hair follicles can be noted as early as 4 days following radiation. Similar effects are noted with x-rays and electron beam radiation. The earliest change is thinning of the hair bulb, especially in the area of the matrix. Two or 3 weeks following irradiation, hairs with tapered shafts are apparent. The number of follicles showing radiation effects is roughly proportional to the dose of radiation. The definitive treatment is the scar excision or, better, a hair transplant restoration (Figure 10.18a and b).

CONCLUSION

Traction alopecia and trichotillomania are types of physical trauma that can lead to alopecia.

Traction alopecia is seen most commonly in black females. Trichotillomania is a traction alopecia related to a compulsive disorder. In the long term, permanent alopecia may occur.

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11 Management of acquired primary cicatricial alopecia

Salvador Villablanca and Juan Ferrando

INTRODUCTION

Human hair involves aspects of self-image, identity, ethnicity, and health, among other attributes. This is why it is no surprise that diseases that cause alopecia can cause altered self-perception and psychosocial interactions.[1]

Alopecia can be classified as cicatricial (or scarring) and noncicatricial. In turn, cicatricial alopecia (CA) is subdivided into primary cicatricial alopecia (PCA) and secondary cicatricial alopecia (SCA). The PCAs represent a rare and heterogeneous group of diseases, clinically characterized by the absence of follicular ostium and histologically by the replacement of hair follicle structures by fibrous tissue making the alopecia irreversible. From a physiopathological point of view, the scar is the end point of reparative fibrosis with permanent destruction of the preexisting tissue.[2–4] In PCA the hair follicle is the main target of the inflammatory process, as evidenced microscopically as a preferential destruction of follicular epithelium and/or adventitial dermis associated with relative preservation of interfollicular reticular dermis.[5,6]

In secondary CA, the destruction of the hair follicle is not the primary pathological event. It results from nonfollicular damage that eventually destroys the follicle. In these cases, the permanent follicular scarring develops when the involved pathological process is close to the follicular unit. Exogenous factors such as trauma (burns, radiation, and traction) and infiltrative and inflammatory endogenous processes (sarcoidosis, pemphigus vulgaris, and scleroderma) may result in secondary scarring alopecia[2,4,7] (Table 11.1).

Primary CA may become a true clinical challenge due to the limited knowledge of the natural history of the disease. Many do not have a known cause. Clinical findings often have limited useful data for the diagnosis, because of overlapping findings and lack of specific signs. This sometimes makes it difficult to distinguish between the different conditions. Also, the clinical and histological characteristics can change over time, finally resulting in most cases in hair replacement with scarring tissue.

EPIDEMIOLOGY

The epidemiology of PCA in the general population is unknown. Two retrospective studies in hair research institutions estimated prevalence between 3.2% and 7.3%.[2,3] In a recent survey performed in the United Kingdom, the estimated incidence of PCA was 6.96 per 1000 new general dermatology referrals per year, which is equivalent to about 9.6 new cases per clinician per year.[8] The ratio between PCA and SCA is estimated at 1:15, and the ratio between neutrophilic and lymphocytic PCA is 1:4.[3]

Between 30% and 40% of patients with PCA in a hair research institution are cataloged as pseudopelade (or

Table 11.1 Causes of Secondary Cicatricial Alopecia

Physical/ chemical agents	• Chemical burns • Insect bites • Mechanical trauma, traction, compression or laceration • Radiation dermatitis • Thermal burns
Dermal granulomatous infiltrations (infectious origin)	• Fungic infections • Protozoa • Tuberculosis • Syphilis • Viral infections
Dermal granulomatous infiltrations (noninfectious)	• Necrobiosis lipoidica • Sarcoidosis • Amyloidosis • Actinic granuloma
Sclerosing disorders	• Lichen sclerosus et atrophicus • Morphea • Scleroderma • Sclerotic porphyria cutanea tarda
Neoplastic infiltrations	• Basal cell carcinoma • Squamous cell carcinoma • Dermatofibrosarcoma protuberans • Lymphoma • Malignant melanoma • Metastasic carcinoma • Adnexal Tumor • etc.
Inherited and congenital disorders	• Aplasia cutis, eccrine hamartoma, incontinencia pigmenti, keratosis pilaris spinulosa decalvans, neurofibromatosis, chondrodysplasia punctata, polyostotic fibrous dysplasia, cutis verticis gyrata, Darier disease, epidermal nevi, epidermolisis bullosa, hair follicle hamartoma, hypotrichosis congenita, ichthyosis (sex-linked recessive), porokeratosis of Mibelli

nonspecific cicatricial alopecia).[2] This means that one-third of cases have no specific diagnosis, becoming a true diagnostic and therapeutic "desert" for both dermatologists and patients. The diagnosis of PCA is not a purely academic exercise, because early treatment of the inflammatory component may prevent the progression of primary scarring alopecia, and the secondary fibrosis that gives the alopecia its irreversibility. Within the PCA with predominantly lymphocytic infiltrate, the most frequent condition varies depending on the different series. First and second places in frequency are always disputed between lichen planopilaris (and its variants) and cutaneous discoid lupus erythematosus, followed by pseudopelade of Brocq (PB).[2,3,9] This difference may be influenced by a discrepancy in the clinicopathological diagnostic criteria between authors, especially in regard to the classical PB (an entity in constant discussion). Among the causes of PCA with an initially neutrophilic infiltrate, we should consider folliculitis decalvans (FD) as the most common form (10% of al PCA), unlike *perifolliculitis abscedens et suffodiens capitis* (less than 5% of PCA).[10]

Table 11.2 Proposed NAHRS Working Classification of Primary Cicatricial Alopecia

Lymphocytic	Chronic cutaneous lupus erythematosus
	Lichen planopilaris
	Classic lichen planus
	Frontal fibrosing alopecia
	Graham-Little syndrome
	Classic pseudopelade (Brocq)
	Central centrifugal cicatricial alopecia
	Alopecia mucinosa,
	Keratosis follicularis spinulosa decalvans
Neutrophilic	Folliculitis decalvans
	Dissecting cellulitis/folliculitis (*perifolliculitis capitis abscedens et suffodiens*)
Mixed	Folliculitis (acne) keloidalis
	Folliculitis (acne) necrotica
	Erosive pustular dermatosis
Nonspecifi	

Source: Adapted from Tan E, Martinka M, Ball N et al., *J Am Acad Dermatol*. 2004;50:25–32.

