FEATURE ARTICLE

CE ANCC
1.5 Contact Hours | 0.5 Pharmacology Contact Hour

A Case of Folliculitis Decalvans With Concomitant Acne Keloidalis Nuchae, Androgenic Alopecia, and Profound Postinflammatory Hyperpigmentation

Mitchell Gleed, Kim Carlson

ABSTRACT: Folliculitis decalvans is a cicatricial alopecia of the parietal scalp and vertex characterized by erythematous, scarred, confluent patches of alopecia with scattered peripheral pustules and scale. It is most common among middle-aged men and is frequently associated with acne keloidalis nuchae. The pathogenesis of folliculitis decalvans is not completely understood, but it likely involves an inappropriate inflammatory response to components of *Staphylococcus aureus*. Folliculitis decalvans is a chronic disease characterized by periods of remission and exacerbation. Patients with long-standing, undertreated disease can experience severe hair loss and postinflammatory hyperpigmentation. Treatment is focused on reducing inflammation and bacterial load using oral antibiotic therapy. Early recognition and treatment is paramount to alleviate symptoms and limit irreversible hair loss.

Key words: Acne Keloidalis Nuchae, Alopecia, Folliculitis Decalvans, Postinflammatory Hyperpigmentation, Scarring Alopecia

Mitchell Gleed, BS, *Arizona College of Osteopathic Medicine, Midwestern University, Glendale, AZ.*
Kim Carlson, RN, MSN, NP-C, DCNP, *MacKenzie Dermatology, Prescott, AZ.*

The authors declare no conflict of interest.

Correspondence concerning this article should be addressed to Mitchell Gleed, BS, 1304 S. 105th Pl. Apt. 1015, Mesa, AZ 85209. E-mail: mgleed66@midwestern.edu

DOI: 10.1097/JDN.0000000000000467

CASE REPORT

History of Present Illness

A 37-year-old African American man presented to the outpatient dermatology clinic complaining of pruritic lesions of the scalp and associated hair loss that had been worsening over the past month. He described his scalp lesions as extremely itchy, mildly painful, and dry; they also frequently would bleed when scratched.

In 2004, the patient began to develop erythematous papules and pustules on his scalp after regularly sharing a hard hat with a coworker. At that time, he was diagnosed with superficial bacterial folliculitis and was treated with a topical antibiotic, which temporarily alleviated his symptoms. The patient stated, however, that the lesions returned quickly and persisted since that time, although the severity waxed and waned.

Past medical history was significant for androgenic alopecia and acne keloidalis nuchae, which he had treated with intralesional corticosteroids and topical betamethasone valerate with mild improvement. He denied any personal history of chronic disease such as diabetes, hypertension, kidney disease, autoimmune disorders, HIV/AIDS, or skin disorders. Family history was positive for androgenic alopecia in his grandfather and was otherwise negative for chronic skin conditions, autoimmune disease, cancer, or other types of alopecia. At the time of presentation, the patient stated he was employed in the construction field, did not smoke, did not use intravenous drugs, and drank alcohol infrequently. He denied current or recent use of any medications or supplements.

Physical Examination

Physical examination demonstrated crops of pustules and follicles with scale on a background of erythema localized to the posterior vertex and superior parietal scalp bilaterally (Figure 1). Patchy hair loss was evident in these regions, as well as ill-defined, asymmetric, homogeneous blue macules scattered throughout the inflamed areas. On dermoscopy, tufted hairs, follicular hyperkeratosis, perifollicular erythema, cicatricial white patches, and irregular blue-black macules were visualized (Figure 2).

A large, keloid-like plaque with pustules and associated hair loss was present at the nape of the neck. Hair loss and thinning spanning from the anterior hairline with bitemporal recession were also apparent.

Assessment, Treatment, and Follow-Up

The differential diagnosis included acute exacerbation of chronic folliculitis decalvans (FD), dissecting cellulitis, central centrifugal cicatricial alopecia, lichen planopilaris, discoid lupus erythematous, and secondary syphilis. At the initial visit, the patient declined laboratory testing, bacterial culture, or biopsy because of financial concerns. Because of the acutely worsening symptoms, the patient was treated empirically for FD with a regimen of doxycycline 100 mg by mouth twice daily for 30 days until a biopsy could be performed to confirm the clinical diagnosis.

