

# Pathology of Primary Cicatricial Alopecia

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and

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# Lecture Focus

- Utility of Proposed Working Classification of Primary Cicatricial Alopecia (2001)
- Histopathologic clues to the diagnosis of primary cicatricial alopecia

## SPECIAL REPORT

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# **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,<sup>a</sup> Wilma F. Bergfeld, MD,<sup>b</sup> George Cotsarelis, MD,<sup>c</sup> Vera H. Price, MD,<sup>d</sup> Jerry Shapiro, MD,<sup>c</sup> Rodney Sinclair, MD,<sup>f</sup> Alvin Solomon, MD,<sup>g</sup> Leonard Sperling, MD,<sup>h</sup> Kurt Stenn, MD,<sup>i</sup> David A. Whiting, MD,<sup>j</sup> 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*

# Proposed Working Classification

“The following classification is considered a working classification to facilitate discussion and sharing of clinical information.”

Olson EA et al. J Am Acad Dermatol 48:103-10, 2003

# Proposed Working Classification

“It is based first on pathologic interpretation and second on typical clinical features.”

Olson EA et al. J Am Acad Dermatol 48:103-10, 2003

# Should classification be based upon inflammatory cell type?

- Lymphocytic
- Neutrophilic
- Mixed

## **Lymphocytic**

Chronic cutaneous lupus erythematosus

Lichen planopilaris (LPP)<sup>4</sup>

Classic LPP

Frontal fibrosing alopecia<sup>5</sup>

Graham-Little syndrome<sup>3</sup>

Classic pseudopelade (Brocq)<sup>6,7</sup>

Central centrifugal cicatricial alopecia<sup>8</sup>

Alopecia mucinosa<sup>3</sup>

Keratosis follicularis spinulosa decalvans<sup>9</sup>

# Lymphocytic 'issue' #1

Alopecic LE does not have any histopathologic resemblance to LPP

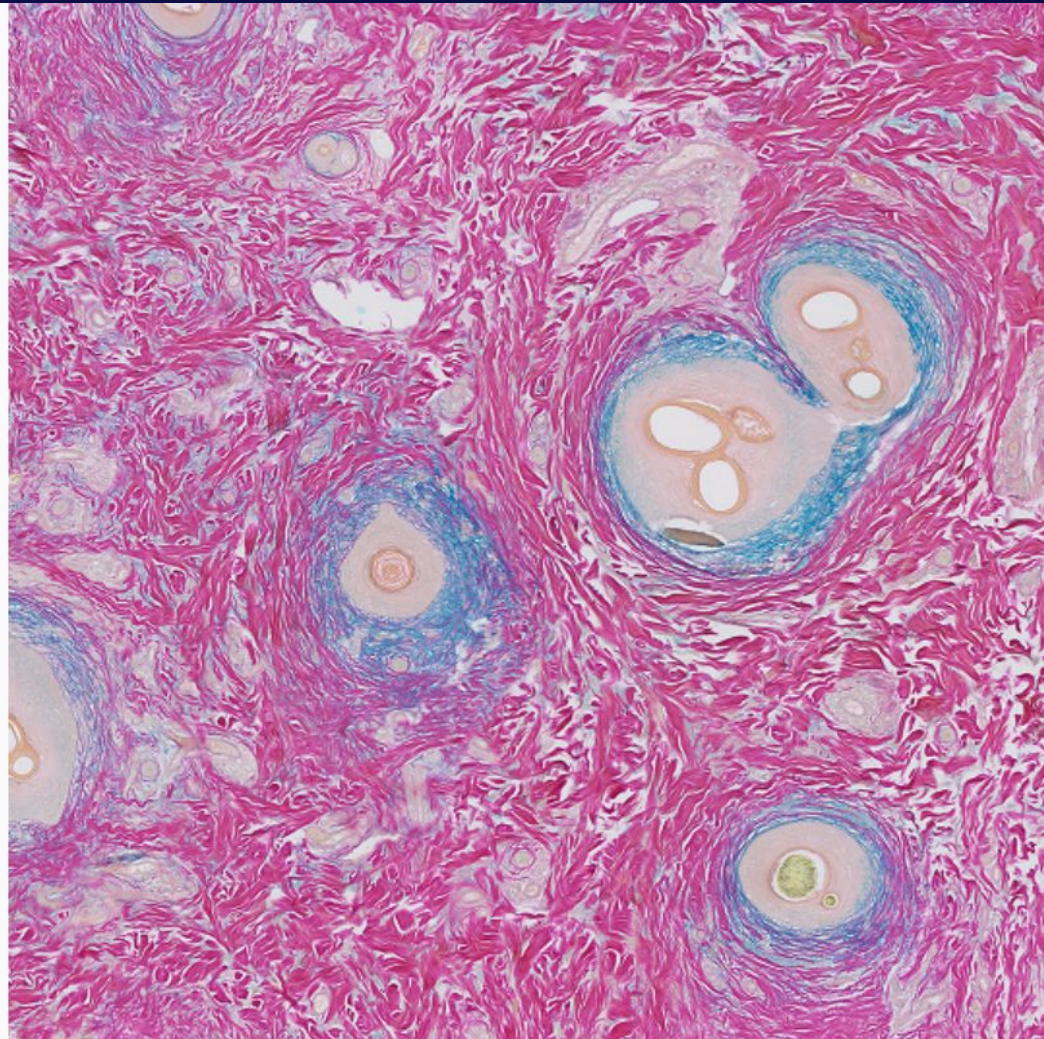
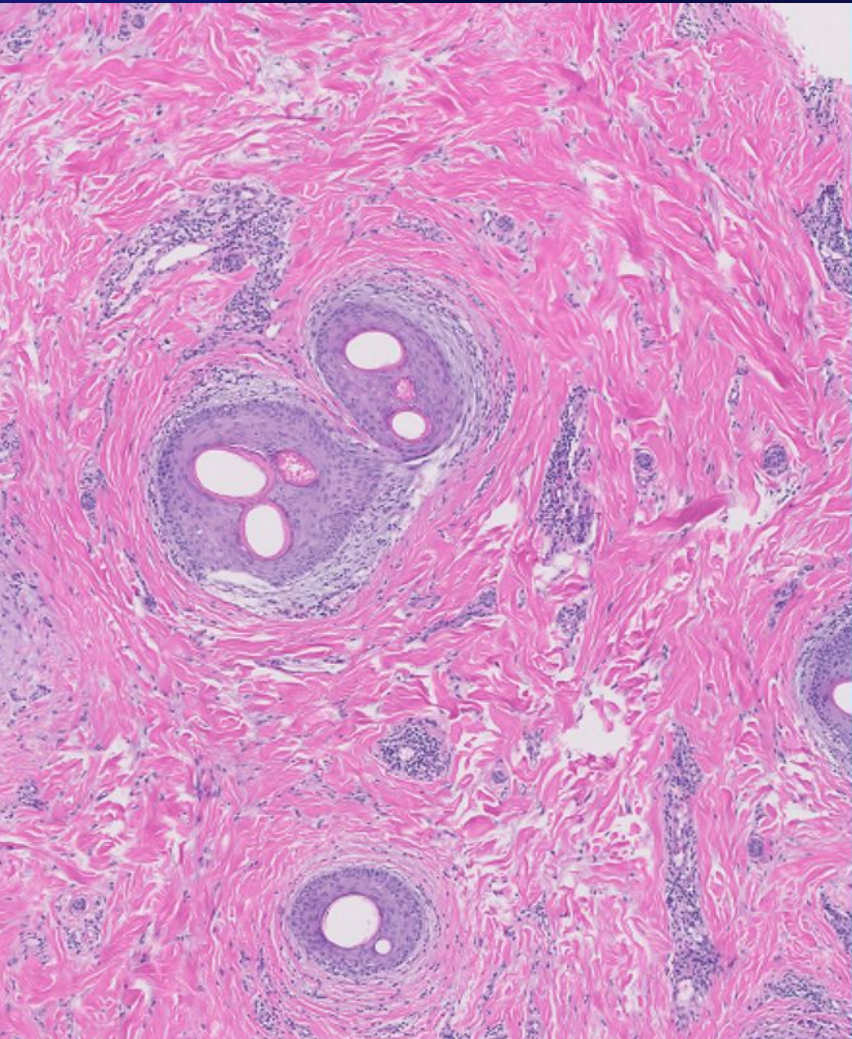
## **Lymphocytic**

