Frontal Fibrosing Alopecia: Cutaneous Associations in Women With Skin of Color

Loren Krueger, MD; Katerina Svigos, BA; Nooshin Brinster, MD; Nada Elbuluk, MD, MSc

PRACTICE **POINTS**

- Frontal fibrosing alopecia (FFA) is associated with lichen planus pigmentosus, especially in patients with skin of color.
- Patients with FFA should be evaluated for additional cutaneous features including facial papules, glabellar red dots, and depressed frontal veins.

Frontal fibrosing alopecia (FFA) was first described as a progressive recession of the frontal hairline in postmenopausal women. Since its initial description, recognition and understanding of FFA has expanded. The condition is now defined as a patterned, symmetric, frontotemporal scarring alopecia that is considered to be histopathologically indistinguishable from lichen planopilaris. Numerous case reports and series have suggested clinical variants of and associations with FFA. In addition to reviewing the literature on FFA's associations, this article includes a case series of 5 women with skin of color (Hispanic and black) who presented with various cutaneous findings in association with FFA, including lichen planus pigmentosus (LPP), facial papules, and eyebrow loss. Recognition of the conditions that can occur in association with FFA in individuals with skin of color is important in further expanding our knowledge and understanding of FFA as a disease entity.

Cutis. 2018;102:335-338.

Frontal fibrosing alopecia (FFA) has been reported in association with lichen planus pigmentosus (LPP) and facial papules.[1-3] Lichen planus pigmentosus is a variant of lichen planus that causes hyperpigmentation of the face, neck, and/or intertriginous areas that may be useful as a clinical indicator in the development of FFA.[1] Facial papules in association with FFA are secondary to fibrosed vellus hairs.[2,3] Currently, reports of concomitant FFA, LPP, and facial papules in women with skin of color are limited in the literature. This case series includes 5 women of color (Hispanic and black) who presented to our clinic with FFA and various cutaneous associations. A review of the current literature on cutaneous associations of FFA also is provided.

Case Reports

Patient 1—A 50-year-old Hispanic woman who was previously presumed to have melasma by an outside physician presented with pruritus of the scalp and eyebrows of 1 month's duration. Physical examination revealed decreased frontal scalp hair density with perifollicular erythema and scale with thinning of the lateral eyebrows. Hyperpigmented coalesced macules (Figure 1A) and erythematous perifollicular papules were noted along the temples and on the perioral skin. Depressed forehead and temporal veins also were noted (Figure 1B). A biopsy of the scalp demonstrated perifollicular and perivascular lymphocytic inflammation and fibrosed hair follicles, and a biopsy of the perioral skin demonstrated perivascular lymphocytic inflammation with melanophages in the

Drs. Krueger and Brinster as well as Ms. Svigos are from New York University School of Medicine, New York. Drs. Krueger and Brinster are from the Ronald O. Perelman Department of Dermatology. Dr. Elbuluk is from the Department of Dermatology, University of Southern California, Los Angeles.
The authors report no conflict of interest.
Correspondence: Loren Krueger, MD, 240 E 38th St, 11th Floor, New York, NY 10016 (loren.krueger@nyumc.org).

papillary dermis. A diagnosis of FFA with LPP was established with these biopsies.

Patient 2—A 61-year-old black woman presented with asymptomatic hair loss along the frontal hairline for an unknown duration. On physical examination the frontal scalp and lateral eyebrows demonstrated decreased hair density with loss of follicular ostia. Fine, flesh-colored, monomorphic papules were scattered along the forehead and temples, and ill-defined brown pigmentation was present along the forehead, temples, and cheeks. Biopsy of the frontal scalp demonstrated patchy lichenoid inflammation with decreased number of follicles with replacement by follicular scars, confirming the diagnosis of FFA.

