Folliculitis Decalvans and Dissecting Cellulitis

18

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Contents

18.1 Etiology

Folliculitis decalvans (FD) is an inflammatory, cicatricial scalp disorder in which the cellular infiltrate is primarily neutrophilic [1]. The most widely accepted theory for the pathogenesis for FD postulates that *Staphylococcus aureus* secretes cytotoxins or superantigens that bind to major histocompatibility complex (MHC) class II molecules [2, 3]. This stimulates T-lymphocytes, leading to release of proinflammatory mediators (IFN-gamma and TNF-alpha) and pro-fibrotic mediators (TGF-beta and IL-4) [4]. An abnormal host immune response is thought to be responsible for allowing this cascade of events to occur. *S. aureus* is usually isolated from mostly all cases of untreated folliculitis decalvans. *S. aureus* is present in 20–30 % of the general population, but less than 0.05 % of these normal carriers suffer from infection.

Dissecting cellulitis or dissecting folliculitis is a chronic, inflammatory scalp disorder also known as perifolliculitis capitis abscedens et suffodiens or Hoffmann disease. It can occur with hidradenitis suppurativa, pilonidal cysts, and acne conglobata as part of the follicular occlusion tetrad. Coexistence with acne conglobata and hidradenitis occur in one-third of patients. These disorders are felt to share a common pathogenesis. Occlusion of the follicles with keratinous and sebaceous material occurs. The follicles then rupture and lead to a neutrophilic inflammatory reaction.

18.2 Epidemiology

Although sufficient epidemiologic studies are not available, FD accounts for approximately 10–11 % of all cases of primary cicatricial alopecias. This disorder occurs in young adults, affecting men more than women and African Americans more than Caucasians [3, 5]. There may be a genetic predisposition, as there are a number of familial cases reported. Dissecting cellulitis usually affects black males in the second to fourth decades of life; however, 10 % of cases have been in white males and more rarely females.

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D. Jackson-Richards, A.G. Pandya (eds.), *Dermatology Atlas for Skin of Color*,
DOI 10.1007/978-3-642-54446-0_18, © Springer-Verlag Berlin Heidelberg 2014

18.3 Clinical Features

Folliculitis decalvans first involves the vertex and occipital scalp, presenting with erythematous perifollicular papules and yellow scale at follicular openings (Figs. 18.1 and 18.2). Pustules, erosions, and hemorrhagic crusts then develop and, finally, scarring alopecia. Tufted folliculitis is seen with multiple hairs emerging from a single follicular orifice (Fig. 18.3). Although tufting is commonly seen in folliculitis decalvans, it is also seen in other cicatricial alopecias. Patients often experience pruritus, pain, and disfiguring alopecia. The course of FD is chronic and treatment is difficult.

Dissecting cellulitis affects the vertex and occipital scalp beginning as perifollicular pustules. This folliculitis progresses into painful nodules and deep-seated abscesses that are interconnected by sinus tracts (Figs. 18.4, 18.5, and 18.6). The formation of sinus tracts are a hallmark finding of dissecting cellulitis and distinguishes it from folliculitis decalvans (Figs. 18.7 and 18.8). Secondary bacterial infection with *S. aureus* is common. This progressive inflammation leads to marked, scarring alopecia. Dissecting cellulitis is a painful chronically relapsing condition that's cosmetically disfiguring. When part of the follicular occlusion tetrad, there is a 30 % risk of HLA-B27 negative spondyloarthropathy associated with SAPHO (synovitis, acne, palmoplantar pustolosis, hyperostosis, osteitis) syndrome.

