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Red face revisited: Disorders of hair growth and the pilosebaceous unit

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Abstract This paper reviews the recent literature on the diseases of the hair and pilosebaceous unit that may cause a red face. We discuss the epidemiology, clinicals, pathogenesis, and therapy of lichen planopilaris with its variants, discoid lupus erythematosus, folliculitis decalvans, dissecting folliculitis, acne keloidalis nuchae, pseudofolliculitis barbae, tinea capitis, tinea barbae, folliculitis of diverse causative factors and inflammatory follicular keratotic syndromes, ulerythema ophryogenes, atrophoderma vermiculatum, keratosis follicularis spinulosa decalvans, and folliculitis spinulosa decalvans.

Introduction

Several illnesses may turn the face red; among them, there are some disorders of hair and pilosebaceous follicle. We discuss the diseases related to the hair that may cause a red face, and we present an update on their epidemiology, clinicals, pathogenesis, and therapy. Most of them are causes of cicatricial alopecia of the scalp, but they can affect also the face, especially the area of the beard.

The conditions presented in this paper are lichen planopilaris with its variants, classic lichen planopilaris, Lassueur Graham-Little Piccardi syndrome, and frontal fibrosing alopecia, as well as fibrosing alopecia in a pattern distribution, which may be a fourth form of lichen planopilaris; discoid lupus erythematosus; folliculitis decalvans; dissecting folliculitis; acne keloidalis nuchae; pseudofolliculitis barbae; tinea capitis; tinea barbae; folliculitis of diverse causative factors and inflammatory follicular keratotic syndromes, including erythromelanosis follicularis faciei et colli; ulerythema ophryogenes; atrophoderma vermiculatum; keratosis follicularis spinulosa decalvans; and folliculitis spinulosa decalvans.

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Lichen planopilaris

Lichen planopilaris, a follicular form of lichen planus, is a rare inflammatory lymphocyte-mediated disorder that selectively involves hair follicles. Lichen planopilaris leads to follicular destruction and, consequently, cicatricial alopecia. Three forms of lichen planopilaris are described: classical lichen planopilaris, Lassueur Graham-Little Piccardi syndrome, and frontal fibrosing alopecia. Fibrosing alopecia in a pattern distribution could be added as a fourth variant.[1,2]

Classic lichen planopilaris

Epidemiology

Classic lichen planopilaris is largely seen in adults. Usually considered a rare disease, it is the most common cause of primary cicatricial alopecia.[1] It is more common in women, and its onset usually occurs between ages 40 and 60 years. Children are less frequently affected.[1,3,4] Association with nonscalp lichen planus is reported in 17% to 28% of cases at presentation and may occur during the course of disease in 50% of patients.[3–5]

Pathogenesis

The cause of lichen planopilaris remains unknown. Based on similar histologic and direct immunofluorescence findings,

it is suggested that lichen planopilaris results from the same pathologic process as lichen planus. An unknown antigenic trigger expressed by follicular keratinocytes appears to initiate the process. The inflammatory reaction occurs mainly around the bulge area and is mediated by T lymphocytes activated by Langerhans cells, which are increased in the dermis and epidermis. Certain drugs are known to trigger disease, but in most cases the cause is unknown.[1,3,5]

Clinical features

Lichen planopilaris frequently occurs on the vertex, but any region of the scalp can be involved. It may also be present at other sites such as the face, affecting eyebrows or beard, or more often, on arms, legs, axillary regions, and pubic regions.[1] At the scalp, there may be solitary or multiple areas of baldness. The alopecic patches stretch centrifugally, sometimes in an irregular manner, and can occasionally evolve to affect the entire scalp. Interestingly, unaffected hairs may be found within the lesion (Figures 1 and 2).[1,5] The most common symptoms and signs are increased hair shedding, pruritus, scale, and scalp tenderness.[4] Disease activity is usually seen in the hair-bearing margin and is perceived as perifollicular erythema and scaling associated with anagen hair release on execution of the pull test.[5]

