

## Cicatricial Alopecia as a Manifestation of Different Dermatoses

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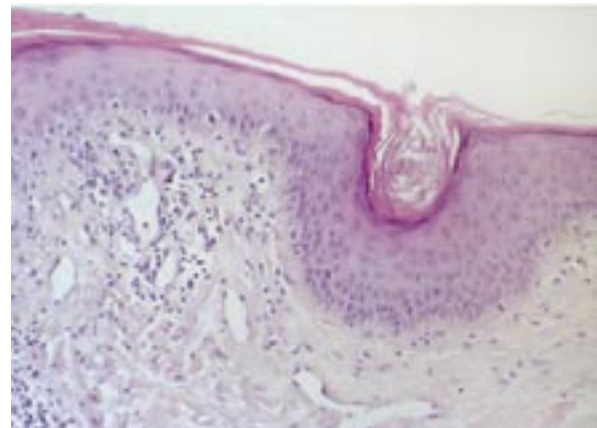
**SUMMARY** There are numerous dermatoses which may cause cicatricial alopecia when localized on the scalp, such as chronic discoid lupus erythematosus (DLE), lichen planus, graft-versus-host disease, dermatomyositis, scleroderma, cicatricial pemphigoid, porphyria cutanea tarda, follicular mucinosis, perifolliculitis capitis abscedens, lichen sclerosus et atrophicus, necrobiosis lipoidica, sarcoidosis, etc. Histologically, cicatricial alopecia is characterized by dermal scarring, along with absent or reduced hair follicles and reduced number of erector pili muscles. According to working classification of cicatricial alopecia by the North American Hair Society, primary cicatricial alopecia may be divided into the following categories: lymphocytic group (e.g., DLE, lichen planopilaris, classic pseudopelade (Brocq), central centrifugal cicatricial alopecia); neutrophilic group (e.g., folliculitis decalvans, dissecting cellulitis); and mixed group (e.g., folliculitis keloidalis). Over a 5-year period, 36 patients with cicatricial alopecia were hospitalized at our Department: DLE (n=27), pseudopelade Brocq (n=3), mucinosis follicularis (n=2), and lichen planopilaris, folliculitis decalvans, folliculitis abscedens and folliculitis keloidalis (one patient each). Clinical evaluation was compared with histopathologic analysis of follicular architecture, as well as with the type, localization and extent of inflammatory infiltrate. Scalp biopsy was considered mandatory in all cases. Our experience indicates the need of more complex research to extend the knowledge about the etiopathogenesis and treatment options for cicatricial alopecia. We hope that this type of alopecia may attract more attention and research in the future.

**KEY WORDS:** cicatricial alopecia; discoid lupus erythematosus; pseudopelade; histopathologic analysis

### INTRODUCTION

Besides noncicatricial (nonscarring) forms, there is "scarring" (cicatricial) alopecia where follicular epithelium is replaced by connective tissue, leading to an irreversible process of scarring (1). Primary cicatricial alopecias encompass a group

of disorders characterized by permanent destruction of the hair follicle (2). They include destruction of the hair follicle with inflammatory process predominantly around the permanent portion of the hair follicle (stem cells of the bulge and the



**Figure 1.** Lupus erythematosus chronicus: hyperkeratosis, dilated follicular ostia filled with keratine, liquefaction degeneration of basal keratinocytes, incontinentia pigmenti, predominantly lymphocytic infiltration at the dermal-epidermal border as well as pericapillary and periadnexal

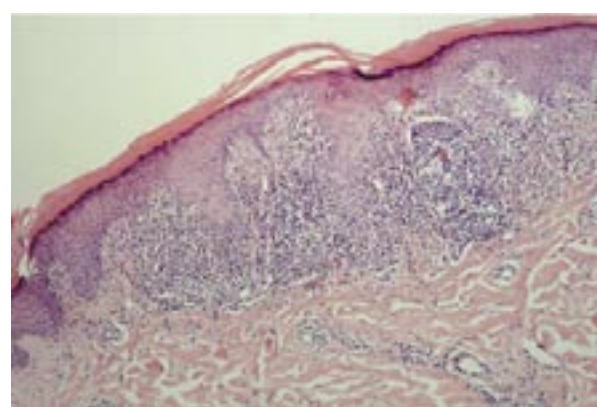
infundibulum), hence resulting in irreversibility of alopecia (3-6).