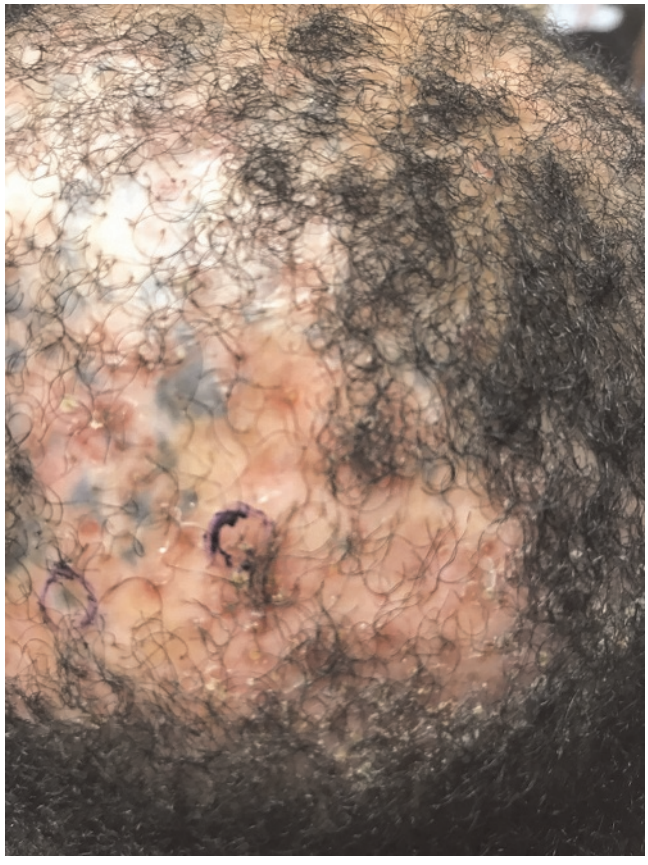

FIGURE 1. Cicatricial alopecia of the left superior parietal scalp.

At follow-up 2 weeks later, the patient reported adherence with his medication regimen and modest improvement of his symptoms. He stated his scalp was no longer itchy or painful. On physical examination, there was visible improvement of the erythema, whereas the remaining findings from the previous visit persisted. At this visit, the patient agreed to limited laboratory and pathological workup. A single 4-mm punch biopsy was performed, which sampled inflamed tissue and dark pigment on the right parietal scalp (Figure 3). A complete metabolic profile, lipid panel, complete blood count with differential, antinuclear antibody, venereal disease research laboratory test, and thyroid-stimulating hormone were ordered, and all returned within normal limits.

Histological analysis of the specimen supported a "lymphocyte-mediated scarring alopecia." The report revealed "mild orthohyperkeratosis, perifollicular fibroplasia, a perivascular and perifollicular lymphocytic infiltrate, a decreased number of hair follicles, numerous melanophages in the upper dermis, and horizontal fibrosis of collagen in the upper and mid dermis." The pathological findings, in the context of the clinical and laboratory findings, were most consistent with the expected findings of chronic FD after initiation of tetracycline therapy. The patient was informed of the results and instructed to complete the course of doxycycline. At 1-month follow-up, the patient no longer had pustules or papules, although scattered blue macules, mild erythema, perifollicular scale, and scarring persisted.

DISCUSSION

FD is a highly inflammatory cicatricial alopecia characterized by a painful, pruritic, suppurative folliculitis of the scalp. FD accounts for an estimated 10% of patients with cicatricial alopecia (Tan, Martinka, Ball, & Shapiro, 2004; Whiting, 2001), occurs most frequently in middle-aged men (Bolduc, Sperling, & Shapiro, 2016), and has no definitive racial predilection. Several familial cases have been reported, suggesting a possible genetic predisposition among some patients (Miguel-Gómez et al., 2018; Otberg, Kang, Alzolibani, & Shapiro, 2008). As in this case, FD is frequently associated with acne keloidalis nuchae (Luz Ramos, Muñoz-Pérez, Pons, Ortega, & Camacho, 1997; Tan et al., 2004; Whiting, 2001).

