FOLLICULITIS DECALVANS – DIAGNOSIS AND TREATMENT DIFFICULTIES

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Summary

Folliculitis decalvans is a rare, inflammatory disease of the scalp, which can evolve to irreversible cicatricial alopecia. Even though the mechanisms of this disease are not fully known, it is considered that Staphylococcus aureus plays an important role in its etiopathogenesis. We report the case of a 60 years old patient that was admitted to our department for multiple pruriginous, alopecic atrophic plaques, with numerous ulcerations covered by serohematic crusts and scales, painful pustules on the vertex and parietal-occipital regions of the scalp. The diagnosis was confirmed by clinical, pathological examination, and trichoscopy, which were the key factors to a successful treatment. The particularity of the case resides in the long, unfavorable evolution, incorrect treatment of the patient, being misdiagnosed for a long time in other dermatological services. All this underlines the complexity of cicatricial alopecia and the difficulty of diagnosis, which may often lead to inappropriate treatment.

Keywords: *cicatricial alopecia, folliculitis decalvans, scalp, staphylococcus aureus.*

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Introduction

Folliculitis decalvans of the scalp, named by Brocq in 1905, described by Quinaquaud in 1888 as "a destructive and epilating folliculitis of the hairy regions", represents an intensive inflammatory subtype of primary scarring alopecia. [1] Cicatricial alopecias are rare diseases described by the irreversible loss of hair follicles replaced by fibrous tissue.

The destructive process of scarring alopecias can be primary when the inflammation is in the center of the follicle, or secondary, caused by a localized inflammatory process of the scalp or post-traumatically, during which the hair follicle is considered an "innocent spectator." In 2001, the workgroup of scarring alopecias from North America classified them into three categories, based on the presence of inflammatory cells:

1. lymphocytic – discoid lupus erythematosus, planopilaris lichen LPP (classic LPP), frontal fibrosing alopecia, Graham-Little syndrome), Brocq pseudopelade, central centrifugal cicatricial alopecia, alopecia mucinosa, follicular decalvans spinulosus keratosis;
2. neutrophilic - folliculitis decalvans, dissecting cellulitis of the scalp;
3. mixed - keloid acne, acne necrotica, erosive pustular dermatosis. [2]

Even though the etiopathogenesis of primary scarring alopecias is not fully known, the main cause is represented by the destruction of epithelial stem cells after an abnormal immune response directed against the autoantigens of specific stem cells or against the surrounding differentiated epithelium, case in which the regenerative potential of the stem cells is exceeded.[3]

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Clinical case

We present the case of a 60-year-old patient, without comorbidities, admitted to the Dermatovenereology Department for multiple painful pustules and ulcerations covered by serohematic crusts, atrophic alopecic plaques diffusely distributed on the scalp, with local pruritus.

The disease debuted approximately 18 years ago and was treated with local corticosteroid infiltrations, which proved inefficient, causing atrophic alopecic plaques with irreversible hair loss. For a long time, especially during the summer months, the patient presented asymptomatic pustules on the scalp. A year ago, pruriginous erythematosus plaques debuted on the scalp, accompanied by painful pustules, for which he was treated, in other dermatology services, with high potency corticosteroids (clobetasol) for three weeks, with an unfavorable evolution, aggravation of the lesions and the debut of numerous pustules, erosions, scales, crusts, and hair loss. Systemic and local antifungal treatment was also associated but was inefficient, so the patient was admitted to our Dermatology Department (Emergency Hospital of Sibiu County). The clinical exam of the patient was within normal limits.

In the parietal-occipital region, on the vertex of the scalp, around the periphery of the alopecic plaques, the patient presented multiple perifollicular pustules and ulcerations of different sizes (diameters of 0.5–3 cm) covered by thick serohematic, brown-yellowish, painful adherent scales and crusts. On the scalp, the patient presented multiple, slightly uneven, pruriginous alopecic atrophic plaques of different shapes and sizes, with an erythematous halo and white center, see (Fig. 1, Fig. 2). The hair pull test was positive. The patient presented a painful erythematosus nodular lesion on the anterior thorax, with a diameter of approximately 1 cm. On both inguinal sides, there were well-defined, pruriginous, erythematosus placards with reticular margins. The scalp's trichoscopy presented areas of alopecia without follicular openings and hair shafts, white dots (dermis fibrosis), "milky-red" erythematosus areas, and porcelain-like structureless white areas, follicular pustules, brown-yellowish scales and crusts, tufted hairs, perifollicular erythema, see (Fig. 3). Laboratory tests showed: mixed dyslipidemia, elevated erythrocyte sedimentation rate (ESR), positive nasal swab test for MSSA Staphylococcus aureus. The bacteriologic exam from the pustules was positive for MSSA Staphylococcus aureus, the antibiogram being the reference method in choosing the appropriate local and systemic treatment. Direct mycological exam, blood count, hepatic and renal function, C3 and C4 complement values, immunoglobulins, lupus cells, rheumatoid factor were negative. Skin biopsy confirmed the diagnosis of folliculitis decalvans, and the histopathological exam showed a cutaneous fragment taped by stratified squa-