ETHIOLOGY

There is a paucity of data in the literature regarding the origin of PCA. In most of the literature, histopathology revealed the presence of inflammation affecting the upper portion of the hair, which would explain the irreversibility of the process, because at this location, stem cells are housed. This place called the protuberance or bulge is located in the infundibulum, where the hair erector muscle inserts. In some situations, the trigger of this inflammatory response is the result of an antigenic stimulation of Langerhans cells that are located in the pilosebaceous unit. Examples of a possible antigenic stimuli would be ultraviolet radiation in the case of lupus erythematosus, certain medications in the case of lichen planopilaris, and *Staphylococcus aureus* in the case of folliculitis decalvans. With the new knowledge in respect to its origin, it is known that there is a loss of immune protection of bulge stem cells,[5,11] a dysfunction in the ability of self-perpetuation of stem cells, increased autoimmune activity enhanced by pro-inflammatory cytokines, and predisposing genetic and environmental factors.[12–14] Recent data also suggest association with an altered lipid metabolism and development of the PCA, where a sebaceous gland dysfunction could play an important role in their pathogenesis. Independent of the initial event, the obliteration or permanent functional impairment of the critical elements for the reconstitution of the follicle results in permanent alopecia.[15–19]

CLASSIFICATION

Currently there are several classifications for PCA, but the most accepted is the one proposed by the North American Hair Research Society (NAHRS).[20] This classification divides the PCA into two groups according to the type of predominant inflammatory cell infiltrate (lymphocytic and neutrophilic), a concept that had been previously suggested by other authors, but was improved by this working group, adding two more subgroups: mixed and nonspecific (Table 11.2). Although there have been debates about whether this classification is satisfactory, it gives us a practical and reasonable view for basic and clinical research.

CLINICAL PATTERNS OF PRESENTATION

There is a big clinical and histopathological overlap between different entities of PCA. There are some forms of PCA whose existence per se is discussed, and among them is the pseudopelade of Brocq.

In daily practice we observe two major clinical patterns of presentations of scarring alopecia, the first correspond to patients with multiple irregular patches of scarring alopecia (Figure 11.1) on the scalp, and the second pattern correspond to patients with a central patch (Figure 11.2) surrounded by several smaller patches of scarring alopecia ("cicatricial satellitosis"). Both types of clinical presentations are final stages of cutaneous processes that previously had affected hair follicles (evidenced or not), finishing with these residual and nonspecific features that do not allow us to elucidate the etiopathogenic origin of it. We call these two classic patterns of scarring alopecia presentation a "footprints in the snow" pattern for the first and "big patch" pattern for the second. If we look at other forms of clinical presentation of scarring alopecia, there are other characteristic patterns that could be called specific patterns: marginal pattern (frontal fibrosing alopecia, and tractional alopecia),

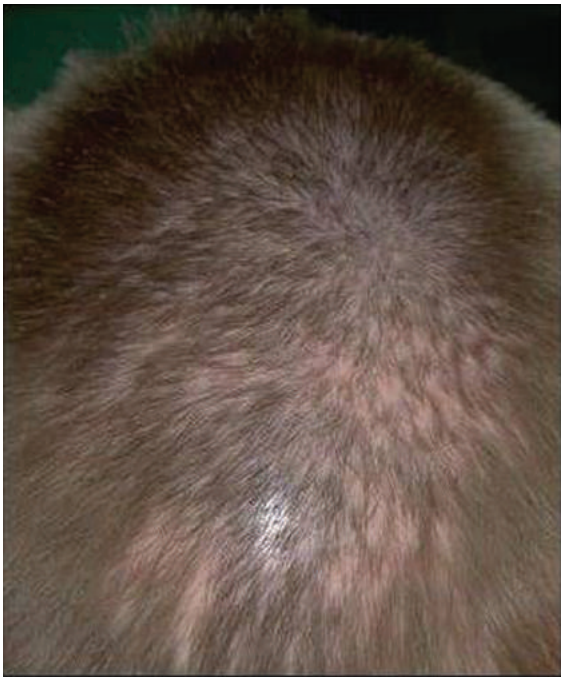

Figure 11.1 **Patches of scarring alopecia as multifocal lesions ("footprints in the snow").**

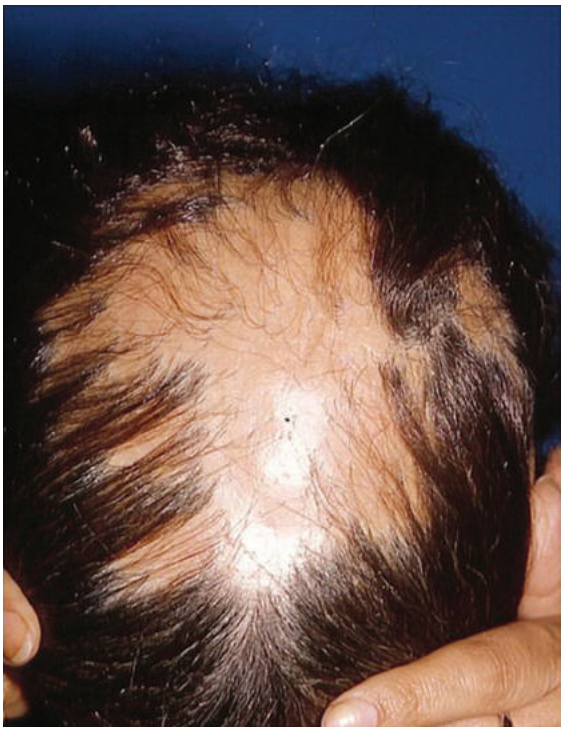

Figure 11.2 **Central patch surrounded by several smaller patches of scarring alopecia at the periphery ("cicatricial satellitosis").**

