Scarring Alopecia

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Proposed Working Classification Primary Cicatricial Alopecia*

- Lymphocytic
- Neutrophilic
- Mixed
- Nonspecific (idiopathic)

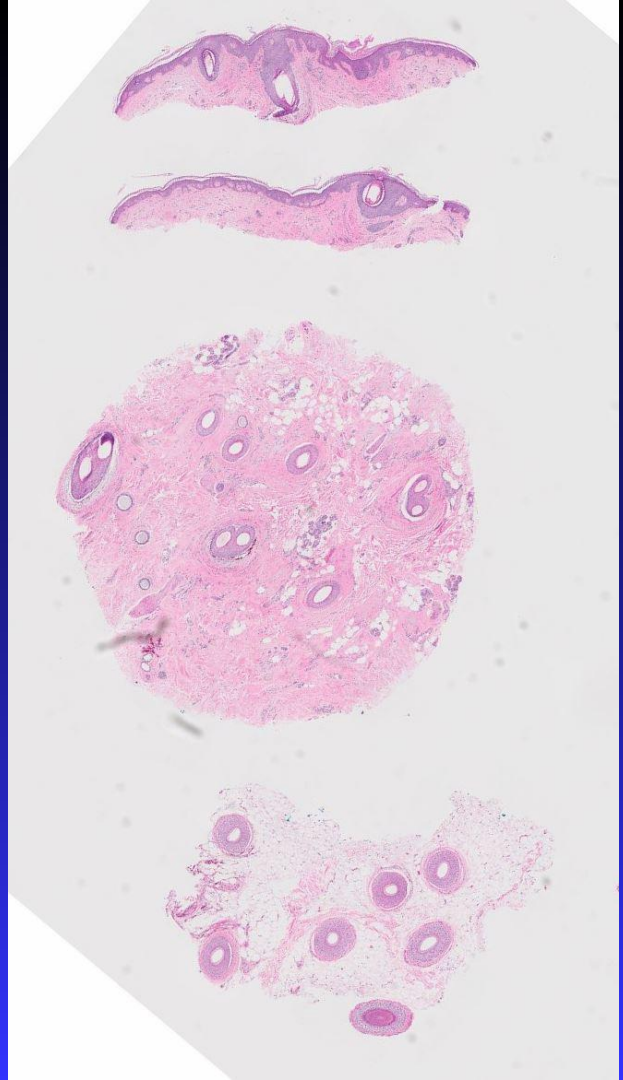
*Olsen E *et al. Summary of North American Hair Research Society (NAHRS) sponsored workshop on cicatricial alopecia, Duke University Medical Center February 10 and 11, 2001. JAAD 48:103-10, 2003.*

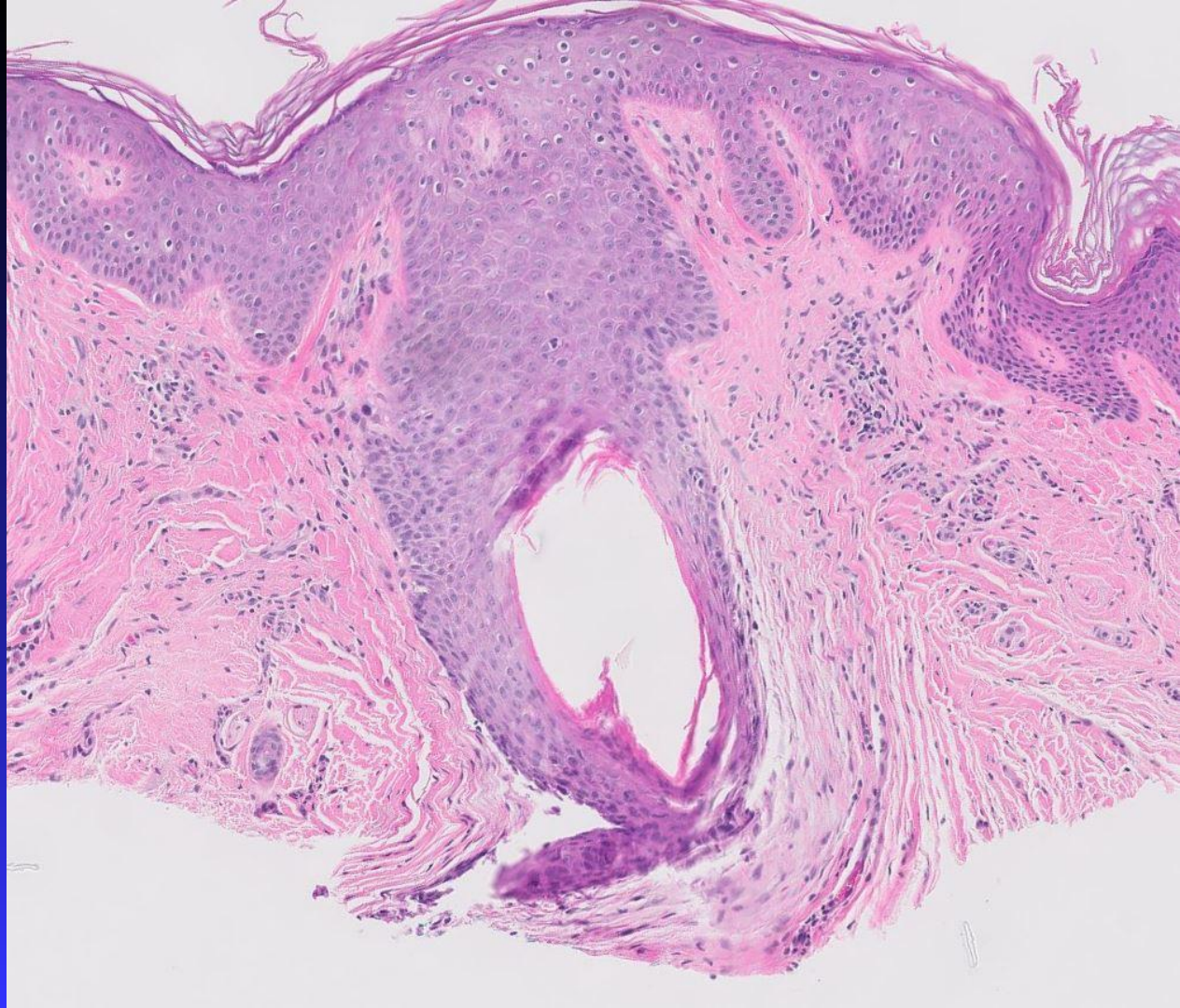
Proposed Working Classification Primary Cicatricial Alopecia

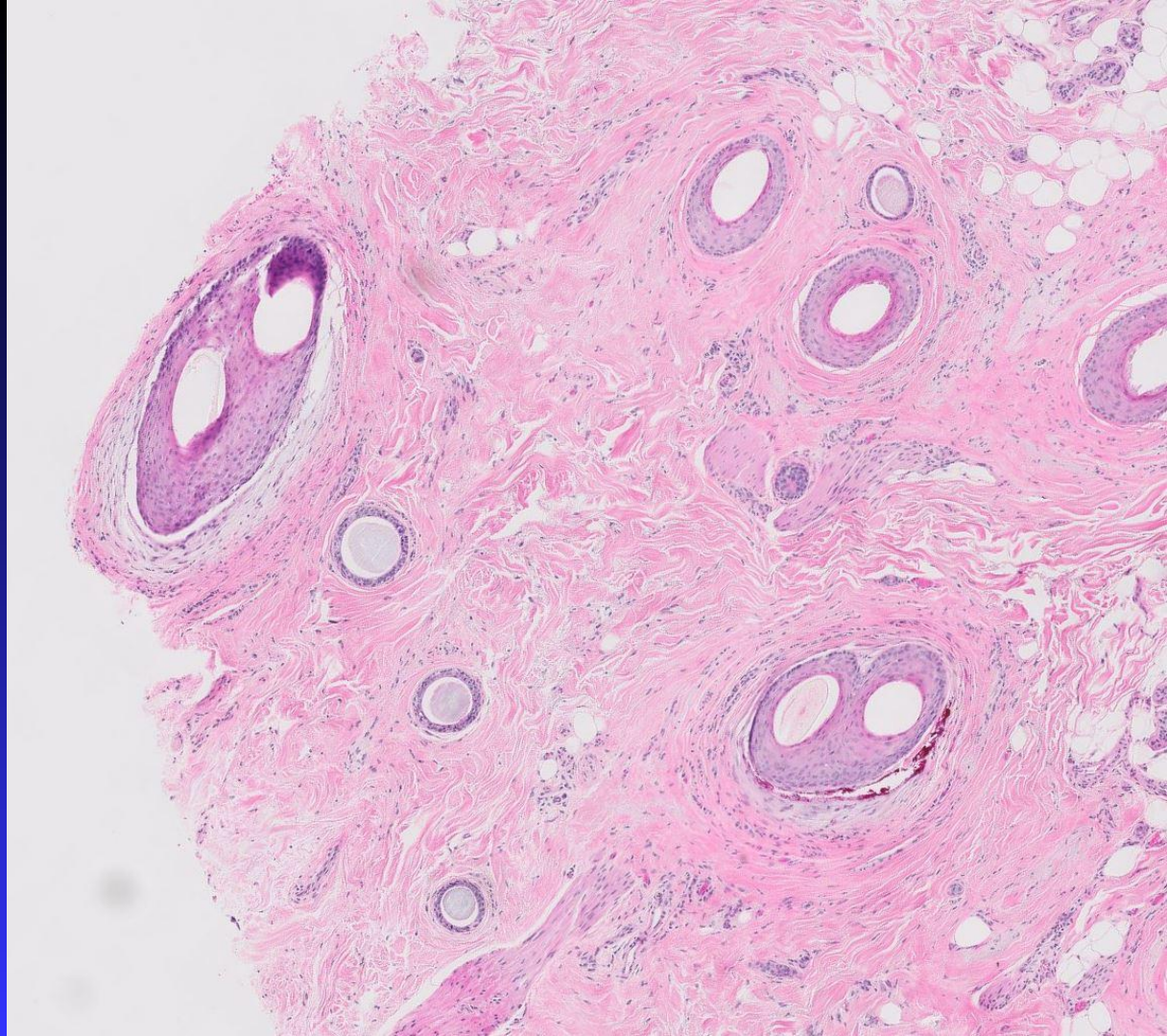
- Lymphocytic
  - Chronic cutaneous lupus erythematosus
  - Lichen planopilaris
    - Classic
    - Frontal fibrosing
    - Graham-Little syndrome
  - Classic pseudopelade (Brocq)
  - Central centrifugal cicatricial alopecia
  - Alopecia mucinosa
  - Keratosis follicularis spinulosa decalvans.

Proposed Working Classification Primary Cicatricial Alopecia

- Neutrophilic
  - Folliculitis decalvans
  - Dissecting cellulitis/folliculitis (perifolliculitis abscedens et suffodiens)
- Mixed
  - Folliculitis (acne) keloidalis.
  - Folliculitis (acne) necrotica.
  - Erosive pustular dermatosis.





