Frontal fibrosing alopecia – A Case report*

Alopecia frontal fibrosante - Relato de caso

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Abstract: Frontal fibrosing alopecia is a kind of progressive and frequently irreversible cicatricial alopecia marked by a lichenoid infiltrate in histology. Since its first description, in 1994, in Australia, some cases have been documented all over the world. The article reports, for the second time in the medical literature, a Brazilian case and reviews the main aspects of this dermatosis.
Keywords: Alopecia; Atrophy; Hair; Menopause

Resumo: A alopécia frontal fibrosante é uma forma de alopécia cicatricial progressiva e, frequentemente, irreversível, marcada por um infiltrado liquenóide na histologia. Desde sua primeira descrição, em 1994, na Austrália, alguns casos têm sido documentados em todo o mundo. O artigo relata, pela segunda vez na literatura, um caso brasileiro e revisa os principais aspectos desta dermatose.
Palavras-chave: Alopécia; Atrofia; Cabelo; Menopausa

INTRODUÇÃO

Described for the first time by Kossard, in 1994,[1] postmenopausal frontal fibrosing alopecia has become less and less unusual than it was supposed to be[2,3]. The disease was initially considered exclusive of postmenopausal women,[1,2] but it has been re-named after reports of cases in women of childbearing age and men [4,5,6,7]. However, it still remains more frequent in women, with an average age of 66 years.[7,8,9]

Its etiology still remains a mystery. The fact that the disease has appeared recently increases the possibility of environmental factors [10] being involved with its etiology. The majority of the cases have been reported in European countries and North America, while a few have been described in Asian countries[11]. Only one report, out of 6 Brazilian cases[8], was found in the medical literature.

CASE REPORT

Female patient, aged 53, white, with menopause for two years seeks medical assistance for hair loss. She complains of hair loss in the frontal region and later, in the eyebrow. Her father presents androgenetic alopecia. She brought a previous biopsy report which showed the presence of pilary follicle, with fibrosis and lymphocytic infiltrate, circling the follicle infundibulum which suggested two hypotheses: alopecia areata or postmenopausal fibrosing alopecia (Figures 1 and 2). She had been treated with minoxidil 2% for six months and topic methylprednisolone (Advantan®), without response to the treatment.

Clinical exam showed frontal alopecia and also hair loss in the eyebrows (Figures 3, 4 and 5), with other normal pilary areas.

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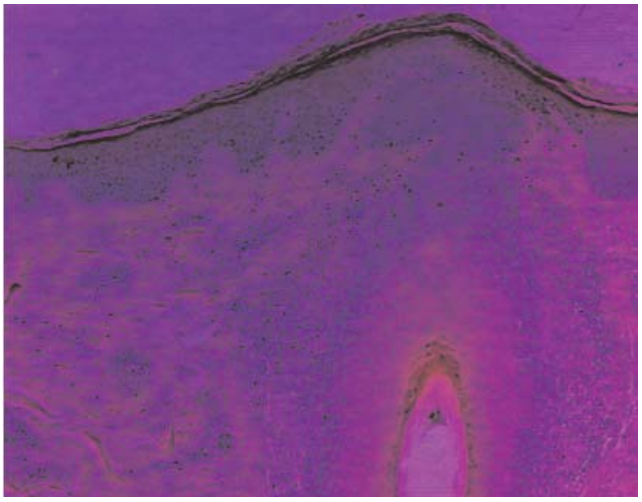

Figure 1: Histologic image stained by HE method , enlarged 40x, showing pilar follicle with fibrosis and lymphocytic infiltrate ,circling the pilary infundibulum

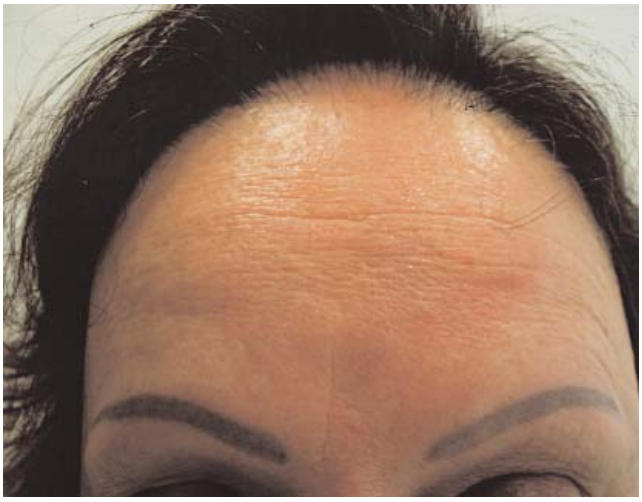

Figure 3: Total bilateral alopecia of the eyebrows

It was then prescribed minoxidil 5%, associated with auxin tricogena 12% in capillary solution and the patient was directed for evaluation for pilary follicles implantation.

