

Alopecia with perifollicular papules and pustules

Our 23-year-old patient thought his hair loss was probably “genetic.” But that didn’t explain the painful pustules.

A 23-YEAR-OLD AFRICAN AMERICAN MAN sought care at our medical center because he had been losing hair over the vertex of his scalp for the past several years. He indicated that his father had early-onset male patterned alopecia. As a result, he considered his hair loss “genetic.” However, he described waxing and waning flares of painful pustules associated with occasional spontaneous bleeding and discharge of purulent material that occurred in the same area as the hair loss.

Physical examination revealed multiple perifollicular papules and pustules on the vertex of his scalp with interspersed patches of alopecia (FIGURE 1). There were no lesions elsewhere on his body and his past medical history was otherwise unremarkable.

- WHAT IS YOUR DIAGNOSIS?
- HOW WOULD YOU MANAGE THIS PATIENT?

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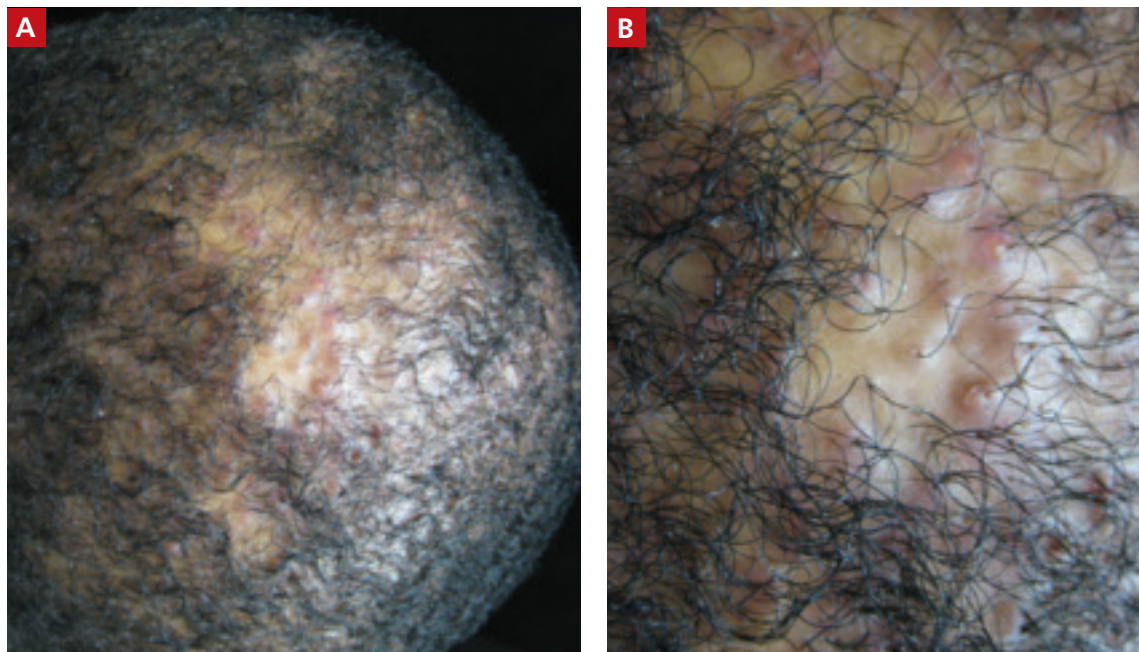
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FIGURE 1

Alopecia with a painful twist



PHOTOS COURTESY OF: OLIVER J. WISCO, DO

This 23-year-old patient said that he had spontaneous bleeding and discharge of purulent material in the area of his hair loss.

➤ **Folliculitis decalvans has been linked to scalp colonization with *S aureus*.**

**Diagnosis:
Folliculitis decalvans**

Folliculitis decalvans (FD) is a highly inflammatory form of scarring alopecia characterized by inflammatory perifollicular papules and pustules. The term scarring alopecia refers to the fact that the follicular epithelium has been replaced by connective tissue, ultimately resulting in permanent hair loss. This manifests clinically as patches of skin without terminal or vellus hairs, whereas a nonscarring alopecia would demonstrate preservation of the vellus hairs. Left untreated, advancing permanent hair loss ensues and may result in an end-stage pattern of tufted folliculitis or polytrichia, where interspersed dilated follicular openings house multiple hairs.

