Patchy Frontal Fibrosing Alopecia: Description of an Incomplete Clinical Presentation

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Established Facts

- Frontal fibrosing alopecia is progressive and affects the vellus hair at the implantation line on the scalp.

Novel Insights

- We present cases with incomplete presentations, namely circumscribed plaques, the absence of inflammatory peripilar signs, and slow progression.

Keywords

Alopecia · Localized alopecia · Circumscribed alopecia

Abstract

We describe 6 incomplete cases of frontal fibrosing alopecia (FFA). All patients were postmenopausal women. Their average age was 60 years; 2 of the women were Caucasian, and the remaining 4 women were of a mixed ethnicity. The disease duration ranged from 3 to 17 years. The patients' lesions presented as irregular alopecic plaques in the frontal and/or temporal regions, with few signs of perifollicular inflammation, slow progression, and progressive loss of the vellus hair in the frontal area. Loss of the eyebrows, facial hyperpigmentation, and loss of the body hair were observed in 3 patients, 1 patient, and 2 patients, respectively. Localized pruritus and dysesthesia were reported in 5 patients and 3 patients, respectively. Dermato-

scopic examination indicated the absence of viable ostia at the centers and edges of the lesions and the absence of peripilar erythema; moreover, peripilar hyperkeratosis was less intense than it would be expected in classical cases of lichen planopilaris (LPP) and FFA. Histological examination revealed that all cases featured the presence of a perifollicular lichenoid infiltrate in the infundibulum and isthmus as well as concentric areas of fibrosis. All samples were subjected to direct immunofluorescence (DIF), which produced negative results. FFA has been recently diagnosed and is considered a subtype of LPP. FFA can present with incomplete clinical manifestations, a histology similar to LPP, and negative DIF findings. The correct diagnosis of FFA-associated lesions is critical for providing appropriate treatment, developing prognostic definitions, and excluding other types of scarring and nonscarring alopecia. The diagnosis of incomplete FFA is essential to avoid diagnostic and therapeutic errors.

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Introduction

Frontal fibrosing alopecia (FFA) is a primary lymphocytic scarring alopecia with a typical clinical pattern of progressive frontotemporal hairline recession and eyebrow loss. This condition, which was first described by Kossard in 1994, mainly affects postmenopausal women.

Pirmez et al. [1] described the first cases of atypical FFA involving the presence of a pseudo-fringe pattern.

At present, FFA tends to be regarded as a widespread disease that affects the vellus hair on the face and body and can be associated with follicular facial papules [2–4].

FFA is considered to be a variant of lichen planopilaris (LPP), and there has been a marked increase in the number of patients diagnosed with FFA in recent years [2–4].

The pathogenesis of FFA is unknown, although an autoimmune reaction appears to play a role [4].

Case Presentation

We examined 6 cases of FFA with incomplete presentations.

All patients described in this study were postmenopausal women. The average age of these women was 60 years; 2 of the women were Caucasian, and the remaining 4 women were of a mixed ethnicity. The disease duration ranged from 3 to 17 years. The patients' lesions presented as irregular alopecic plaques that affected the frontal and/or temporal regions, with few signs of perifollicular inflammation and slow progression but progressive loss of the vellus hair in the frontal area (Fig. 1). Loss of the eyebrows, facial hyperpigmentation, and loss of the body hair were observed in 3 patients, 1 patient, and 2 patients, respectively. Localized pruritus and dysaesthesia were reported in 5 patients and 3 patients, respectively. Dermatoscopic examination revealed the absence of viable ostia at the centers, and on the edges of lesions, the presence of peripilar erythema and peripilar hyperkeratosis that was less intense than it would be expected for classical LPP and FFA (Fig. 2). With respect to the presence of associated autoimmune diseases, 5 patients presented with hypothyroidism and were using levothyroxine; 1 patient had previously presented with discoid erythematosus lupus (DEL) on the scalp, which was currently under control; and 1 patient was HIV positive and was receiving antiretroviral therapy, with good viral and immunological control.

Fig. 1. Alopecia patches in frontal and temporal areas, preserving the frontal implantation hairline and eyebrows (a–e) and affecting the eyebrows (f).

Fig. 2. Trichoscopy in the alopecic area. Lack of follicular ostia, white dots, and transition to the normal area. Very discrete or absent perifollicular erythema and scales, usually seen in lichen planopilaris or frontal fibrosing alopecia.

Color version available online

Fig. 3. Horizontal section of the transition between alopecic and nonalopecic areas (hematoxylin and eosin). Reduction or absence of follicular units and sebaceous glands. Perifollicular lymphocytic lamellar fibrosis.

Color version available online

Discussion

Histological examination revealed that all cases featured the presence of perifollicular lichenoid infiltrates in the infundibulum and isthmus as well as concentric areas of fibrosis; these manifestations have been described in previous studies of LPP/FFA (Fig. 3) [2–4]. All samples were subjected to direct immunofluorescence (DIF), which produced negative results. This findings contrast with those of prior studies that found negative DIF results in all cases of FFA but positive DIF results and the presence of colloid bodies in certain cases of LPP [3].

The cases in our series presented with incomplete manifestations, including unevenly distributed irregular alopecic plaques, the presence of lonely hairs, few inflammatory signs, and slower progression compared with that observed in classical forms of FFA.

Knowledge of these incomplete/atypical forms of FFA is essential for the differential diagnosis of scarring and nonscarring alopecia, the determination of prognostic definitions, and the development of appropriate therapies.

Statement of Ethics

All patients agreed with the publication of their history and photos and signed the consent form.

Disclosure Statement

The authors have no conflicts of interest to disclose.

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