Patient 3—A 47-year-old Hispanic woman presented with hair loss of the frontal scalp and bilateral eyebrows with associated burning of 2 years' duration. Physical examination demonstrated recession of the frontotemporal hairline with scattered lone hairs and thinning of the eyebrows. Innumerable flesh-colored papules were present on the forehead and temples (Figure 2A). Glabellar and eyebrow erythema was noted (Figure 2B). Biopsy of

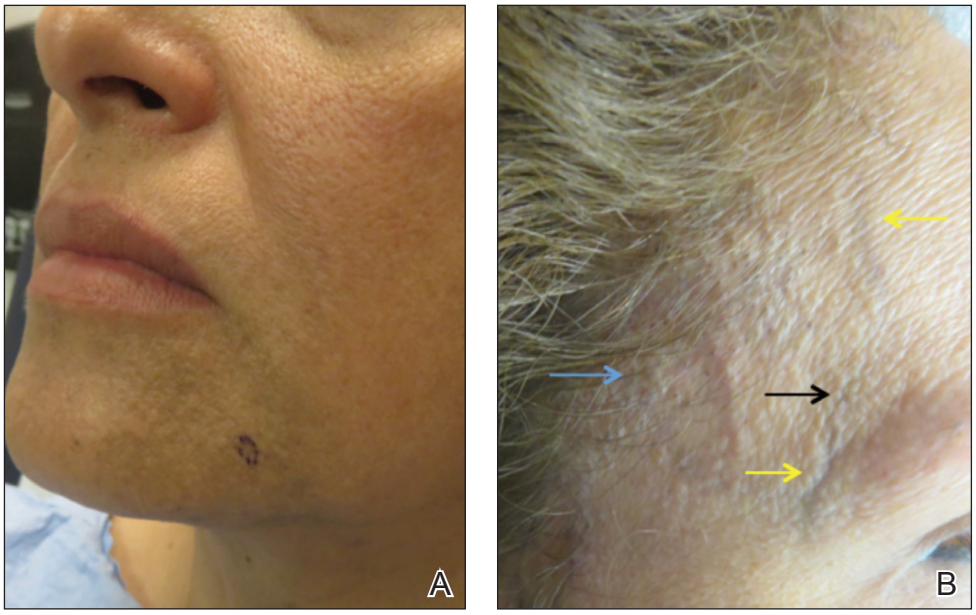

FIGURE 1. Frontal fibrosing alopecia with hyperpigmented coalesced macules around the mouth (A). Perifollicular papules on the temples (black arrow), erythematous perifollicular papules at the frontal hairline (blue arrow), and depressed veins on the forehead and temples (yellow arrows) also were noted (B).

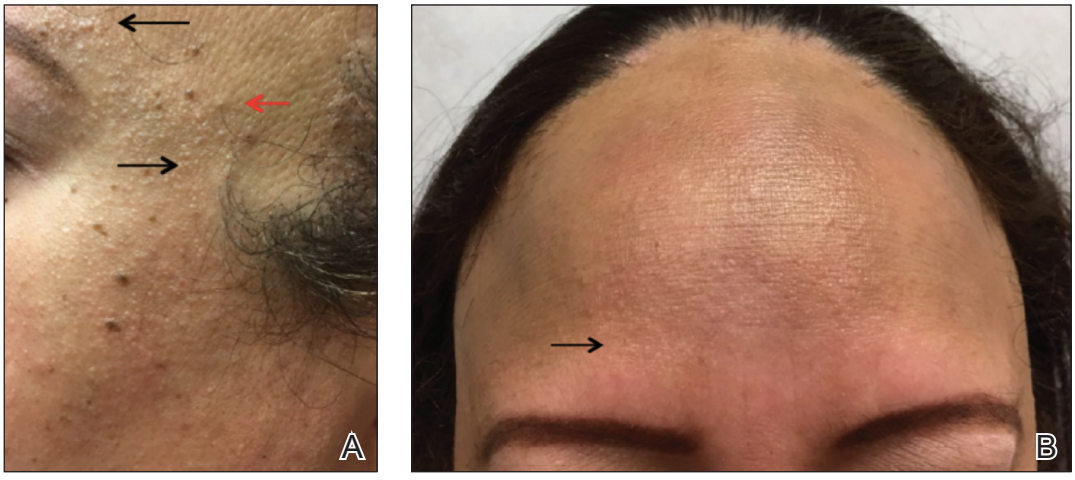

FIGURE 2. Frontal fibrosing alopecia with recession of the temporal hairline with visible lone hairs (red arrow) and scattered flesh-colored papules on the temples (black arrows)(A). Glabellar and eyebrow erythema also was noted with flesh-colored papules on the forehead (black arrow)(B). The eyebrows were notably drawn in due to decreased hair density, and the central frontal hairline was recessed.

