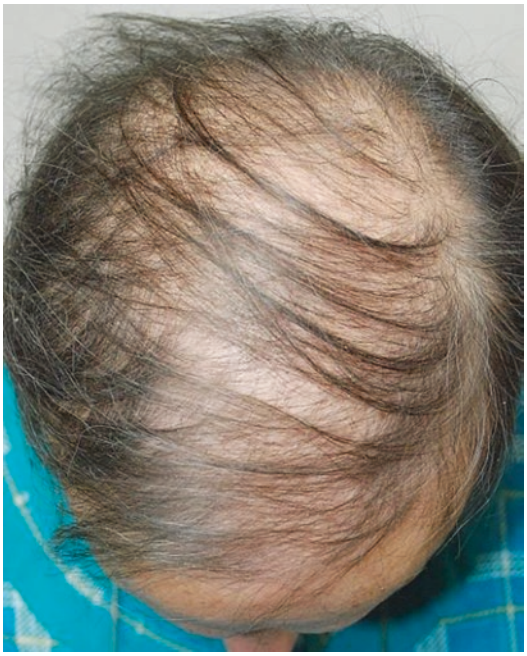
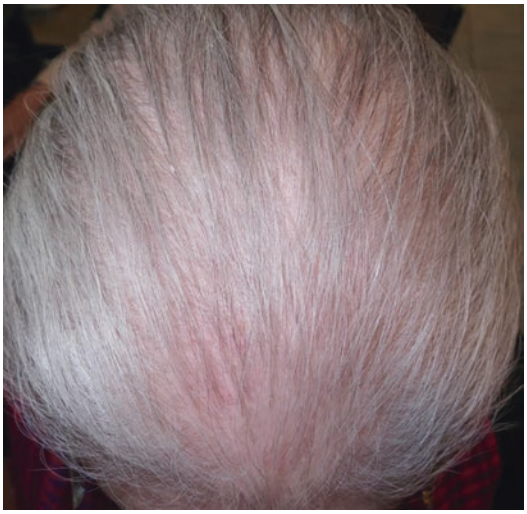
Disorders of Hair

35

Lidia Rudnicka and Anna Waśkiel-Burnat



1. A 65-year-old woman with hair loss limited to frontal area for 5 years.
   (a) What is your diagnosis?
   (b) What is the management?



2. A 76-year-old woman with diffuse hair loss for 2 weeks (with positive pull test). She had undergone cholecystectomy 3 months ago.
   (a) What is your diagnosis?
   (b) What is the prognosis in this condition?
   (c) What is the management?

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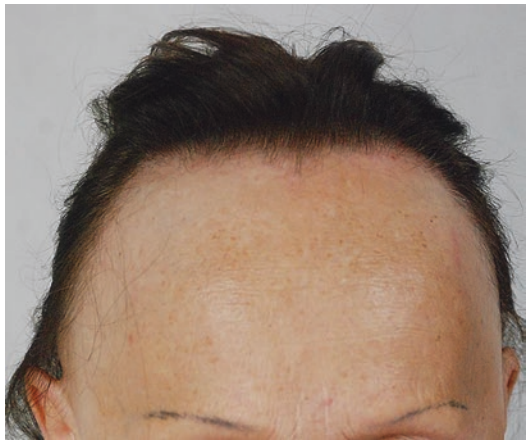
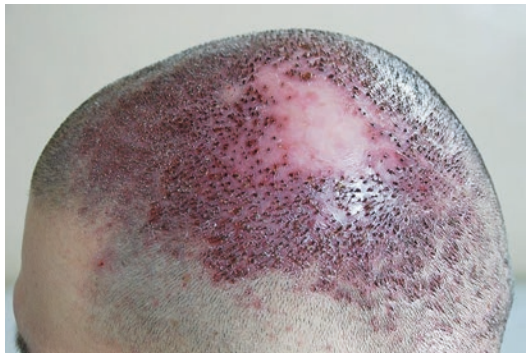
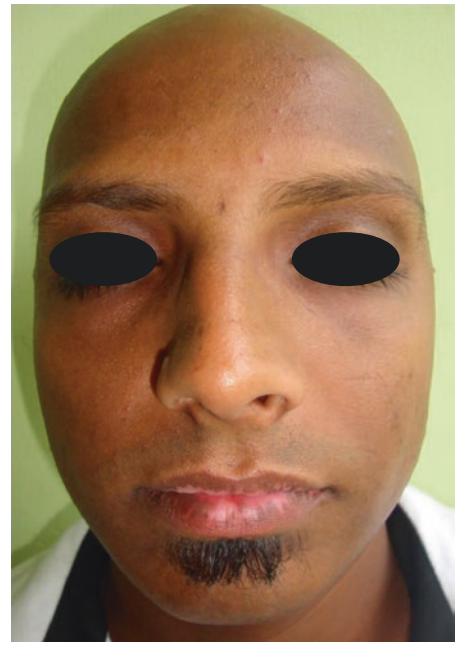
R. R. Ranawaka et al. (eds.), *Atlas of Dermatoses in Pigmented Skin*,
https://doi.org/10.1007/978-981-15-5483-4_35



3. A 51-year-old woman with hair and eyebrow loss for 6 months.
   (a) What is your diagnosis?
   (b) What are the key clinical features that support your diagnosis?
   (c) How do you confirm it?
   (d) What are the treatment options for this disease?



4. A 34-year-old man with hair loss for 1 year. He complained of a lot of purulent secretions.
   (a) What is your diagnosis?
   (b) What are the key clinical features that support your diagnosis?
   (c) How do you confirm the diagnosis?
   (d) What is your management?



(photographed by Dr Ranthilaka R. Ranawaka)

5. A 32-year-old man had total hair loss on the scalp and body for 3 years.
   (a) What is the diagnosis?
   (b) What other diseases can coexist?

Answers

1. (a) Female androgenetic alopecia.
   (b) Topical minoxidil and 5-alpha reductase inhibitors (finasteride or dutasteride). Other treatment options include oral minoxidil, spironolactone, cyproterone acetate, and flutamide.
2. (a) Acute telogen effluvium.
   (b) The disease is a self-limited condition.
   (c) Surgery is the most probable cause of hair loss. No treatment is necessary. Observation is recommended.
3. (a) Frontal fibrosing alopecia.
   (b) The disease most commonly affects postmenopausal women. Recession of fronto-temporal hairline with coexisted eyebrow loss is observed.

(c) Histological examination is the gold-standard diagnostic method. Trichoscopy can be useful to avoid scalp biopsy.
(d) The treatment options include antimalarials, finasteride/dutasteride, tetracyclines, retinoids, topical minoxidil, topical corticosteroids, intralesional corticosteroids, and topical calcineurin inhibitors.

4. (a) Folliculitis decalvans.
(b) The disease most commonly occurs in middle-aged men. Characteristic clinical feature is the presence of tufted hairs with purulent secretion.
(c) Histological examination is the gold-standard diagnostic method. Trichoscopy can be useful to avoid scalp biopsy.
(d) Systemic antibiotics (tetracyclines, first-generation cephalosporins, combination of rifampicin with clindamycin), isotretinoin, topical antiseptic agents, topical or intralesional corticosteroids.

5. (a) Alopecia universalis.
(c) This man has vitiligo on lips; alopecia areata may be associated with other autoimmune diseases, e.g., autoimmune thyroiditis and lupus erythematosus.

35.1 Non-scarring Disorders of Hair Growth

35.1.1 Androgenetic Alopecia (Pattern Hair Loss)

Definition Androgenetic alopecia (pattern hair loss) is the most common form of non-scarring hair loss, characterized by progressive miniaturization of terminal scalp hair with a pattern distribution (Kalidyadan et al. 2013; Lolli et al. 2017).

Clinical Features In women, diffuse central thinning with the frontal hairline spared, prominent frontal scalp thinning with a Christmas tree-like pattern, or recession of the hairline along the bilateral temporal regions are observed (Figs. 35.1, 35.2 and 35.3). In men, the vertex and fronto-temporal areas are the most prominently affected (Fig. 35.4) The disease affects up to 80% of Caucasian men and 50% of women with prevalence increasing with age (Lolli et al. 2017). It is less commonly reported in Asian and African population (Kaliyadan et al. 2013).

Investigations The diagnosis of pattern hair loss is commonly established based on patient history and clinical examination. Pull test is negative. Trichoscopy and histopathological examination may be useful to confirm the diagnosis. In women with clinical features of hyperandrogenism, laboratory tests are recommended (Lolli et al. 2017).

Histopathology The histopathological features of androgenetic alopecia include increased number of miniaturized hair follicles, reduced size of sebaceous glands, decreased anagen to telogen ratio, increased number of follicular stelae, and perifollicular inflammation around the upper portion of the hair follicle with or without perifollicular fibrosis (Kaliyadan et al. 2013; Lolli et al. 2017).

Pathogenesis Androgenetic alopecia is characterized by a progressive miniaturization of terminal hair follicles that results in an increased proportion of vellus follicles. The mechanism of follicular transformation is not fully clarified. However, the role of androgens and genetic susceptibility has been suggested (Kaliyadan et al. 2013).

Prognosis and Treatments In androgenetic alopecia, hair loss progresses with disease duration. On the contrary to male pattern hair loss, in women complete hair loss is not typically observed. Topical minoxidil is usually first-line therapy in both men and women. In male pattern hair loss, 5-alpha reductase inhibitors (finasteride or dutasteride) are recommended. In female androgenetic alopecia, 5-alpha reductase inhibitors (finasteride or dutasteride), spironolactone, cyproterone acetate, and flutamide are used. Other therapeutic option is hair transplantation. Recently, the effectiveness of oral minoxidil and platelet-rich plasma (PRP) in treatment of androgenetic alopecia has been described (Lolli et al. 2017).

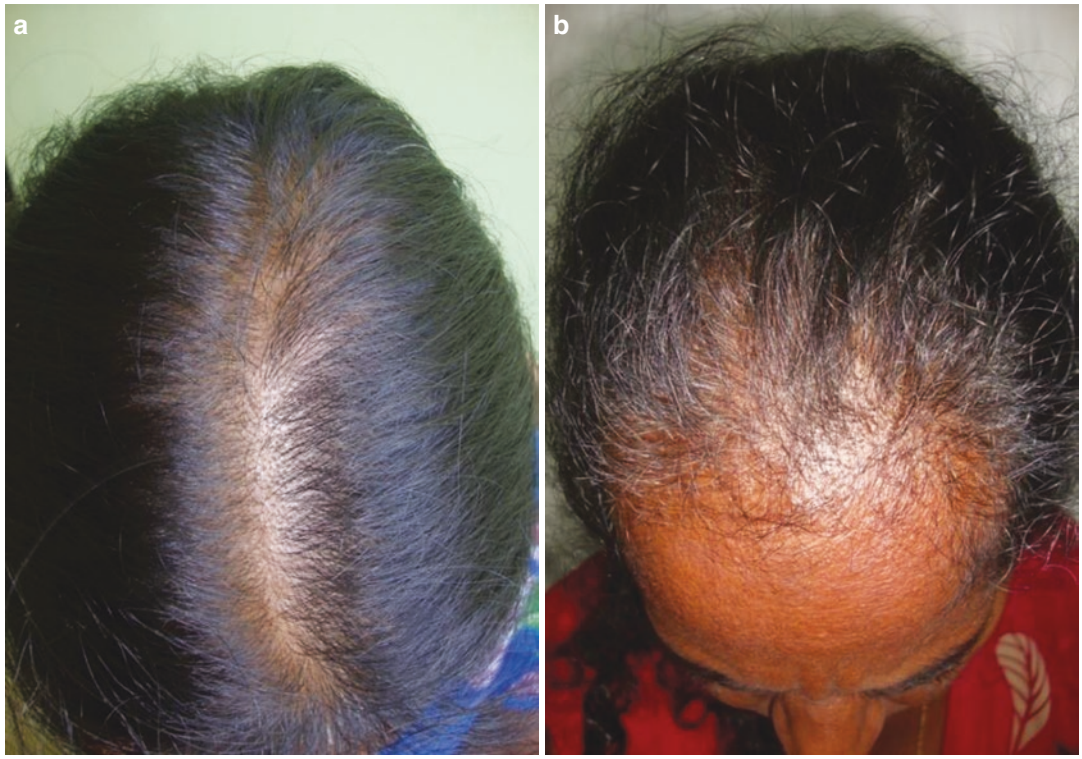


Fig. 35.1 (**a**, **b**) Female androgenetic alopecia—early to moderate (photographed by Dr Ranthilaka R. Ranawaka)

