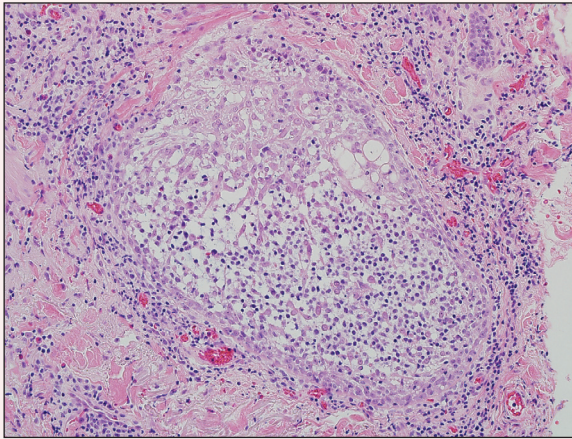


Pruritic Papules on the Scalp and Arms

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H&E, original magnification $\times 20$.

A 60-year-old man presented with a 3-month history of itchy bumps on the scalp and arms. He also noticed some patches of hair loss in these areas. He had no history of other skin conditions and was otherwise healthy with no other medical comorbidities.

The best diagnosis is:

- eosinophilic pustular folliculitis
- follicular mucinosis
- folliculotropic mycosis fungoides
- lupus erythematosus
- pityrosporum folliculitis

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The authors report no conflict of interest.

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Folliculotropic Mycosis Fungoides

Folliculotropic mycosis fungoides (FMF) is a variant of mycosis fungoides (MF) that occurs mostly in adults with a male predilection. The disease clinically favors the head and neck. Patients commonly present with pruritic papules that often are grouped, alopecia, and frequent secondary bacterial infections. Less commonly patients present with acneiform lesions and mucinorrhea. Patients often experience more pruritus in FMF than in classic MF, which can provide a good means of assessing disease activity. Disease-specific 5-year survival is approximately 70% to 80%, which is worse than classic plaque-stage MF and similar to tumor-stage MF.¹

Treatment of FMF differs from classic MF in that the lesions are less responsive to skin-targeted therapies due to the perifollicular nature of dermal infiltrates. Superficial skin lesions can be treated with psoralen plus UVA (PUVA) therapy. Other options include PUVA in combination with interferon alfa or retinoids and local radiotherapy for solitary thick tumors; however, in patients who have more infiltrative skin lesions or had PUVA therapy that failed, total skin electron beam therapy may be required.²

On histologic examination, there typically is perivascular and periadnexal localization of dermal infiltrates with varied involvement of the follicular epithelium and damage to hair follicles by atypical small, medium, and large hyperchromatic lymphocytes with cerebriform nuclei. Mucinous degeneration of the follicular epithelium can be seen, as highlighted on Alcian blue staining, and a mixed infiltrate of eosinophils and plasma cells often is present (quiz image and Figure 1). Frequent sparing of the epidermis is noteworthy.²⁻⁴ In most cases, the neoplastic T lymphocytes are characterized by a CD3⁺CD4⁺CD8⁻ immunophenotype as is seen in classic MF. Sometimes an admixture of CD30⁺ blast cells is seen.¹

Histologic differential considerations for FMF include eosinophilic pustular folliculitis (EPF), primary follicular mucinosis, lupus erythematosus, and pityrosporum folliculitis.

Eosinophilic pustular folliculitis has several clinical subtypes, such as classic Ofuji disease and immunosuppression-associated EPF secondary to human immunodeficiency virus. Histologically, EPF is characterized by spongiosis of the hair follicle epithelium with exocytosis of a mixed infiltrate of lymphocytes and eosinophils extending from the sebaceous gland and its duct to the infundibulum with formation of hallmark eosinophilic pustules (Figure 2). Infiltration of neutrophils in inflamed lesions generally is seen.

