Cicatricial Alopecia

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Antonella Tosti, Eric L. Maranda,
and Bianca Maria Piraccini

Key Points

- Cicatricial alopecia is a permanent condition that cannot be reversed by treatment.
- Early diagnosis of primary cicatricial alopecia is imperative and it should be treated aggressively to reduce permanent scarring.
- Dermoscopy may assist in selecting the optimal site of biopsy for pathological diagnosis.
- Improper lipid metabolism in lichen planopilaris leads to a toxic build-up of lipids in the sebaceous gland that may be initiating the inflammatory response.
- Peroxisome proliferator-activated receptor-γ, a master transcription factor that regulates the expression of genes involved in lipid metabolism, is aberrant in lichen planopilaris.
- Pioglitazone, a peroxisome proliferator-activated receptor-γ agonist, may be a novel means to effectively treat lichen planopilaris.
- In discoid lupus erythematous, autoreactive Granzyme B-positive CD8+ T cells are recruited to the hair follicles resulting in epidermal, dermal, and follicular destruction.
- Antimalarial drugs are the treatment of choice for discoid lupus erythematous.
- Retinoids may be used as an alternative treatment for patients with discoid lupus erythematous that do not respond to topical glucocorticoids, sunscreens and tacrolimus.
- Studies using field emission scanning electron microscopy and confocal laser scanning microscopy suggest that bacterial biofilm organization may play a role in the pathogenesis of folliculitis decalvans.
- Keratosis follicularis spinulosa decalvans is a rare, inherited or sporadically acquired X-linked disorder with a

A. Tosti (✉) • E.L. Maranda
Department of Dermatology and Cutaneous Surgery, Miller School of Medicine, University of Miami, Miami, FL, USA
e-mail: antonella.tosti@unibo.it

B.M. Piraccini
Division of Dermatology, Department of Specialized, Experimental and Diagnostic Medicine, University of Bologna, Bologna, Italy
e-mail: biancamaria.piraccini@unibo.it

A.D. Katsambas et al. (eds.), *European Handbook of Dermatological Treatments*,
DOI 10.1007/978-3-662-45139-7_15, © Springer-Verlag Berlin Heidelberg 2015

mutation in the MBTPS2 gene mapped to Xp21,13-p22.2.
- Global gene expression profiles in lichen planopilaris and pseudopelade of Brocq are distinct from one another and should be considered as separate conditions.
- Dysregulation of connective tissue metabolism in localized scleroderma leads to an excess deposition of collagen.
- Early stages of localized scleroderma are characterized by an elevation of Th1/Th17-associated cytokines and Th2-associated cytokines during the late stages of the disease.
- The downregulation of microRNAs, in particular miRNA let-7a, may perpetuate the fibrosis seen in localized scleroderma.
- Localized scleroderma usually resolves spontaneously in a few years.

Definition and Epidemiology

A large number of scalp disorders may destroy the hair follicles and result in cicatricial alopecia (Table 15.1). These include diseases that primarily affect the hair follicles as well as diseases that affect the dermis and secondarily cause follicular destruction. Once established, cicatricial alopecia is a permanent condition that cannot be reversed by treatment. The differential diagnosis between the diseases that cause cicatricial alopecia requires a pathological examination. The site of biopsy is crucial for pathological diagnosis and can be better selected using dermoscopy. Current treatment options of primary cicatricial alopecia are limited as the precise mechanisms that trigger the diseases are still unknown should be treated aggressively. Early diagnosis and treatment is necessary in order to avoid diffuse follicular destruction. When treating cicatricial alopecia, one must explain to the patient that hair that has been lost will not regrow. Surgical treatment of cicatricial alopecia includes excision of the scarring area after tissue expansion or hair transplantation. The latter technique is complicated by the poor recipient conditions due to the reduced blood perfusion present in scar tissue that may impair graft survival. There is also the possibility that grafting may precipitate relapses through a Koebner phenomenon. In many cases, the treatment selected is based on anecdotal and individual preferences, due to the limited number of double-blind, randomized studies completed and the small number of cases. This chapter discusses optimal management and treatment of some inflammatory scalp disorders that commonly cause primary cicatricial alopecia.