Chronic cutaneous lupus erythematosus  
Lichen planopilaris (LPP)<sup>4</sup>



LPP

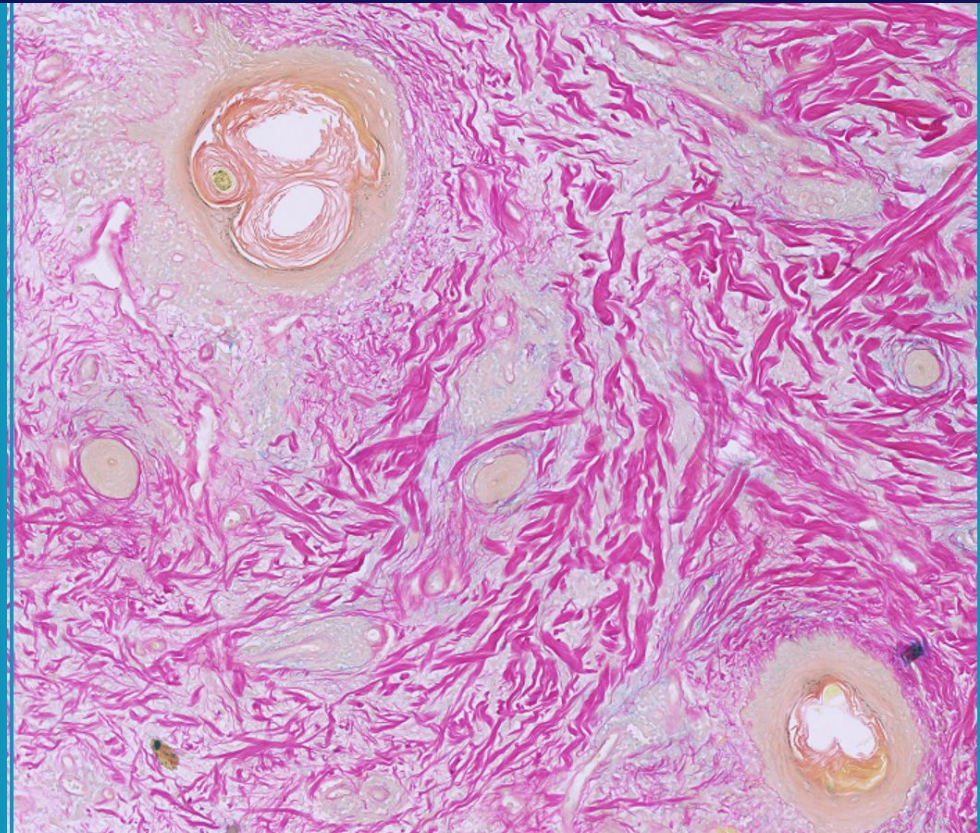
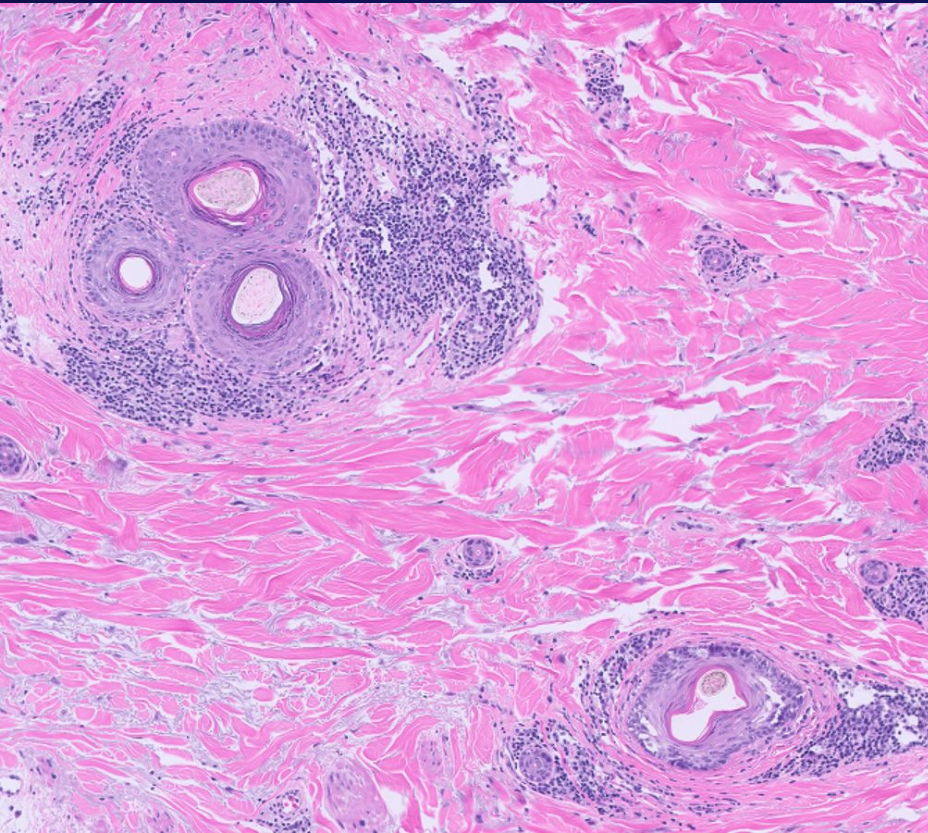
Prominent perifollicular fibroplasia