follicular pattern (lichen planopilaris, decalvant folliculitis, alopecia parvimaculata, acne necrotica, and tufted folliculitis), inflammatory-abscedens pattern (pustular erosive dermatosis of the scalp, perifolliculitis capitis abscedens et suffodiens), and the diffuse pattern (acute lichen planus pilaris, and the red scalp syndrome).

Primary cicatricial alopecia with an initially lymphocytic infiltrate

In early stages of primary scarring alopecia associated with lymphocytic infiltrates, the clinical features are fairly characteristic. The causes are listed in Table 11.2.

CUTANEOUS DISCOID LUPUS ERYTHEMATOSUS

Scalp involvement occurs in many cases of chronic cutaneous discoid lupus erythematosus (CDLE), but in just 10%–20% of cases as a single manifestation. It affects mainly women and usually begins between 20 and 50 years of age.[11] The lesions, which occur mainly on the sun-exposed areas of the face and ears and anywhere on the scalp, are clinically characterized by erythematous maculopapules that extend in a centrifugal way, with plugging of follicular ostia and formation of adherent scale that may be hyperkeratotic (Figure 11.3). In the evolved lesions, an atrophic component with telangiectasias is added and pigmentary changes can be seen. The early, active lesions may be sensitive and pruritic. The developed lesion exhibits a scaly crust with follicular spines attached to the under surface. The diagnosis is facilitated by the presence of lesions of CDLE in other locations, but if these lesions are not present, the differential diagnosis with follicular lichen planopilaris can be difficult, and also should include tinea capitis, psoriasis, and alopecia mucinosa. The lesion may resolve but tends

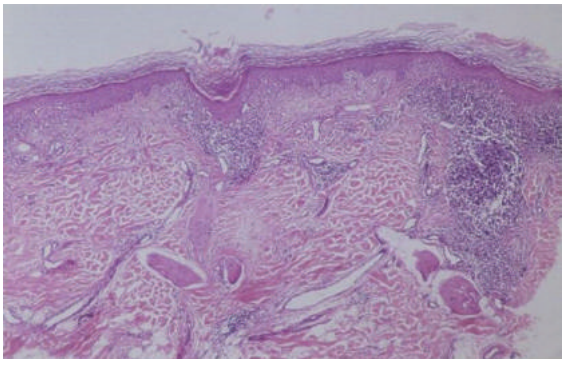

Figure 11.3 **Histology of chronic discoid lupus erythematosus with involvement of the scalp. Hematoxylin and eosin stain, ×40. Atrophic epidermis with interface dermatitis changes with the presence of necrotic keratinocytes and a perivascular and periadnexal inflammatory infiltrate with lymphocyte predominance.**

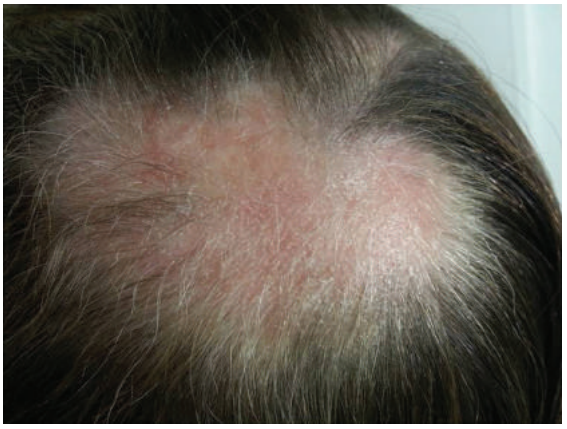

Figure 11.4 **Chronic discoid lupus erythematosus with involvement of the scalp. Erythematous patch with plugging of follicular ostia and formation of adherent scale that may be hyperkeratotic. Lesion activity is primarily in the center of the plate.**

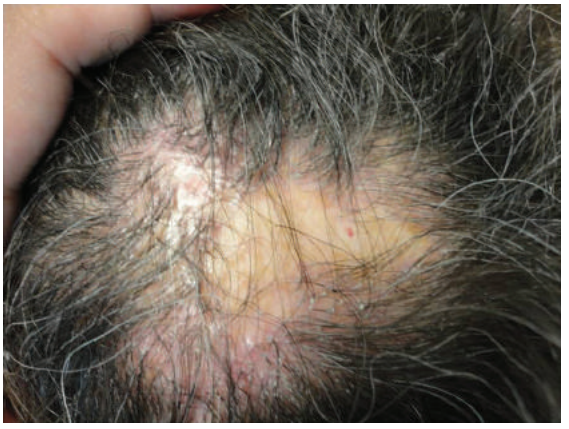

Figure 11.5 **Classical lichen planopilaris. Characterized by perifollicular erythema and hyperkeratotic follicular papules that are diffusely distributed, with isolated follicles respected in the center.**

to recur centrally, well within the previously affected atrophic patch rather than at its margin. From the histopathological point of view, cicatricial alopecia due to CDLE is characterized by perifollicular infiltrate with a lichenoid pattern.

Lupus erythematosus is characterized by the presence of vacuolar changes in the follicular epithelium and the interfollicular epidermis, with apoptotic keratinocytes, thickening of the basal membrane, the presence of dermal mucin, and periadnexal and perivascular infiltrate in the dermis (Figure 11.4). Dyskeratosis is minimal to moderate.[21–23]

LICHEN PLANOPILARIS

Lichen planopilaris is considered a variant of lichen planus and has three recognized variants: the classic follicular lichen planus, Piccardi–Lassueur–Graham-Little syndrome, and the frontal fibrosing alopecia with a marginal distribution pattern as a topographic variation.

