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10 AÑOS

Reclasificando las Alopecias: Una guía práctica para el diagnóstico

Curtis T. Thompson, M.D. Estados Unidos

Reclasificando las Alopecias: Una guía práctica para el diagnóstico

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Oregon Health and Science University

SPECIAL REPORT

Summary of North American Hair Research Society (NAHRS)–sponsored Workshop on Cicatricial Alopecia, Duke University Medical Center, February 10 and 11, 2001

Elise A. Olsen, MD,[a] Wilma F. Bergfeld, MD,[b] George Cotsarelis, MD,[c] Vera H. Price, MD,[d] Jerry Shapiro, MD,[e] Rodney Sinclair, MD,[f] Alvin Solomon, MD,[g] Leonard Sperling, MD,[h] Kurt Stenn, MD,[i] David A. Whiting, MD,[j] and the members of the Workshop on Cicatricial Alopecia* *Durham, North Carolina; Cleveland, Ohio; Philadelphia, Pennsylvania; San Francisco, California; Vancouver, British Columbia; Melbourne, Australia; Atlanta, Georgia; Bethesda, Maryland; Skillman, New Jersey; and Dallas, Texas*

Del año 2001

Proposed working classification of primary cicatricial alopecia

Lymphocytic

- Chronic cutaneous lupus erythematosus
- Lichen planopilaris (LPP)[4]
  - Classic LPP
  - Frontal fibrosing alopecia[5]
  - Graham-Little syndrome[3]
- Classic pseudopelade (Brocq)[6,7]
- Central centrifugal cicatricial alopecia[8]
- Alopecia mucinosa[3]
- Keratosis follicularis spinulosa decalvans[9]

Neutrophilic

- Folliculitis decalvans[3]
- Dissecting cellulitis/folliculitis[3] (*perifolliculitis abscedens et suffodiens*)

Mixed

- Folliculitis (acne) keloidalis[9]
- Folliculitis (acne) necrotica[3]
- Erosive pustular dermatosis[10]

Nonspecific

Cambios probables a la clasificación:

- Alopecia fibrosante con distribución en patrón—añadiendo
- Pseudopelada de Brocq—retentiendo
- Alopecia cicatricial centrifuga central (ACCC)—sigue siendo una entidad distinta.
- Eliminando los trastornos que no sean alopecia cicatricial primaria
- Foliculitis decalvante—cambiando clasificación a ambos neutrofílico y linfocito
- Etapa temprana y tardía de cada trastorno cicatricial

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Arch Dermatol. 1994 Jun;130(6):770-4.

Postmenopausal frontal fibrosing alopecia. Scarring alopecia in a pattern distribution.

Kossard S[1].

Author information Papers

Erratum in
Arch Dermatol 1994 Nov;130(11):1407.

Abstract

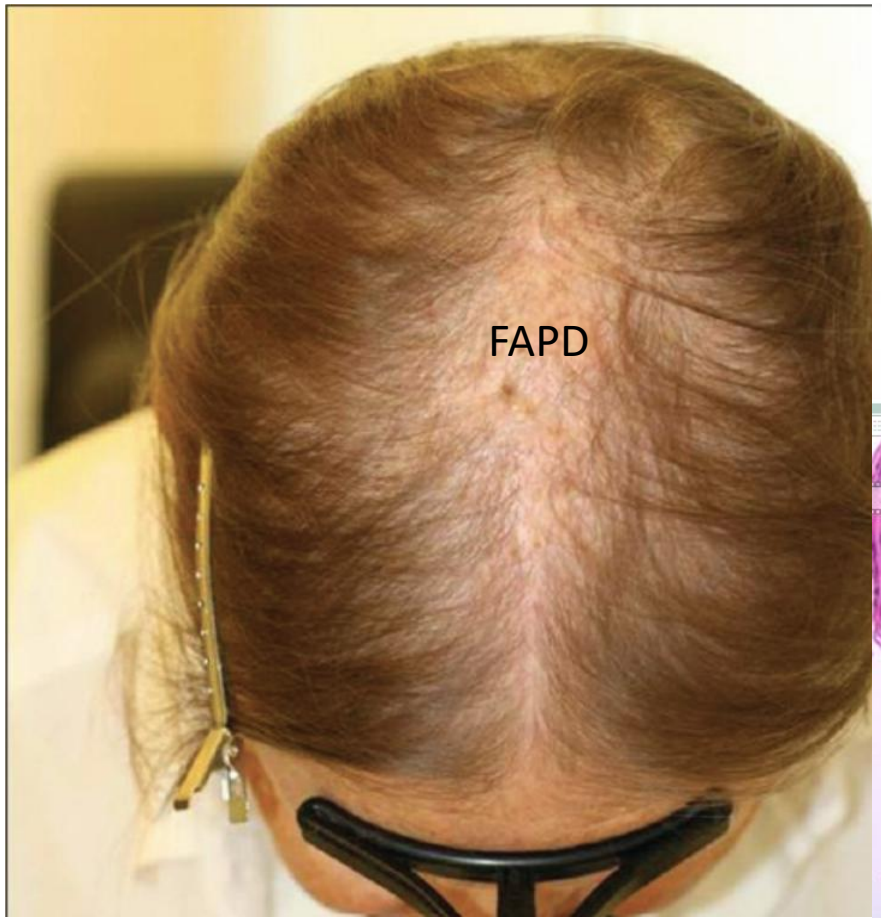
Alopecia Fibrosante con Distribución en Patrón

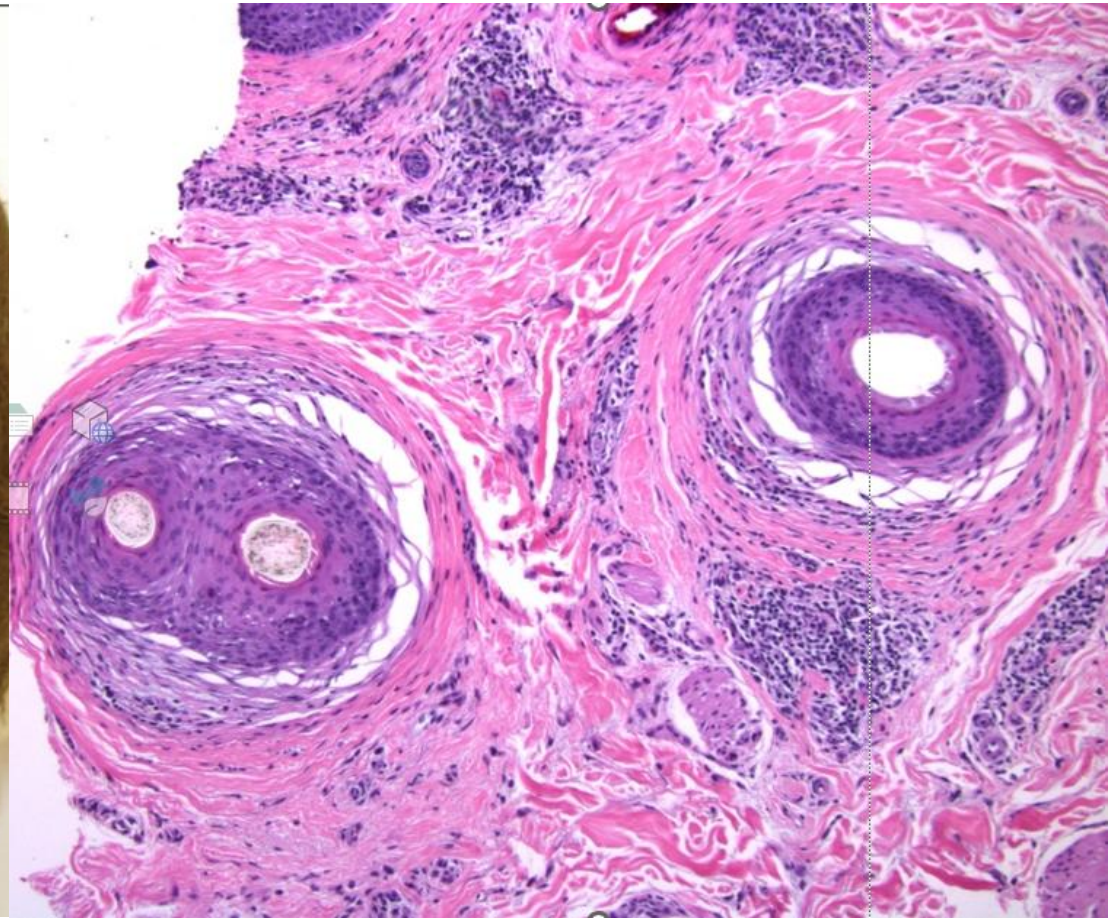
FAPD

OBSERVATIONS: The six postmenopausal women developed a progressive frontal hairline recession that was associated with perifollicular erythema within the marginal hairline, producing a frontal fibrosing alopecia extending to the temporal and parietal hair margins. Scalp biopsy specimens from the frontal hair margin showed perifollicular fibrosis and lymphocytic inflammation concentrated around the isthmus and infundibular areas of the follicles. Immunophenotyping of the lymphocytes showed a dominance of activated T-helper cells. Clinical review of all six cases showed a progressive marginal alopecia without the typical multifocal areas of involvement seen in lichen planopilaris or pseudopelade. None of the patients had mucous membrane or skin lesions typical of lichen planus. Hormonal studies, in five patients, showed no elevated androgen abnormalities.

CONCLUSIONS: Progressive frontal recession in postmenopausal women may show clinical features of a fibrosing alopecia. The histologic findings are indistinguishable from those seen in lichen planopilaris. However, the absence of associated lesions of lichen planus in all six women raises the possibility that this mode of follicular destruction represents a reaction pattern triggered by the events underlying postmenopausal frontal hairline recession.

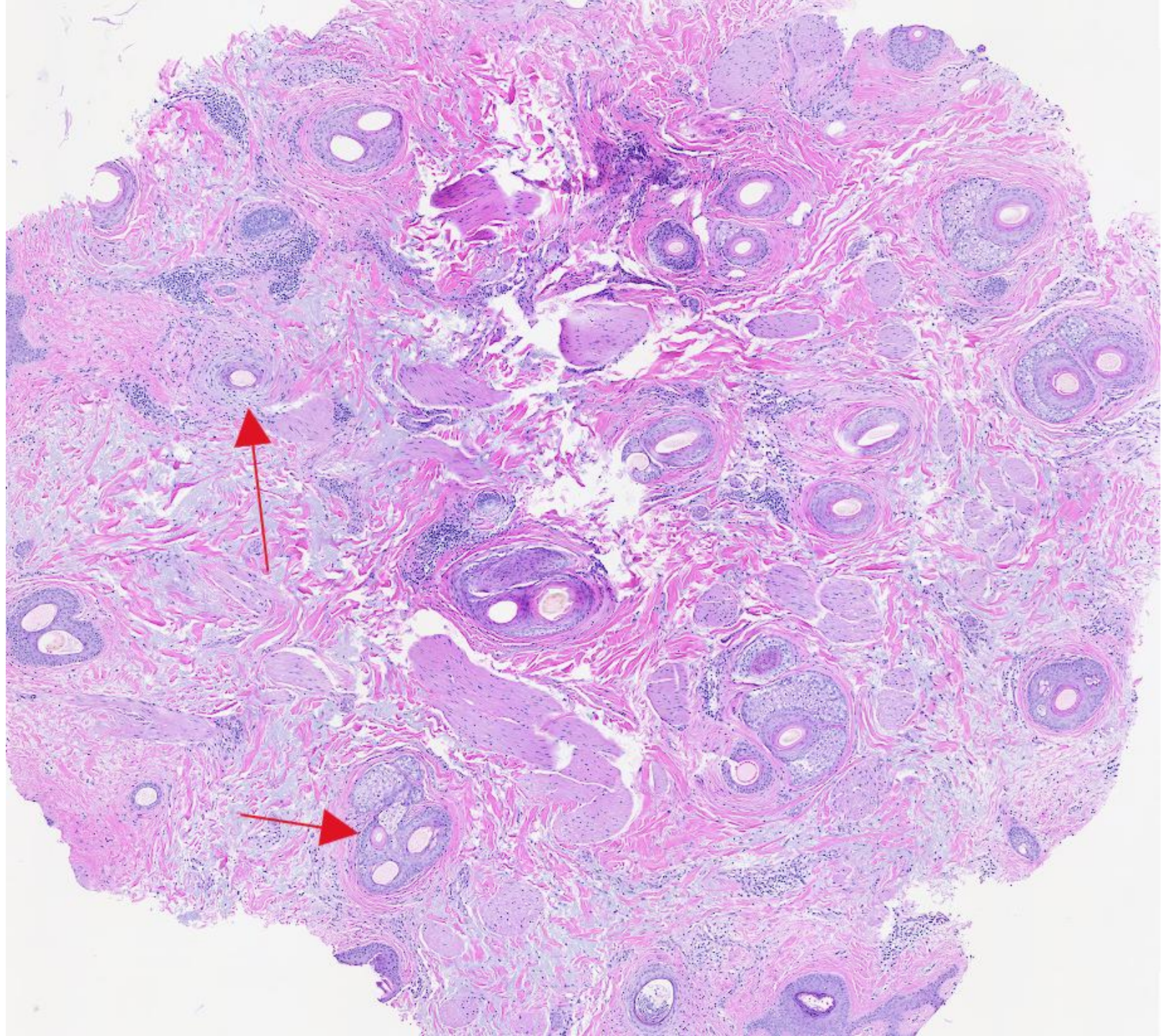
Alopecia fibrosante con distribución en patrón





Alopecia fibrosante con distribución en patrón

- Liquen Plano Pilar (LPP) con una presentación con caída difusa del cabello de patrón femenino.
- Subyacente la calvicie de patrón femenino (CPF)
  - Androgénico (<60 años)
  - Senectud (>60 años)



Alopecia fibrosante con distribución en patrón

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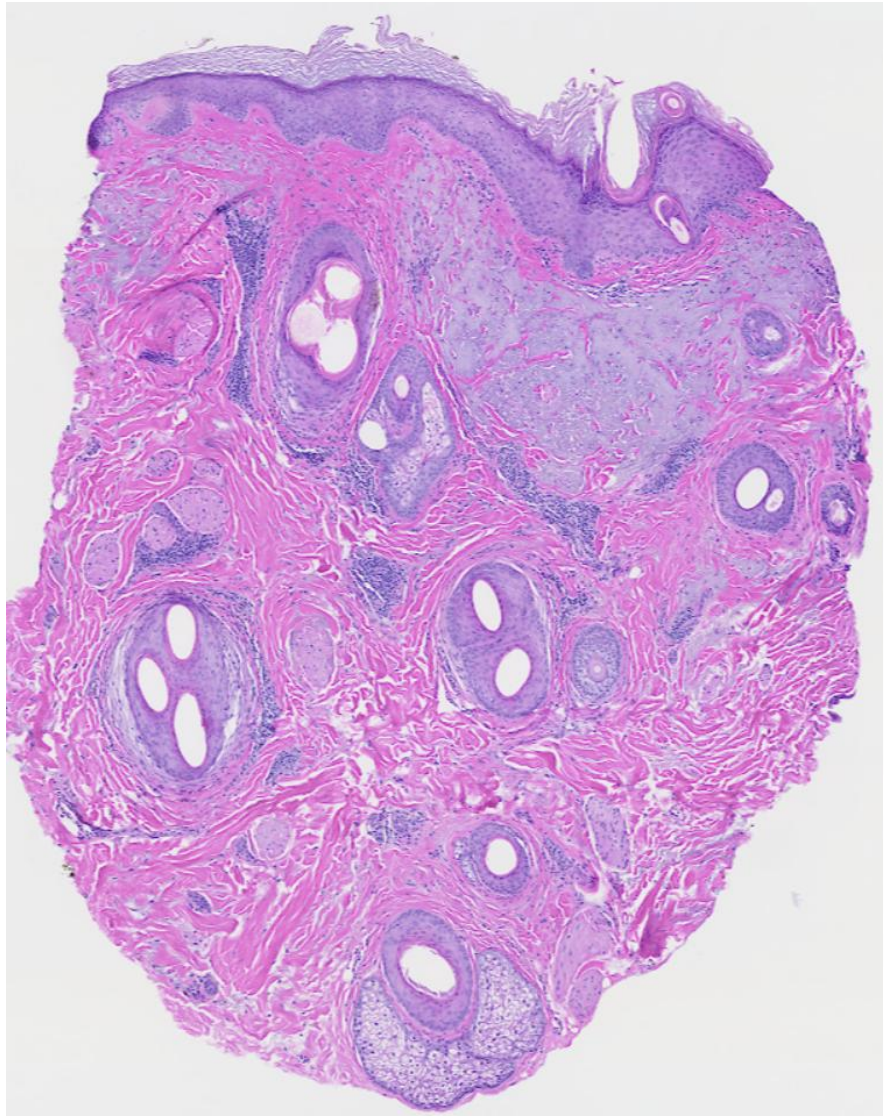


Alopecia fibrosante con distribución en patrón

Tinte para retoque de raíces del cabello (Polvo)

