Frontal fibrosing alopecia: when the treatment is worse than the disease

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Summary

Frontal Fibrosing Alopecia (FFA) is a type of primary lymphocytic scarring alopecia, clinically characterized by a distinctive pattern of progressive frontotemporal hairline recession and eyebrow loss. The etiopathogenesis of FFA is unknown, but it is widely considered a variant of lichen planopilaris usually, but not exclusively, affecting postmenopausal women. Current treatment aim to stop the disease progression, but no standardized treatment are recommended, and best results from observational studies have been reported for 5-α reductase inhibitors and intralesional corticosteroids, followed by antimalarials and calcineurin inhibitors. Here, we report a 52-year-old postmenopausal woman with a 7-year history of FFA, who experienced a severe worsening after treatment with intralesional triamcinolone acetonide, for the occurrence of steroid induced skin atrophy. Whether the physician has undervalued the well-known side effect of intralesional steroids or the peculiar fibrosing tendency of the disease has played a role is difficult to assess. No previous report of steroid induced atrophy in FFA was retrieved from the current literature. This case draws attention on the importance to properly estimate the consequences of medical interventions in disorders with exclusive aesthetic impact.

KEY WORDS: frontal fibrosing alopecia; cicatricial alopecia; follicular lichen planus.

Introduction

Frontal Fibrosing Alopecia (FFA), firstly described by Kossard in 1994, is an increasingly common form of acquired primary lymphocytic scarring alopecia, characterized by fronto-temporal hairline recession, frequently accompanied by eyebrow loss (1). Clinical presentation is distinctive, for the loss of follicular units on an uniformly pale skin, with perifollicular erythema and follicular keratinization of the remaining hairs at the hairline, well-demarcated from the tanned respected front-line (2-7). FFA is considered a variant of lichen planopilaris (LPP), mainly for the similar histopathological features, such as a reduced number of hair follicles and perifollicular lymphoid cell infiltrate with perifollicular fibrosis (2, 8). Speculation about a hormonal origin, mainly affecting postmenopausal women or with a history of hysterectomy, and early menopause was questioned for the publication of cases in men and premenopausal women; but somewhat proposed again by the efficacy of 5-α reductase inhibitors and topical minoxidil treatment (9, 10). No evidence-based treatment is recommended for FFA, and observational studies suggest similar efficacy for 5-α reductase inhibitors, antimalarials, calcineurin inhibitors, and intralesional steroids, which remain a very popular choice in daily practice (2, 11-13). Treatment aim at stopping the recession, but the disease stabilize with time in most of the patients, with or without treatment, and it is not clear whether the natural history of the disease is really improved by interventions. Thus, FFA is often a frustrating condition, for both patient and dermatologist. We report a case in which the treatment (intralesional steroid injections) was worse than the disease (FFA).

Case report

A 52-year-old postmenopausal woman presented with a 7-year history of asymptomatic fronto-parietal hair recession and marked decrease of the eyebrows. Family history for FFA or LPP was negative and laboratory tests including thyroid function, serological markers for viral hepatitis B and C, screening for syphilis infection and antinuclear antibodies were negative. She reported prior treatment, about 6 months before our evaluation, with oral dutasteride 0.5 mg daily and intralesional triamcinolone acetonide, every 15-20 days, for a total of 5 injections. Band-like irregularity of the frontal skin surface had appeared 3 months before our observation, and had slowly but progressively widen. Physical examination revealed a band of symmetrical...
recession of the frontoparietal hairline extending to the preauricular areas, suggestive of frontal alopecia, but associated with multiple patches of thin pale skin (Figure 1). The atrophic patches, adherent to the subcutaneous planes, and often overlapped by the superficial veins, were surrounded by the normal frontal skin, which by contrast appeared protruding of several millimeters. Worst atrophic lesions involved the eyebrows, completely altering the facial physiognomy. There were not perifollicular erythema and follicular keratinization within the marginal hairline, residual hairs appearing normal, although reduced in density. There was no involvement of the trunk, extremities, mucosae or nails. Stating the apparent remission of the frontal fibrosing alopecia, we decide to wait and see, locally applying a silicon based gel, 100% dimethicone, normally used for hypertrophic scars. After 6 months a certain improvement had occurred (Figure 2), especially on the frontal and right sides of the hairline, but with persistence of a wide atrophic plaque on the left side, extending to the temporal periocular region, and bilaterally disfiguring the eyebrows. No apparent relapse of FFA was noted. The patient was than referred to the plastic surgeon for the correction of the superior palpebral defects.