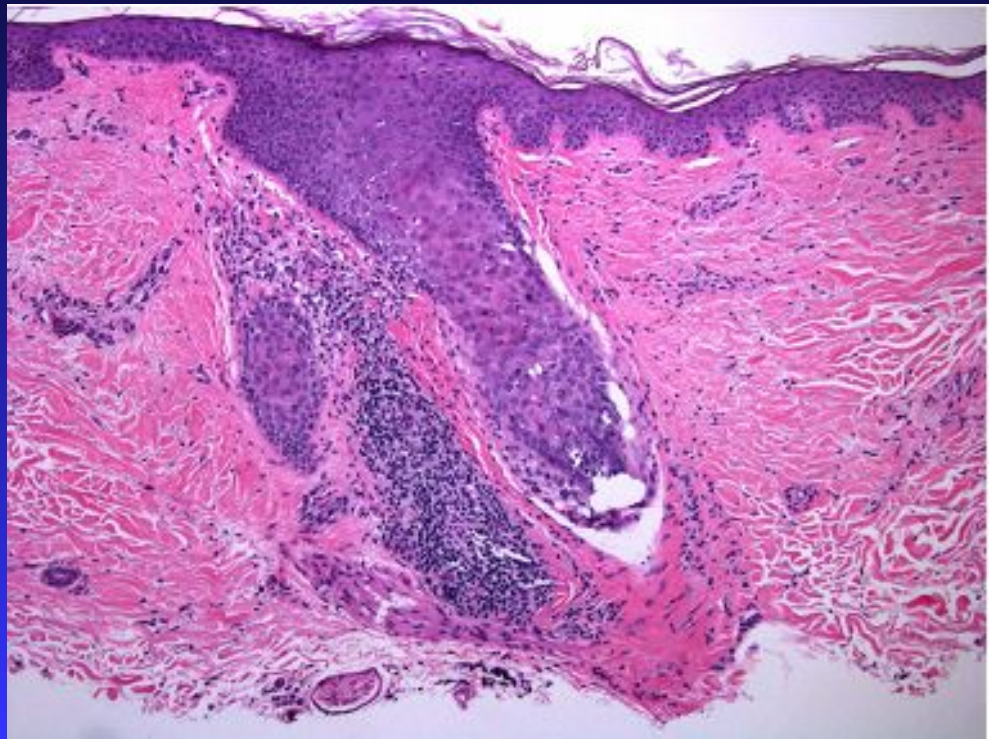


Frontal fibrosing variant of lichen planopilaris

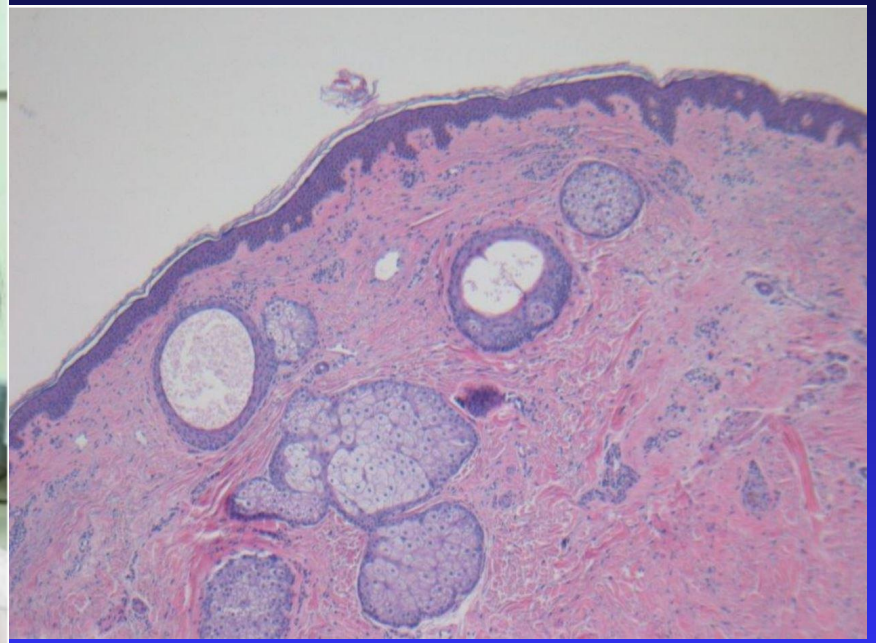
- Superficial perifollicular lymphocytes at the level of the infundibulum or isthmus
- Gray-staining, perifollicular fibrosis may or may not be present
- Follicular loss (low follicular density) is usually present. On the eyebrows, follicular epithelium may be entirely lost before any perifollicular fibrosis forms.

Frontal fibrosing variant of lichen planopilaris

- Fibrosis may be absent
- May require close clinical correlation
  - Hairline recession
  - ‘Lonely hairs’
  - Eyebrow loss
  - Facial papules



Papular eruption in FFA



Frontal fibrosing variant of lichen planopilaris

For 2mm punches
of eyebrows

Embed skin surface down
Level through block with unstained

Frontal fibrosing variant of lichen planopilaris

- Differential
  - Female pattern hair loss (androgenetic alopecia) with superimposed seborrheic dermatitis
  - Alopecia areata

Frontal fibrosing variant of lichen planopilaris

- Incidence increased markedly in 1990s
- Most perimenopausal women but also younger and in men
- Targets smaller hairs of body (eyebrow, body hair)

Br J Dermatol. 2016 Oct;175(4):762-7. doi: 10.1111/bjd.14535. Epub 2016 Jun 30.

Frontal fibrosing alopecia: possible association with leave-on facial skin care products and sunscreens; a questionnaire study.

Aldoori N[1], Dobson K[1], Holden CR[1], McDonagh AJ[1], Harries M[2], Messenger AG[3].

Author information

Abstract

BACKGROUND: Since its first description in 1994, frontal fibrosing alopecia (FFA) has become increasingly common, suggesting that environmental factors are involved in the aetiology.

OBJECTIVES: To identify possible causative environmental factors in FFA.

METHODS: A questionnaire enquiring about exposure to a wide range of lifestyle, social and medical factors was completed by 105 women with FFA and 100 age- and sex-matched control subjects. A subcohort of women with FFA was patch tested to an extended British standard series of allergens.

RESULTS: The use of sunscreens was significantly greater in the FFA group compared with controls. Subjects with FFA also showed a trend towards more frequent use of facial moisturizers and foundations but, compared with controls, the difference in frequencies just failed to reach statistical significance. The frequency of hair shampooing, oral contraceptive use, hair colouring and facial hair removal were significantly lower in the FFA group than in controls. Thyroid disease was more common in subjects with FFA than controls and there was a high frequency of positive patch tests in women with FFA, mainly to fragrances.

CONCLUSIONS: Our findings suggest an association between FFA and the use of facial skin care products. The high frequency of sunscreen use in patients with FFA, and the fact that many facial skin care products now contain sunscreens, raises the possibility of a causative role for sunscreen chemicals. The high frequency of positive patch tests in women with FFA and the association with thyroid disease may indicate a predisposition to immune-mediated disease.

Frontal fibrosing alopecia in men: an association with facial moisturizers and sunscreens

DOI: 10.1111/bjd.15311

Dear Editor, Frontal fibrosing alopecia (FFA) was first described by Kossard in 1994 in six postmenopausal women.[1] FFA remained rare during the 1990s, but in the last 10–15 years it has become increasingly common, a phenomenon observed worldwide. The recent onset and apparently rising incidence of FFA suggest involvement of environmental factors in the aetiology. We previously reported a questionnaire study in women with FFA that asked about a wide range of medical, social and environmental exposures. The results suggested an association between FFA and leave-on facial products, including moisturizers and sunscreens.[2] However, although the regular use of moisturizers was greater in women with FFA, these products are used by most women and we were unable to show a significant difference in their use between women with FFA and similarly aged controls. The use of primary sunscreens was significantly greater among women with FFA than in controls, but we were not able to assess whether patients were also exposed to sunscreens from other sources.

We have therefore repeated our questionnaire study in men with FFA, as we anticipated that their use of leave-on facial skincare products would be lower than in women.

As FFA is rare in men, patients were recruited from across the U.K. and one case was recruited from Belgium. In all cases the diagnosis was made by a clinician with special expertise in hair disease, and it was supported by histology in most cases. The clinical diagnosis was based on scarring alopecia affecting the frontal hairline causing recession of the hairline. Additional features included loss of eyebrows, follicular erythema of the frontal hairline and loss of sideburn and beard hair. Male controls aged 35–80 years were recruited from three sites (Sheffield, Salford and Glasgow). The patients completed a questionnaire similar to that used in our female study, but inviting more detailed information on the use of facial skincare and hair care products. Male patients with FFA were asked about the timing and distribution of hair loss, but otherwise the questionnaires completed by both groups were identical.

Seventeen men with FFA and 73 controls were recruited. The mean age of onset of hair loss in the patients with FFA was 54·5 years (range 35–77). All had loss of hair from the frontal hairline, and 16 (94%) had lost eyebrows. Twelve men (71%) reported loss of hair from the beard and 13 (76%) reported loss of hair from the limbs. All men with FFA reported using facial moisturizers, compared with 40% in the control group. Facial moisturizers were used at least twice a week by 94% of patients with FFA, but by only 32% of controls ($P < 0·001$) (Table 1). Sixteen patients reported using moisturizers for a period consistent with their use prior to the onset of FFA. The use of primary sunscreens by men with FFA was significantly more common than by controls. Overall 35% of men with FFA reported using a sunscreen at least twice a week all year round, compared with 4% of controls ($P = 0·0012$).

When moisturizers containing sunscreen chemicals were included in the analysis, at least 71% of men with FFA applied a product containing a sunscreen at least twice a week all year

Table 1 Reported use of skincare and hair care products by patients with frontal fibrosing alopecia (FFA) and controls

	Patients with FFA	Controls	P-value
Number of patients	17	73	
Age (years), mean (range)	63·1 (42–80)	59·1 (37–79)	
Age at onset of hair loss (years), mean (range)	54·5 (35–77)		
Facial moisturizer[a]	16 (94)	23 (32)	< 0·001
Primary sunscreen[b]	6 (35)	3 (4)	0·0012
Sunscreen[b]	12 (71)	8 (11)	< 0·001
Facial cleanser[a]	4 (24)	5 (7)	0·066
Facial scrub[a]	0	0	
Facial mask[a]	0	0	
Aftershave[a]	7 (41)	28 (39)	1·00
Shampoo[a]	13 (76)	62 (85)	0·27
Conditioner[a]	4 (24)	13 (18)	0·73
Hair spray[a]	1 (6)	2 (3)	0·48
Hair mousse[a]	0	0	
Hair gel[a]	2 (12)	10 (14)	1·00
Hair dye[c]	2 (12)	3 (4)	0·26

Values are n (%) unless stated otherwise. [a]Twice a week or more frequently. [b]Twice a week or more frequently all year round. [c]At least once a year. Sunscreen includes exposure to sunscreen chemicals in primary sunscreens and moisturizers. Analyses were performed after excluding subjects who failed to answer the question. Frequencies in the FFA and control groups were compared using Fisher's exact test.

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Sunscreen in FFA

- Oxybenzone and Avobenzone introduced late 1980s
- Zinc oxide and titanium dioxide
  - Oral lichen planus associated with dental metal

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SOLEIL SYNCHROTRON

Accueil > Actualités > Crème solaire, nanoparticules et alopécie frontale – un lien de cause à effet ?

Crème solaire, nanoparticules et alopécie frontale – un lien de cause à effet ?