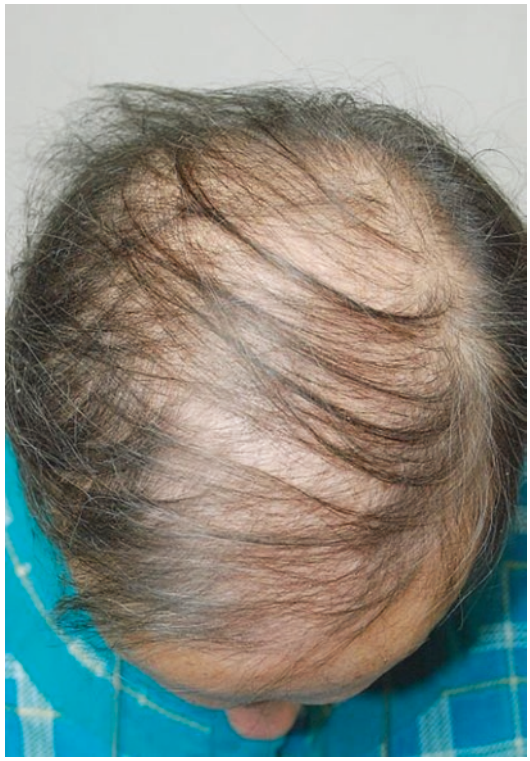


Fig. 35.2 Female androgenetic alopecia—late (photographed by Dr Anna Waśkiel-Burnat)

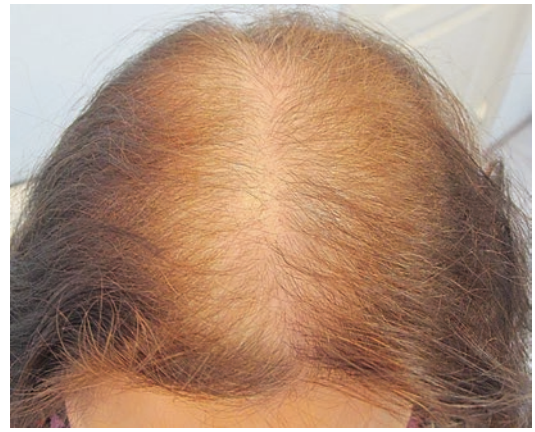


Fig. 35.3 Female androgenetic alopecia (photographed by Dr Adriana Rakowska, MD, PhD, Dermatologist, Department of Dermatology, Medical University of Warsaw, Poland)

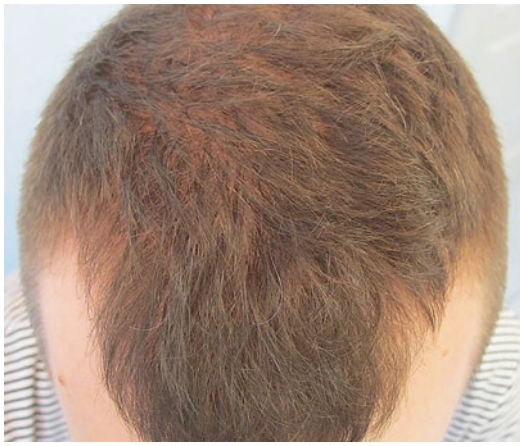


Fig. 35.4 Male pattern hair loss (photographed by Dr Anna Waśkiel-Burnat)

Differential Diagnosis Telogen effluvium, diffuse alopecia areata, central centrifugal cicatricial alopecia, frontal fibrosing alopecia, fibrosing alopecia in a pattern distribution, traction alopecia (Lolli et al. 2017).

35.1.2 Telogen Effluvium

35.1.2.1 Acute Telogen Effluvium

Definition Acute telogen effluvium is non-scarring hair loss that results from abnormal shift in follicular cycle which lasts less than 6 months (Grover and Khurana 2013).

Clinical Features The disease is characterized by diffuse, non-scarring hair loss (Fig. 35.5) that developed within 2–3 months after exposure to the triggering factor. Women are more commonly affected compared to men (Grover and Khurana 2013).

Investigations The diagnosis of acute telogen effluvium is mainly established based on patient history and physical examination. Hair pull test is positive. Trichoscopy, trichogram, and histopathological examination may be useful to confirm diagnosis. In the cases of not obvious patients' history, laboratory tests may be required (Grover and Khurana 2013).

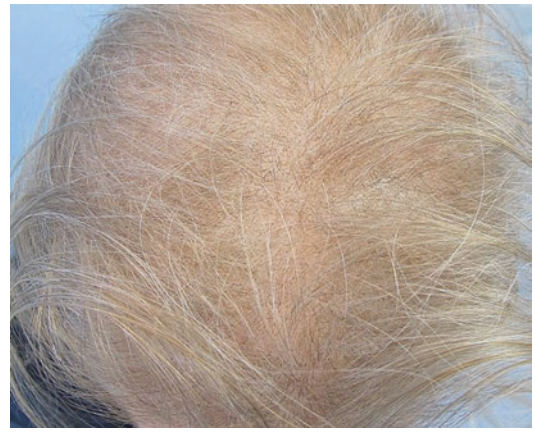


Fig. 35.5 Acute telogen effluvium (photographed by Dr Adriana Rakowska, MD, PhD, Dermatologist, Department of Dermatology, Medical University of Warsaw, Poland)

Histopathology An increased proportion of telogen follicles is the characteristic histopathologic finding (Grover and Khurana 2013).

Pathogenesis Telogen effluvium is characterized by increased percentage of hair follicles in the telogen phase resulting in subsequent prominent hair shedding. There are five functional types of telogen effluvium suggested: immediate anagen release, delayed anagen release, immediate telogen release, short anagen syndrome, and delayed telogen release. The most common triggering factors of acute telogen effluvium include febrile illness, surgery, childbirth, rapid weight loss, drugs, supplements or toxins, inflammatory conditions of the scalp, and significant emotional stress (Grover and Khurana 2013).

Prognosis and Treatments Acute telogen effluvium is a self-limited condition. Complete hair loss is not observed (Grover and Khurana 2013).

Differential Diagnosis Androgenetic alopecia, diffuse alopecia areata, anagen effluvium, loose anagen syndrome (Grover and Khurana 2013).

35.1.2.2 Chronic Telogen Effluvium

Definition Chronic telogen effluvium is non-scarring hair loss resulting from abnormal shift

in follicular cycle and persists more than 6 months (Grover and Khurana 2013).

Clinical Features It is characterized by diffuse, non-scarring hair loss (Fig. 35.6). Women are predominantly affected (Grover and Khurana 2013).

Investigations The diagnosis of chronic telogen effluvium is mainly established based on patient history, physical examination, and positive hair pull test. Trichoscopy, trichogram, and histopathological examination may be also used to confirm diagnosis. Laboratory tests including complete blood count, complete metabolic panel, thyroid-stimulating hormone, and ferritin are recommended to identify the underlying disease (Grover and Khurana 2013).

Histopathology An increased proportion of telogen follicles is the characteristic histopathologic finding (Grover and Khurana 2013).

Pathogenesis Telogen effluvium is characterized by increased percentage of hair follicles in the telogen phase resulting in subsequent prominent hair shedding. There are five functional types of telogen suggested: immediate anagen release, delayed anagen release, immediate telogen release, short anagen syndrome, and delayed telogen release. It may be primary or secondary to various conditions. The most common causes of secondary chronic telogen effluvium include thyroid disorders, iron deficiency, metabolic disorders, restricted diet, nutritional deficiencies, connective tissue diseases, drugs, and HIV infections. The disease may also be triggered by acute telogen effluvium. In primary chronic telogen effluvium, no specific triggering agent is identified (Grover and Khurana 2013).

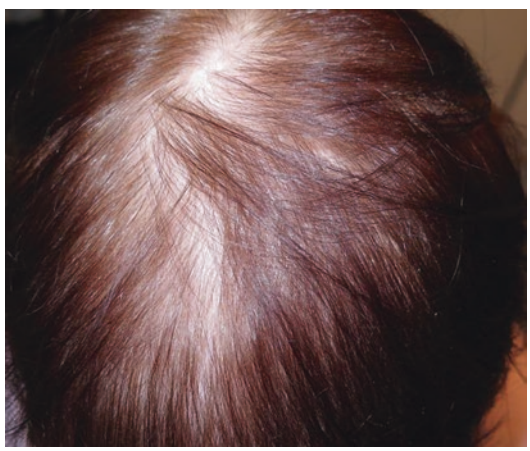


Fig. 35.6 Chronic telogen effluvium (photographed by Prof. Lidia Rudnicka)

Prognosis and Treatment Complete hair loss is not observed. Identifying and treating the underlying condition is the most important. The effectiveness of topical or oral minoxidil has been described (Grover and Khurana 2013).

Differential Diagnosis Androgenetic alopecia, diffuse alopecia areata, anagen effluvium, loose anagen syndrome (Grover and Khurana 2013).

35.1.3 Alopecia Areata

Definition: Alopecia areata is an autoimmune form of non-scarring hair loss that may affect any hair-bearing area (Messenger et al. 2016; Juárez-Rendón et al. 2017).

Clinical features: Alopecia areata is characterized by the presence of hair loss areas within the skin which remains normal (Figs. 35.7, 35.8, 35.9, 35.10, 35.11, 35.12, and 35.13). Although the scalp is most commonly affected, hair loss can also be observed in other hair-bearing areas (such as eyebrows, eyelashes, pubic and axillary areas). Based on the extent of hair loss, alopecia areata is classified as: patchy alopecia areata with partial scalp hair loss, alopecia areata totalis with complete scalp hair loss and alopecia areata universalis with complete scalp and body hair loss. Alopecia areata can be associated with several autoimmune diseases; thyroiditis, lupus erythematosus and vitiligo (Messenger et al. 2016; Juárez-Rendón et al. 2017).

Investigations: The diagnosis of alopecia areata is mainly established based on patient history and physical examination. Trichoscopy and histopathological examination may be also used to confirm diagnosis (Messenger et al. 2016; Juárez-Rendón et al. 2017).

Histopathology: Anagen follicles at the margins of expanding patches of alopecia areata characteristically show a perifollicular and intrafollicular inflammatory cell infiltrate, concentrated in and around the hair bulb (Messenger et al. 2016; Juárez-Rendón et al. 2017).

Pathogenesis: The exact pathophysiology of the disease is not fully understood. However, it is suggested that alopecia areata is caused by an autoimmune reaction to the hair follicles due to both genetic and environmental factors Prognosis and treatment: The course of alopecia areata is unpredictable. Spontaneous remission can occur. In some patients relapsing episodes, progression to extensive loss, and prolonged courses with the lack of regrowth are observed. The most important negative prognostic factors are the extent of hair loss, an ophiasis pattern of hair loss, a long duration of the disease, onset of the disease before puberty, atopy, a positive family history, the presence of other autoimmune diseases and nail involvement. Treatment options depends on severity of hair loss and age of the patient (Strazzulla et al. 2018; Peloquin and Castelo-Soccio 2017). Topical or intralesional corticosteroids are usually first-line treatment options. Other therapeutic modalities include: systemic corticosteroids, contact immunotherapy, phototherapy, immunosuppressant agents (cyclosporine, methotrexate, azathioprine), JAK inhibitors

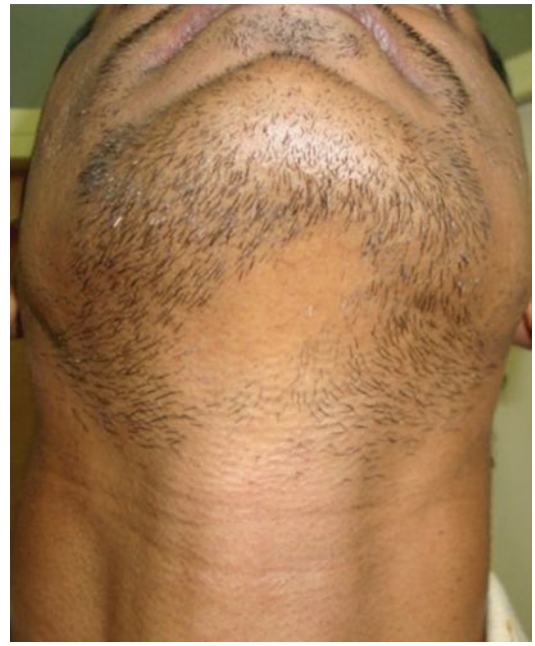


Fig. 35.8 Patchy alopecia areata on the beard area (photographed by Dr Ranthilaka R. Ranawaka)

