Unmasking a T cell lymphoma: Folliculotropic mycosis fungoides with a gamma-delta phenotype

Jessica Tran, BBA,[a] Francisco Vega, MD, PhD,[b] and Madeleine Duvic, MD[a]
Houston, Texas

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INTRODUCTION

Mycosis fungoides (MF) with a gamma-delta cell lineage is a poorly understood T cell lymphoma that seems to be different from classic primary cutaneous gamma-delta T cell lymphoma.[1] Here, we describe a rare case of clinically consistent and biopsy-proven folliculotropic MF with a gamma-delta T cell phenotype that was locally exacerbated by the use of a face mask.

Abbreviation used:

MF: mycosis fungoides

CASE PRESENTATION

In November 2018, a 52-year-old female began working as an operating room nurse. Within a week, she began developing intensely pruritic red-brown plaques with a peau d'orange appearance on her cheeks, which was worse on the left cheek (Fig 1).

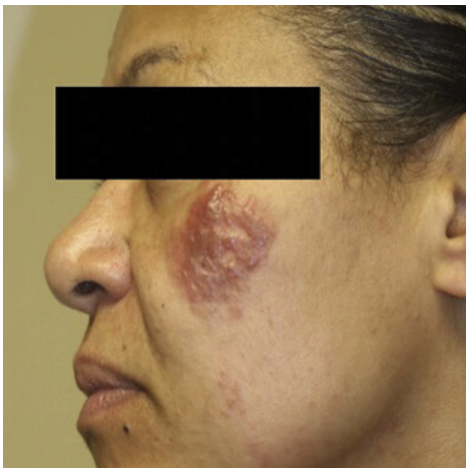

Fig 1. Folliculotropic MF with a gamma-delta phenotype. Red-brown plaques with a peau d'orange appearance on the left cheek. *MF*, Mycosis fungoides.

The lesions persisted for several months. During this period, the patient went on vacation for several days and noted that the lesions cleared almost completely. However, upon returning to work, the lesions progressed again. Three dermatologists diagnosed allergic contact dermatitis and prescribed

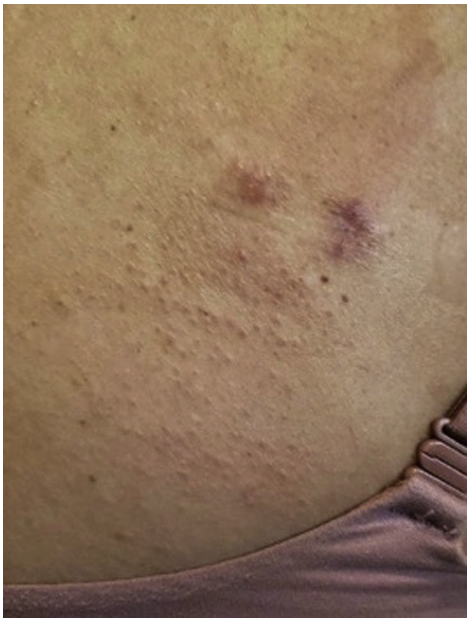

Fig 2. Folliculotropic MF with a gamma-delta phenotype. Red-brown papules and plaques on the posterior aspect of the right trunk. *MF*, Mycosis fungoides.

From the Department of Dermatology[a] and the Department of Hematopathology,[b] The University of Texas, MD Anderson Cancer Center.

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Correspondence to: Jessica Tran, BBA, 3400 Montrose Blvd, Houston, TX 77006. E-mail: jessica.tran@bcm.edu.

