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Acneiform follicular mucinosis: an indolent follicular mucinosis variant unrelated to mycosis fungoides?

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Summary

Follicular mucinosis (FM) can present as an acneiform eruption, and is usually a benign variant of primary FM unrelated to cutaneous T-cell lymphoma (CTCL). We report two cases of women in their twenties who presented with an acneiform rash on the face, arms and back. In both cases, pathological evaluation of the facial papules revealed predominantly mucinous degeneration of the follicular epithelium, with insufficient lymphocytic infiltration or atypia to diagnose mycosis fungoides. These cases are similar to previous reports of acneiform FM. As none of the reported cases progressed to CTCL, we consider that overdiagnosis and overtreatment should be avoided in acneiform FM, but recommend long-term follow-up.

Follicular mucinosis (FM) is an uncommon condition, which is characterized by mucin deposition within the follicular unit. It is classified into two forms: a primary benign idiopathic form, and a secondary form that is usually associated with cutaneous T-cell lymphoma (CTCL), mainly folliculotropic mycosis fungoides (FMF).¹ FMF can present as acneiform lesions, comedones or cysts, and histologically shows a prototypical pattern of a folliculotropic lymphoid infiltrate sparing the epidermis with or without follicular mucinosis. We describe two patients who were referred for evaluation of an acneiform rash and correlative histological findings of FM, which raised concerns about the possibility of FMF.

Report

Patient 1 was a 28-year-old woman who presented with a 4-year history of an asymptomatic facial skin-coloured papules (Fig. 1). She had previously been treated for acne rosacea and folliculitis (oral minocycline and cephalexin; topical pimecrolimus and metronidazole) with no effect.

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Patient 2 was a 26-year-old woman referred with a pruritic papular rash on her face, upper arms and back that waxed and waned for over a year. Slight improvement was achieved with acne therapies (doxycycline and topical retinoids).

Both patients denied any constitutional symptoms. On physical examination, neither presented with patches, plaques, tumours or comedones, and no lymphadenopathy was identified.

A biopsy was taken from the facial papules of each patient, and histological examination revealed follicular mucinosis and a perifollicular lymphocytic infiltrate lacking cytological atypia (Fig. 2). In Patient 2, two other biopsies were taken from the shoulder and the back, both of which were found to have nonspecific perivascular and periadnexal lymphocytic infiltrates with focal spongiosis. T-cell receptor (TCR) gene rearrangement studies were negative.

For both patients, a diagnosis of acneiform FM was made, based on the clinicopathological findings.

FM presenting as an acneiform eruption had been described in few case reports and small case series. This unusual primary FM variant clinically presents in young patients, usually women, as asymptomatic or pruritic skin-coloured to erythematous papules on the cheeks, forehead, neck and arms. The typical patient describes a chronic rash with periodic flare-ups despite multiple treatments for acne, acne rosacea or folliculitis. Acneiform FM is diagnosed based on histopathology showing follicular mucinosis and a perifollicular lymphocytic infiltrate that does not meet the criteria for MF. A total of 16 patients have been reported in the literature; no progression to MF was reported for any of these, even after long follow-up periods in excess of 20 years.¹⁻⁶ The clinical presentation, treatment and reported outcome for previously published acneiform FM cases are summarized in Table 1.

Based on these data, we decided to treat both of our patients conservatively; the first patient was treated with oral isotretinoin and topical corticosteroids, while the second patient was monitored only. After follow-up periods of 8 months, both patients still presented with occasional flare-ups, but no morphological changes or systemic symptoms were noted.

The presence of FM always raises the concern for FMF and causes confusion and anxiety, particularly in young patients with no CTCL history. Cerroni *et al* suggested that idiopathic FM may represent an indolent localized variant of MF⁷ but they specifically referred to the different clinical appearance of acneiform FM and suggested that it might be a variant of acneiform dermatoses or a reaction pattern of the follicular epithelium unrelated to MF.⁸

Recent studies have shown that FMF does not necessarily imply a poor prognosis,⁹ defining a subgroup of patients with early skin-limited FMF that have a favourable prognosis, with 5-year and 10-year disease specific survival rates of 96% and 93%, respectively.⁹ Acneiform FM, if considered as a MF-associated variant, could eventually be included within this indolent FMF group.

FM may also be seen as a reactive histological pattern in a wide range of nonlymphoid skin disorders with a benign outcome, including acne vulgaris.¹⁰

In conclusion, FM may present in young adults as an acneiform eruption with an unknown aetiology. It has a chronic course, and response to therapy is often poor. We suspect that acneiform FM is more prevalent than reported as it mimics common inflammatory and infectious dermatoses, and facial biopsies are often avoided. Recognizing the indolent nature of this variant of FM should prevent overdiagnosis and overtreatment. As the data on this variant is limited, we recommend long-term follow-up for these patients (once a year at least) and repeated biopsies of suspicious lesions with new clinical morphology.