L'alopécie frontale fibrosante (AFF) est une pathologie décrite pour la première fois il y a une vingtaine d'années. Elle est caractérisée notamment par une alopécie cicatricielle sur la zone antérieure du cuir chevelu et affecte principalement les femmes après la ménopause. Son origine est inconnue mais, le nombre de cas recensés ne cessant d'augmenter, on suspecte l'implication de facteurs environnementaux qui restent à définir.

Des dermatologues des Hôpitaux Bichat et Rothschild ont, pour la première fois, observé la présence de nanoparticules de dioxyde de titane (TiO_2) le long de follicules pileux d'une patiente atteinte d'AFF qui utilise quotidiennement, depuis 15 ans, des écrans solaires contenant du TiO_2. .

Brunet-Possenti F *et al.* Detection of titanium nanoparticles in the hair shafts of a patient with frontal fibrosing alopecia. JEADV 2018 Mar26, doi; 10.1111/jdv.14967 (Epub ahead of print).

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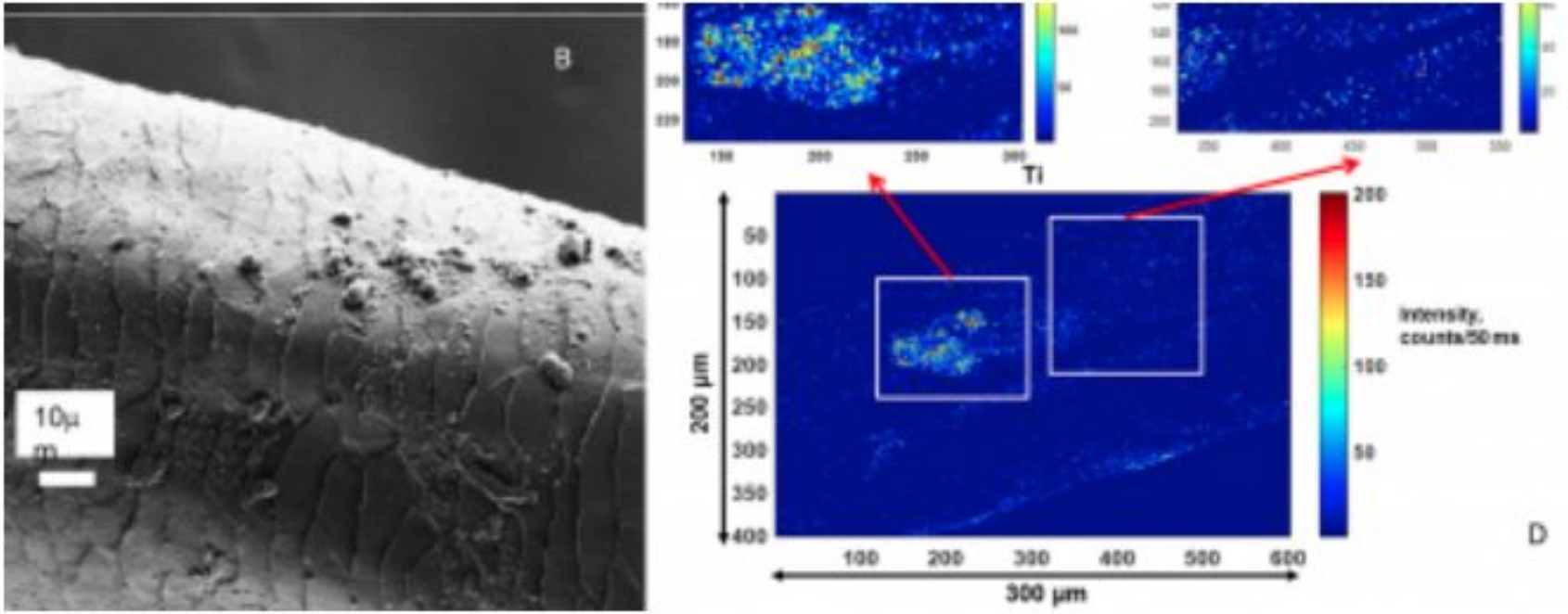

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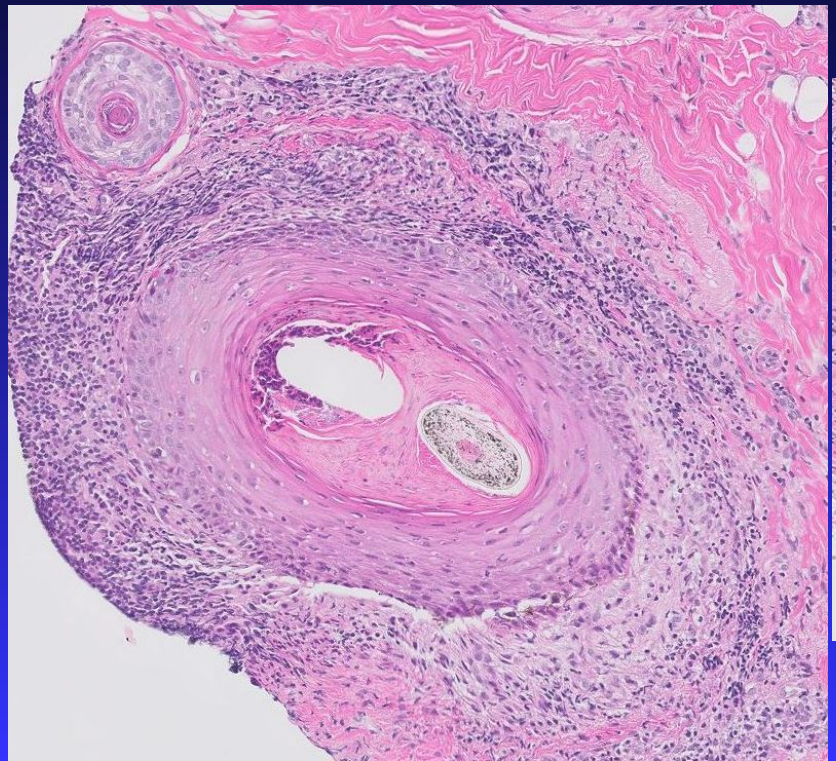
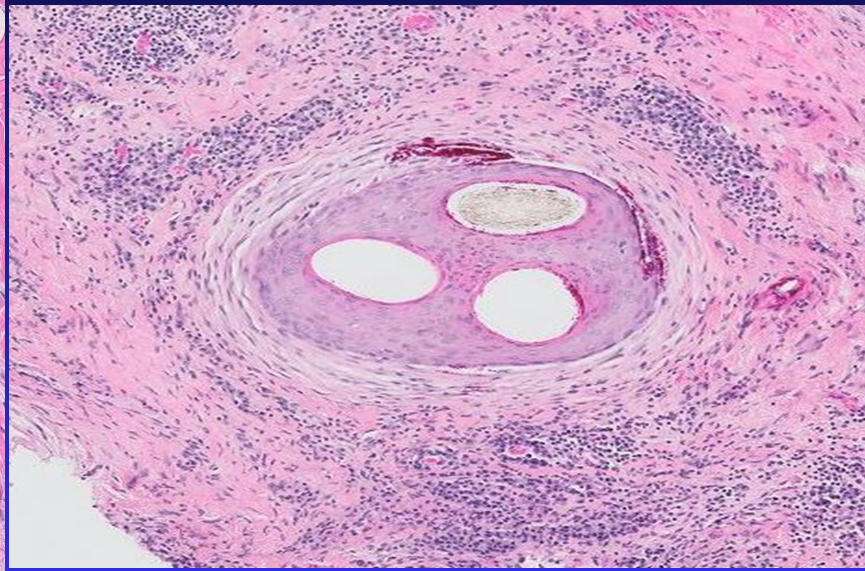
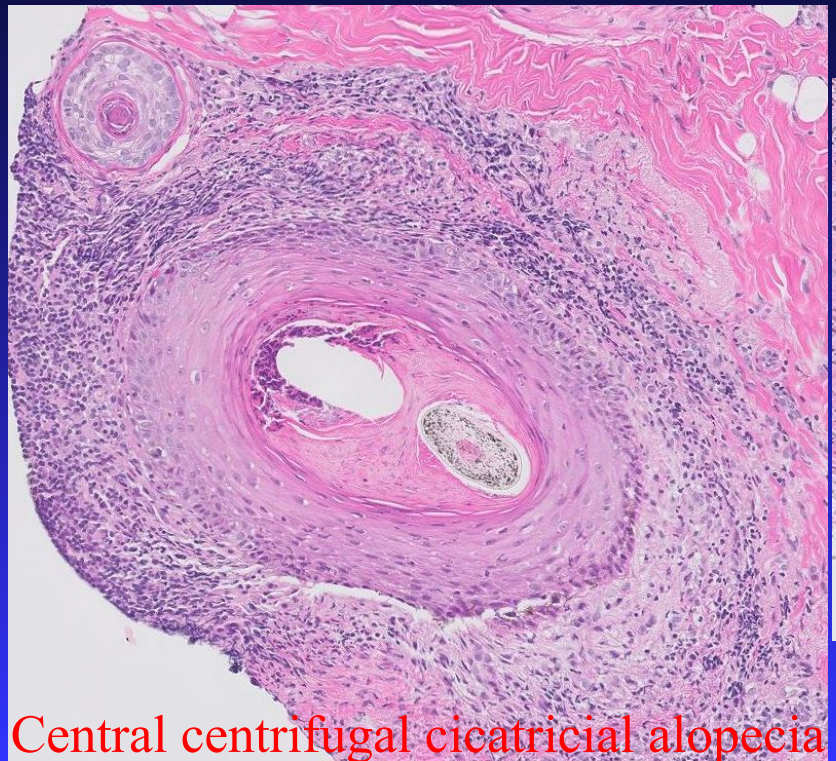
- 53 y/o female of African descent with an alopecic patch on crown of head

Central Centrifugal Cicatricial Alopecia CCCA

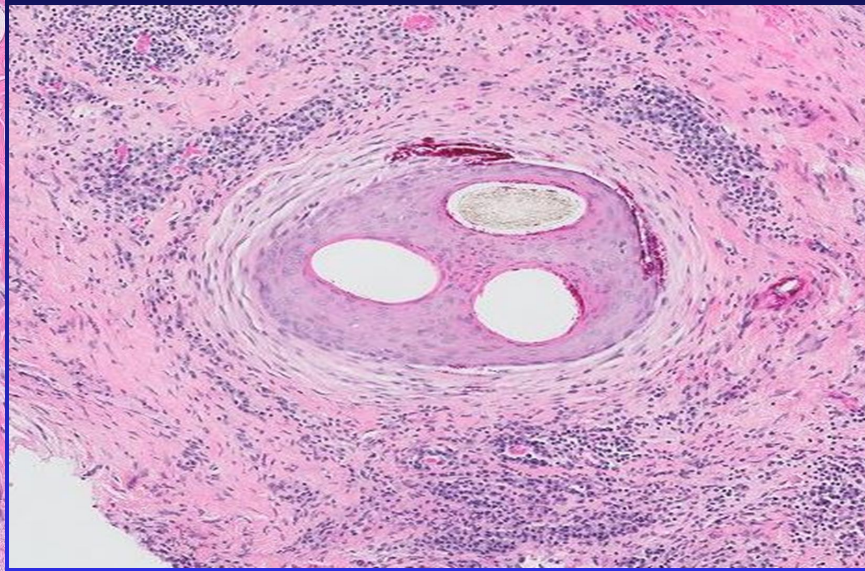
- Elusive entity with no specific etiology
  - Hair care practices likely induces disease
    - Heat
    - Traction
    - Chemicals







Central centrifugal cicatricial alopecia



Lichen planopilaris

Immunohistochemical characterization of Lichen planopilaris (LPP) and Central centrifugal cicatricial alopecia (CCCA)

Retrospective review of 55 cases

Background: What is the major problem that prompted the study?