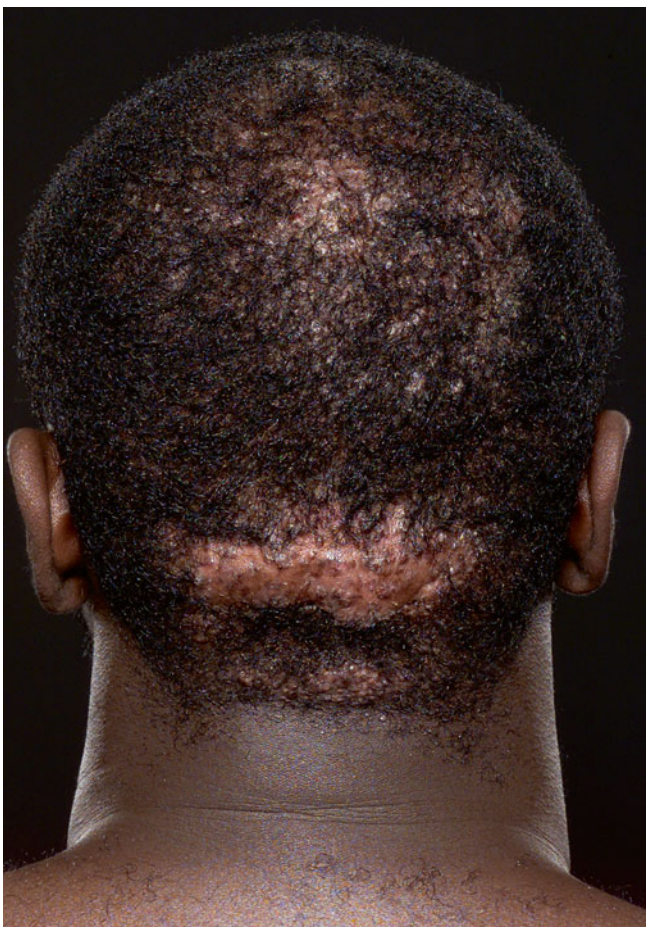

Fig. 18.1 Folliculitis decalvans of scalp along with acne keloidalis nuchae on occiput in an African American male

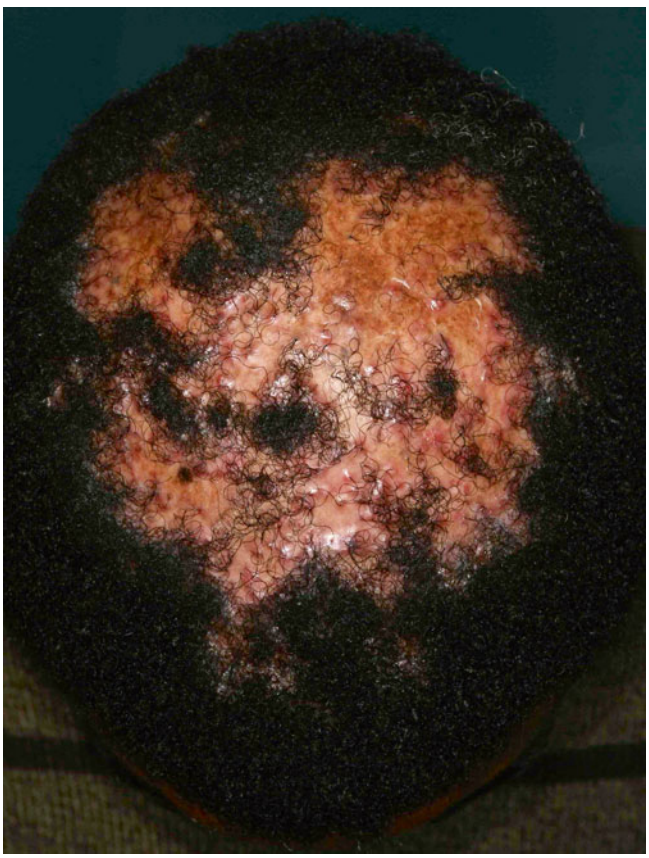

Fig. 18.2 Folliculitis decalvans in an African male (Courtesy of Service de Dermatologie, APHP Hospital Saint-Louis, Paris, France)