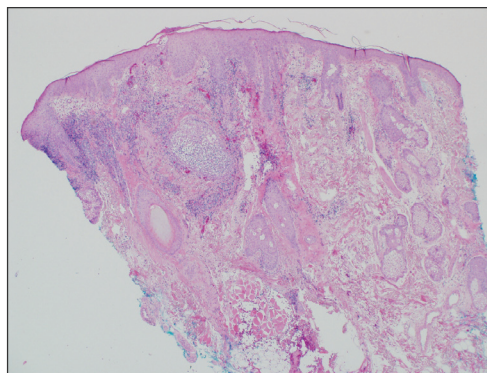


Figure 1. Follicular and perivascular dermal infiltrates in folliculotropic mycosis fungoides (H&E, original magnification $\times 4$).

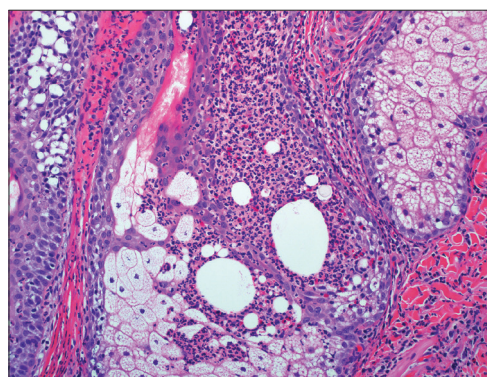


Figure 2. Eosinophilic pustular folliculitis with spongiosis of the hair follicle epithelium and exocytosis of a mixed infiltrate of lymphocytes and eosinophils (H&E, original magnification $\times 20$). Photograph courtesy of Brian Swick, MD (Iowa City, Iowa).

Eosinophilic pustular folliculitis is an important differential for FMF, as follicular mucinosis has been observed in lesions of EPF.⁵ Both EPF and FMF can exhibit eosinophils and lymphocytes in the upper dermis, spongiosis of the hair follicle epithelium, and mucinous degeneration of follicles,⁶ though lymphocytic atypia and relatively fewer eosinophils are suggestive of the latter.

Primary follicular mucinosis (PFM) tends to occur as a solitary lesion in younger female patients in contrast to the multiple lesions that typically appear in older male patients with FMF. Histologically, PFM usually manifests as large, cystic, mucin-filled spaces and polyclonal perivascular and periadnexal lymphocytic infiltrate without notable cellular atypia or epidermotropism (Figure 3). Because follicular

mucinosis is a common feature of FMF, its distinction from PFM can be challenging and often is aided by the absence of cellular atypia and relatively mild lymphocytic infiltrate in the latter.⁷

Cutaneous lupus erythematosus with its characteristic folliculocentric lymphocytic infiltration and

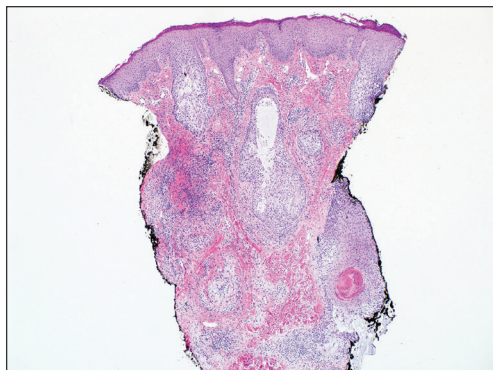


Figure 3. Follicular mucinosis with mucin-filled cystic spaces (H&E, original magnification ×4). Photograph courtesy of Brian Swick, MD (Iowa City, Iowa).

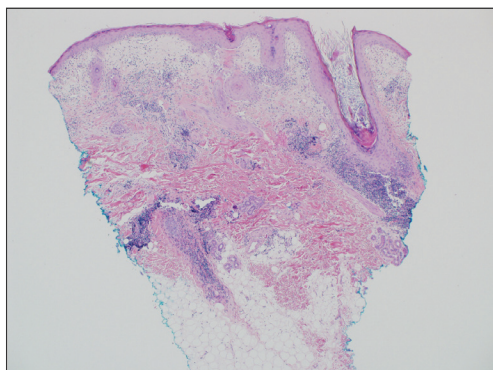


Figure 4. Cutaneous lupus erythematosus with interface dermatitis as well as a perivascular and periadnexal lymphocytic infiltrate with follicular plugging (H&E, original magnification ×4). Photograph courtesy of Brian Swick, MD (Iowa City, Iowa).