Clinically, the predominant lesions are perifollicular pustules and scale, with or without inflammatory papules, on a background of erythema near the periphery of areas of alopecia (Otberg et al., 2008). The pustules may be pruritic or painful, but they are frequently asymptomatic. The patches of alopecia are smooth and lack follicular orifices—these patches grow larger as the disease progresses. The lesions are distributed primarily among the vertex and parieto-occipital scalp (Otberg et al., 2008), and involvement of locations such as the beard and neck

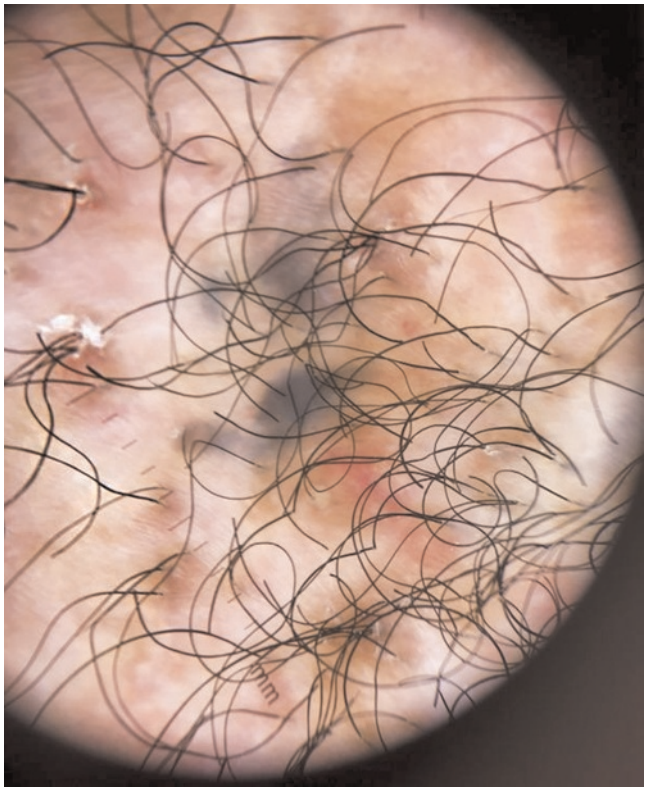

FIGURE 2. Dermoscopy of an area of cicatricial alopecia and deep postinflammatory hyperpigmentation.

is less common (Karakuzu, Erdem, Aktas, Atasoy, & Gulec, 2001). Trichoscopic findings classically include follicular hyperkeratosis, perifollicular erythema, tufted hairs, hair diameter variability, and white patches of scarring (Fernández-Crehuet et al., 2017). Tufted hair, the emergence of several hair shafts from a single follicle, is a classic finding in FD and may be associated with severe diseases (Miguel-Gómez et al., 2018); however, this finding is not specific to FD and can be found in other cicatricial alopecias (Bolduc et al., 2016).

The inflammation of FD is predominantly neutrophil mediated (Olsen et al., 2003). The immune response of FD is so similar to that of other cicatricial alopecias that it is even sometimes considered a subtype of central centrifugal cicatricial alopecia (Sperling, Solomon, & Whiting, 2000), although this concept is disputed (J. Powell & Dawber, 2001). *Staphylococcus aureus* is implicated in the pathogenesis of FD, but it remains unclear whether the clinical manifestations are because of bacterial superinfection, immune response to degenerating follicular components, or abnormal host response to the bacterial toxins (Sperling et al., 2000).