Classical lichen planopilaris (LPP) usually appears in middle-aged women, in the form of pruritic (or painful) perifollicular erythema followed by acuminate, spinous, hyperkeratotic follicular papules that are diffusely distributed, but predominantly in the center of the scalp, which resolve leaving erythematous, atrophic polygonal-shaped patches of cicatricial alopecia areas (with intact follicles inside) with little tendency to coalesce (Figures 11.5 and 11.6).[10,21,24] The disease tends to be multifocal, with slow and progressive extension (usually with active lesions in the periphery) marked by exacerbations, which can determine an intense hair fall. Typical lesions of lichen planus are rarely observed in other skin sites or mucosa (bipolar lichen planus).[25] When the disease begins, erosions are rare, and the association with CDLE had been described. The differential diagnosis includes CDLE, alopecia areata, pseudopelade of Brocq, alopecia mucinosa, and folliculitis decalvans (although in the latter it is common to observe micropustules).

From a histopathological point of view, LPP is characterized by lichenoid infiltrate focused at the isthmus and the follicular infundibulum, with infundibular hyperkeratosis and underlying hypergranulosis, often sebaceous glands are atrophic or absent, interfollicular epidermis can be involved, and incontinentia

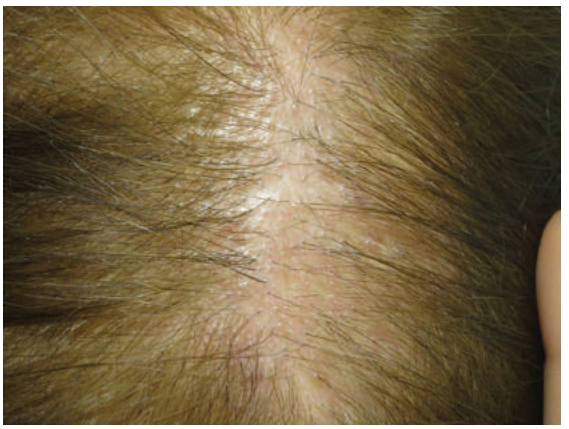

Figure 11.6 **Evolved classical lichen planopilaris. Erythematous, atrophic patches of cicatricial alopecia areas (with intact follicles inside of it) with little tendency to coalesce.**

pigmenti should be intense. Dyskeratosis is moderate to prominent. Exocytosis of cells into the epidermis is common. Perieccrine infiltrates are absent, and perivascular infiltrate may be minimal, superficial, and perifollicular. Epidermal and dermal mucin is usually absent (Figure 11.7). In evolved lesions, a destruction of the follicle with foreign body granulomas is observed, and eventually longitudinal and concentric lamellar fibrosis paths corresponding to existing follicles with minimal inflammatory infiltrate are present. In residual lesions, it is often impossible to make a specific diagnosis. In LPP diffuse dermal fibrosis is not observed, the latter being a characteristic change in CDLE.[21–23]

Piccardi–Lassueur–Graham-Little syndrome is characterized by progressive frontal scarring alopecia of the scalp associated with loss of axillary and pubic hair and rapid development of follicular keratoses on the trunk and occasionally in the lower extremities, histopathologically consistent with follicular lichen planus. It is possible that this condition is closer to a form of scarring follicular keratosis than to lichen planus or lichen planopilaris.

Frontal fibrosing alopecia (FFA), originally described by Kossard in 1994,[26] mainly affects postmenopausal women over 50 years, and is characterized by atrophic recession of the frontotemporal hairline, with remaining isolated follicles, giving it a distinctive look. The frontotemporal recession exposes pearly white looking skin that has been previously protected from the dermatoheliosis. In the recession or progression line of alopecia, perifollicular erythema and infundibular hyperkeratosis can be observed, being indistinguishable from those of classic LPP. Most patients also present with scarring alopecia in parietal areas, preauricular areas, and eyebrows, which may be accompanied by erythema (Figure 11.8). The association with androgenetic alopecia is common in postmenopausal women, and occasionally concomitant involvement of lichen planus has been described at other sites, which, together with histopathological findings,