5A/MEDIUM ASH BROWN

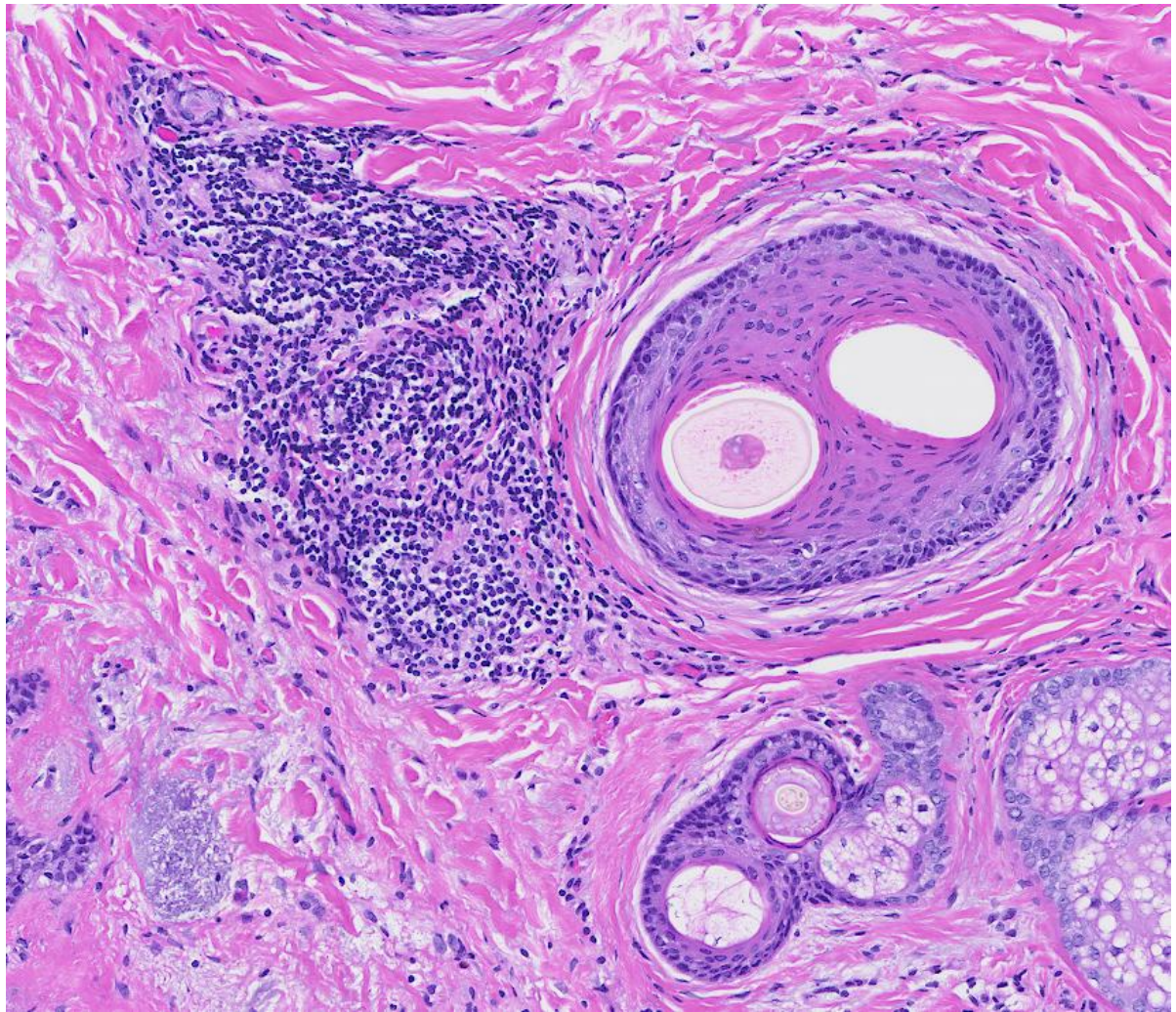
WATER, C12-15 PARETH-3, AMMONIUM HYDROXIDE, OLETH-10, DILINOLEIC ACID, COCAMIDE MEA, LINOLEAMIDOPROPYL DIMETHYLAMINE DIMER DILINOLEATE, STEARETH-21, BEHENTRIMONIUM CHLORIDE, POLYQUATERNIUM-22, SODIUM SULFATE, FRAGRANCE, RESORCINOL, ERYTHORBIC ACID, p-PHENYLENEDIAMINE, CARAMEL, p-AMINOPHENOL, m-AMINOPHENOL, IRON OXIDES, MICA, SODIUM SULFITE, N,N-BIS(2-HYDROXYETHYL)-p-PHENYLENEDIAMINE SULFATE, 1-NAPHTHOL, SODIUM METASILICATE, EDTA, SARGASSUM FILIPENDULA EXTRACT, HYPNEA MUSCIFORMIS EXTRACT, GELLIDIELA ACEROSA EXTRACT, TITANIUM DIOXIDE.

Cambios probables a la clasificación:

- Alopecia fibrosante con distribución en patrón—añadiendo
- Pseudopelada de Brocq—retentiendo
- Alopecia cicatricial centrifuga central (ACCC)—sigue siendo una entidad distinta.
- Eliminando los trastornos que no sean alopecia cicatricial primaria
- Foliculitis decalvante—cambiando clasificación a ambos neutrofílico y linfocito
- Etapa temprana y tardía de cada trastorno cicatricial

Pseudopelada de Brocq

Pseudopelada de Brocq



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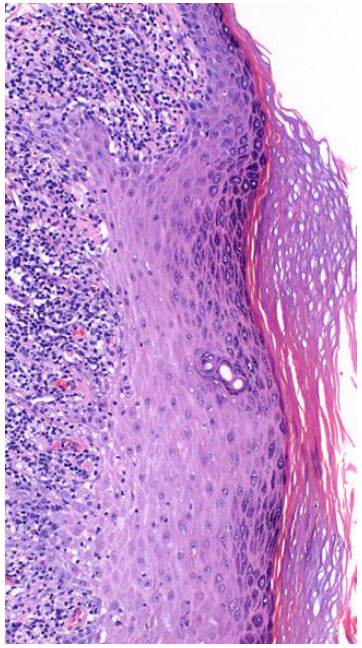
LPP versus ACCC

La pregunta:

Son LPP y ACCC idénticos en histopatología?

Descamación prematura de la vaina radicular interna (PDIRS)
=
Liquen plano cambia del epithelia follicular

Son LPP y ACCC idénticos en histopatología?



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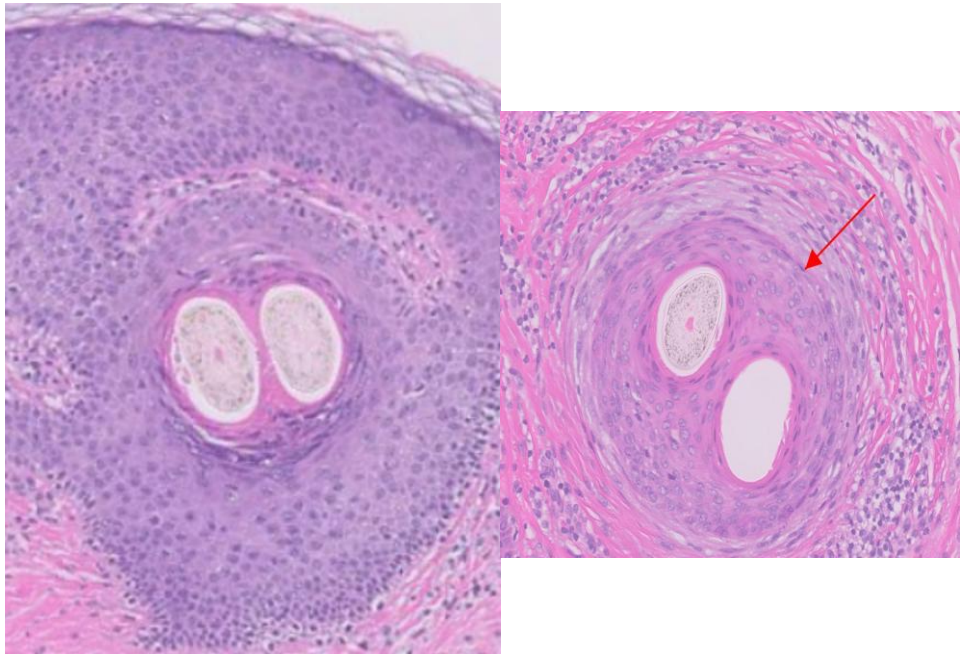


LPP versus ACCC

- Jordan CS, Chapman C, Kolivras A, Roberts JL, Thompson NB, Thompson CT. Clinicopathologic and immunophenotypic characterization of lichen planopilaris and central centrifugal cicatricial alopecia: A comparative study of 51 cases. *J Cutan Pathol*. 2020;47(2):128-134.

Conclusión: No existe una diferencia entre LPP y ACCC

Premature Desquamation of the Inner Root Sheath in Noninflamed Hair Follicles as a Specific Marker for Central Centrifugal Cicatricial Alopecia

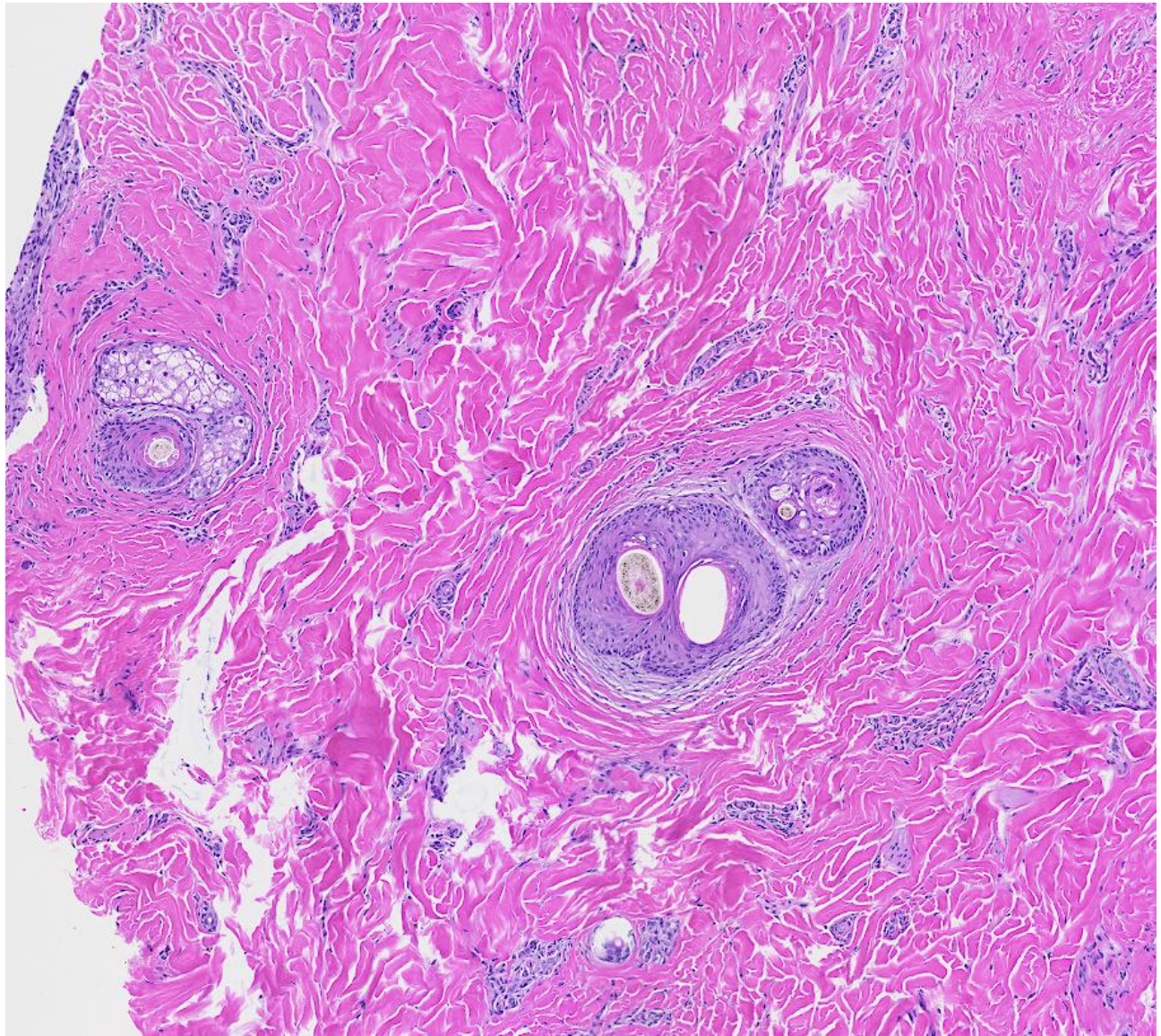
Timothy Tan, DO, Joan Guitart, MD,†‡ Pedram Gerami, MD,†‡ and Pedram Yazdan, MD†*

TABLE 2. Cases of PDIRS in Inflamed and Noninflamed Hair Follicles

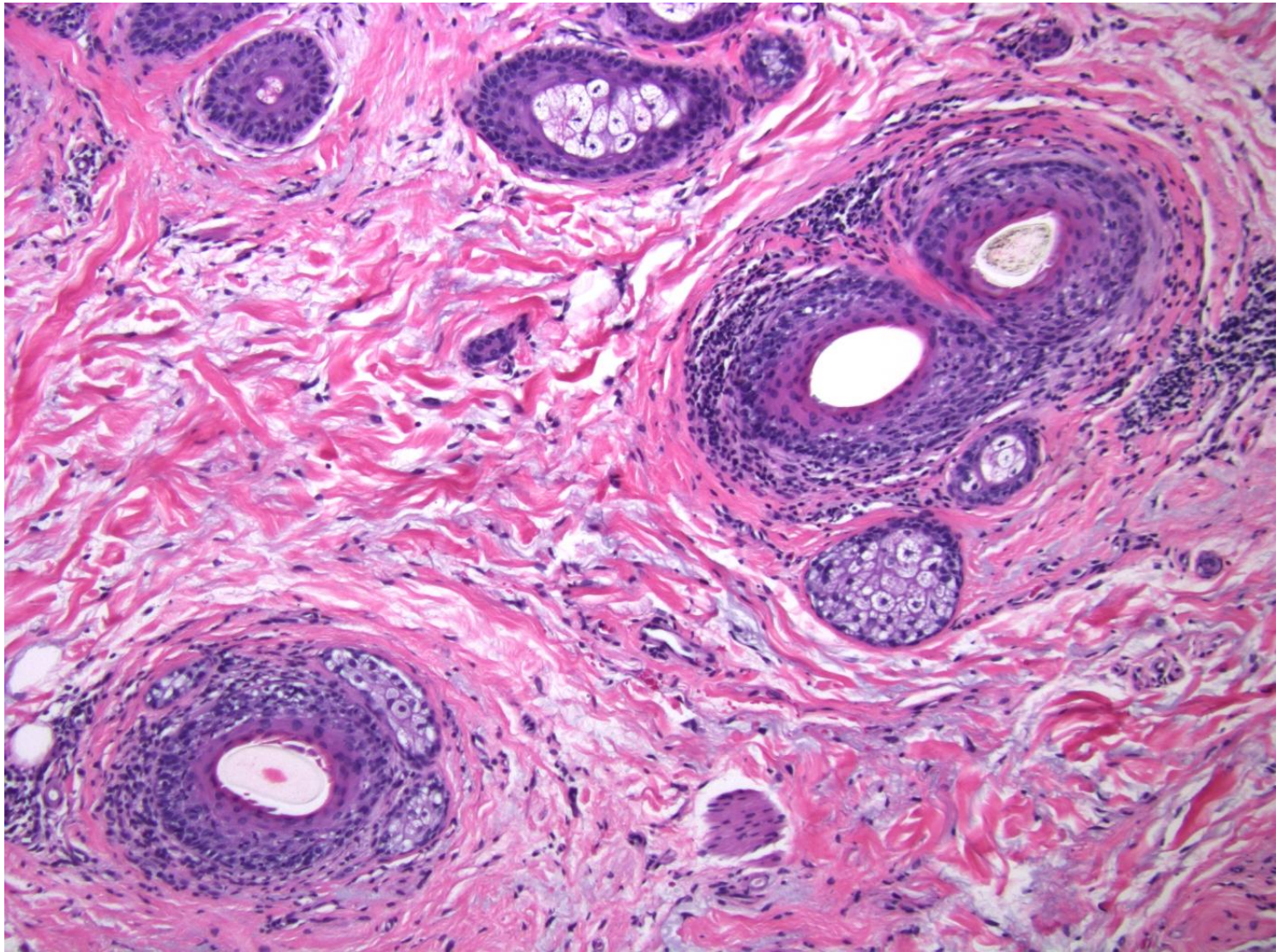
Type of Alopecia	Total Cases With PDIRS, n	Cases With PDIRS in Inflamed Follicles, n (%)	Cases with PDIRS in Noninflamed Follicles, n (%)
Cicatricial alopecia			
CCCA	111	30 (27)	81 (73)
LPP	44	44 (100)	0 (0)
DLE	3	3 (100)	0 (0)
AKN	1	1 (100)	0 (0)
FD	24	21 (87)	3 (13)
Noncicatricial alopecia			
AGA	1	0 (0)	1 (100)
AA	1	1 (100)	0 (0)
PsA	2	0 (0)	2 (100)

Tan T, Guitart J, Gerami P, Yazdan P. Premature Desquamation of the Inner Root Sheath in Noninflamed Hair Follicles as a Specific Marker for Central Centrifugal Cicatricial Alopecia. Am J Dermatopathol. 2019 May;41(5):350-354..