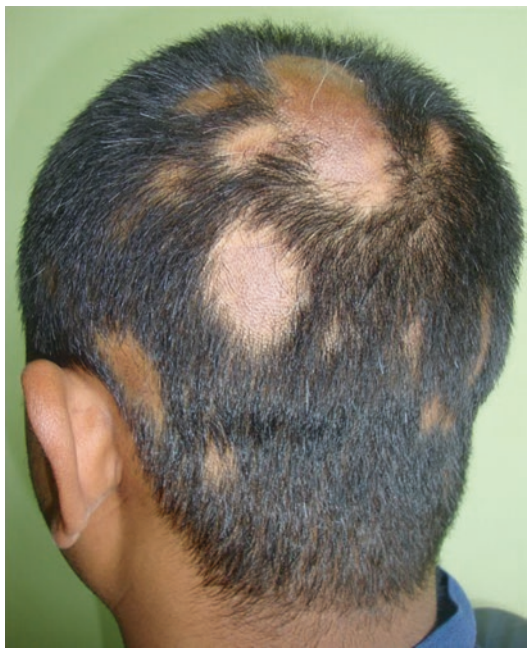


Fig. 35.7 Patchy alopecia areata on the scalp in a young man (photographed by Dr Ranthilaka R. Ranawaka)

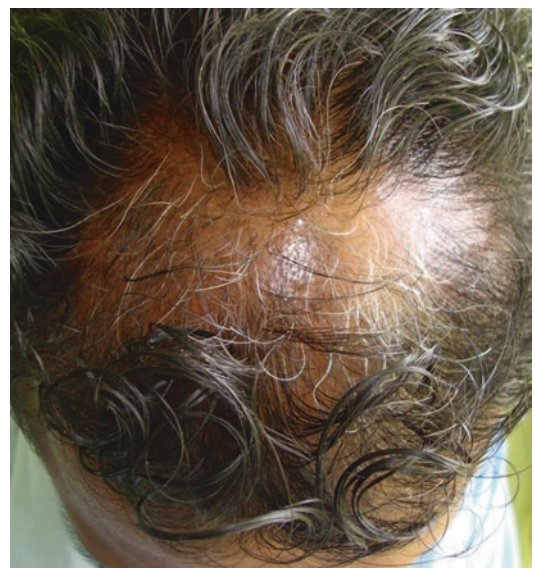


Fig. 35.9 A 47-year-old man with gray hair showing spared white hair. The disease process appears preferentially to affect pigmented hair, so that non-pigmented or white hair is spared (photographed by Dr Ranthilaka R. Ranawaka)

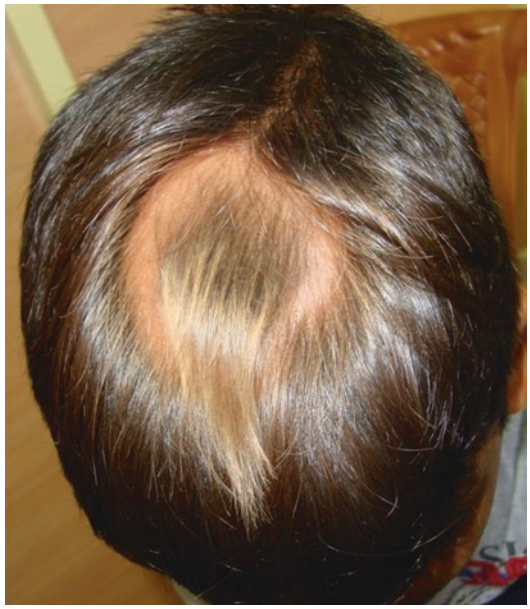


Fig. 35.10 During re-growth phase hair may be non-pigmented or hypopigmented. But hair pigmentation usually recovers completely (photographed by Dr Ranthilaka R. Ranawaka)

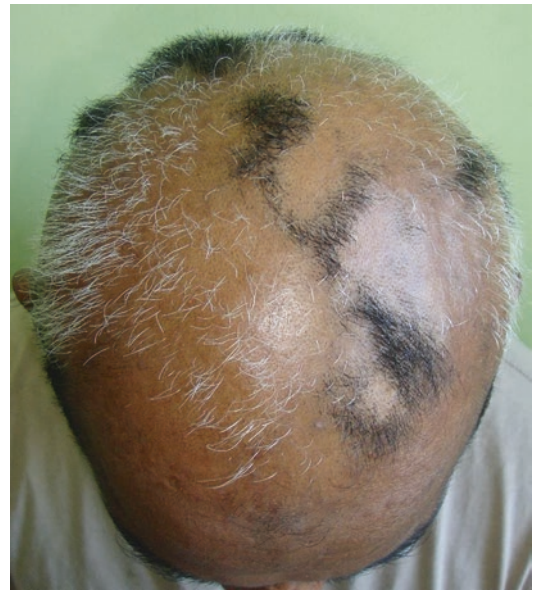


Fig. 35.11 Alopecia totalis showing re-growing hairs which are white in a 29-year-old man (photographed by Dr Ranthilaka R. Ranawaka)

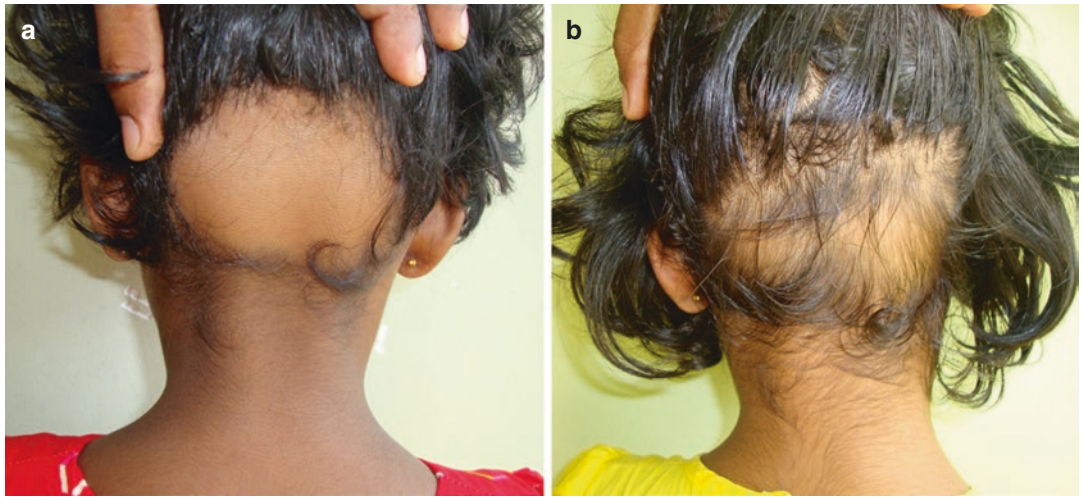


Fig. 35.12 Ophiasis pattern in a 9-year-old girl (photographed by Dr Ranthilaka R. Ranawaka). The prognosis is less favorable when onset occurs during childhood and in ophiasis pattern. (**a**) At presentation. (**b**) Hair growth after 6 months of treatment with monthly superficial liquid nitrogen cryotherapy and daily potent topical steroids

(topical and systemic) and topical minoxidil. Non-medical treatments include wigs or hairpieces; men tend to shave their heads; semipermanent tattooing can be helpful to disguise the loss of eyebrow (Spano and Donovan 2015).

Differential diagnosis: tinea capitis, trichotillomania, cicatricial alopecia and syphilitic alopecia (Messenger et al. 2016; Juárez-Rendón et al. 2017).

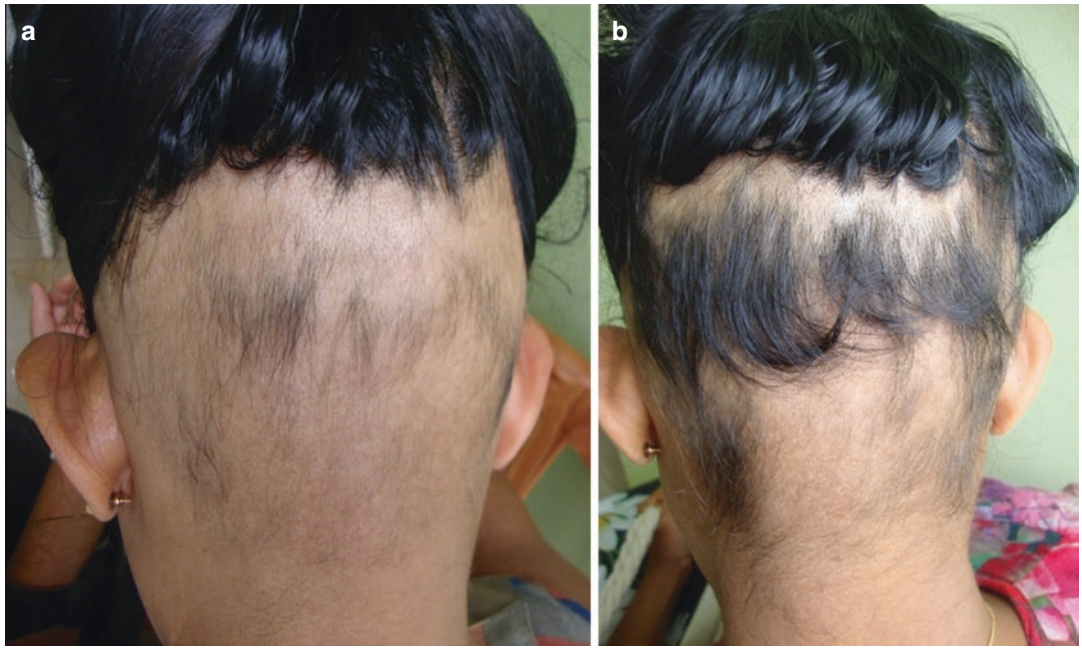


Fig. 35.13 Ophiasis pattern in a 37-year-old woman for more than 3 years (photographed by Dr Ranthilaka R. Ranawaka). She had poor response to treatments with multiple modalities: oral dexamethasone pulses, monthly superficial liquid nitrogen cryotherapy, and daily topical 5% minoxidil lotion. (**a**) At presentation. (**b**) After 3 months of therapy with intralesional steroids monthly and topical minoxidil daily. She refused oral drugs

35.2 Scarring Disorders of Hair Growth

35.2.1 Acquired Scarring Alopecia

Definition Acquired scarring alopecia (secondary scarring alopecia) refers to the destruction of hair follicles secondary to inflammatory processes, neoplastic conditions, or physical trauma (Fanti et al. 2018).

Clinical Features Scarring hair loss limited to area of the underlying condition is observed (Figs. 35.14, 35.15, 35.16, 35.17, and 35.18).

Investigations The diagnosis of acquired scarring alopecia is established based on patients' history and clinical picture. Additional examination (such as histopathological or direct immunofluorescence test) may be helpful to identify the underlying disease (Kanti et al. 2018).

Histopathology A histopathological features of the initial disease with coexisting scarring are observed.

Pathogenesis Acquired scarring alopecia results from inflammatory processes and mechanical damage of the surrounding tissue that subsequently affect and destroy the hair follicles (Fanti et al. 2018; Kanti et al. 2018).

The causes of acquired scarring alopecia include physical or chemical trauma (burns, toxic substances), radiation, infections (bacterial, fungal, or viral), malignant and benign tumors, genodermatoses (aplasia cutis congenita, ectodermal dysplasia, ichthyosis, epidermolysis bullosa, Darier's disease), granulomatous disease (sarcoidosis), autoimmune disease (localized scleroderma, cicatricial pemphigoid, lichen sclerosus, graft-versus-host disease), "deposition" dermatoses (amyloidosis, mucinosis), and inflammatory diseases (psoriasis, pityriasis amiantacea) (Fanti et al. 2018, Kanti et al. 2018).

Prognosis and Treatment Treatment of the underlying condition is necessary. Hair transplantation may be effective (Kanti et al. 2018).

