INTRODUCTION

Frontal fibrosing alopecia (FFA) represents a peculiar condition with a quasi-symmetrical, marginal alopecia along the frontal and temporal hairline with scarring. Steven Kossard is credited with the original description in 1994,[1] when he reported six postmenopausal women with progressive frontal hairline recession that was associated with perifollicular erythema within the marginal hairline, producing a frontal fibrosing alopecia extending to the temporal and parietal hair margins. Scalp biopsy specimens revealed histologic features that were indistinguishable from those seen in lichen planopilaris.[2] Ultimately, the condition has been recognized to represent a more generalized rather than localized process of inflammatory scarring alopecia, with extension beyond the frontotemporal hairline to include the parieto-occipital hairline, involve peculiar facial papules as evidence of facial vellus hair involvement,[3] and loss of peripheral body hair.[4,5] Furthermore, lichen planus-type nail involvement has been reported,[6] again pointing to a close relationship of FFA to lichen planus.

Originally considered to be an uncommon condition, the frequency of FFA has globally increased exponentially, to include premenopausal women and men,[7–11] while its etiology has remained obscure. Familial cases of FFA[12–14] point to the possible contribution of hereditary factors, maybe related to androgenetic alopecia. We report the first case of connubial frontal fibrosing alopecia in a genetically unrelated couple pointing to the possibility of a common environmental exposure in the etiology of the condition. Our observation may be fortuitous, considering the high frequency of female frontal fibrosing alopecia. Nevertheless, the incidence of male frontal fibrosing alopecia has remained low with a consequently low statistical probability of random occurrence of the condition in a marital couple. We, therefore, suggest to systematically include the hair condition of marital partners in the patient history of patients with frontal fibrosing alopecia, to elucidate the actual frequency of connubial frontal fibrosing alopecia and maybe a common causative agent or hair grooming practice.

Key words: Causative environmental agent or grooming practice, connubial frontal fibrosing alopecia, female frontal fibrosing alopecia, male frontal fibrosing alopecia

Case Report of Connubial Frontal Fibrosing Alopecia

Ricardo da Silva Libório, Ralph M Trüeb

Department of Dermatology and Radiotherapy, Botucatu Medical School, Paulista State University, UNESP, São Paulo, Brazil, ¹Center for Dermatology and Hair Diseases Professor Trüeb, Zurich, Switzerland

ABSTRACT

Since its original report in 1994, frontal fibrosing alopecia has become increasingly common, attracting the attention of the medical community and giving rise to speculations on its etiology, specifically the possibility of environmental factors. Familial cases of frontal fibrosing alopecia point to the possible contribution of hereditary factors maybe related to androgenetic alopecia. We report the first case of connubial frontal fibrosing alopecia in a genetically unrelated couple pointing to the possibility of a common environmental exposure in the etiology of the condition. Our observation may be fortuitous, considering the high frequency of female frontal fibrosing alopecia. Nevertheless, the incidence of male frontal fibrosing alopecia has remained low with a consequently low statistical probability of random occurrence of the condition in a marital couple. We, therefore, suggest to systematically include the hair condition of marital partners in the patient history of patients with frontal fibrosing alopecia, to elucidate the actual frequency of connubial frontal fibrosing alopecia and maybe a common causative agent or hair grooming practice.

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Originally considered to be an uncommon condition, the frequency of FFA has globally increased exponentially, to include premenopausal women and men,[7–11] while its etiology has remained obscure. Familial cases of FFA[12–14] point to the possible contribution of hereditary factors, maybe related to androgenetic alopecia, while a recent questionnaire-based study suggested a possible association of FFA with the use of facial skin care products, particularly sunscreens, both in women,[15] and in men.[16] We report the first case of connubial FFA in a genetically unrelated couple pointing again to the possibility of a common environmental exposure in the etiology of the condition.

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CASE REPORTS

Case 1

An 83-year-old Caucasian male originally presented in 2010 at the Center for Dermatology and Hair Diseases with a 6-month history of loss of eyebrows and receding frontotemporal hairline associated with pruritus.

The clinical examination revealed a marginal alopecia along the frontotemporal hairline and loss of eyebrows. The affected scalp skin was pale and smooth, with loss of follicular orifices, perifollicular erythema, and follicular keratosis.

A diagnosis of male frontal fibrosing alopecia was made and treatment was started with 1% topical pimecrolimus bid along the frontotemporal hairline and 5% topical minoxidil bid for the centroparietal scalp area.

At the time point of presentation of his wife, 7 years later, the marginal alopecia had extended to converge with advanced androgenetic-type alopecia (Hamilton–Norwood VII) [Figure 1].

