Postmenopausal frontal fibrosing alopecia

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SUMMARY

:

The authors present a 55-year-old female patient as a new case of fibrosing alopecia with typical localization and clinical form. The diagnosis was based on histological findings, circumscribed form of the disease, lack of the internal organ involvement and unsuccessful aggressive therapy. Recently a similar case was reported in a male patient.

KEY WORDS

Alopecia, postmenopausal, fibrosing

Postmenopausal frontal fibrosing alopecia (PFFA) is a new clinical entity that can be regarded as a version of lichen plano-pilaris with fronto-temporal, and more rarely, eyebrow localization. Kossard et al. (1) was the first to describe the first six cases in elderly Australian female patients. Later on, a clinical and immune-histological evaluation of another 10 cases (2), as well as further cases (3) have been published. This alopecia with characteristic localization and clinical picture was also observed by others. Till now over 20 cases have been reported in the literature (4,5). The present paper deals with our first typical case of such alopecia.

Case report

The 55-year-old female patient had uterus extirpation, at the age of 40 she also suffered from depression. Treatment included saliciylic acid (Aspirin-protect), propranololium chloratum (Huma-Pront), and seroxate paroxetin (Seroxate). A year before admission the patient observed a strip-shaped loss of hair on her temples and forehead. Due to loss of hair, the side shift of the hairline was about 2 cm. Despite local treatment with different hair-lotions and intrafocally applied steroid injections, the hair-loss was still in progress.

At admission: strip-shaped alopecia along the hairy skin of the forehead and temples, with small rare erythematous patches of follicular localization; whitish-scarred areas with closed follicles extended over most of the skin affected with alopecia. The border of scarred alopecia was not sharply demarcated, since short, stiff hair still grow in the scarred areas. Foci with signs of scarring penetrate into the healthy scalp (Fig. 1, 2). The process did not affect the lashes and the eyebrows; furthermore, we failed to find any sign of lichen planus on the unaffected scalp, on the the body, mucosa, on the vulva, or nails.

Routine laboratory tests failed to reveal any pathological changes; the serum iron level was normal. Hormon levels: TSH, FT4, FSH, LH, prolactin, serum testosterone, and SHBG levels were normal. No alter-

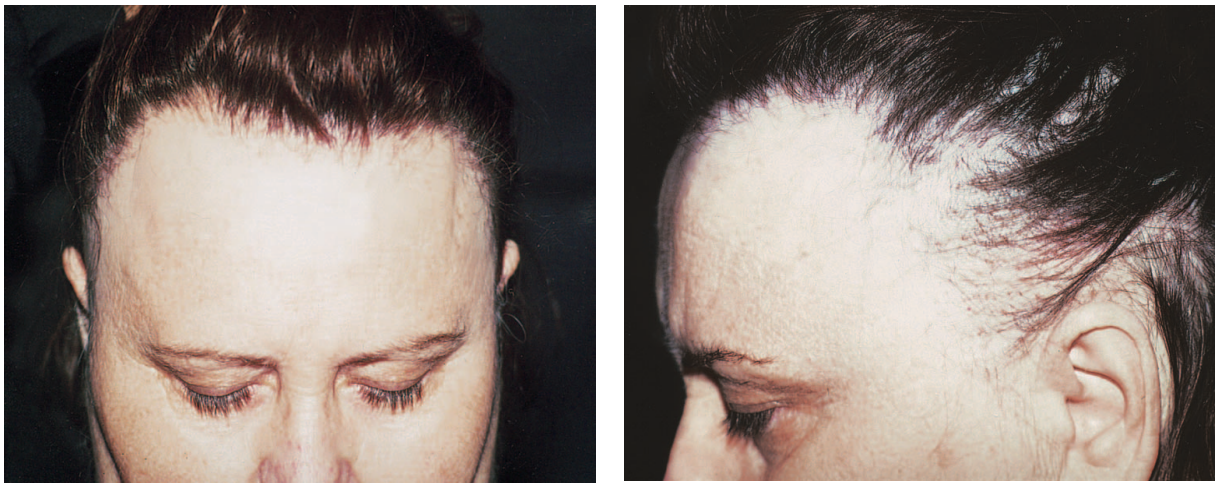


Figure 1-2. The fronto-temporal hairline is typically shifted "upward" with visible signs of scarring. A full, clearly visible temporal scarred line has not yet developed; many follicles still contain hairs.

ations suggesting focal pathology were found.