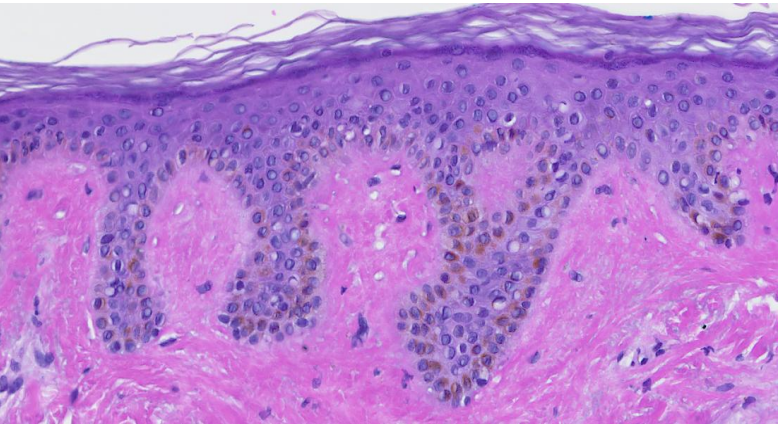
Alopecia cicatricial centrifuga central (ACCC)

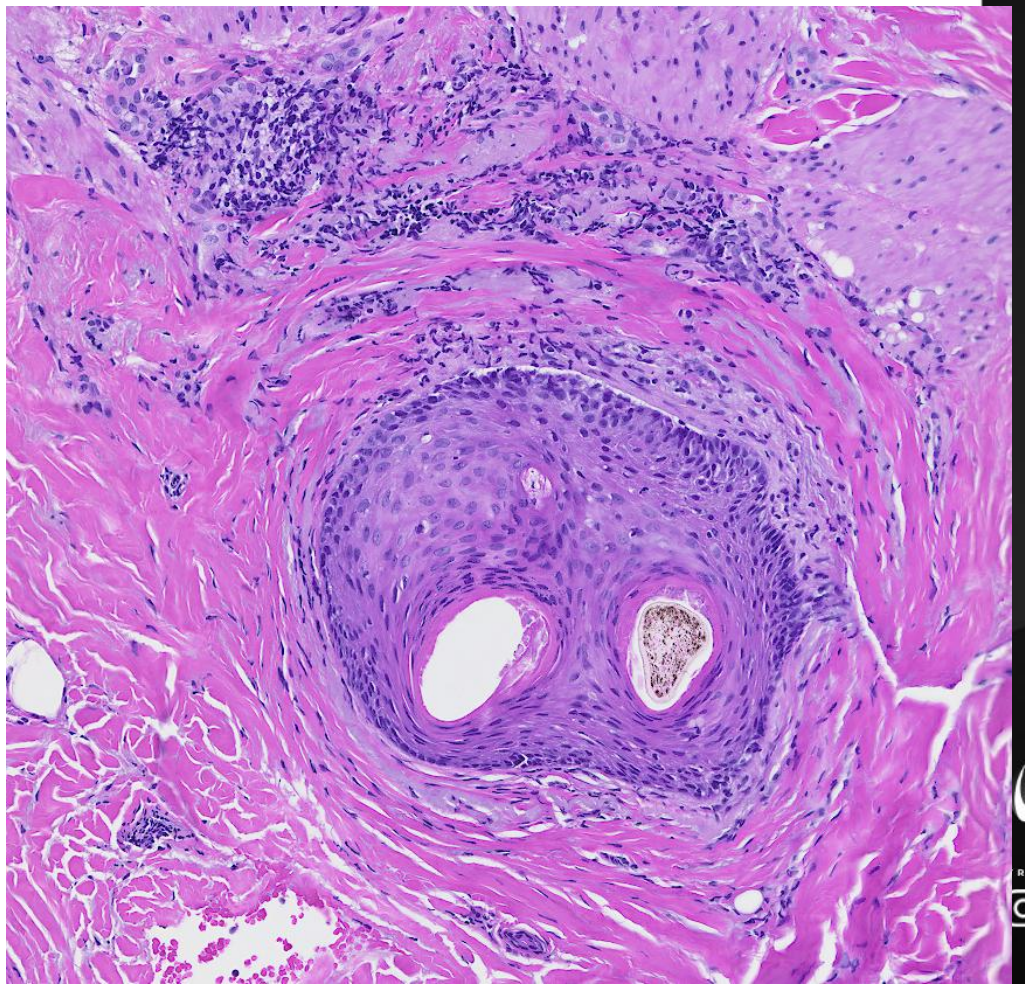


En LPP, hay linfocitos intraepiteliales



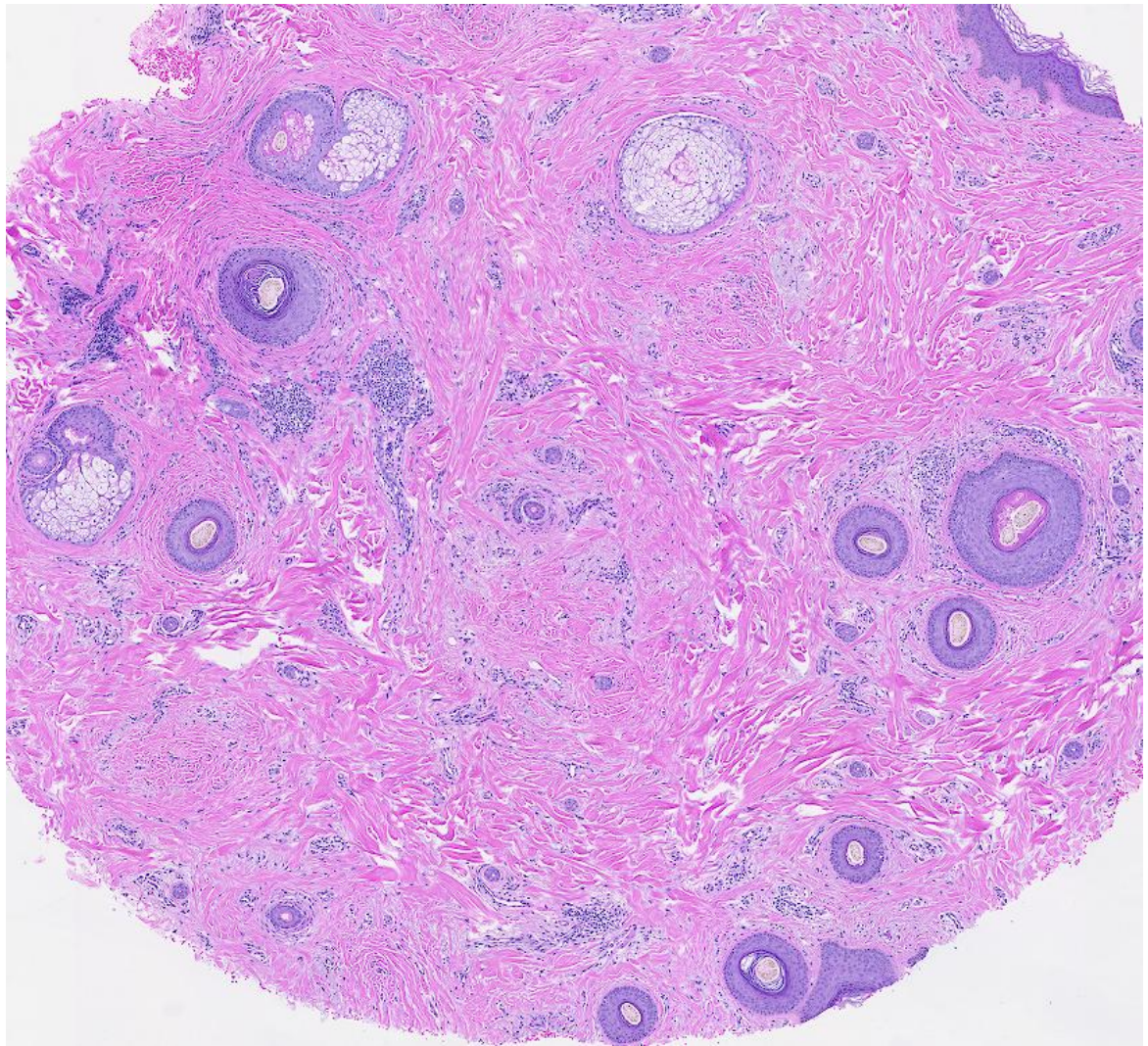
ACCC—Atrofia folicular excéntrica



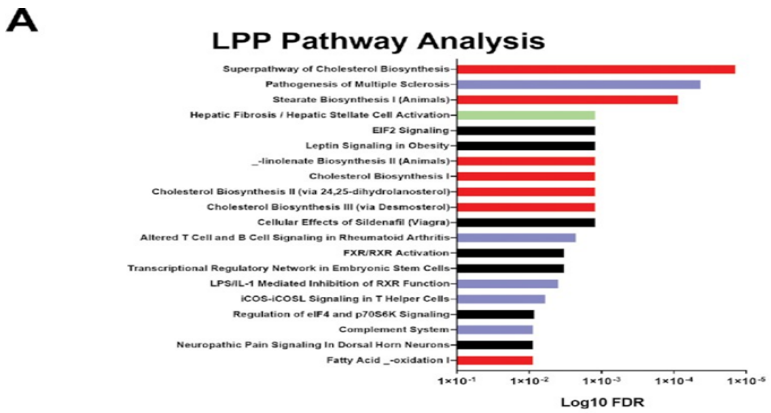


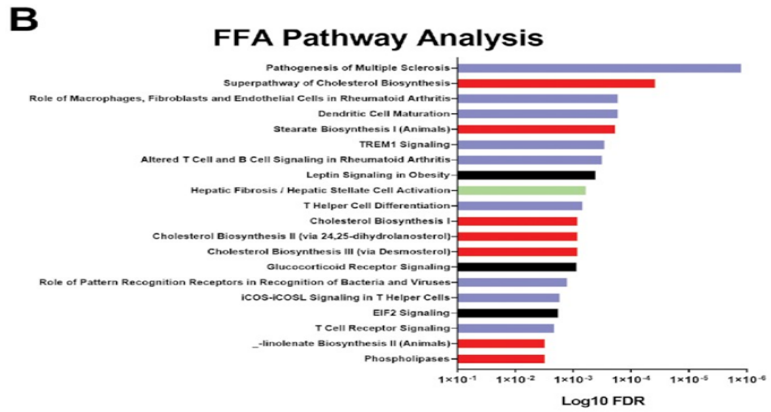
Alopecia cicatricial centrifuga central (ACCC)

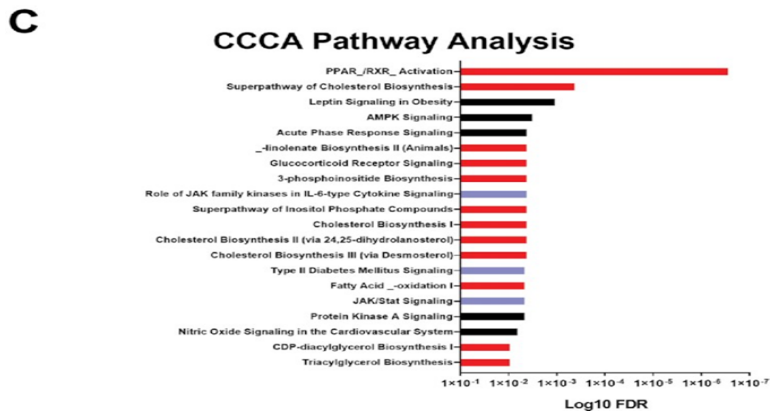
- Menos inflamación
- Más en etapa terminal
- Paciente de ascendencia africana



Comparten vías desreguladas en la biosíntesis del colesterol . . .







Eddy H C Wang, Isha Monga, Brigitte N Sallee, James C Chen, Alexa R Abdelaziz, Rolando Perez-Lorenzo, Lindsey A Bordone, Angela M Christiano, Primary cicatricial alopecias are characterized by dysregulation of shared gene expression pathways, *PNAS Nexus*, Volume 1, Issue 3, July 2022, pgac111.

LPP versus ACCC

- Clínicas especializadas en la pérdida de cabello en mujeres de ascendencia africana.
  - Es importante confirmar la presencia de una alopecia cicatricial primaria.
  - Es importante descartar otras causas (Polilla de la cabeza, etc.)
  - Existen tres presentaciones clinicas:
    - ACCC
    - LPP
    - ACCC/LPP indeterminado

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Eliminar los trastornos que no sean alopecia cicatricial primaria

- Trastornos congénitos (Existen tantos tipos)
  - Keratosis follicularis spinulosa decalvans
- Envejecimiento/daño solar
  - Dermatosis pustulosa y erosive del cuero cabelludo (DEPCC)
- Acneiforme
  - Folliculitis (acne) necrotica
- Trastornos linfoproliferativos
  - Alopecia mucinosa (follicular mucinosis)