The various clinical features and differences in terminology have led to difficulties in defining consistent clinicopathologic correlation (2). There are a series of dermatoses which may cause cicatricial alopecia when localized on the scalp, such as chronic discoid lupus erythematosus (DLE), lichen planus (LP), graft-versus-host disease (GVHD), dermatomyositis (in its chronic, poikilodermic phase), scleroderma (especially in the *en coup de sabre* form), cicatricial pemphigoid, porphyria cutanea tarda, perifolliculitis capitis abscedens, lichen sclerosus et atrophicus, necrobiosis lipoidica, sarcoidosis, follicular mucinosis, etc. (3,7,8).

Histologically, cicatricial alopecia is characterized by dermal scarring, often relatively deep, along with absent or reduced hair follicles and reduced number of erector pili muscles. They represent a diverse group of diseases characterized

by the lack of follicular ostia and irreversible alopecia. Currently, various classification schemes are available for cicatricial alopecias. Thus, cicatricial alopecias may be classified into congenital cicatricial alopecias (genodermatoses: Darier's disease, sex linked ichthyosis, dystrophic epidermolysis bullosa, incontinentia pigmenti, polyostotic fibrous dysplasia, HHHH syndrome (hypotrichia, hyperkeratosis, hydrocystomas, hypodontia), obstetric traumas, nevi (organoid and epidermal), simplex hereditary hypotrichoses, follicular keratosis, aplasia cutis congenita, etc.

Another group of cicatricial alopecia are acquired cicatricial alopecias. This type of alopecia may be the result of physical traumas (chronic traction, avulsion, electrical or thermal burns, freezing, ionizing radiation, neurotrophic ulcers), chemical traumas (alkali, acids), cutaneous infections (fungal, viral, bacterial, zooparasitoses), or tumors (benign, malignant). Acquired cicatricial alopecias



**Figure 2.** Lichen planopilaris: follicular hyperkeratosis and dilated follicular ostia, typical lichenoid striped infiltration which erodes basal layers of the epidermis, incontinentia pigmenti

Table 1. Distribution of alopecia areata in our patients according to histopathological findings

CICATRICAL ALOPECIA-TYP	DISEASE	No/ %	SEX	AGE	AVERAGE AGE
			F:M		
LYMPHOCYTIC	Lupus erythematosus chronicus	27/75	19:8	20-70	49,9
	Lichen planopilaris	1/2,8	F	31	
	Pseudopelade	3/8,3	F	40-54	49,3
NEUTROPHILIC	Folliculitis decalvans	1 / 2,8	M	24	
	Folliculitis abscedens	1/2,8	M	20	
MIXED	Folliculitis keloidalis	1 / 2,8	M	37	
MUCINOUS/CICATRICAL	Follicular mucinosis	2/5,5	1:1	42,57	49,5

also include acquired cicatricial alopecias in special dermatoses (primary cicatricial alopecia), such as DLE, LP, GVHD, sarcoidosis, linear morphea, porphyritic alopecia, perifolliculitis capitis abscedens et suffodiens, mucinosis follicularis, pseudopelade Brocq, Pinku's fibrosing alopecia, erosive pustular dermatosis of the scalp, etc.

Currently, there is also a classification scheme of cicatricial alopecias according to the North American Hair Society (NAHRS) (7). According to the working classification of cicatricial alopecia, primary cicatricial alopecia can be divided into the following categories: lymphocytic group (e.g., DLE, lichen planopilaris (LPP), classic pseudopelade (Brocq), central centrifugal cicatricial alopecia); neutrophilic group (e.g., folliculitis decalvans, dissecting cellulitis); and mixed group (e.g., folliculitis keloidalis). There is also a group of nonspecific cicatricial alopecia, defined as an idiopathic scarring alopecia with inconclusive clinical and histopathologic findings (7).

The aim of this study was to determine the causes of scarring alopecia by clinical and histopathologic characteristics in patients hospitalized at our Department during a 5-year period.

### PATIENTS AND METHODS

The study was conducted at University Department of Dermatology and Venereology, Sestre milosrdnice University Hospital, Zagreb, Croatia. The patients with scarring alopecia were monitored over a 5-year period (2001-2005). We performed prospective clinical and histopathologic evaluation of clinically typical primary cicatricial alopecias. Biopsy specimens were obtained from early stages of alopecia lesions.

Over the 5-year period, 36 patients with cicatricial alopecia were hospitalized at our Department (Table 1). A total of 36 patients with cicatricial alopecia were included in the study. The examina-

tion included history data, duration of the disease, previous treatment, trichogram, antinuclear antibodies, histopathologic analysis, and direct immunofluorescence. The diagnoses were set upon analysis of these data.

