60-Year-Old Male with Painful Patches of Hair Loss on the Scalp

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Abstract

Folliculitis decalvans (FD) is a rare, primary, neutrophilic, cicatricial alopecia that usually occurs in young to middle-aged adults. FD initially presents as a distinct follicular pustule or papule that may be tender, painful, or pruritic. This area begins to slowly expand creating irregularly shaped patches of scarring alopecia as the follicular orifice is lost. Active disease continues peripherally in an expanding pattern. FD has a waxing and waning course with periods of more intense disease presentation followed by periods of reduced activity. FD almost exclusively affects the scalp with preferential activity at the vertex and occiput. Another notable characteristic of FD is tufting of hair characterized by multiple hairs (5–20) emerging from the same dilated follicle. The first step in the diagnosis of FD involves gathering a general medical history and a bacterial infection history as *Staphylococcus aureus* is commonly cultured from the active lesions. Following this, a thorough examination of the scalp should be performed to look for follicular ostia, perifollicular erythema, or follicular hyperkeratosis. Treatment for FD has traditionally focused on controlling inflammation and the microbiota through the use of steroids and antibiotics. Destruction of hair follicles results in permanent hair loss; therefore, therapy is targeted at active lesions to prevent future permanent hair loss. Photodynamic therapy (PDT) with red light as well as immunotherapies are potential treatments usually in those patients who are refractory to steroids and antibiotics. Both of these treatment options

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lack robust evidence and are associated with mixed outcomes. As FD can be a painful, distressing, disease that is scarring in nature, patient counseling is essential for comprehensive patient care.

Keywords

Folliculitis decalvans · Cicatricial alopecia · Hair tufting · Staphylococcus aureus · Photodynamic therapy

A 60-year-old male presented with tender, firm lesions on the central scalp of 6 months duration. He reported pustules throughout his scalp that he would pick at constantly. He was concerned because he now noticed hair loss within the patches and thickening of the skin. He denied hair loss elsewhere. He tried ketoconazole shampoo with no improvement.

On physical examination, there were irregular, atrophic patches of hair loss with surrounding pustules at the periphery that were tender to touch. Hair tufting was present. The eyebrows and eyelashes were of normal density. The fingernails were normal.

Based on the clinical case description, what is the most likely diagnosis?

1. Discoid lupus erythematosus
2. Dissecting cellulitis
3. Tinea capitis
4. Folliculitis decalvans

Diagnosis

Folliculitis decalvans.

Discussion

Folliculitis decalvans (FD) is a primary neutrophilic cicatricial alopecia [1]. Primary cicatricial alopecia is characterized by inflammatory destruction of the hair follicle, resulting in permanent hair loss, in contrast to secondary cicatricial alopecia, where non-follicular disease results in the follicle destruction (e.g., pemphigus vulgaris, burns) [2]. FD is considered a rare cause of alopecia, and in a large multicenter retrospective study was found to account for 3% of 3133 diagnoses of alopecia and 11% of 840 diagnoses of cicatricial alopecia [3]. FD presents in both men and women but shows male predominance. FD usually occurs in young to middle-aged adults and more frequently in African-American patients than in Caucasian patients [1, 4].

Clinically, FD initially presents as a distinct follicular pustule or papule that may be tender, painful, or pruritic (Figs. 13.1 and 13.2). Expansion ensues as adjacent papulopustules appear on the periphery of the erythematous lesion. Slowly, irregularly shaped patches of scarring alopecia occur, and the follicular orifice is lost. Active disease continues peripherally in an expanding pattern. Yellow-gray scales may be present, especially around the follicles, as well as follicular hyperkeratosis, erosions, and hemorrhagic crusts. FD has a waxing and waning course with periods of more intense disease presentation followed by periods of reduced activity [1]. FD may affect any area of the scalp but preferentially affects the vertex and occiput and often presents with multifocal lesions [2]. It almost exclusively involves the scalp, though there have been rare cases affecting the beard, face, and nape of the neck [5]. Another notable characteristic of FD is the tufting of hair characterized by multiple hairs (5–20) emerging from the same dilated follicle [2]. Despite its classification as a neutrophilic alopecia, FD may possibly be a continuum of disease with eventual lichenoid features depending on levels of bacterial biomarkers [6].