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References

1. Brown HA, Gibson LE, Pujol RM, et al. Primary follicular mucinosis: long-term follow-up of patients younger than 40 years with and without clonal T-cell receptor gene rearrangement. *J Am Acad Dermatol.* 2002; 47:856–62. [PubMed: 12451369]
2. Brau-Javier CN, Santos-Arroyo AE, De Sanctis-Gonzalez IM, Sanchez JL. Follicular mucinosis presenting as an acneiform eruption: a follow-up study. *Am J Dermatopathol.* 2013; 35:792–6. [PubMed: 24257190]
3. Muscardin LM, Capitanio B, Fagnoli MC, Maini A. Acneiform follicular mucinosis of the head and neck region. *Eur J Dermatol.* 2003; 13:199–202. [PubMed: 12695140]
4. Passaro EM, Silveira MT, Valente NY. Acneiform follicular mucinosis. *Clin Exp Dermatol.* 2004; 29:396–8. [PubMed: 15245540]
5. White FN, Bergstresser PR, Lamontagne D, Boswell JS. Acneiform follicular mucinosis responding to hydroxychloroquine. *Arch Dermatol.* 2011; 147:130–1. [PubMed: 21242413]
6. Wittenberg GP, Gibson LE, Pittelkow MR, el-Azhary RA. Follicular mucinosis presenting as an acneiform eruption: report of four cases. *J Am Acad Dermatol.* 1998; 38:849–51. [PubMed: 9591801]
7. Cerroni L, Fink-Puches R, Back B, Kerl H. Follicular mucinosis: a critical reappraisal of clinicopathologic features and association with mycosis fungoides and Sezary syndrome. *Arch Dermatol.* 2002; 138:182–9. [PubMed: 11843637]
8. Cerroni L, Kerl H. Primary follicular mucinosis and association with mycosis fungoides and other cutaneous T-cell lymphomas. *J Am Acad Dermatol.* 2004; 51:146–7. author reply 147–8. [PubMed: 15243544]
9. van Santen S, Roach RE, van Doorn R, et al. Clinical staging and prognostic factors in folliculotropic mycosis fungoides. *JAMA Dermatol.* 2016; 152:992–1000. [PubMed: 27276223]
10. Mir-Bonafe JM, Canueto J, Fernandez-Lopez E, Santos-Briz A. Follicular mucinosis associated with nonlymphoid skin conditions. *Am J Dermatopathol.* 2014; 36:705–9. [PubMed: 25072678]

Learning points

- FM is an uncommon condition characterized by mucin deposition within the follicular unit.
- FM may present in young adults as an acneiform eruption with an unknown aetiology.
- Acneiform FM has a chronic course, and response to therapy is often poor.
- None of the previously reported cases of acneiform FM progressed to cutaneous T-cell lymphoma.
- Recognizing the indolent nature of this variant of FM should prevent overdiagnosis and over-treatment; however, as the data on this variant are limited, long-term follow-up is recommended.

Figure 1.

(a–c) Patient 1 (a). Multiple skin-coloured papules on the cheeks; no comedones visible. (b,c) Mucin within the folliculosebaceous units with a mild perfollicular and intraepithelial lymphocytic infiltrate with no obvious atypia. No epidermotropism visible. Haematoxylin and eosin; original magnification (b) $\times 20$; (c) $\times 200$.

Table 1
Summary of acneiform follicular mucinosis cases in our study and the literature

Reference	Patient	Sex	Age, years	Location of lesions	Morphology of lesions	Itch	Clonality	Treatment	Outcome	Follow-up, months
Current study	1*	F	28	Cheeks, mandibles, forehead	Multiple skin-coloured papules	No	NP	Observation	PR	6
	2*	F	26	Cheeks, lateral upper arms, back	Multiple skin-coloured papules	Yes	Negative	Tretinoin, hydrocortisone	PR	8
	3	F	22	Forehead, cheeks, neck	Multiple 2–4 mm noninflammatory papules	No	Positive	Tretinoin gel, pentoxifylline, isotretinoin	PR	88
Brown <i>et al.</i> , 2002, ¹	4	M	31	Cheeks, chin, forehead, neck	Multiple 2–4mm papules	No	Positive	Nitrogen mustard, minocycline	PR	86
Wittenberg <i>et al.</i> , 1998 ⁶	5	F	35	Cheeks, chin, temples, forehead	Acneiform lesions	Yes	Negative	Clobetasol cream, isotretinoin	PR	280
Brau-Javier <i>et al.</i> , 2013 ²	6	M	29	Face (mainly cheeks, and forehead), neck	Erythematous or skin-coloured facial papules Associated patchy noncicatricial alopecia in one patient	Yes; in 7/8	Performed in 1/8; positive	NM	AD	60
Muscardin <i>et al.</i> , 2013 ³	7	F	13						CR	48
	8	F	30						AD	60
	9	F	26						CR	36
	10	M	43						CR	36
	11	F	49						AD	12
	12	F	45						AD	12
	13	F	16						CR	12
	14	F	45	Cheeks, chin, forehead	Multiple skin-coloured papulocystic lesions	No	Negative	Observation	PR	36
	15	F	30	Face, neck	Multiple 2–5 mm erythematous papulocystic lesions	No	Negative		PR	24
	16	M	36	Forehead, Temples, retro-auricular	Multiple red or pale isolated papules and nodules	Yes	NP	Prednisone	CR	7
Passaro <i>et al.</i> , 2004 ⁴	17	F	10	Forehead, cheeks, temples, arms	Firm erythematous papules	No	NP	Hydroxychloroquine	CR	6
White <i>et al.</i> , 2011 ⁵	18	F	21	Forehead, temples, mandibles	Skin-coloured papules	Yes	NP	Benzoyl peroxide, tetracycline	PR	190
Wittenberg <i>et al.</i> , 1998 ⁶										

AD, active disease; CR, complete response; NM, not mentioned; NP, not performed; PR, partial response