DISCUSSION

Frontal fibrosing alopecia is a form of slowly progressive alopecia,varying from one to ten years, and cicatricial that presents symmetrical retreat from the frontal and temporal borders of the hair implantation [7,8,12,13]. There is only one case described in the medical literature of development on the posterior occipital line.[3]

Although less frequent, it has already been reported reduction of hair on the axillae, arms, legs, chest and pubic region. However inflammatory signs were not found in these areas.[6,8,10] Androgenetic alopecia, causing difuse reduction on the density of the hair, could be simultaneously observed, in some cases.[6]

The affected skin presents itself as pale, atrophic and without follicular holes. It is evident the contrast between the pigmentation of the area of alopecia, uniformly pale and the hyperpigmentation (by the solar damage of the skin immediately anterior). The bilateral hair loss of the eyebrows is a common finding and it can happen before or after hair loss. As for the hairline, it is possible to observe hyperkeratosis and erythematous-violaceous coloration, in some follicular holes, being this an early sign and better observed with magnifying glasses. These clinical conditions are considered typical of

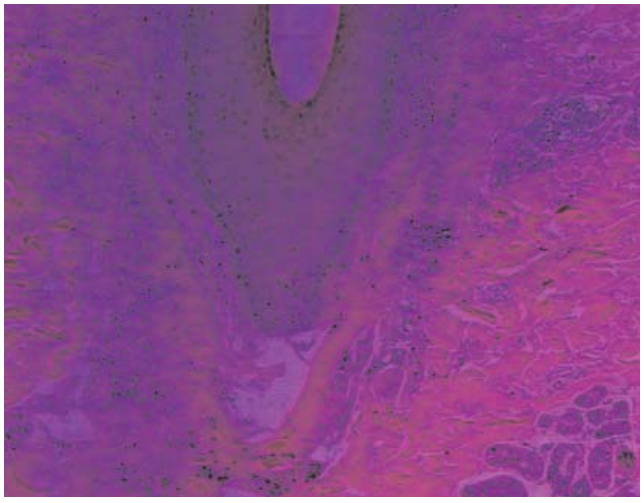

Figure 2: Histologic image stained by HE method , enlarged 100x, showing pilar follicle with fibrosis and lymphocytic infiltrate ,circling the pilary infundibulum

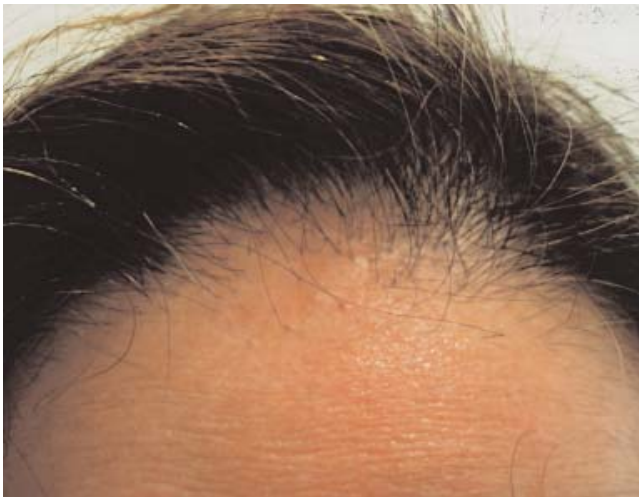

Figure 4: Frontal alopecia with atrophy of the skin and reduced number of pilar follicles

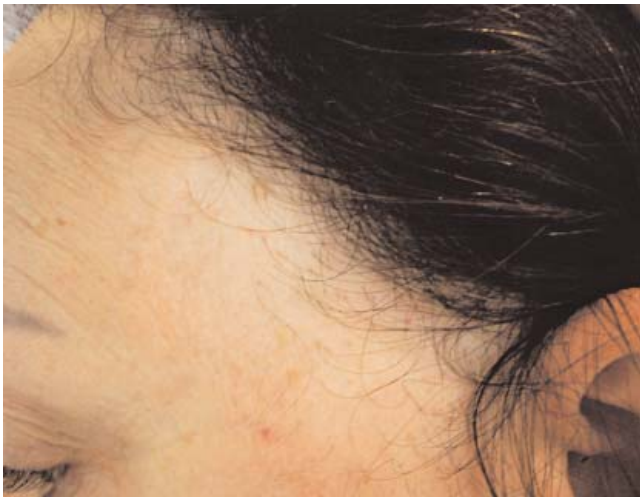

Figure 5: Hair rarefaction on the temporal region

frontal fibrosing alopecia [2,3,6,7,8,9,10].

Laboratorial investigation as mentioned in the literature such as CBC (Complete Blood Count), serum parameters of sexual hormones, TSH, antithyroid hormones, FAN, VDRL and anti-DNA, were always either negative or normal [2,6,7,8,9,12].