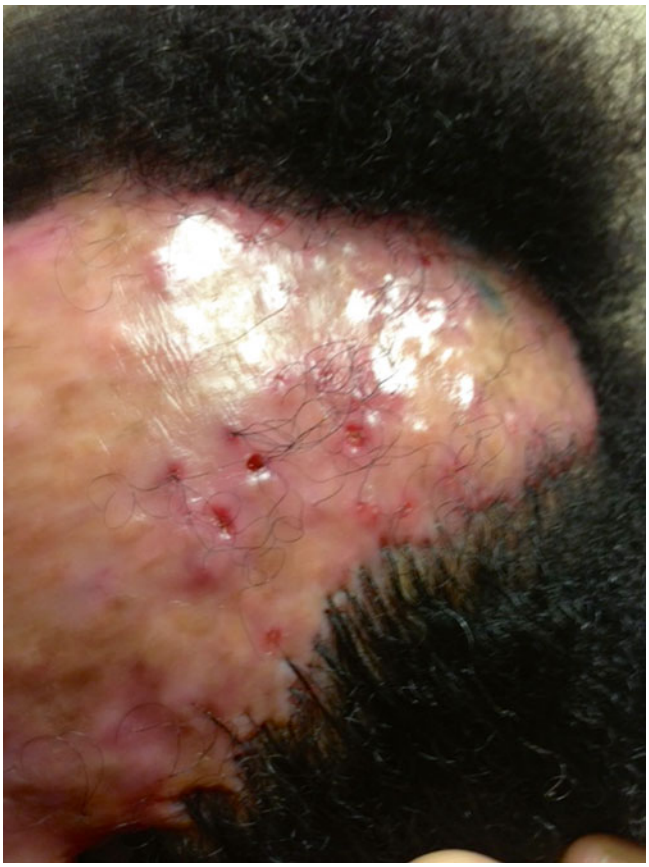

Fig. 18.3 Severe folliculitis decalvans keloidal scarring and tufted folliculitis in an African American male

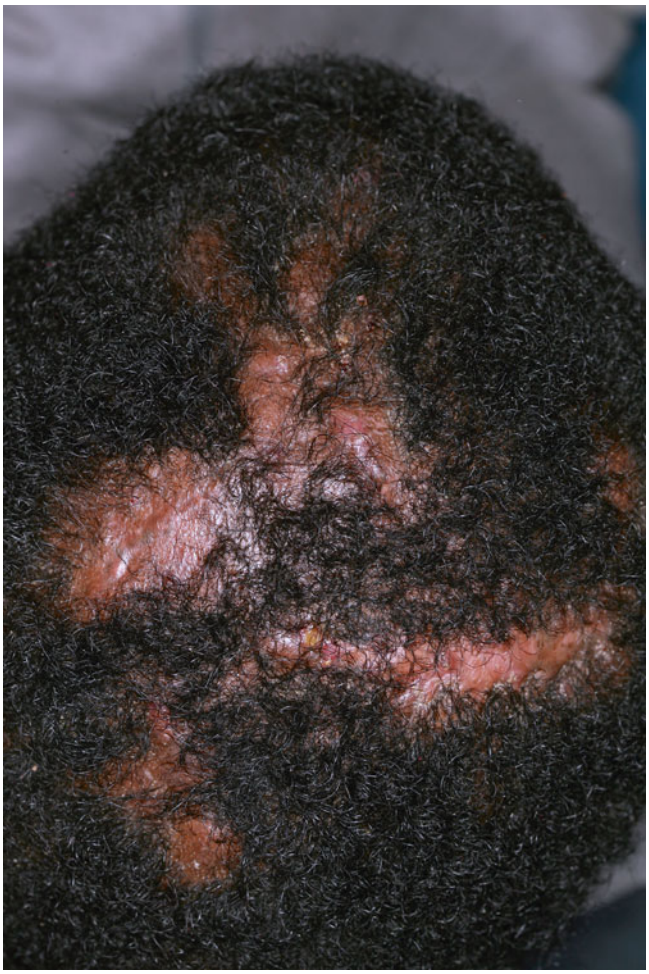

Fig. 18.4 Dissecting cellulitis in an African male (Courtesy of Service de Dermatologie, APHP Hospital Saint-Louis, Paris, France)