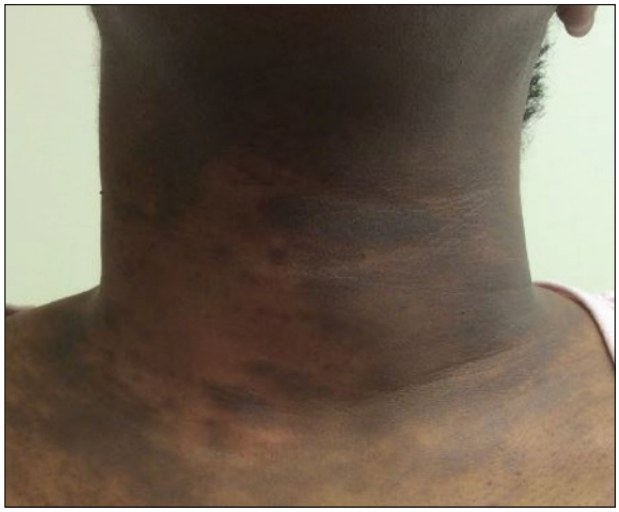

FIGURE 3. Diffuse and coalescing brown-gray macules and patches on the neck consistent with lichen planus pigmentosus.

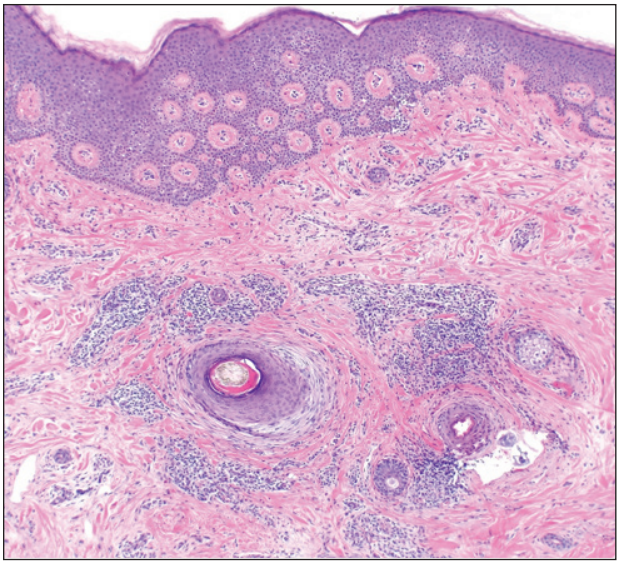

FIGURE 4. Representative photograph demonstrating a diminished number of hair follicles with partial loss of sebaceous glands. There was perifollicular fibroplasia and interface inflammation along the basement membrane of the follicular epithelium with exocytosis of lymphocytes. Low-grade vacuolar alteration also was seen along the dermoepidermal junction (H&E, original magnification ×100).

the frontal scalp demonstrated decreased terminal anagen hair follicles with perifollicular lymphoid infiltrate and fibrosis, consistent with a diagnosis of FFA. The patient was started on oral hydroxychloroquine 400 mg once daily, and 3 months later hyperpigmentation of the forehead and perioral skin was noted. The patient reported that she had facial hyperpigmentation prior to starting hydroxychloroquine and declined a biopsy.

Patient 4—A 40-year-old black woman presented with brown pruritic macles of the face, neck, arms, and forearms of 4 years' duration. She also reported hair loss on the frontal and occipital scalp, eyebrows, and arms. On physical examination, ill-defined brown macules and patches were noted on the neck (Figure 3), face, arms, and forearms. Decreased hair density was noted on the frontal and occipital scalp with follicular dropout and perifollicular hyperpigmentation. Biopsy of the scalp demonstrated perivascular lymphocytic inflammation with sparse anagen follicles and fibrous tracts, and biopsy of the neck revealed superficial perivascular inflammation with numerous melanophages in the upper dermis; these histopathologic findings were consistent with FFA and LPP, respectively.

Patient 5—A 46-year-old black woman with history of hair loss presented with hyperpigmentation of the face and neck of 2 years' duration. On physical examination decreased hair density of the frontal and vertex scalp and lateral eyebrows was noted. Flesh-colored papules were noted on the forehead and cheeks, and confluent dark brown patches were present on the temples and neck. Three punch biopsies were performed. Biopsy of the scalp revealed lymphocytic inflammation with surrounding fibroplasia with overlapping features of FFA and central centrifugal cicatricial alopecia (Figure 4). Biopsy of the neck revealed vacuolar interface dermatitis. Additionally, biopsy of a facial papule revealed lichenoid inflammation involving a vellus hair follicle. Clinical and histopathological correlation confirmed the diagnosis of FFA with LPP and facial papules.