### RESULTS

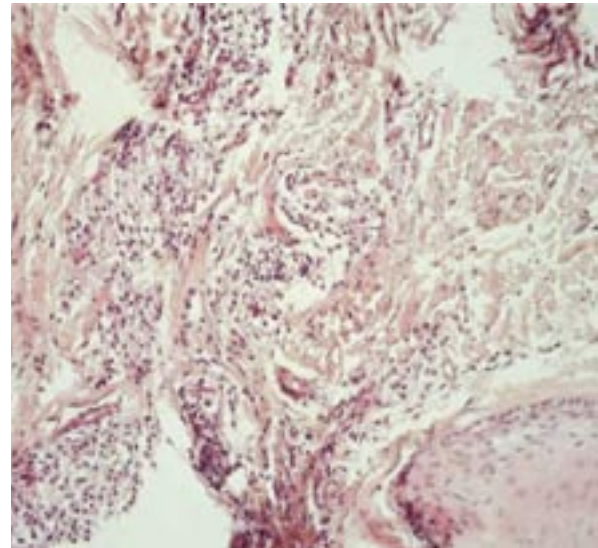
Over the 5-year period, we treated 36 patients with cicatricial alopecia by the previously mentioned procedures: DLE (n=27), pseudopelade Brocq (n=3), mucinosis follicularis (n=2), lichen planopilaris (n=3), and folliculitis decalvans, folliculitis abscedens and folliculitis keloidalis (one patient each) (Table 1). Thus, most of our patients had DLE (n=27), followed by pseudopelade Brocq (n=3) and mucinosis follicularis (n=2). Results of clinical and histopathologic examinations are shown in Figures 1-6. Patients were treated according to recommendations for the respective histologic type of alopecia (Table 2).

### DISCUSSION

The term cicatricial alopecia represents a group of disorders characterized by the final pathway of replacement of the follicular structure by fibrous tissue (9). In dermatological practice, the term cicatricial alopecia is considered convenient because it is clear from the beginning that there

Table 2. Therapy recommendations

LYMPHOCYTIC CICATRICAL ALOPECIA	NEUTROPHILIC/MIXED CICATRICAL ALOPECIA
Corticosteroids	Antibiotics
Antimalarials	Isotretinoin
Isotretinoin	Topical minoxidil
Methotrexate	Surgical reduction
Topical minoxidil	
Hair transplantation	

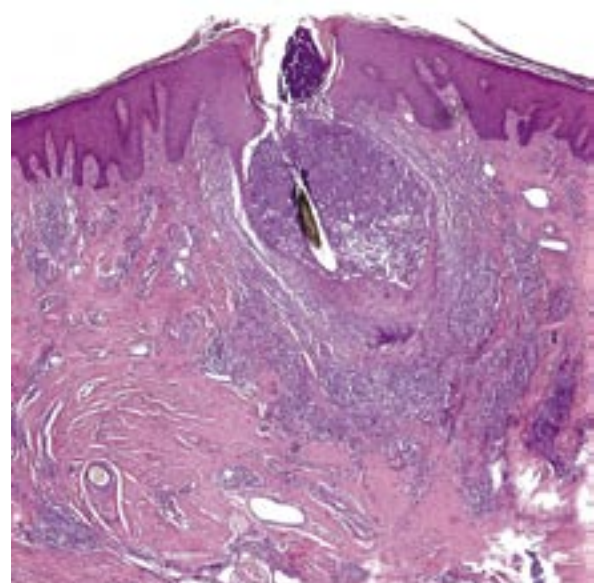
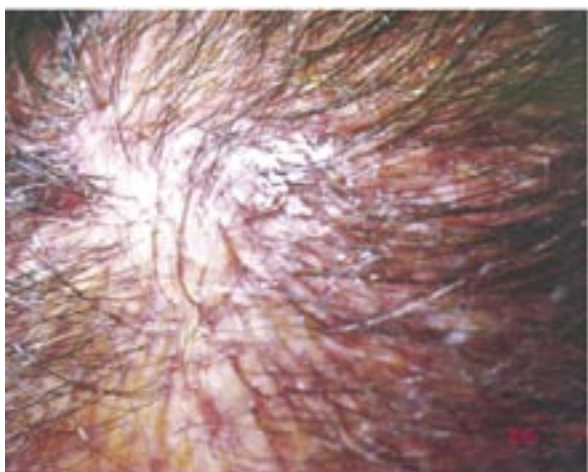