Punch biopsy is the preferred method for pathologic evaluation of alopecia. In many cases, a second sample can be taken via punch biopsy for vertical sectioning, although it is often unnecessary when suspicion for FD is high (Tailor et al., 2013). Bacterial culture of pustules with antimicrobial sensitivity analysis is also recommended to direct treatment decisions, as most cases of FD will grow *Staphylococcus aureus* (Otberg et al., 2008). Histological findings will commonly demonstrate follicular neutrophilic pustules, foreign-body giant cells, follicular tufting, keratin aggregation, and damaged or absent sebaceous glands. In long-standing diseases, a mixed infiltrate composed of plasma cells, neutrophils, and lymphocytes may be present, which may lead to difficulty in histologic discrimination between the various lymphocytic cicatricial alopecias (Otberg et al., 2008; Weedon, 2009). The lack of histologic neutrophil predominance in the presented case is most likely because of the chronic nature of the patient's disease as well as expected reduction in neutrophils as a result of starting tetracyclines (Gabler & Creamer, 1991); ideally, biopsy would be performed before treatment for an accurate diagnosis.

Diagnosis of FD is best made when the clinical picture, patient history, and histologic findings are considered collectively and other forms of alopecia are excluded. Superficial bacterial folliculitis of the scalp will lack the scarring alopecia seen in FD (Goldberg, 2018). Although pustules and inflammatory papules are often present in both dissecting cellulitis and FD, the presence of fluctuant nodules, abscesses, and sinus tracts is more consistent with dissecting cellulitis (Otberg et al., 2008). Erosive

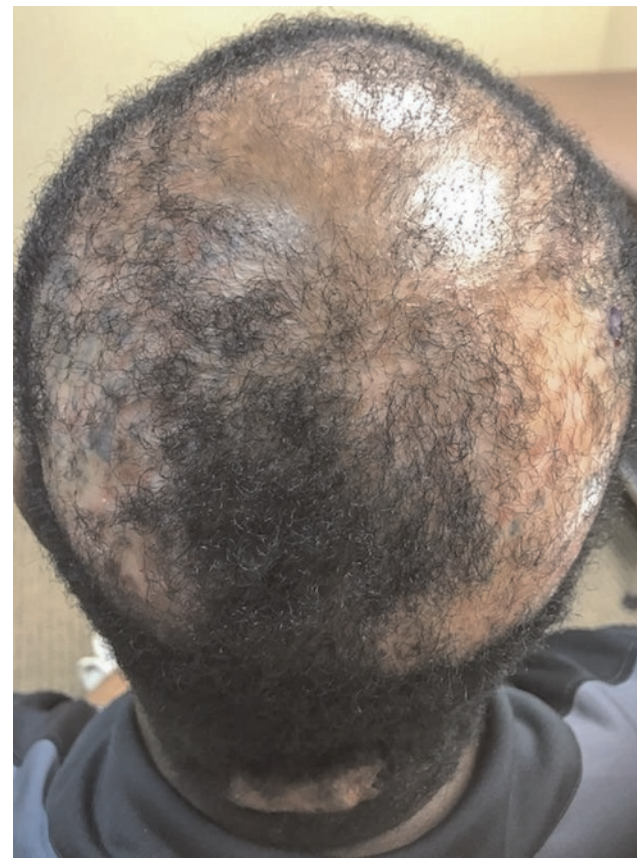

FIGURE 3. Reduction in erythema and number of pustules after 2 weeks of 100-mg doxycycline twice daily.

pustular dermatosis of the scalp features sterile pustules, erosions, and plaques, and it is most likely to occur in the older adults after scalp surgery or trauma. Advanced tinea capitis can mimic FD, although pustules are less likely to dominate clinically in tinea capitis, and fungal etiology is readily excluded with a potassium hydroxide preparation, biopsy, or fungal culture. Central centrifugal cicatricial alopecia typically lacks the erythema and scale seen in FD and is more likely to occur in women of African descent. Discoid lupus erythematosus of the scalp is distinguished from FD by the presence of erythematous scaly plaques and is easily differentiated with histological analysis (Goldberg, 2018). Lichen planopilaris features erythema and perifollicular scaling, similar to FD, but is far less likely to feature pustules, and a histological finding of lichenoid dermatitis will support the diagnosis of lichen planopilaris (Sperling et al., 2000). The ''moth-eaten'' pattern of alopecia observed in secondary syphilis may resemble the pattern of patchy hair loss in FD, but the hair loss of secondary syphilis is nonscarring and typically lacks the classic pustules or papules seen in many cicatricial alopecias (Stary & Stary, 2012).