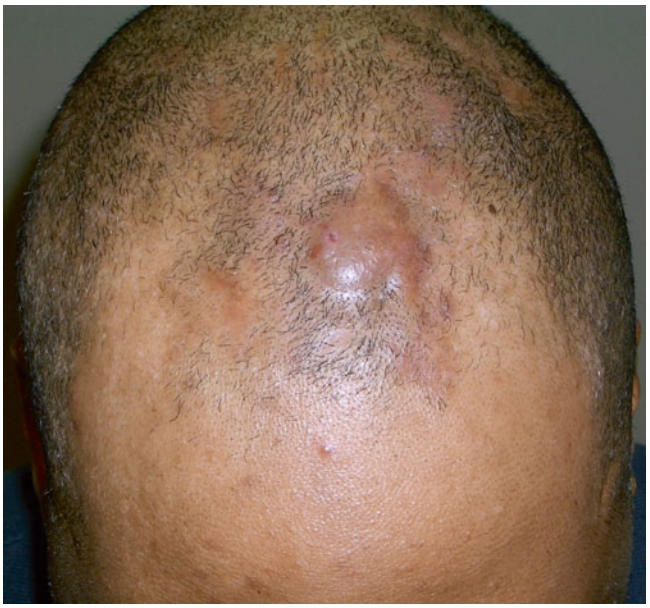

Fig. 18.5 Dissecting cellulitis in an African American male (Courtesy of Dr. Iltefat Hamzavi)

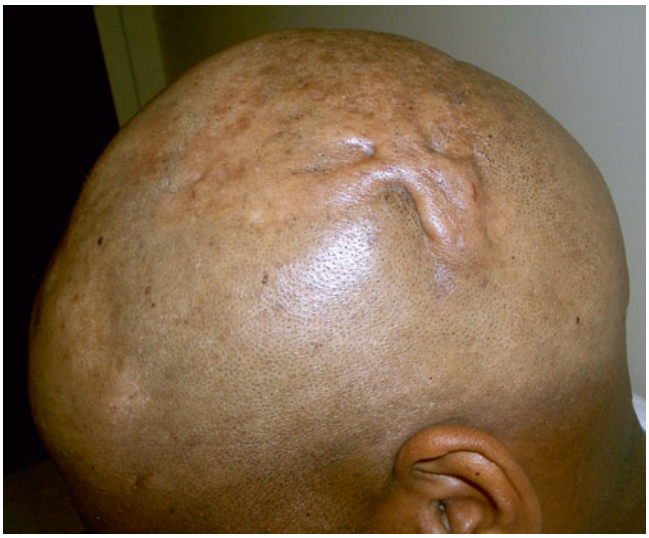

Fig. 18.7 Dissecting cellulitis with sinus tracts in an African American male (Courtesy of Dr. Iltefat Hamzavi)

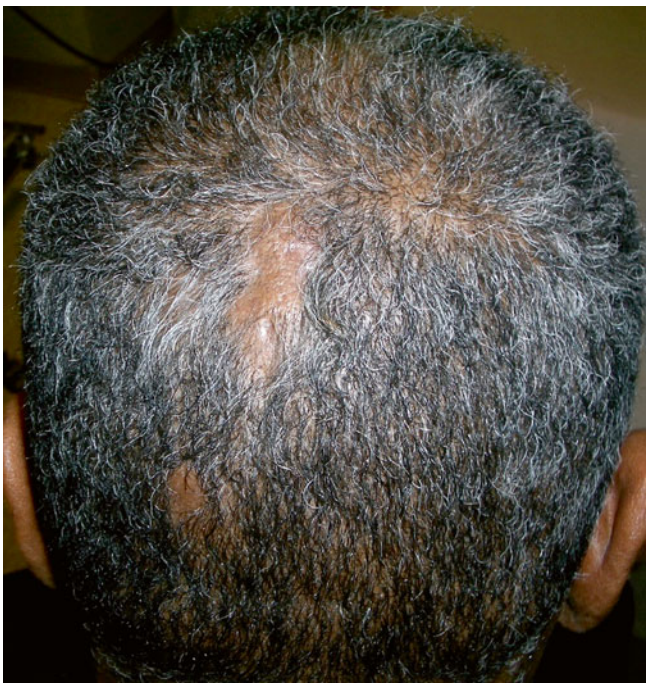

Fig. 18.6 Dissecting cellulitis in a Middle Eastern male (Courtesy of Dr. Iltefat Hamzavi)