Case 2

The patient’s 82-year-old wife presented only 7 years later at the Center for Dermatology and Hair Diseases with again a 6-month history of thinning of sideburns and eyebrows.

Clinical examination revealed initial loss of sideburns and thinning of left eyebrow [Figure 2a and b]. Dermatoscopic examination revealed perifollicular erythema, follicular keratosis, and loss of follicular ostia of affected temporal areas and along the frontal hairline, associated with diversity of hair shaft diameters within the central hair part. A diagnosis of initial frontal fibrosing alopecia with comorbid androgenetic alopecia was made.

A lesional biopsy performed by an external dermatologist was interpreted as lichen planopilaris and consistent with the clinical diagnosis of frontal fibrosing alopecia.

Medication with 0.5 mg oral dutasteride proposed by an external dermatologist was declined by the patient, and treatment was started with 1% topical pimecrolimus bid along the frontotemporal hairline and 5% topical minoxidil bid for the centroparietal scalp area.

On follow-up after 3 months of respective treatment, dermatoscopic examination showed lesser diversity of hair shaft diameter of the central hair part and no signs of follicular inflammation along the frontal hairline. The partial loss of sideburns remained unchanged.

DISCUSSION

Since its original report in 1994,[1] FFA has become increasingly common, attracting the attention of the medical community and giving rise to speculations on its etiology, specifically the possibility of environmental factors.

Steven Kossard is credited with the original description of the condition,[1] and eventually its nosological classification as a frontal variant of lichen planopilaris,[2] though there is circumstantial evidence of its existence well before 1994,[17] and the observation of cutaneous lupus erythematosus presenting as FFA[18] suggests that the pattern of clinical disease presentation may be more specific for the condition than the underlying inflammatory autoimmune reaction, whether lichen planus or lupus erythematosus. Ultimately, it has been discussed to what extent a background of androgenetic alopecia may contribute to the particular...
clinical presentation of a pattern-type of cicatricial hair loss. The reports on familial cases of FFA point to the possibility of a genetic background. Nevertheless, the localization of FFA in androgen-independent areas, the lack of evidence of associated androgenetic alopecia in some cases of FFA, and a limited success rate of anti-androgen therapy, including 5α-reductase inhibitors, all point to the fact that androgenetic alopecia represents only a facultative comorbidity of FFA.

Our observation of connubial FFA in a genetically unrelated couple, though both had evidence of associated androgenetic alopecia, points to the possibility of a common exposure in the etiology of the condition.

A recent questionnaire-based study of both women and men suffering from FFA suggests a possible association between FFA and the use of facial skin care products, particularly sunscreens. Nevertheless, the proposed cause-relationship remains doubtful for a number of reasons: First, the probably first case of FFA described in 1929 by the celebrated Swedish physician Axel Munthe (1857–1948) in his account of “The Story of San Michele” with “an exceptionally high and narrow forehead, no eyebrows” predates the use of chemical sunscreens since it was only in 1946 that Austrian-born chemist and mountaineer Franz Greiter (1919–1985) introduced with the product “Gletscher Crème” what may have been the first effective modern sunscreen that subsequently became the basis for the company Piz Buin (named after the respective mountain at the Swiss–Austrian border). Second, the study results may have been biased through patient and question selection, as well as confounding factors. Third, the statistics of the respective study have been scrutinized formally and found not to be convincing. The authors also noted an association with thyroid disease, as originally pointed out in 2003 by Trüeb, and a high frequency of positive patch tests, which they interpreted as indicative of a predisposition to immune-mediated disease. With regard to the type of immune-mediated hypersensitivity reaction, there may exist an analogy to the association of oral lichen planus with positive dental patch-testing, though the authors found positive patch tests in women with FFA mainly to fragrances, which again may be questionable in its cause-relationship due to the more frequent exposure found in women with FFA than in the controls.

Finally, our observation of connubial FFA may be fortuitous, considering the high frequency of female FFA. Nevertheless, the incidence of male FFA has remained low with a consequently low statistical probability of random occurrence of the condition in a marital couple. Moreover, there exists an analogy of our observation with connubial androgenetic alopecia in a female involuntarily exposed to topical testosterone gel used by her spouse. We therefore suggest to systematically include the hair condition of marital partners in the patient history of patients with FFA, to elucidate the actual frequency of connubial FFA, and maybe a common culprit, whether an environmental agent or hair grooming practice that has as yet to be identified.

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**Conflicts of interest**

There are no conflicts of interest.

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