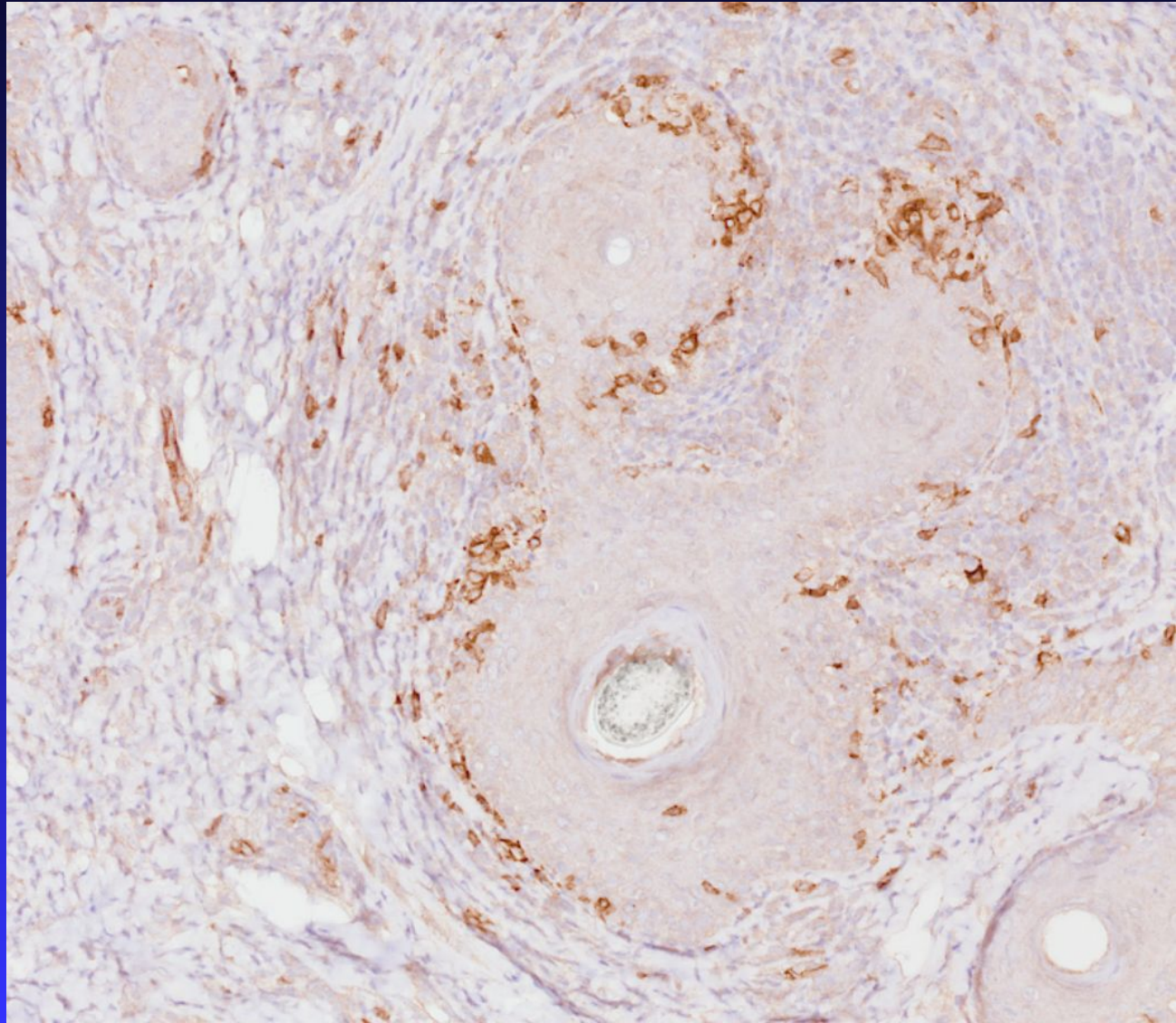
# Alopecic LE

## No perifollicular fibroplasia

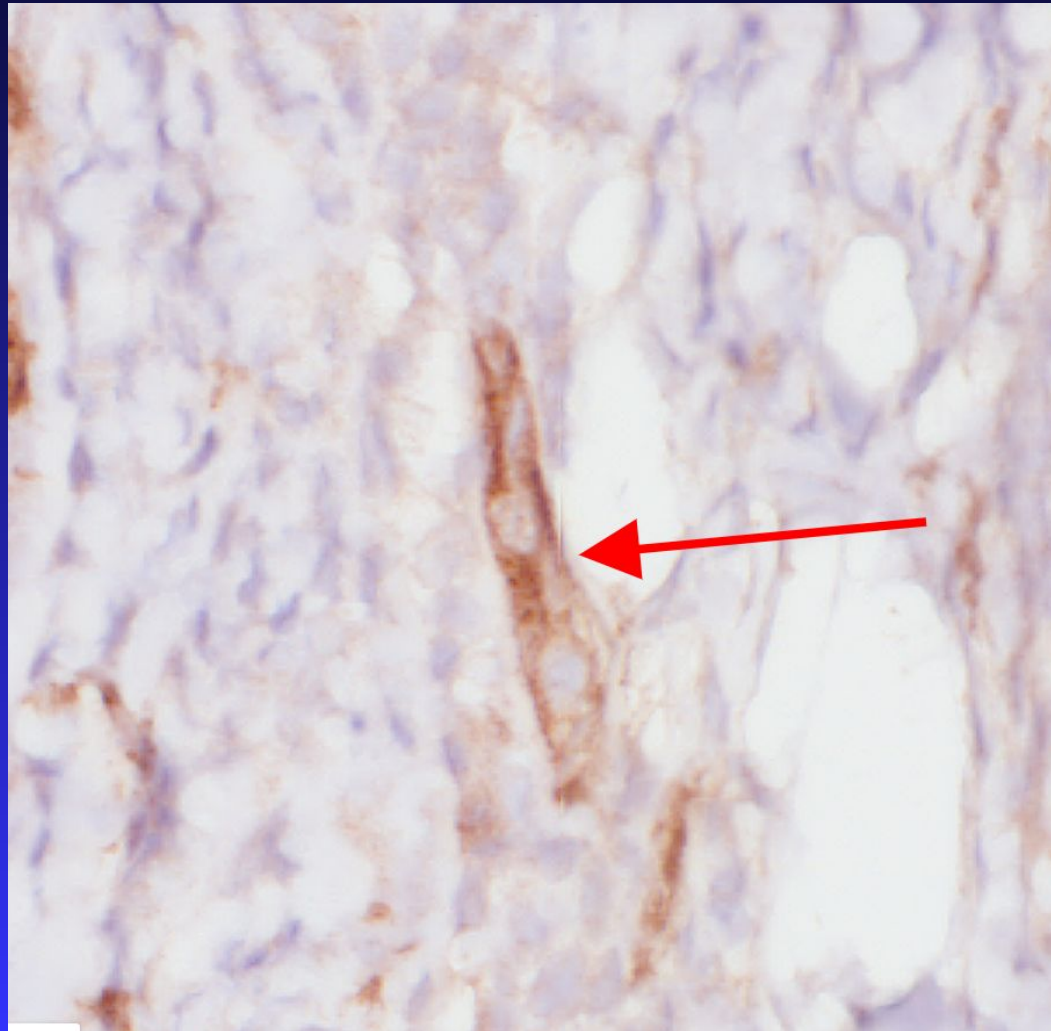




# Alopecic LE—CD123 very useful

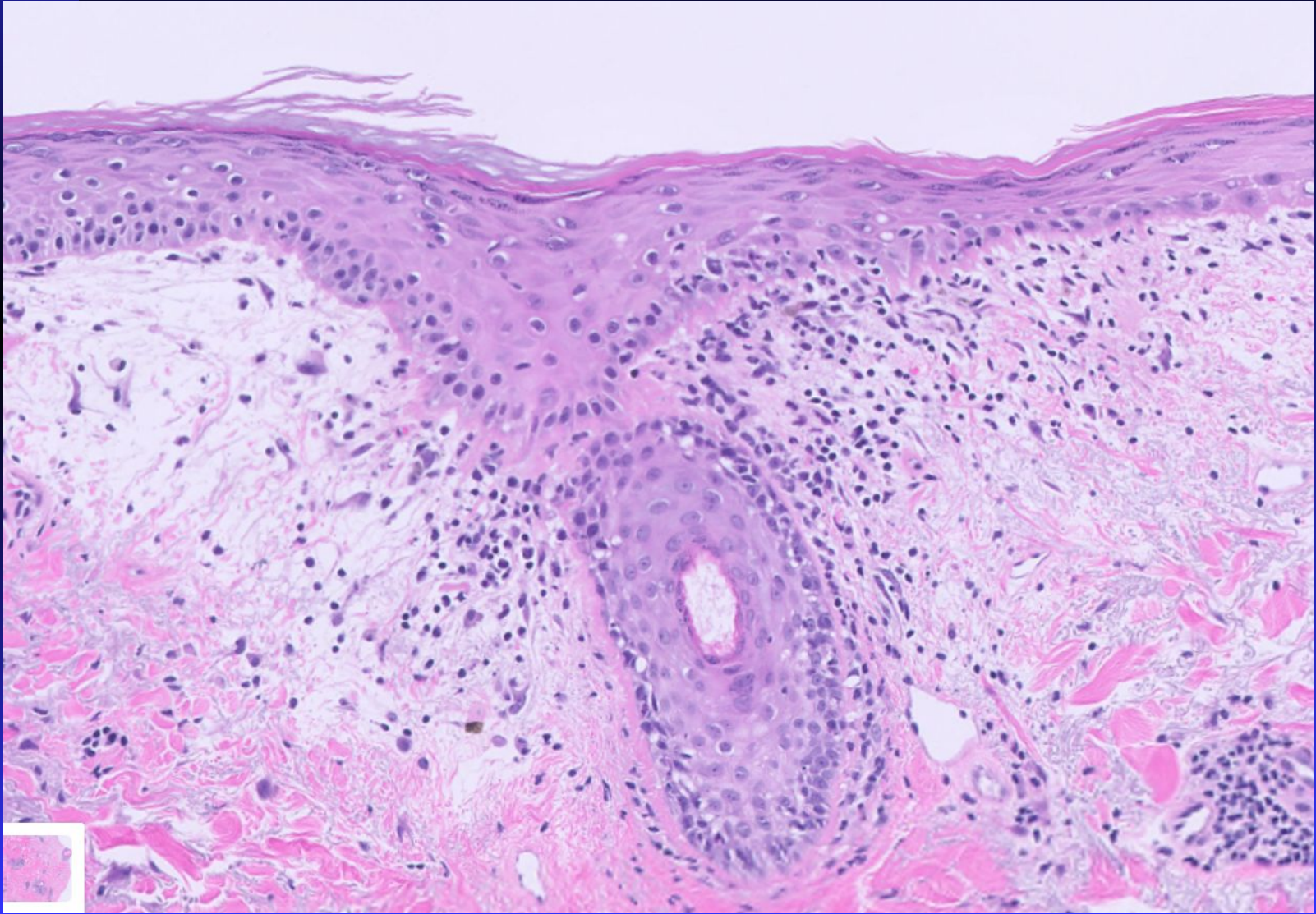


CD123 warning—endothelial+



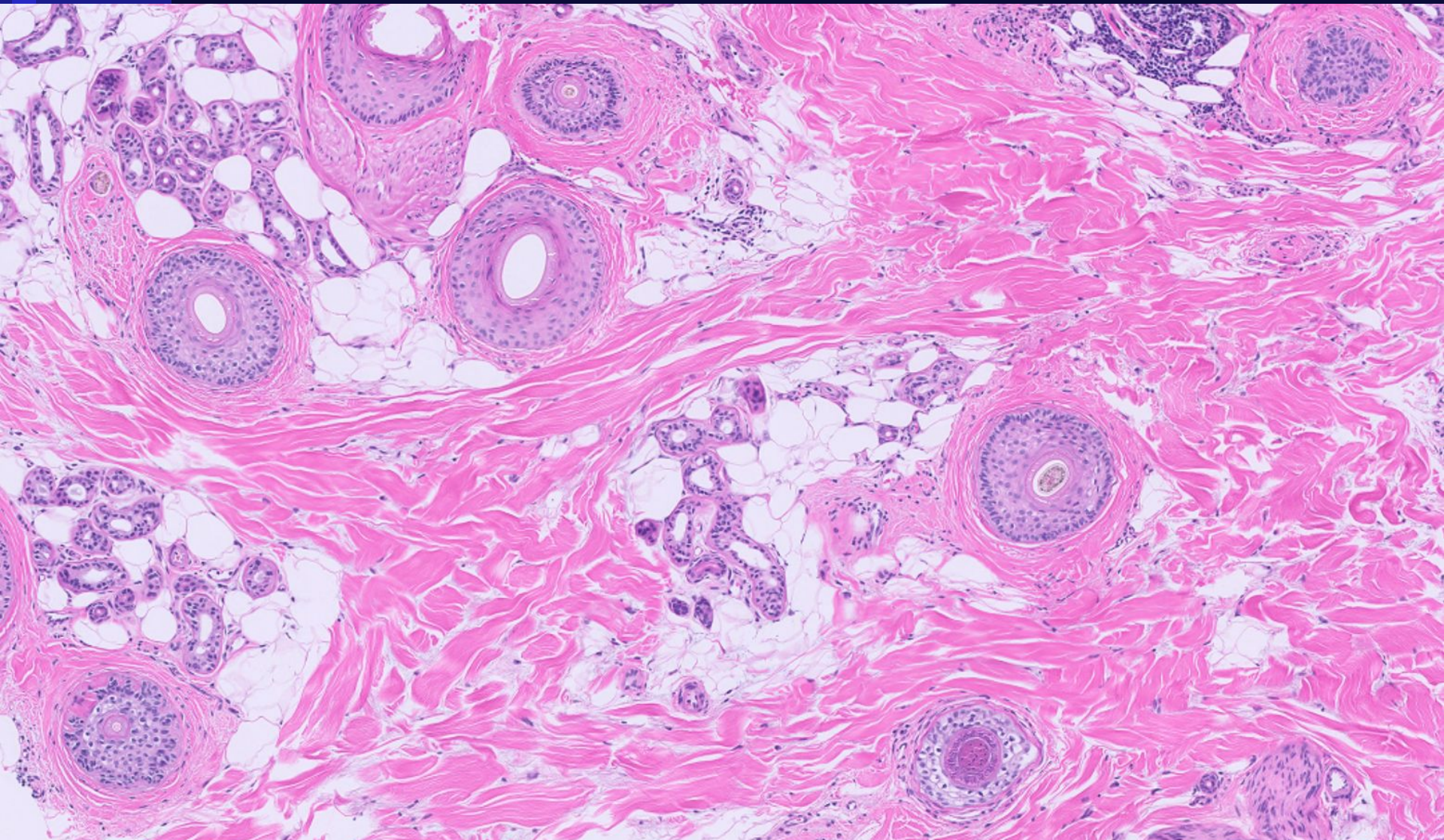