- Central centrifugal cicatricial alopecia (CCCA)
  - Poorly defined histological entity
  - Clinical variant of lichen planopilaris (LPP)?
- Lack of definition of CCCA
- What is “Premature desquamation of the inner root sheath?”

Methods: How was the study done?

- Retrospective study using archival tissue of specimens of LPP and CCCA from CTA Lab in Portland and Hospital St. Pierre in Brussels between 2014 and December 2016.
- Histologic assessment including perifollicular fibrosis, depth/location and magnitude of lymphocytic infiltrate, premature desquamation of inner root sheath, and catagen/telogen shift.
- Immunohistochemical profile including CD3, CD4, CD8, CD20, CD68, CD123, myeloperoxidase, and CK15 was performed on all samples.

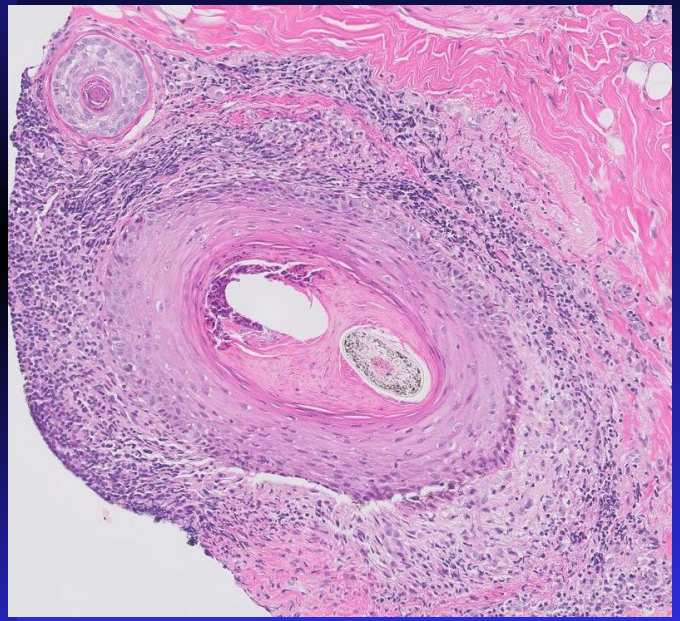
Results: What are the most important findings?

- No difference between LPP and CCCA was identified with the panel of immunohistochemical stains or histologically.
- 'Premature desquamation of the inner root sheath'
  - "Squamatization' of the follicular epithelium secondary to the lichenoid interface process
  - Pattern similar to the epidermal changes of lichen planus.

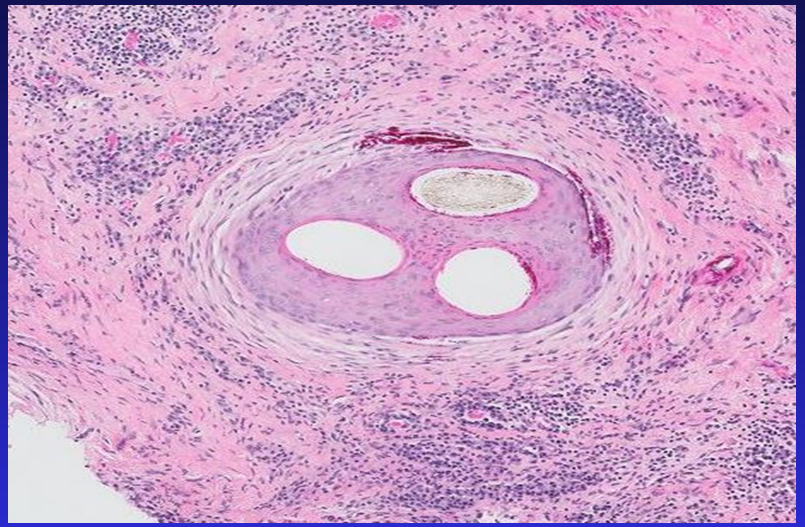
CCCA

- Gray-staining, perifollicular fibrosis at the level of the superficial isthmus and infundibulum
- Perifollicular lymphocytes at the same level as the fibrosis which may scatter into the follicular epithelium
- Squamotization of the follicular epithelium, especially the basalis, in the area of inflammation with variable loss of the inner root sheath
- Compound follicles may or may not be present
- Near absence of catagen- and telogen-phase follicles

CCCA = LPP Histologically



CCCA



LPP

Premature desquamation of the inner root sheath

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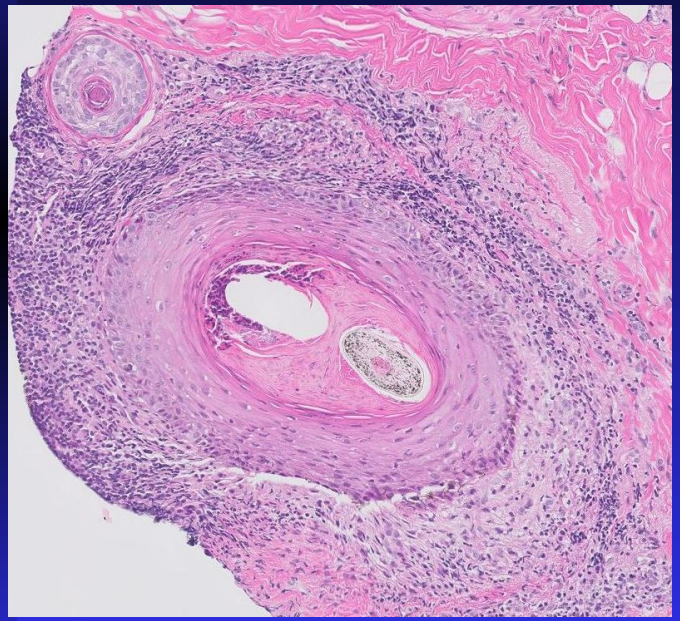
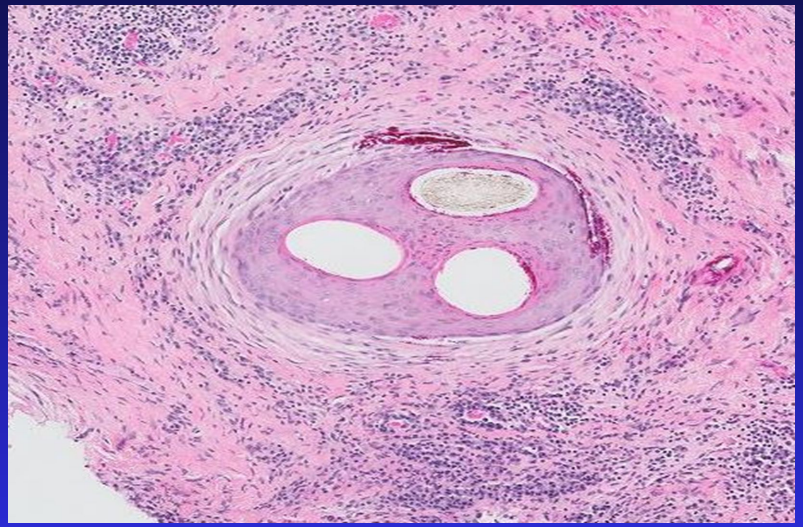
Squamatization of the follicular epithelium





Premature desquamation of the inner root sheath

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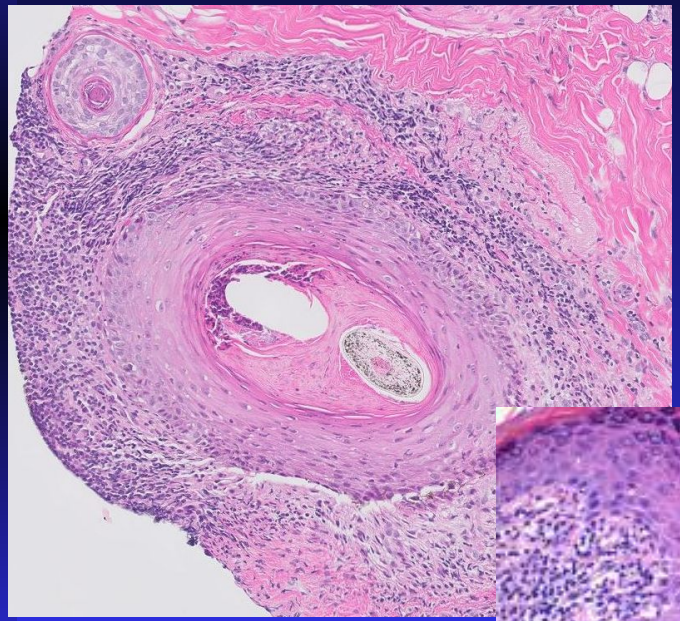
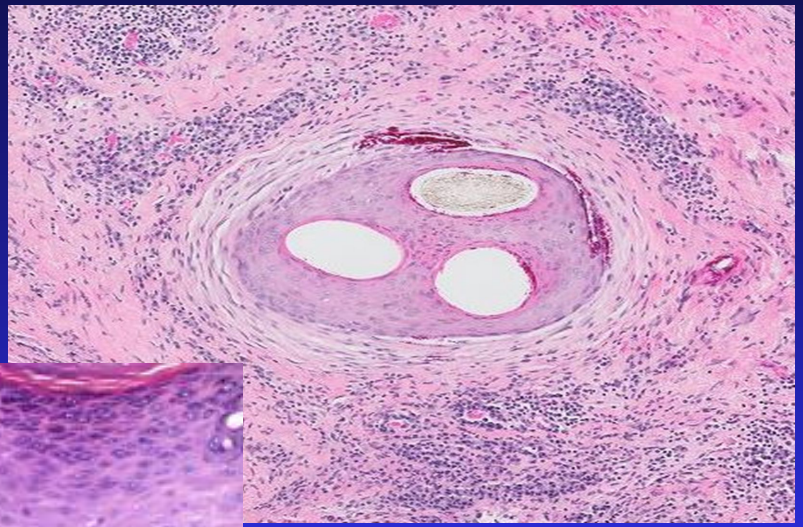
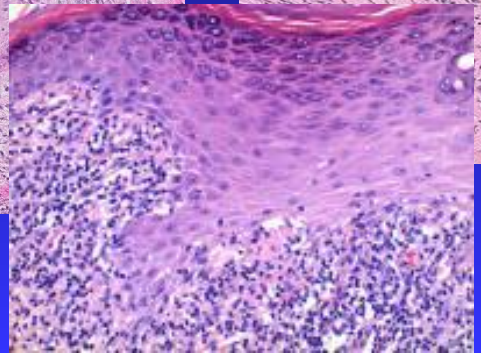
Squamatization of the follicular epithelium







Conclusion: What is the single most important conclusion?

- For diagnostic purposes, CCCA and LPP share identical immunohistological staining and histological features
- Study strongly supports that CCCA is one of several clinical variants of LPP.
- LPP and CCCA have identical histopathological and immunohistochemical findings.