Table 15.1 Causes of cicatricial alopecias

Primary cicatricial alopecia
Lichen planopilaris
Frontal fibrosing alopecia
Fibrosing alopecia in a pattern distribution
Discoid lupus erythematosus
Keratosis follicularis spinulosa decalvans
Folliculitis decalvans
Central centrifugal scarring alopecia
Acne keloidalis nuchae
Secondary cicatricial alopecia
Traction alopecia
Bacterial or fungal infection
Localized scleroderma
Radiation
Pemphigoid
Chemical or physical injuries
Scalp metastases

Clinical Presentation and Treatment

Lichen Planopilaris

Lichen planopilaris (LPP) is the most common cause of cicatricial alopecia and is considered the follicular form of lichen planus. Microarray analysis has recently shown a reduction in the

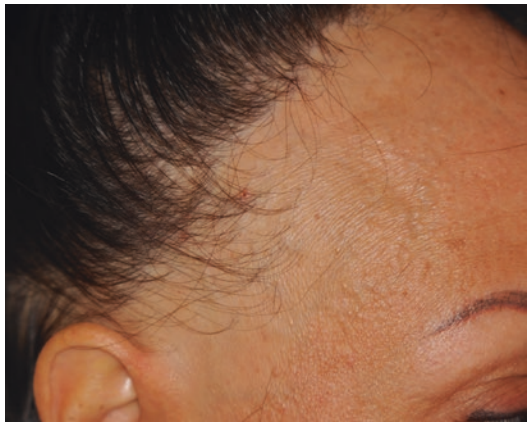

Fig. 15.1 Frontal fibrosing alopecia: hairline recession and loss of eyebrows. Also note prominence of temporal veins and papular lesions

expression of genes necessary for lipid metabolism and peroxisome biogenesis in LPP. Improper lipid metabolism in the sebaceous gland results in a toxic build-up of lipids that may be initiating the inflammatory response in this condition. It was hypothesized that peroxisome proliferator-activated receptor-γ (PPAR-γ), a master transcription factor that regulates these processes, is aberrant in LPP. To confirm the role of PPAR-γ in the pathogenesis of LPP, PPAR-γ was deleted in the follicular stem cells of mice, producing a phenotype similar to cicatricial alopecia. This finding supports the role of PPAR-γ in the pathogenesis of cicatricial alopecia and modulation of this pathway may be a new option to treat LPP.

The clinical variants of lichen planopilaris include classic lichen planopilaris, frontal fibrosing alopecia (Fig. 15.1), fibrosing alopecia in a pattern distribution and Graham-Little syndrome.

Classic lichen planopilaris presents as follicular destruction with a multifocal or central distribution and rarely involves the entire scalp. Patients usually seek medical advice once they have noticed one or several patches of hair loss. A certain degree of itching and burning is frequently reported. The pull test from these areas typically reveals anagen hairs with hyperkeratotic sheaths.

Frontal fibrosing alopecia causes progressive recession of the frontal hairline and loss of the eyebrows. The new hairline is uneven and lacks vellus follicles, while the alopecic area shows less signs of photoaging, compared to the skin of the forehead. This clinical variant most commonly presents in postmenopausal women. The pathogenesis of FFA is still unknown but the role of environmental toxins is suggested by epidemiology.

Treatment

Treatment results in complete remission of disease in about one-third of patients. Partial remission is obtained in another third, while the disease progresses despite treatment in the remaining patients.

Systemic Treatment

1. Steroids represent our treatment of choice. We utilize intramuscular triamcinolone acetonide at the dosage 0.5–1 mg/kg/month. Steroids are gradually tapered when active lesions disappear. This may require 6–12 months.
2. Azathioprine 100 mg/day is useful in association with systemic steroids in severe cases.
3. Systemic cyclosporine 3 mg/kg/day in our experience is scarcely effective.
4. Oral hydroxychloroquine at 200 mg, twice daily for 6–12 months is a second-line treatment. Dosage should not exceed 6.5 mg/kg.
5. Pioglitazone, a peroxisome proliferator-activated receptor (PPAR)-γ agonist is effective at a dosage of 15 mg daily. Liver function tests should be performed before starting therapy. Increased risk of bladder cancer has been reported in women taking the medication for more than 1 year.
6. Finasteride 1 mg/day in men and 2.5–5 mg/day for women or dutasteride 0.5 mg daily is useful in fibrosing alopecia in a pattern distribution and in frontal fibrosing alopecia.
7. Low-dose excimer 308-nm laser, two to three times a week, is useful in reducing the inflammation both in LPP and FFA.
8. We do not recommend retinoids for the treatment of LPP or FFA.

