Progressive Erythematous Facial Rash

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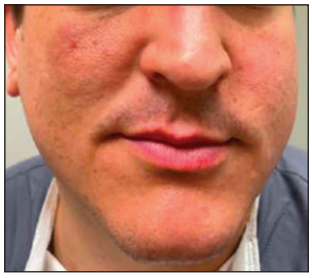

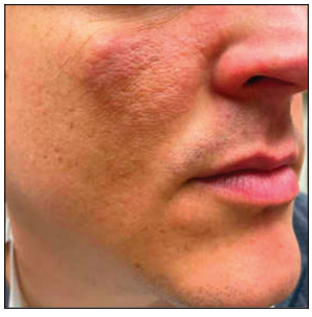

A 32-year-old man presented to the dermatology clinic for evaluation of a progressive erythematous facial rash of 4 years' duration. The patient reported some worsening with increased face mask wear during the COVID-19 pandemic. On occasion, fluid could be expressed when the area on the right cheek was compressed. Physical examination revealed a well-demarcated erythematous plaque on the right cheek. The patient also reported intermittent mild involvement of the nose and left cheek. He initially was treated with triamcinolone and ketoconazole cream for several months, but the rash persisted. Given the chronicity and worsening of the eruption, a punch biopsy from the right cheek with immunohistochemical staining was obtained.

WHAT'S YOUR **DIAGNOSIS?**

a. contact dermatitis
b. cutaneous lupus
c. follicular mucinosis
d. rosacea
e. seborrheic dermatitis

PLEASE TURN TO PAGE E53 FOR THE DIAGNOSIS

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THE **DIAGNOSIS**: Follicular Mucinosis

Histologic examination of the hematoxylin and eosin–stained sections of the biopsy revealed an overall moderately dense, perivascular, and perifollicular lymphocytic infiltrate with follicular intraepidermal mucin (Figure). Immunohistochemical staining showed that the lymphocytic infiltrate was predominantly CD4+ over CD8+, with moderate loss of CD7 and absence of CD20 expression. Positive T-cell receptor (TCR) gene rearrangements were detected for both TCRγ and TCRβ. The clinical features along with the histopathologic findings suggested a diagnosis of follicular mucinosis (FM) with concern in the differential for folliculotropic mycosis fungoides.

Follicular mucinosis, also known as alopecia mucinosa, is an uncommon inflammatory disorder characterized by follicular degeneration due to the accumulation of mucin within the pilosebaceous unit.[1] This condition manifests clinically as indurated plaques and/or follicular papules most often on the face, neck, and scalp.[2] It is further categorized as primary vs secondary FM. Primary idiopathic FM, which can further be subdivided into acute or chronic, tends to follow a more benign course, whereas secondary FM usually is associated with underlying inflammatory or neoplastic conditions, most commonly mycosis fungoides, a cutaneous T-cell lymphoma.[1,2] In cases of secondary FM, treatment of the underlying cause often leads to resolution of symptoms. Regular follow-up is warranted in either classification.[1,3]

The initial differential diagnosis for this patient included contact dermatitis associated with mask use, with possible underlying seborrheic dermatitis or rosacea; however, the rash persisted and worsened after treatment with topical triamcinolone and ketoconazole. After the diagnosis of FM was made, the patient was started on topical betamethasone and tacrolimus with good response.

A referral to hematology/oncology revealed that the patient had primary FM and possible stage 1A folliculotropic mycosis fungoides with limited skin involvement (<10% body surface area). On physical examination, no palpable cervical or axillary lymphadenopathy were noted. Flow cytometry for lymphoma was negative with no lymphoid or blast population detected. Laboratory workup and positron emission tomography/computed tomography were unremarkable. The patient had rapid improvement with a more potent topical steroid but also was given tacrolimus ointment 0.1% for residual findings. His disease remained stable without progression at 1-year follow-up.

Contact dermatitis typically manifests as an eczematous eruption that appears on an anatomic location that was exposed to or came into contact with allergens or irritants.[4] Contact dermatitis was less likely in our patient due to the lack of acute or subacute spongiosis and lymphocyte exocytosis. Rosacea is a chronic inflammatory dermatosis that presents as recurrent episodes of flushing or transient erythema, persistent erythema, phymatous changes, papules, pustules, and telangiectasia[5]; however, rosacea was less likely in our patient due to the histopathologic and immunohistochemical findings that were suggestive of FM on punch biopsy. Cutaneous lupus generally is associated with photosensitivity and manifests as erythema over the malar eminences and bridge of the nose with sparing of the nasolabial folds.[6] Seborrheic dermatitis manifests as erythematous macules or patches with scale and associated pruritis on the scalp, eyebrows, eyelids, and nasolabial folds.[7] This condition was less likely in our patient due to the persistence and worsening of the facial erythematous dermatitis despite the use of ketoconazole cream as well as no evidence of spongiosis, shoulder parakeratosis, vascular changes, or presence of microorganisms such as *Malassezia* species.

Due to the relatively rare nature of this condition as well as a wide variety of other more common etiologies for an erythematous dermatitis of the cheeks, the diagnosis of FM may be delayed or missed entirely. Physicians must have a high index of suspicion to diagnose properly and

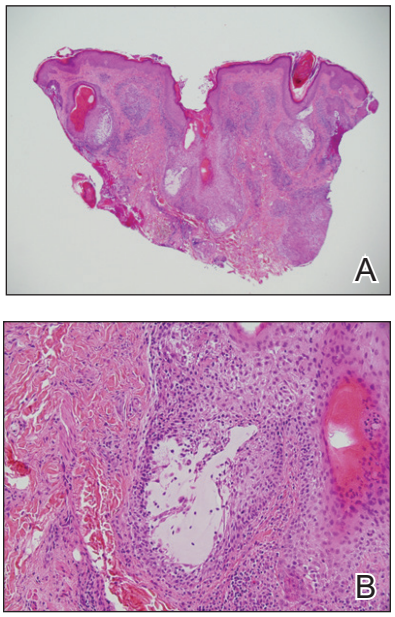

FIGURE. A and B, Moderately dense, perivascular, and perifollicular lymphocytic infiltrate with follicular intraepidermal mucin (H&E, original magnification ×4 and ×20).

biopsy if necessary. This photoquiz serves as an important reminder to physicians to keep uncommon diseases on their differential, especially when the patient's symptoms do not respond to treatment.

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