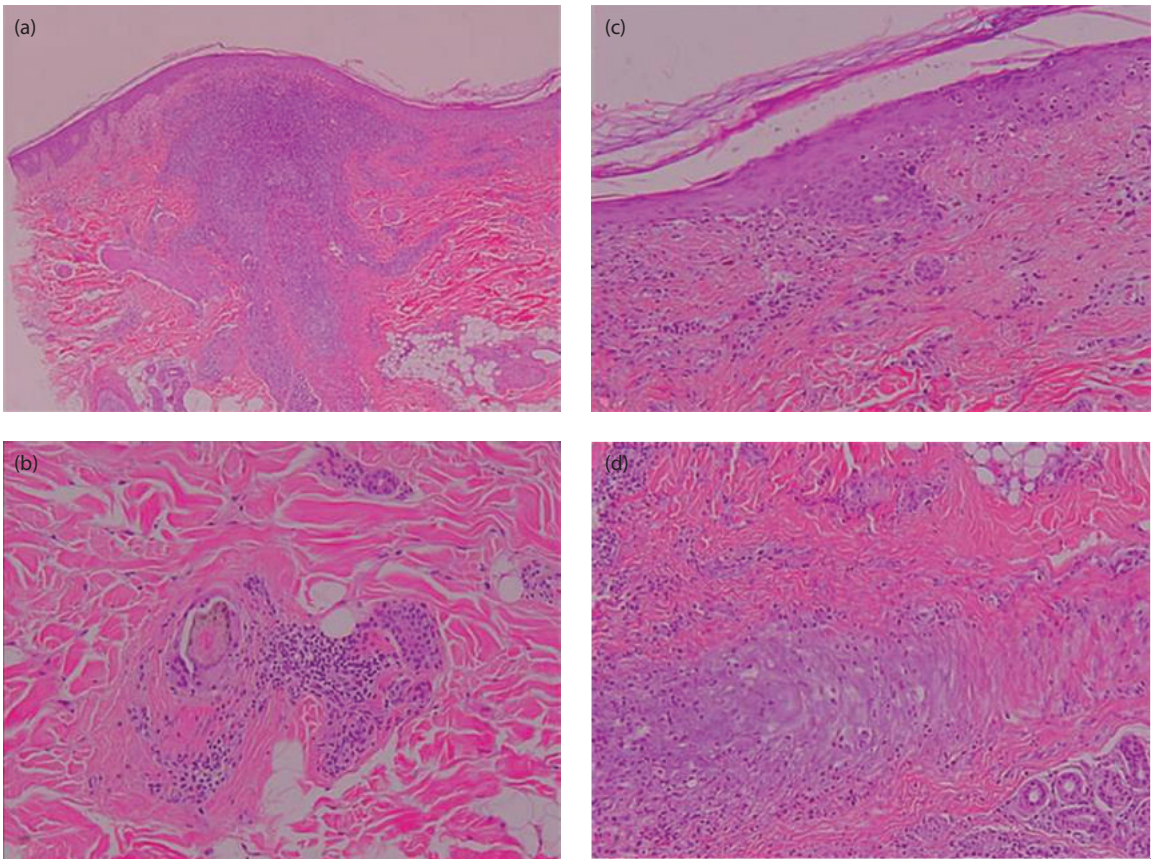

Figure 11.7 **Histology of lichen planopilaris (hematoxilin and eosin stain). (a) (×40) Perifollicular lymphocytic infiltrate primarily affecting the follicular isthmus and infundibulum. (b) (×100) Vacuolar changes in the epidermis next to the follicle. (c) (×100) Lymphocytic infiltrate surrounding a follicle with incipient scarring. (d) (×100) Follicle with necrotic keratinocytes.**

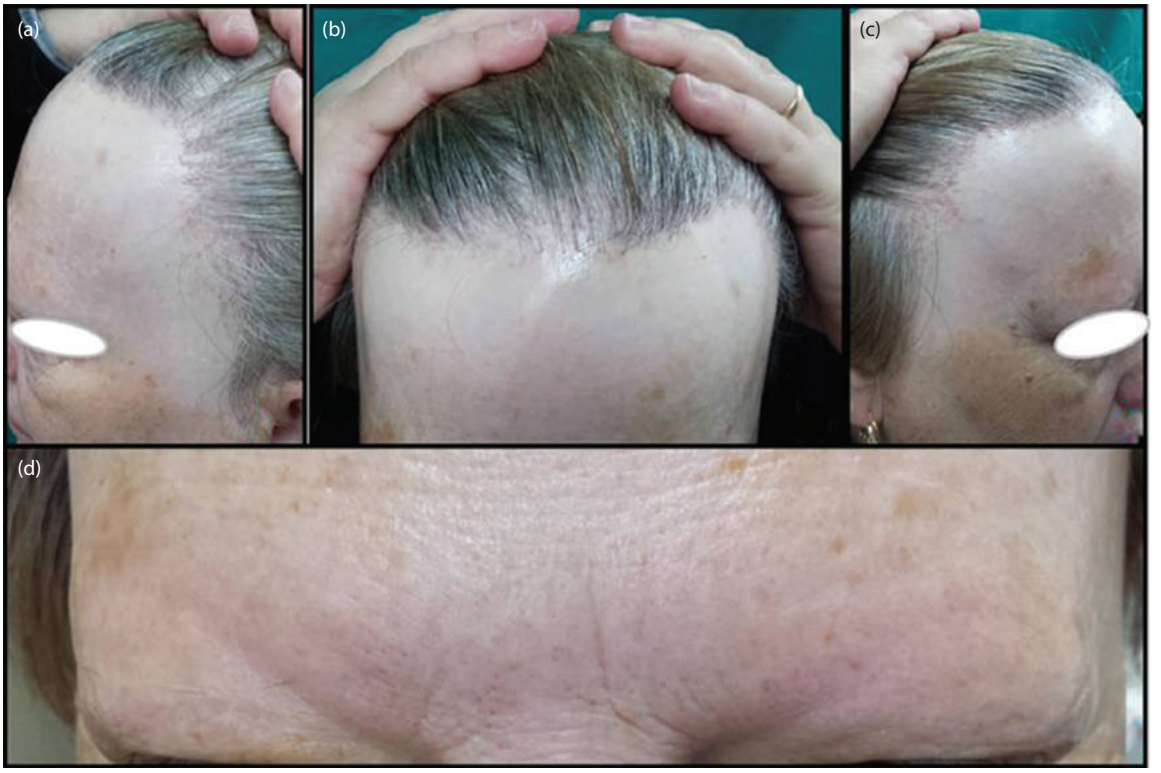

Figure 11.8 **Frontal fibrosing alopecia. Characteristic marginal pattern affecting the temporal (a and c) and frontal region (b). It is typically the partial or total involvement of the eyebrows (d).**

justifies the nosological grouping of these entities. Small papules of the face have also been described accompanying FFA, with the characteristic changes of LPP on the facial involved vellus.[27]