Affected areas commonly include the vertex and occipital scalp. Common symptoms include pain, itching, burning, and occasionally spontaneous bleeding or discharge of purulent material.¹

FD generally occurs in young and middle-aged African Americans with a slight predominance in males. It accounts for 11% of all primary scarring alopecias.^{2,3} The etiology of this inflammatory process is not fully understood; however, scalp colonization with *Staphylococcus aureus* has been implicated as a contributing factor.⁴ Other reports suggest patients may have an altered host immune response and/or genetic predisposition for this condition.^{2,3}

The differential includes various scarring, nonscarring alopecias

Since clinical findings of FD can range from relatively nonspecific mild disease at its onset to the end stage described above, a detailed patient history is needed. The following scarring and nonscarring alopecias should be considered in the differential diagnosis: dissecting cellulitis of the scalp, central centrifugal cicatricial alopecia (CCCA), acne keloidalis nuchae, erosive pustular dermatosis, lichen planopilaris (LPP), inflammatory tinea capitis, and secondary syphilis.

■ **Dissecting cellulitis of the scalp** is a distinctive, often debilitating disease commonly seen in young adult African American

men. It is considered part of the follicular occlusion tetrad that also includes hidradenitis suppurativa, acne conglobata, and pilonidal cysts. It presents as a scarring alopecia with firm scalp nodules that rapidly develop into boggy, fluctuant, oval to linear sinuses that may eventually discharge purulent material.

In contrast to FD, dissecting scalp cellulitis lesions interconnect via sinus tract formation so that pressure on one fluctuant area may result in purulent discharge from perforations several centimeters away.⁵ Although both dissecting cellulitis and FD are considered primary neutrophilic scarring alopecias, the presence of true sinus tract formation can be a distinguishing finding.

■ **CCCA** is the most common form of scarring alopecia among African Americans and is particularly seen among African American women.⁵ It generally presents on the scalp vertex like FD, but it is much less inflammatory and typically causes only mild pruritus or tenderness of the involved areas.

Although numerous theories have been suggested, the etiology is unknown. The pathogenesis is thought to be associated with premature desquamation of the inner root sheath, which can be demonstrated on biopsy. Also seen histologically is lymphocytic perifollicular inflammation and polytrichia.⁶

■ **Acne keloidalis nuchae** is also a scarring alopecia. It is seen most commonly in African American men and presents as keloid-like papules and plaques with occasional pustules characteristically on the occipital scalp and posterior neck. In contrast to FD, acne keloidalis nuchae papules coalesce and may form firm, hairless, protuberant keloid-like plaques that may be painful and cosmetically disfiguring. The cause of acne keloidalis nuchae is unknown.

Shaving or cutting tight curly hair too short and subsequently having the new hair curve back and penetrate the skin may be the precipitating event. Thus, a history of close shaving should make one suspect this diagnosis. Histologic analysis reveals a chronic, predominantly lymphocytic folliculitis with eventual follicular destruction.

■ **Erosive pustular dermatosis** is a rare disorder that primarily affects the elderly. It is characterized by a chronic amicrobial pustule-

lar dermatosis with extensive boggy, crusted, erosive plaques on the scalp resulting in scarring alopecia. Most cases have an onset after the age of 40. Therefore, age of onset may help differentiate between erosive pustular dermatosis and FD.

The cause of erosive pustular dermatosis is unknown. It is thought to be related to local trauma, such as chronic sun exposure, occurring months to years prior to the onset of lesions or as an autoimmune process.⁶ Histologic specimens show nonspecific changes including parakeratosis or hyperkeratotic scale with atrophy or erosion of the epidermis, while an inflammatory infiltrate with lymphocytes and plasma cells is found in the dermis.

■ **LPP** is seen more commonly in women than men, and Caucasians are more often affected than African Americans. It presents with erythema, perifollicular scale, and scattered patches of scarring alopecia. Half of involved cases develop concomitant clinical features of lichen planus. When present, these characteristics may help distinguish it from FD and other scarring alopecias.⁶

The etiology of LPP is unknown, but is thought to be similar to the presumed cause of lichen planus: a T-cell-mediated autoimmune response that damages basal keratinocytes.⁵ Histologic findings include a band-like mononuclear cell infiltrate obscuring the interface between follicular epithelium and dermis at the superficial part of the follicle with occasional interfollicular epidermal changes consistent with lichen planus.

■ **Inflammatory tinea capitis** is a common dermatophyte infection of the scalp that affects children and adults alike. Typically, it is easily distinguished from FD. However, severe cases may result in a highly inflammatory pustular eruption with alopecia—with or without a kerion—which can make differentiation difficult.