# Alopecia Lupus Erythematosus



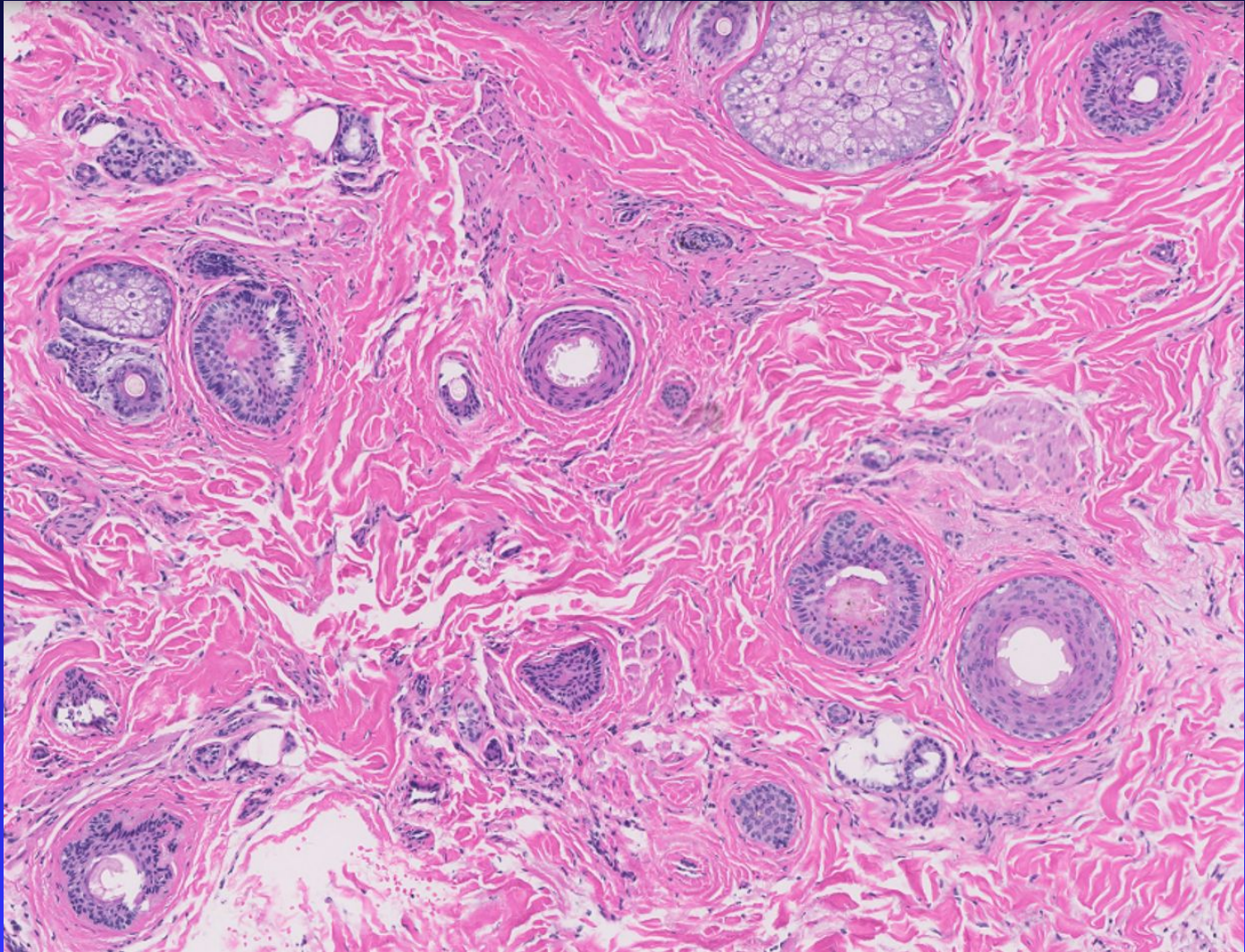


# Alopecic LE



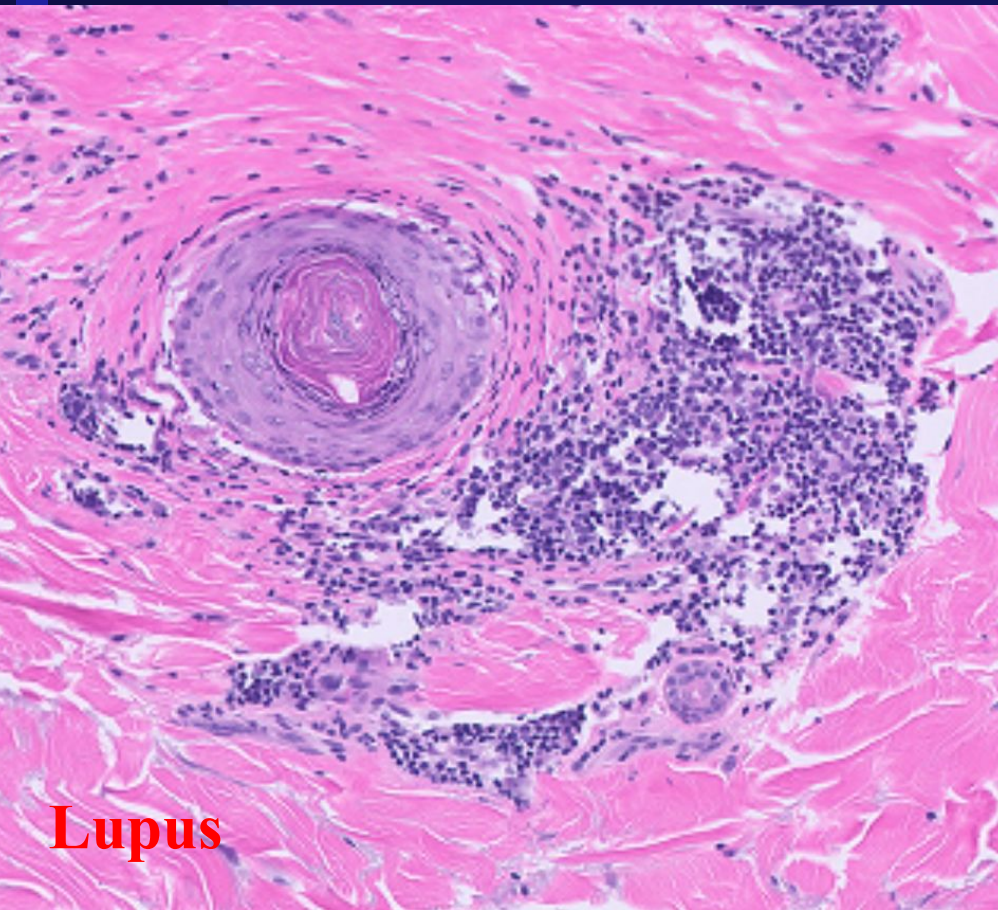


# Alopecic LE resembles subacute AA

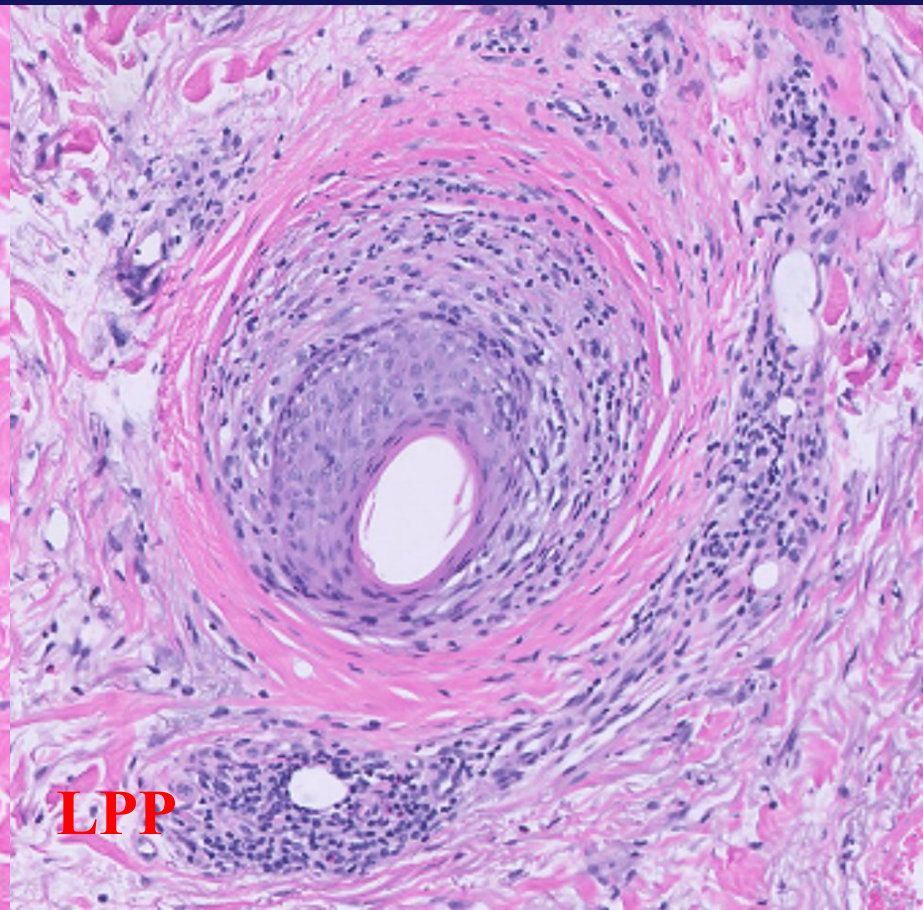




Should such histologically disparate entities be grouped together simply because they have a lymphocytic infiltrate?



