Facial milia-like eruption in a patient with alopecia

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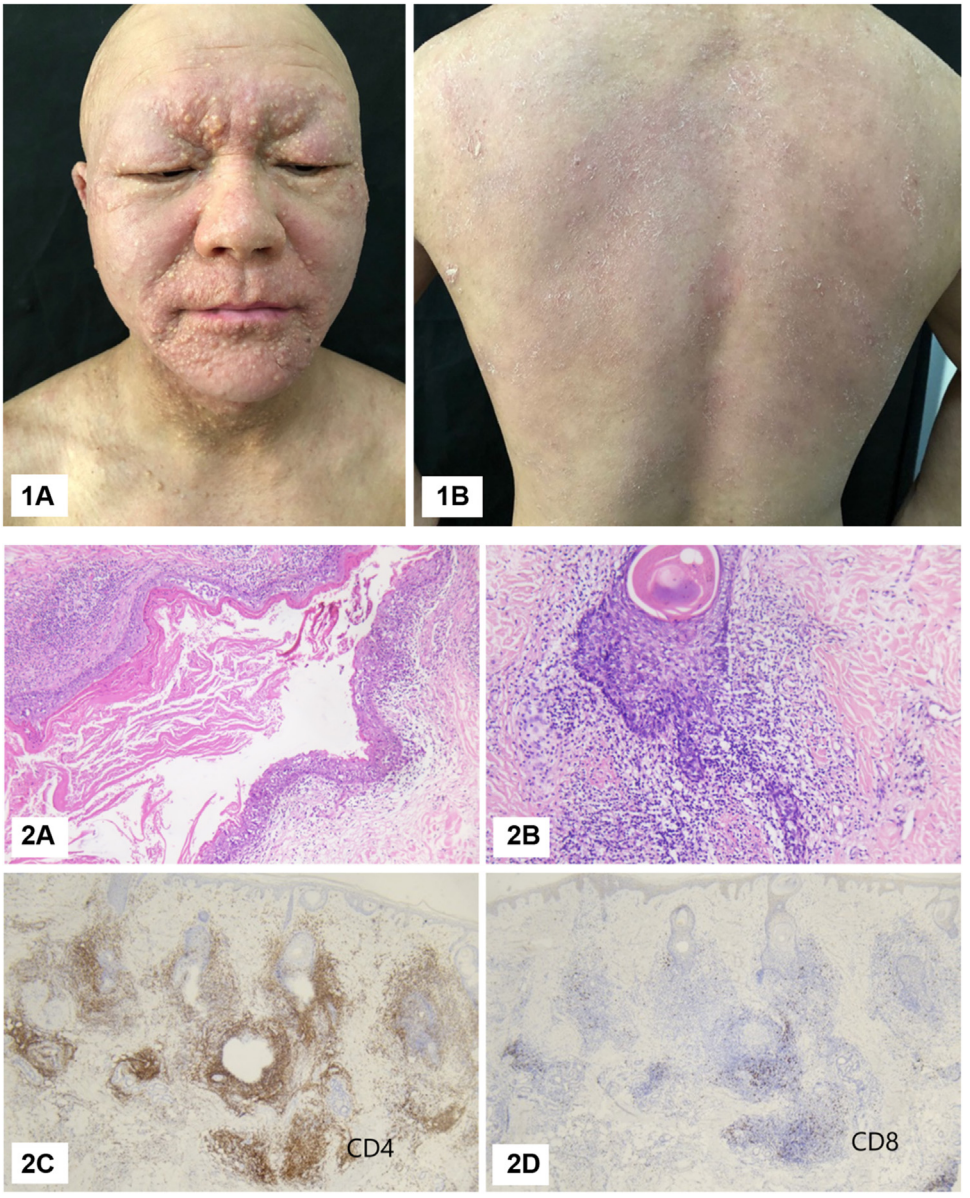

A 36-year-old male patient presented with yellow papules on the face/neck and scalp of 7-years' duration. Physical examination revealed grouped papules with small keratinous cysts or milia-like lesions on the face/neck and scalp (Fig 1, *A*). The patient also had multiple discrete erythematous plaques covered with nonadherent scales involving 50% of the body surface (Fig 1, *B*) and loss of all body hair, including eyebrows, eyelashes, armpit hair, and pubic hair. Biopsies of chin and scalp were performed (Fig 2, *A-D*).

Question 1: What is the most likely diagnosis?