Histological findings: biopsy made from the skin with alopecia was assessed in two section plains. Both vertical and horizontal sections showed scarred follicles. In some samples scars in the horizontal plain showed onion leaves appearance. Scars have replaced almost all the follicles. Signs of lymphocyte infiltration were scarcely noted; there were no hydropic degeneration and no sebaceous glands (Fig. 3, 4). Conclusion: histological findings correspond to the late stage postmenopausal frontal fibrosing alope-

Figure 3. Scarred follicles with onion leave-shaped fibrosis at cross-section, HE 40X.

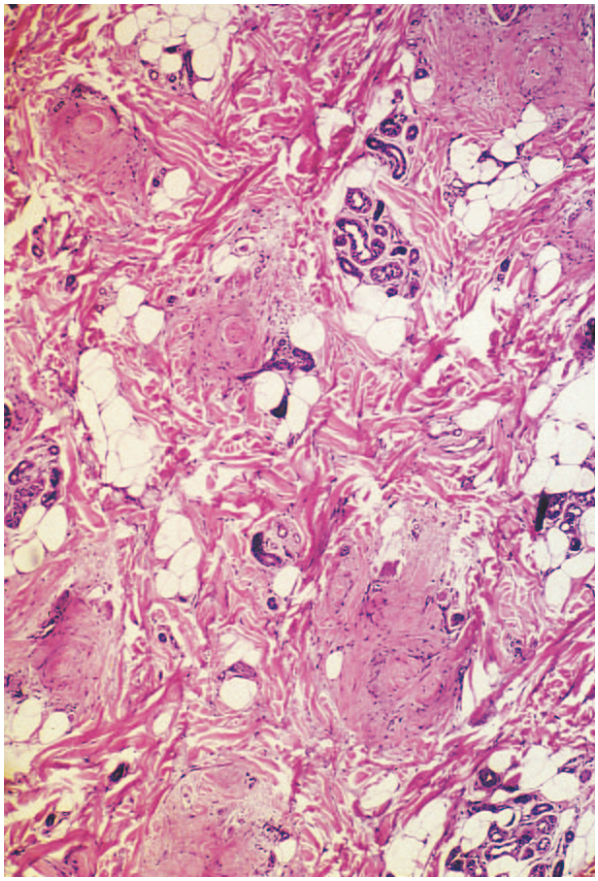
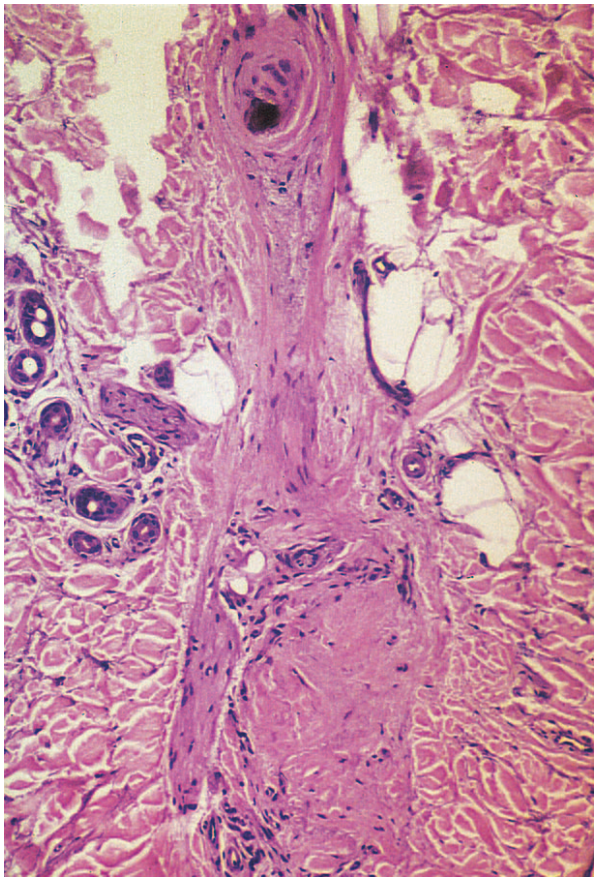


Figure 4. Longitudinal section reveals replacement of follicles by fibrotic bundles, HE 160X.



cia. No lichenoid alterations affecting follicular openings that are characteristic of the active stage of the process were found. The typical band-like lymphocytic infiltrate at the epidermal border as well as other tipical microscopic symptoms were missing.

On two occasions the patient received steroid infiltration treatment. Oral treatment included antiandrogenic hormone replacement therapy: estradiol valerate and cyproteron acetate (Climen 28) and as well as chloroquin (Delegil). Regain hair lotion was applied externally. After a six-month long observation period no signs of progression were found.