**Lupus**



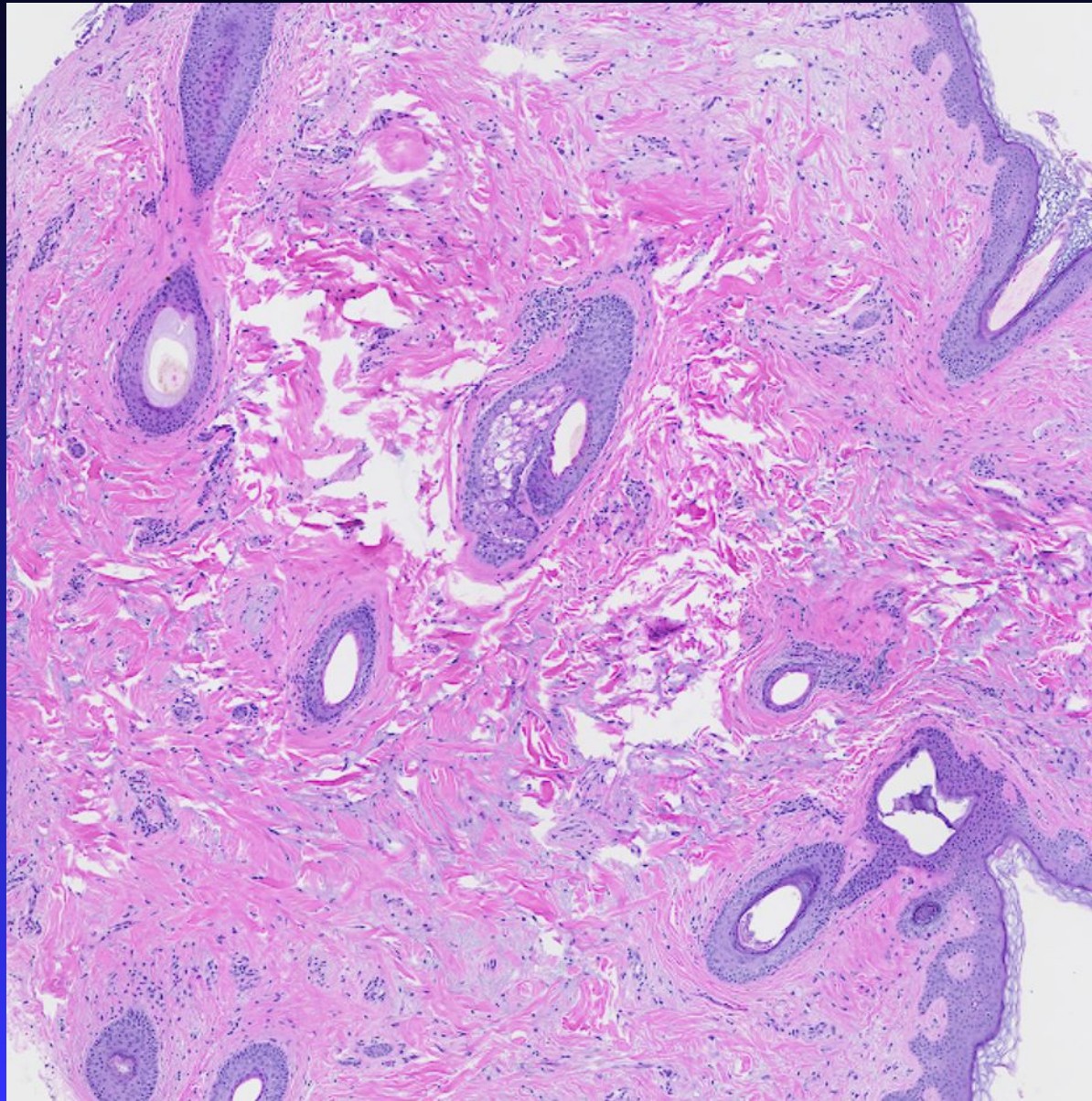
**LPP**



# Lymphocytic 'issue' #2

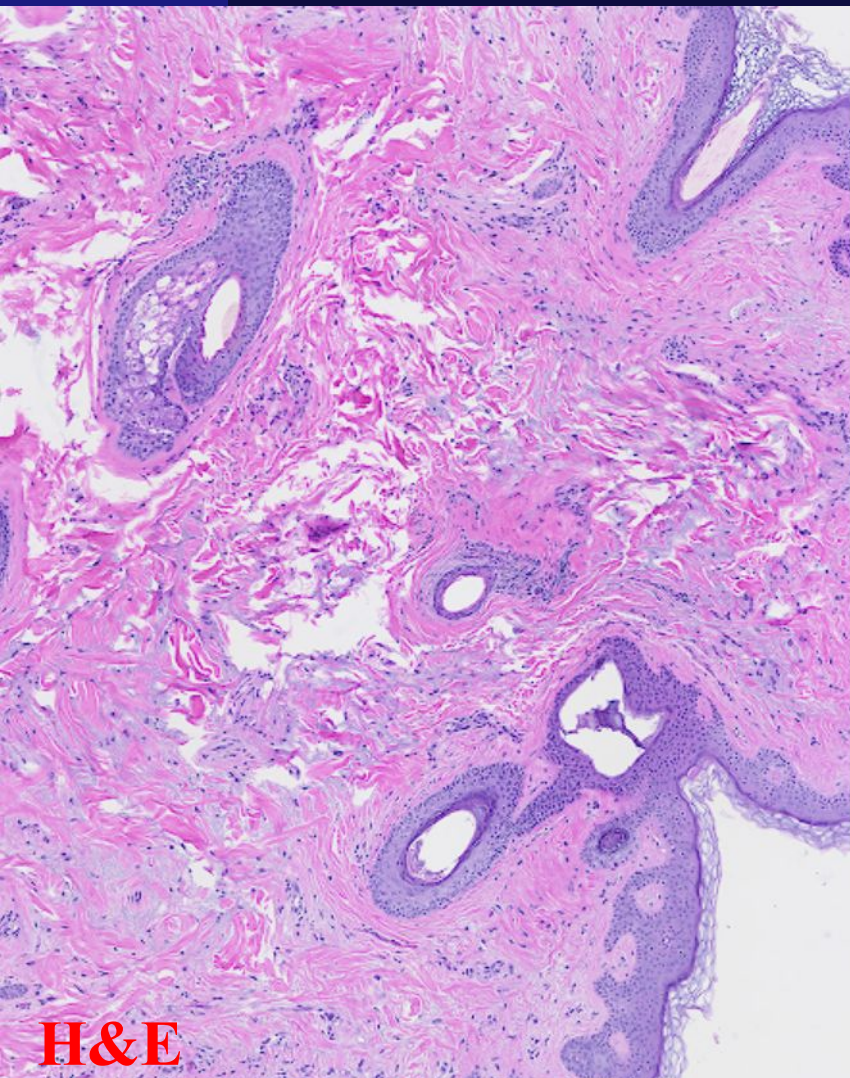
- Frontal fibrosing alopecia often has minimal perifollicular scarring (confusing to dermatopathologists)

# FFA



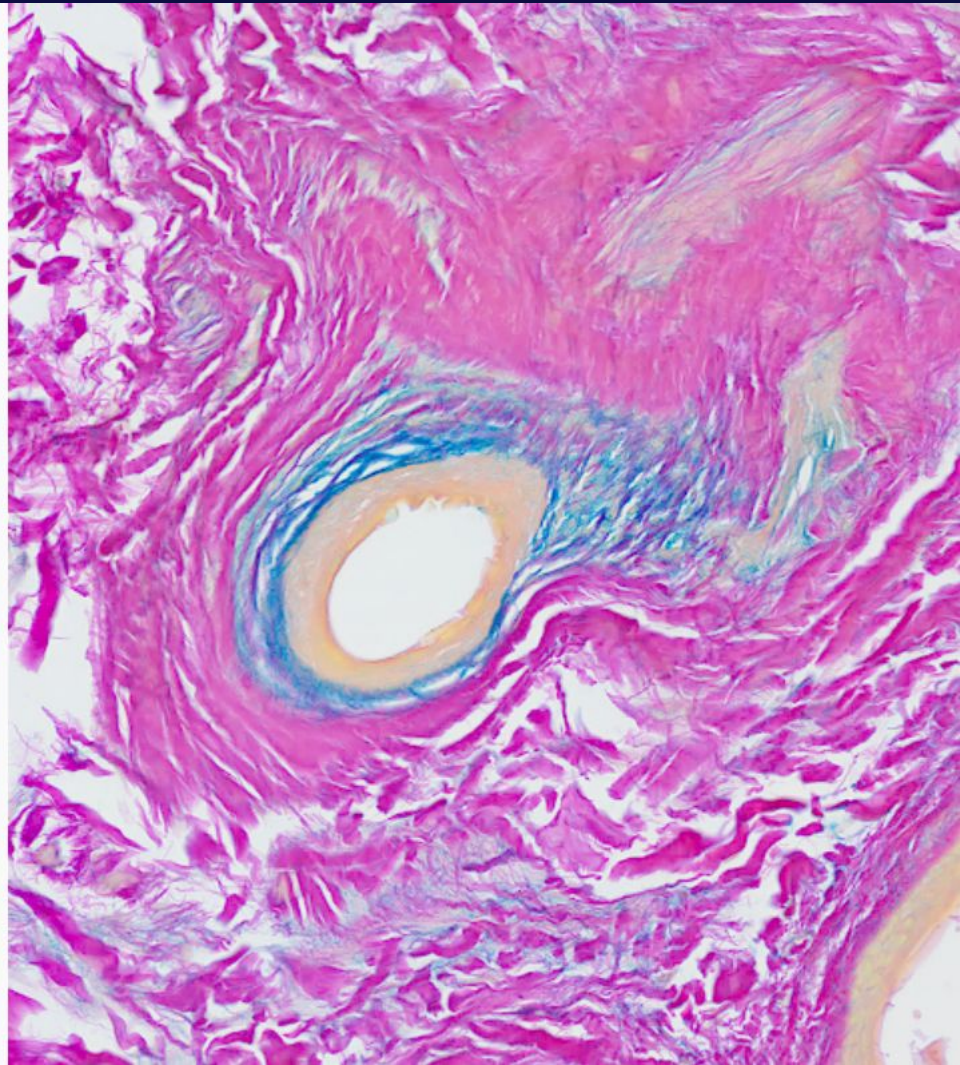
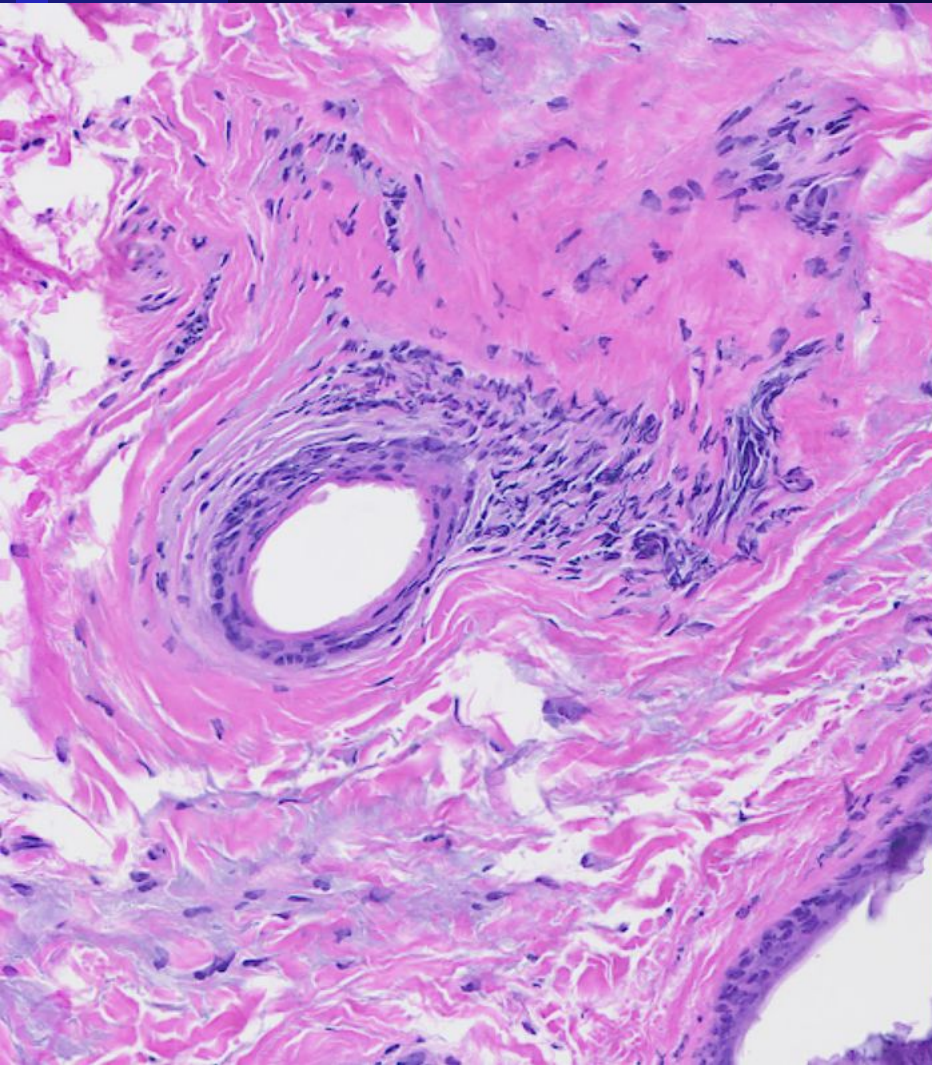


# FFA





# Colloidal Iron of great utility in subtle LPP/FFA

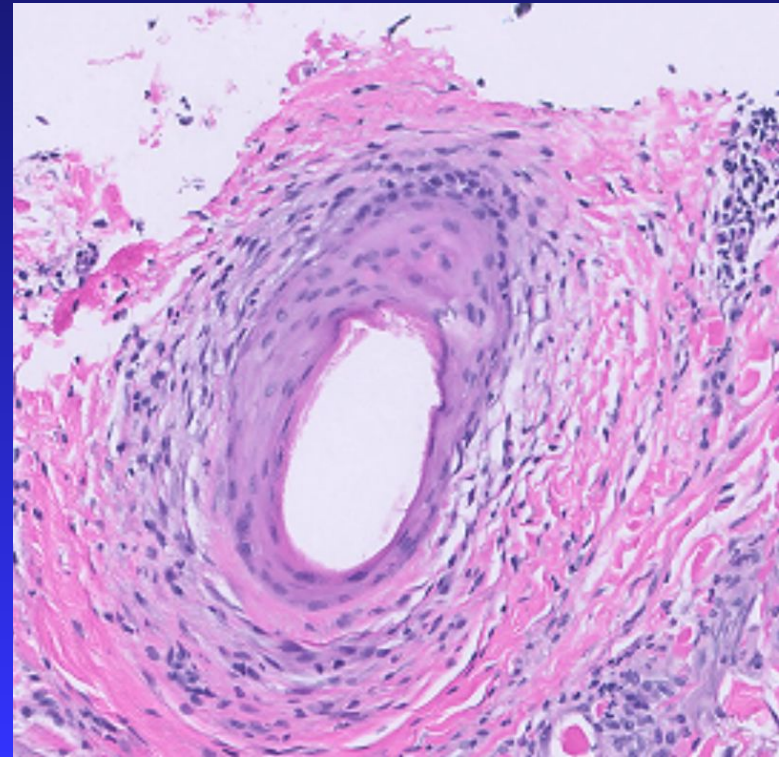


# Lymphocytic 'issues' #2

Are LPP and CCCA are histopathologically identical?