Histologically, FD commonly presents with an intrafollicular and perifollicular neutrophilic infiltrate. As the disease progresses, this infiltrate spreads into the surrounding adventitial dermis. Foreign body giant cells may be present and are localized to ectopic portions of the hair shaft [2]. In late stages of the disease, chronic inflammation results in the presence of more plasma cells and fewer neutrophils. These advanced lesions may also present with dermal fibrosis [2]. On dermoscopy,

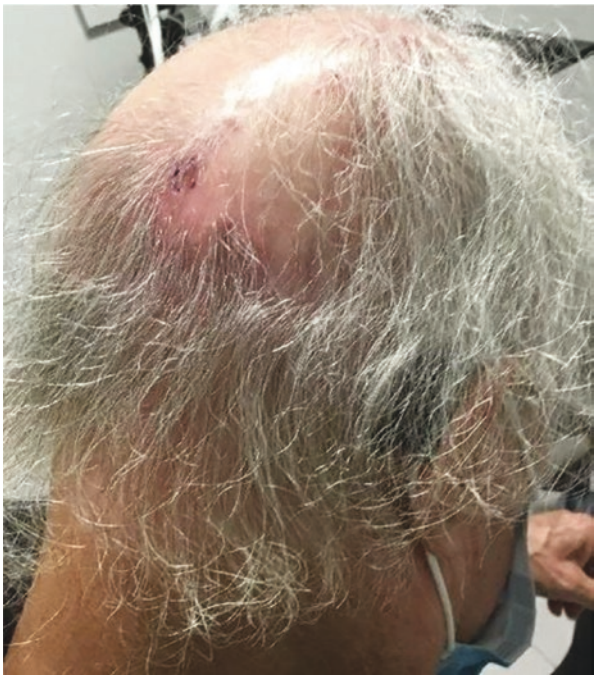

Fig. 13.1 Tender erythematous follicular pustule. Image courtesy of Dr. Melissa Piliang and Janine Sot, MBA medical photographer

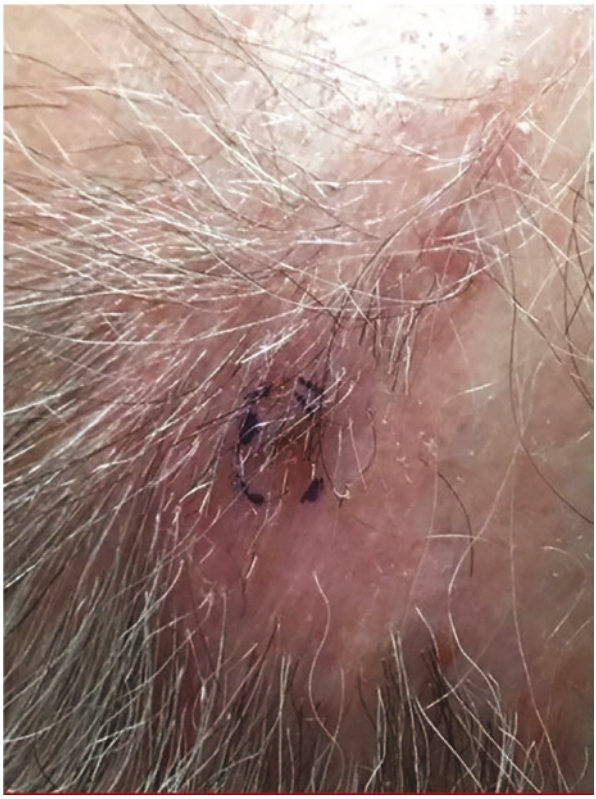

Fig. 13.2 Magnified image of the tender, erythematous follicular pustule that is the initial finding in FD. Image courtesy of Dr. Melissa Piliang and Janine Sot, MBA medical photographer

FD presents as fused follicular infundibula and thickened interfollicular keloid-like areas with tufted hairs [7].

While there are multiple theories on the etiology of FD, the pathophysiology is unknown. Many of the theories involve *Staphylococcus aureus* as it is commonly cultured from FD pustules. This finding has driven the use of antibiotics as the principal treatment for FD. However, it is important to note that while *S. aureus* is commonly cultured from active lesions, FD is not simply a bacterial infection. One study proposed that the epidermal barrier may be defective in FD. This study compared the superficial and subepidermal microbiota and found *S. aureus* colonized in 16 out of 20 patients in non-lesional, subepidermal skin in patients with lesions. As *S. aureus* was present in non-lesioned subepidermal skin, the implication is that FD could be due to a rupture of the epidermal barrier resulting in the invasion of opportunistic bacteria [8]. Other theories range from a congenital abnormality of follicular orifices to a deficiency in the host immune response to *S. aureus* autoimmune process [9]. The postulation that the host cell-mediated immunity is defective may explain the effectiveness of biological medications such as adalimumab in some refractory cases of FD [10]. More research is required before the etiology of FD can be determined conclusively.