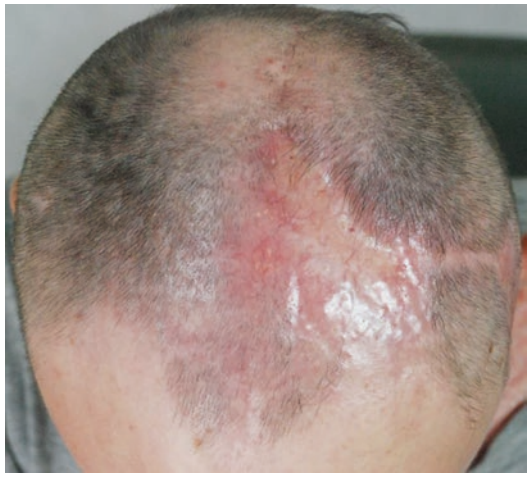


Fig. 35.14 Scarring alopecia secondary to lichen sclerosus on scalp (photographed by Dr Anna Waśkiel-Burnat)

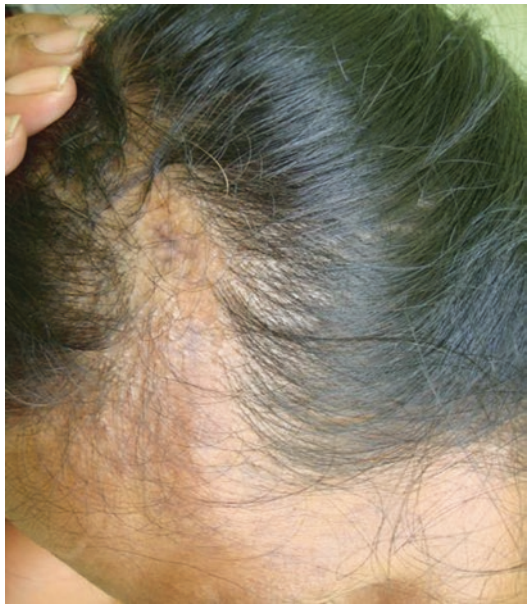


Fig. 35.15 Scarring alopecia secondary to linear morphoea (photographed by Dr Ranthilaka R. Ranawaka)

Differential Diagnosis Chronic lupus erythematosus, pseudopelade of Brocq, central centrifugal cicatricial alopecia, folliculitis decalvans, dissecting cellulitis (Fanti et al. 2018).

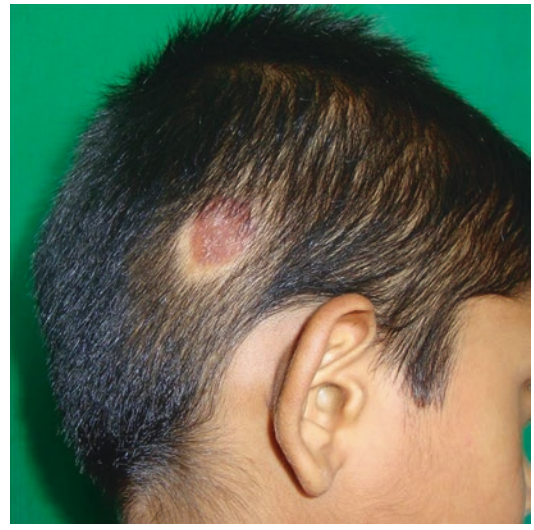


Fig. 35.16 Scarring alopecia secondary to a healed kerion (photographed by Dr Ranthilaka R. Ranawaka)

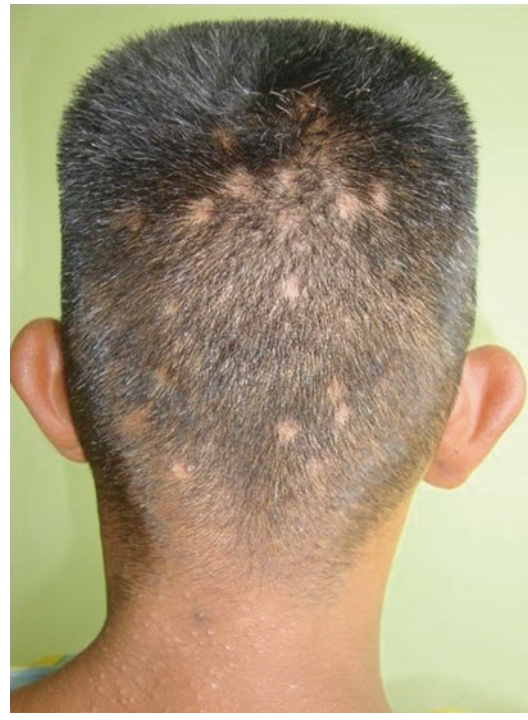


Fig. 35.17 Patchy scarring alopecia secondary to bacterial folliculitis (photographed by Dr Ranthilaka R. Ranawaka)

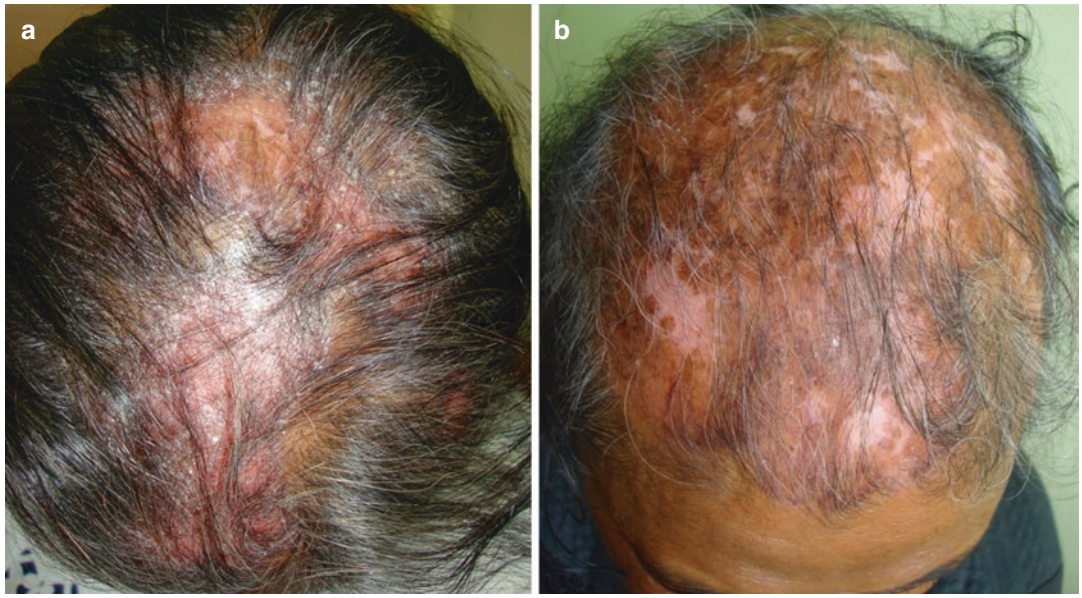


Fig. 35.18 Scarring alopecia secondary to sarcoidosis on scalp (**a**) before treatments (**b**) healed with scarring alopecia (photographed by Dr Ranthilaka R. Ranawaka)

35.2.2 Non-specific Scarring Alopecia

Definition Non-specific scarring alopecia is defined as an idiopathic scarring alopecia with inconclusive clinical and histopathologic findings or the end stage of inflammatory scarring alopecias (Fanti et al. 2018; Kanti et al. 2018).

Clinical Features Scarring hair loss areas are observed (Fanti et al. 2018; Kanti et al. 2018).

Investigations Diagnosis of non-specific scarring alopecia is established in absence of clinical and histopathological findings characteristic for other causes of scarring hair loss (Kanti et al. 2018).

Differential Diagnosis Lichen planopilaris, chronic lupus erythematosus, pseudopelade of Brocq, central centrifugal cicatricial alopecia, folliculitis decalvans, dissecting cellulitis (Fanti et al. 2018; Kanti et al. 2018).

35.2.3 Lichen Planopilaris

Definition Lichen planopilaris, a variant of lichen planus, is primary lymphocyte-mediated

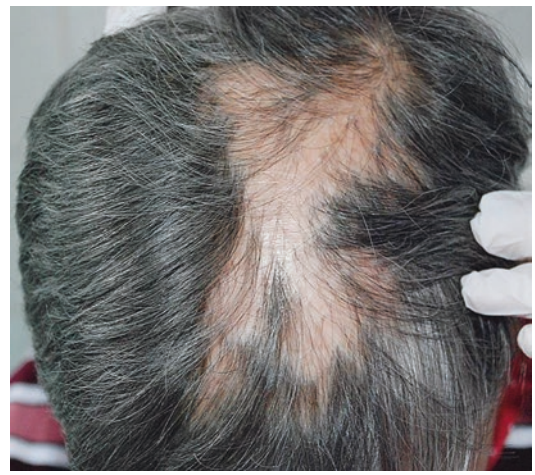


Fig. 35.19 Lichen planopilaris (photographed by Dr Joanna Golińska, MD, Resident of Dermatology and Venereology, Department of Dermatology, Medical University of Warsaw, Poland)

form of cicatricial alopecia (Kanti et al. 2018).

Clinical Features Classic lichen planopilaris presents as cicatricial hair loss areas with presence of perifollicular erythema and follicular hyperkeratosis (Figs 35.19, 35.20 35.21 and 35.22). It predominantly affects the vertex or the parietal areas. The lesions are commonly associated with itching, burning, or scalp tenderness.

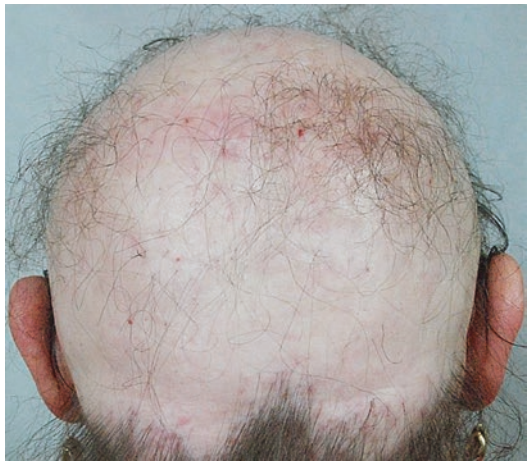


Fig. 35.20 Lichen planopilaris (photographed by Dr Patrycja Gajda, MD, Resident of Dermatology and Venereology, Department of Dermatology, Medical University of Warsaw, Poland)

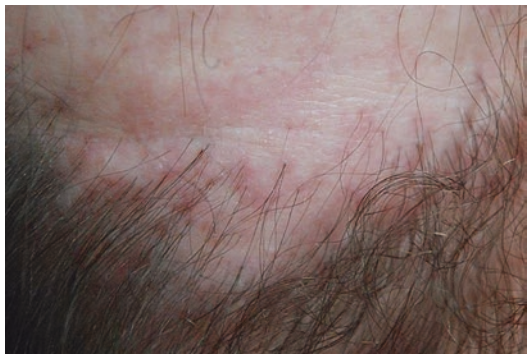


Fig. 35.21 Lichen planopilaris closer view (photographed by Dr Joanna Golińska, MD, Resident of Dermatology and Venereology, Department of Dermatology, Medical University of Warsaw, Poland)

Women are more commonly affected compared to men. The typical age of onset is between 40 and 60 years (Fanti et al. 2018; Kanti et al. 2018).

Investigations The diagnosis of classic lichen planopilaris is established based on clinical picture and a histopathological examination. Trichoscopy may be useful to avoid scalp biopsy (Fanti et al. 2018).

Histopathology In histopathological examination, dense lymphocytic infiltrate and fibrosis around the infundibulum and isthmus of the hair follicle often with lichenoid interface dermatitis involving the upper follicle and loss of sebaceous glands are observed (Fanti et al. 2018; Kanti et al. 2018).

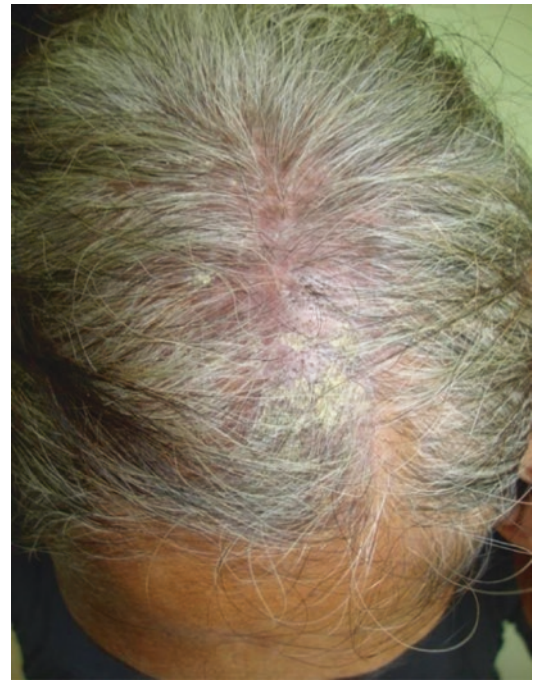


Fig. 35.22 Lichen planus causing scarring alopecia in a 66-year-old woman who had generalized lichen planus (photographed by Dr Ranthilaka R. Ranawaka)

Pathogenesis The pathogenesis of lichen planopilaris remains to be not fully elucidated. However, an autoimmune etiology is generally accepted (Kanti et al. 2018).