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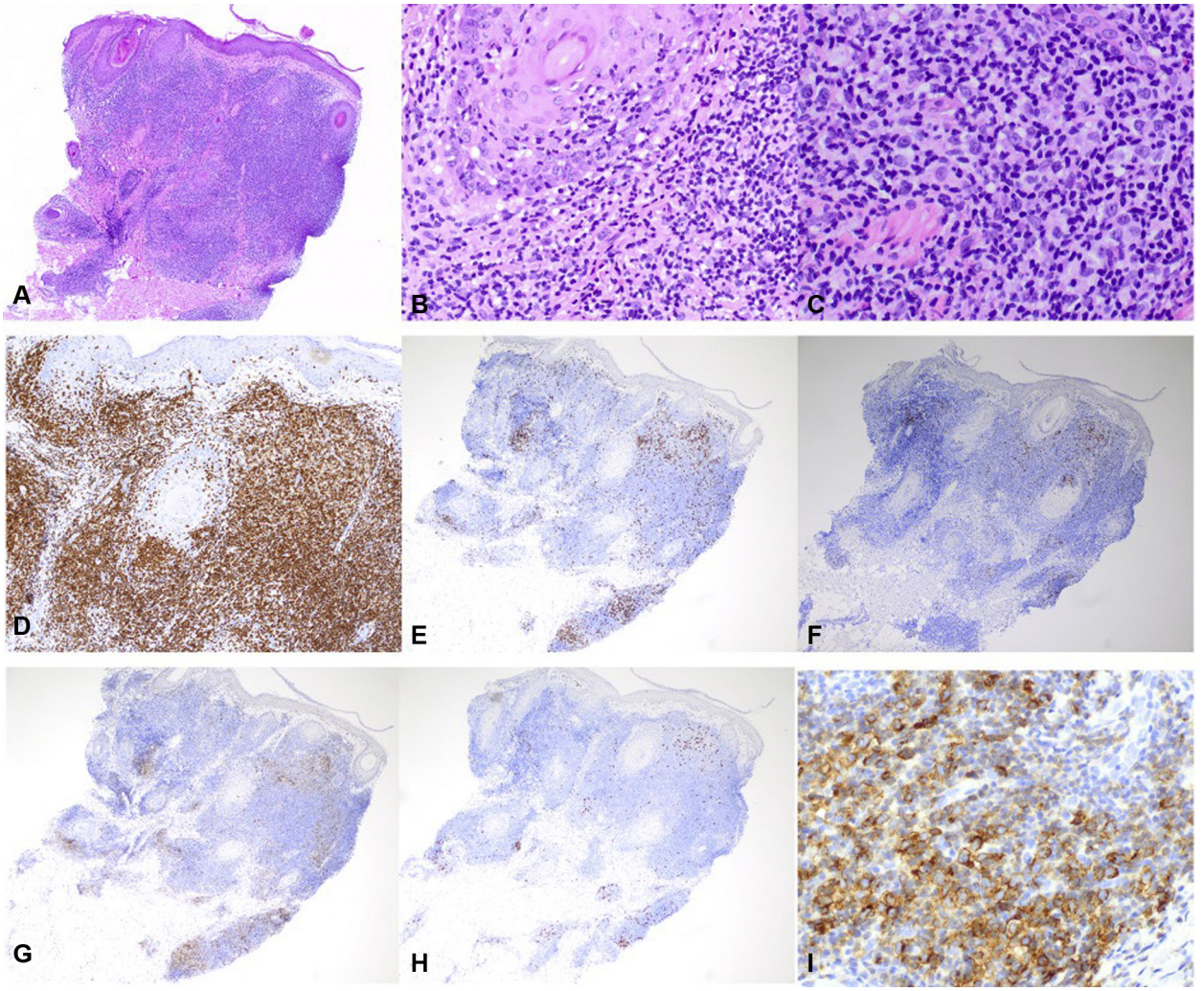

Fig 3. Folliculotropic MF with a gamma-delta phenotype from skin biopsy of left cheek. **A**, Dense diffuse atypical lymphoid infiltrate in the dermis. **B**, Prominent folliculotropism in addition to focal epidermotropism. **C**, The infiltrate is composed mostly of intermediate atypical lymphoid cells with scattered large lymphoid cells. **D**, The tumor cells strongly express CD3. Note that CD3 highlights the folliculotropism. The tumor cells have the following aberrant immunophenotype: $CD5^-$ (**E**), $CD7^-$ (**F**), $CD4^-$ (**G**), and $CD8^-$ (**H**). **I** CD30 highlights a small subset of the large lymphoma cells and a subset of the small lymphoid cells. The expression and pattern of CD30 is not the expected for $CD30^+$ lymphoproliferative disorders involving the skin. *MF*, Mycosis fungoides.

various topical steroids and oral antibiotics for the patient, none of which were effective. The patient reported that the only time the lesions improved was when she took time off from work and did not wear a face mask for approximately 1 week. Eventually, the lesions spread to her left arm and right posterior trunk (Fig 2).

A biopsy of the 42 × 38-mm^2 plaque on the left cheek was performed in June 2019, and subsequent biopsies of plaques on her left arm and posterior trunk were performed in August 2019. Histological sections (Figs 3 and 4) demonstrated neoplastic T cells that varied in size, with irregular nuclear contours, epidermotropism, and perivascular but mainly folliculotropic distribution. The tumor cells were $CD3^+$, $CD4^-$, and $CD8^-$.[2] T cell receptor rearrangement testing on the specimens from the posterior trunk and arm were $TCRdelta^+$ and $TCRbeta^-$ (BF1). Cytologic atypia, in combination with the aberrant immunophenotype of the cells, supported a neoplastic T cell population. These pathologic findings in combination with the clinical features supported the diagnosis of folliculotropic MF with a gamma-delta T cell phenotype.