# Lymphocytic 'issues' #2

Premature desquamation of inner root sheath is difficult to understand and appears to be seen in LPP





## Lymphocytic 'issue' #3

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. doi:10.1111/cup.13592

## Lymphocytic 'issue' #3

Flamm A, Moshiri AS, Roche F, et al.  
Characterization of the inflammatory  
features of central centrifugal cicatricial  
alopecia. *J Cutan Pathol*.  
2020;47(6):530-534.  
doi:10.1111/cup.13666

Only studied CCCA cases (no comparison)

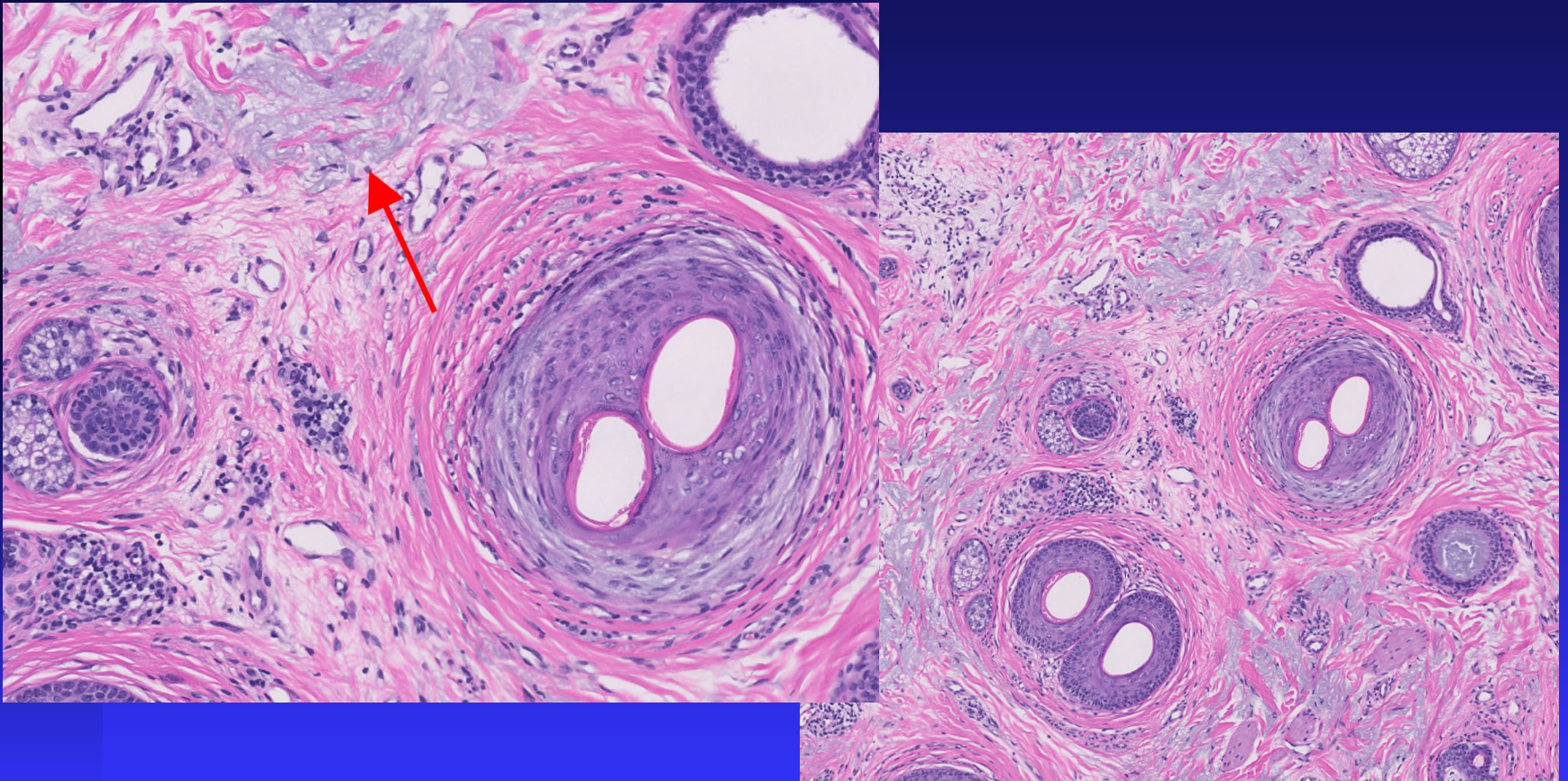


# Lymphocytic 'issue' #4

- Fibrosing Alopecia in a Pattern Distribution (FAPD)
  - ◆ Is this diffuse LPP in patients with FPHL (senescence)?

# Lymphocytic 'issue' #4

- FAPD—LPP with solar elastosis





# Diffuse LPP (FAPD) etiology?



## Lymphocytic (and Neutrophilic) ‘issue’ #5

Folliculitis decalvans often has few to no neutrophils and looks like LPP

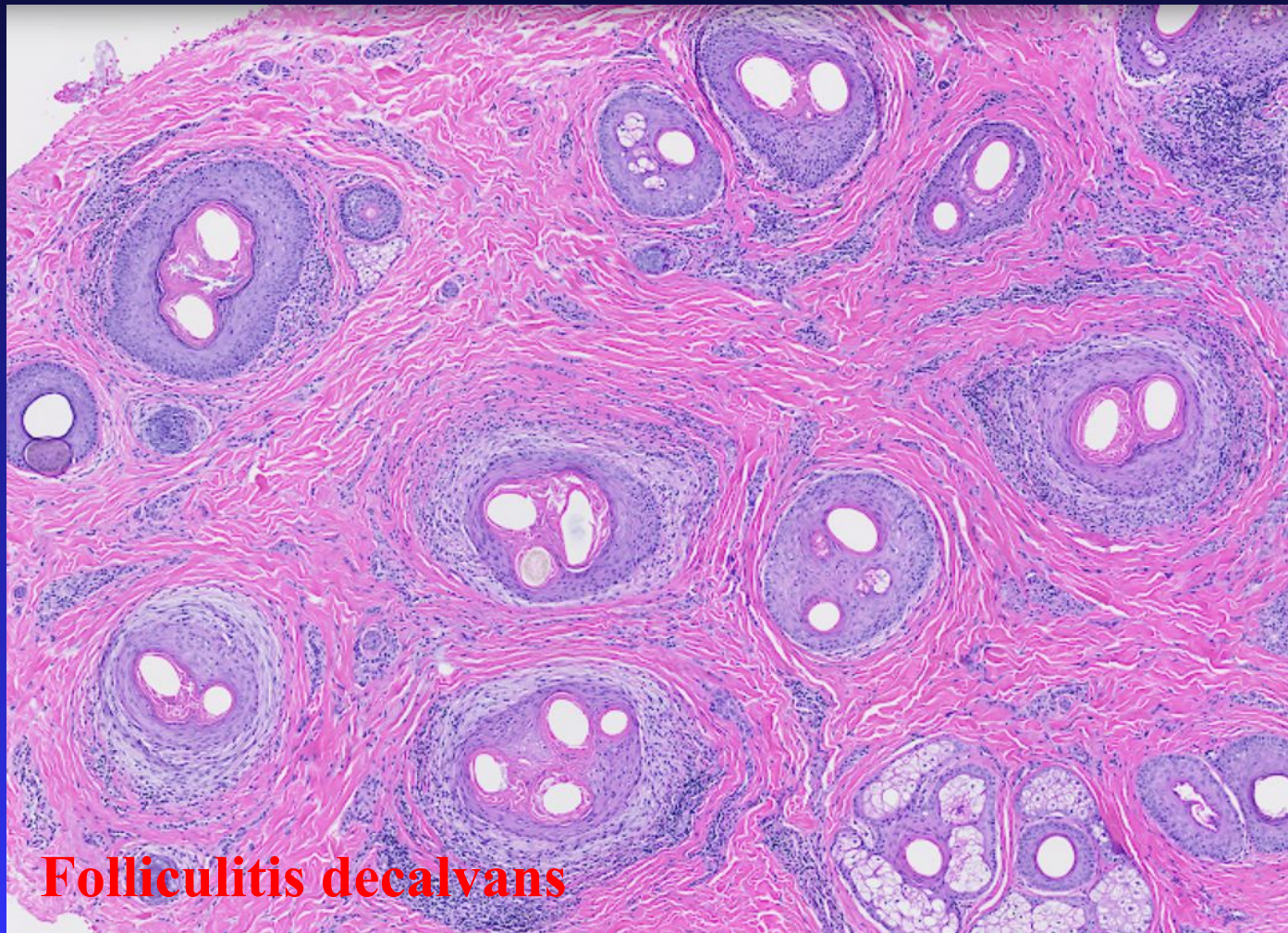
### **Neutrophilic**

Folliculitis decalvans<sup>3</sup>

Dissecting cellulitis/folliculitis<sup>3</sup> (*perifolliculitis  
abscedens et suffodiens*)



