the slide and concluded the diagnosis of MF. Accordingly, histopathologic examination of skin biopsy from the back lesion revealed a lichenoid band-like infiltrate of inflammatory cells along the basal layer with basal layer vacuolar changes. There were some atypical lymphocytes in the basal layer with hyperchromatic nuclei and perinuclear halo, and scattered dyskeratotic cells. Also, some melanophages in the superficial dermis were identified (Figure 2A–E).

He was referred for further evaluation. Immunohistochemistry staining revealed a predominance of CD4+ lymphocytes rather than CD8+ lymphocytes and decreased CD5 and CD7 lymphocytes among intraepidermal lymphocytes. No Sézary cell was seen on flow cytometric evaluation. There was no organomegaly or lymphadenopathy in the abdominopelvic sonography or spiral computed tomography scan (CT scan). T-cell receptor gene rearrangement revealed a clonal band of amplification favoring the neoplastic process.

Following the clinicopathological and molecular assessments, a diagnosis of lichenoid MF with a stage IIA (T2N1M0B0) was rendered and subsequently treated with NBUVB three times a week. Lesions resolved significantly in 8 months after the NBUVB initiation with remaining hyperpigmented patches, implying the response of the lichenoid MF to phototherapy (Figure 3A–H). At this writing, the patient was undergoing maintenance NBUVB therapy once weekly. The patient consented to report his images and information.

3 | DISCUSSION

Besides the classic presentation of patches, plaques, and tumors, MF may show atypical features mimicking other dermatologic entities clinically and histopathologically, making it known as the great masquerader.[4–6]

Herein, we presented a case with clinical manifestations of MF, whose histopathology assessments revealed marked LP features reported as LP by various dermatopathologists despite MF being the first clinical impression by dermatologists. After four subsequent biopsies with histopathological diagnoses of LP and MF, we could not reach a definite diagnosis. Clinical presentations and immunohistochemical evaluations helped us distinguish lichenoid MF from its major histopathological simulant, LP.

Lichenoid MF is a rare histopathological variant of MF that presents as a lichenoid band-like infiltrate of inflammatory cells, typical of LP. Very few cases of lichenoid MF have been reported in the literature.[3,7,8] A study investigating the clinical characteristics of 223 Korean patients with MF found 3 (1.3%) cases with a lichenoid subtype of MF; all were middle-aged women with early-stage MF who had the symptoms for over 13 years.[9] Another study reported a middle-aged woman with a typical presentation of a papulosquamous form of syphilis whose lesions were accompanied by poikilodermatous plaques refractory to syphilis medications.