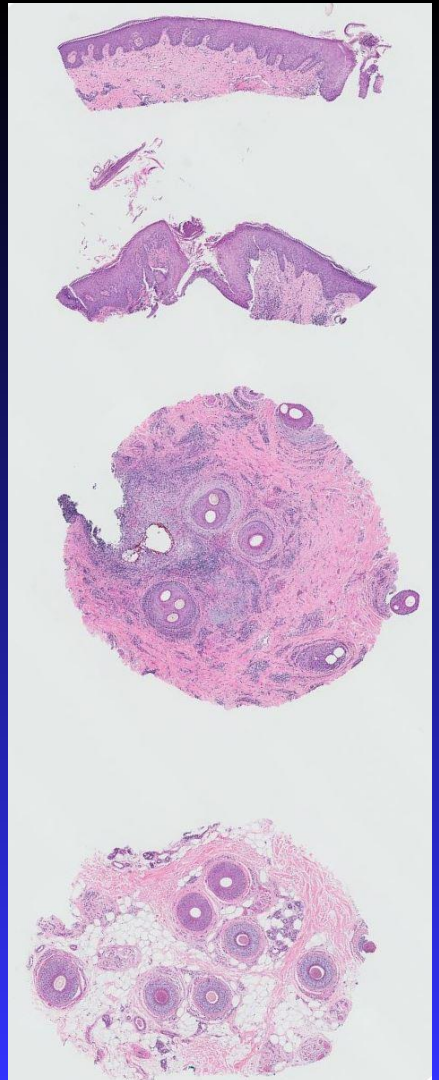
CCCA variant of LPP

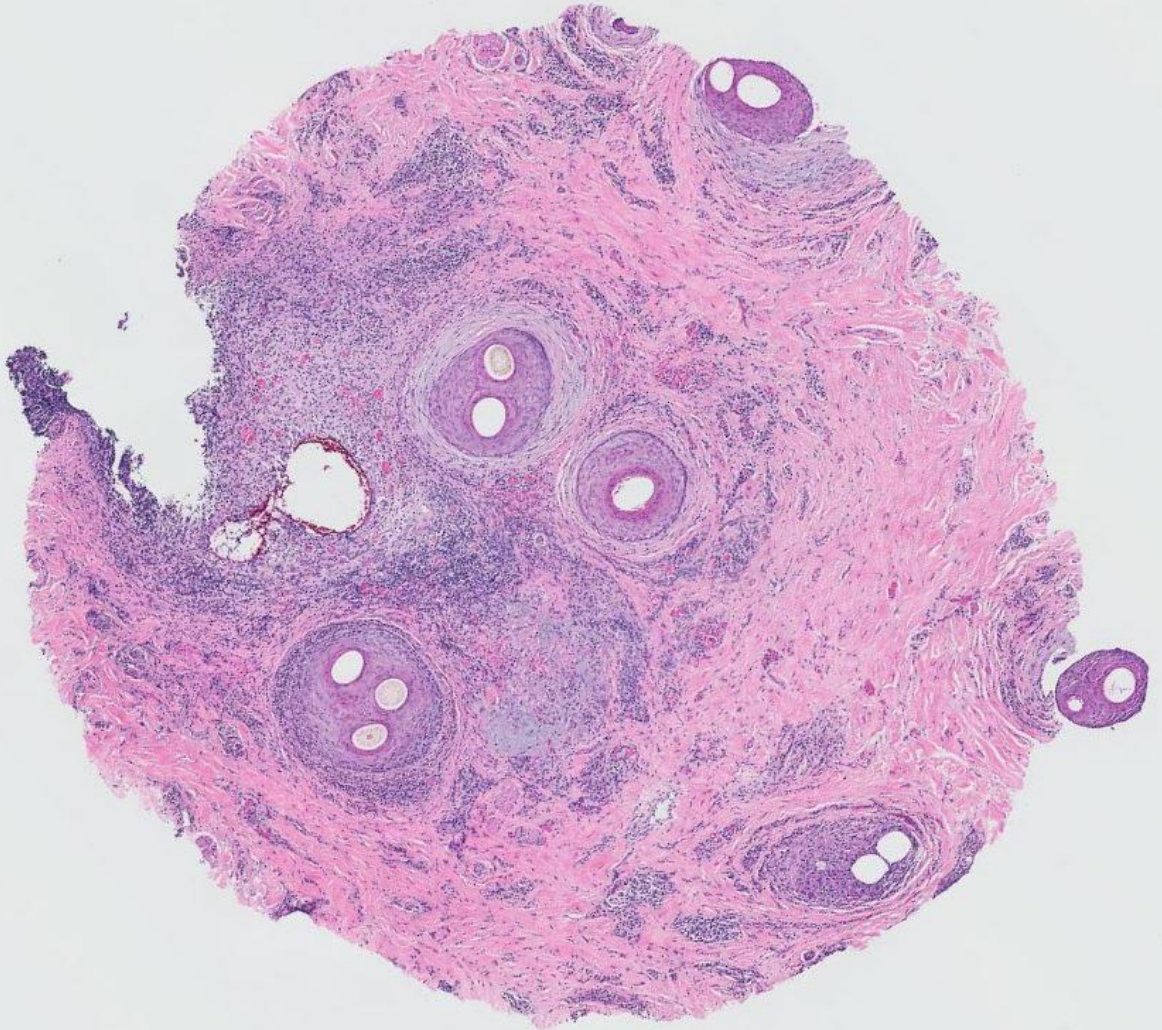
- Differential
  - Folliculitis decalvans
    - Mostly occurs in men whereas CCCA is in women
    - Interfollicular inflammation
    - Epidermal acanthosis

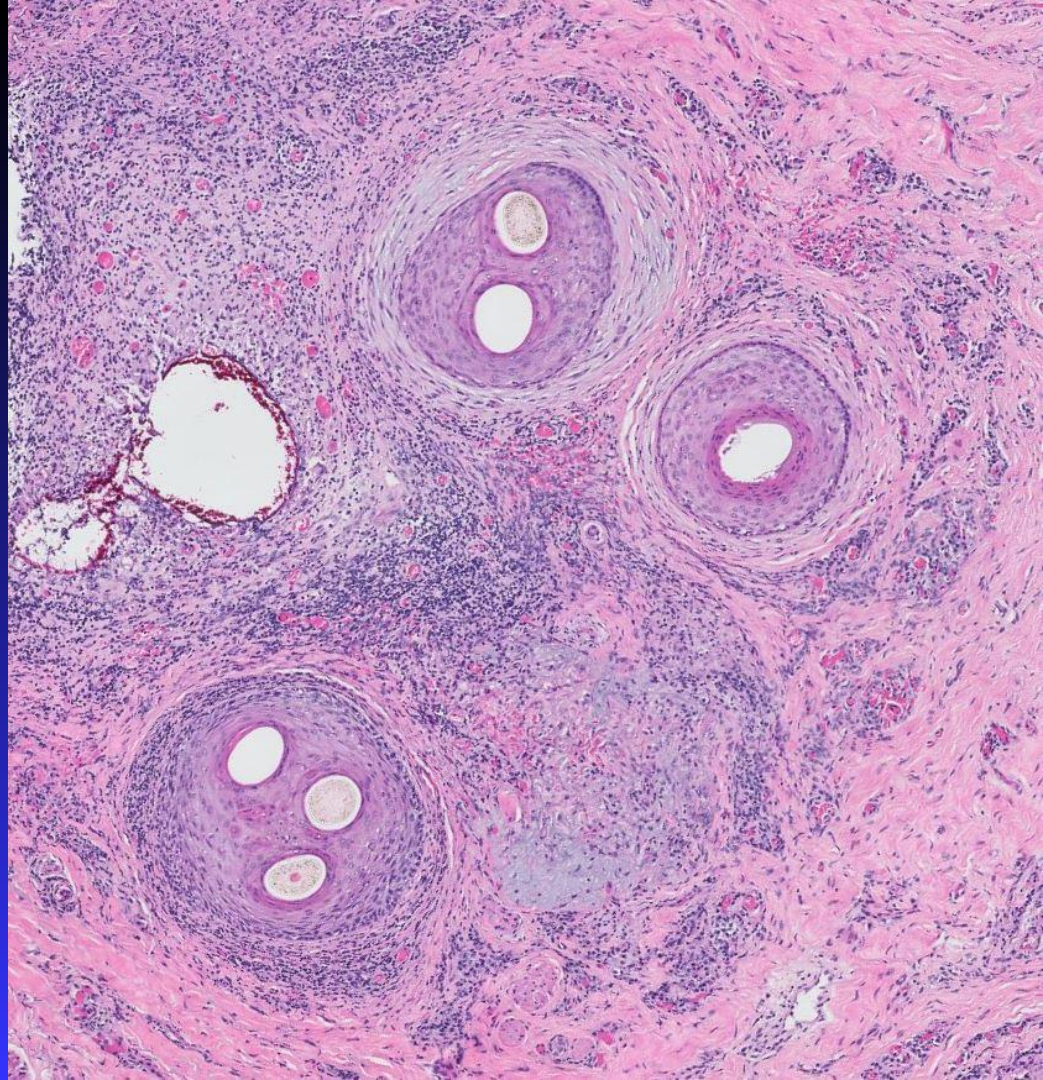
Subtypes of LPP

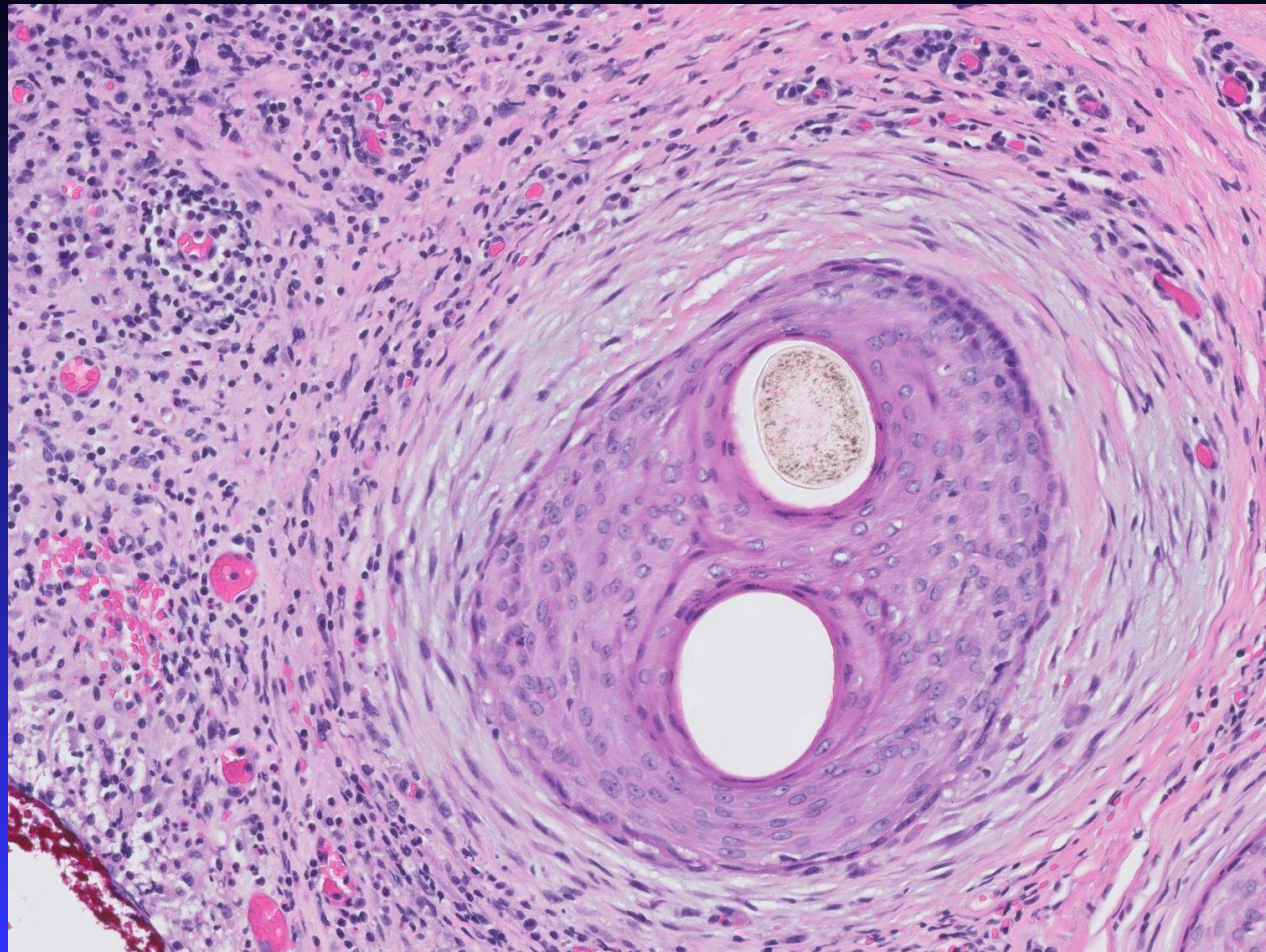
- Classic
- Graham Little Piccardi Lasseur syndrome
- Frontal fibrosing alopecia
- CCCA

