Postmenopausal frontal fibrosing alopecia

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Summary

Recently a new entity, postmenopausal frontal fibrosing alopecia, was added to the established subtypes of scarring alopecias affecting postmenopausal women. This condition is characterized by a progressive frontal hairline recession associated with scarring. We studied the clinical and histopathologic features in four women with this disorder. Of note, a history of bilateral oophorectomy in two of them appears to be a new association. All four cases had frontoparietal recession of the hairline and two of them also had loss of their eyebrows. None of our four patients had any mucous membrane or other skin lesions. Histological examination showed perifollicular fibrosis and lymphocytic inflammation around the isthmus and infundibular areas of the follicles. No effective treatments have emerged for this type of postmenopausal alopecia, but progression of the hair loss and scarring appears to be self-limiting. We believe that this condition is a distinct clinicopathological variant of lichen planopilaris.

Report

Recently, a form of alopecia in postmenopausal women characterized by a typical band-like loss of hair on the frontoparietal scalp associated with scarring has been described. The disorder has been considered to be a distinct variant of lichen planopilaris. Here, we report four new cases. Two of them are the first reported cases with frontal fibrosing alopecia following bilateral oophorectomy.

All four women presented with frontal recession extending to the marginal temporal and parietal areas of the scalp associated with scarring.

Case 1

A 44-year-old-woman presented with a progressive band-like loss of hair over a period of 1 year (Fig. 1a). Her family history was unremarkable. She had undergone a hysterectomy with bilateral oophorectomy 8 years previously. The fronto-parietal band of alopecia was 3 cm wide. No androgenetic hair thinning was

observed. She also had alopecia of the eyebrows. There was no associated scalp scaling or itch. She was treated with fluocinolone acetonide 0.05% topical cream applied twice daily for 1 year without improvement.

Case 2

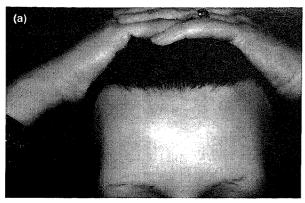
A 75-year-old woman had slowly progressive alopecia for more than 2 years. She was not on any medication and was otherwise well. The hair margin had receded 4 cm from the original hairline. The symmetrical band of marginal alopecia was uniformly pale with loss of the follicular orifices, in contrast with the sun damaged skin on her forehead. She did not have alopecia at other sites, although diffuse androgenetic hair thinning of the scalp was noted. She was treated with oral prednisone (1 mg/kg per day) for 3 months; the hair loss ceased but further progression occurred after corticosteroids were stopped.

Case 3

A 43-year-old Afro-Caribbean woman presented with fronto-temporal alopecia, that had slowly progressed over a period of 1 year. She had undergone a hysterectomy with bilateral oophorectomy 2 years previously. The hair margin had receded by about 3 cm. The vertex

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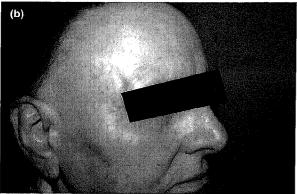


Figure 1 (a) Case 1. Fronto-temporal scarring alopecia showing marked recession of hairline and loss of eyebrows. (b) Case 4. Pale skin in area of frontal recession associated with scarring and loss of eyebrows.

did not show any alopecia or hair thinning as seen in androgenetic alopecia, neither were there any signs of patchy alopecia suggestive of lichen planopilaris or pseudopelade. She denied any history of scalp trauma or hair traction. She was asymptomatic. Follow-up at 6 months showed no improvement with application of 2% minoxidil solution applied twice daily.

Case 4

The patient was a 67-year-old woman who presented with frontal hair loss and marked alopecia of the eyebrows (Fig. 1b) that had occurred over 1 year. She was not on any medication and was asymptomatic. The fronto-parietal band of alopecia was 4 cm wide. Mild androgenetic alopecia was also observed. Initially, she was thought to have alopecia areata and was treated with triamcinolone acetonide 0.025% topical cream applied twice daily for 6 months, but without improvement. Finally, the histopathological features revealed the evidence of cicatricial alopecia.

Laboratory studies in all patients, including full blood count, erythrocyte sedimentation rate, thyroid function tests and fasting blood glucose were normal. Antinuclear antibodies were negative in all cases and serum androgen levels were also normal in all women.