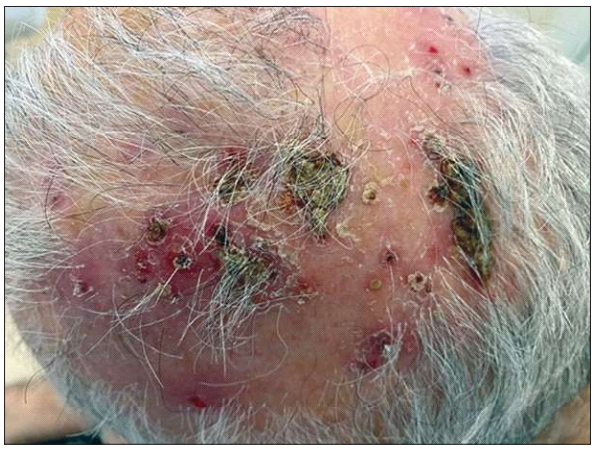

Figure 1. Folliculitis decalvans – on admission.

Figure 2. Folliculitis decalvans – after initiation of treatment.

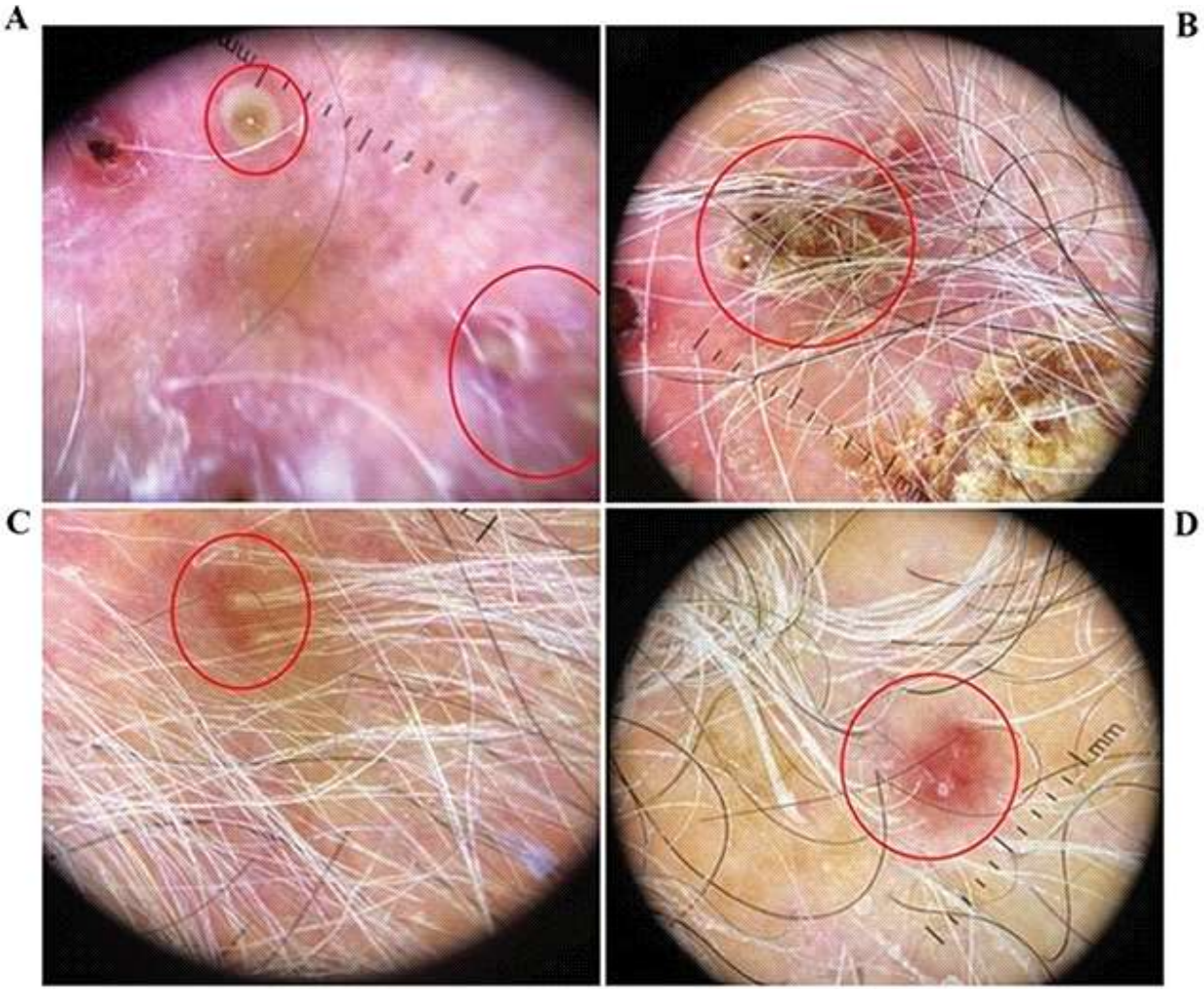

Figure 3. Trichoscopy of folliculitis decalvans. Legend: A. Follicular pustules (red circles), B. Ulcerations covered by yellow-brown scales and crusts (red circle), C – Tufted hairs (red circle), D. Perifollicular erythema (red circle).

mous epithelium with focal areas of parakeratosis, slight acanthosis, faded epidermal ridges, intact dermal-epidermal junction. There was a rich, diffusely located lymphocytic inflammatory infiltrate and numerous mature plasmocytes with a profound extension and destruction of follicles in the papillary and reticular dermis, (see Fig. 4).

After thorough clinical and paraclinical investigations, the patient was diagnosed with folliculitis decalvans of the scalp, multiple superinfected ulcerations with MSSA Staphylococcus aureus, hidradenitis of the anterior thorax, inguinal intertrigo candidosis, rhinitis with MSSA Staphylococcus aureus, mixed dyslipidemia.