The first step in the diagnosis of FD involves gathering a general medical history and a bacterial infection history. Following this, a thorough examination of the scalp should be performed to look for follicular ostia, perifollicular erythema, or follicular hyperkeratosis. Measurements, bacterial cultures, and biopsies of the active lesions should be taken to determine the diagnosis [9].

Treatment

Due to the erythematous pustules with abnormal bacterial findings, treatment for FD traditionally has focused on controlling inflammation and the microbiota through the use of steroids and antibiotics [10]. Treatment of FD is most effective for patients with active disease. Destruction of hair follicles results in permanent hair loss; therefore, therapy is targeted at active lesions to prevent future permanent hair loss. As FD can be a painful, distressing, disease that is scarring in nature, patient counseling is essential. Additionally, camouflaging techniques (e.g., wigs, caps, hairpieces, etc.) are possible solutions, but they can be a reservoir for *S. aureus*; therefore, cleansing with antiseptics and frequent switching between headpieces is important.

Current evidence of therapies is primarily based on observational studies or small sample sizes that lack external validity. One comparative case-control study was performed that focused on the impact of *S. aureus* on FD before and after an anti-staphylococcal treatment of rifampicin and clindamycin [8]. *S. aureus* was found initially in 0/20 of the control patients and 16/20 of the patients with FD. After treatment, 70% of patients had a decrease in pustule and crust formation and approximately 60% of patients had a decrease in the peripilar erythema. The *S. aureus* colonies almost completely disappeared, only remaining positive in two patients. Despite efficacious treatment, peripilar erythema persisted in about 80% of patients to a moderate degree [8].

For mild to moderate FD, topical antibiotics and topical and intralesional steroids are first-line therapies and can be used especially for those desiring to avoid the systemic side effects of antibiotics and steroids. The potency of the topical steroids is such that they can be applied locally 2 or 3 times per week in conjunction with topical antibiotics to treat FD of slight or moderate severity. Treatment with intralesional steroids is among the most effective methods for treating FD, and they can be used once every 3 months for slight inflammation in mild to moderate FD. Oral antibiotics, such as tetracyclines or azithromycin, may also be administered for mild to moderate FD [11, 12].

As FD becomes more severe, the above treatments may still be considered. However, for refractory cases, a combination of rifampicin with clindamycin is more effective with a longer duration of response. Clindamycin 300 mg twice daily with rifampin 300 mg twice daily are dosed for 12 weeks. Systemic corticosteroids may also be effective to control FD [11, 12]. Isotretinoin is beneficial for FD that is not well controlled with previous treatments, and other cases demonstrate that it can

result in complete remission of FD and may warrant further consideration as a primary treatment [12, 13].

Photodynamic therapy (PDT) with red light is another potential treatment, though it lacks robust evidence and is associated with mixed outcomes [14]. To date, the largest study of PDT in FD selected 13 patients with previously poor outcomes using various topical and oral agents and placed them on a course of PDT with no other therapy. All showed improvement during treatment, and at 12-month follow-up, nine patients maintained well-controlled symptoms while four relapsed [15]. Insufficient evidence supports using PDT as first-line therapy, but it may be beneficial to use in refractory cases of FD.

Lastly, there have been reports of various immunotherapies namely secukinumab, adalimumab, hydroxychloroquine, and infliximab successfully treating FD, and one case utilizing apremilast [11, 16, 17]. All of these patients were refractory to traditional treatments and responded favorably to their respective immunotherapy. This may indicate an underlying defect of the immune system in individuals with FD, especially in refractory cases. Additionally, the efficacy of platelet-rich plasma (PRP) was demonstrated in the treatment of two cases of refractory FD. Further research is needed to understand the successful symptomatic control achieved with PRP, but its antimicrobial and anti-inflammatory effects are promising for FD treatment [18].

Key Points

- Folliculitis decalvans (FD) is a rare cause of alopecia identified by active erythematous disease on the periphery of lesions.
- Despite bacteria in active pustules, a simple bacterial infection is not the sole cause of FD manifestation.
- Initial treatment is topical/oral antibiotics, like tetracyclines, and rifampicin/clindamycin and corticosteroids.
- Future therapies, especially in refractory cases, may involve PDT and various immunotherapies.

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