Prognosis and Treatment The course of lichen planopilaris is usually slowly progressive. Spontaneous remission may occur. Topical corticosteroids and intralesional corticosteroid injections are commonly used as first-line therapy for classic lichen planopilaris. Other treatment options include antimalarials, systemic steroids, tetracyclines, pioglitazone, retinoids, methotrexate, cyclosporine, and mycophenolate mofetil(Racz et al. 2013; Assouly and Reygagne 2009).

Differential Diagnosis Discoid lupus erythematosus, pseudopelade of Brocq, central centrif-

ugal cicatricial alopecia, folliculitis decalvans, dissecting cellulitis, alopecia areata.

35.2.4 Frontal Fibrosing Alopecia

Definition Frontal fibrosing alopecia, a variant of lichen planopilaris, is lymphocytic, primary scarring alopecia (Kanti et al. 2018).

Clinical Features The disease is characterized by a symmetrical, progressive recession of frontal or fronto-temporal hairline frequently associated with eyebrow hair loss (Figs. 35.23, 35.24 and 35.25). Postmenopausal women are predominantly affected. Perifollicular erythema, hyperkeratosis, and symptoms of itching and burning are typically observed (Kanti et al. 2018; Fanti et al. 2018).

Investigations The diagnosis of frontal fibrosing alopecia is established based on clinical picture and histopathological examination. Trichoscopy may be useful to avoid scalp biopsy (Fanti et al. 2018; Kanti et al. 2018).

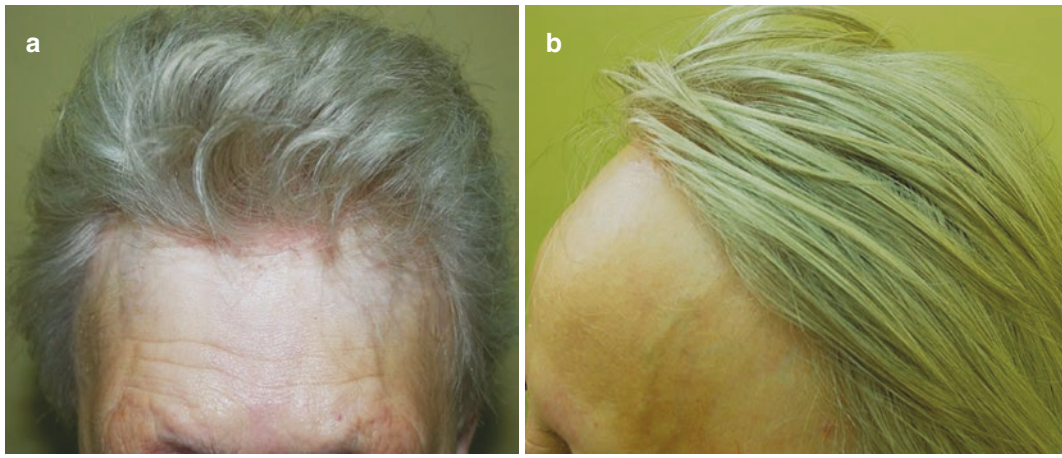


Fig. 35.23 (**a**, **b**) Frontal fibrosing alopecia (photographed by Dr Anna Waśkiel-Burnat)

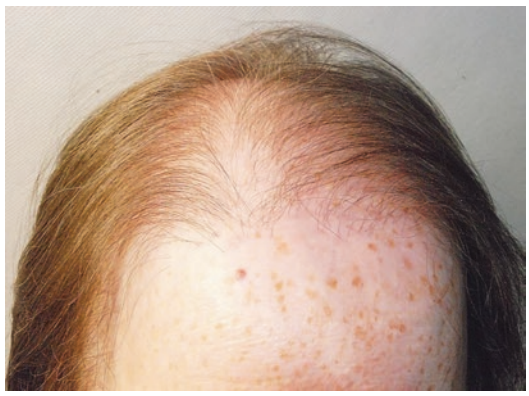


Fig. 35.24 Frontal fibrosing alopecia with female androgenetic alopecia (photographed by Dr Anna Waśkiel-Burnat)

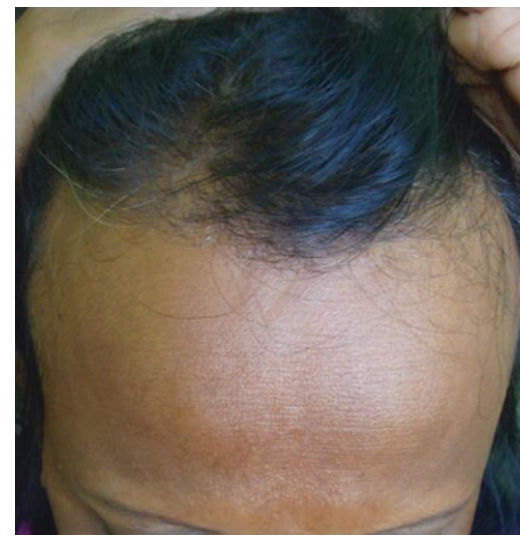


Fig. 35.25 Frontal fibrosing alopecia with female androgenetic alopecia in a 45-year-old woman (photographed by Dr Ranthilaka R. Ranawaka)

Histopathology Histopathological examination of frontal fibrosing alopecia is consistent with lichen planopilaris (Fanti et al. 2018).

Pathogenesis The pathogenesis of frontal fibrosing alopecia is not fully understood. However, the role of androgens has been suggested (Kanti et al. 2018).

Prognosis and Treatment The disease is typically slowly progressive. Spontaneous stabilization may occur. Treatment options for frontal fibrosing alopecia include antimalarials, finasteride/dutasteride, retinoids, tetracyclines, topical minoxidil, topical corticosteroids, intralesional corticosteroids, and topical calcineurin inhibitors (Racz et al. 2013).

Differential Diagnosis Alopecia areata, androgenetic alopecia, cicatricial marginal alopecia, traction alopecia.

35.2.5 Graham-Little Syndrome

Definition Graham-Little syndrome is a variant of lichen planopilaris (Assouly and Reygagne 2009).

Clinical Features The disease is characterized by a triad consisting of cicatricial patchy scalp alopecia (Fig. 35.26), non-cicatricial axillary and pubic hair loss, and lichenoid follicular eruptions. Middle-aged women are most commonly affected (Assouly and Reygagne 2009).

Investigations The diagnosis of Graham-Little syndrome is established based on clinical picture and a histopathological examination. Trichoscopy and dermoscopy may be useful to avoid skin biopsy (Assouly and Reygagne 2009).

Histopathology Histopathological examination of Graham-Little syndrome is consistent with lichen planopilaris (Fanti et al. 2018).

Pathogenesis The pathogenesis of Graham-Little syndrome is not fully understood. However, it is considered as autoimmune, lymphocyte-mediated disease (Kanti et. al 2018).

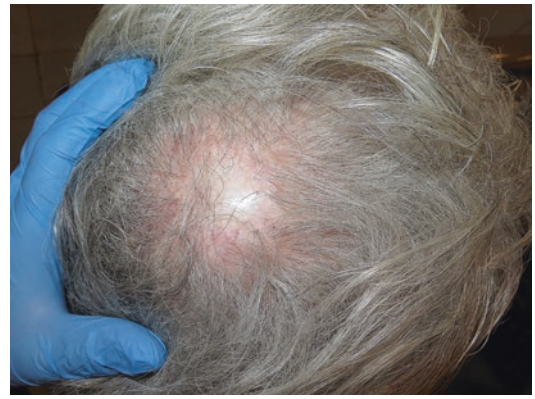


Fig. 35.26 Scarring alopecia in a patient with Graham-Little syndrome (photographed by Prof. Lidia Rudnicka)

Prognosis and Treatment The course of disease is slowly progressive and often chronic. Treatment of Graham-Little syndrome is often difficult and unsuccessful. Therapeutic options include topical or intralesional corticosteroids, systemic corticosteroids, retinoids, psoralen plus ultraviolet A (PUVA) phototherapy, and cyclosporine (Vashi et al. 2011).

Differential Diagnosis Classic lichen planopilaris, frontal fibrosing alopecia, lichen spinulosus, alopecia mucinosa, discoid lupus erythematosus, pseudopelade of Brocq, pityriasis rubra pilaris, sarcoidosis, psoriasis, parapsoriasis, and drug eruptions (Assouly and Reygagne 2009).

35.2.6 Discoid Lupus Erythematosus

Definition Discoid lupus erythematosus, a variant of chronic cutaneous lupus erythematosus, is a form of lymphocytic primary cicatricial alopecia (Kanti et al. 2018).

Clinical Features The disease is characterized by circumscribed erythematous indurated plaques with peripheral scaling and central scarring (Figs. 35.27 and 35.28). When the adherent scale is

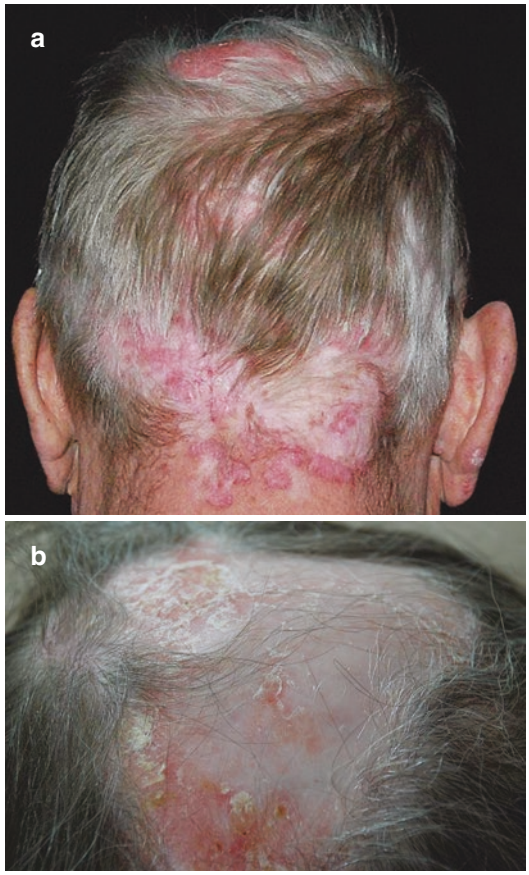


Fig. 35.27 (**a**, **b**) Discoid lupus erythematosus (photographed by Prof. Lidia Rudnicka)

removed, follicular plugging is typically observed (carpet tack sign). Telangiectasias, atrophy, depigmentation, and hyperpigmentation are other characteristic findings. Discoid lupus erythematosus occurs between 20 and 40 years of age. Women are more commonly affected than men (Kanti et al. 2018; Fanti et al. 2018).

Investigations The diagnosis of discoid lupus erythematosus is established based on clinical picture, a histopathological examination, and direct immunofluorescence test. Trichoscopy may be useful to avoid scalp biopsy. Antinuclear antibody (ANA) titers are positive in 15% to 45% of cases. Systemic lupus erythematosus should be excluded (Kanti et al. 2018; Fanti et al. 2018).

Histopathology In histopathology, follicular hyperkeratosis with a dense, partially perifollicular infiltrate of lymphocytes and histiocytes with hydropic degeneration of the basal layer are detected (Kanti et al. 2018).

Pathogenesis Discoid lupus erythematosus is an autoimmune condition that may occur as isolated cutaneous lesions or as a part of systemic lupus erythematosus (Fanti et. al 2018).