In October 2019, the patient was referred to MD Anderson Cancer Center for treatment and exhibited

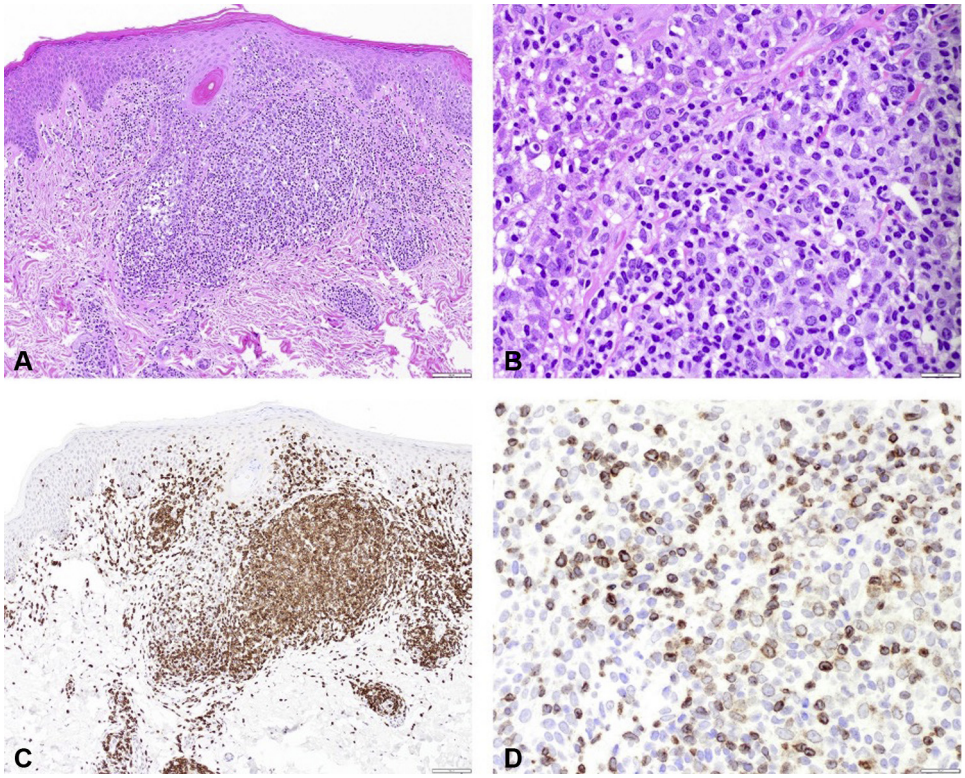

Fig 4. Folliculotropic MF with a gamma-delta phenotype from skin biopsy of the left arm. **A** The neoplastic infiltrate exhibits variable epidermotropism, folliculotropism, and perivascular distribution. **B** The neoplastic cells are variable in size, ranging from small to scattered intermediate to large. **C** The tumor cells express CD3. **D** The tumor cells are variably TCRdelta^{+} and CD30^{+} and TCRbeta^{-} (BF1), CD4^{-}, CD5^{-}, CD7^{-}, CD8^{-}, CD20^{-}, CD56^{-}, TIA1^{-}, granzyme B^{-}, and EBER^{-} (not shown). Note that the smaller tumor cells have irregular nuclear contours as highlighted by the TCRdelta immunostain. Also, note that the intermediate to large neoplastic cells have dimmer expression of TCRdelta. *MF*, Mycosis fungoides.

complete clearance of her cheek lesions following local radiation and treatment with oral bexarotene. She was transferred out of the OR and subsequently remained in clinical remission for 4 months.

However, in the wake of the current Coronavirus disease 2019 pandemic, the patient began wearing a 3M N95 mask for short periods of time and reported that she felt a burning sensation on her cheeks within 15 minutes of wearing the mask. Within 1 week, she developed erythematous and indurated nodules on her cheeks. The patient was advised to use cloth masks until her follow-up visit, and topical tacrolimus was prescribed. Within 2 months of treatment, the lesions improved significantly.

DISCUSSION

The temporal relationship between exposure to a face mask and the subsequent worsening of the patient's cheek lesions on multiple occasions is unusual. However, this may be coincidental. There have been reports of antigens, such as those from *Staphylococcus aureus*, causing local expansion or homing of neoplastic T cells in patients with cutaneous T cell lymphoma.[3] However, in the case of this patient, the spread of the lesions to her trunk and extremities does not support the theory of a local stimulus. Additionally, a reaction to a face mask would more likely indicate a reactive disease rather than a malignancy. Therefore, it is possible that some other factor may be the cause.

Although folliculotropic MF with a gamma-delta phenotype has a much more favorable prognosis than classic primary cutaneous gamma-delta T cell lymphoma (28.6% vs. 67% mortality), there is a subset within the former that exhibits aggressive behavior.[1]

Because of the potentially aggressive nature of folliculotropic MF with a gamma-delta phenotype, early diagnosis and treatment are imperative.[1]

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