Frontal Fibrosing Alopecia Associated with Lichen Planus Pigmentosus: Not Only in Dark Phototypes

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Established Facts

- Different associated facial clinical signs have been described within frontal fibrosing alopecia.
- The association of frontal fibrosing alopecia with lichen planus pigmentosus is mainly described in dark phototypes.

Novel Insights

- Frontal fibrosing alopecia and lichen planus pigmentosus can also occur in lower Fitzpatrick phototypes.

Keywords

Frontal fibrosing alopecia · Lichen planus pigmentosus

Abstract

The association between frontal fibrosing alopecia and lichen planus pigmentosus was first described in African women. Later, most reports about this association involved dark-skinned patients. Here, we describe 5 cases of frontal fibrosing alopecia associated with lichen planus pigmentosus in light-skinned women from Argentina. Our communication highlights the strength of both entities' association also in lower Fitzpatrick phototypes.

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Introduction

Frontal fibrosing alopecia (FFA) is commonly described as a lichen planopilaris variant. It is not yet clear if these are distinct entities or belong to a spectrum of the same pathology [1]. FFA has a high aesthetic impact on patients, often affecting their quality of life severely. Since the first report in 1994 by Kossard [2], many series of patients have been published. It seems to be an epidemic type of alopecia, increasing in incidence in the last decades and mainly involving postmenopausal women. Because of the rise in its incidence, many environmental triggers had been postulated, but none had been proved as causal.

Table 1. Patients' characteristics

Pa- tient	Age, years	Gen- der	Fitz- patrick photo- type	Ethnicity	Family history (AFF)	Age at onset of LPiig, years	Facial involvement (LPiig)	Neck involvement (LPiig)	Clinical patterns of hyper- pigmentation	Age at onset of FFA, years	FFA severity [10]	Other variants of lichen planus	Skin biopsy	Scalp biopsy	Treatment	Evo- lution	Meno- pausal state	Age at meno- pause, years	Eyebrow alopecia	Alopecia of other anatomic sites	Facial papules	Associated medical diseases
1	62	F	III	Caucasian	No	62	Bilateral hyperpigmented macules on her temples, malar area, and superior eyelids	Yes	Multiple pigmented macules	62 (4 months later)	Mild (I)	No	Yes, neck; interface dermatitis and pigment incontinence (Fig. 1)	Yes	Finasteride, hydroxy- chloroquine, clobetasol, lightening cream, and sunscreen	Pro- gression of facial lesion	Post	38	Yes	No	No	Hyper- tension
2	45	F	II	Caucasian	No	42 (months later)	Bilateral periocular hyperpigmented macules that converged, forming a mask (upper eyelid involvement), and the lesions also involved the chin	Yes	Multiple pigmented macules	42	Severe (V)	No	Yes, neck; interface dermatitis with focal basal cell vacuolization and melanophages	Yes	Finasteride, hydroxy- chloroquine, clobetasol, lightening cream, and sunscreen	Stabili- zation	Pre		Yes	No	Yes	Metabolic syndrome
3	83	F	III	Caucasian	No	79	Bilateral hyperpigmented reticular tan macules on her face (upper eyelids involvement) (Fig. 2)	Yes	Reticular	75	Severe (IV)	No	Yes, neck; interface dermatitis and pigment incontinence	No	No (denied)	Lost to follow-up	Post	45	Yes	No	No	Hyper- tension
4	50	F	III	Caucasian	Yes	50	Flares of painful erythematous facial macules (cheeks) that evolved to hyperpigmented macules. She received rosacea treatment (minocycline and topical metronidazole); facial papules	No	Multiple pigmented macules	Un- known	Mild (III)	No	Yes, cheek; interface dermatitis and pigment incontinence	No	Meprednisone 40 mg qo, topical 0.1% tacrolimus, sunscreen, finasteride, clobetasol, 5% minoxidil, and methotrexate 15 mg qw folic acid	Improve- ment	Pre		Yes	No	Yes	Hypo- thyroidism
5	57	F	III	Caucasian	Un- known	44	Facial hyperpigmented macules (upper eyelids involvement) (Fig. 3)	Yes	Reticular	47	Mild (III)	No	Yes, cheek; interface dermatitis and pigment incontinence	No	Meprednisone 40 mg qo, tacrolimus, lightening cream, and sunscreen	Pro- gression of facial lesions	Post	48	Yes	No	Yes	Hypo- thyroidism

AFF, frontal fibrosing alopecia; LPiig, lichen planus pigmentosus.

Color version available online

Fig. 1. Skin biopsy taken from the neck. HE $\times 10$. Interface dermatitis and pigment incontinence. HE, hematoxylin and eosin.