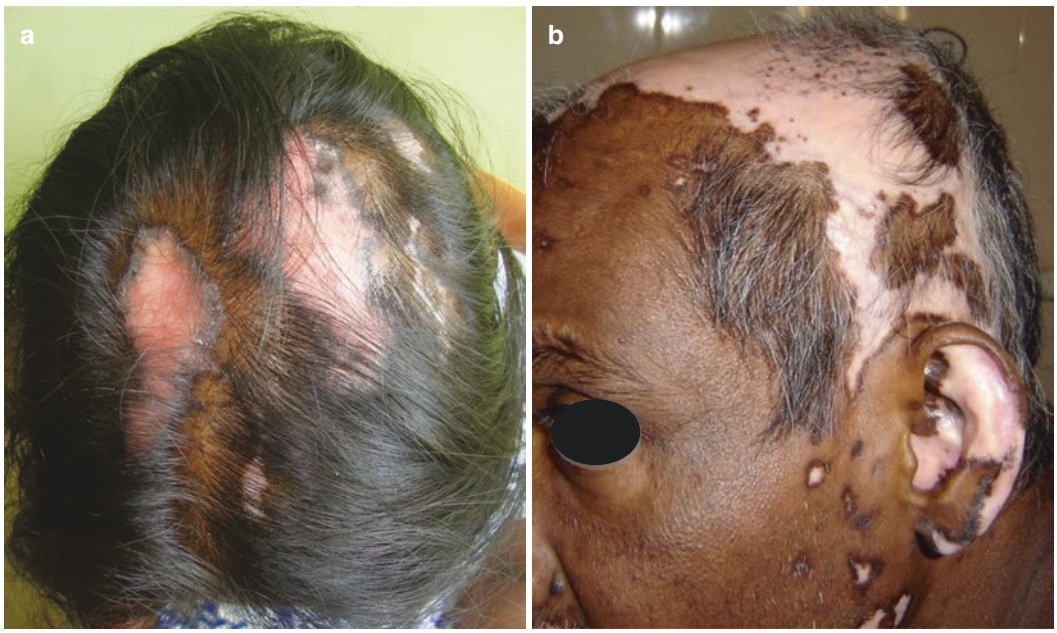


Fig. 35.28 (**a**, **b**) Discoid lupus erythematosus in a 34-year-old woman and a 45-year-old man (photographed by Dr Ranthilaka R. Ranawaka)

Prognosis and Treatment Photoprotection is paramount in treatment of discoid lupus erythematosus. Most commonly, topical or intralesional corticosteroids, antimalarials, and systemic corticosteroids are recommended. Other treatment options include retinoids, dapsone, thalidomide, methotrexate, azathioprine, and cyclosporine (Fanti et al. 2018; Kanti et al. 2018).

Differential Diagnosis Psoriasis, tinea capitis, folliculitis decalvans, lichen planopilaris, dissecting cellulitis (Fanti et al. 2018).

35.2.7 Pseudopelade of Brocq

Definition Pseudopelade of Brocq is a lymphocytic primary scarring alopecia (Kanti et al. 2018).

Clinical Features Clinically it presents with multiple small flash-toned alopecic areas with irregular borders without hyperkeratosis and inflammatory signs (sometimes reminiscent of "footprints in the snow"). The lesions are usually located on the vertex and parietal areas. The condition most commonly affects women between the ages of 30 and 50 (Kanti et al. 2018; Fanti et al. 2018).

Investigations The diagnosis of pseudopelade of Brocq is commonly established based on clinical picture and a histopatological examination. Trichoscopy may be useful to avoid scalp biopsy (Kanti et al. 2018).

Histopathology Histological examination shows non-specific changes consistent with the end stage of a cicatricial alopecia (Fanti et al. 2018).

Pathogenesis There is still no clear consensus whether pseudopelade of Brocq is a distinct entity or represents the end stage of any given cicatricial scalp disorder (Kanti et al. 2018, Fanti et al. 2018).

Prognosis and Treatment The disease is typically slowly progressive. In case of continued disease activity or progression, treatment options are the same as for lichen planopilaris. Spontaneous subsidence may occur (Fanti et al. 2018; Kanti et al. 2018).

Differential Diagnosis Psoriasis, tinea capitis, folliculitis decalvans, lichen planopilaris, dissecting cellulitis (Fanti et al. 2018).

35.2.8 Central Centrifugal Cicatricial Alopecia

Definition: Central centrifugal cicatricial alopecia is a form of primary lymphocytic cicatricial alopecia (Messenger et al. 2016; Herskovitz and Miteva 2016).

Clinical features: The disease is characterized by a single area of cicatricial alopecia in the vertex region with slow, often symmetric peripheral progression. Perifollicular hyperpigmentation and/or erythema, polytrichia and islands of unaffected skin with the affected area are observed (Figs. 35.29 and 35.30). Hypersensitivity, pruritus, and/or a burning sensation can be reported. Central centrifugal cicatricial alopecia mainly affects women of African ethnicity. The female to male ratio is approximately 3:1. (Messenger et al. 2016; Herskovitz and Miteva 2016).

Investigations: The diagnosis of central centrifugal cicatricial alopecia is commonly established based on clinical picture and histopathological examination. Trichoscopy may be useful to avoid scalp biopsy (Messenger et al. 2016; Herskovitz and Miteva 2016).

Histopathology: A lymphocytic infiltrate and premature loss of the inner root sheath (Messenger et al. 2016; Herskovitz and Miteva 2016).

Pathogenesis: The pathogenesis of central centrifugal cicatricial alopecia is unknown. Various

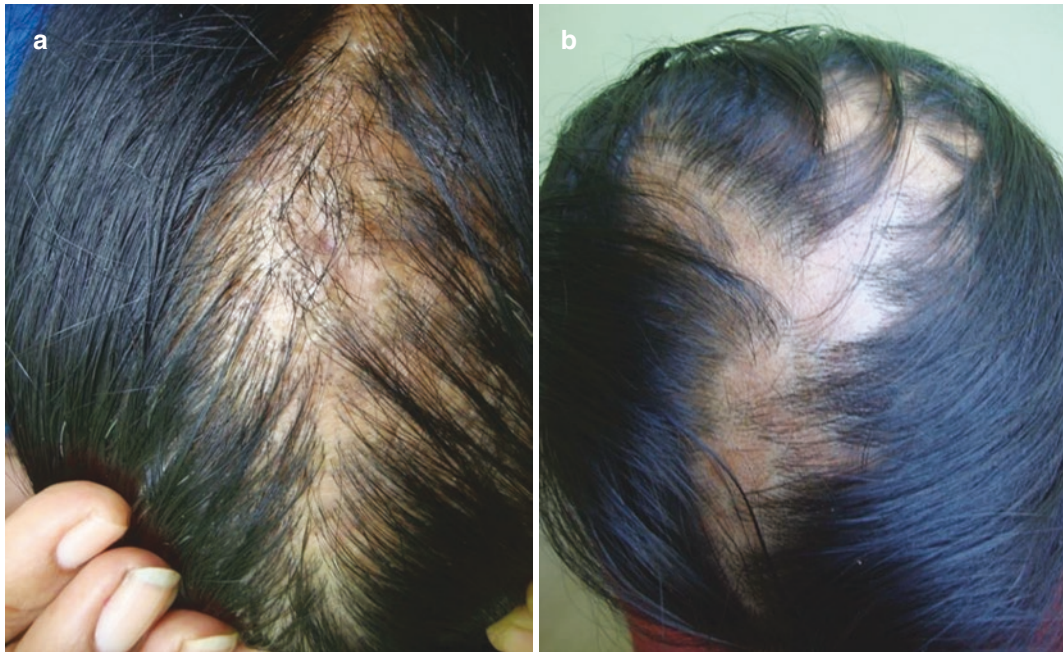


Fig. 35.29 (**a, b**) Central centrifugal cicatricial alopecia in two women aged 25 and 29 years (photographed by Dr Ranthilaka R. Ranawaka)

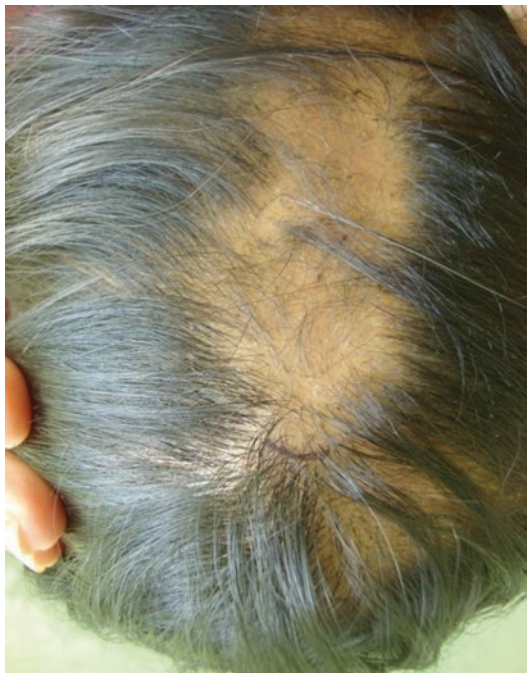


Fig. 35.30 Central centrifugal cicatricial alopecia in a 49-year-old woman (photographed by Dr Ranthilaka R. Ranawaka)

etiological factors (genetic predisposition, use of chemical straighteners, traction hairstyles, bacterial or fungal infections of the scalp) have been suggested (Messenger et al. 2016; Herskovitz and Miteva 2016).

Prognosis and Management: Spontaneous remission after a few years is usually reported. Topical and intralesional corticosteroids may be effectivw. Other therapeutic options include systemic antibiotics (tetracyclines), systemic corticosteroids, antimalarials, mycophenolate mofetil, and cyclosporine A (Messenger et al. 2016; Herskovitz and Miteva 2016).

Differential diagnosis: traction alopecia, trichotillomania, androgenic alopecia, other forms of cicatricial alopecia (Messenger et al. 2016; Herskovitz and Miteva 2016).

35.2.9 Folliculitis Decalvans

Definition Folliculitis decalvans is a primary form of cicatricial alopecia with a neutrophilic infiltrate (Fanti et al. 2018).

Clinical Features Folliculitis decalvans is initially characterized by follicular papules and pustules, especially in the vertex and occipital region (Fig. 35.31). The beard region and other body hairs (neck, axillae, and pubic region) are less frequently affected. It typically occurs in young to middle-aged adults. Men are more commonly affected than women (Fanti et al. 2018, Kanti et al. 2018).

Investigations The diagnosis of folliculitis decalvans is established based on clinical picture and a histopathological examination. Trichoscopy may be useful to avoid scalp biopsy (Kanti et al. 2018).

Histopathology Dilatation of the follicular ostium with intra- and perifollicular neutrophilic infiltrate is mainly observed. Follicular tufts may be detected (Kanti et al. 2018).

Pathogenesis The role of *Staphylococcus aureus* antigens in pathogenesis of folliculitis decalvans has been suggested (Fanti et al. 2018).

Prognosis and Treatment First-line treatment consists of topical (mupirocin, fusidic acid, erythromycin) or systemic antibiotics (tetracyclines, first generationcefalosporines, combination of rifampicin with clindamycin), topical antiseptic agents with or without topical or intralesional corticosteroids. Isotretinoin can also be useful. To prevent recurrences, all tufted hair follicles can be surgically removed if feasible (Fanti et al. 2018; Kanti et al. 2018).

Differential Diagnosis Bacterial folliculitis, tinea capitis, dissecting cellulitis, erosive pustular dermatosis of the scalp, lichen planopilaris, central centrifugal cicatricial alopecia (Fanti et al. 2018).

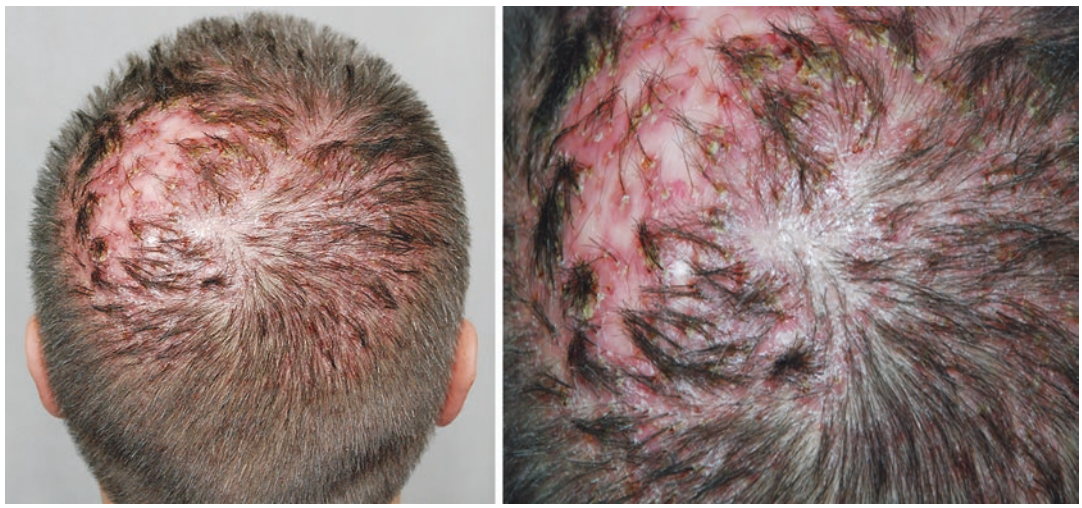


Fig. 35.31 Folliculitis decalvans (photographed by Dr Mariusz Sikora, MD, PhD, Dermatologist, Department of Dermatology and Venereology y, Medical University of Warsaw, Poland)

35.2.10 Dissecting Cellulitis

Definition Dissecting cellulitis, or perifolliculitis capitis abscedens et suffodiens, is a neutrophilic scarring hair loss. It is a part of the follicular inclusion triad along with hidradenitis suppurativa and acne conglobata (Fanti et al. 2018; Kanti et al. 2018).