Diagnostic examination

Trichoscopy allows better appreciation of the signs of disease activity, as well as the perception that, in lichen planopilaris, perifollicular scaling tends to form tubular structures, considered highly characteristic of the disease.[6] Resolution of associated symptoms and the absence of visible inflammation do not, however, automatically imply that hair loss has been arrested.[7] Trichoscopy of the alopecic area may also reveal white dots indicating fibrotic tracts and pinpoint white dots, which are unspecific and represent openings of eccrine sweat ducts better visualized in patients of higher skin phototypes.[6,8] Blue-gray dots in a target pattern may be present, corresponding to pigment incontinence secondary to perifollicular interface dermatitis.[9] The presence of a pigmented network in a honeycomb pattern seen either in dark-skinned patients or in sun-exposed areas of the scalp is an important feature to distinguish lichen planopilaris from discoid lupus erythematosus, because in the former, the inflammatory process is restricted to the hair follicle, thus preserving the interfollicular architecture.[10] Elongated, parallel-oriented blood vessels at the margin of some active lichen planopilaris lesions have also been described[6] (Figure 3).

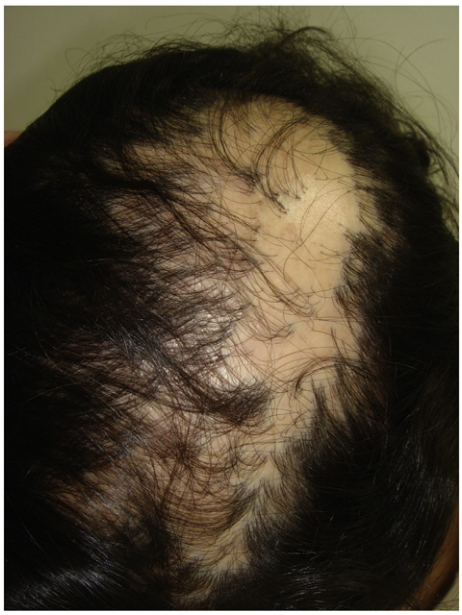

Fig. 1 Lichen planus pilaris: alopecic patch affecting the parietal region and vertex.

Pathology

Histologic features consistent with early, active lichen planopilaris are follicular lymphocytic interface dermatitis, scattered dyskeratotic and necrotic keratinocytes in the basal layer, infundibular hyperkeratosis and hypergranulosis, and atrophic or absent sebaceous glands. Pigmentary incontinence may be prominent. In late-stage lesions, inflammation can be minimal or absent, and fibrous tracts take the place of destroyed hair follicles.[5] Loss of elastic tissue staining in a wedge-shaped configuration involving the upper third of the fibrous tract and associated destruction of the elastic sheath that marginates the fibrous tracts is a useful feature in late-stage lesions.[11] Direct immunofluorescence can be helpful in ambiguous cases, although it is not uncommonly negative. "Shaggy" or "patchy" deposition of fibrinogen and clumped IgM or, less commonly, IgA and C3, is seen along the follicular basement membrane zone.[3,5]

Treatment

Therapy is often not satisfactory.[1,3] First-line treatment consists of intralesional or potent topical corticosteroids. As second-line therapy, oral corticosteroids, oral or topical ciclosporin, or oral tetracycline may be tried. Oral retinoids,

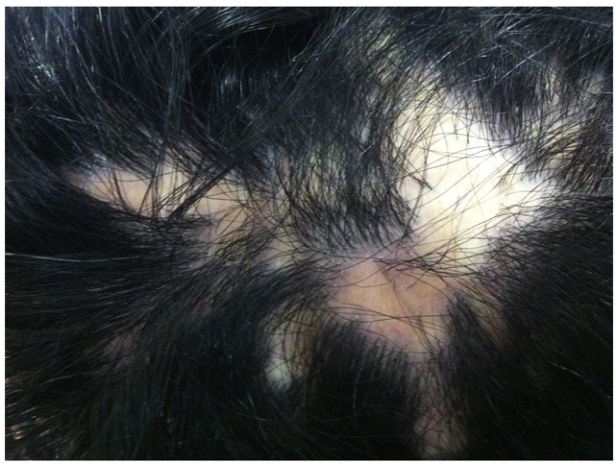

Fig. 2 Lichen planus pilaris: multiple alopecic patches at the vertex. Perifollicular erythema and scaling at the hair-bearing margin, indicating disease activity.