

## LETTER TO THE EDITOR

## Urticaria-like follicular mucinosis: four new cases of a controversial entity

### Editor

Primary follicular mucinosis (PFM) is a rare idiopathic disorder characterized by mucin deposition inside the hair follicles.<sup>1</sup> It usually presents with follicular papules or infiltrated plaques with prominent follicular openings associated with alopecia.<sup>1</sup> Unusual presentations include alopecia areata-like, scarring alopecia, folliculitis, acneiform eruptions and linear lesions following Blaschko lines.<sup>2,3</sup>

In 1980, a new clinical variant called Urticaria-like follicular mucinosis (ULFM) has been reported, presenting with a cyclic eruption of urticarial papules and plaques on the face on a 'rosacea-like or seborrhoeic' background.<sup>4</sup> Since then, only six cases have been subsequently described.<sup>4-9</sup>

We report four new cases which occurred to one 40-year-old female, two 46-year-old females and one 60-year-old male. All cases presented with a cyclic eruption of itching, erythematous, slightly scaling papules and plaques on the face and the upper chest (Fig. 1). Laboratory investigations including anti-nuclear antibodies were not contributory. Histopathology showed typical features of follicular mucinosis (FM) (Fig. 2). *Demodex folliculorum* was never found. The infiltrate was made up of CD3 + lymphocytes without a significant increase of CD4 + cells. T-cell receptor gene analysis using Polymerase



**Figure 1** Clinical aspect of Urticaria-like follicular mucinosis: erythematous, urticarial, slightly scaling papules, merging into small plaques on the face.

Chain Reaction technique revealed a T-cell polyclonal pattern in two cases. Direct immunofluorescence was negative in one case. Topical corticosteroids and metronidazole and systemic antibiotics, hydroxychloroquine, levocetirizine and total sun block were ineffective. Pimecrolimus was effective in one patient, but was interrupted due to a pustular eruption. Spontaneous resolution occurred to two patients.

Urticaria-like follicular mucinosis differs from classical PFM because lesions are not fixed, not follicular and not associated with alopecia. This entity has been defined as urticaria-like due to the transient course and the associated itch. Characteristically, lesions wax and wane irregularly over a period that can vary from a few months to 15 years.<sup>5</sup>

Although face and neck are the commonest sites, the upper part of the trunk can be affected.<sup>7,8</sup>

Urticaria-like follicular mucinosis had been described only in two women<sup>6,7</sup> and, adding our three female new cases, we demonstrate that this entity is not exclusive to male gender. The relationship with sunlight is controversial.<sup>4,5</sup> In our series, lesions worsened with sun exposure in one case.

The diagnosis of ULFM is often delayed as the dermatitis is easily misinterpreted as urticaria, Jessner's lymphocytic infiltration of the skin, lupus tumidus, seborrhoeic dermatitis, rosacea, atopic dermatitis and allergic contact dermatitis. Histopathology clarifies the issue showing a typical FM pattern.

The main microscopic differential diagnosis is lymphoma-associated FM. However, ULFM has never been associated with lymphoma and it lacks the clinical and histological criteria that favours an associated lymphoma, in particular mycosis fungoides (MF).<sup>10</sup> A monoclonal rearrangement has been found in one of three (our two included) cases of ULFM,<sup>9</sup> but has not been associated with a progression to MF.

The etiopathogenesis of ULFM remains unknown. Considering the clinical similarity with seborrhoeic dermatitis or rosacea and the fact that FM has been recognized as a histological epiphenomenon in a variety of dermatoses, one can hypothesize that ULFM is a histological variant of these skin diseases, eventually triggered by long-term use of topical corticosteroids. However, FM as a histological feature has been reported to occur neither in rosacea nor in seborrhoeic dermatitis.

A validated treatment does not exist. In our third patient, we observed a remarkable initial improvement of his lesions with topical pimecrolimus. Spontaneous healing may also occur.

Dermatologists should be aware that PFM can manifest with different clinical expressions, not always characterized by alopecia, including ULFM.