**Discussion**

Skin atrophy after local corticosteroid injections is a well-known and dread side effect, especially in highly aesthetic impact areas (14-16). However, the development of linear patchy areas of skin atrophy on the hairline and frontal skin was not retrieved on English language literature (medline search from 1981 to 2016). Skin atrophy occurs for a direct anti-proliferative effect on keratinocytes and fibroblasts, perturbing the extracellular matrix protein metabolism, combined with enhanced vasoconstriction, with resultant local tissue hypoxia (17). A lymphatic spread of the steroidal suspension is also involved to explain the typically linear pattern of the lesions (18, 19). The natural tendency towards atrophy of the FFA, which is included among primary scarring alopecia, might have contributed to the extension and disfiguring results of the steroidal local injections. Besides, this type of intervention is among the most popular to treat FFA, and the adverse event should have been observed previously. Probably the steroid concentration was not opportunely diluted, but the patient had not medical report or further information on the performed procedure.

*Figure 1 - A band of symmetrical recession of the frontoparietal hairline extending to the preauricular areas, suggestive of fibrosing frontal alopecia associated with multiple patches of thin pale skin configuring a cobblestone appearance.*

*Figure 2 - The same patient showing some improvement after 6-month follow up.*
Our patient was typical for FFA presentation, with postmenopausal onset of a 2 cm band of symmetrical fronto-tempo-parietal hairline recession, and wide eyebrows loss. A grade II severity of FFA was diagnosed, following recent classification proposed by Vañó-Galváan et al. (4). Family history was negative, as in the majority of FFA patients (2, 4). At referral, there were no signs of active disease, such as perifollicular erythema and follicular keratinization within the marginal hairline. The patient refused to take a biopsy for histological examination, hardly upset by her disfiguring condition. Histopathological features of FFA are generally indistinguishable from lichen planopilaris, with a lichenoid lymphocytic infiltrate, with vacuolar degeneration of basal keratinocytes, mainly localized to the isthmus, infundibular and bulge portions of the hair follicles (2, 8). Lamellar or perifollicular fibrosis is observed around the hair follicle and there is a reduction in the number of hair follicles and replacement by fibrous tracts. According to Tosti et al., differently from LPP, in FFA the lymphocytic infiltrate and fibrosis affect selectively the intermediate and the vellus-like follicles of the frontal margin. The reason for this selective involvement is still unknown (3). Dermoscopy is a valuable tool to support clinical diagnosis (20, 21), but in our patient, we did not document perifollicular erythema, and peripilar casts, further confirming the apparent inactivity of the disease.

The previous oral dutasteride treatment, as well as the various cycles of local and finally the intralesional steroids might have induce the remission of FFA. Currently, no evidence-based treatment protocols exist for the treatment of FFA, but a recent systematic review suggests that 5-alfa-reductase inhibitors, finasteride and dutasteride are the most effective drugs in stabilizing the disease, followed by intralesional and topical corticosteroids (2, 12). It is otherwise known that the disease tends to stabilize with time, and no conclusions about the real effects of the treatments have been reached for FFA (2, 10-13).

In conclusion, the dramatic iconography underlines the risk of a widely diffused procedure, especially in very visible areas, such as the face. Skin atrophy generally improve with time, and discontinuation of the steroid, but any residual damage has a high aesthetic impact on the patient’s quality of life, and self-image. Consequences are more severe, considering the natural course of the FFA, and the absence of evidence-base treatment guidelines. Clinicians should be very well aware of this side effect of intralesional steroids, when treating FFA, and always remember the Hippocratic Oath: «Primum nil nocere».

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