Histopathologic analysis frequently shows lymphocytic infiltrate, around the pilary follicles, with lichenoid reaction, noticeble reduction in the number of follicles and fibrosis.[7,10] It is believed that the destruction of the external root of the sheath at the isthhmus – where it is believed that the stem cells reside- is responsible for the the irreversible pattern of cicatricial alopecia.[3] The direct immunoflorescence was negative in all cases.[3,7]

The majority of the authors consider frontal fibrosing alopecia a localized clinical variant of lichen planopilaris , affecting predominantly the fronto-parietal capillary margin because the clinical and microscopic findings - cicatricial alopecia and periffolicular erythema would be indistinguishable.[3,6,9] Microscopic findings common to both include: inflammatory lynphocytic infiltrate (involving the isthmus) and the infundibulum of the pilary follicles, the presence of apoptotic cells (on the external of the root of the sheath)and a concentric fibrosis (around the follicles) that would result in its destruction, with subsequent cicatricial alopecia. The characteristic findings of frontal fibrosing alopecia would be more prominent apoptosis and smaller inflammation than in lichen planopilaris, side by side with spared interfolicular epidermis.[3] Immuno histochemical studies showed lynphocytic infiltrate, composed of lynphocites T CD4 and CD8 equally.[8] However, the age, most frequently postmenopausal, for the development of fibrosing alopecia and its characteristically frontal localization contrast with the classical multifocal coalescent areas of cicatricial alopecia in lichen planus. Besides that, lichen planus is associated with evidences of lichen lesions, in other sites, in more than fifty per cent of the patients and, as for frontal fibrosis alopecia, other skin lesions are usually absent.[7] It is not clear yet if this clinical entity represents a variant of lichen planopilaris with selective topography or another distinct type of lymphocytic cicatricial alopecia[3,12].

The diagnoses to be dismissed include: alopecia areata, cutaneous and systemic lupus erythematosus, traction alopecia, androgenetic alopecia, pseudopelade of Brocq and high central capillary family line [6,7,9]. Dermatoscopy is a diagnostic tool especially useful to distinguish frontal fibrosing alopecia from alopecia areata. [14]

The etiology remains a mystery.[2,3] Its premature begining in a patient , with precocious menopause, suggests that hormonal changes as a consequence of menopause , might play a role in its etiology. [10] However, there is no association with hormonal abnormalities and there is no response either to hormone replacement therapy.[2] The response observed in the treatment with finasteride suggests that the androgens might be partially responsible for the pathogenesis of the disease. [10,12] It is speculated that the pilary follicles of the fronto-temporal scalp in response to the changes occured during the menopause can be programmed differently to cell death or start to express neoantigens that invoke an autoimmune lymphocyte-T mediated response, leading to follicular destruction[2,12].

Little is known about the history of frontal fibrosing alopecia. The frontal retreat can progress up to half of the scalp or more. However, the progression of the lesion is variable.[10] It seems that the disease is self-limited but, in the majority of the cases, the level of progression before stabilization is unpredictable.[2,10,12]

As it is an irreversible disease, the objective of the treatment should be to avoid its progression. However, its treatment is still a chalenge. Most reports affirm that there is no effective therapy for such type of alopecia.[2,7,9,10] Treatments that have been tried include the use of : topic corticosteroids, intralesional and systemic ; topic retinoids; oral isotretinoin, topic minoxidil; hydroxychloroquine and finasteride.[2,10] Corticoids are considered the best therapeutics, especially in early inflammatory stages but , relapses of disease usually occur when there is interruption in the treatment[6]. The intralesional triancinolone acetonide provides a response rate of 40%, but it may worsen fibrosis and atrophy in advanced stages[12]. Oral finasteride (2,5mg/day), combined with minoxidil 2%,

stopped the progression of alopecia, in some pacients, after 12-18 months of treatment.[12] In any study it was possible to determine if the estabilization of the disease was a result of the therapy or part of the natural evolution of the disease. [10] A study published by the European Academy of Dermatology and Venereology [12] suggests that the combination of oral dutasteride with topic inhibitor of calcineurin may represent a safe and effective therapeutics to frontal fibrosing alopecia. In this study,[12] the use of oral dutasteride, 0,5 mg/day, for 6 months, associated with cream pimecrolimus 1%, twice a day, for 3 months, promoted a significant regrowth of the eyebrows and axillae and moderate improvement on the scalp. The only adverse effect reported was constipation. No recurrent case was observed 6 months after the end of the treatment.

Frontal fibrosing alopecia has become a more common diagnosis. The fact that it has appeared recently increases the possibility of environmental factors being involved with its etiology.[10] Most cases have been reported in European countries and North America while only a few cases have been described in Asian countries. [11] Only one report, out of 6 Brazilian cases[8], was found in the medical literature. This condition seems to be still little recognized or its diagnosis frequently confused with other types of alopecia in clinical practice all over the world. From a correct diagnosis, prognostic factors and better strategic therapeutics might be established. ❑

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