Discussion

Establishment of an accurate diagnosis and etiopathogenesis of frontal fibrosing alopecia is still a difficult clinical task (3,9,10). Within different forms of scarring alopecia, attention is mainly focused on lichen pilaris that includes lichenplanopilaris and Graham-Little's syndrome. Frontal fibrosing alopecia can be regarded a new member of this group of diseases.

Lichen planopilaris affects the hairy skin as scattered foci of scarring alopecia with erythema; as a rule, perifollicular desquamation is another concomitant sign of the disease. Diagnosis must be verified by histological findings. Extensive follicular keratotic papules resembling keratosis pilaris cover large surfaces of the trunk, and affect the hairy skin, armpits, pubic hair, and the eyelids of the patient. Later these alterations cause scarring alopecia that can affect the skin and the hairy skin, and is characterized by typical multifocal localization, rapid progress, and therapy resistance. Current literature defines this pathology as follicular keratosis, which is a special version of lichen plano-pilaris (8, 9,10).

Recently this group has been extended by including postmenopausal frontal fibrosing alopecia; in contrast to Kossard, this disease is not so rare. This dermatosis develops only in postmenopausal women at an average age of 67 years. It is interesting however, that this form of alopecia was reported in a male patient as well (6). If more male patients will be diagnosed in the future, the term "postmenopausal" should be abandoned. The disease may have a progressive course, it can develop within 1-10 years. The frontal pale strip of scarred skin can be several cm wide. Later on the closed and scarred of follicular openings can conceal the original perifollicular erythema. In patients originally described by Kossard, the skin, nails, and the mucosa were not involved (1,2). Other authors, however, reported buccal, vulvar, and other forms of cutaneous lichen (4,7). As stated in the original publication of Kossard, in its later stage the pathological process can also affect the brows (1,2).

Laboratory tests remain within the normal range, and no signs of focal changes or drug and hormonal interactions are observed. In some cases, antinuclear positivity was reported in the dilution range between 1:40 – 1:160 (2).

At the onset, histological findings indicate follicular fibrosis, which can penetrate deep into the reticular dermis, as well as lymphocyte infiltration. Vacuolar degeneration can be found in the basal layer. Lack of lymphocyte infiltration and perifollicular fibrosis are typical findings of the later stage of this pathological process. Since the histological picture is scarcely differentiated from that of lichen plano-pilaris, many authors, including Kossard, regard this form as a special fronto-temporal manifestation of lichen plano-pilaris. Immune-histological findings were negative, while immune-histochemistry revealed activated T-lymphocytes and a complete lack of B-lymphocytes (2).

Though the etiology of the disease is still unknown, it is assumed that the lichenoid tissue reaction might be the cause of the follicular destruction. Furthermore, factors that favor androgenic alopecia can also contribute to the development of this lichen-planopapillar form with specific localization (9).

Postmenopausal frontal fibrosing alopecia is an irreversible process with a slow course, although cases with spontaneous cessation of the process were reported. No effective therapy has yet been described. In cases with rapid progress, medium-dose steroid and chloroquin therapy resulted in a transient improvement of the symptoms (2). Other treatment modalities (including internal use of griseofulvin, isotretinoin, or application of local steroids, vitamin A acid, intralesional corticosteroids) failed to improve the course of the disease. Similarly, hormone replacement therapy was ineffective. Considering the assumed pathogenesis, these therapeutic attempts should not be fully abandoned; the otherwise necessary hormone replacement therapy with antiandrogen containing drugs (Climen 28) can be included in the treatment protocol.

The disease must be differentiated from other forms of fibrosing alopecia (DLE, folliculitis decalvans, keloid acne) and lichen pilaris. Of non-fibrosing alopecias differentiation from ophiasis that mainly affect the lateral and posterior areas of the hairy skin, is also of importance. Furthermore, the disease should be differentiated from traction alopecias that lead to progressive miniaturization of the follicles, and from the familiar high frontal hairline, which is more frequent in women. As a rule, androgenic alopecia in female patients does not shift the frontal hairline, but rather affects the vertex and the temples (8,9,10).

Analysis of our case and the data of the literature allow us to suggest that the frontal fibrosing alopecia is a special fronto-temporal version of lichen planus. Clinical diagnosis is verified by the presence of a typical whitish, scarred strip of alopecia, and by histological findings revealing fibrosing follicles. The disease has a progressive course and as a rule, is resistant to therapy. Selection of therapy requires caution since cases of spontaneous healing are reported, and meanwhile adequate hairstyling helps the patient to conceal minor areas of alopecia.

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