Fibrosing alopecia with a female androgenetic alopecia pattern distribution has been described in women about 60 years of age, proving the presence of a scarring alopecia with perifollicular erythema, loss of ostia and follicular keratosis, sometimes painful, located in the central portion of the scalp. Histologically, a lymphomononuclear infiltrate is observed in the isthmic portion of some follicles, with perifollicular lamellar fibrosis, and in a variable percentage of biopsied cases, interface dermatitis and basal vacuolar change with apoptosis of keratinocytes (Figure 11.9). Although the differential diagnosis must be made with the pseudopelade and follicular degeneration syndrome (central centrifugal scarring alopecia), this is probably either a topographic variant FFA or a hybrid form of alopecia, combining female androgenetic alopecia with a LPP.[3,10,28,29]

PSEUDOPELADE OF BROCQ

Although the concept of "pseudopelade" is used to refer to the final stage of various scarring alopecias, to avoid confusion, it has been proposed that the term be abandoned.[22,30,31] The clinical definition of the disease according to Ross et al.[10] is of a chronic form of scarring alopecia in white women that is typically presented with outbreaks of multifocal lesions (lenticular, "moth-eaten," or "footprints in the snow" pattern) predominantly of central distribution (vertex). Characteristically it presents with small size, geometrical configuration, minimal perifollicular erythema, and ivorian hypopigmentation, occasionally showing a slight depression (Figure 11.10). From the histopathological view, the Braun-Falco et al.[32] series highlights the presence of a normal epidermis with fibrous tracts in the subcutaneous tissue, the absence or decrease in number of sebaceous glands, and the absence of marked inflammation, significant follicular plugging, or widespread scarring. Although it has been described as an atrophy of the epidermis, it does not correspond to an interface dermatitis with vacuolar change. It should be considered then as a histopathological exclusion diagnosis in the context of characteristic signs. The final evolution of pseudopelade of Brocq gives clinical and histopathological features indistinguishable from other "burned" primary scarring alopecia. In summary, with this term, both forms of scarring alopecia are recognized, the small patches and the conventionally large central patch, as both are considered a final cicatricial stage of a previous inflammatory follicular process not detected at its early stage.[33,34]

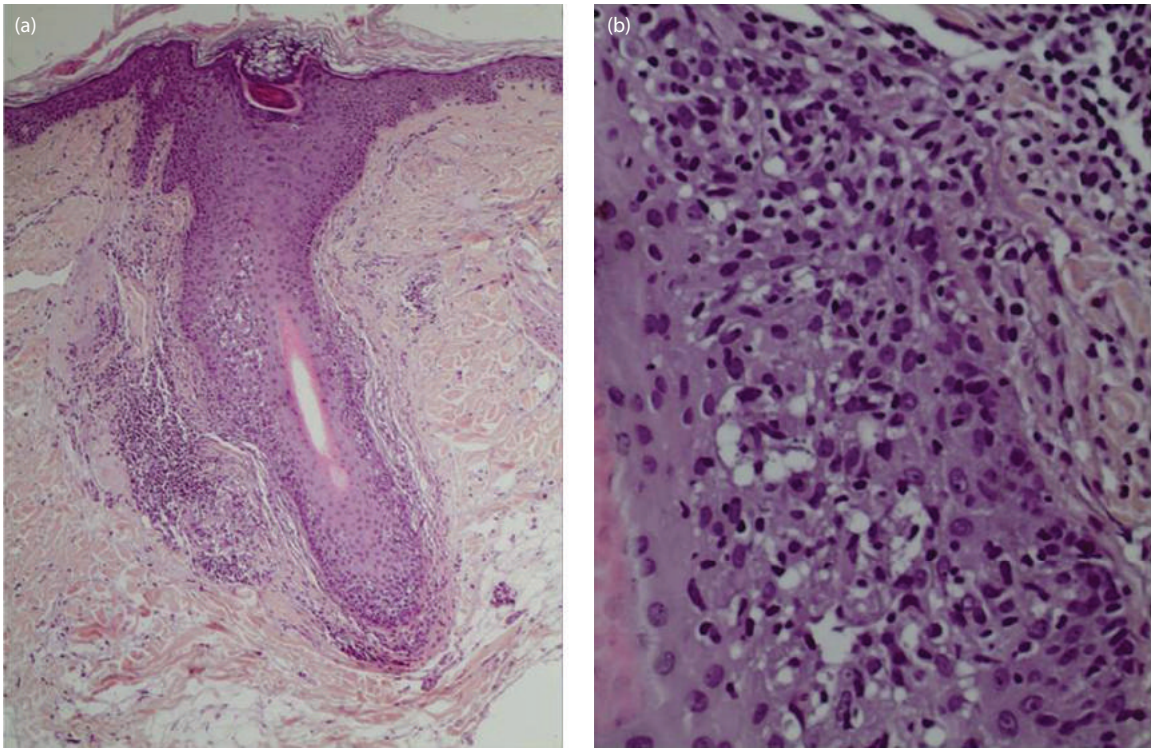

Figure 11.9 **Histology of frontal fibrosing alopecia (hematoxylin and eosin stain). (a) (×40) lymphocytic infiltrate surrounding the hair follicle. (b) (×100) Hair follicle with vacuolization of basal layer and lymphocytic infiltrate.**

CENTRAL CENTRIFUGAL CICATRICIAL ALOPECIA

The follicular degeneration syndrome was initially described in African American women who used hot combs to straighten their hair, and was then regarded as a form of traumatic alopecia. The clinical term of *central centrifugal cicatricial alopecia* (CCCA) has been set to include cases that occur in white individuals with a similar pattern of alopecia characterized by a central alopecia patch with centrifugal extension. CCCA commonly affects middle-age black women, starting as a central cicatricial alopecia over the vertex, which gradually spreads outward in a centrifugal direction (Figure 11.11).[10,35] Histopathologically it is considered characteristic of this entity of perifollicular chronic inflammation, concentric lamellar fibroplasias with eccentric epithelial atrophy (more marked in isthmus and infundibulum). Premature desquamation of the internal follicular sheath (although this is not a consistent or specific finding of this entity) and eventual destruction of the follicular epithelium, with acute and chronic perifollicular inflammation are noted, but sparse and peri-eccrine infiltrates are absent. Central centrifugal cicatricial alopecia is characterized by preservation of the elastic sheath surrounding the fibrous tracts. Dyskeratosis is absent, and pigment incontinence is minimal. Epidermal and dermal mucins are absent. The differential diagnosis includes androgenetic alopecia and trichotillomania.[35]