Topical Treatment

1. Topical 2 % or 5 % minoxidil may prevent fibrosis and is useful in association with systemic steroids. It also induces thickening of the remaining hair and allows better coverage of the alopecic area.
2. High-potency topical steroids can be prescribed in association with systemic treatment. However, they may aggravate skin atrophy, particularly in FFA.
3. Topical tacrolimus 0.1 % is useful as it does not cause skin atrophy. In lichen pigmentosus, topical tacrolimus 0.03 % twice daily can improve the pigmentation of the lesions.

Discoid Lupus Erythematosus

Discoid lupus erythematous (DLE) is the most frequent form of chronic cutaneous lupus erythematosus that leads to primary cicatricial alopecia. While it was previously shown that type 1 IFN production drives the inflammatory processes in DLE, new studies show that autoreactive Granzyme B-positive CD8+ T cells are recruited to the hair follicles, resulting in epidermal, dermal, and follicular destruction. Additionally, scientists have found a variant of a gene that may exert a strong genetic influence on the risk of developing DLE, independently from systemic involvement. It was found that the coding variant rs1143679 of ITGAM was highly associated with DLE patients without systemic involvement and with SLE patients that exhibited discoid rashes.

Diagnosis of DLE is strongly suggested by the presence of photosensitivity and "discoid" erythematous papules with follicular hyperkeratosis. As the disease progresses, the plaques enlarge and spread centrifugally and eventual atrophy with telangiectasia become evident. DLE may also present with cysts and comedones. Patients most commonly complain of single or multiple patches of hair loss, burning and scalp tenderness. Direct immunofluorescence is useful for the diagnosis and is often necessary for differentiating DLE from other primary lymphocytic cicatricial alopecias. All patients with discoid lupus erythematosus should be examined for systemic lupus erythematosus. Complications include the development of squamous cell carcinoma.

Treatment

DLE usually responds to treatment.

Systemic Treatment

1. Antimalarial drugs are the treatment of choice. Hydroxychloroquine 200 mg twice a day or chloroquine (200 mg/day) can be prescribed alone or in association with systemic steroids. Treatment lasts for at least 3 months and is then tapered to the lowest effective dose.
2. Quinacrine may be used in combination with other antimalarials in patients that have resistance or only partially respond to chloroquine or hydroxychloroquine. It may also be used as a monotherapy in patients with preexisting eye problems that contraindicate the use of chloroquine or hydroxychloroquine. The dosage of quinacrine as monotherapy or in combination therapy is 100 mg/day, for approximately 9 months.
3. Systemic steroids: initial dosages should be 40–60 mg/day of oral prednisone or 0.5–1 mg/month of intramuscular triamcinolone acetonide. Steroids are gradually tapered when active lesions disappear. This may require 3–6 months.
4. Thalidomide (100–300 mg/day) is a possible alternative. Low-dose thalidomide is an effective treatment for refractory DLE, but its benefits need to be balanced against the potential adverse effects.
5. Retinoids may be used as an alternative treatment for DLE patients that do not respond to topical glucocorticoids, sunscreens and tacrolimus.

Intralesional Steroids

Triamcinolone acetonide (5 mg/ml sterile saline solution) can be useful in localized lesions.

Topical Treatment

1. High-potency topical steroids can be utilized when the disease is circumscribed to a small area of the scalp.

2. Tacrolimus (0.1 %) ointment can be applied twice daily.
3. Topical 2 % or 5 % minoxidil may prevent fibrosis and can be prescribed in association with systemic steroids.
4. Topical tocoretinate 0.25 % ointment applied twice daily has been shown to improve erythema after 1 month. Improvements in skin pigmentation and atrophy can be observed after 12 months of treatment. Alternatively, topical tazarotene 0.05 % gel applied once daily at bedtime may show improvements.

