Case Report

Linear Circumscribed Scleroderma-Like Folliculitis Decalvans: Yet Another Face of a Protean Condition

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ABSTRACT

Since Quinquaud’s original report of folliculitis decalvans (FD), further clinical variants have been described on the basis of common histopathological and microbiological findings. Histopathology reveals a neutrophilic primary scarring alopecia, and microbiological studies invariably reveal pathogenic strains of *Staphylococcus aureus*. The presence of thickening of lesional skin in FD has been previously described. We report a new presentation of FD, clinically mimicking linear circumscribed scleroderma of the scalp. Overlapping features of the scarring alopecias may blur the distinction between different conditions that ultimately share the common final pathway of replacement of follicle by fibrous tissue. Therefore, a careful patient history, clinical examination including dermoscopy, microbiological studies, and a scalp biopsy for histopathology, and immunofluorescence studies are prerequisites to an accurate diagnosis and appropriate treatment of the respective condition. The case is presented to illustrate the clinical variability in presentation of FD and to underline the necessity of performing a biopsy for an accurate diagnosis in the scarring alopecias.

Key words: Folliculitis decalvans, folliculitis decalvans with linear arrangement, linear circumscribed scleroderma, linear circumscribed scleroderma-like folliculitis decalvans

INTRODUCTION

*I can add colours to the chameleon, Change shapes with Proteus for advantages*

—William Shakespeare

The scarring alopecias represent a group of diverse disorders with permanent destruction of the pilosebaceous unit and irreversible hair loss. Where there is no obvious physical/chemical injury or acute infectious etiology, clinical differential diagnosis of scarring alopecia is often difficult. The clinical inspection is of limited usefulness in establishing a diagnosis. Overlapping features may blur the distinction between different conditions that ultimately share the common final pathway of replacement of follicle by fibrous tissue. Therefore, a careful patient history, clinical examination including dermoscopy, microbiological studies, and a scalp biopsy for histopathology, and immunofluorescence studies are prerequisites to an accurate diagnosis and appropriate treatment of the respective condition.[³]

Linear circumscribed scleroderma of the scalp and folliculitis decalvans (FD) represent two disparate scarring alopecias that are characterized by distinctive clinical, dermoscopic, microbiological, and histopathological features. A case of linear circumscribed scleroderma-like FD is presented to illustrate the clinical variability in presentation of FD and to underline the necessity of performing a biopsy for an accurate diagnosis in the scarring alopecias.

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CASE REPORT

A 39-year-old female patient was referred by her dermatologist because of a focal scarring alopecia of the vertex region with an unspecific histopathologic finding of fibrosing alopecia.

Clinical inspection revealed a 1.5 cm × 4 cm area of white porcelain-colored plaque of scarring alopecia with thickened skin in a linear pattern [Figure 1].

Dermoscopic examination of the active border showed discrete tufting, perifollicular scaling, erythema, telangiectasia, and tubular silver-white hair casts.

Histological examination revealed hyperplasia of the interfollicular epidermis with hair follicles merging to form hair tufts and a perifollicular-mixed inflammatory infiltrate with a large number of plasma cells [Figure 2a]. In the elastin stain, the dermal elastic network was effaced, consistent with diffuse dermal fibrosis [Figure 2b].

Borrelia serology tested positive for Borrelia burgdorferi IgG with 21.0 U/mL (normal: <16 U/mL) and negative for B. burgdorferi IgM.

The patient was treated with 200 mg oral doxycycline daily and topical clobetasol propionate lotion.

DISCUSSION

FD, as originally described by Quinquaud in 1888, represents a chronic and recurrent pustulofollicular scalp inflammation usually of the crown area with exudative crusted areas and grouped follicular pustules at the hair-bearing margin and centrifugal progression with central scarring. Since Quinquaud’s original report, further clinical variants have been described on the basis of common histopathological and microbiological findings. Histopathology reveals a neutrophilic primary scarring alopecia, and microbiological studies invariably reveal pathogenic strains of Staphylococcus aureus although a Gram-negative bacterial folliculitis may evolve in patients who receive prolonged courses of antibiotic therapy or use antibacterial topicals that selectively inhibit Gram-positive organisms.