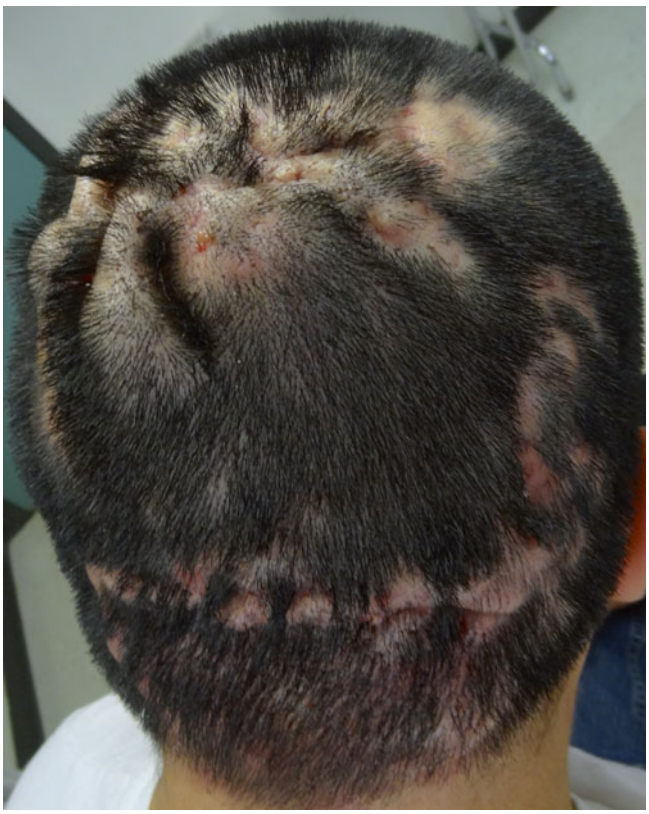

Fig. 18.8 Dissecting cellulitis with sinus tracts in a Hispanic male

18.4 Histopathologic Features

Histopathology of early lesions of FD shows keratin aggregation and a dilated infundibulum with an intrafollicular neutrophilic infiltrate [3, 5]. Even in early lesions there is a loss of sebaceous glands. More advanced lesions show a mixed infiltrate of neutrophils, lymphocytes, and plasma cells. Granulomatous inflammation with foreign body giant cells is seen around ectopic hair shafts. Follicular tufts with multiple hairs merging into one infundibulum are often seen. End-stage lesions exhibit fibrosis. Biopsies of dissecting cellulitis show distension of the follicular infundibula with a perifollicular neutrophilic infiltrate of the upper and middle portion of the hair follicle. Follicular rupture occurs and the infiltrate becomes a mixed pattern that includes lymphocytes, histiocytes, and plasma cells. Abscesses form in the adventitial dermis. Late-stage disease shows sinus tracts that are partially lined by squamous epithelium and dense fibrous scarring replacing follicular units.

18.5 Differential Diagnosis

Differential diagnosis of FD includes acne keloidalis nuchae, which affects males, especially African American males, disproportionately. Both disorders can have similar clinical and histopathologic features. Lichen planopilaris, discoid lupus, pseudopelade of Brocq, and central centrifugal cicatricial alopecia are lymphocytic primary cicatricial alopecias. Pustules are not the primary lesion in those disorders unless there is secondary bacterial infection. Tinea capitis can cause hair loss and pustules can be seen in inflammatory cases. Fungal cultures to rule out a dermatophyte infection such as kerion are recommended in all cases. Differential diagnosis of dissecting cellulitis includes folliculitis decalvans and acne keloidalis nuchae as well. These conditions may demonstrate perifollicular pustules and scarring alopecia; however, large abscesses and sinus tracts are usually not seen. Folliculotropic mycosis fungoides can sometimes resemble dissecting cellulitis and can be ruled out with a skin biopsy.