General Measures

Patients should wear a hat to avoid sun exposure.

Folliculitis Decalvans (FD)

This term is utilized to include a spectrum of scalp disorders characterized by painful acute inflammatory changes with or without pustules (Fig. 15.2). Eventually, relapsing inflammatory episodes result in irregularly shaped areas of cicatricial alopecia and tufted folliculitis. Although *Staphylococcus aureus* may frequently be isolated from the pustules, FD is not an infective condition. Some cite local immunological deficits as a predisposition to this condition, while others propose the cause to be an abnormal host response against staphylococcal antigens or toxins. Recent studies suggest that bacterial biofilm organization may play a role in the pathogenesis. Biofilm presence in the root sheaths and on the hair shafts within the lesions was confirmed using field emission scanning electron microscopy and confocal laser scanning microscopy.

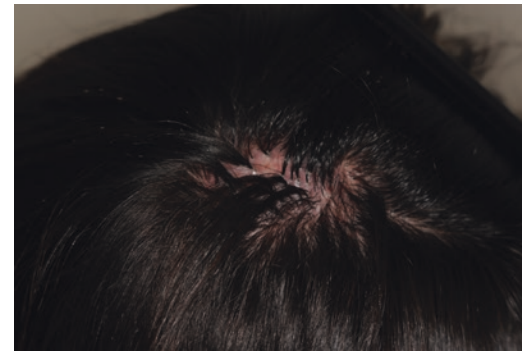

Fig. 15.2 Folliculitis decalvans: scarring alopecia and tufted folliculitis

Treatment

Although the inflammatory scalp lesions usually subside with treatment, the condition has a chronic course with frequent relapses.

Systemic Treatment

1. Oral antibiotics are the treatment of choice. Possible alternatives include erythromycin 1 g/day, clarithromycin 500 mg/day, cephalosporins or tetracyclines (minocycline 100 mg/day or hydroxytetracycline 200 mg/day). All of these are usually effective in arresting the inflammatory process, but relapses are common as soon as the treatment is interrupted. Combination therapy with fusidic acid 1,500 mg/day for 3 weeks and zinc sulphate 400 mg/day resulted in permanent remission in three patients (Abeck et al. 1992). The association of rifampicin 300 mg and clindamycin 300 mg taken both orally twice daily for 10 weeks is probably the most effective treatment (Powell et al. 1999, Powell and Dawber 2001). This may be due to the fact that rifampicin is an effective antistaphylococcal agent in addition to its immunomodulatory properties. This regimen produces long-term remissions in most patients. This particular association is required to avoid development of rifampicin resistance that is common when using this antibiotic alone.
2. Isotretinoin 0.5–1 mg/kg/day is effective in some patients but actually worsens the scalp inflammation in most cases.
3. TNF-alpha blockers are an option in recalcitrant cases. Infliximab at a dose of 5 mg/kg of body weight can improve symptoms after three infusions.

A short course of systemic and topical steroids, associated with oral antibiotics, can be utilized to suppress inflammation.

Topical Treatment

1. Shampoos containing antibacterial agents or 2 % ketoconazole may be helpful in preventing relapses.
2. Photodynamic therapy with methyl aminolevulinate has been shown to be effective in some cases.
3. Shaving of the scalp may improve the disease.

Keratosis Follicularis Spinulosa Decalvans (KFSD)

KFSD is a rare, inherited or sporadically acquired X-linked disorder with a mutation in the MBTPS2 gene, mapped to Xp21,13-p22.2. A more severe form of KFSD that is inherited in an autosomal dominant fashion has recently been proposed (Bellet et al. 2008). MBTPS2 codes for an intramembrane zinc metalloprotease necessary for the cleavage of sterol regulatory element-binding proteins (SREBPs). This genetic condition usually becomes evident in infancy and worsens into adulthood. Clinical diagnosis is suggested by the presence of follicular keratotic papules and pustules involving the vertex followed by progressive alopecia. Its severity considerably varies from patient to patient and may involve follicular papules on the eyebrows and cheeks. Due to the clinical and genetic heterogeneity observed in patients with KFSD, clear diagnosis is frequently challenging.