In 1947, Laymon described cicatrizng seborrheic eczema as a condition of scarring alopecia combining features of seborrheic dermatitis and FD.

In 1977, Smith and Sanderson and at the same time Metz and Metz reported on tufted hair folliculitis, whereby the former considered the condition to be secondary to follicular inflammation and destruction, while the latter deemed the inflammation to be secondary to preexisting nevoid hair bundles. Since the original report of tufted hair folliculitis, there has been considerable controversy whether the condition represents a distinctive entity or an unspecific finding secondary to a variety of inflammatory and scarring alopecias. Based on clinical, histopathological, and microbiological criteria, the general consensus is that the

Figure 1: White porcelain-colored plaque of scarring alopecia with thickened skin in a linear pattern

Figure 2: (a) Histopathology (H and E): Hyperplasia of the interfollicular epidermis with hair follicles merging to form hair tufts, and a perifollicular-mixed inflammatory infiltrate with a large number of plasma cells. (b) Elastin stain: effaced dermal elastic network, consistent with diffuse dermal fibrosis
condition represents a variant of FD. Secondary tufting of hair follicles due to other inflammatory conditions of the scalp is usually less marked with <5 hair shafts per tuft, and dermoscopic features allow for differentiation. Histopathological studies reveal perifollicular inflammation around the upper portions of the follicles sparing the hair root level. Within areas of inflammation, several follicles converge toward a common follicular duct with a widely dilated opening. It is believed that the development of atrophy with loss of adnexal structures in classical FD or of hair tufts in tufting folliculitis depends on the depth and destructive potential of the inflammatory process, with sparing of the bulge area preserving an intact lower portion of hair follicles in the case of tufted hair folliculitis.

Very rarely, FD can present with an extensive ulcerated appearance of the scalp that has been reported to have been misinterpreted as a laceration of the scalp at the forensic scene.

Ultimately, the pseudopeladic state as originally defined by Degos in 1954 represents the nonspecific end stage of a variety of at least 60 types of cicatricial alopecias, including FD. It presents with a large area of scarring with irregular borders.

The pathogenesis and clinical presentation of FD are determined by three factors: (1) the infectious pathogen, (2) the incubatory microenvironment, and (3) the host immune response.

Occasionally, FD has been linked to a specific immune deficiency, ectodermal dysplasia, or hair transplantation, either autologous or synthetic, impairing the immune defense mechanisms of the hair follicle. Traditionally, the medical focus has been on the condition of either the hair or the scalp. Indeed, the proximate structural arrangement of the scalp and hair leads to an interdependent relationship between the two. The role of the scalp as an incubatory environment has only recently received appropriate attention. In fact, seborrhea, dandruff, and seborrheic dermatitis of the scalp are frequently associated with hair loss and superficial pustular folliculitis that nevertheless may eventually evolve to cicatrizing seborrheic eczema.

Chiarini et al. proposed that the infection of hair follicles with S. aureus induces an intense peri- and intrafollicular migration of neutrophils, recruited by innate immunity mechanisms, involving interleukin-8 (IL-8). Furthermore, T-lymphocytes may be activated either by microbial antigens through processing by Langerhans cells or by superantigens through the Vβ domain of the T-cell receptor with consecutive release of pro-inflammatory such as interferons alpha-gamma and tumor necrosis factor-alpha and profibrotic mediators such as transforming growth factor-beta, beta-fibroblast growth factor, IL-1β, and II-4, resulting in both inflammation and fibrosis.

The observation of simultaneous occurrence of secondary cutis verticis gyrata, FD, and folliculitis keloidalis nuchae in a male patient of African origin with dreadlocks underlines a putative pathogenic role of traction or trauma and the activation of different fibroblast growth factor members.

According to Pujol's hypothesis, tufted hair folliculitis observed in FD and in folliculitis keloidalis nuchae is secondary to follicular damage and caused by inflammatory cytokine secretion. Accordingly, tufted hair folliculitis has been reported following scalp injury (scalp laceration).