Postinflammatory hyperpigmentation (PIH) is a less common complication of FD—particularly to the degree shown in this case—although it is frequently observed in other cutaneous inflammatory reactions such as acne, psoriasis, lichen planus, and atopic dermatitis (Callender, St Surin-Lord, Davis, & Maclin, 2011; Lacz, Vafaie, Kihiczak, & Schwartz, 2004). PIH is caused by abnormal melanin retention and can involve both the dermis and epidermis, or it can also be limited to one or the other. Melanin deposits of PIH tend to appear increasingly blue with increasing distance from the skin surface because short-wavelength light (blue) scatters and is reflected more readily than long-wavelength light (red) as visible light passes through a medium, a phenomenon known as the Tyndall effect (Weismann & Lorentzen, 2006). As a result, epidermal PIH appears tan to brown clinically, whereas dermal PIH appears dark brown or blue-gray, as observed in this case. Inflammatory mediators play a significant role by stimulating epidermal melanocyte proliferation, increasing melanin synthesis by melanocytes, and promoting melanin transfer to surrounding keratinocytes (Lacz et al., 2004; Weismann & Lorentzen, 2006).

In dermal PIH, inflammation results in the destruction of the basal layer and melanophages to accumulate in the upper dermis, which phagocytize the basal keratinocytes and melanocytes resulting in dermal melanin retention. (Lacz et al., 2004). Classic histological findings of dermal PIH include increased number of melanophages in the upper dermis, increased pigment deposition in the dermis, decreased pigmentation of the epidermis, and dermal perivascular lymphocytic infiltration (Park et al., 2017). Dermal PIH is notoriously difficult to treat and self-resolves less often than epidermal PIH. Effective treatment of PIH typically involves topical medications and procedural therapy, including chemical peels and phototherapy, although the first step is to prevent and treat the underlying inflammatory condition (Kaufman, Aman, & Alexis, 2018).

Treatment of FD is centered on eliminating pathogenic bacteria and reducing the inflammatory response. Oral antibiotics are the mainstay of treatment for FD. Tetracyclines are effective first-line therapy, particularly in mild-to-moderate or treatment-naive cases. A recommended regimen is 50–100 mg of doxycycline or minocycline given twice daily for 1–3 months or longer according to patient response (Bunagan, Banka, & Shapiro, 2015; Goldberg, 2018; Miguel-Gómez et al., 2018). In severe cases or cases refractory to first-line therapy, rifampicin 300 mg in combination with clindamycin 300 mg twice daily (to prevent bacterial resistance to rifampicin) for an initial course of 10 weeks is a common regimen (Miguel-Gómez et al., 2018; J. J. Powell, Dawber, & Gatter, 1999). In case of intolerance, allergy, or resistance, other oral antibiotics to consider include cephalexin, azithromycin, fusidic acid, trimethoprim–sulfamethoxazole, and ciprofloxacin. (Bunagan et al., 2015; Miguel-Gómez et al., 2018). Topical antibiotics including mupirocin, fusidic acid, erythromycin, and clindamycin have shown efficacy when combined with topical or intralesional corticosteroids in patients who have mild disease or prefer to avoid systemic therapy.

Alternative or supplemental therapies include zinc gluconate, zinc sulfate, oral glycyrrhizin, oral isotretinoin, dapsone, oral fusidic acid, acitretin, oral L-tyrosine, photodynamic therapy, tacrolimus, tumor necrosis factor-alpha inhibitors, and intravenous human immunoglobulin; however, these therapies either have shown inconsistent efficacy or have been used in a small number of cases (Bolduc et al., 2016; Ismail, Ralph, & Murphy, 2015; Miguel-Gomez, Vano-Galvan, Perez-Garcia, Carrillo-Gijon, & Jaen-Olasolo, 2015). Surgical hair transplants are discouraged in FD; only patients who have been disease-free for several years and are not being actively treated should be considered for hair transplant candidacy (Rose & Shapiro, 2004). Long-term management involves tapering oral antibiotics to the lowest dose that maintains remission in patients who quickly revert after initial therapy. Conservative measures, such as avoidance of caps and wigs (which often act as reserves for *Staphylococcus aureus*), are also important in maintaining remission (Goldberg, 2018; Otberg et al., 2008).