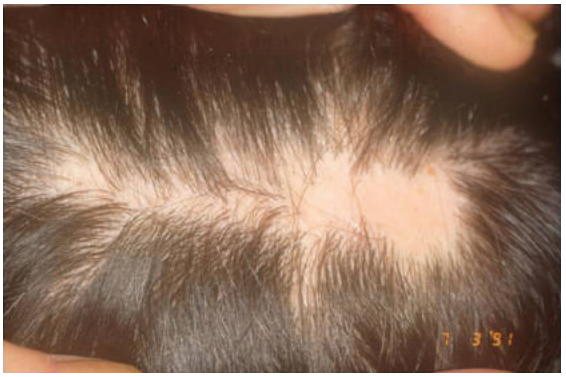

Figure 11.10 **Classic pseudopelade of Brocq. Small size patches of geometrical configuration, with minimal perifollicular erythema and ivorian hypopigmentation, occasionally showing a slight depression.**

ALOPECIA MUCINOSA

Alopecia mucinosa is usually benign and self-limiting in children and young adults, although it occasionally

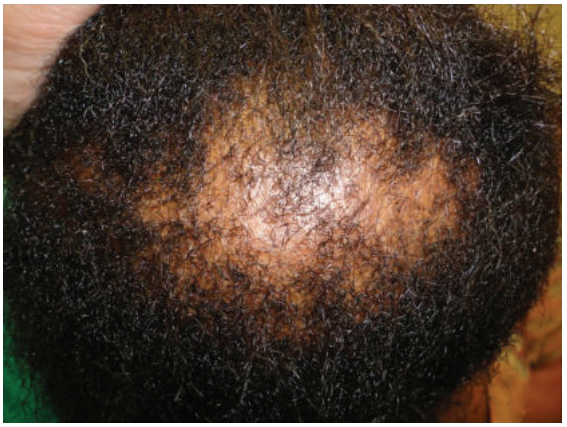

Figure 11.11 **Central centrifugal cicatricial alopecia characterized by a central cicatricial patch of alopecia over the vertex, which gradually spreads outwards in a centrifugal direction.**

results in permanent patchy alopecia. It may be associated with lymphoproliferative disorders in older patients with multiple lesions. It is characterized by scaly erythematous infiltrate plaques formed by the confluence of follicular papules, with alopecia and dilation of the ostium follicular, localized predominantly on the face, shoulders, neck, and scalp. Histopathologically, the entity is defined by the presence of follicular mucinosis, accompanying a lymphomononuclear infiltrate that in many cases (30% of cases in adults) has a clonal origin, and corresponds to a cutaneous T cell lymphoma, of relatively indolent course in many cases.[10,36]

Keratosis follicularis spinulosa decalvans

Keratosis follicularis spinulosa decalvans (KFSD) was described by Siemens in 1926. The disease is grouped with the follicular atrophic facial keratosis and the atrophoderma vermicularis under the name of *keratosis pilaris decalvans*. However, some authors postulate that they might be different stages of the same disease.

The sex-linked inheritance of KFSD has been proposed by several authors. The most severe manifestations have been observed in boys, thus confirming a sex-linked inheritance. In genetic mapping studies, the KFSD is located on chromosome Xp21.2-p22.2. Female carriers often show signs of the disease but are affected in a milder form. This type of inheritance can be explained by the inactivation escapade mechanism of X-linked genes. Some authors believe that KFSD is transmitted in autosomal dominant form with a variable expression. However, some cases may occur sporadically.[37]

They have described three stages in the development of KFSD. The onset occurs in early childhood with photophobia and follicular keratosis, then comes an active phase of progressive scarring alopecia that mainly affects the parietal and occipital region, as well as eyebrows, especially in its outer third, and eyelashes, this being a distinguishing characteristic of the disease. After puberty it may also affect axillary and pubic regions. The hair shows a dry, rough, short, and fragile aspect. This process continues until early adolescence when the progressive scarring alopecia stops and photophobia improves. During adolescence, the disease is frequently associated with hyperkeratosis of palms and soles. Other clinical features that occur in this syndrome include atopia, photophobia, and corneal abnormalities.[38]

The histological findings include intrafollicular abscesses, perifollicular and perivascular infiltrate that may contain numerous plasma cells, follicular horny plugs, and hypoplasia, or absence of sebaceous glands. The latter is a fundamental pathological substrate of the disease. The scanning electron microscope reveals a fragile constitution hair with intense severe cuticular abnormalities.

The disease has been associated with mental retardation in a few cases, sensorineural hearing loss, delayed growth, repeated bacterial infections, hyperaminoaciduria, nail changes, microcephaly, arachnodactyly, and seizures.[39]

The differential diagnosis should take into account monilethrix, trichothiodystrophy, Netherton syndrome, and ectodermal dysplasia. Although it is a rare genodermatosis, KFSD should always be considered in all cases of hyperkeratosis with congenital alopecia. The treatment of such cases should be started as early as possible (especially in the inflammatory phase) in order to delay and minimize the cicatricial phase.