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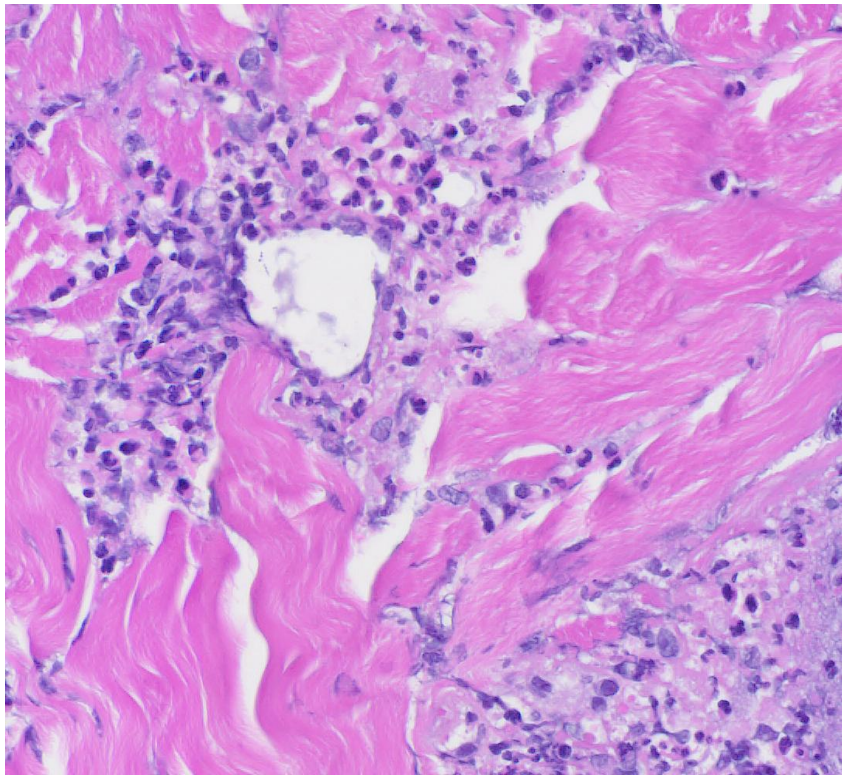
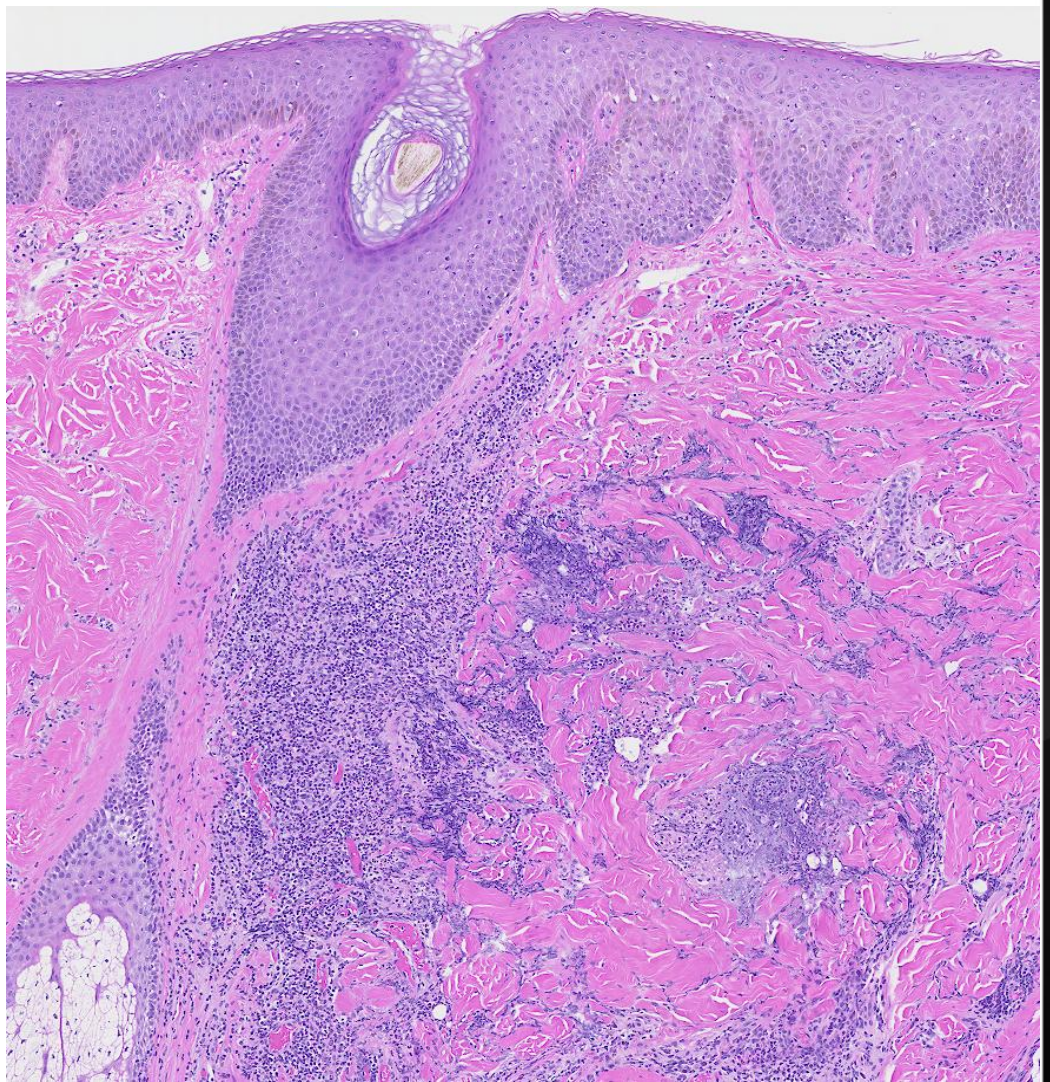
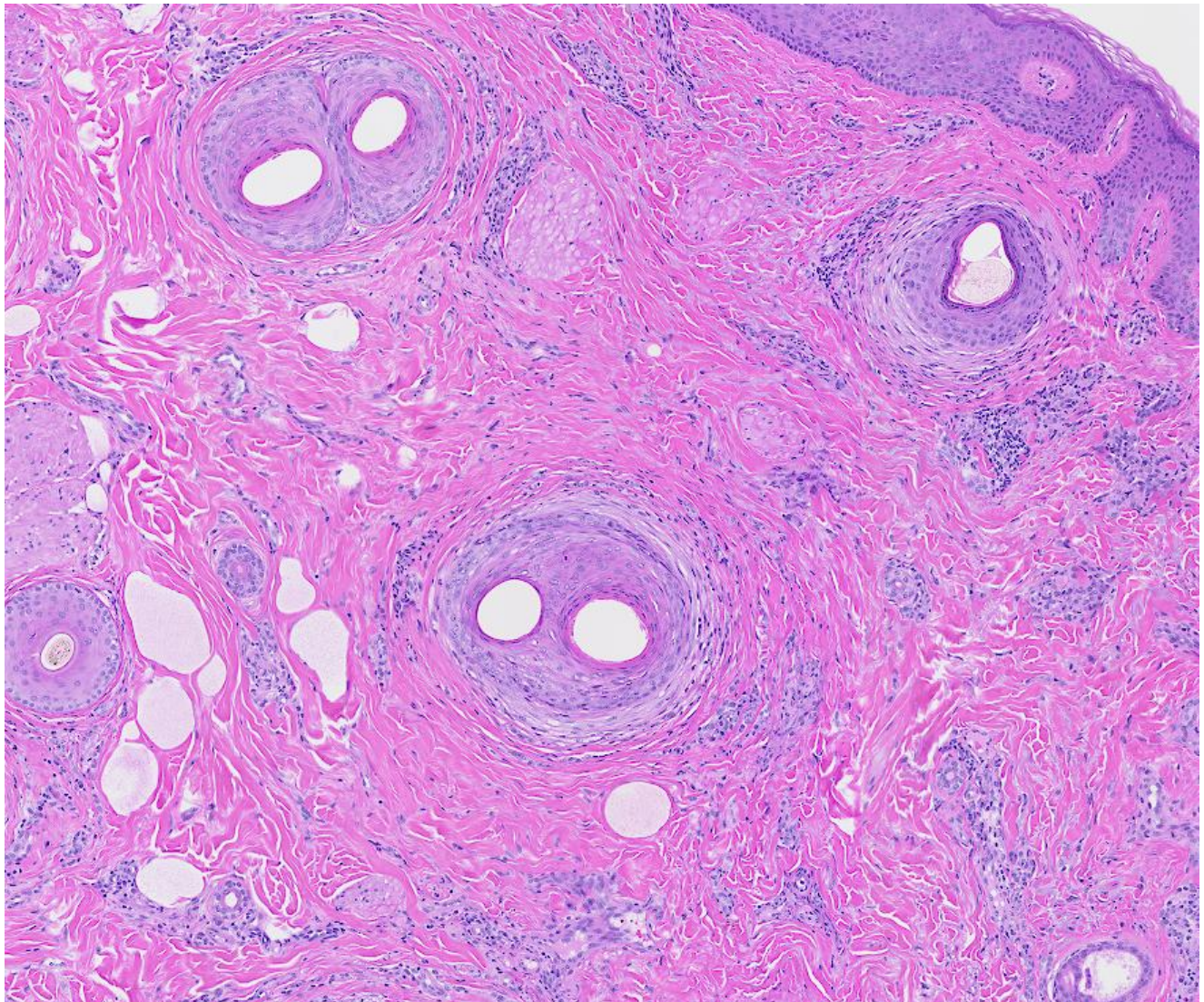
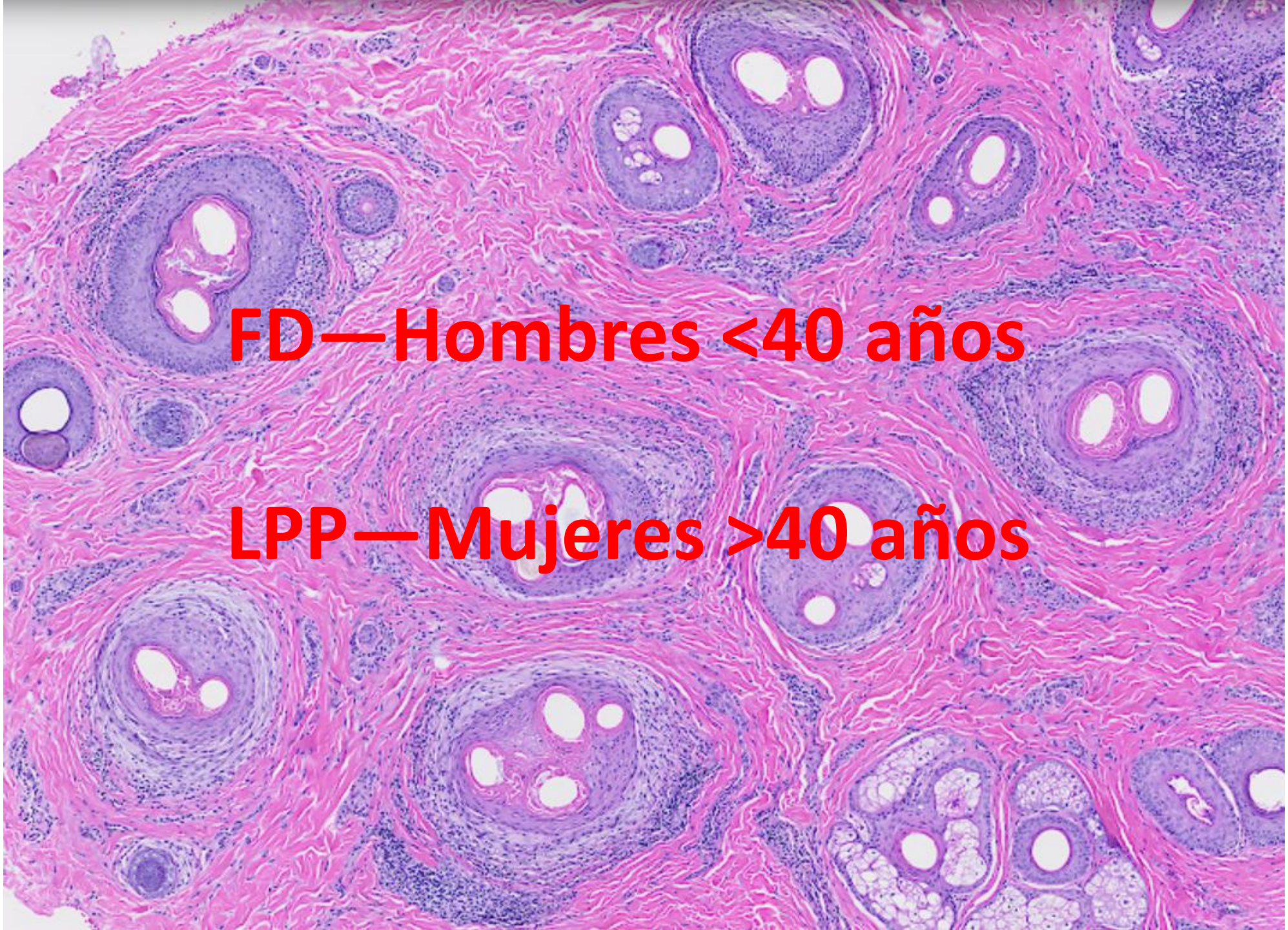
Foliculitis decalvante
En etapa temprana--Neutrofilica



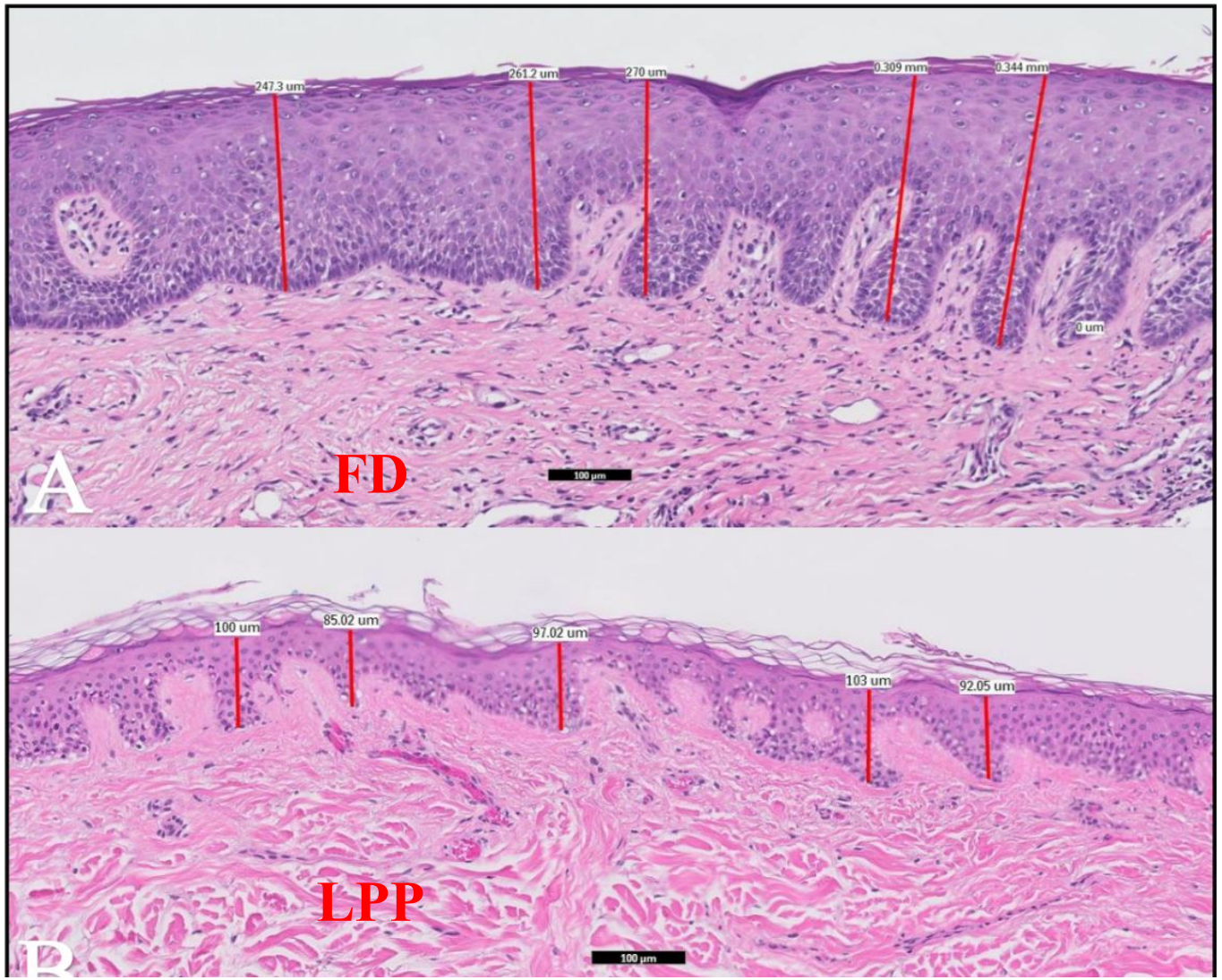


Folliculitis decalvante
En etapa avanzada--Linfocítica





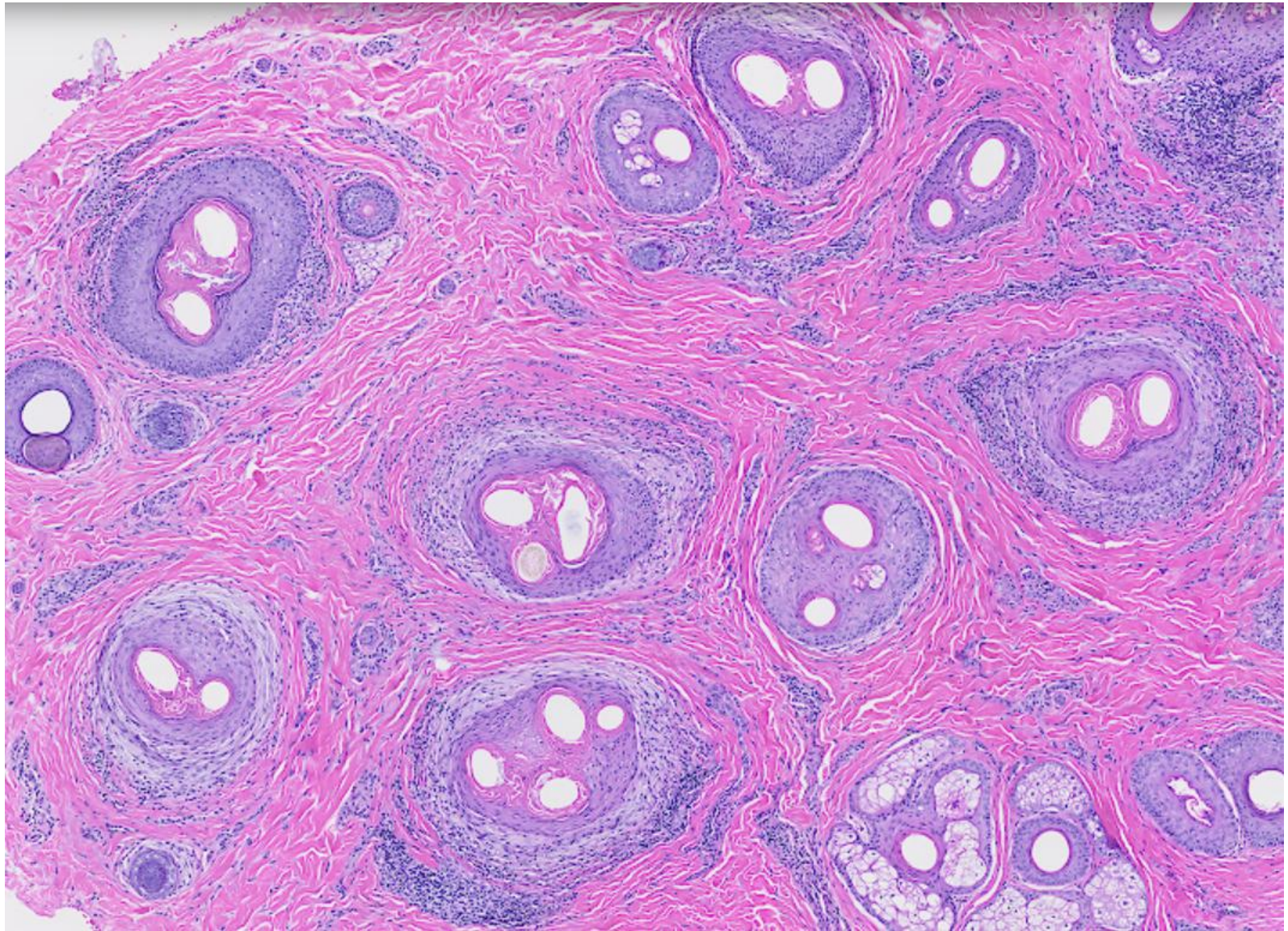
Espesor epidérmico
Folliculitis decalvante vs LPP



Bohnett MC, Kolivras A, Thompson AA, Thompson CT. *J Cutan Pathol*. 2021;48(6):816-818.