The patient was treated with systemic antibiotics (100 mg Doxycycline 2 tablets/day for two weeks, then 1 tablet/day up to 3 months) and probiotics. The local treatment included the antiseptics, cleaning of the area with saline, antibiotic ointments (respecting the antibiogram), keratolytics, emollients, with a favorable evolution, remission of scales and crusts, epithelialization of the ulcerations, fading of local erythema, and improvement of the symptomatology in a matter of days, (see Fig. 5). The local maintenance treatment for the next

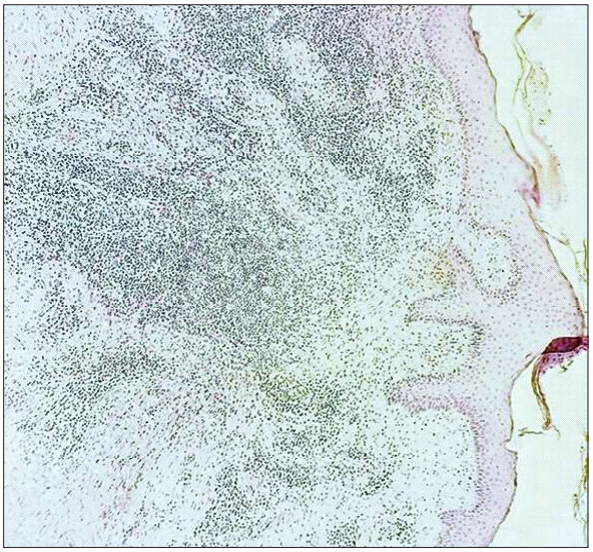

Figure 4. Histopathological exam.

months included: antiseptics, antibiotics, retinoids, keratolytics, exfoliants.

The lesion on the anterior thorax was treated with ichthyolated ointment combined with antibiotics after the disinfection of the area. The inguinal intertrigo was treated with Sabouraud solution. In order to eradicate the nasal infection with Staphylococcus aureus, we used an intranasal topical antibiotic treatment.

After a month of treatment, the patient no longer presented active lesions of folliculitis, and the erythema was barely visible, (see Fig. 6).

Discussions

Folliculitis decalvans represents approximately 11% of the total of primary scarring alopecias – neutrophilic group, next to dissecting cellulitis of the scalp.

