Disseminated Small Papules on the Face: A Quiz

Victor OLSAVSZKY[1], Cyrill GÉRAUD[1–3] and Moritz FELCHT[1,3]*
*[1]Department of Dermatology, Venereology, and Allergology, University Medical Center and Medical Faculty Mannheim, Heidelberg University, and Center of Excellence in Dermatology, Mannheim, [2]Section of Clinical and Molecular Dermatology, Medical Faculty Mannheim, Heidelberg University, Mannheim, and [3]European Center for Angioscience, Medical Faculty Mannheim, Heidelberg University, Mannheim, Germany. *E-mail: moritz.felcht@umm.de*

A woman in her 40s presented to our outpatient department with progressive facial skin lesions that had appeared 7 years earlier. She had also experienced loss of eyebrow hair and recession of the frontoparietal hairline 3-4 years ago. She had numerous disseminated, non-confluent, pinhead-sized, skin-coloured papules on the cheeks, infraorbital and perioral regions (Fig. 1a, b). Her anterior and temporal hairline was visibly receding, revealing discrete keratotic follicular papules on the forehead (Fig. 1c). Her eyebrows had been cosmetically tattooed with permanent make-up. A 9-month treatment with topical and oral isotretinoin led to a significant improvement in the facial lesions, but treatment was discontinued due to elevated liver enzymes and epistaxis. The patient also reported itching of the papules for the last 2 weeks. Skin biopsies were taken from the left orbital region and the right temple (Fig. 2).

What is your diagnosis? See next page for answer.

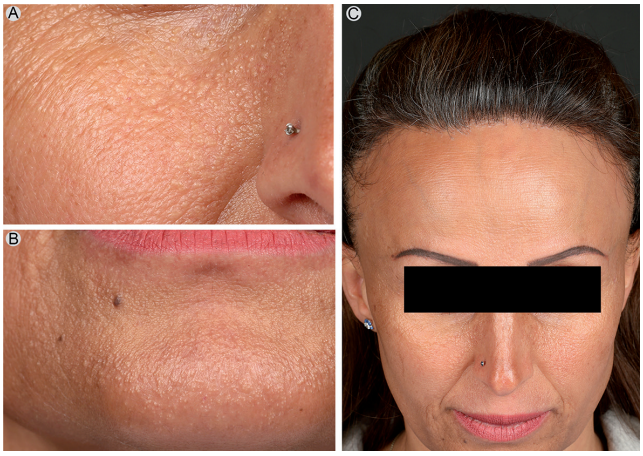

Fig. 1. (a, b) Close-up of the papules on (a) infraorbital and (b) perioral regions. (c) Receding frontoparietal hairline with keratotic follicular papules on the frontal line. Eyebrow hair loss concealed by permanent make-up.

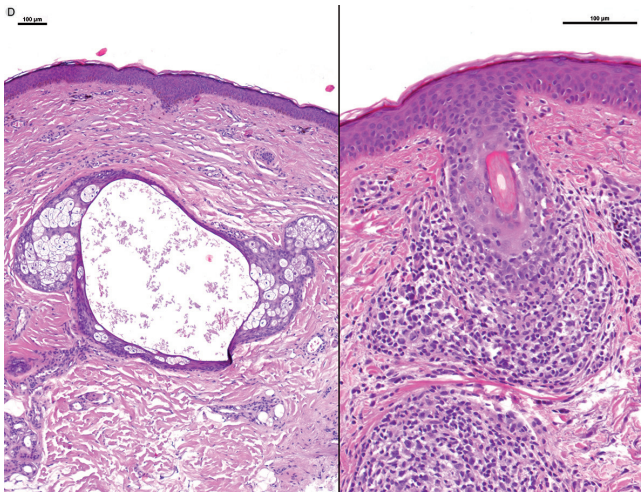

Fig. 2. Haematoxylin-eosin stained skin biopsy specimen. Left: comedo-like hyperkeratosis. Right: follicular interface dermatitis of a vellus hair. (Scale bar: 100 µm).

doi: 10.2340/00015555-3146
Acta Derm Venereol 2019; 99: 625–626

ANSWERS TO QUIZ

Disseminated Small Papules on the Face: A Commentary

Acta Derm Venereol 2019; 99: 625–626.

Diagnosis: **Facial keratosis pilaris-like papules in frontal fibrosing alopecia**

A biopsy specimen taken from the left lateral orbital region revealed follicular dilatation with comedo-like hyperkeratosis, as well as follicular interface dermatitis of vellus hair follicles (Fig. 2). A second biopsy taken from the right temple confirmed the diagnosis of frontal fibrosing alopecia by revealing infundibular interface dermatitis with atrophy and perifollicular fibrosis (data not shown). Blood analysis revealed positive antinuclear and anti-centromere antibodies. The patient denied any Raynaud symptoms and there were no signs of sclerodactyly. The patient was given clobetasol solution locally on the hairline and hydroxychloroquine in an alternate-day dosing, 2 tablets of 200 mg each 1 day and 1 tablet the next day, with a 3-month follow-up.