Foliculitis decalvante versus LPP

Folículos en penacho (>3 tallos)



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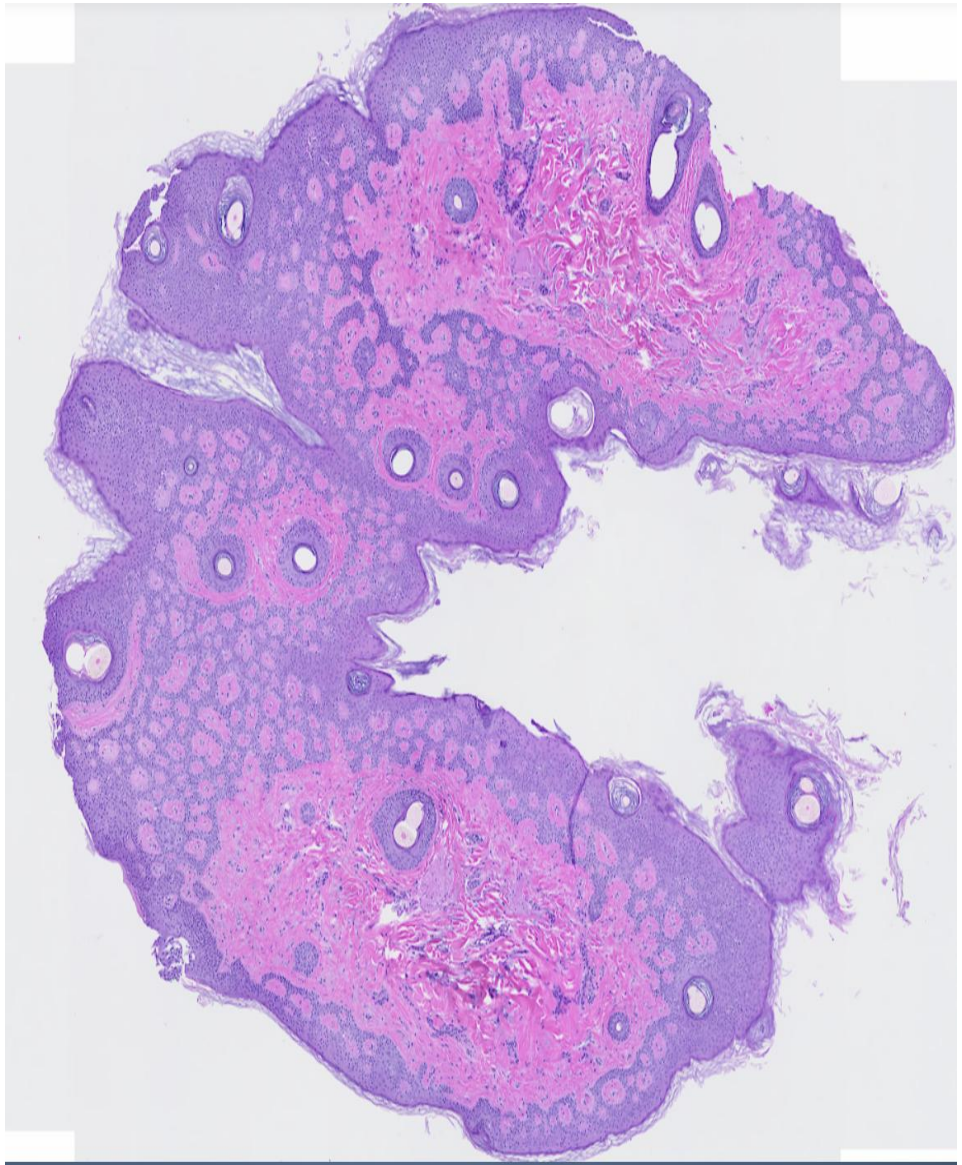
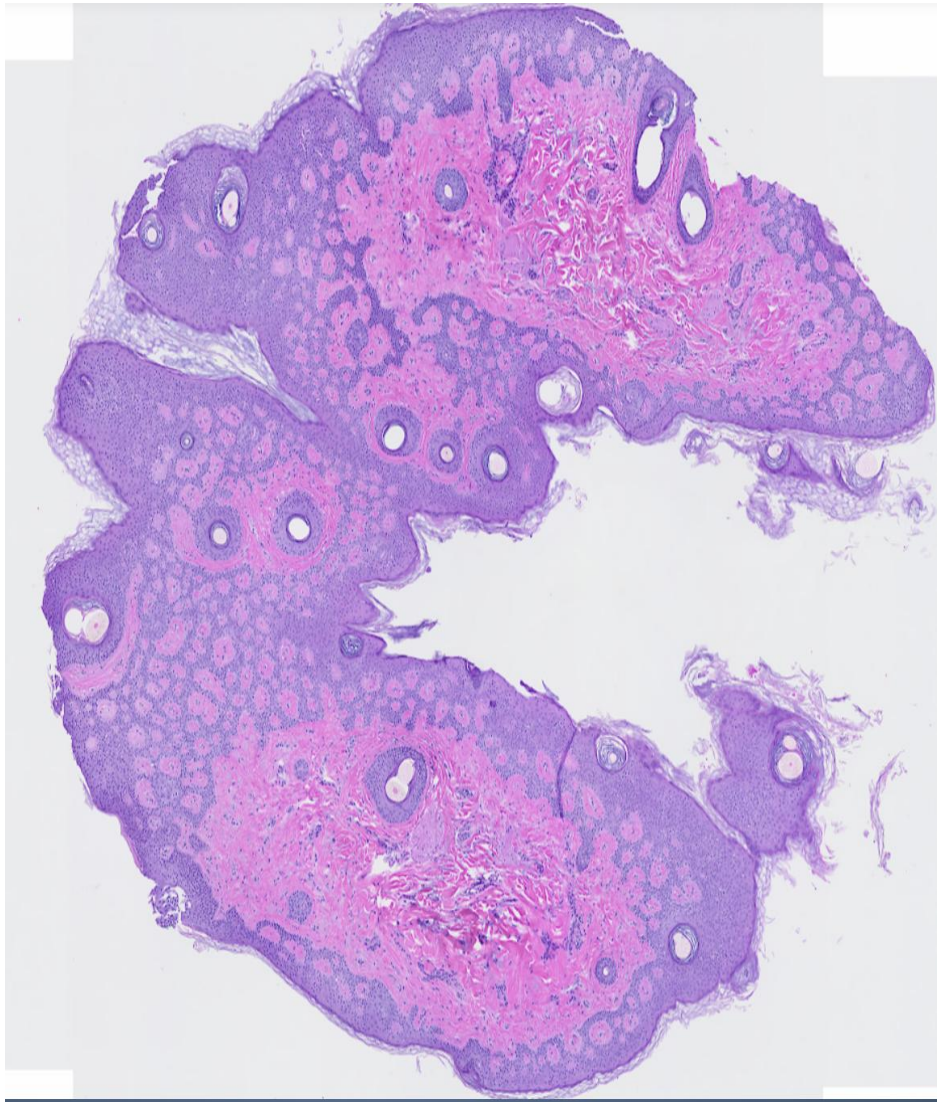
Algunas pistas diagnósticas . . .

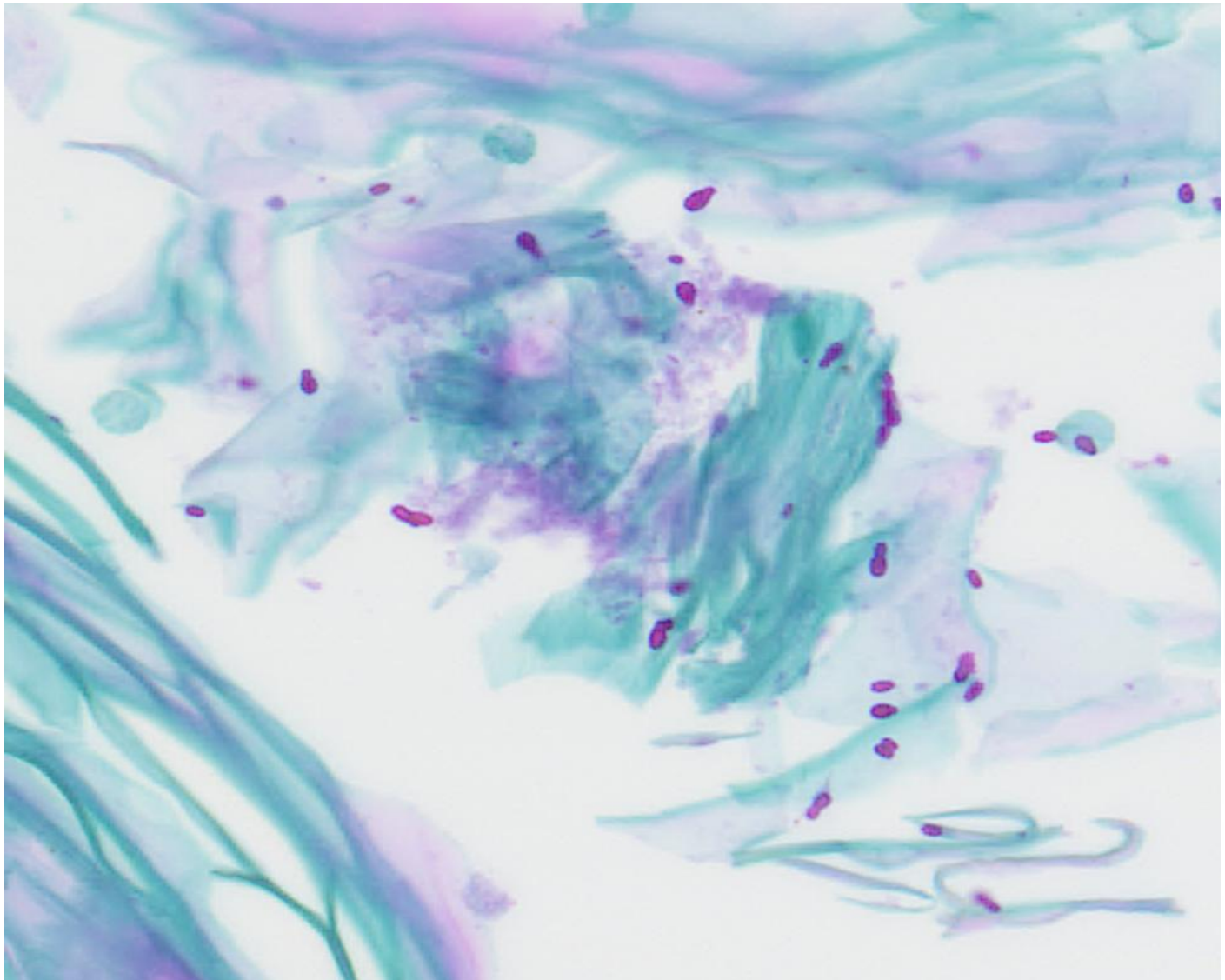


Pista #1

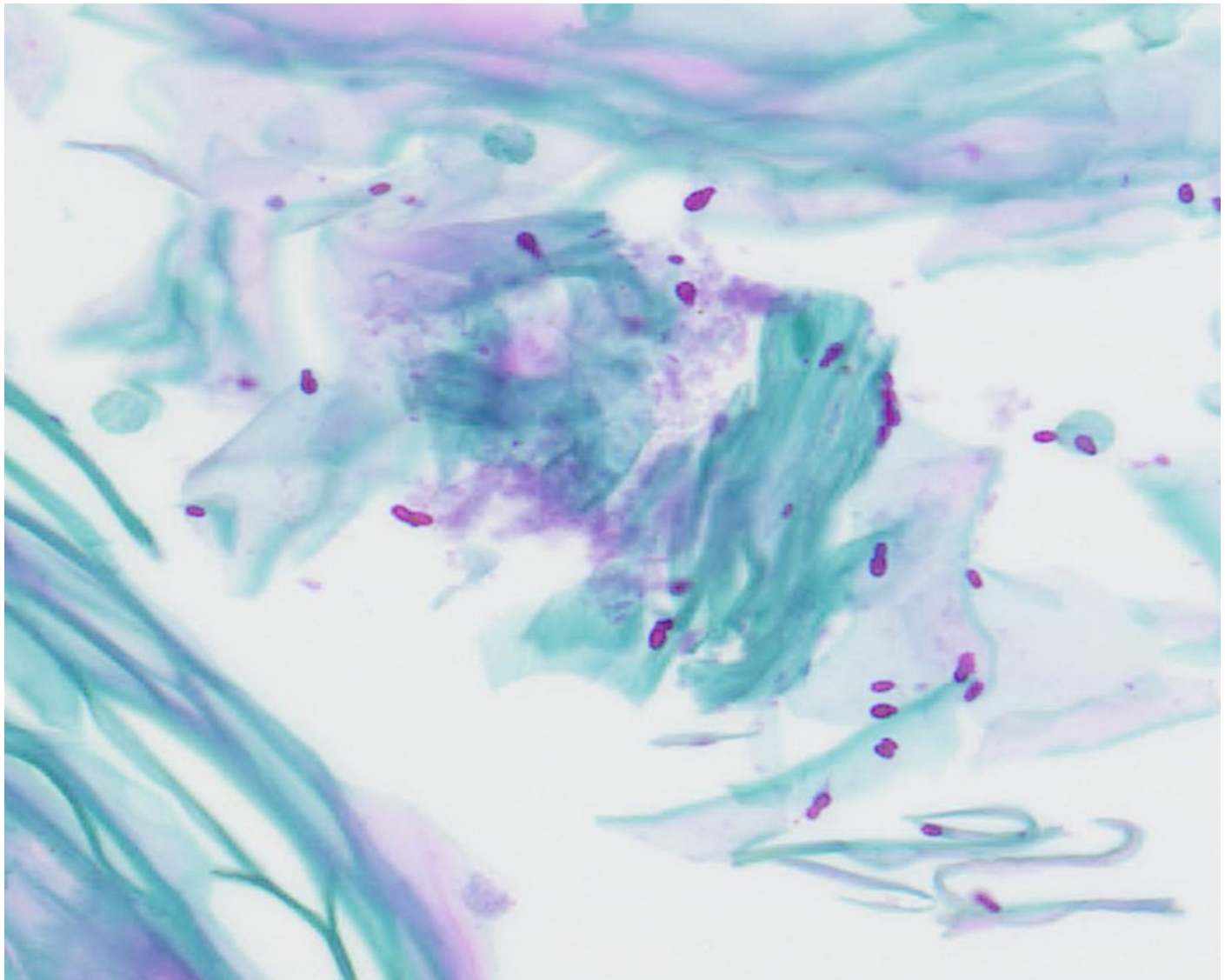
Hiperqueratosis (no parakeratosis) es una pista para el diagnóstico de la dermatitis seborreica del cabello.



Levadura malassezia



Levadura malassezia



Histologic absence of yeast as a clue for classic lichen planopilaris, fibrosing alopecia in a pattern distribution, and frontal fibrosing alopecia: A cross-sectional observational study

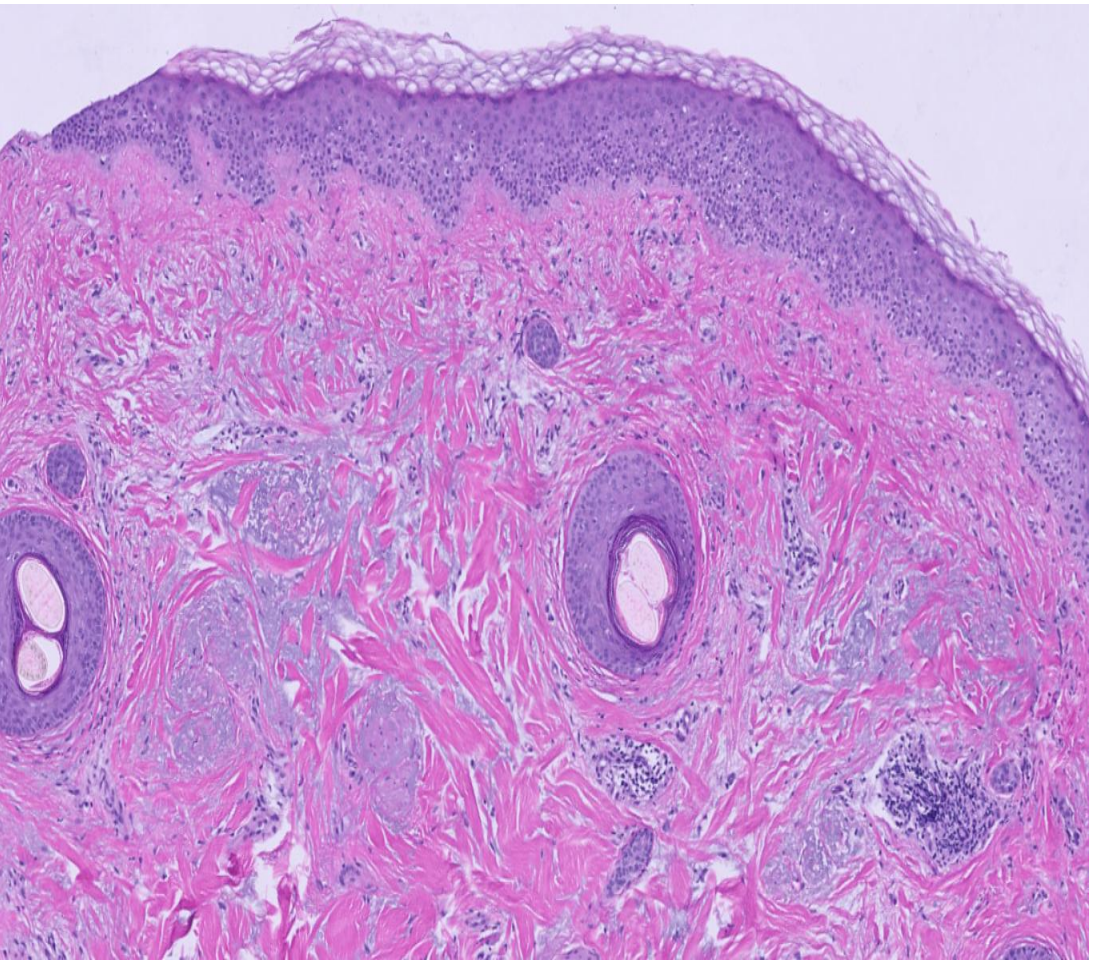
Kimberly Williams, BS [a] · Antonella Tosti, MD [a] · Curtis T. Thompson, MD [b,c]

JAAD International, Volume 19, 10 - 11

There was a distinct difference between the absence and presence of yeast between cases of LPP/FAPD/FFA and FPHL. In LPP/FAPD/FFA, 98.5% (68/69) of cases had no identifiable yeast. In contrast, in FPHL 50% (34/68) of cases had identifiable yeast ($P < .001$).