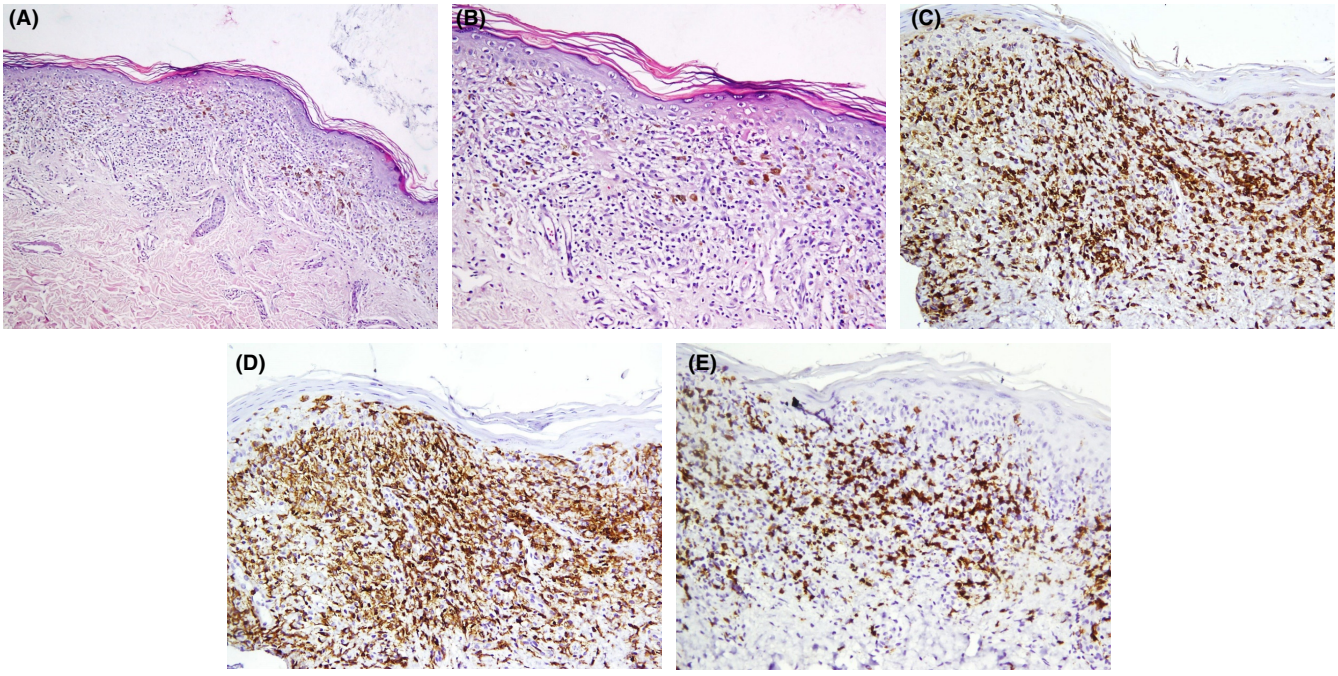

FIGURE 2 Histopathologic examination revealed band-like lichenoid lymphocytic infiltrate along the basal layer (A. H&E X40). Some atypical basally-located lymphocytes with perinuclear halo (B. H&E X200). Immunohistochemistry study revealed CD3-positive intraepidermal lymphocytes (C) and predominance of CD4 positive intraepidermal lymphocytes (D) rather than CD8 positive intraepidermal lymphocytes (E).