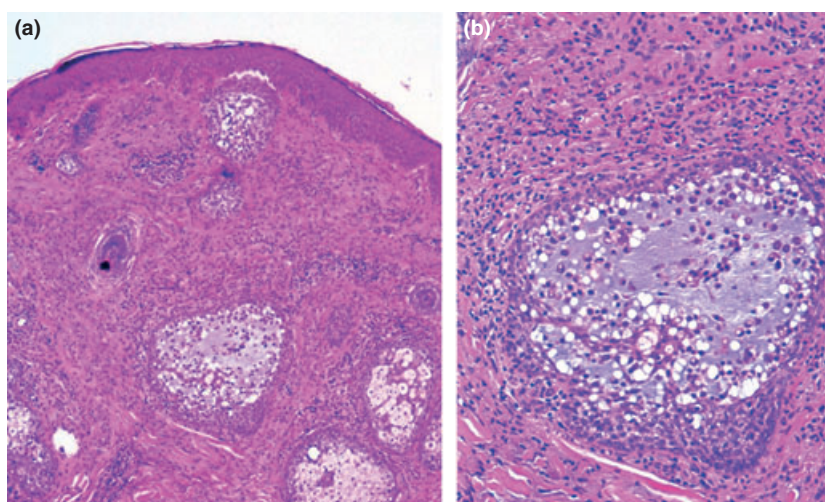


**Figure 2** Histological aspect of Urticaria-like follicular mucinosis. Mucin deposits are present in the follicular epithelium, dissociating follicular keratinocytes and forming large cavities, together with a superficial and deep perivascular and periadnexal lymphocytic infiltrate (H&E, a 4× and b 10×).

E. Cinotti,<sup>†,\*</sup> D. Basso,<sup>†</sup> P. Donati,<sup>‡</sup> A. Parodi,<sup>†</sup>  
F. Rongioletti<sup>†,§</sup>

<sup>†</sup>Section of Dermatology, DISSAL, University of Genoa, Italy

<sup>‡</sup>San Gallicano Dermatological Institute, Rome, Italy

<sup>§</sup>Department of Pathology, University of Genoa, Italy

\*Correspondence: Elisa Cinotti. E-mail: elisacinotti@gmail.com

## References

- 1 Pinkus H. Alopecia mucinosa; inflammatory plaques with alopecia characterized by root-sheath mucinosis. *Arch Derm* 1957; **76**: 419–424.
- 2 Passaro EM, Silveira MT, Valente NY. Acneiform follicular mucinosis. *Clin Exp Dermatol* 2004; **29**: 396–398.
- 3 Tosti A, Fanti PA, Peserico A *et al*. Linear alopecia mucinosa along Blaschko lines. *Acta Derm Venereol* 1992; **72**: 155–156.
- 4 Enjolras O, Guillemette J, Hewitt J. Dermatose ortiée avec mucinose folliculaire. *Ann Dermatol Venereol* 1980; **107**: 491–495.
- 5 Crovato F, Nazzari G, Nunzi E *et al*. Urticaria-like follicular mucinosis. *Dermatologica* 1985; **170**: 133–135.
- 6 Dommergues C, Rybojad M, Menanteau Y *et al*. Cas pour diagnostic. *Rev Eur Dermatol MST* 1989; **1**: 431–433.
- 7 Tupker RA, van der Meer JB, Groote AD *et al*. Urticaria-like follicular mucinosis in a young female patient. *Acta Derm Venereol* 1997; **77**: 323–324.
- 8 Harthi FA, Kudwah A, Ajlan A *et al*. Urticaria-like follicular mucinosis responding to dapsone. *Acta Derm Venereol* 2003; **83**: 389–390.
- 9 Marrou K, Skowron F, Philippot V *et al*. Urticaria-like mucinosis. *Ann Dermatol Venereol* 2006; **133**: 605–606.
- 10 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.

DOI: 10.1111/j.1468-3083.2012.04701.x