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Peculiar hypertrichosis in a patient affected by Frontal fibrosing alopecia with pseudo “fringe sign”

Sara GRASSI MD[1], Marta CARLESIMO MD[1], Maria Caterina FORTUNA MD[1], Alfredo ROSSI MD, PhD[1]

[1]Department of Internal Medicine and Medical Specialties, "La Sapienza" University of Rome, Italy.

Corrisponding author:
Sara Grassi, M.D.
Department of Internal Medicine and Medical Specialties,
"Sapienza" University of Rome,
Piazzale Aldo Moro, 5, 00185 Roma RM
e-mail sara.grassi03@gmail.com

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Letters to the Editor

Frontal fibrosing alopecia (FFA) was firstly described by Kossard S. in 1994 as scarring alopecia characterized by progressive and symmetric recession of frontal or frontotemporal hairline, frequently associated with partial or complete eyebrow loss, and with facial skin-coloured papules.[1] Incidence of FFA has remarkably increased in the last decade and its pathophysiology is still mainly unknown. Other than the typical presentation with symmetrical frontotemporal hairline recession, some unusual clinical pattern have been recently described.[2,3]

We report a case of a 60-years-old otherwise healthy Caucasian female who presented with a band of frontotemporal hair loss. Pronounced bitemporal hairline retrocession with maintenance of rim along the temporal hairline was observed (Figure 1A,C,D). Thinning of lateral eyebrow aspects were also present. Trichoscopical examination revealed absence of follicular openings, some scattered terminal hairs, and mild perifollicular erythema at margins of involved hairlines (Figure 1B). On the basis of clinical and trichoscopical findings, diagnosis of FFA was assessed. Treatment with galenic lotion composed of 2% minoxidil, 0.08% hydrocortisone butyrate, 0.05% 17α-estradiol, 16% ciclosilicone pentamer, 96° alcohol to apply 2 ml/die was prescribed.

At the follow up visit, after 6 months of treatment, the patient presented with increased hair at the anterior margins of the frontotemporal scarring band, configuring a localized hypertrichosis. In the context of scarring area a few scattered terminal hairs were present (Figure 2).

Recently, Pirmez R.[2] reported in 7 patients a pattern of FFA presenting with more pronounced bitemporal involvement with unusual retention of the hairline, configuring the so called pseudo "fringe sign". We observed this peculiar pattern in a postmenopausal female otherwise healthy patient. To date there are no universal treatment guidelines for FFA. A few findings on different treatment approaches have been reported; topical or intralesional steroids, topical minoxidil lotion, oral tetracyclines, hydroxychloroquine, pimecrolimus and hormonal treatments (finasteride, dutasteride) have shown efficacy in order to stabilize the disease in small case series.[4,5] Recently,

the efficacy of topical galenic lotion that combined 2% minoxidil, with 0.08% hydrocortisone butyrate, 0.05% 17α-estradiol, in 16% ciclosilicone pentamer and 96° alcohol has been reported.[6] Minoxidil promotes hair growth, beyond the vasodilatory effect, seems to have angiogenetic properties, the ability to increase cell proliferation and DNA synthesis, and to inhibit collagen synthesis, antiandrogen and immunosuppressive effects. Hydrocortisone butyrate, a II class steroid, aims to block the inflammatory process, induced by lymphocytic perinfundibular and peristhmus infiltrate. Moreover, oestrogens operate as potent hair growth modulators and as hair protective factors 17α-estradiol is an oestrogen that has an effect on the measurable aromatase activity.

Hypertrichosis is a well-known side effect of minoxidil topical solution (MTS). The mechanism for MTS-mediated hypertrichosis is actually not well known, also if different hypotheses have been proposed, such as accidental manual or fomites (e.g. pillows) spread of the post-application minoxidil residue on face and/or hands, a dose-dependent effect resulting from systemic absorption, and a possible run-off of the applied dose.[7]

In our case hypertrichosis induced by MTS appeared with peculiar features, with a remarkable increasing of vellus-like adjacent the alopecic area, and of terminal hair in the context of scarring band. To the best of our knowledge, hypertrichosis with this features has never been reported in patients affected by FFA.

We may speculate that the possible explanation of the peculiar hypertrichosis observed in our case may be searched in histopathologic features of the disease. Indeed, vellus-like hair follicles are irreversibly replaced by fibrosis in the area affected, so that minoxidil hair growth-induced may effect only the adjacent non-affected hair follicles. The presence of terminal-like hair follicle in the context of fibrotic area could be explained by the fact that the inflammatory lymphocyte infiltrate in FFA affects and destroys mainly hair follicles in the upper part of the dermis, as it has been shown by some Authors.[5] To date, the target to which activated lymphocytes are directed is still unknown,

but probably it should be searched among proteins selectively expressed in vellus and intermediate hair follicles, at least as regards the initial stages of the disease.

Further studies are needed for a better comprehension of pathophysiological mechanisms of FFA in order to develop effective therapeutic strategies for this scarring condition whose incidence is continuously increasing.

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Figure legends

Figure 1. Bilateral frontotemporal hairline recession with more pronounced bitemporal involvement in a 60-years-old female patient. Note the presence of a rim of hairline sparing along the temporal region (*red arrows*) (**A**). Trichoscopical examination reveal single-hair pilosebaceous units, mild perifollicular erythema and scaling (*blue arrows*), white patches with lack of follicular openings (*green circle*), (**B**). The residuated alopecic area show atrophic pale features, typical of frontal fibrosing alopecia with few scattered terminal hair "*lonely hair*" (**C, D**).

Figure 2. An increasing of hair at the anterior margins of the frontotemporal scarring band (*red arrows*) at the follow up visit, after 6 months of treatment with topical lotion composed galenic lotion composed of 2% minoxidil, 0.08% hydrocortisone butyrate, and 0.05% 17α-estradiol can be observed. Localized hypertrichosis (**A**). Trichoscopical examination scant/absence of perifollicular erythema and scaling (**B**). Note the presence of few scattered terminal hair (*yellow arrows*) in the context of scarring area (**C**, **D**).