A more recent study provided evidence of the presence of bacterial biofilms in the infrainfundibular part of human scalp hair follicles in FD. A biofilm is any group of microorganisms in which cells stick to each other on a surface. These adherent cells are frequently embedded within a self-produced matrix of extracellular polymeric substance. Biofilms form on living or nonliving surfaces and in the case of FD on the surfaces of the hair shaft. Bacteria living in a biofilm usually have significantly different properties from free-floating bacteria of the same species, as the dense and protected environment of the film allows them to cooperate and interact in various ways. One benefit of this environment is increased resistance to antibiotics, as the dense extracellular matrix and the outer layer of cells protect the interior of the community. Biofilms have been found to be involved in a wide variety of microbial infections in the body, and more recently, it has been noted that bacterial biofilms may impair cutaneous wound healing and reduce topical antibacterial efficiency in healing or treating infected skin wounds. The presence of a bacterial biofilm at the interface of the hair shaft may provide an explanation for the chronicity and high relapse rate of FD.

With time, FD tends to develop clinical and dermoscopic features of lichen planopilaris (LPP). More important tufting, and on histopathology, a more diffuse pattern of effaced dermal elastic fibers versus selective loss of elastic fibers at the site of selectively destroyed hair follicles, in combination with a more important number of plasma cells in the inflammatory infiltrate helps distinguish late phases of FD from LPP. It can be assumed that through the destruction of hair follicles in the course of the primary
infectious disease, follicular antigens are exposed and give rise to an autoimmune reaction. In fact, LPP is regarded to be a T cell-mediated autoimmune reaction in response to some antigenic challenge with apoptosis of the follicular epithelial cells. Harries et al.\[28\] provide the first evidence that LPP may result from an immune privilege collapse of the hair follicle’s epithelial stem cell niche. Where a causal or triggering agent is identified, this is termed a lichenoid reaction rather than lichen planus; therefore, the term chronic lichenoid phase of FD is proposed for this presentation of FD.

We report yet another presentation of FD, clinically mimicking linear circumscribed scleroderma of the scalp.

Linear circumscribed scleroderma or linear morphea of the scalp is a form of localized scleroderma that characteristically affects the scalp to produce a line of ivory- or porcelain-colored thickened skin with loss of hair. The thickening and hardening of the skin and underlying subcutaneous tissue result from excessive collagen deposition. On histopathological examination, the dermal elastic network remains preserved.\[26\] The cause is unknown although reports of morphea coexisting with other systemic autoimmune diseases support that morphea is also a condition of autoimmune origin. It has been proposed that linear morphea follows Blaschko’s lines, giving rise to the hypothesis that in patients with linear morphea, susceptible cells are present in a mosaic state, and exposure to some trigger factor, including trauma, may result in the development of this condition.\[29\]

Kim et al.\[30\] originally reported tufted hair folliculitis in a linear arrangement and again discussed Pujol’s trauma hypothesis. We present the second case of histopathologically proven FD in linear arrangement, though with a linear circumscribed scleroderma-like appearance due to thickening of the epidermis. The presence of thickening of lesional skin in FD has been previously commented on. In a retrospective histological analysis of 26 patients with FD, the authors found follicular hyperkeratosis in 77%, hyperplasia of the interfollicular epidermis in 92% with a psoriasiform aspect in 88%, plasma cells in infiltrate in 92%, in 42% in large quantities, and hair tufting in 54%.\[31\]

We propose adding linear circumscribed scleroderma-like FD to the protean clinical manifestations of FD, and as a subset of FD with linear arrangement, as presented in Table 1.

### Table 1: Clinical variants of folliculitis decalvans

| Condition | Description |
|-----------|-------------|
| Classical folliculitis decalvans of Quinquaud | |
| Cicatrizing seborrhoeic eczema | |
| Tufted hair folliculitis | |
| Ulcerative folliculitis decalvans | |
| Folliculitis decalvans with linear arrangement | Tufted hair folliculitis with linear arrangement |
| Linear scleroderma-like folliculitis decalvans | |
| Folliculitis decalvans associated with immune deficiency | |
| Ectodermal dysplasia with clefing | |
| Cutis verticis gyrata and folliculitis keloidalis nuchae | |
| Hair transplantation, either autologous or synthetic | |
| End-stage folliculitis decalvans | |
| Chronic lichenoid phase of folliculitis decalvans | |
| Pseudopeladic state (of Degos) resulting from folliculitis decalvans | |

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### Conflicts of interest

There are no conflicts of interest.

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