INTRODUCTION

Frontal fibrosing alopecia (FFA) represents a peculiar condition with a quasi-symmetrical, marginal alopecia along the frontal and temporal hairline with scarring. When FFA was first described by Axel Munthe in 1929,[1] it must have been sporadic and ignored, until Kossard’s original report in 1994 of six postmenopausal women affected by the condition.[2] Since then, cases have multiplied worldwide exponentially, resulting in FFA eventually representing the most frequent cause of primary scarring alopecia these days. The pattern of hair loss and frequent involvement of eyebrows has a substantial psychological impact on patients, since the high forehead, the receding temporal hairline and the missing eyebrows are reminiscent of the hairstyles of the 15–16th centuries. Indeed, it has been proposed that the Duchess of Urbino may have suffered from the condition[3] while it is more probable that we are dealing with fashionable or cultural forms of frontal pseudoalopecia[4,5] than a true pathologic condition, since the hair was then shaved around the hairline or plucked at the temples and the napes of the neck, and the condition later disappeared from the arts.

Obviously, there is a need for effective therapy, while the loss of hair is usually permanent, due to a lichen planopilaris (LPP)-like inflammation with fibrosis.[6] Due to the localization of the alopecia, a hair prosthesis is rather inept. Therefore, the last resort is hair transplantation surgery. Since the underlying pathology is usually LPP, reservations and caveats have been expressed with respect to the risk of köbnerization phenomena following hair transplantation surgery. An important question that arises is how the lichenoid tissue reaction pattern is generated around the hair follicles in FFA. Follicles with some form of damage or malfunction might express cytokine profiles that attract inflammatory cells to assist in damage repair or in the initiation of apoptosis-mediated organ deletion. Alternatively, an as yet unknown antigenic stimulus from the damaged or malfunctioning hair follicle might initiate a lichenoid tissue reaction in the immunogenetically susceptible individual. Therefore, it might be expected that the transplantation of whole healthy hair follicles might less give rise to an inflammatory reaction than the disease itself, as revealed in our case report of successful hair transplantation in FFA.

Key words: Frontal fibrosing alopecia, hair transplantation, lichenoid tissue reaction
We report a patient with FFA, in whom autologous hair transplantation was successfully performed despite evidence of active disease.

CASE REPORT

A 57-year-old female patient suffering from FFA since 9 years presented for a regular follow-up examination 2 years after successful autologous hair transplantation (follicular unit extraction) at the frontal hairline [Figure 1a-d]. Dermoscopic examination revealed evidence of persistent follicular inflammation (perifollicular erythema and casts) at the site of FFA [Figure 2a], whereas the hair transplants did not [Figure 2b]. The patient was initially treated with 1% pimecrolimus cream b.i.d. along the frontal hairline, and later with dermoscopically guided intralesional triamcinolone acetonide at the sites of active disease, as formerly described.[10,11]

DISCUSSION

Kossard himself interpreted FFA as a frontal variant of LPP on the basis of his histopathological studies.[6] LPP is regarded to be a T-cell-mediated autoimmune reaction that triggers apoptosis of the follicular epithelial cells. This autoimmune process is thought to be in response to some antigenic challenge, but a specific antigen has yet not been identified. Harries et al.[12] provide the first evidence that LPP may result from an immune privilege collapse of the hair follicle’s epithelial stem cell niche. Where a causal or triggering agent is identified, this is termed a lichenoid reaction rather than lichen planus. The etiology of FFA has remained elusive, though some observations may provide clues to its pathogenesis. The predilection for postmenopausal women and the pattern of alopecia have suggested hormonal factors, which so far have not been identifiable. Moreover, the condition has meanwhile also been described in premenopausal women and in men,[13-17] though with a significantly lesser frequency. The observation of both familial FFA[18-20] and more recently, connubial FFA[21] suggest both genetic and environmental factors, related to either androgenetic alopecia[22] or to the use of cosmetics and sunscreens,[23,24] though the latter has been challenged.[25]

An important question that arises is how the lichenoid tissue reaction pattern is generated around the hair follicles in FFA. Follicles with some form of damage or malfunction might express cytokine profiles that attract inflammatory cells to assist in damage repair or in the initiation of apoptosis-mediated organ deletion. Alternatively, an as yet unknown antigenic stimulus from the damaged or malfunctioning hair follicle might initiate a lichenoid tissue reaction in the immunogenetically susceptible individual.[26] Remarkably, in healthy murine skin clusters of perifollicular macrophages have been described as perhaps indicating the existence of a physiological program of immunologically controlled hair follicle degeneration by which malfunctioning follicles are removed by programmed organ deletion.[27] It has been proposed that various forms of clinically perceptible, permanent alopecia might represent pathological exaggeration of this type of programmed organ deletion, resulting in a lichenoid tissue reaction pattern and scarring. Therefore, it might be expected that the transplantation of whole healthy hair follicles might less give rise to an inflammatory reaction than the disease itself, as revealed in our case report of successful hair transplantation in FFA.

In any case, care should be given that the donor area for transplantation is checked for evidence of follicular inflammation,[20] follicular unit test grafting should precede the procedure,[29,30] and the patient should be carefully followed up posttransplantation, while at the same time active disease in the proximity of the transplants is actively

Figure 1: Frontal fibrosing alopecia (a) before, and (b-d) after autologous hair transplantation (performed by Beatrice Banholzer, Zurich, Switzerland)

Figure 2: Dermoscopic view on (a) active frontal fibrosing alopecia (perifollicular erythema and follicular casts), and (b) hair transplants without evidence of inflammation despite the active disease
treated, preferably with a topical calcineurin inhibitor that does not impair wound healing.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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**Conflicts of interest**

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