Skin biopsy specimens from the area of frontal alopecia in all four patients showed similar histopathological findings. The number of hair follicles was markedly reduced and replaced by fibrous tracts. Mild to moderate lymphocytic infiltration was present around the isthmus and infundibular areas of the follicles. There was also perifollicular lamellar fibrosis. We also found increased fibrous tracts in the subcutis that extended throughout the reticular dermis at the sites of destroyed follicles. The interfollicular epidermis was normal, without signs of lichenoid inflammation (Fig. 2). Direct immunofluorescence in all cases was negative.

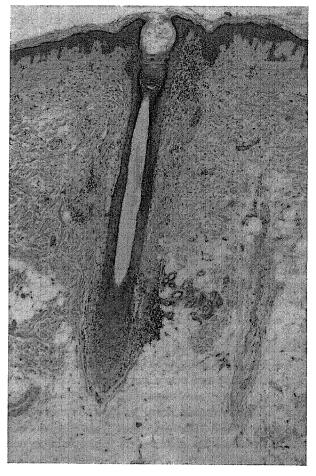


Figure 2 Horizontal section of scalp biopsy specimen demonstrating perifollicular lymphocytic inflammation at the isthmus with sparing of the interfollicular epidermis. (haematoxylin & eosin, original magnification \times 100).

In 1994 Kossard reported six postmenopausal women with frontal fibrosing alopecia;1 only 27 reports have been described since then.²⁻⁷ Two patients were premenopausal but the vast majority of women have been postmenopausal, aged between 50 and 85 years (the average age was 66 years). Typically, they presented with a band-like area of scarring alopecia 3-7 cm wide that evolved over the course of 6 months to 10 years. Alopecia of the eyebrows was described in 14 patients and only four patients had other skin or mucous membrane lesions. Specifically, Feldman et al. reported one patient with postmenopausal frontal fibrosing alopecia and oral lichen planus,2 Trueb et al. described one patient with vulvar lichen sclerosus and another patient with lichen planopilaris of the scalp,5 and Faulkner et al. described a case of frontal fibrosing alopecia with cutaneous lichen planus.7 Hormonal levels were normal in all cases and antinuclear antibodies were negative except in four patients with levels ranged between 1:40 and 1:160 with anti-DNA antibodies negative in all cases. 1.3

One of the most important aspects of postmenopausal frontal fibrosing alopecia is its differential diagnosis. Lichen planopilaris of the scalp is usually associated with multifocal areas of scarring alopecia that may coalesce. However, it appears that postmenopausal fibrosing alopecia represents a localized clinical variant of lichen planopilaris predominantly affecting the fronto-parietal hair margin.^{3,8} The differences between both conditions are the postmenopausal age and specific location in frontal fibrosing alopecia in contrast with the classic form of lichen planopilaris. Also, multifocal lichen planopilaris may be associated with evidence of lichen planus at other sites in up to 50% of patients,⁷ but in the majority of the women with frontal fibrosing alopecia described there is no associated multifocal pattern or other evidence of lichen planus. Moreover, the scarring alopecia associated with the Piccardi-Lassueur-Graham-Little syndrome has been described as multifocal and patchy and has not been specifically defined as a frontal fibrosing alopecia. 9,10 Possible confusion with alopecia areata only occurred in one of our patients who had a sudden onset of hair loss in the scalp and eyebrows. Histologically, in contrast with alopecia areata, scalp biopsy in postmenopausal fibrosing alopecia demonstrates a lichenoid lymphocytic reaction in the upper follicles, while in alopecia areata the lymphocytic infiltrate is peribulbar. 3,11 Distinction from lupus erythematosus, traction alopecia and androgenetic alopecia is usually possible on clinicopathological grounds. $^{9-11}$

Treatment of postmenopausal frontal fibrosing alopecia is usually disappointing, although oral steroids may temporarily slow the course. Nevertheless, Kossard noted that alopecia in some patients may stabilize with time.³

We think that this form of scarring alopecia is more frequent than the current literature suggests and that postmenopausal frontal fibrosing alopecia represents a localized form of lichen planopilaris with loss of the fronto-parietal implantation hairline and in some cases, loss of the eyebrows.

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