Treatment

In most patients the disease does not respond to treatment.

Systemic Treatment

1. Acitretin (0.5–0.75 mg/kg/day) may be effective in some cases. Isotretinoin 1 mg/kg/day is usually not effective.
2. Dapsone 100 mg/day can be tried in patients who do not respond to retinoids.
3. Oral antibiotics (tetracyclines, macrolides or rifampicin) as treatment for folliculitis decalvans are rarely effective.

Topical Treatment

1. Topical steroids and keratolytics may partially improve the symptoms.
2. Intense pulsed light (IPL) and pulse dye laser (PDL) has improved the condition in some patients.

Pseudopelade of Brocq

Pseudopelade of Brocq was classically thought of as not a separate entity but a representation of the final outcome of advanced lichen planopilaris. Yu et al. (2010), using significance analysis of microarrays, showed that the global gene expression profiles in lichen planopilaris and pseudopelade of Brocq are distinct from one another. This data suggest different etiologies accounting for these two disorders, which should be considered as separate conditions. The scalp presents multiple atrophic, irregular areas involving the vertex with little to no signs of inflammation. Mild pruritus and involvement of the beard area have also been reported. Clinicopathological features of classic pseudopelade of Brocq show nonspecific findings. There have been case reports of alopecia areata clinically mimicking this disease (Kittridge et al. 2010). The pull test from these areas typically reveals anagen hairs during active disease.

Patients with this diagnosis may fall into two possible categories:

- Patients in whom the disease has already spontaneously remitted
- Patients in whom the biopsy has been wrongly taken from an atrophic area

Currently, no established protocols exist for treating this condition.

Treatment

No chemical medical treatment is available.

Surgical autologous hair transplantation may be considered in patients with stable disease. Initial test transplantation of approximately 50 grafts should be done to exclude the possibility of reactivating the disease during the larger grafts.

Localized Scleroderma

Localized scleroderma (LE) is a connective tissue disorder in which a dysregulation of connective tissue metabolism leads to an excess deposition of collagen. Early stages of the disease are characterized by an elevation of Th1/Th17-associated cytokines and Th2-associated cytokines during the late stages of disease, resulting in fibrosis. Furthermore, microRNAs (miRNAs) may perpetuate the fibrosis seen in LE. Studies have shown that the miRNA let-7a, in particular, is downregulated in LE lesions (Makino et al. 2013; Etoh et al. 2013). Interestingly, miRNA injection of let-7a into the mouse model of bleomycin-induced dermal sclerosis improved the skin fibrosis. Insight into the role of miRNA in the pathogenesis of localized scleroderma may aid in the development of novel therapeutics to treat this disease.

Localized scleroderma of the scalp presents as a slowly progressing irregular patch of hair loss. The skin often shows a certain degree of erythema or pigmentation in the absence of follicular keratosis or scaling. The patch is often not completely bald but presents some vellus or intermediate hairs. Severe atrophy with involvement of the hypodermis and muscles is a feature of frontoparietal linear scleroderma ("en coup de sabre").

Treatment

Localized scleroderma usually resolves spontaneously in a few years.

Systemic Treatment

1. Systemic calcitriol at the dosage of 0.50–0.75 mg/day is effective in localized scleroderma of the scalp including frontoparietal linear scleroderma.
2. Oral methotrexate at 15 mg/m^2 (maximum 20 mg/week) for 6 months may be combined for the first 3 months with oral prednisone (1 mg/kg/day, maximum 50 mg/day) as a single morning dose.
3. Ultraviolet (UV) A1 phototherapy is a possible option.

Topical Treatment

1. Topical 2 % or 5 % minoxidil.
2. Calcipotriol lotion may be effective.
3. Tacrolimus 0.1 % ointment twice a day is a possible option.

Patients with frontal linear scleroderma ("en coup de sabre") can gain functional and aesthetic results from surgical volumetric restoration with fat grafting.

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