Clinical Features Clinically patients present with multiple firm, dome-shaped violaceous papules in the vertex and occipital region, which may coalesce to form plaques and nodules and eventually abscesses and sinus tracts (Figs. 35.32 and 35.33). Aspiration yields pus, serous or hemorrhagic fluid. Bacterial and fungal cultures are usually negative. The condition leads to patchy alopecia and ultimately atrophy or hypertrophic scars. The vertex and occipital area are mainly affected. The disease predominantly occurs in African American men of 20–40 years of age (Fanti et al. 2018; Kanti et al. 2018).

Investigations The diagnosis of folliculitis decalvans is established based on clinical picture and a histopathological examination. Trichoscopy may be useful to avoid scalp biopsy. Bacterial and fungal cultures are usually negative (Kanti et al. 2018).

Histopathology Histopathological examination shows a lymphocytic infiltrate in the lower dermis extending into the subcutis. Abscesses consisting of neutrophils, lymphocytes, and plasma cells as well as granulomatous foreign body reactions are observed (Kanti et al. 2018; Fanti et al. 2018).

Pathogenesis The precise pathogenesis of dissecting cellulitis is unknown; however, the role of hyperkeratosis, follicular occlusion, and subsequent inflammation has been described (Kanti et al. 2018).

Prognosis and Treatment The treatment options include isotretinoin, systemic corticosteroids, antibiotics (ciprofloxacin, clindamycin, rifampin, and trimethoprim/sulfamethoxazole), and tumor necrosis factor (TNF) inhibitors (Kanti et al. 2018).

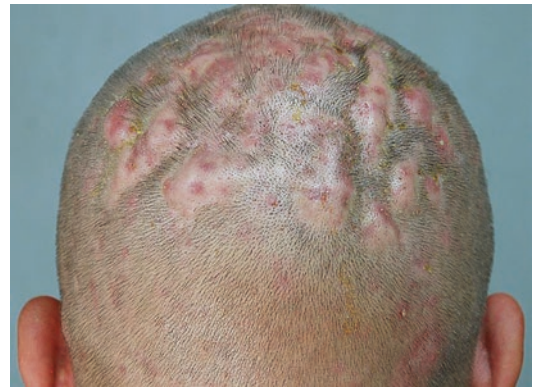


Fig. 35.32 Dissecting cellulitis (photographed by Dr Anna Waśkiel-Burnat)

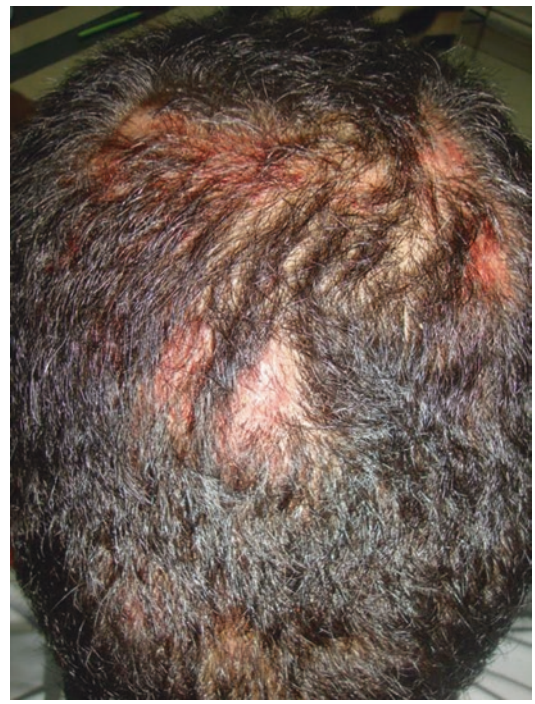


Fig. 35.33 Dissecting cellulitis (photographed by Dr Ranthilaka R. Ranawaka)

Differential Diagnosis Folliculitis decalvans, tinea capitis, cutis verticis gyrata, epidermal cysts, alopecic and aseptic nodules of the scalp (Fanti et al. 2018).

35.3 Abnormalities of Hair Shaft

Netherton Syndrome
Definition: Netherton syndrome is a rare, hereditary disorder of cornification (Saleem et al. 2018).

Clinical features: The disease consists of the triad of features: congenital ichthyosiform erythroderma, trichorrhexis invaginata, and allergic manifestations with elevated serum levels of immunoglobulin (Saleem et al. 2018) (Fig. 35.34).

Investigations: Diagnosis of Netherton syndrome is usually based on clinical picture and hairs examination. Histopathological examination can be useful. To confirm diagnosis genetic examination is required (Saleem et al. 2018).

Histopathology: In histopathological examination psoriasiform hyperplasia, with skin inflammation, thinning, and a diminished granular layer are detected (Saleem et al. 2018).

Pathogenesis: Netherton syndrome is autosomal recessive disorder caused by mutations in the serine protease inhibitor of Kazal type 5 gene (Saleem et al. 2018).

Prognosis and treatment: Symptoms of Netherton syndrome tend to improve with age. Increased risk of skin cancer is observed.

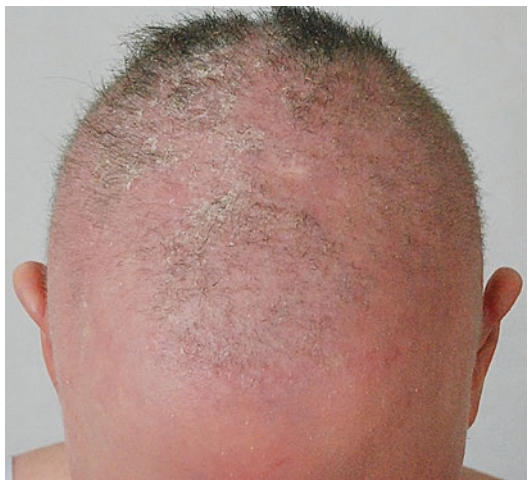


Fig. 35.34 Netherton syndrome (photographed by Dr Anna Waśkiel-Burnat)

Treatment modalities are emolients, topical corticosteroids, phototherapy and retinoids (Saleem et al. 2018).

Differential diagnosis: autosomal recessive congenital ichthyosis, erythrodermic psoriasis, atopic dermatitis, acrodermatitis enteropathica, primary immunodeficiency syndromes, peeling skin syndrome type B, seborrheic dermatitis, exfoliative ichthyosis (Saleem et al. 2018).

35.4 Excessive Growth of Hair

35.4.1 Hypertrichosis

Hypertrichosis is defined as excessive hair growth anywhere on the body that is not androgen dependent. Hypertrichosis is classified as localized or generalized, and congenital or acquired (Figs. 35.35, 35.36, 35.37, 35.38, and 35.39).

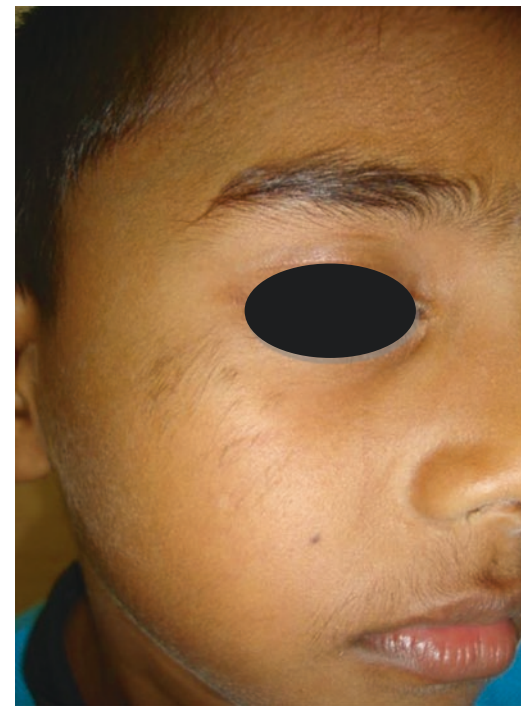


Fig. 35.35 Congenital localized hypertrichosis since birth in a 9-year-old boy (photographed by Dr Ranthilaka R. Ranawaka)

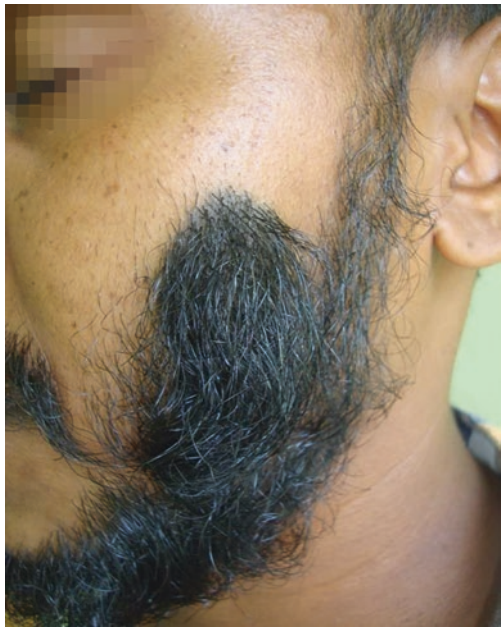


Fig. 35.36 Nevoid hypertrichosis in a 29-year-old man (photographed by Dr Ranthilaka R. Ranawaka). Nevoid hypertrichosis refers to a localized well-circumscribed area of hypertrichosis. It can occur in a hair-bearing site such as the scalp with a localized area of marked increased density of terminal hair fibers or in areas normally lacking terminal hairs

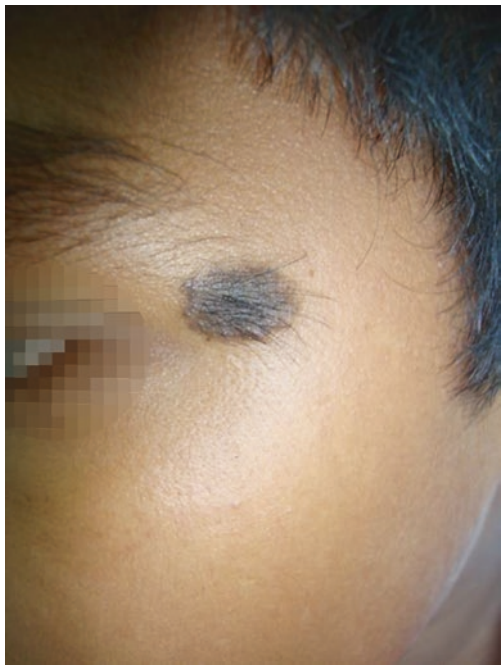


Fig. 35.37 Congenital melanocytic nevi with hypertrichosis (photographed by Dr Ranthilaka R. Ranawaka)

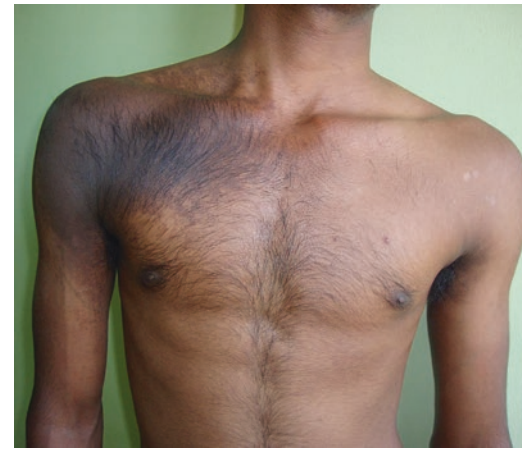


Fig. 35.38 Becker nevus with localized hypertrichosis (photographed by Dr Ranthilaka R. Ranawaka)

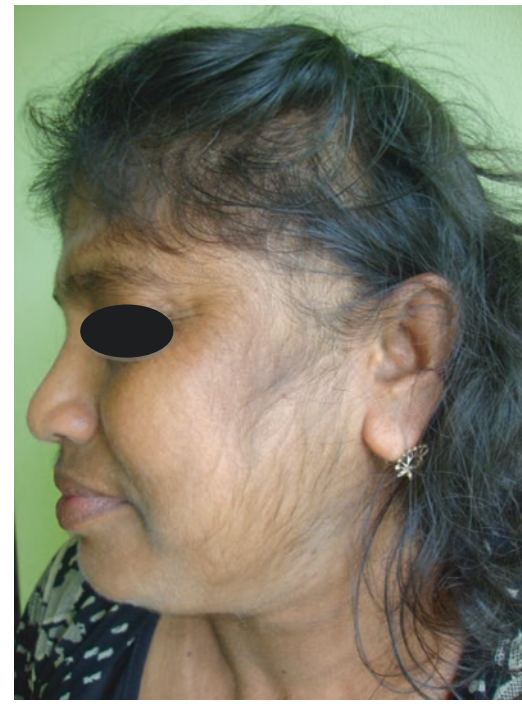


Fig. 35.39 Acquired hypertrichosis on the forehead and the face following 5% minoxidil lotion applied daily for 1 month to the occipital scalp (photographed by Dr Ranthilaka R. Ranawaka)

35.4.2 Hirsutism

Hirsutism is the presence of excess terminal hair in women inandrogen-dependent sites (a male distribution pattern) (Figs. 35.40, 35.41 and 35.42) It is associated with increased level of circulating androgens (whose sourceis primarily the ovary or adrenal gland e.g. polycystic ovary syndrome) or an enhanced end-organ response to androgens.