- 98.5% de LPP/FAPD/FFA sin levadura
- 50% de CPF tiene levadura

Sin sebo = Sin levadura



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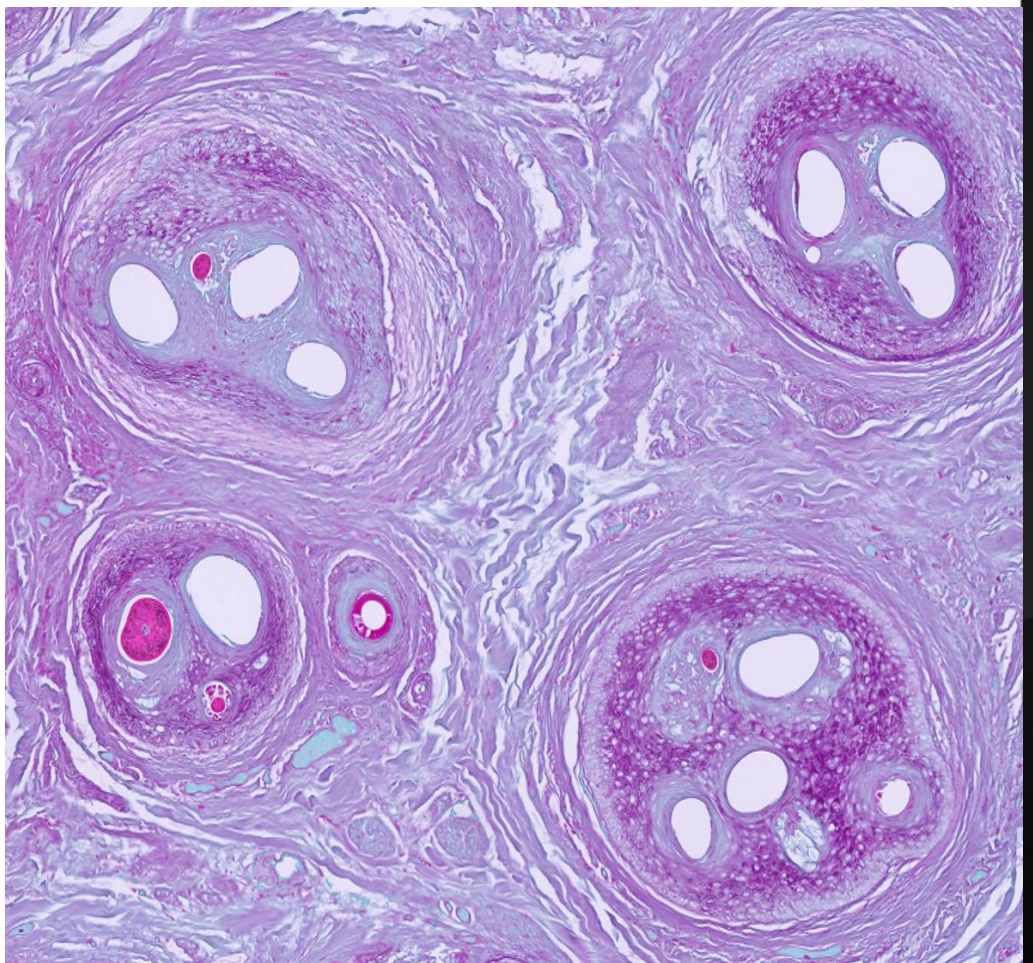
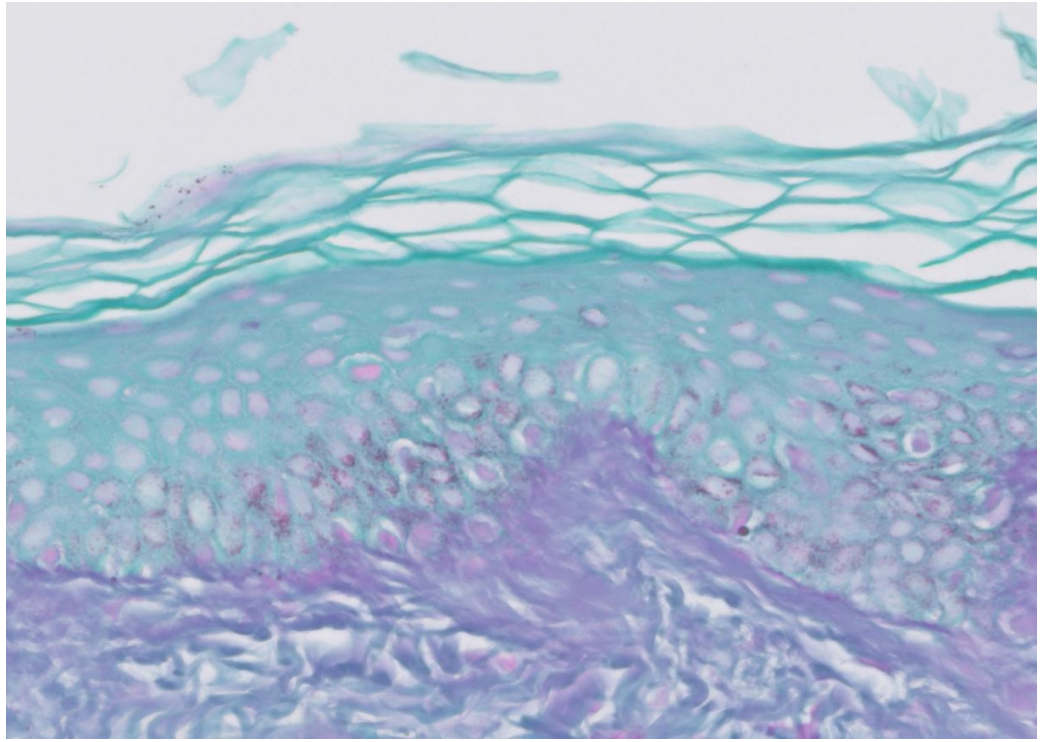


La ausencia de levadura es una pista al diagnóstico de LPP/AFF/ACCC/FAPD



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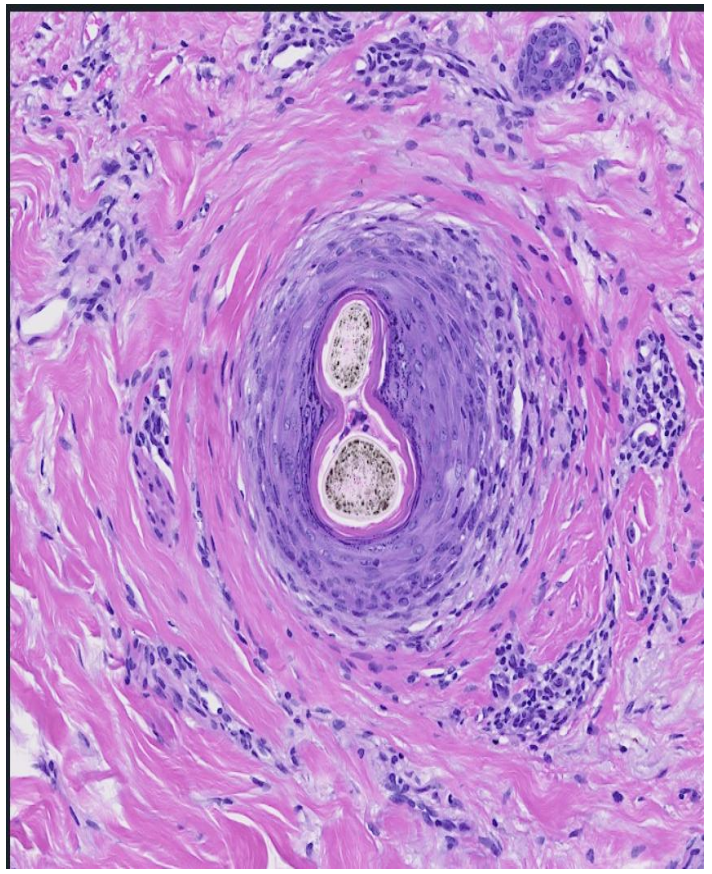
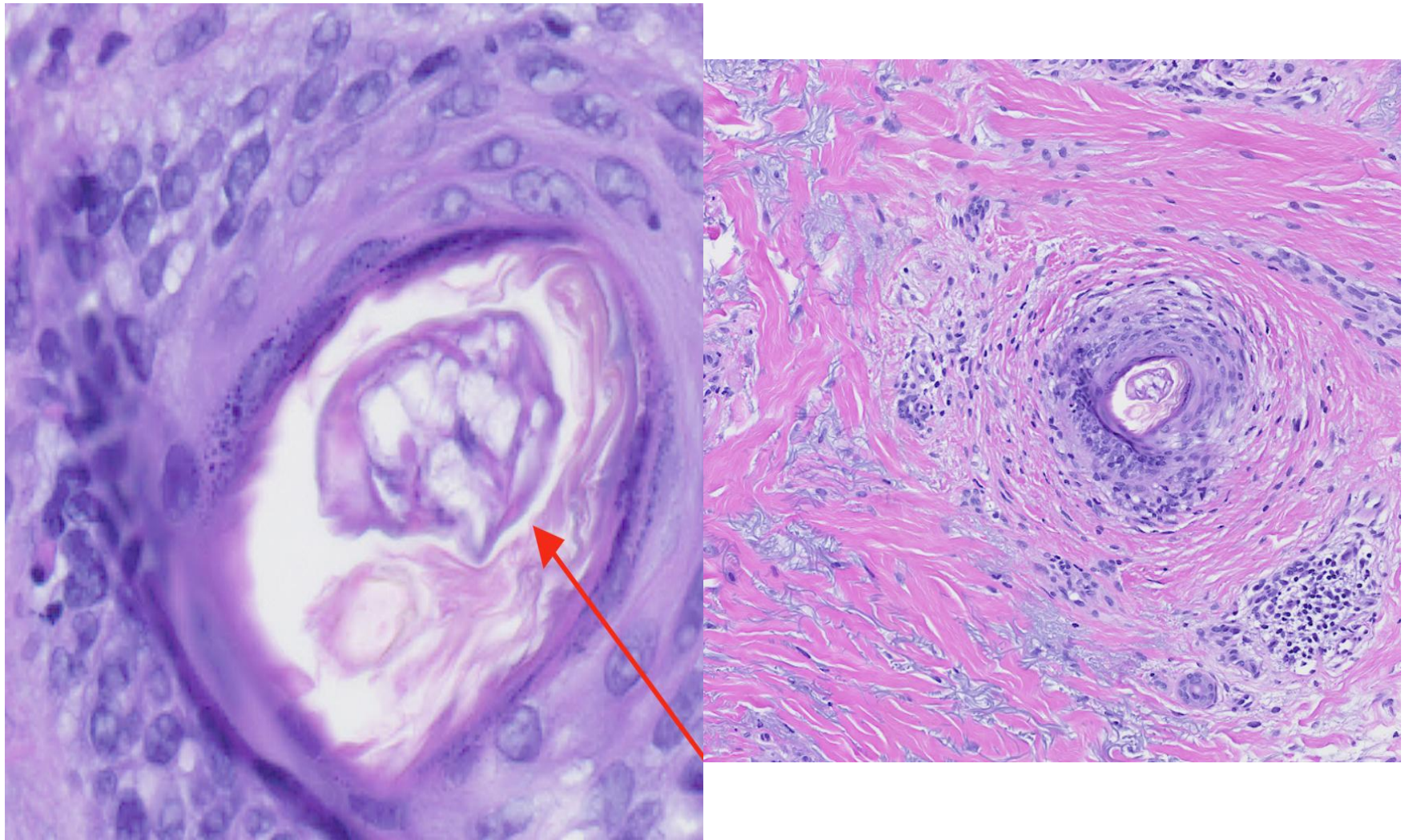
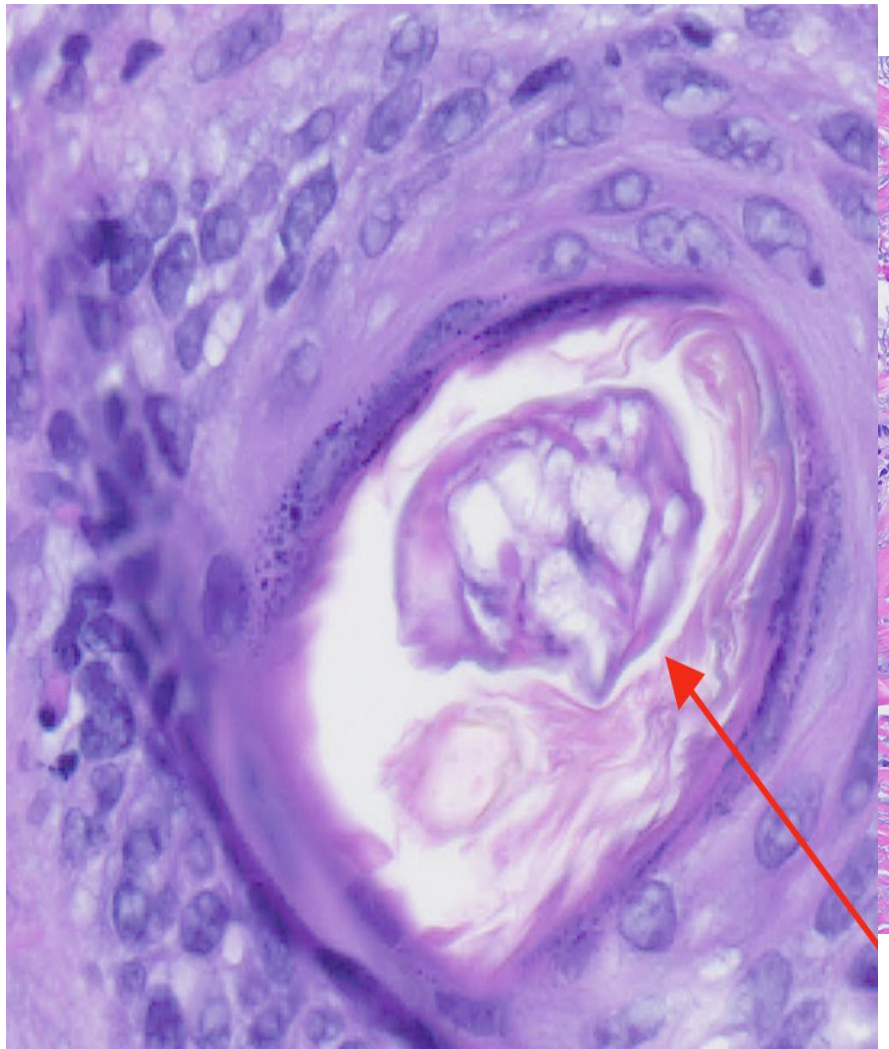


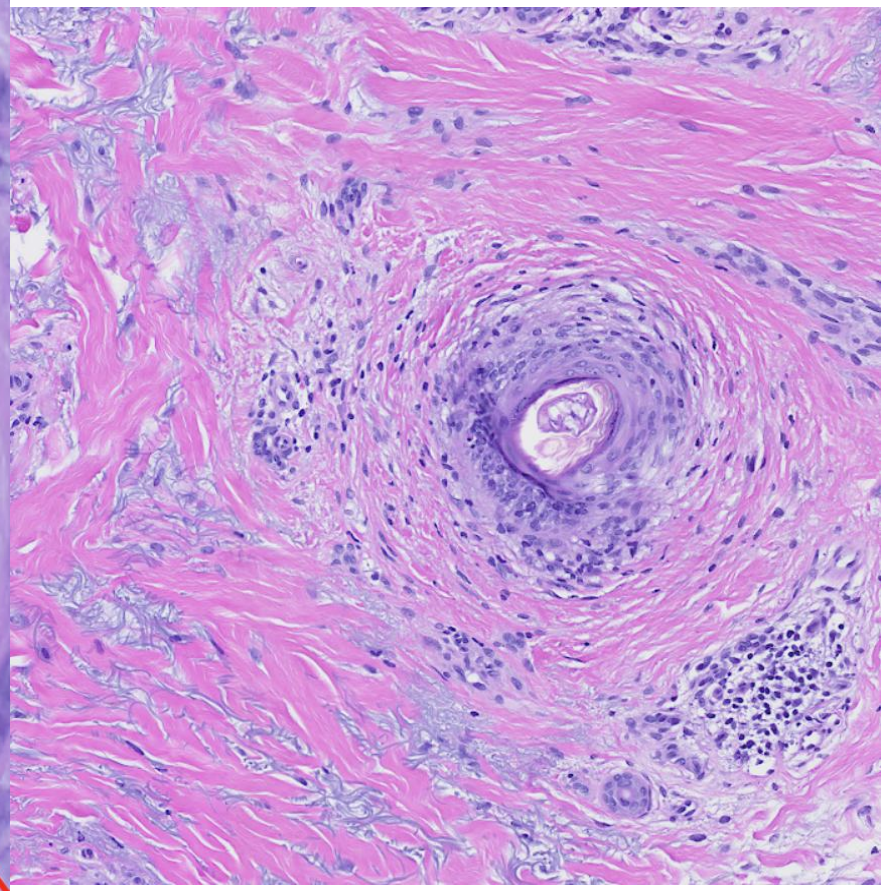
Pista #2
Demodex es una causa común de inflamación y prurito.