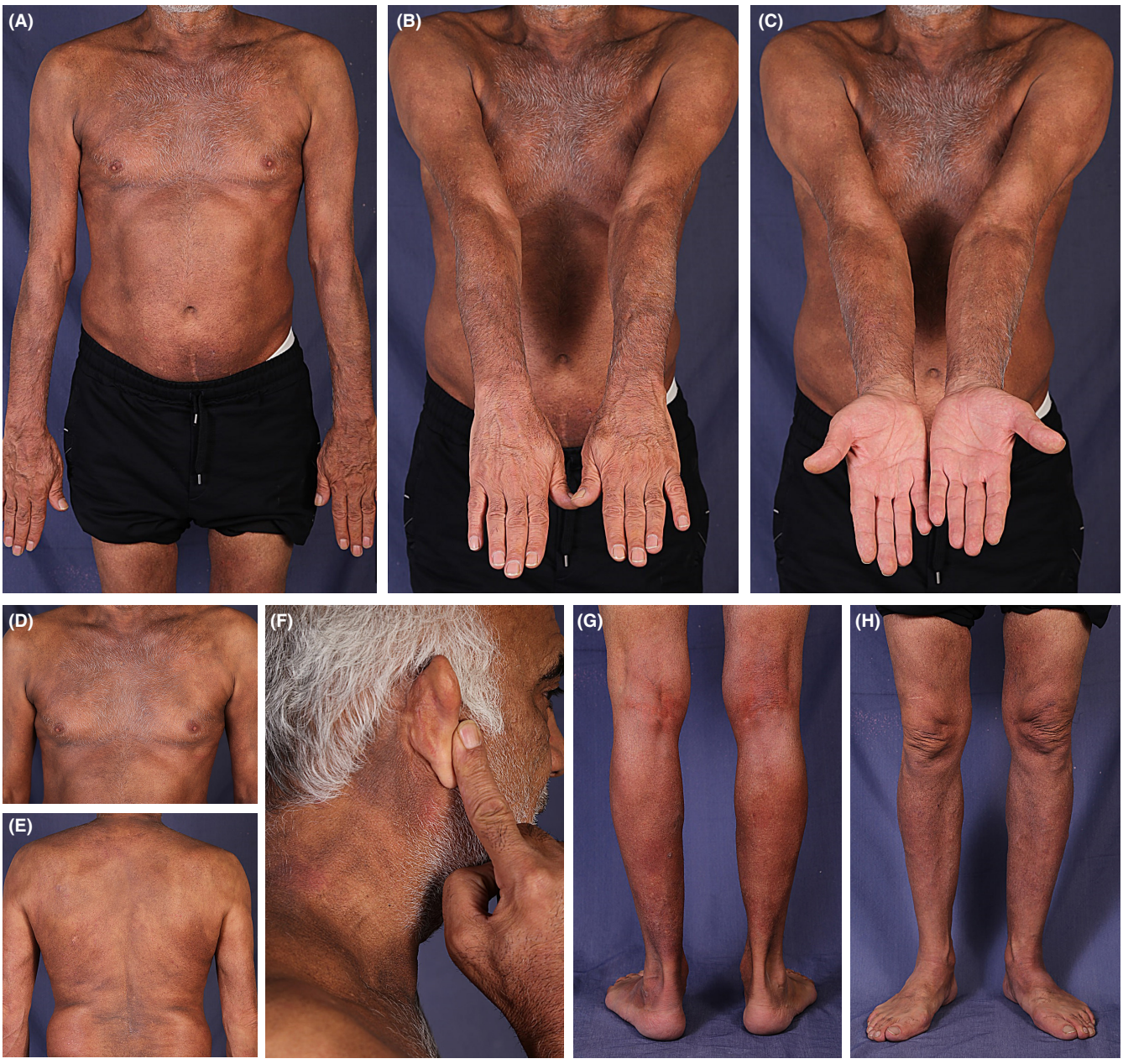

FIGURE 3 (A–H) The patient's clinical images after 8 months of narrowband UVB therapy. Notice the hyperpigmented patches (post-inflammatory hyperpigmentation) and the response of his lichenoid MF to the treatment.

Further biopsies after 2 years showed band-like lymphocyte-rich inflammation at the dermo-epidermal junction, supportive of MF. She responded to acitretin with plaques resolution in 3 months.[3]

A case series reported that 50% of their cases with lichenoid MF experienced an accelerated course of the disease, implying a poor prognosis for this variant.[7] Contrarily, based on another study, the disease status of three reported lichenoid MF was complete and partial remission of the disease.[9] Our case also responded to the treatment and did not show a progressive course after the 5 years since the start of his disease and after the 2 years following the start of his treatment.

Histopathologic diagnosis of early stages MF is challenging and can easily be misdiagnosed with other inflammatory dermatoses. The histopathological characteristics of early stages MF often reveal a patchy lichenoid or band-like infiltrate; however, some features like dermal fibrosis, haloed lymphocytes, and basilar epidermotropism would be a clue in the diagnosis of MF.[10] Nevertheless, marked lichenoid band-like infiltrate of inflammatory cells along the basal layer may seriously hinder the diagnosis of MF. Lichenoid presentations may be caused by a cell-mediated immune response provoked by the T cells, resulting in the cytotoxicity of target keratinocytes. This histological picture of lichenoid

MF may be confused with benign dermatoses like LP, same as our case. It is the time when other diagnostic methods, together with several biopsy-taking, enter the game. The finding of a monoclonal T-cell receptor gene rearrangement becomes necessary for diagnostic accuracy. MF generally has an overwhelming predominance of CD4+T cells in the infiltrate, as was found in our case.[7] Moreover, clinicopathologic correlation is mandatory to establish a proper diagnosis in the early stages of MF to have a good prognosis.

This case report highlights how peculiar findings of MF should be tightly followed up with sequential biopsies. It also demonstrated the importance of undertaking immunohistochemistry methods where indicated.

In conclusion, lichenoid MF is a rare variant of cutaneous T-cell lymphoma. The histopathology of this variant showed a marked lichenoid band-like infiltrate, mimicking LP. This can pose a significant diagnostic challenge to dermatologists and pathologists. Clinical presentations, histopathology features, and occasionally immunohistochemistry assessments are paramount for correct and early diagnosis.

AUTHOR CONTRIBUTIONS

Nika Kianfar: Conceptualization; data curation; writing – original draft. **Alireza Ghanadan:** Investigation; methodology; resources. **Zeinab Aryanian:** Validation; visualization. **Ifa Etesami:** Conceptualization; project administration; writing – review and editing.

FUNDING INFORMATION

None.

DATA AVAILABILITY STATEMENT

The data are available upon reasonable request from the corresponding author.

CONSENT

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

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How to cite this article: Kianfar N, Ghanadan A, Aryanian Z, Etesami I. Lichenoid mycosis fungoides: Report of a case with lichen planus-like histopathologic features. *Clin Case Rep*. 2023;11:e8347. doi:10.1002/ccr3.8347