In contrast to FD, the alopecia associated with tinea capitis is usually non-scarring, although this depends on the extent and depth of infection. Also, tinea capitis may present with either discrete patches or involve the entire scalp, whereas FD is usually localized to the vertex or occiput (as noted earlier). Correct diagnosis can be accomplished by means of light microscopy and fungal culture.

■ **Secondary syphilis** is usually a sexually transmitted disease, but it can also be acquired perinatally. It often presents with a “moth-eaten” alopecia and should be considered when examining patients with patchy alopecia such as that seen in FD. These lesions manifest 3 to 10 weeks after the onset of primary syphilis. Early in its course, the condition is reversible, but if it becomes chronic, the condition will cause a scarring alopecia.

The presence of other stigmata, including a generalized pruritic papulosquamous eruption with involvement of the palms and soles, mucosal lesions ranging from superficial ulcers to large gray plaques, and condylomata lata, should help to differentiate syphilis from FD.

Serologic tests such as rapid plasma reagin and venereal disease research laboratory assays are often preferred for routine screening. If the index of suspicion is high, confirmatory testing with direct antibody assays such as a microhemagglutination assay or fluorescent treponemal antibody absorption test is indicated.

Biopsy is needed for the diagnosis

Two scalp biopsies should be performed to make the diagnosis. Recommended guidelines for sampling the scalp include performance of 4-mm punch biopsies extending into the fat at 2 different clinically active sites.⁷ One biopsy should be processed for standard horizontal sectioning, but the second biopsy should be bisected vertically, with half sent for histologic examination and the other half for tissue culture (fungal and bacterial). An additional subsequent biopsy for direct immunofluorescence may also be considered if the initial biopsies are nondiagnostic.

Bacterial and fungal cultures collected from an intact pustule on the scalp with a standard culture swab should also be undertaken with pustular disease. If scale is present, a potassium hydroxide examination can help establish the diagnosis of a fungal etiology.

Doxycycline, intralesional corticosteroids are the first line of Tx

Management of FD can be difficult, and long-term treatment is often necessary. You'll need

➤ **Management of folliculitis decalvans can be difficult, and long-term treatment is often necessary.**

➤ **Our patient responded well to oral doxycycline 100 mg twice daily and clobetasol 0.05% topical solution to the affected area in the morning and evening.**

to explain to patients that their current hair loss is permanent and that the goal of treatment is to decrease inflammation and prevent further balding.

After initial bacterial cultures and sensitivities are obtained, primary treatment is aimed at eliminating *S aureus* colonization. Often, this requires oral antibiotic therapy, most commonly doxycycline 100 mg twice daily⁵ (strength of recommendation [SOR]: C). Topical antibiotics, however, may be used in mild cases; options include 2% mupirocin, 1% clindamycin, 1.5% fusidic acid, or 2% erythromycin applied twice daily¹ (SOR: C). In recalcitrant cases, a common treatment regimen includes oral rifampin 300 mg and clindamycin 300 mg twice daily for 10 weeks¹ (SOR: C).

Adjunctive topical and intralesional corticosteroids may help reduce inflammation and provide symptomatic relief from itching, burning, and pain. Topical class I or II corticosteroids can be used twice daily, whereas intralesional triamcinolone acetonide (combined with topical and/or oral antibiotics) may be administered every 4 to 6 weeks, starting at a concentration of 10 mg/mL¹ (SOR: C). Oral corticosteroids should only be considered for highly active and rapidly progressive symptoms.

Dapsone may also be considered as a treatment option for FD due to its antimicrobial activity and anti-inflammatory action directed toward neutrophil metabolism. Relapse, however, is frequent after treatment withdrawal¹ (SOR: C).

Improvement, but anticipated chronicity

We prescribed oral doxycycline 100 mg twice daily for our patient, as well as clobetasol 0.05% topical solution, to be applied to the affected area in the morning and evening.

We told our patient that FD is a chronic relapsing disorder and that while we could not make the condition go away completely, we could control it. We advised the patient to follow up every 2 months for the next 6 months, then every 6 months to ensure there was no progression or need to change the treatment regimen.

The patient's symptoms improved after the first 2 months. After weaning the patient off doxycycline over a 6-month period, we planned to transition the patient to topical clindamycin solution twice daily.

In some cases, the patient can be weaned off oral antibiotics once the condition is controlled, but for most patients, continuous systemic therapy is needed. **JFP**

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Strength of recommendation (SOR)

- A** Good-quality patient-oriented evidence
- B** Inconsistent or limited-quality patient-oriented evidence
- C** Consensus, usual practice, opinion, disease-oriented evidence, case series

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