CONCLUSION

FD, as well as other inflammatory cicatricial alopecias, is important to recognize clinically as early intervention is critical to prevent long-standing complications such as significant irreversible hair loss and PIH. Optimal outcomes are achieved with administration of oral antibiotics, implementation of conservative management strategies, and regular follow-up. ■

REFERENCES

Bolduc, C., Sperling, L. C., & Shapiro, J. (2016). Primary cicatricial alopecia: Other lymphocytic primary cicatricial alopecias and neutrophilic and mixed primary cicatricial alopecias. *Journal of the American Academy of Dermatology*, *75*(6), 1101–1117. doi:10.1016/j.jaad.2015.01.056

Bunagan, M. J., Banka, N., & Shapiro, J. (2015). Retrospective review of folliculitis decalvans in 23 patients with course and treatment analysis of long-standing cases. *Journal of Cutaneous Medicine and Surgery*, *19*(1), 45–49. doi:10.2310/7750.2014.13218

Callender, V. D., St Surin-Lord, S., Davis, E. C., & Maclin, M. (2011). Postinflammatory hyperpigmentation: Etiologic and therapeutic considerations. *American Journal of Clinical Dermatology*, *12*(2), 87–99. doi:10.2165/11536930-000000000-00000

Fernández-Crehuet, P., Vañó-Galván, S., Molina-Ruiz, A. M., Rodrigues-Barata, A. R., Serrano-Falcón, C., Martorell-Calatayud, A., … Camacho-Martínez, F. M. (2017). Trichoscopic features of folliculitis decalvans: Results in 58 Patients. *International Journal of Trichology*, *9*(3), 140–141. doi:10.4103/ijt.ijt_85_16

Gabler, W. L., & Creamer, H. R. (1991). Suppression of human neutrophil functions by tetracyclines. *Journal of Periodontal Research*, *26*(1), 52–58. doi:10.1111/1600-0765.ep13695977

Goldberg, L. J. (2018). Folliculitis decalvans. Retrieved from https://www.uptodate.com/contents/folliculitis-decalvans

Ismail, N., Ralph, N., & Murphy, G. (2015). Intravenous human immunoglobulin for treatment of folliculitis decalvans. *Journal of Dermatological Treatment*, *26*(5), 471–472. doi:10.3109/09546634.2015.1024599

Karakuzu, A., Erdem, T., Aktas, A., Atasoy, M., & Gulec, A. I. (2001). A case of folliculitis decalvans involving the beard, face and nape. *The Journal of Dermatology*, *28*(6), 329–331. doi:10.1111/j.1346-8138.2001.tb00143.x

Kaufman, B. P., Aman, T., & Alexis, A. F. (2018). Postinflammatory hyperpigmentation: Epidemiology, clinical presentation, pathogenesis and treatment. *American Journal of Clinical Dermatology*, *19*(4), 489–503. doi:10.1007/s40257-017-0333-6

Lacz, N. L., Vafaie, J., Kihiczak, N. I., & Schwartz, R. A. (2004). Post-inflammatory hyperpigmentation: A common but troubling condition. *International Journal of Dermatology*, *43*(5), 362–365. doi:10.1111/j.1365-4632.2004.02267.x

Luz Ramos, M., Muñoz-Pérez, M. A., Pons, A., Ortega, M., & Camacho, F. (1997). Acne keloidalis nuchae and tufted hair folliculitis. *Dermatology*, *194*(1), 71–73. doi:10.1159/000246063

Miguel-Gomez, L., Vano-Galvan, S., Perez-Garcia, B., Carrillo-Gijon, R., & Jaen-Olasolo, P. (2015). Treatment of folliculitis decalvans with photodynamic therapy: Results in 10 patients. *Journal of the American Academy of Dermatology*, *72*(6), 1085–1087. doi:10.1016/j.jaad.2015.02.1120