Comment

Current understanding of FFA as a progressive, lymphocytic, scarring alopecia has expanded in recent years. Clinical observation suggests that the incidence of FFA is increasing[4]; however more epidemiologic data are needed. Frontal fibrosing alopecia presents clinically with symmetrical frontotemporal hair loss with lone hairs. Trichoscopy reveals perifollicular hyperkeratosis, perifollicular erythema, and follicular plugging in 72%, 66%, and 44% of cases, respectively.[5] In one study (N=242), patients were classified into 3 clinical patterns of FFA: pattern I (linear) showed bandlike loss of frontal hair with normal density directly behind the hairline; pattern II (diffuse) showed loss of density behind the frontal hairline; and pattern III (double line) showed a pseudo–"fringe sign" appearance. The majority of patients were classified as either pattern I or II, with pattern II predicting a poorer prognosis.[6]

Frontal fibrosing alopecia is increasingly recognized in men, with prevalence as high as 5%.[1] Facial hair involvement, particularly of the upper lip and sideburns, is an important consideration in men.[7] Most studies suggest that 80% to 90% of affected women are postmenopausal,[8] though a case series presented by Dlova[1] identified 27% of affected women as postmenopausal. The coexistence of premature menopause and hysterectomy in FFA patients suggests a hormonal contribution, but this

association is still poorly understood.[8] Epidemiologic data on ethnicity in FFA are sparse but suggest that white individuals are more likely to be affected. Frontal fibrosing alopecia also may be misdiagnosed as traction alopecia in Hispanic and black patients.[8]

It is prudent for physicians to assess for and recognize clinical clues to severe forms of FFA. A 2014 multicenter review of 355 patients identified 3 clinical entities that predicted more severe forms of FFA: eyelash loss (madarosis), loss of body hair, and facial papules.[8] Madarosis occurs due to perifollicular inflammation and fibrosis of eyelash hair follicles. Similarly, perifollicular inflammation of body hair was present in 24% of patients (N=86), most commonly of the axillary and pubic hair. Facial papules form due to facial vellus hair inflammation and fibrosis and were identified in 14% of patients (N=49).[8] These clinical findings may allow providers to predict more extensive clinical involvement of FFA.

Frontal fibrosing alopecia and LPP occur concomitantly in up 54% of patients, more commonly in darker-skinned patients.[1,9,10] Lichen planus pigmentosus frequently occurs on the face and neck, most commonly in a diffuse pattern, though reticulated and macular patterns also have been identified.[11] In some patients, LPP precedes the development of FFA and may be useful as a herald sign[1]; therefore, it is important for dermatologists to evaluate for signs of FFA when evaluating those with LPP. Thorough evaluation in patients with skin of color also is important because FFA may be misdiagnosed as traction alopecia.

Additional cutaneous associations of FFA include eyebrow loss, glabellar red dots, and prominent frontal veins. Eyebrow loss occurs secondary to fibrosis of eyebrow hair follicles and has been found in 40% to 80% of patients with FFA; it is thought to be associated with milder forms of FFA.[8] Glabellar red dots correlate with histopathologic lymphocytic inflammation of vellus hair follicles.[12] Additionally, frontal vein prominence has been described in FFA and is thought to be secondary to atrophy in this scarring process, perhaps worsened by local steroid treatments.[13] Mucocutaneous lichen planus, rosacea, thyroid disease, vitiligo, and other autoimmune disorders also have been reported in patients with FFA.[14]

Conclusion

Concomitant FFA, LPP, and facial papules have been rarely reported and exemplify the spectrum of cutaneous associations with FFA, particularly in individuals with skin of color. Clinical variants and associations of FFA are broad, including predictors of poorer prognosis such as eyelash loss and vellus hair involvement seen as facial papules. Lichen planus pigmentosus is well described in association with FFA and may serve as a herald sign that frontal hair loss should not be mistaken for traction alopecia in early stages. Eyebrow loss is thought to represent milder disease. It is important for dermatologists to be aware of these findings to understand the breadth of this disease and for appropriate evaluation and management of patients with FFA.

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