Mainly, this pathology affects young people and adults, mostly afro-americans than Caucasians, with a predilection for males. [4]

The etiopathogenesis of folliculitis decalvans is not fully known. It is considered that Staphylococcus aureus (SA) plays a very important part because it can be found in most patients with untreated folliculitis of this type. It can act through bacterial superantigens that stimulate the immune system, as an abnormal response from the host to the staphylococcal toxins or as a secondary infection. It is well known that in order to treat and limit the

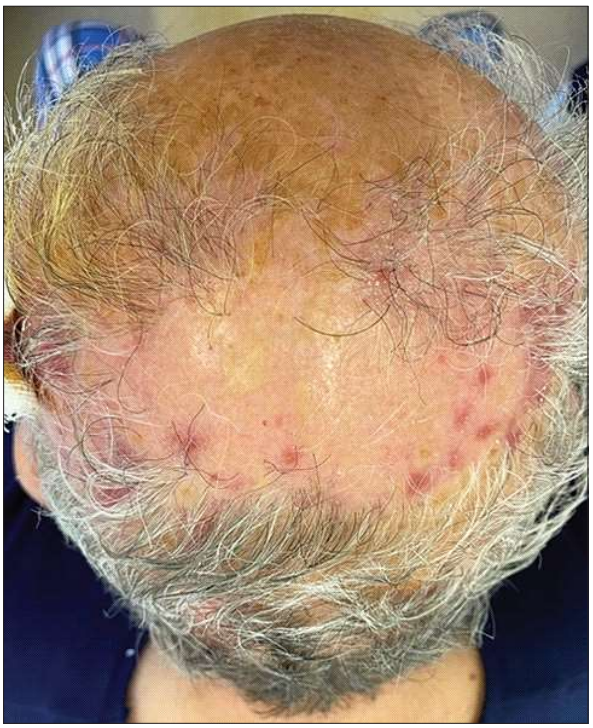

Figure 5. Evolution after six days of treatment.

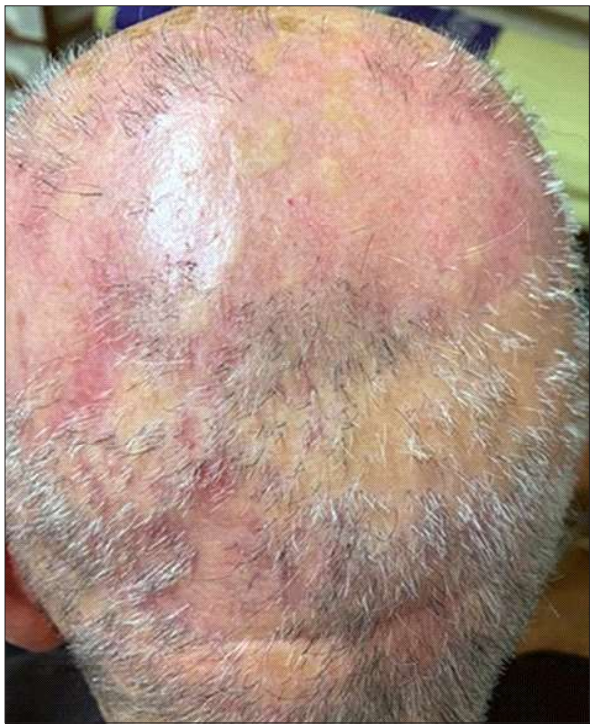

Figure 6. Evolution after one month of treatment.

disease's evolution, it is very important to eradicate Staphylococcus aureus. [4] [5]

Antigen-presenting cells, such as Langerhans cells, process Staphylococcus aureus (SA) and activate T lymphocytes. SA enterotoxins can act as superantigens and directly activate T lymphocytes through the V domain of the T lymphocyte receptor. Neutrophils can be recruited by immune mechanisms via interleukin (IL) 8. T lymphocytes can promote inflammation mainly by interferon (IFN), tumor necrosis factor (TNF), and fibrogenesis through the transforming growth factor (TGF) and through the basic fibroblast growth factor (b-FGF)), IL 1 and IL 4. Fibroblasts can overproduce the extracellular matrix, which accumulates in the dermis. Molecules can also be involved in the pathophysiological process: ELAM, endothelial leukocyte adhesion molecule, ICAM, intercellular adhesion molecule, VCAM, vascular cell adhesion molecule, etc. [5]

A genetic theory was also postulated since there were some familial cases reported. [6]