Pista #2

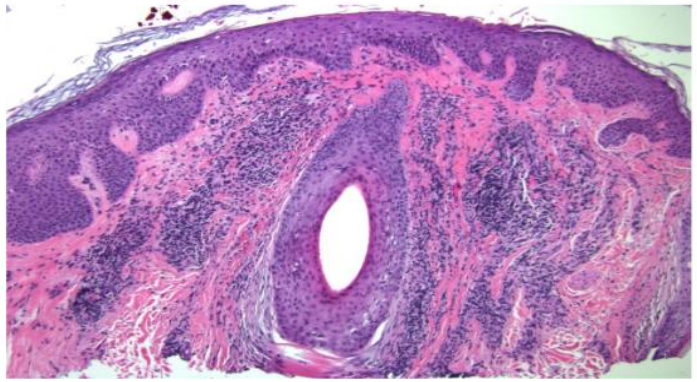
Es necesario explanar la causa de la inflamación perifolicular para prevenir un diagnóstico de una alopecia cicatricial sutil

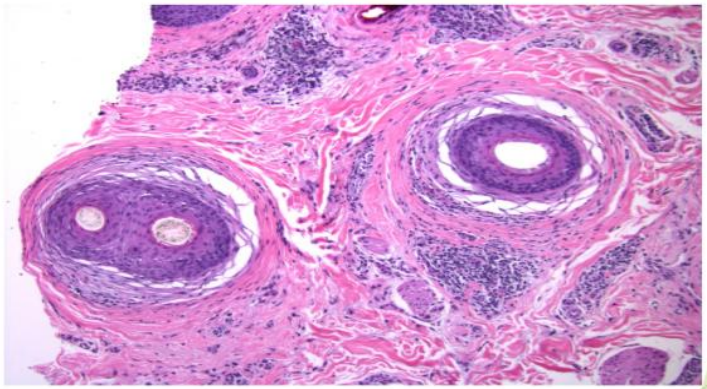




Pista #3
casi total ausencia de los folículos en etapa catágena o telógena.

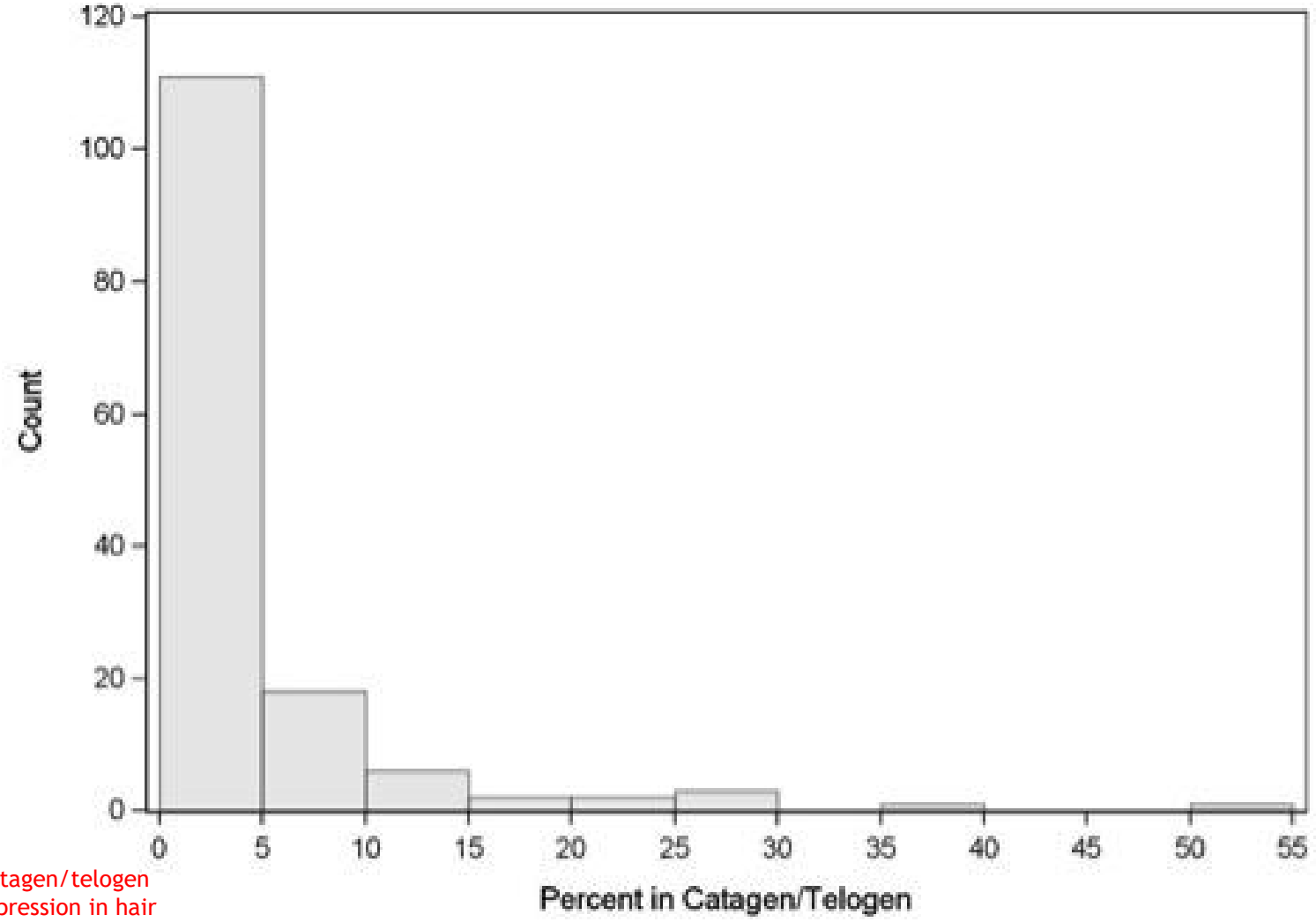
Lichen Planopilaris (LPP)





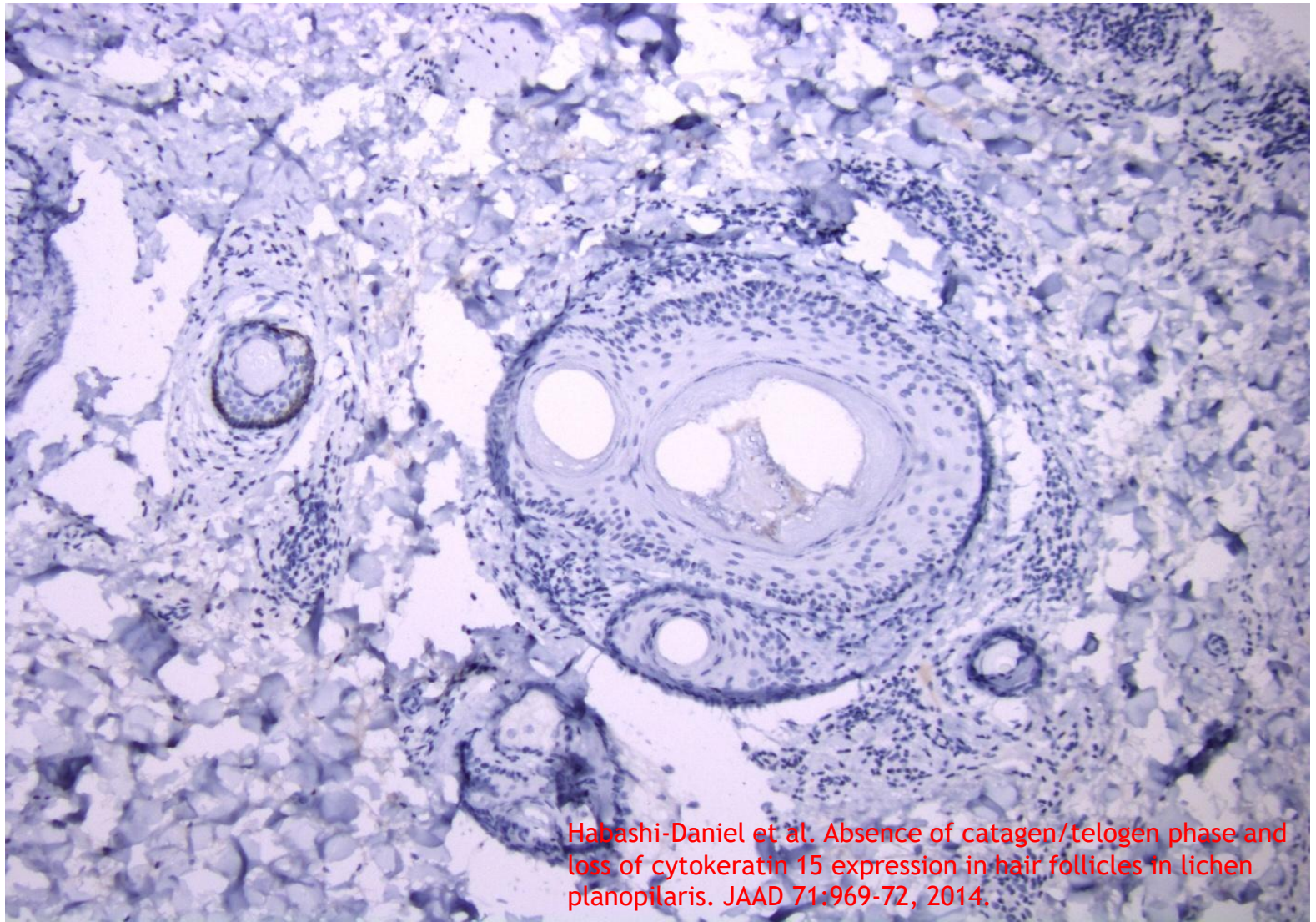
Loss of catagen/telogen phase follicles

Casi total ausencia de los folículos en etapa catágena o telógena en LPP



Habashi-Daniel et al. Absence of catagen/telogen phase and loss of cytokeratin 15 expression in hair follicles in lichen planopilaris. JAAD 71:969-72, 2014.

La pérdida de citoqueratina 15 en células madre del folículo



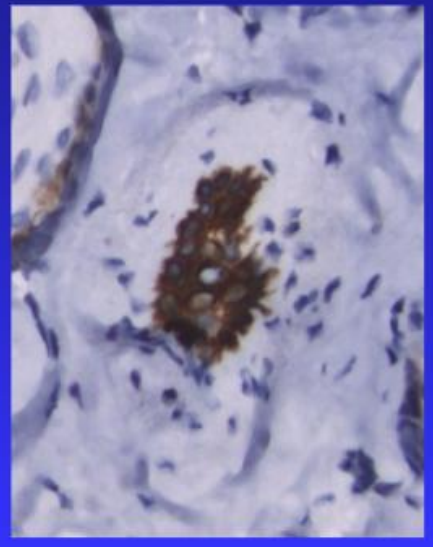
Habashi-Daniel et al. Absence of catagen/telogen phase and loss of cytokeratin 15 expression in hair follicles in lichen planopilaris. JAAD 71:969-72, 2014.

Lichen Planopilaris Progression

Loss of CK15+ stem cells

↓

Disappearance of follicle when cycle into catagen



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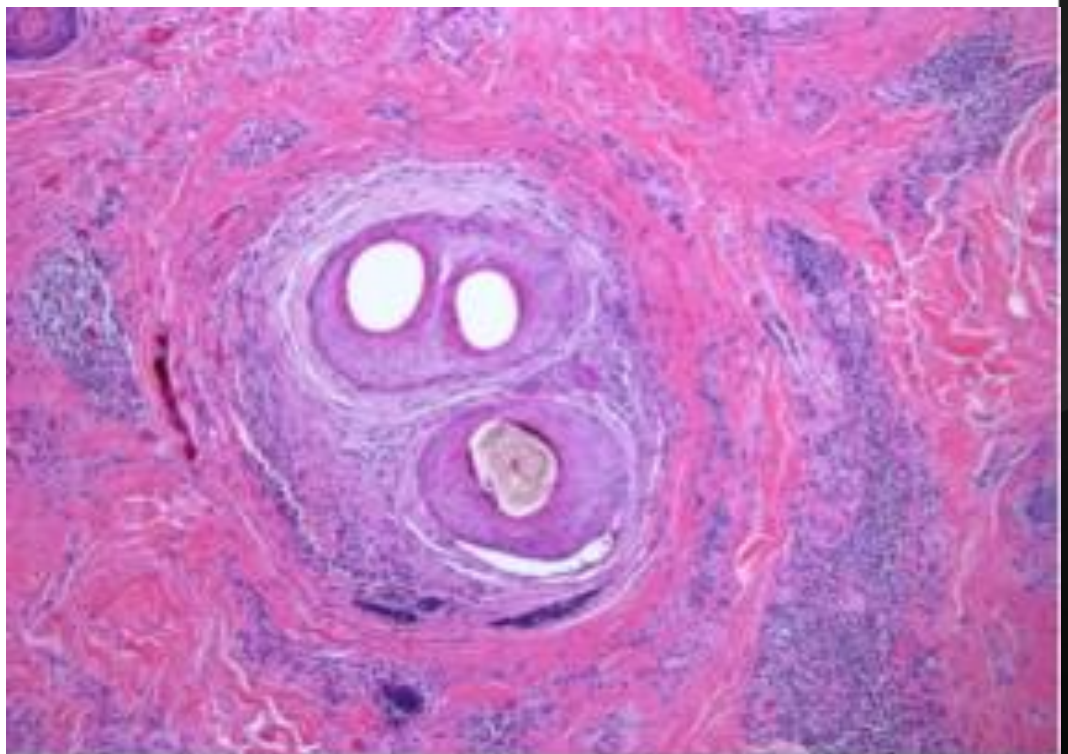
Clinical progression despite immunosuppressive treatment

Unas mas pistas . . .

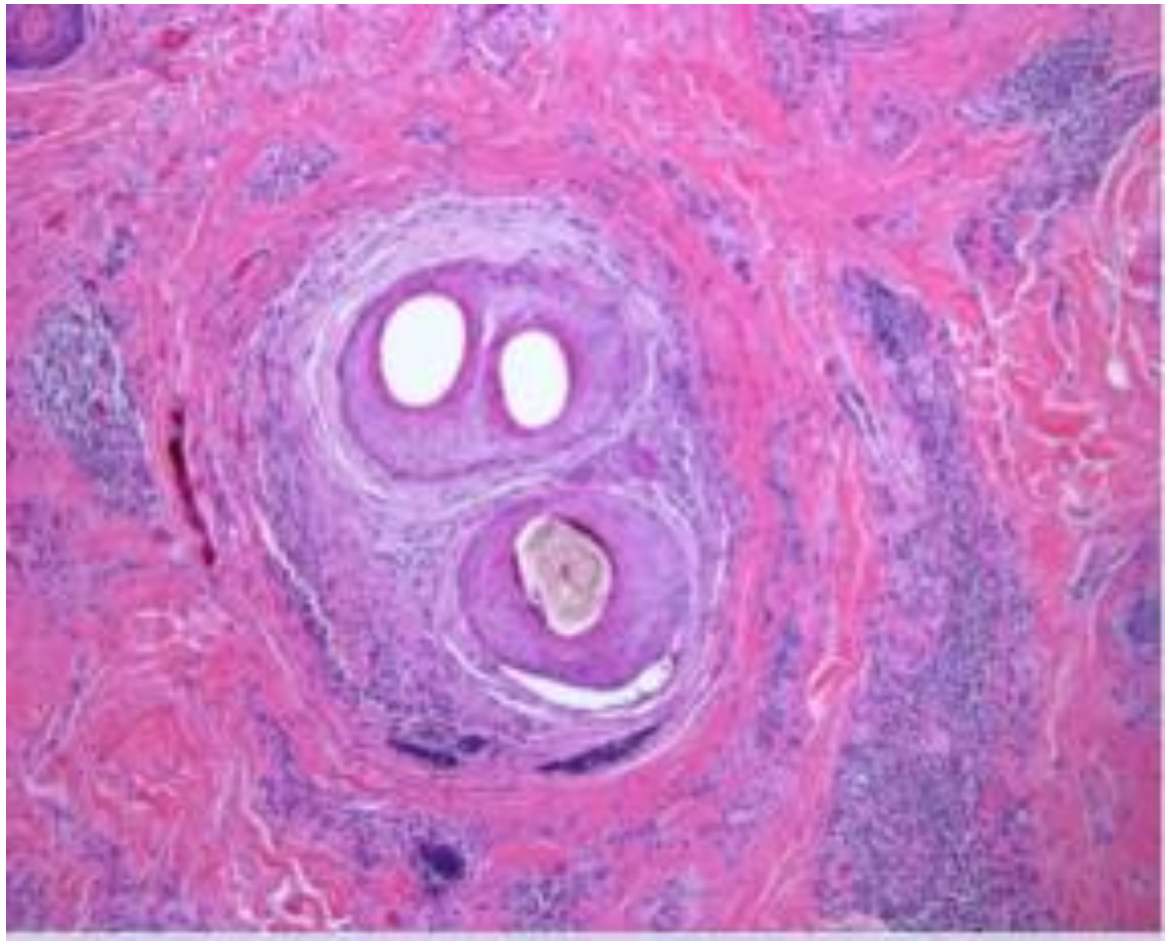
- La información más importante de la clínica:
  - Una calva o la pérdida difusa?
  - ‘Pull test’ positiva o la caída del pelo?
- Tricotilomanía siempre tiene liquen simple crónico (LSC).

La salida común de los tallos es normal y es importante que no haga un diagnosis del LPP.