35.5 Miscellaneous Conditions

35.5.1 Disseminate and Recurrent Infundibulofolliculitis

Definition: Disseminate and Recurrent Infundibulofolliculitis is a rare itchy follicular skin condition (Rekha et al. 2019; Hay 2016).

Clinical features: The disease is characterized by the presence of asymptomatic skin-colored, monomorphic, follicular papules usually distrib-

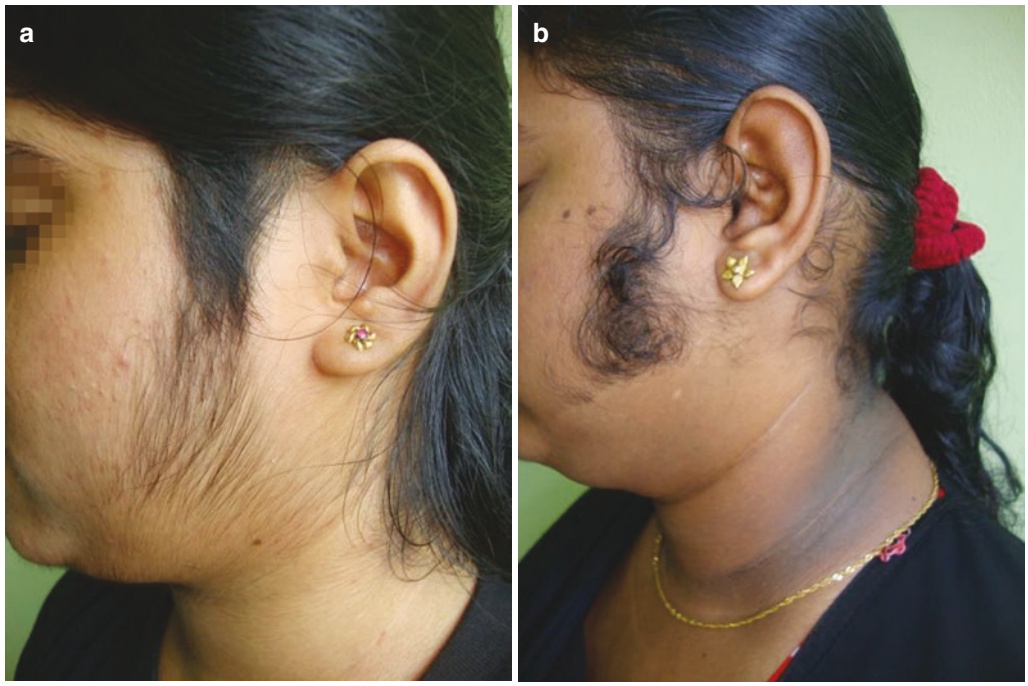


Fig. 35.40 (**a**, **b**) Two women with hirsutism aged 21 and 19 years. Both had other features of PCOS: obesity, acne, seborrhea, acanthosis nigricans, and glucose intolerance (photographed by Dr Ranthilaka R. Ranawaka)

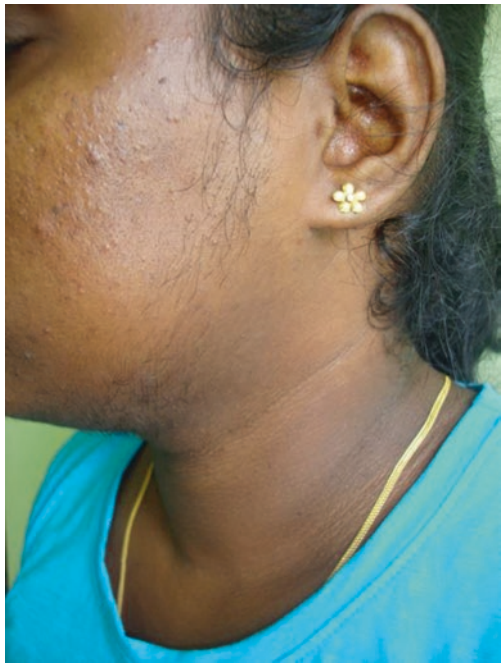


Fig. 35.41 An 18-year-old woman with hirsutism. She also had other features of polycystic ovary disease: obesity, acanthosis nigricans, acne, menstrual irregularities, and impaired blood glucose (photographed by Dr Ranthilaka R. Ranawaka)

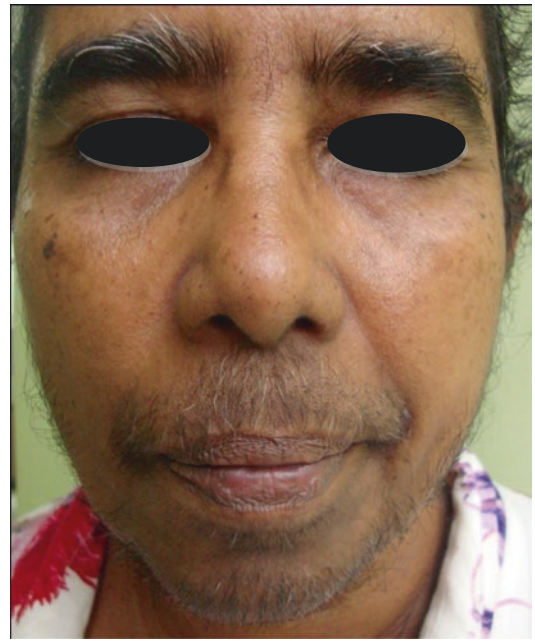


Fig. 35.42 A 54-year-old woman with hirsutism (photographed by Dr Ranthilaka R. Ranawaka)

uted on the trunk and extremities. It is most commonly affects young, dark-skinned individuals (Rekha et al. 2019; Hay 2016).

Investigations: Diagnosis of disseminate and recurrent infundibulofolliculitis is based on clinical picture and a histopathologic examination (Rekha et al. 2019).

Histopathology: Typical histopathologic feature of disseminate and recurrent infundibulofolliculitis is perifollicular mononuclear infiltrate concentrated around the infundibular part of the follicle (Rekha et al. 2019).

Pathogenesis: The pathogenesis of the disease is unknown (Rekha et al. 2019).

Prognosis and treatment: The disease is self-limited. Treatment options are topical corticosteroids, tretinoin, oral vitamin A, isotretinoin, and phototherapy (Rekha et al. 2019) (Figs. 35.43 and 35.44).

Differential diagnosis: keratosis pilaris, lichen spinulosus (Rekha et al. 2019).

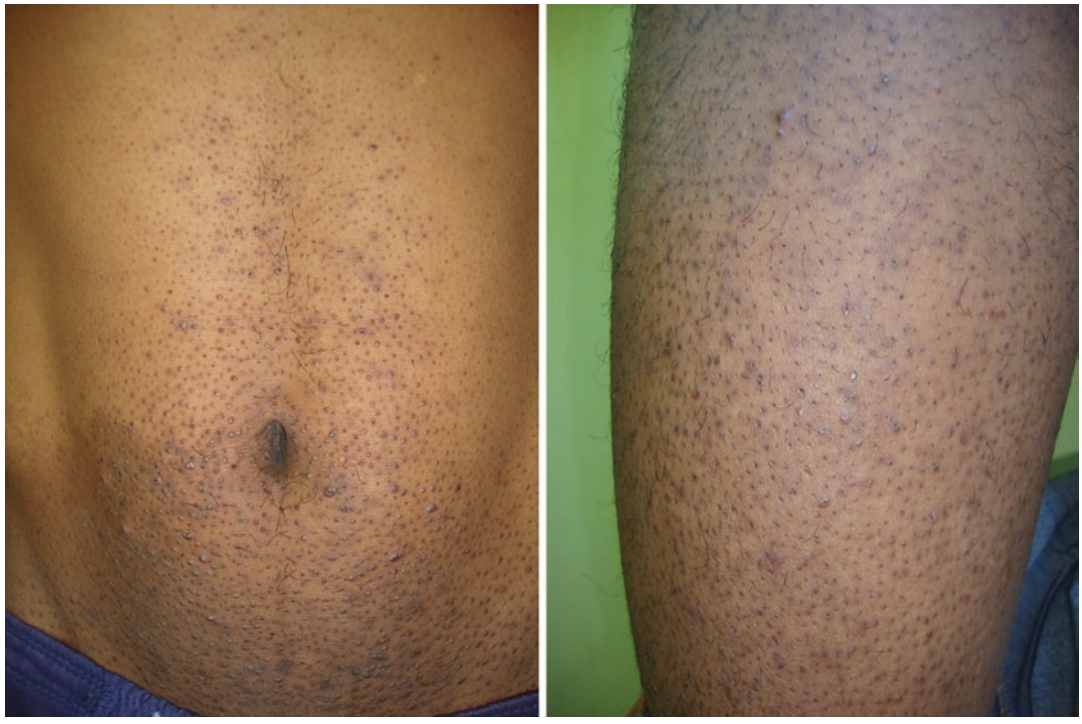


Fig. 35.43 Disseminate and recurrent infundibulofolliculitis in a 19-year-old boy (photographed by Dr Ranthilaka R. Ranawaka)

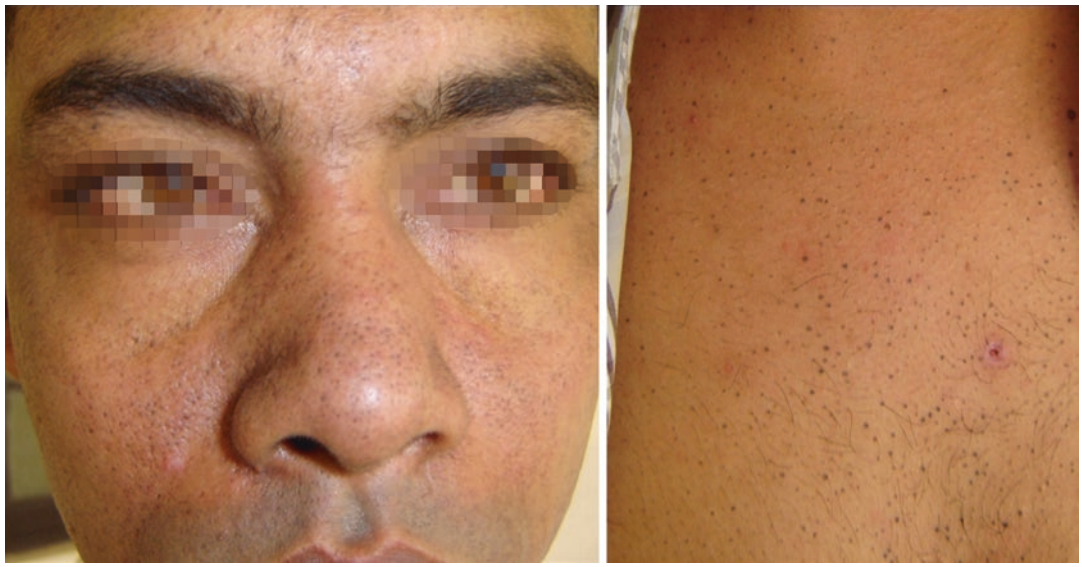


Fig. 35.44 This 29-year-old man gets inflammation around the hair follicles permanently destroying the hair follicles. Histopathology showed perifollicular inflammatory cell infiltrate (photographed by Dr Ranthilaka R. Ranawaka)

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