- 42 y/o man with pustular eruption on scalp and hair loss









Neutrophil-poor Folliculitis decalvans

- Mostly in young men
- Pustules, redness, swelling, tufted follicles
- Staphylococcus aureus often identified
- Acne keloidalis nuchae is likely FD initiated and exacerbated by occlusive headware

Neutrophil-poor Folliculitis decalvans

- Classified in the “Neutrophilic” group in the North American Hairs Research Society (NAHRS) classification

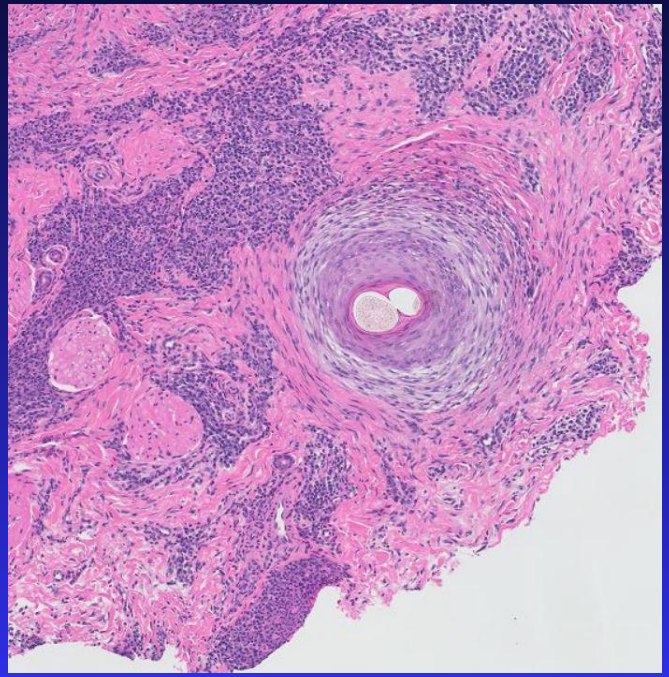
Neutrophil-poor Folliculitis decalvans

- Variable density, mixed-cell type inflammatory cell infiltrate with lymphocytes, plasma cells, macrophages and neutrophils, present in a perifollicular and interstitial distribution
- If neutrophils are not evident, a myeloperoxidase IHC study may be of utility
- Variable amount of perifollicular fibrosis—sometimes none is present

Neutrophil-poor Folliculitis decalvans

- Differential
  - Lichen planopilaris
  - Tinea capitis
  - Dissecting folliculitis/cellulitis

LPP or the lymphocytic-variant of FD?



J Cutan Pathol. 2017 Apr;44(4):352-357. doi: 10.1111/cup.12892. Epub 2017 Jan 27.

Epidermal psoriasiform hyperplasia, an unrecognized sign of folliculitis decalvans: A histological study of 26 patients.

Matard B[1], Cavelier-Balloy B[2], Reygagne P[1].

Author information

Abstract

BACKGROUND: Follicular hyperkeratosis along with hyperplasia of the follicular and interfollicular epithelia are major histopathological characteristics of hidradenitis suppurativa (HS). The presence of an occasional thickening of lesional skin in some folliculitis decalvans (FD) patients and histological similarities between FD and HS led us to look for epidermal hyperplasia and follicular hyperkeratosis in FD patients.

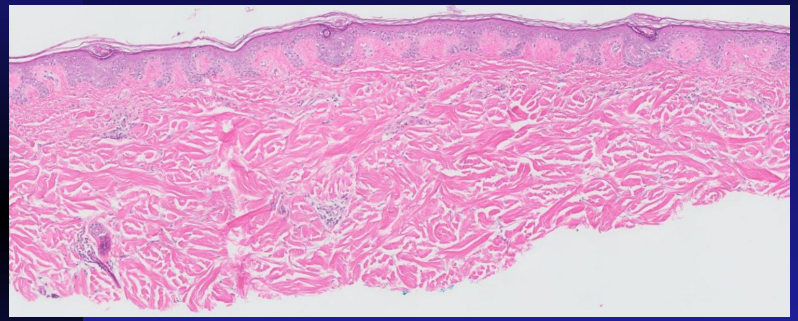
PATIENTS AND METHOD: We performed a retrospective histological analysis of 26 patients with FD.

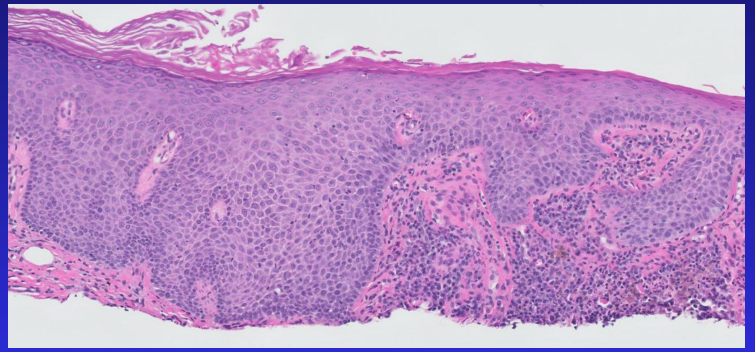
OBJECTIVE: We sought to find out whether the presence of hyperplasia of the interfollicular epidermis and of the follicular epithelia could be verified in FD, with reference to the work of von Laffert et al. concerning HS.

RESULTS: The main quantitative and qualitative data were: follicular hyperkeratosis (77%), hyperplasia of the interfollicular epidermis (92%) with a psoriasiform aspect (88%), atrophy of the follicular epithelia (85%), plasma cells in infiltrate (92%) in large quantities (42%), follicular microcysts (60%), atrophy of the sebaceous glands (85%) and polytrichia (54%).

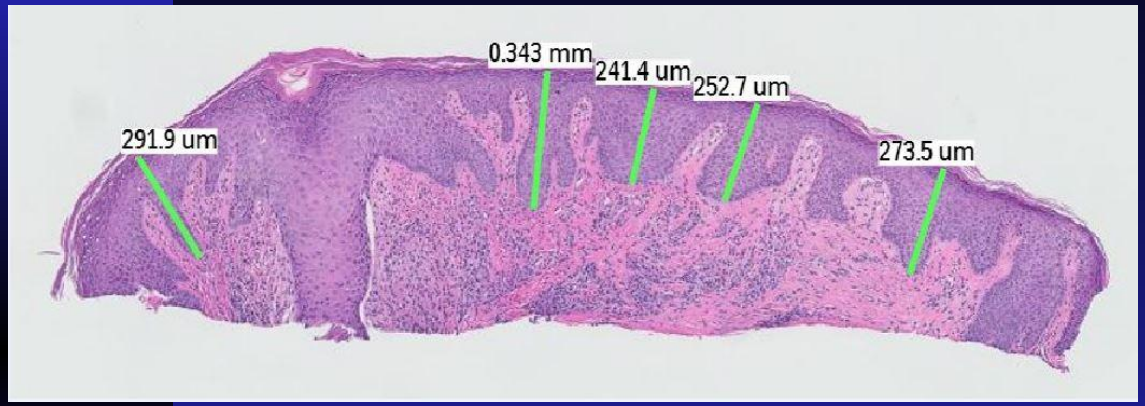
CONCLUSION: Epidermal hyperplasia, sometimes psoriasiform and follicular microcysts, are significant histological signs of FD, which have been ignored until now although they seem very common.

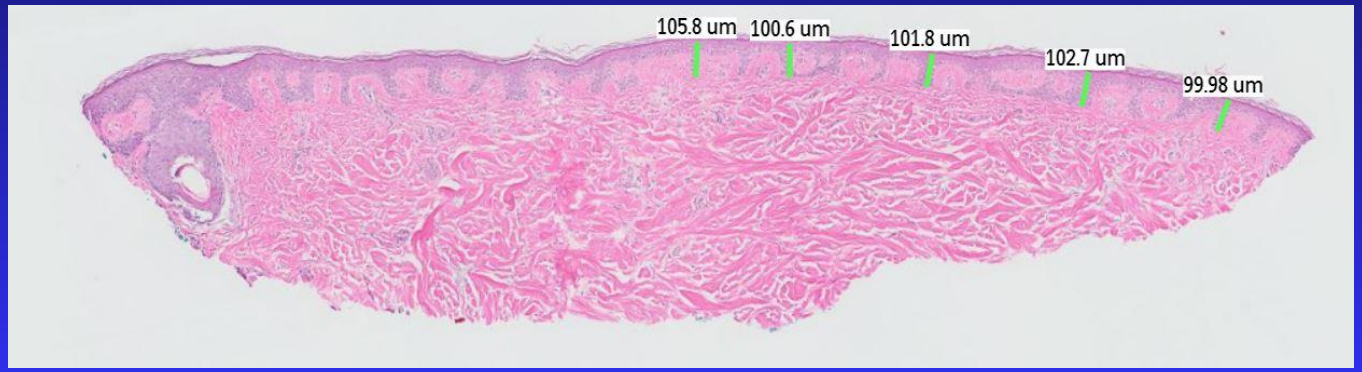
Folliculitis decalvans or LPP?