Anamnesis, clinical examination, trichoscopy, bacterial cultures, and scalp biopsy can help the clinician establish the diagnosis. Clinically, the disease debuts with macules, perifollicular erythematous papules, and hyperkeratosis (possibly), pustules, accompanied by pain, pruritus, and/or burning sensation. The lesions evolve clinically with the onset of yellow/gray scales, erosions, and ulcerations, yellow/hemorrhagic crusts, alopecic plaques. The evolution of the disease in the absence of treatment is chronic, progressive with the repeated reappearance of pustules, destruction of hair follicles, and the formation of skin-colored, erythematous or ivory white, smooth atrophic alopecia plaques of different sizes and shapes, with pustules on the periphery. Old lesions have a scarred appearance, without active lesions of folliculitis. [7]

The histopathological examination is the gold standard in differentiating the types of scarring alopecias. It is recommended to perform a scalp biopsy of the alopecic plaques with a 4 mm punch-biopsy tool in a clinically active area. [8] According to the classification of scarring alopecias, folliculitis decalvans belongs to the neutrophilic group, we specify that neutrophils are present in the inflammatory infiltrate at the onset of the disease. In contrast, in patients with advanced disease, the inflammatory infiltrate may be mixed. Thus, in folliculitis decalvans, the initial lesions show keratin aggregation at the infundibulum with dense acute perifollicular neutrophilic infiltrate in the upper part of the follicle. In evolution, the intrafollicular and perifollicular inflammatory infiltrate consists of neutrophils, lymphocytes, histiocytes, and plasmocytes in the dermis, with the onset of follicular destruction. Later, histopathological examination describes scarring perifollicular fibrosis with fibrous trajectories that replace the hair follicle, tufted hairs, interstitial dermal fibrosis, hypertrophic scars. The sebaceous glands are destroyed early on. [9]

Trichoscopy can provide important clues for an early diagnosis. The specific criteria for trichoscopy in primary scarring alopecias are: loss of follicular ostium, white dots and plaques, scales, and erythema around the hair shafts. Common trichoscopy elements in folliculitis decalvans are: tufted hairs, which represent the emergence of 5-20 hairs from a dilated follicle, are highly specific criteria for this type of folliculitis, perifollicular erythema, follicular hyperkeratosis, white plaques, perifollicular pustules. Perifollicular tubular scales, crusts, wooded vessels, erosions may also be present. [10]

The differential diagnosis of folliculitis decalvans of the scalp may be difficult due to the evolving clinical polymorphism. We excluded the other pathologies that are part of primary scarring alopecias, secondary scarring alopecia, alopecia areata, trichotillomania, secondary syphilis. [4] [9]

Folliculitis decalvans is a chronic pathology with many relapses and is very difficult to treat. The treatment is usually long-lasting, with irreversibly destroyed hair follicles, but if initiated at the onset of the disease, the evolution of the disease may be slowed down. The role of treatment is to stop inflammation and follicular destruction while preventing irreversible alopecia. There is still no therapeutic consensus due to a small number of studies, low incidence of the disease. However, antibiotics are considered the first line of treatment for antibacterial and anti-inflammatory effects, as we

previously mentioned the role of Staphylococcus aureus in the etiopathogenesis of the disease. Thus, therapeutic options may include: topical and systemic antibiotics (doxycycline, rifampicin, clindamycin, azithromycin), topical and systemic corticosteroids, topical and systemic vitamin A derivatives, dapsone, antiseptics, photodynamic therapy (PDT), tacrolimus, etc. [11] [12]

The particularity of this case is that the patient was misdiagnosed for a very long time, so the targeted treatment could not be initiated to limit the evolution of the disease; topical corticosteroid therapy aggravated the lesions, antifungal treatment was ineffective, and the evolution of the disease was unfavorable, with extensive irreversible hair loss. Another particularity is the association of decalcifying folliculitis with suppurative hidradenitis, an inflammatory condition that affects the sweat glands, in which inflammation and Staphylococcus aureus play a decisive role.

Conclusions

- Folliculitis decalvans is a chronic disease with multiple relapses that has a major impact on the patient's quality of life: discomfort, psychological effects, disfigurement, lowering self-esteem.
- In the other scarring alopecias and folliculitis decalvans, a fast diagnosis is extremely important to slow down the evolution of the disease and its complications: scarring alopecia with irreversible loss of hair.
- The improvement of inflammation and stopping the progression of this pathology are the most important therapeutic targets. Eradication of Staphylococcus aureus has a very important role in limiting the disease.
- New studies are needed to understand the pathogenesis and therapeutic options for a better management of this type of alopecia.

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Conflict of interest
NONE DECLARED

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