Color version available online

Fig. 2. Case 3: hyperpigmented reticular tan macules involving the face and neck.

Color version available online

Fig. 3. Case 5: facial and neck hyperpigmented macules in a reticular distribution.

Different associated clinical signs have been described, including depression of the frontal veins [3] and facial papules [4], among others. Within the lichenoid spectrum, associations with cutaneous lichen planus [5], mucosal lichen planus, and lichen planus pigmentosus (LPPig) have been reported. The latter supports a shared pathologic pathway between FFA and lichen planopilaris.

Clinical features of LPPig, an uncommon variant of lichen planus, were first described in 1974 by Bhutani et al. [6] in Indian patients. The association between FFA and LPPig was first described by Dlova in 2013 in African women [7]. Since then, most publications about this association involved dark-skinned patients. Pirmez et al. [8] described the clinical and dermoscopic features of LPPig in 37 patients with FFA, most of them with high phototypes. Romiti et al. [9] reported 2 skin phototype-II cases in a series of 16 mainly darker skin phototype patients. Vaño-Galván et al. [10] published in 2014 one of the largest series of AFF in Caucasian patients. They did not report any LPPig association. This may be an indirect sign that LPPig is not common in light-skinned patients, given that most of the patients were Spanish. We report 5 cases of FFA associated with LPPig in light-skinned Argentine women.

Case Report/Case Presentation

We reviewed charts of all FFA patients seen at the Hair Clinic of CEMIC, Buenos Aires Skin, and CIAL from 2008 to 2018 in Buenos Aires and Bahía Blanca (Argentina). Evaluation of 111 patients diagnosed with FFA seen in these clinics revealed 5 patients with the diagnosis of LPPig on the face and neck. FFA was confirmed by scalp biopsy in 2 patients and LPPig by skin biopsy in all of them (Table 1; Fig. 1–3).

Discussion and Conclusion

All patients were women with light skin phototypes (Fitzpatrick phototypes II, III), aged 45–83 years. The age at onset of LPPig ranged from 42 to 79 years and of FFA from 42 to 69 years. In 3 patients, LPPig preceded FFA (4 months to 4 years). Two patients were premenopausal, and 4 had extra-facial skin involvement (neck) of LPPig. One patient had a maternal history of FFA.

Patient number 4 was diagnosed previously as rosacea because of the facial erythema, as reported by López-Pestaña et al. [11]. Some of these patients are sometimes misdiagnosed with erythematous rosacea, although they never had inflammatory papules or pustules. As published by Gavazzoni Dias et al. [12], 4 women in this series had hyperpigmentation of the upper lid, a clue to the diagnosis of facial LPPig that helps its differentiation from other dermatoses such as melasma.

The clinical variants of LPPig were reticulated in 2 patients and comprised multiple pigmented macules in the others. All the patients had eyebrow alopecia, and facial papules were present in 3 women. Recently, Mervis et al. [13] reported a significant association between Hispanic/Latino ethnicity, facial papules and premenopausal status, which may portend susceptibility to a more severe disease.

Although it was not systematically recorded in our series, dermoscopy is a great tool to help in the diagnosis of LPPig, as described by Pirmez and other authors [8]. Our patients were treated with the reported options for FFA, 5-alpha-reductase inhibitors and other drugs borrowed from the treatment of lichen planopilaris. Treatment of LPPig is generally disappointing; our patients received topical clobetasol, tacrolimus, lightening creams, and sunscreens. Two patients developed new facial pigmented lesions in spite of treatment.

The inflammatory phase of the skin lesions that affected our patient in case 4, although uncommon, should remind us that the presence of inflammatory cells in the early phases of LPPig supports the indication for early

treatment to avoid chronic persistent pigmentation [14]. Early intervention with systemic steroids or other agents such as methotrexate can probably help prevent hyperpigmentation, as in our case. Also, because of the hyperpigmentation risk associated with antimalarials, caution should be taken when using these drugs for FFA.

This brief communication highlights the strength of the FFA and LPPig association also in lower Fitzpatrick phototypes. As described by Mervis et al. [13], patients who have worse prognostic factors, such as Latino ethnicity, facial papules, and premenopausal status, may deserve a more aggressive treatment option.

Regarding the association, we can think that both entities are within the lichenoid spectrum. We also wondered which patients can develop skin pigmentation, obviously patients with dark skin and perhaps lower phototypes with high facultative pigmentation capacity.

Statement of Ethics

All subjects gave their written informed consent to publish their cases (including publication of images).

Disclosure Statement

The authors have no conflicts of interest to declare.

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Author Contributions

All authors contributed equally to this manuscript.

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