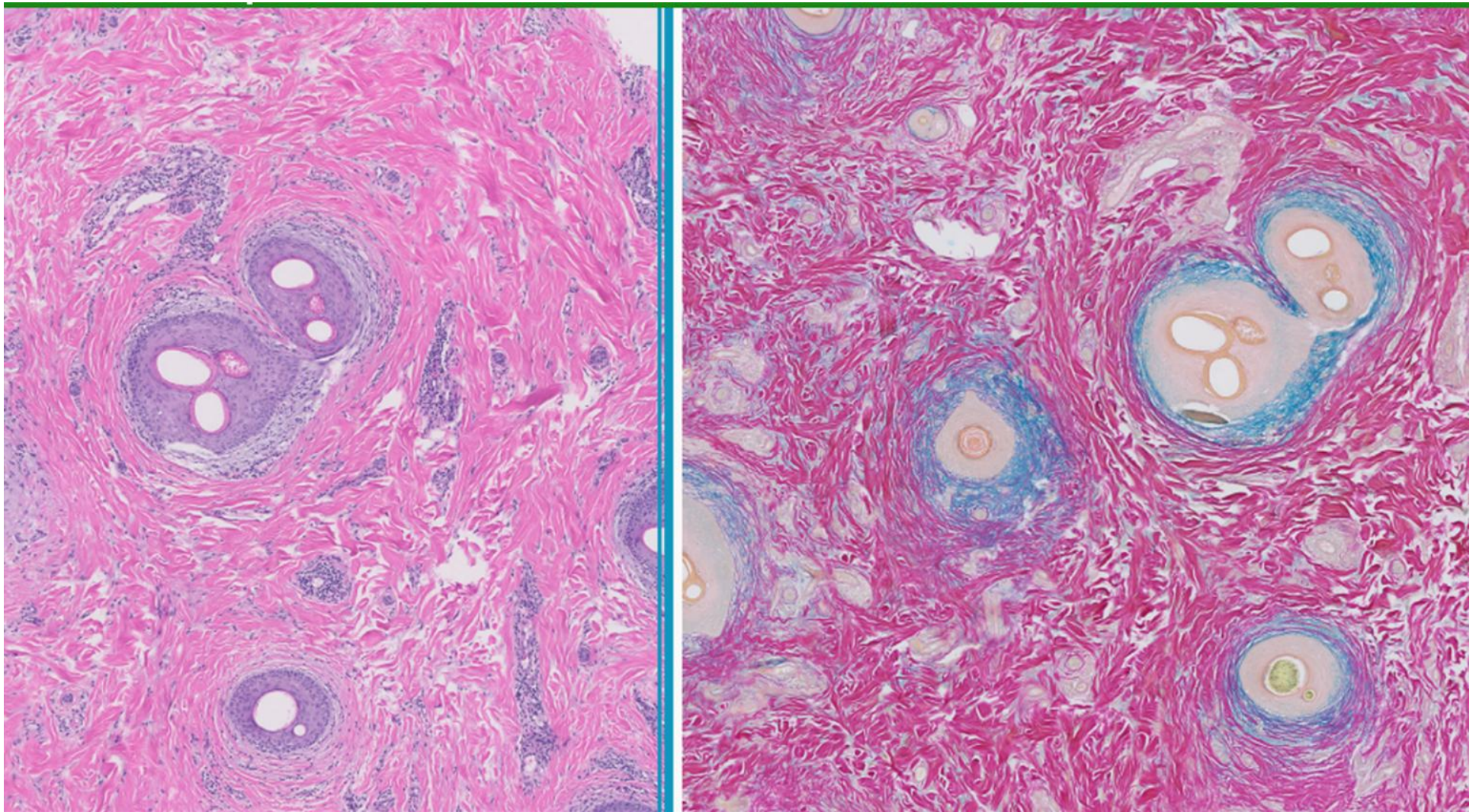
≠



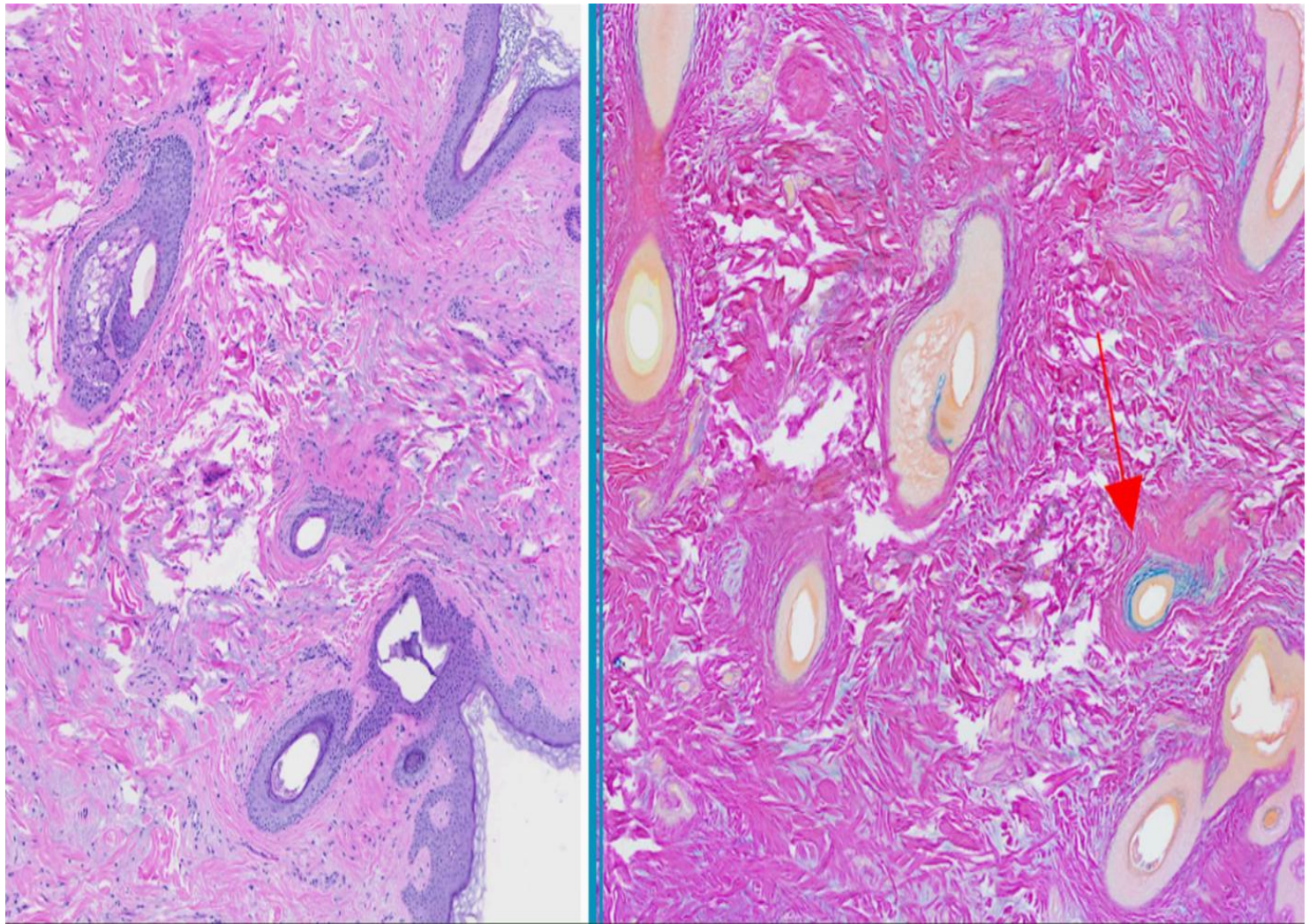
Una lesión acneiforme en resolución tiene la histopatología del LPP o FD.



La tinción especial “Hierro Coloidal” identifica muy bien la fibrosis perifolicular en la alopecia cicatricial.



La tinción ‘Hierro Coloidal’ identifica la fibrosis perifolicular sutil en casos de AFF.



La causa de AFF

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.

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

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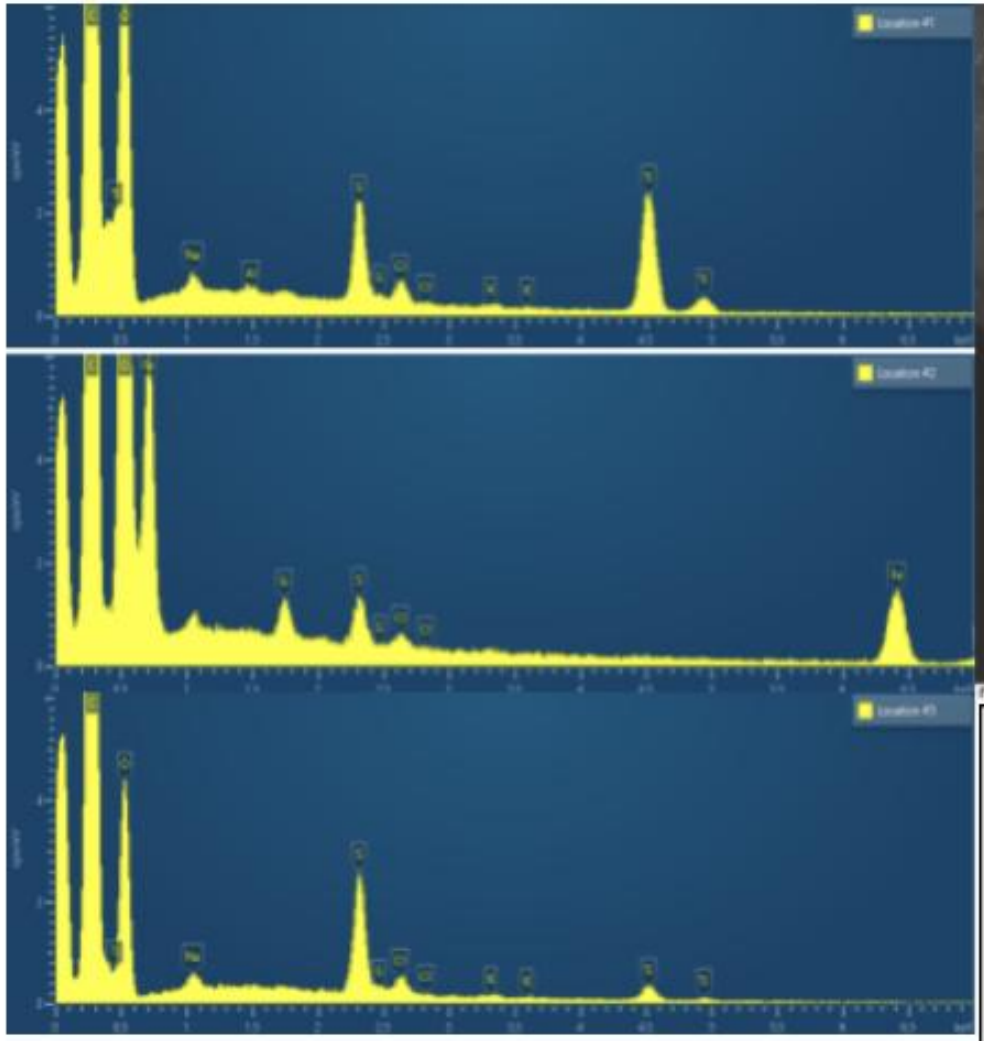

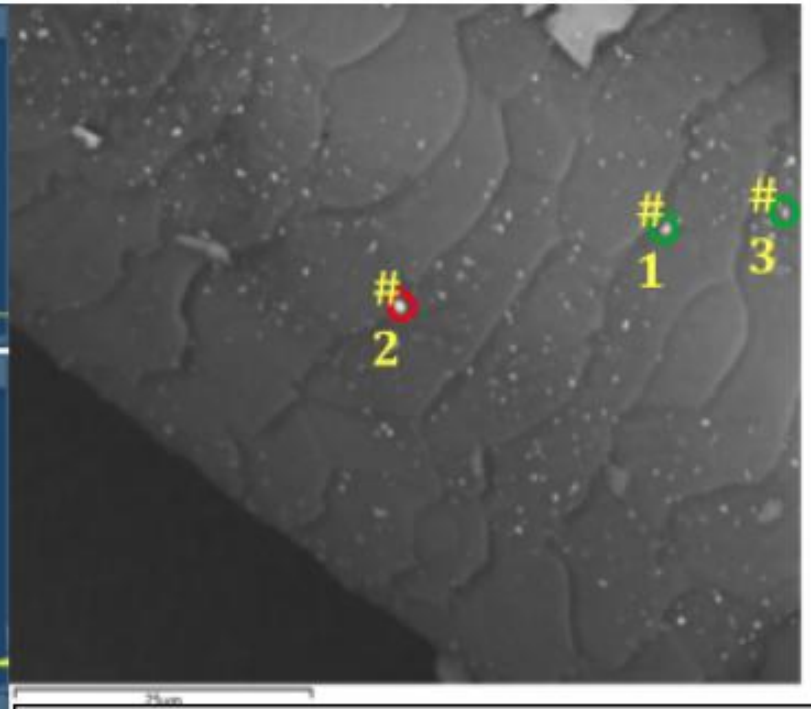

Fig. 1 Right: a backscattered electron image taken from one of typical hair shafts with SEM showing the presence of particles with brighter contrast on a hair shaft; Left: EDX spectra collected from particle 1-3 showing the presence of Ti species on particle 1 and 3.

Sur

La causación de AFF

Meta-Analysis > Arch Dermatol Res. 2023 Oct;315(8):2313-2331.
doi: 10.1007/s00403-023-02604-7. Epub 2023 Apr 4.

Frontal fibrosing alopecia and personal care product use: a systematic review and meta-analysis

Olivia Kam [1], Sean Na [2], William Guo [3], Christina I Tejeda [3], Tara Kaufmann [3]

Un sumario de nuevas investigaciones.
1,248 pacientes de FFA
1,459 controles negativos.

La causación de FFA
(9 investigaciones)

- Protector solar OR 3.02
  - 4.61 (hombres)
  - 2.74 (mujeres)
- Crema facial
  - 5.07 (hombres)
  - 1.58 (mujeres)

Kam O, Na S, Guo W, Tejeda CI, Kaufmann T. Frontal fibrosing alopecia and personal care product use: a systematic review and meta-analysis. Arch Dermatol Res. 2023 Oct;315(8):2313-2331.

Sin asociación

- Limpiador y tónico facial, aftershave
- Base de maquillaje
- Champú, acondicionador, espuma, gel, tinte, productos químicos para alisar o rizar

Kam O, Na S, Guo W, Tejeda CI, Kaufmann T. Frontal fibrosing alopecia and personal care product use: a systematic review and meta-analysis. Arch Dermatol Res. 2023 Oct;315(8):2313-2331.

Meta-Analysis > JAMA Dermatol. 2025 Mar 1;161(3):310-314.
doi: 10.1001/jamadermatol.2024.6434.

Epistasis of ERAP1 With 4 Major Histocompatibility Complex Class I Alleles in Frontal Fibrosing Alopecia: A Genome-Wide Association Study Meta-Analysis

- 6668 pacientes incluidos
  - 1585 AFF (femeninas europas)
  - 5083 controles

Una predisposición genética a AFF

- Genome-wide significant associations at 4 genomic loci
  - HLA-A*11:01, HLA-A*33:01, HLA-B*07:02, and HLA-B*35:01.
  - ERAP1 gene mutation at 5q15

Rayinda T, Dand N, McSweeney SM, Christou E, Ung CY, Stefanato CM, Fenton DA, Harries M, Palamaras I, Tidman A, Holmes S, Koutalopoulou A, Ardern-Jones M, Kaur M, Papanikou S, Chasapi V, Vañó-Galvan S, Saceda-Corralo D, Melián-Olivera A, Azcarraga-Llobet C, Lobato-Berezo A, Bustamante M, Sunyer J, Starace MVR, Piraccini BM, Wiss IP, Senna MM, Singh R, Hillmann K, Kanti-Schmidt V, Blume-Peytavi U, McGrath JA, Simpson MA, Tziotzios C. Epistasis of ERAP1 With 4 Major Histocompatibility Complex Class I Alleles in Frontal Fibrosing Alopecia: A Genome-Wide Association Study Meta-Analysis. JAMA Dermatol. 2025 Mar 1;161(3):310-314.

Resumen de la causación de AFF

- Protector solar (mujeres y hombres)
- Crema facial (hombres)
- Una predisposición genetica
  - 40% con uno de los 4 loci genómicos
    - HLA-A*11:01, HLA-A*33:01, HLA-B*07:02, and HLA-B*35:01.
    - (ERAP1 gen a 5q15)

Hipotesis
Son todos resultado de la misma química?

- LPP
- AFF
- FAPD
- ACCC
- Liquen plano pigmentoso

Otra Hipótesis

- ¿Permite la protección solar que se produzca una inflamación crónica?
- De ser así, diferentes productos de protección solar podrían favorecer la LPP y la FFA, especialmente en personas genéticamente susceptibles.

¡Muchas Gracias!

- University of Miami
  - Antonella Tosti, Betty Nguyen and Kimberly Williams
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  - Paradi Mirmirani and Vera Price
- Belgium
  - Athanassios Kolivras
- Mexico
  - Abril Martinez Velasco, Daniel Asz-Sigal, and Hassiel Ramirez
- Cicatricial Classification Working Group
  - Elise Olsen, Wilma Bergfeld, Dirk Elston, Lynne Goldberg, Eleanor Knopp, Mariya Miteva, John Seykora, Michelle Schneider, Len Sperling

curtis@ctapathology.com

Mahalo!

¡Gracias!

Merci beaucoup!