ORIGINAL ARTICLE

Facial lesions in frontal fibrosing alopecia (FFA): Clinicopathological features in a series of 12 cases

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Background: Facial lesions in frontal fibrosing alopecia (FFA) have been poorly described in published series.

Objective: We sought to describe facial lesions in FFA.

Methods: We reviewed our series of 55 cases of FFA, selecting 12 cases with clinically significant facial lesions. We performed a histologic study of these lesions.

Results: In addition to the observations already described in the literature such as facial papules or follicular red dots, we observed perifollicular and diffuse erythema, sometimes with a reticular pattern, and the gradual appearance of pigmented macules on facial skin. Biopsy specimens from the areas with facial erythema showed perifollicular and interfollicular lymphocytic infiltrate and fibrosis around vellus hair follicles. Histologic evaluation of pigmented macules sometimes exhibited an increased epidermal pigmentation and on occasions, pigmentary incontinence.

Limitations: More patients are needed to determine the prevalence of these lesions in FFA.

Conclusion: On facial skin of patients with FFA, we can observe papules or perifollicular erythema secondary to vellus hair follicle involvement. We describe diffuse erythema, owing to follicular and interfollicular lichenoid infiltrate, and the gradual appearance of pigmented macules, which could be secondary to an increased epidermal pigmentation or to pigmentary incontinence. (J Am Acad Dermatol http://dx.doi.org/10.1016/j.jaad.2015.08.020.)

Key words: alopecia; face; frontal fibrosing alopecia; lichen planus; papules; vellus hair.

Frontal fibrosing alopecia (FFA), first described by Kossard[1] in 1994, is a form of permanent alopecia involving hair follicles in the frontotemporal area of the scalp, with hairline recession and eyebrow loss, affecting mostly postmenopausal women. It is now considered a particular form of lichen planopilaris, as it may damage follicles of any size, that is, vellus, intermediate, and terminal,[2] anywhere in the integument, and during any phase of the hair cycle.[3] Its recognition has substantially increased in recent years, with ever longer series of published cases.[4] Regarding facial injuries, the appearance of facial papules secondary to the involvement of vellus hair follicles was first reported by Abbas et al[5] in 2007 and later by Donati et al[6] in 2011. In 2014, Pirmez et al[7] described the follicular red dots in the glabellar and forehead areas in 6 of their 69 patients with FFA, although these lesions can also be observed on the skin of eyebrows[8] and cheeks.[9] They present as perifollicular erythema with or without follicular keratosis, and histologically exhibit perifollicular lichenoid inflammation. Recently, Dlova,[10] Rao et al,[11] and Berliner et al[12] reported the coexistence of FFA and lichen planus

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pigmentosus on facial skin in 27 patients, 24 of African origin, 1 of Indian origin, and 2 of Hispanic origin. They conclude that this association is very common in dark races, in which lichen planus pigmentosus is more prevalent.

In our series of 55 women given the diagnosis of FFA in the Department of Dermatology of the Donostia University Hospital in San Sebastian, Spain, from May 2005 to May 2013, we have observed 12 cases (21.8%) with clinically significant facial lesions. We describe the lesions and the histologic findings of their facial injuries.

CAPSULE SUMMARY

- Facial lesions reported in frontal fibrosing alopecia include papules and perifollicular erythema.
- We describe diffuse erythema as a result of interfollicular lichenoid infiltrate and the gradual appearance of pigmented macules.
- As these are subtle findings, clinicians should look for facial erythema and pigmented macules in these patients.

CLINICAL CASES

Twelve women with FFA presented with facial skin lesions (Table I). They all were Caucasian, with a mean age of 54 years, 7 of them premenopausal. The hairline recession was minor in 5 cases, and moderate in 7. They showed total or partial eyebrow alopecia and substantial vellus hair loss on limbs. In 11 of 12 patients, noninflammatory papules were observed over the temples, cheeks, or chin. In some cases there were scattered papules with keratin-filled dilated infundibula, simulating open comedones (keratosis pilaris-like papules). Histologic examination of these lesions from 7 patients showed perifollicular lichenoid infiltrate around vellus follicles (Fig 1).

In 9 of 12 cases, perifollicular erythema, sometimes with follicular keratosis, was evident in the glabellar, forehead, eyebrow, or cheek regions. In 7 patients the erythema was more diffuse, especially over the eyebrows and cheeks (Fig 2; available at http://www.jaad.org). Some of these patients referred a burning sensation, and had previously been given a diagnosis of rosacea, although they had never shown papules or pustules. Three patients (cases 7, 8, and 9) presented with generalized erythema on facial skin and neck, adopting a reticular pattern, more visible over the zygomatic arch. Histologic evaluation showed destruction of vellus hair follicles, interfollicular lichenoid infiltrate with patchy damage to the basal layer, and pigmentary incontinence. There were no mucin deposits or thickening of the basal membrane, and direct immunofluorescence produced negative findings (Fig 3; available at http://www.jaad.org). The erythema disappeared over the months and we noticed the gradual appearance of blue-gray or brown perifollicular macules, sometimes with a targetlike pattern, more evident on dermoscopy (Fig 4).

Table I. Clinical cases

Case	Age, y	Menopause	Disease duration, y	Frontotemporal hairline recession*	Eyebrow loss	Vellus limb loss	Facial papules	Perifollicular or diffuse erythema	Pigmented macules	Other diseases
1	44	No	1	Grade I	Yes	Yes	Yes	No	No	No
2	47	No	4	Grade I	Yes	Yes	Yes	Yes: eyebrows, cheeks	No	Birdshot disease
3	49	No	3	Grade II	Yes	Yes	Yes	No	Yes	Thalassemia minor
4	59	Yes	4	Grade II	Yes	Yes	Yes	No	No	No
5	60	Yes	2	Grade II	Yes	Yes	Yes	Yes: eyebrows, cheeks	No	No
6	43	No	3	Grade I	Yes	Yes	Yes	Yes: nose, cheeks	Yes	Hypothyroidism
7	42	No	4	Grade II	Yes	Yes	Yes	Yes: forehead, zygomatic arches, cheeks, neck	Yes	No
8	73	Yes	10	Grade III	Yes	Yes	Yes	Yes: forehead, zygomatic arches, cheeks, neck	Yes	No
9	63	Yes	3	Grade II	Yes	Yes	Yes	Yes: zygomatic arches, cheeks	Yes	No
10	57	Yes	6	Grade II	Yes	Yes	No	Yes: glabellar, cheeks	Yes	Hypothyroidism
11	57	Yes	8	Grade I	Yes	Yes	Yes	Yes: cheeks	Yes	Hypothyroidism
12	58	Yes	3	Grade I	Yes	Yes	Yes	Yes: eyebrows, cheeks	Yes	No

*Grade I, <1 cm; grade II, 1-2.99 cm; grade III, 3-4.99 cm; grade IV, 5-6.99 cm; grade V, >7 cm (from Vañó-Galván et al[4]).