18.6 Treatment

Folliculitis decalvans often affects individuals for years and treatment is difficult. Topical treatment alone is not sufficient. Various oral antibiotics are used but relapse is common. Oral doxycycline, minocycline, clarithromycin, sulfamethoxazole-trimethoprim, and fusidic acid have been used alone and in combination. Rifampin is a very good antistaphylococcal agent, but resistance often occurs when used alone. A combination of rifampin 300 mg twice daily and clindamycin 300 mg twice daily has been used successfully and has reportedly given the best outcomes, with remission for months to years after treatment. Patients must be monitored for hepatitis, oral contraceptive failure, interaction with warfarin, hemolytic anemia, and thrombocytopenia when using rifampin. Pseudomembranous colitis is a side effect of clindamycin. Topical antibiotics such as mupirocin, clindamycin, fusidic acid, and erythromycin have also been used with oral antibiotics [2, 3, 6]. Intralesional, topical, and oral corticosteroids are used to reduce inflammation when used with oral antibiotics. Other reported treatments include dapsone, isotretinoin, topical tacrolimus [7], and ND-YAG laser [8].

Dissecting cellulitis is chronic and recalcitrant to a number of treatments and multiple modalities are usually necessary [9]. Oral antibiotics such as doxycycline and ciprofloxacin have been used, but relapse is common [10]. Concomitant use of topical antibiotics and intralesional and oral corticosteroids is useful [11]. Large abscesses should be incised and drained. Scerri et al. have reported long-standing remission with oral isotretinoin, 1 mg/kg/day for 4 months followed by a maintenance dosage of 0.75 mg/kg/day for 5–7 months [12]. Adalimumab has been used successfully to control dissecting cellulitis in a number of reported cases [13]. When medical treatment fails, follicular destruction with radiation, lasers, or surgical resection with skin grafting may be necessary.

References

1. Olsen EA, Bergfeld WF, Cotsarelis G, et al. Workshop on Cicatricial Alopecia. Summary of North American Hair Research Society (NAHRS)-sponsored Workshop on Cicatricial Alopecia. Duke University Medical Center, Feb 10 and 11, 2001. J Am Acad Dermatol. 2003;48:103–10.
2. Powell JJ, Dawber RP, Gatter K. Folliculitis decalvans including tufted folliculitis: clinical, histological and therapeutic findings. Br J Dermatol. 1999;140:328–33.
3. Whiting DA. Cicatricial alopecia: clinico-pathological findings and treatment. Clin Dermatol. 2001;19:211–5.
4. Chiarini C, Torchia D, Bianchi B, Volpi W, Caproni M, Fabbri P. Immunopathogenesis of folliculitis decalvans. Am J Clin Pathol. 2008;130:526–34.
5. Tan E, Martinka M, Ball N, Shapiro J. Primary cicatricial alopecias: clinicopathology of 112 cases. J Am Acad Dermatol. 2004;50:25–32.
6. Otberg N, Kang H, Alzolibani A, Shapiro J. Folliculitis decalvans. Dermatol Ther. 2008;21:238–44.
7. Bastida J, Valeron-Almazon P, Santana-Molina N, Medina-Gil C, Carretero-Hernandez G. Treatment of folliculitis decalvans with tacrolimus ointment. Int J Dermatol. 2012;51:216–20.
8. Parlette E, Kroeger N, Ross V. Nd:YAG laser treatment of recalcitrant folliculitis decalvans. Dermatol Surg. 2004;30:1152–4.
9. Mundi JP, Marmon S, Fischer M, Kamino H, Patel R, Shapiro J. Dissecting cellulitis of the scalp. Dermatol Online J. 2012;18(12):8.
10. Otberg N. Primary cicatricial alopecias. Dermatol Clin. 2013;31(1):155–66. W.B. Saunders.
11. Somani N, Bergfeld WF. Cicatricial alopecia: classification and histopathology. Dermatol Ther. 2008;18:12.
12. Scerri L, et al. Dissecting cellulitis of the scalp: response to isotretinoin. Br J Dermatol. 1996;134:1105.
13. Sukhatme SV, Lenzy YM, Gottlieb AB. Refractory dissecting cellulitis of the scalp treated with adalimumab. J Drugs Dermatol. 2008;7(10):981–3.