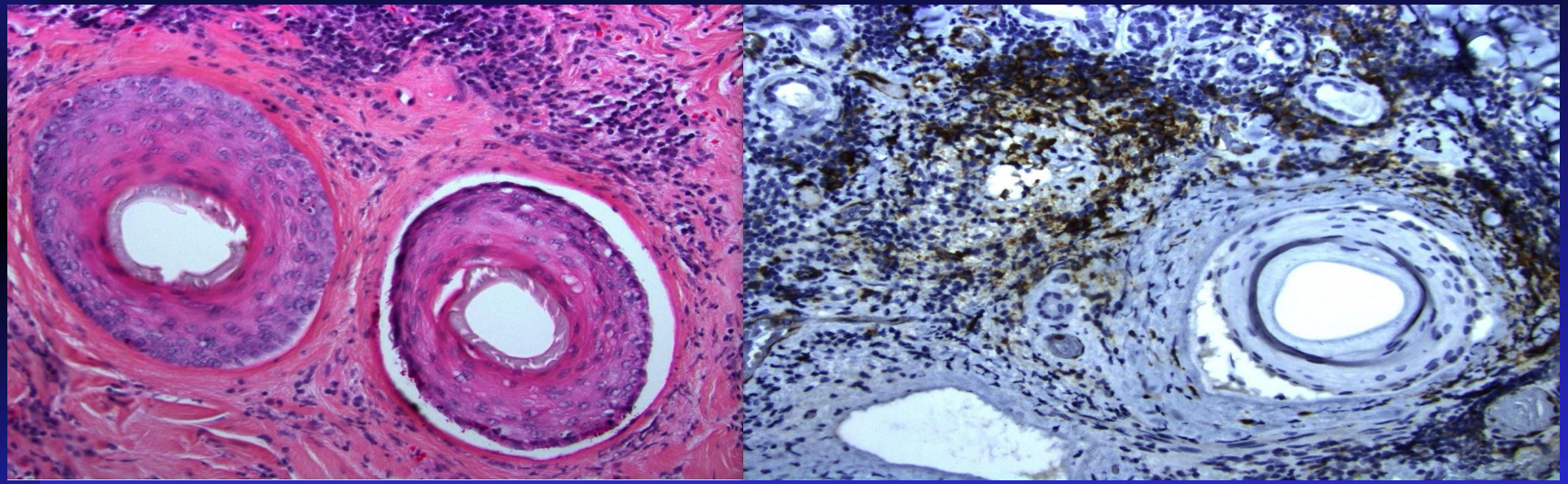


Folliculitis decalvans or LPP?

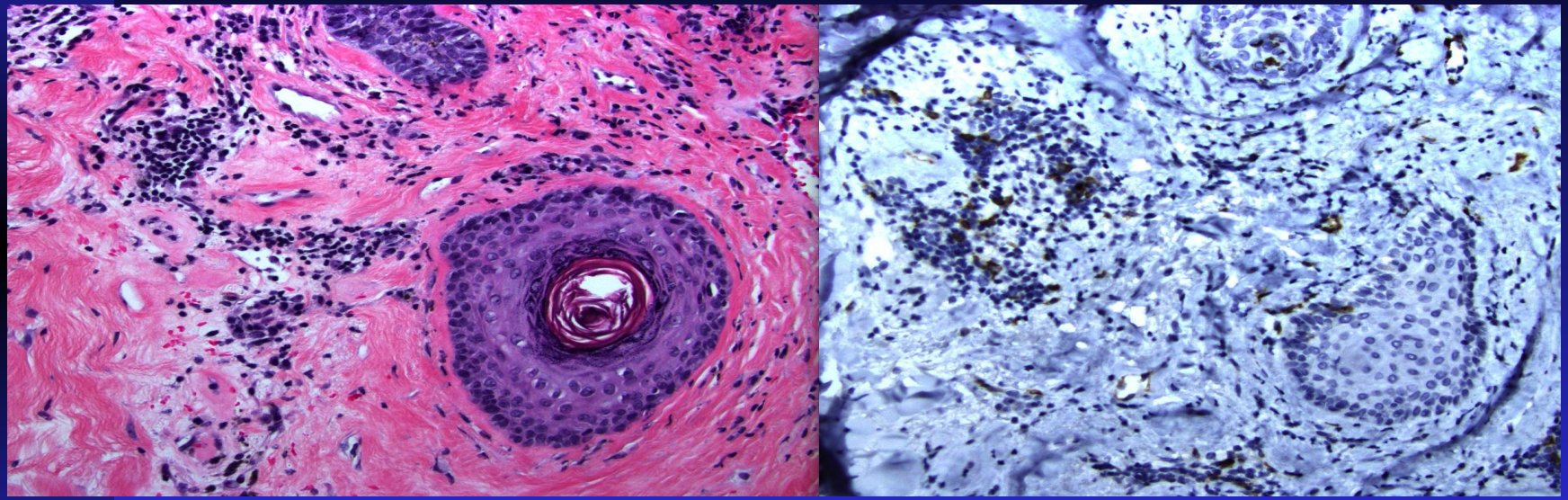




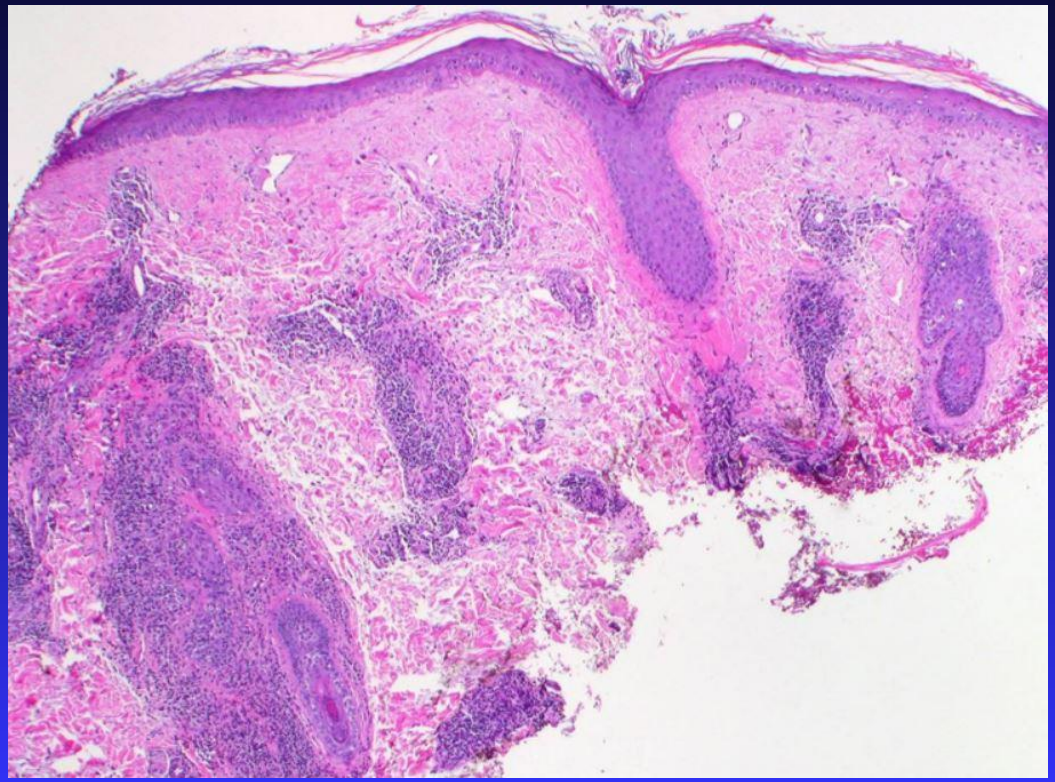
Clusters of CD123 = Lupus



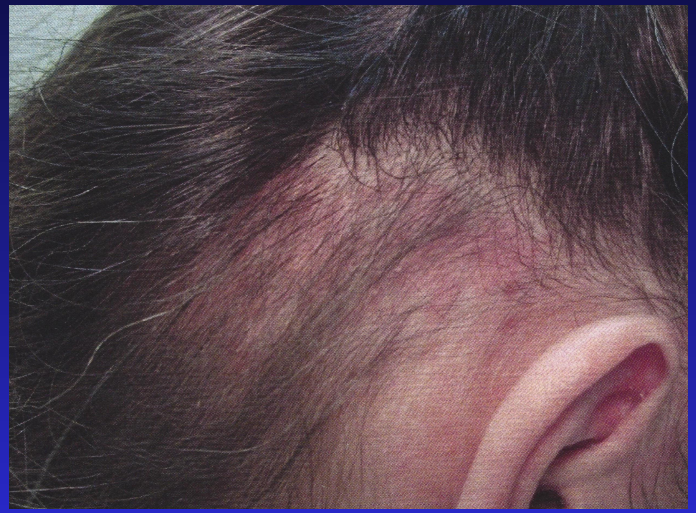
CD123 positive in endothelium

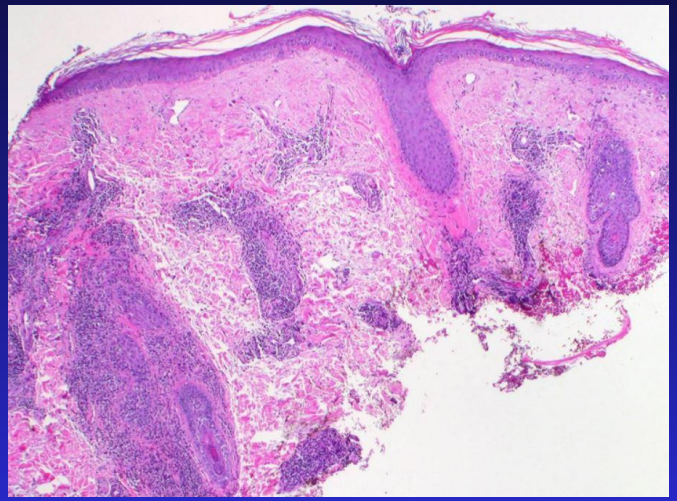


Alopecia areata-like pattern



Systemic lupus erythematosus
AA-like pattern



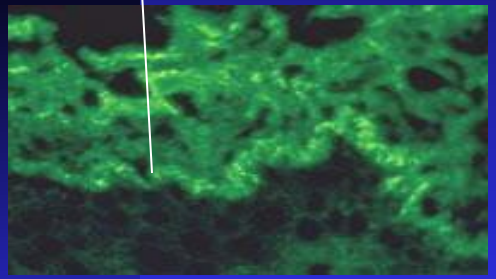


Journal of American Academy of Dermatology 2010; 63: 333-6

Sperling LC, Cowper SE, Knopp EA. An Atlas of Hair Pathology with Clinical Correlations. 2nd ed. Boca Raton (FL): Taylor and Francis Group, 2012.

Lupus Erythematosus

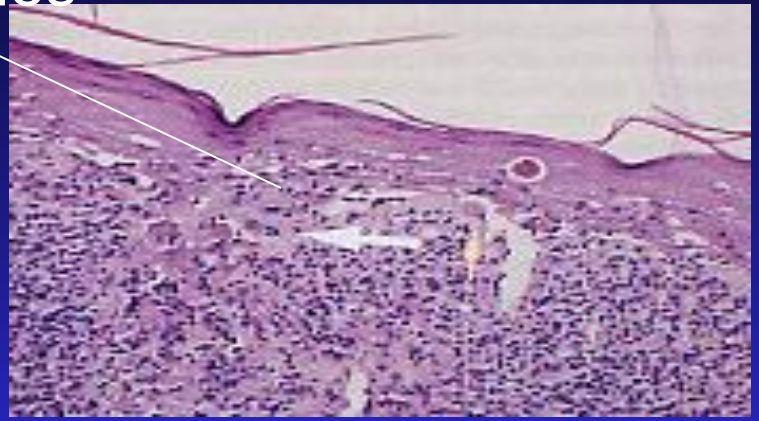
Lupus band in 76% of patients



- Linear/ granular
- IgG, IgM, C3 at DEJ

Lichen Planopilaris.