**Figure 3.** Pseudopelade Brocq: extensive lymphocytic perifollicular infiltration leading to the replacement of follicle with the fibrous streak

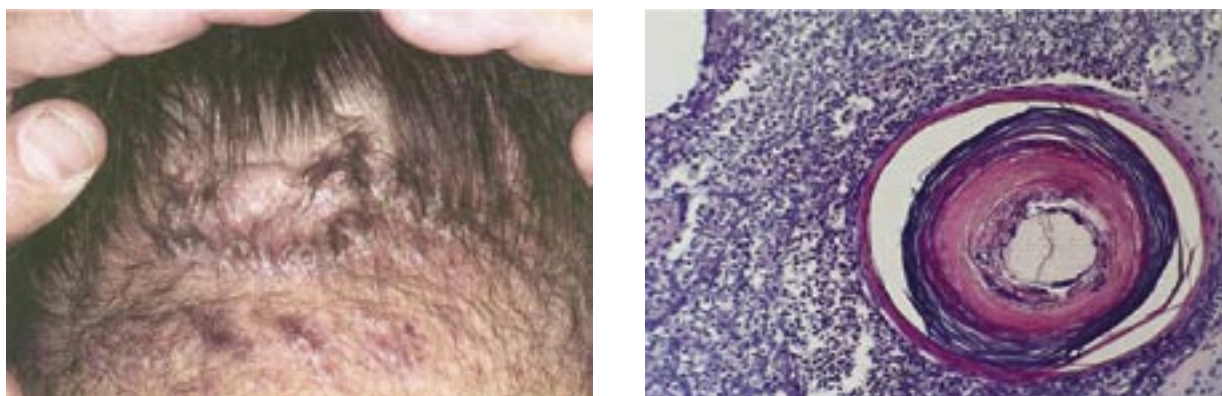
are no follicles, as they have been replaced by fibrosed or slack dermal tissue (3). There are different hypotheses based on the etiological factors involved in the development of cicatricial alopecia (9). It has been suggested that cicatricial alopecias have a genetic component (predisposition), but environmental factors also play a role in its development, including chemical contributions and infective factors (viral, bacterial, fungal). Experimental studies have also confirmed cicatricial alopecias in laboratory mice (spontaneous or induced), similar to that seen in humans. There is some evidence that cicatricial alopecia results from a defect in the hair follicle structural proteins or an abnormal immune response. There are also some arguments indicating that cicatricial alopecia is a secondary

result of pathologic fragments of follicle-nutrient vessels and/or nerves, or the result of disruption in the mesenchymal/epithelial interactions which are necessary for normal cycling (9). However, these etiologic factors have not yet been elucidated and need to be tested, confirmed or excluded in the future.

Besides primary cicatricial alopecias, there are several causes of secondary cicatricial alopecia such as certain types of dermatophyte infections, infiltrative processes (e.g., metastatic cancers), and trauma (e.g., radiation or burns). There are



**Figure 4.** Folliculitis decalvans: destructive chronic or subacute folliculitis with perifollicular chronic inflammatory cells



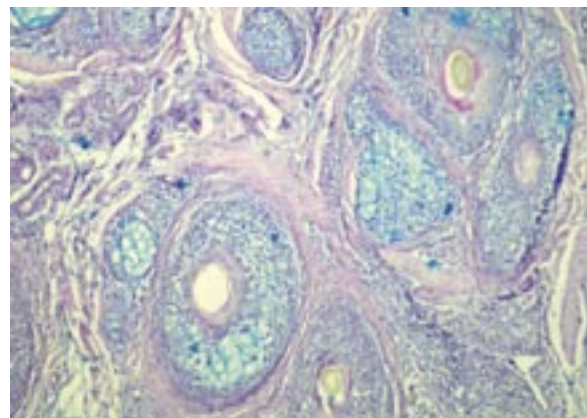
**Figure 5.** Folliculitis abscondens: destructive acute folliculitis with neutrophils and suppuration

many other environmental factors suspected of invoking or contributing to the development of permanent alopecia, such as chemicals used to straighten or curl hair in selected susceptible populations (9). The classification of primary cicatricial alopecias on the basis of pathology provides further diagnostic and investigational procedures (10).