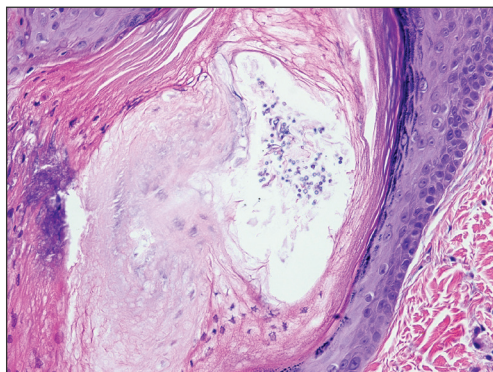


Figure 5. Pityrosporum folliculitis with budding yeast forms (H&E, original magnification ×40). Photograph courtesy of Brian Swick, MD (Iowa City, Iowa).

associated dermal mucin also qualifies as a potential differential possibility for FMF; however, the perivascular and periadnexal pattern of lymphocytic infiltration as well as the localization of mucin to the reticular dermal interstitium^{8,9} are key histopathologic distinctions (Figure 4). Furthermore, although the histologic presentation of lupus erythematosus can be variable, it also classically shows interface dermatitis, basement membrane thickening, and follicular plugging.

Pityrosporum folliculitis is the most common cause of fungal folliculitis and is caused by the *Malassezia* species. On histology, there typically is an unremarkable epithelium with plugged follicles and suppurative folliculitis. Serial sections of the biopsy specimen often are required to identify dilated, follicle-containing, budding yeast cells (Figure 5). Organisms are located predominantly within the infundibulum and orifice of follicular lumen, are positive for periodic acid–Schiff, and are diastase resistant.¹⁰

REFERENCES

1. Willemze R, Jaffe ES, Burg G, et al. WHO-EORTC classification for cutaneous lymphomas. *Blood*. 2005; 105:3768-3785.
2. van Doorn R, Scheffer E, Willemze R. Follicular mycosis fungoides, a distinct disease entity with or without associated follicular mucinosis. a clinicopathologic and follow-up study of 51 patients. *Arch Dermatol*. 2002;138:191-198.
3. DeBloom J 2nd, Severson J, Gaspari A, et al. Follicular mycosis fungoides: a case report and review of the literature. *J Cutan Pathol*. 2001;28:318-324.
4. Flaig MJ, Cerroni L, Schuhmann K, et al. Follicular mycosis fungoides: a histopathologic analysis of nine cases. *J Cutan Pathol*. 2001;28:525-530.
5. Fujiyama T, Tokura Y. Clinical and histopathological differential diagnosis of eosinophilic pustular folliculitis. *J Dermatol*. 2013;40:419-423.
6. Lee JY, Tsai YM, Sheu HM. Ofuji's disease with follicular mucinosis and its differential diagnosis from alopecia mucinosa. *J Cutan Pathol*. 2003;30:307-313.
7. Rongioletti F, De Lucchi S, Meyes D, et al. Follicular mucinosis: a clinicopathologic, histochemical, immunohistochemical and molecular study comparing the primary benign form and the mycosis fungoides-associated follicular mucinosis. *J Cutan Pathol*. 2010;37:15-19.
8. Vincent JG, Chan MP. Specificity of dermal mucin in the diagnosis of lupus erythematosus: comparison with other dermatitides and normal skin. *J Cutan Pathol*. 2015;42:722-729.
9. Yell JA, Mbuagbaw J, Burge SM. Cutaneous manifestations of systemic lupus erythematosus. *Br J Dermatol*. 1996;135:355-362.
10. Durdu M, Ilkit M. First step in the differential diagnosis of folliculitis: cytology. *Crit Rev Microbiol*. 2013;39:9-25.