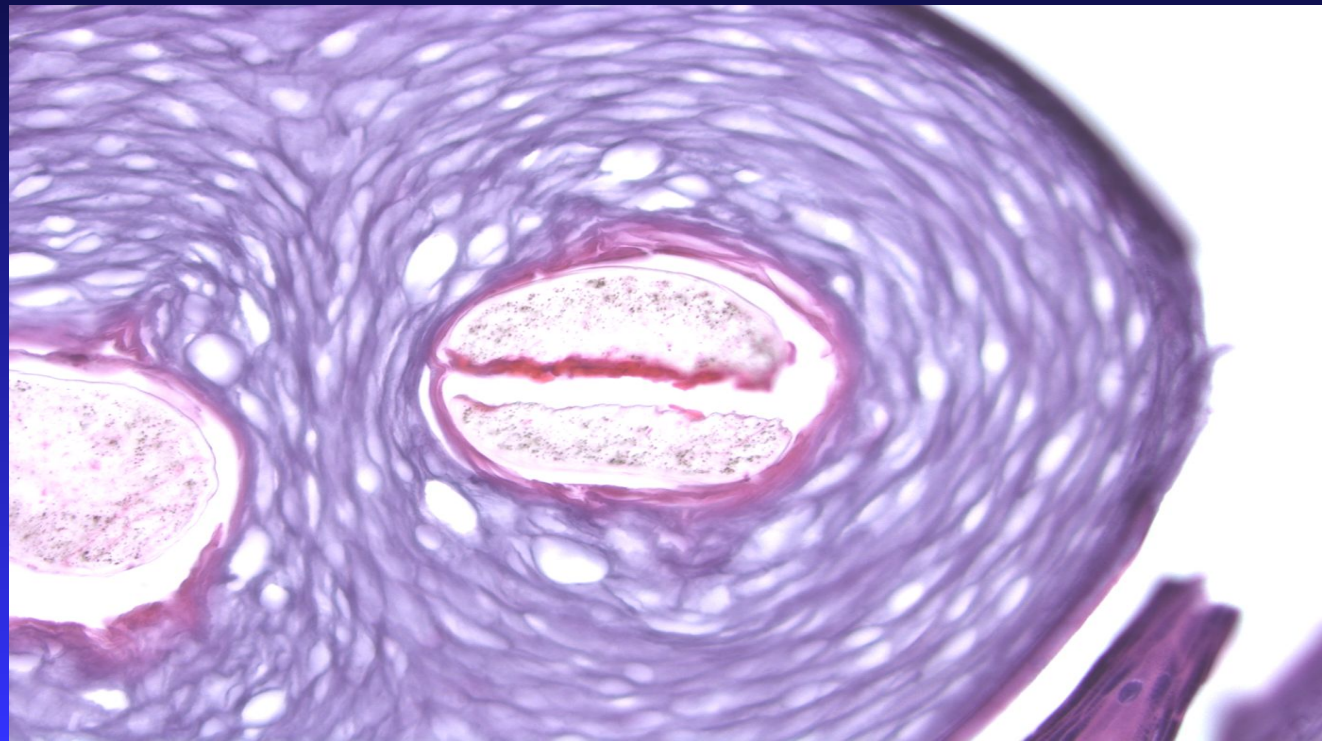
Colloid bodies



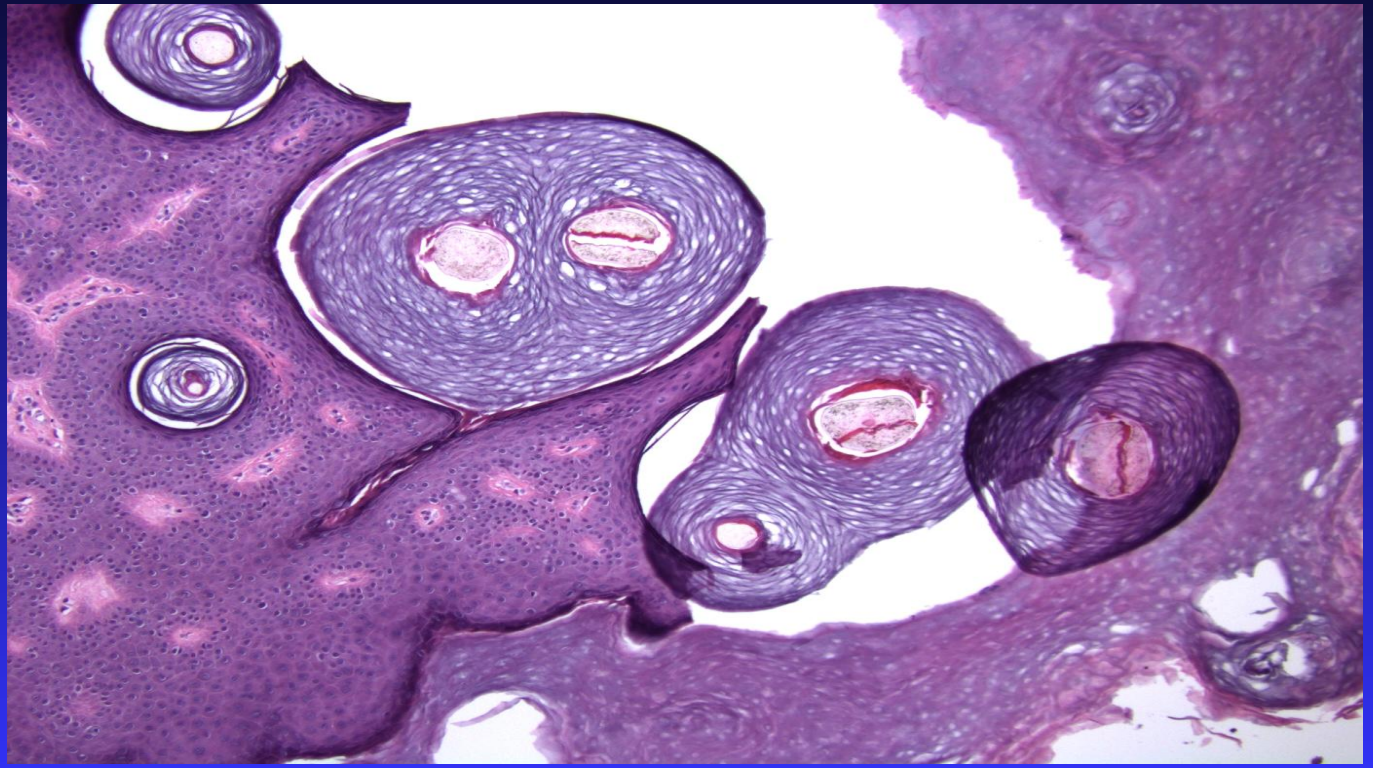
- Globular fluorescence of IgM, IgA and C3 (not IgG)

Trichotillosis: Hamburger Sign

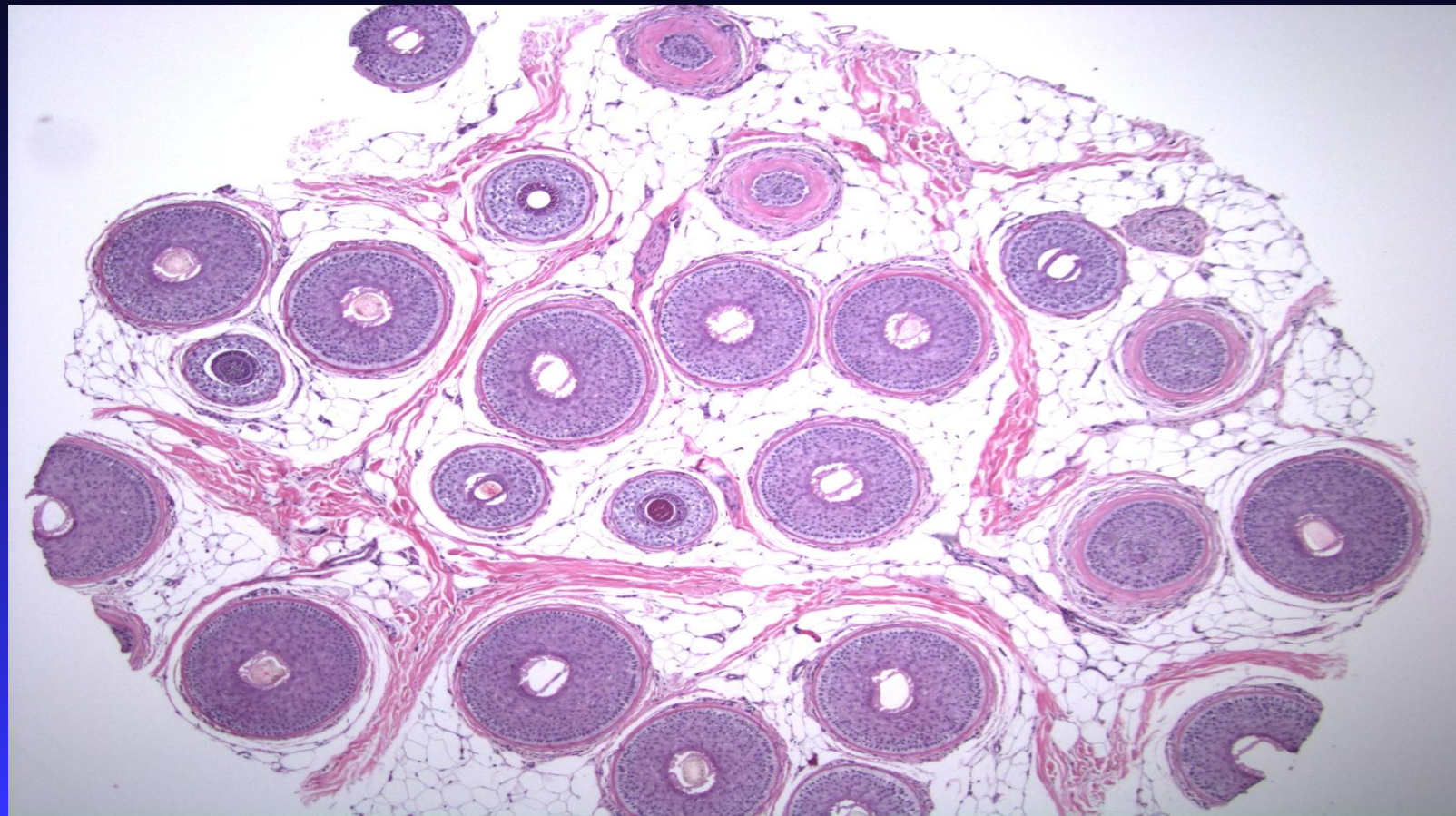
Royer MC and Sperling LC. Splitting hairs: the 'hamburger sign' in Trichotillomania. J Cutan Pathol. 33Suppl 2:63-4, 2006.



Lichen simplex chronicus



Trichotillosis—Catagen shift



Thank you!