However, most of our patients with cicatricial alopecias were those with DLE. In chronic DLE, cicatricial alopecia is the result of chronic inflammatory infiltrate which destroys the follicle and cutaneous adnexa. It is present in the form of single or multiple patches of variable size, with pigmented periphery and atrophic central skin, which tends to grow slowly. It has been noticed that 35%

of DLE patients have cicatricial alopecias, long-standing DLE in almost all of them (11). One can also find this type of alopecia in systemic LE (11). Histologically, there is a reduction in the size of sebaceous glands with perifollicular lymphocytic infiltrate, which is more obvious in the middle portion at the level of sebaceous gland (12). Classic treatment includes topical fluorinated corticosteroids, systemic or intralesional corticosteroids, but antimalarial drugs and sulfones may be effective as well. Recently, it has been reported that the use of thalidomide in this process may result in regression of alopecic patches on the scalp (13). Pseudopelade is non-inflammatory progressive and idiopathic cicatricial alopecia, which is characterized by small erythematous alopecic areas located on the scalp tending to confluence. Thus, a larger alopecic patch with dentated edges is formed, with some centrally dispersed hairs, occasionally with true retractile follicular depressions and intact peripheral hair implantation line (13,14).

Follicular mucinosis (alopecia mucinosa) is characterized by very pruriginous infiltrated ery-



**Figure 6.** Follicular mucinosis: pilosebaceous degeneration associated with the deposits of mucine I, with inflammatory infiltrates and histiocytes, and variable number of eosinophils

thematous patches, consisting of a group of follicular papules, localized on the face, shoulders, neck and scalp, and causing definite alopecia (3). In approximately 20% of cases it accompanies mycosis fungoides, considered to be an early stage of mycosis fungoides in elderly people (15). A linear form of mucinosis follicularis has been described on the scalp, following Blasckho's lines, associated with lichen striatus, a process in which one can see follicular mucinosis (16). Dermatopathologically, one can see mucine in the outer root sheath as well as in sebaceous glands.

Scalp involvement within LP leads to an inflammatory process with characteristic erythematous infiltrates around the follicular openings associated with scarring alopecia, lichen planopilaris (3,8). The cutaneous features are typical and therefore easy to diagnose clinically and dermatopathologically, but only in the early stages of the diseases. Other rarer cases of cicatricial alopecias in our patients included folliculitis, such as folliculitis decalvans, folliculitis abscedens and folliculitis keloidalis.

We consider that in patients with cicatricial alopecias it is important to examine clinical and pathologic features along with therapeutic options for the disease (10). It is very useful to perform histopathologic analysis of follicular architecture, type, location, and extent of the inflammatory infiltrate, as well as the presence or absence of sebaceous glands (2). It is also important to distinguish the role of pathological findings of cicatricial alopecias, the histological type of cicatricial alopecia (lymphocytic or neutrophilic) in particular, which has great influence on therapy. Analyzing the types of cicatricial alopecia among our patients, we found a greater proportion of lymphocytic cicatricial alopecias *versus* neutrophilic cicatricial alopecias, which is consistent with the data from the UBC Hair Clinic and results of Whiting (he found a proportion of 4:1 (4,7).

There is a variety of therapeutic modalities for patients with lymphocytic cicatricial alopecias (7). These include potent topical (class I or II), intralesional or oral corticosteroids; antimalarials (chloroquine or hydroxychloroquine); and isotretinoin. In our study, the patients were mostly treated with potent topical and/or intralesional corticosteroids (triamcinolone acetonide). The patients with active disease or rapid progression may be treated by systemic corticosteroids (prednisone, initial dose of 1 mg/kg with gradual tapering over 6 to 8 weeks). Intermittent short courses of prednisone

were frequently required to control acute flares of both LE and LPP (7). Steroids are frequently combined with other systemic agents to achieve remission.

Generally, the treatment of neutrophilic cicatricial alopecia includes broad-spectrum antibiotics (tetracyclines, minocycline, cloxacillin, erythromycin, or broad-spectrum cephalosporin) as first choice therapy. Frequently intralesional/potent corticosteroids may be added to curb acute inflammation (7).

## CONCLUSION

The high variability in the clinical presentation of cicatricial alopecias makes the diagnosis and treatment a challenging issue in everyday dermatological practice. An accurate diagnosis may be achieved through attentive clinicopathologic evaluation with histopathologic analysis of follicular architecture and changes of sebaceous glands, and type, location and extent of the inflammatory infiltrate. We consider scalp biopsy mandatory in all cases. Our experience indicates the need of more complex research to extend the knowledge about the etiopathogenesis and treatment possibilities in cicatricial alopecia. We hope that this type of alopecia may attract more attention and research efforts in the future.

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Elida cream, for skin care; year 1935.  
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