Primary cicatricial alopecia with an initially neutrophilic infiltrate

The pathogenesis of the PCA with an initially neutrophilic infiltrate is unknown. It is hypothesized that *Staphylococcus aureus* may be involved because it is often isolated in pustules, but it is not clear if it is a primary or a secondary process. Other suggested causes include a possible role of bacterial superantigens or a defect in cellular immunity. Currently, the theory is that both folliculitis decalvans and dissecting folliculitis (an even acne keloidalis nuchae) may represent a different spectrum of the same disease, and depending on the difference in follicular anatomy and host immune response, one of the clinical manifestations will predominate.[40,41]

FOLLICULITIS DECALVANS

Folliculitis decalvans or Quinquaud folliculitis is a destructive and suppurative folliculitis that affects young adults of both sexes. It presents with outbreaks of multifocal, erythematous, and hyperkeratotic follicular pustules that surround multiple, slowly expanding round or oval areas of alopecia on the scalp (Figure 11.12). Sometimes it affects several neighboring follicles, which

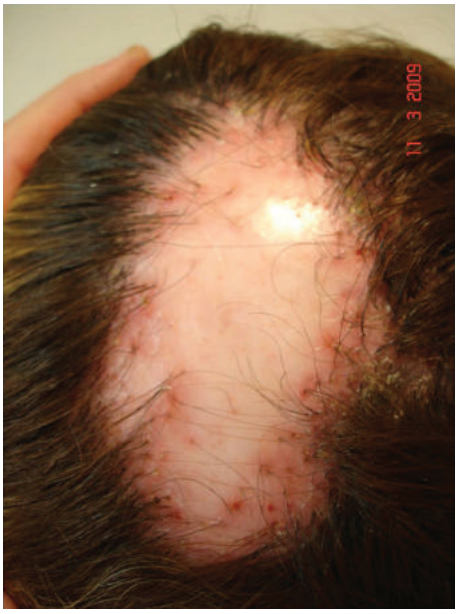

Figure 11.12 **Folliculitis decalvans. A slowly expanding oval area of alopecia on the scalp with multiple hyperkeratotic follicular pustules and crust at the periphery.**

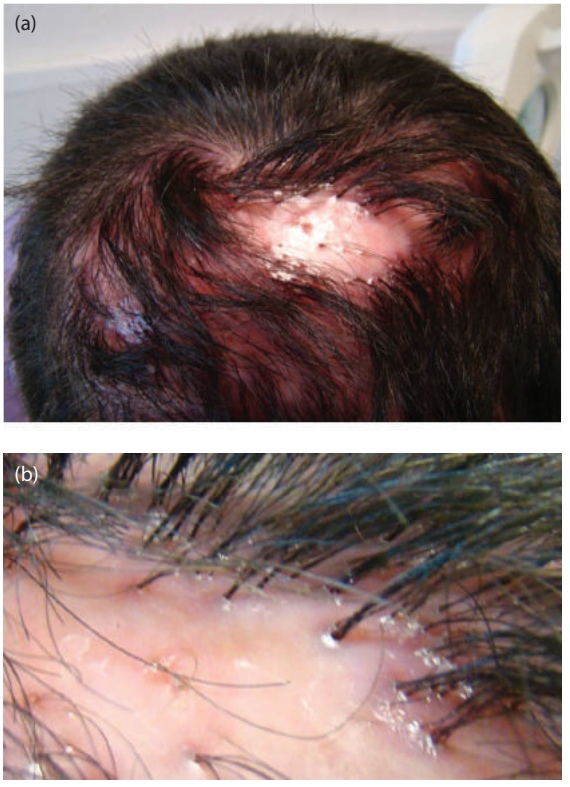

Figure 11.13 **Tufted folliculitis is a peculiar form of folliculitis decalvans where several neighboring follicles are joined in a common follicular ostium with several hairs, forming scarring alopecia patches (a) with the hairs emerging in a "bunched" way (b).**

are joined in a common follicular ostium with several hairs, forming scarring alopecia plates with these hairs emerging in a "bunched" way, which is known as tufted folliculitis (Figure 11.13). The etiological role of *Staphylococcus aureus* is under discussion, which usually appears in bacteriological cultures. Differential diagnosis must be made primarily with other bacterial folliculitis, but also fungal or viral folliculitis, CCCA, and acne necrotica. It tends to affect the vertex area and may also affect other hairy areas beyond the scalp such as the beard, pubic and axillary areas, and the inner thighs. It runs a very long course, and it is sometimes complicated by the fact that even after pustules disappear, the progressive cicatricial alopecia can continue. Histopathological examination shows intra- and perifollicular abscesses affecting the upper and middle portions of the follicle with an initial neutrophilic infiltration, with eventual follicular destruction, foreign body granuloma formation, and perifollicular dermal fibrosis (Figure 11.14). Sinus tract formation is absent, but eventually tufts of hairs may emerge through the same enlarged follicular infundibulum.[42,43]

DISSECTING FOLLICULITIS (PERIFOLLICULITIS CAPITIS ABSCEDENS ET SUFFODIENS)

Also called Hoffman disease, this is an element of the acne tetrad syndrome, along with acne conglobata, hidradenitis suppurativa, and cysta pilaris. A common etiology based on follicular obstruction with follicular rupture and secondary infection has been proposed. This most often affects young black men, and is located essentially in the vertex and occiput. Initial lesions consist on confluent pustules that form fluctuating or firm nodules, with painful suppurative follicular orifice and abscess formation, which eventually lead to scarring alopecia with sinus tracts and keloids production (Figure 11.15). Although spontaneous regression can occur, the occurrence of chronic relapses with active elements at the edges is characteristic.[44,45] The diffe ential diagnosis is limited and must be made with Celso kerion.

Histopathologically, infundibular distention is observed with formation of intra- and perifollicular abscesses. Follicular perforation determines the production of a mixed infiltrate, composed by plasma cells and foreign body granulomas in the perifollicular dermis and superficial fat. Abscesses are lined partially by squamous epithelium that mark off the sinus tracts, that eventually lead to extensive fibrosis in the dermis and subcutaneous tissue, with possible occurrence of keloids.[10,46]