Postmenopausal frontal fibrosing alopecia was first described by Kossard in 1994 (1). This disease is usually regarded as a variant of lichen ruber follicularis, due to similar histopathological findings (2). The term postmenopausal was subsequently renounced, since it may affect premenopausal women as well as men (3). Clinically, perifollicular erythema and hyperkeratosis of the frontal hairline occur, but the temporal and occipital regions may also be affected. Thinning or complete hair loss of the eyebrows is highly characteristic (4). Other affected areas may include the axillae and pubic region.

Abbas et al. (5) first reported involvement of vellus hair follicles, in the form of pinpoint, skin-coloured, follicular keratotic papules of the face, and related it to the Piccardi-Lassueur-Graham-Little syndrome. Histological examination of the facial papules shows a perifollicular infiltrate around the vellus follicles, accompanied by hyperkeratotic dilated, comedo-like, infundibula, reminiscent of keratosis pilaris papules (6). Other histological features include perifollicular fibrosis and follicular atrophy of the isthmus (7). Direct immunofluorescence is negative.

The incidence of facial papules in frontal fibrosing alopecia has been reported as between 14% and 21%, although some authors argue that they may often not be recognized by clinicians (3, 6). Furthermore, Pedrosa et al. reported another type of yellow facial papule also associated with frontal fibrosing alopecia, which were histologically distinct, consisting of hypertrophic sebaceous glands (8). Most patients report itching of the facial lesions or a burning sensation. Some may present with an erythema on the face, reminiscent of rosacea (6).

To date there is no known effective local therapy for facial papules associated with frontal fibrosing alopecia. Improvements have been described after systemic corticosteroids (i.e. 0.5 mg/kg prednisone), antimalarials (hydroxychloroquine, 400 mg/day) and oral isotretinoin (10 mg/day) (7, 9). While the use of 5-alpha-reductase inhibitors, such as finasteride and dutasteride, showed a response in almost 50% of patients with frontal fibrosing alopecia, there have been no reports in the literature regarding their benefits for the associated facial papules (3).

The patient described here presented progressive facial lesions as the primary complaint. Therefore, multiple clinicians, consulted previously, failed to correlate these lesions with the patient's frontal fibrosing alopecia. We believe that keratosis pilaris-like papules of the face represent a commonly unrecognized characteristic finding of frontal fibrosing alopecia and do not represent an unusual variant of Piccardi-Lassueur-Graham-Little syndrome. Dermatologists should be aware of this association and should examine the facial skin of patients presenting with frontal fibrosing alopecia thoroughly. On the other hand, the frontal hair-line and eyebrows should be examined when patients present with small pinhead papules of the face as their primary complaint.

REFERENCES

1. Kossard S. Postmenopausal frontal fibrosing alopecia. Scarring alopecia in a pattern distribution. Arch Dermatol 1994; 130: 770–774.
2. Kossard S, Lee MS, Wilkinson B. Postmenopausal frontal fibrosing alopecia: a frontal variant of lichen planopilaris. J Am Acad Dermatol 1997; 36: 59–66.
3. Vano-Galvan S, Molina-Ruiz AM, Serrano-Falcon C, Arias-Santiago S, Rodrigues-Barata AR, Garnacho-Saucedo G, et al. Frontal fibrosing alopecia: a multicenter review of 355 patients. J Am Acad Dermatol 2014; 70: 670–678.
4. Anzai A, Donati A, Valente NY, Romiti R, Tosti A. Isolated eyebrow loss in frontal fibrosing alopecia: relevance of early diagnosis and treatment. Br J Dermatol 2016; 175: 1099–1101.
5. Abbas O, Chedraoui A, Ghosn S. Frontal fibrosing alopecia presenting with components of Piccardi-Lassueur-Graham-Little syndrome. J Am Acad Dermatol 2007; 57: S15–18.
6. Lopez-Pestana A, Tuneu A, Lobo C, Ormaechea N, Zubizarreta J, Vildosola S, et al. Facial lesions in frontal fibrosing alopecia (FFA): clinicopathological features in a series of 12 cases. J Am Acad Dermatol 2015; 73: 987 e981–986.
7. Donati A, Molina L, Doche I, Valente NS, Romiti R. Facial papules in frontal fibrosing alopecia: evidence of vellus follicle involvement. Arch Dermatol 2011; 147: 1424–1427.
8. Pedrosa AF, Duarte AF, Haneke E, Correia O. Yellow facial papules associated with frontal fibrosing alopecia: a distinct histologic pattern and response to isotretinoin. J Am Acad Dermatol 2017; 77: 764–766.
9. Flores-Terry MA, Garcia-Arpa M, Franco-Munoz M, Gonzalez-Ruiz L. Facial papules in frontal fibrosing alopecia: good response to isotretinoin. Actas Dermosifiliogr 2018; 109: 831–833.