A. Folliculotropic mycosis fungoides

B. Acne conglobata

C. Sézary syndrome

D. Lupus comedonicus

E. Lupus miliaris disseminatus faciei

Answer:

A. Folliculotropic mycosis fungoides (FMF)—Correct. Histologic examination of the chin demonstrated multiple infundibular cysts with a folliculotropic lymphoid infiltrate without a background of epidermotropic lymphocytes (Fig 2, *A*). The scalp biopsy also exhibited perifollicular atypical lymphocytes with folliculotropism but no overlying epidermotropism (Fig 2, *B*). Immunohistochemical stains revealed CD3+ CD4+ CD5+ CD7+ CD30- CD20- T cells with a marked shift in the CD4-to-CD8 ratio (CD4 shown in Fig 2, *C* and CD8 shown in Fig 2, *D*). Molecular test of the scalp lesion demonstrated a monoclonal T-cell receptor gene rearrangement. A diagnosis of FMF was made.

B. Acne conglobata—Incorrect. Although acne conglobata may present similarly on the face, histopathologic features and the results of immunohistochemical stains can exclude this diagnosis.

C. Sézary syndrome (SS)—Incorrect. There is clinical and histomorphological similarity between SS and FMF. However, folliculotropic atypical T cells with a marked shift in the CD4-to-CD8 ratio and without blood involvement make SS less likely.

D. Lupus comedonicus—Incorrect. Lupus comedonicus shows lichenoid interface dermatitis and comedo-like dilatation of follicular infundibula histologically. Unlike FMF, there are no atypical lymphocytes involving the follicles.[1]

E. Lupus miliaris disseminatus faciei (LMDF)—Incorrect. In LMDF, the patient develops an abrupt onset of red, yellow, or brown papules and nodules on the face. The histologic hallmark of LMDF is palisaded granulomas with central caseous necrosis,[2] unlike the pathological features of our patient.[2]

Question 2: Which of the following characteristic CANNOT be used to distinguish early/indolent FMF from advanced/aggressive FMF?

A. Clinical distribution

B. Pruritus

C. Eosinophils

D. Depth of infiltrates & density of perifollicular infiltrate

E. Degree of folliculotropism

Answer:

A. Clinical distribution—Incorrect. Advanced FMF characteristically involves the head and neck, while early-stage lesions present mainly on the trunk and limbs, although about one-third of the patients have concurrent head/neck lesions.[3]

B. Pruritus—Incorrect. Pruritus is more common in advanced-stage than in early-stage FMF. Patients with keratosis pilaris–like lesions type of FMF or juvenile patients with early-stage FMF only feel minor itching.[3]

C. Eosinophils—Incorrect. Consistent with classic mycosis fungoides (MF), the number of eosinophils in advanced-stage FMF is almost 6 times higher than those in the early-stage subgroup.[3]

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D. Depth of infiltrates & density of perifollicular infiltrate—Incorrect. In advanced-stage FMF, infiltrates are significantly deeper and perifollicular infiltrates are more obvious than early-stage lesions. Early-stage lesions show infiltration limited to the adventitial perifollicular dermis, whereas the infiltration of advanced lesions has a nodular to diffuse pattern extending to the reticular dermis.[3]

E. Degree of folliculotropism—Correct. Unlike classic tumor-stage MF, there was no difference in the degree of folliculotropism between early-stage and advanced-stage FMF.[3]

Question 3: What is the best treatment for this patient?

A. Hydroxychloroquine

B. Tetracycline

C. Allogeneic stem-cell transplantation

D. Combine interferon with UVA1

E. Highly potent corticosteroids

Answers:

A. Hydroxychloroquine—Incorrect. Antimalarials such as hydroxychloroquine are effective in the treatment of lupus comedonicus, but not MF. For resistant cases, combination of antimalarials with intralesional steroids and manual extraction may have efficacy.

B. Tetracycline—Incorrect. Tetracycline is the first-line treatment for LMDF but not MF. Histologic caseous necrosis may be induced by inflammatory and immune responses to Demodex. Thus, ornidazole tablets can be used against Demodex infection in LMDF.

C. Allogeneic stem-cell transplantation—Incorrect. Allogeneic stem-cell transplantation can be used as an alternative treatment for extracutaneous FMF or advanced-stage SS. It has been reported that transplantation at an early phase of SS was associated with a lower risk of relapse, while 3-year overall survival was not statistically significant.[4]

D. Combine interferon with UVA1—Correct. Patients with advanced-stage FMF often receive combined treatment (psoralen plus ultraviolet A plus interferon, retinoids, or radiotherapy). Studies showed that UVA1 penetrates more deeply than psoralen plus ultraviolet A.[5] In our case, the lymphocytes penetrated deep into the follicle, so we opted to treat the patient with interferon and UVA1.

E. Highly potent corticosteroids—Incorrect. Early-stage FMF could be treated with highly potent corticosteroids. However, this patient is more likely to belong to the advanced subgroup. So, corticosteroids are not the best treatment option.

Abbreviations used:

FMF: folliculotropic mycosis fungoides
LMDF: lupus miliaris disseminatus faciei
SS: Sézary syndrome

Conflicts of interest

None disclosed.

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