## Lymphocytic (and Neutrophilic) 'issue' #5

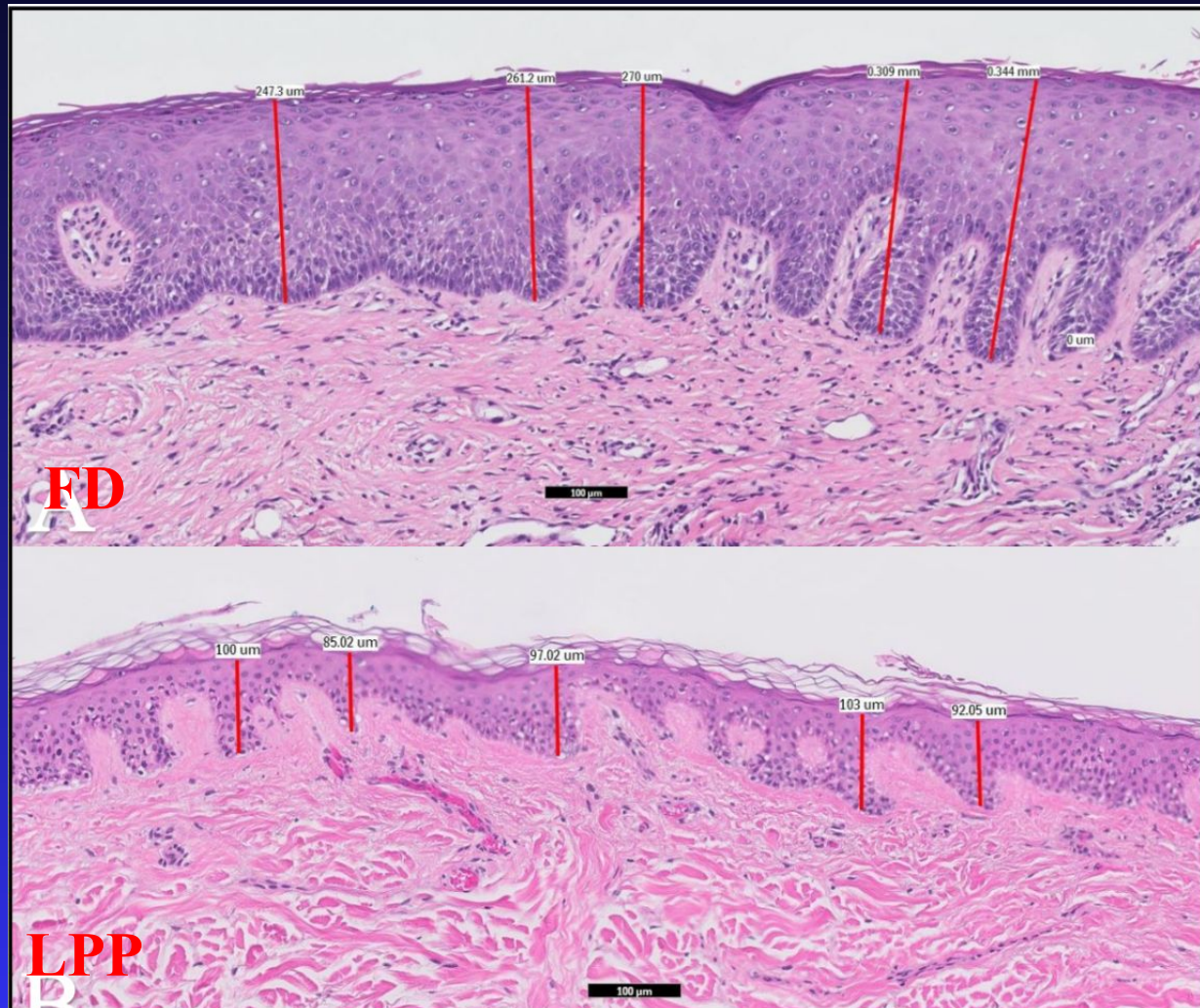


**Folliculitis decalvans**



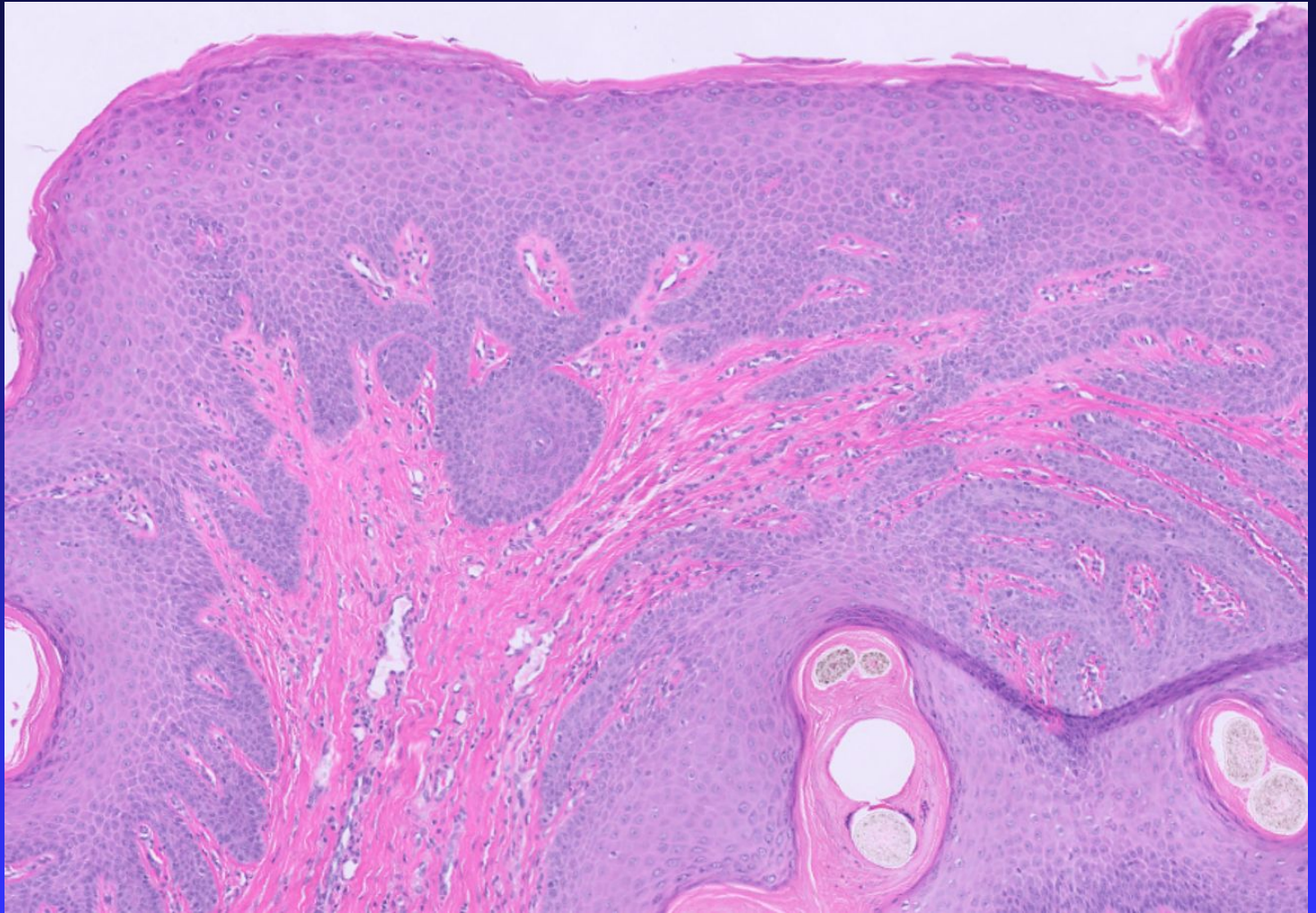
# Epidermal Thickness

## Folliculitis decalvans vs LPP



# Epidermal Thickness

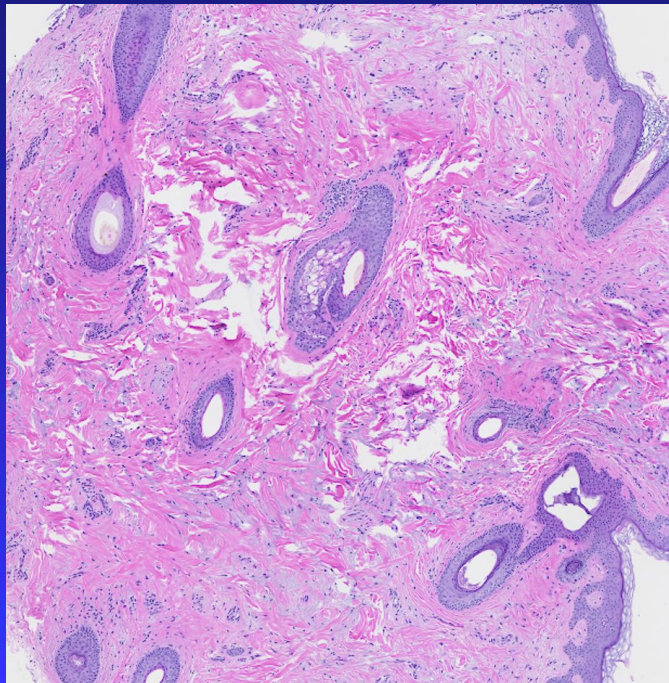
## Folliculitis decalvans





# Lymphocytic 'issue' #6

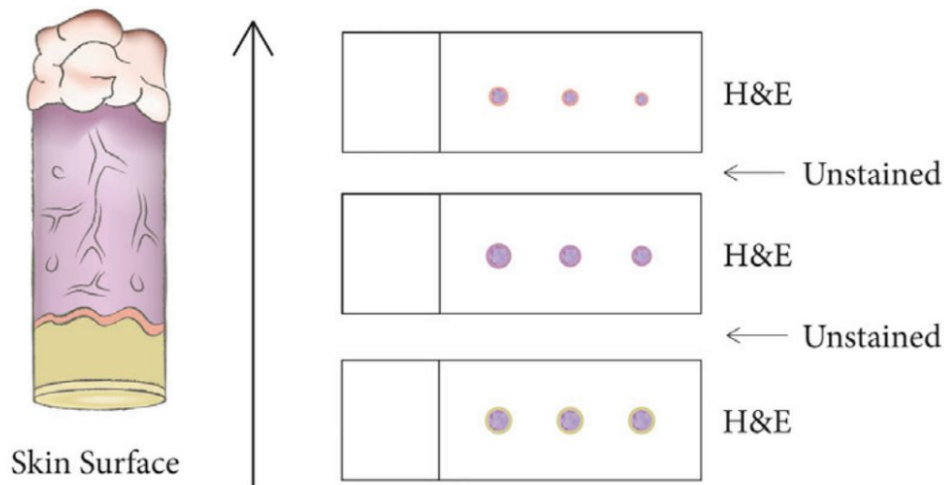
Pseudopelade of 'Brocq' has disappeared as a histopathologic entity—always find inflammation with newer





# A method for more precise sampling of the scalp and eyebrows in frontal fibrosing alopecia

Curtis T. Thompson, MD,<sup>a,b,c</sup> and Antonella Tosti, MD<sup>d</sup>  
*Portland, Oregon, and Miami, Florida*



3 slides total with 9 cross  
sections; 3 sections per slide

1. Tissue is embedded epidermis-down
2. Step through entire block on initial H&E stains
3. Obtain unstained slides

# Lymphocytic 'issue' #7

Alopecia mucinosa (follicular mucinosis)—Should a lymphoproliferative disorder be considered a cause of primary cicatricial alopecia?