Miguel-Gómez, L., Rodrigues-Barata, A. R., Molina-Ruiz, A., Martorell-Calatayud, A., Fernández-Crehuet, P., Grimalt, R., … Vañó-Galván, S. (2018). Folliculitis decalvans: Effectiveness of therapies and prognostic factors in a multicenter series of 60 patients with long-term follow-up. *Journal of the American Academy of Dermatology*, *79*(5), 878–883. doi:10.1016/j.jaad.2018.05.1240

Olsen, E. A., Bergfeld, W. F., Cotsarelis, G., Price, V. H., Shapiro, J., Sinclair, R., … Workshop on Cicatricial Alopecia (2003). Summary of North American Hair Research Society (NAHRS)-sponsored Workshop on Cicatricial Alopecia, Duke University Medical Center, February 10 and 11, 2001. *Journal of the American Academy of Dermatology*, *48*(1), 103–110. doi:10.1067/mjd.2003.68

Otberg, N., Kang, H., Alzolibani, A. A., & Shapiro, J. (2008). Folliculitis decalvans. *Dermatologic Therapy*, *21*(4), 238–244. doi:10.1111/j.1529-8019.2008.00204.x

Park, J. Y., Park, J. H., Kim, S. J., Kwon, J. E., Kang, H. Y., Lee, E. S., & Kim, Y. C. (2017). Two histopathological patterns of postinflammatory hyperpigmentation: Epidermal and dermal. *Journal of Cutaneous Pathology*, *44*(2), 118–124. doi:10.1111/cup.12849

Powell, J. J., Dawber, R. P., & Gatter, K. (1999). Folliculitis decalvans including tufted folliculitis: Clinical, histological and therapeutic findings. *The British Journal of Dermatology*, *140*(2), 328–333. doi:10.1046/j.1365-2133.1999.02675.x

Powell, J., & Dawber, R. R. (2001). Folliculitis decalvans and tufted folliculitis are specific infective diseases that may lead to scarring, but are not a subset of central centrifugal scarring alopecia. *Archives of Dermatology*, *137*(3), 373–374.

Rose, P., & Shapiro, R. (2004). Transplanting into scar tissue and areas of cicatricial alopecia in hair transplantation. In Unger W., & Shapiro R. (Eds.), *Hair transplantation* (4th ed., pp. 600–606). New York, NY: Marcel Dekker.

Sperling, L. C., Solomon, A. R., & Whiting, D. A. (2000). A new look at scarring alopecia. *Archives of Dermatology*, *136*(2), 235–242. doi:10.1001/archderm.136.2.235

Stary, A., & Stary, G. (2012). Sexually transmitted infections. In Bolognia J. L., Jorizzo J. L., & Schaffer J. V. (Eds.), *Dermatology* (3rd ed. Vol. 2, p. 1371).

Tailor, A., Asher, R. G., Craig, P. J., Groves, R. W., Fenton, D. A., & Stefanato, C. M. (2013). The current state of play in the histopathologic assessment of alopecia: Two for one or one for two? *Journal of Cutaneous Pathology*, *40*(3), 298–304. doi:10.1111/cup.12075

Tan, E., Martinka, M., Ball, N., & Shapiro, J. (2004). Primary cicatricial alopecias: Clinicopathology of 112 cases. *Journal of the American Academy of Dermatology*, *50*(1), 25–32. doi:10.1016/j.jaad.2003.04.001

Weedon, D. (2009). Weedon's skin pathology. In Houston M., & Davies B. (Eds), (3rd ed., pp. 397). London, England: Churchill Livingstone.

Weismann, K., & Lorentzen, H. F. (2006). Dermoscopic color perspective. *Archives of Dermatology*, *142*(9), 1250. doi:10.1001/archderm.142.9.1250

Whiting, D. A. (2001). Cicatricial alopecia: Clinico-pathological findings and treatment. *Clinics in Dermatology*, *19*(2), 211–225.