## **Proposed working classification of primary cicatricial alopecia**

### **Lymphocytic**

Chronic cutaneous lupus erythematosus

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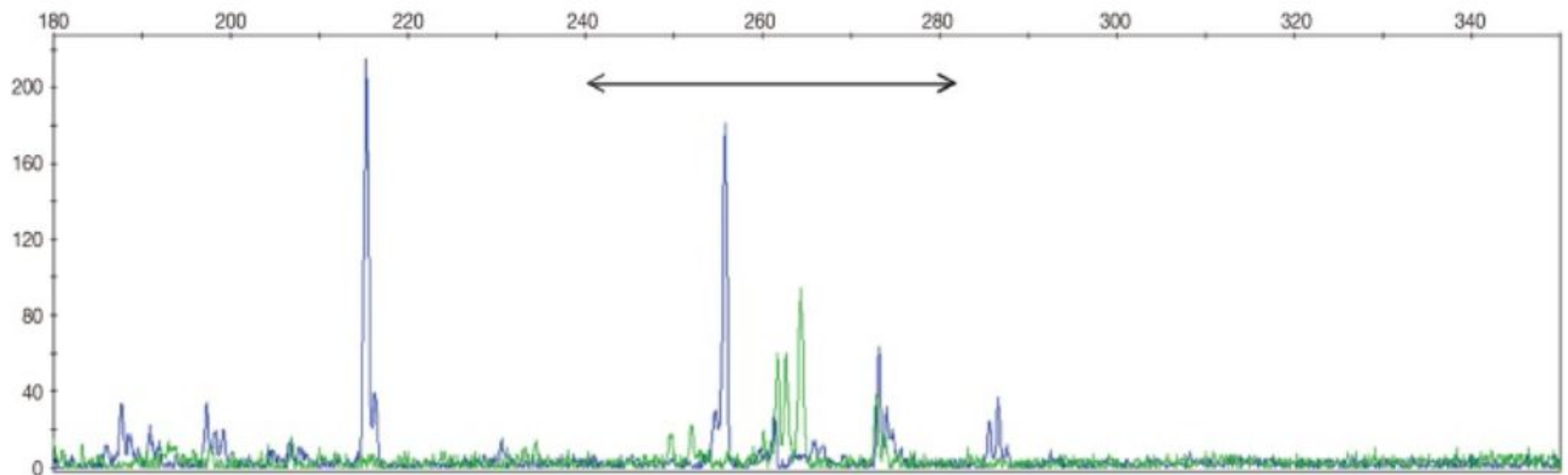
Central centrifugal cicatricial alopecia<sup>8</sup>

Alopecia mucinosa<sup>3</sup>

Keratosis follicularis spinulosa decalvans<sup>9</sup>

# Lymphocytic ‘issue’ #7

Alopecia mucinosa (follicular mucinosis)—Should a lymphoproliferative disorder be considered a cause of primary cicatricial alopecia?



# Lymphocytic 'issue' #8

Should cicatricial hair loss from congenital/genetic conditions be considered a primary cicatricial alopecia?

## **Proposed working classification of primary cicatricial alopecia**

### **Lymphocytic**

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# Lymphocytic 'issue' #8

- Keratosis follicularis spinulosa decalvans
  - ◆ Autosomal recessive mutation  
(desmoglein DSG4 gene)
- Many more conditions would have to be added.

# Mixed 'issue' #9

- Folliculitis (acne) keloidalis
  - ◆ No histologic keloid—only fibrosis
  - ◆ Confusing to pathologist

# Mixed 'issue' #10

- Folliculitis (acne) necrotica
  - ◆ Suppurative folliculitis with gram positive bacteria
    - ◆ Does this cause a primary cicatricial alopecia?
    - ◆ Is this just a part of the follicular occlusion triad?

## **Mixed**

Folliculitis (acne) keloidalis<sup>9</sup>

Folliculitis (acne) necrotica<sup>3</sup>

Erosive pustular dermatosis<sup>10</sup>



# Mixed 'issue' #11

- Erosive pustular dermatosis
  - ◆ Almost entirely elderly scalp with end-stage male pattern hair loss/androgenetic alopecia
    - ◆ Severe solar elastosis, ulceration and actinic keratoses
    - ◆ Collagen gone and failure to maintain epithelialization.
    - ◆ Does not appear to be a primary follicular process

# **Proposed working classification of primary cicatricial alopecia**

## **Lymphocytic**

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## **Mixed**

Folliculitis (acne) keloidalis<sup>9</sup>

Folliculitis (acne) necrotica<sup>3</sup>

Erosive pustular dermatosis<sup>10</sup>

## **Nonspecific**

# 2024 Proposed Classification of Primary Cicatricial Alopecia

- Cell type?
- Pathogenetic source?
  - ◆ Autoimmune (include pemphigus v.)
  - ◆ Exogenous chemicals (poorly understood)
  - ◆ Congenital/genetic (many types)
  - ◆ Neoplastic
  - ◆ Aging—not primary cicatricial process
- Histopathologic similarity?



# Histopathologic similarity?

- Perifollicular fibrosis
  - ◆ LPP, FFA, CCCA, FD
- Catagen/telogen shift
  - ◆ LE
- Deep dermal infiltrate
  - ◆ LE, Dissecting folliculitis
- Interfollicular interface
  - ◆ LE

¡Muchas gracias!



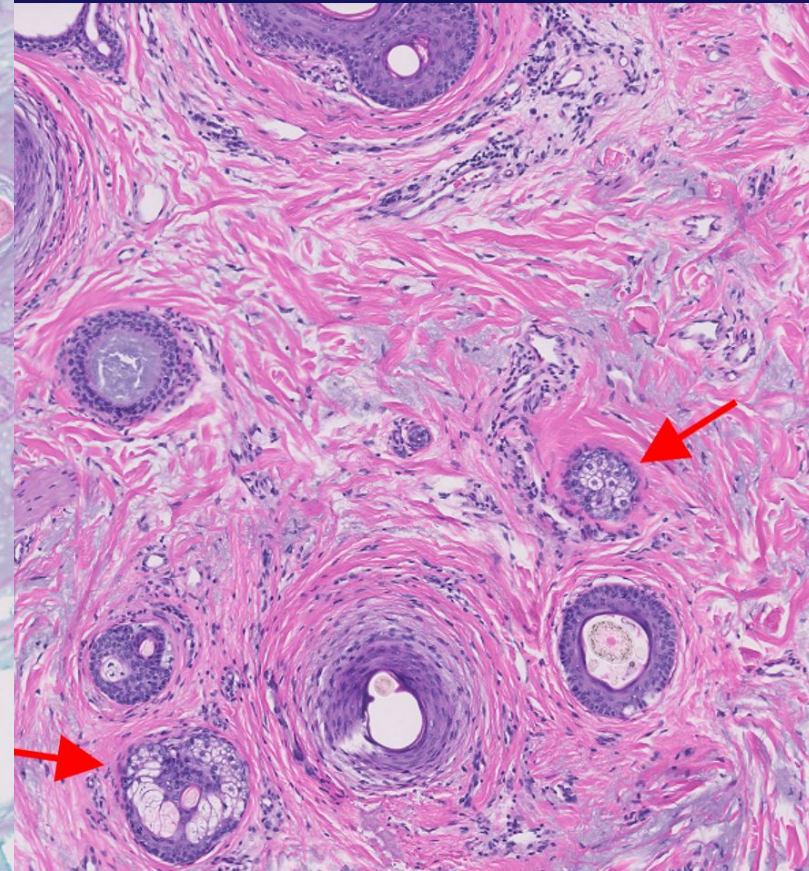
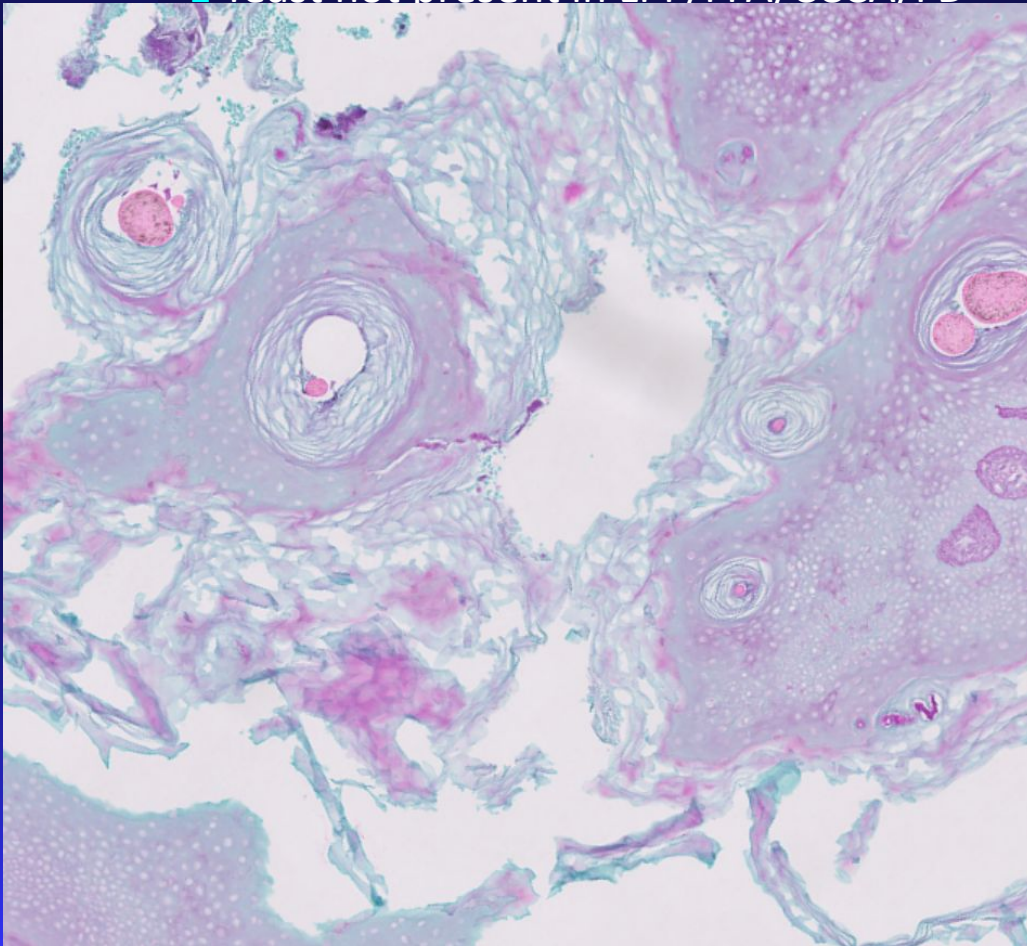
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# Clue

- Yeast not present in LPP/FFA/CCCA/FD





Absence of yeast  
Absence of sebaceous glands

