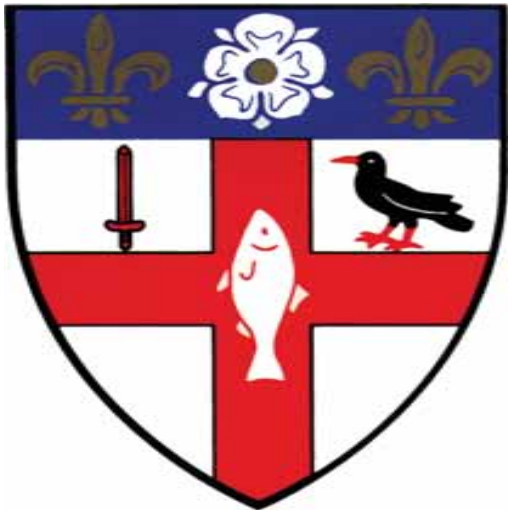
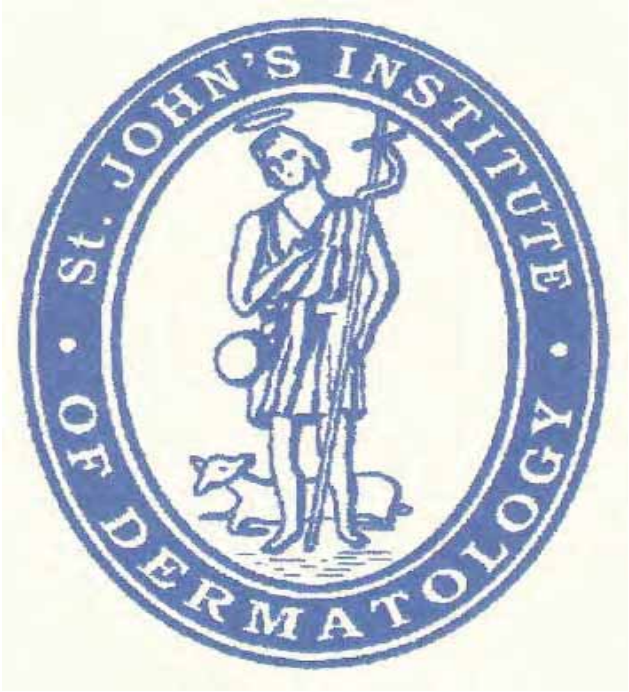




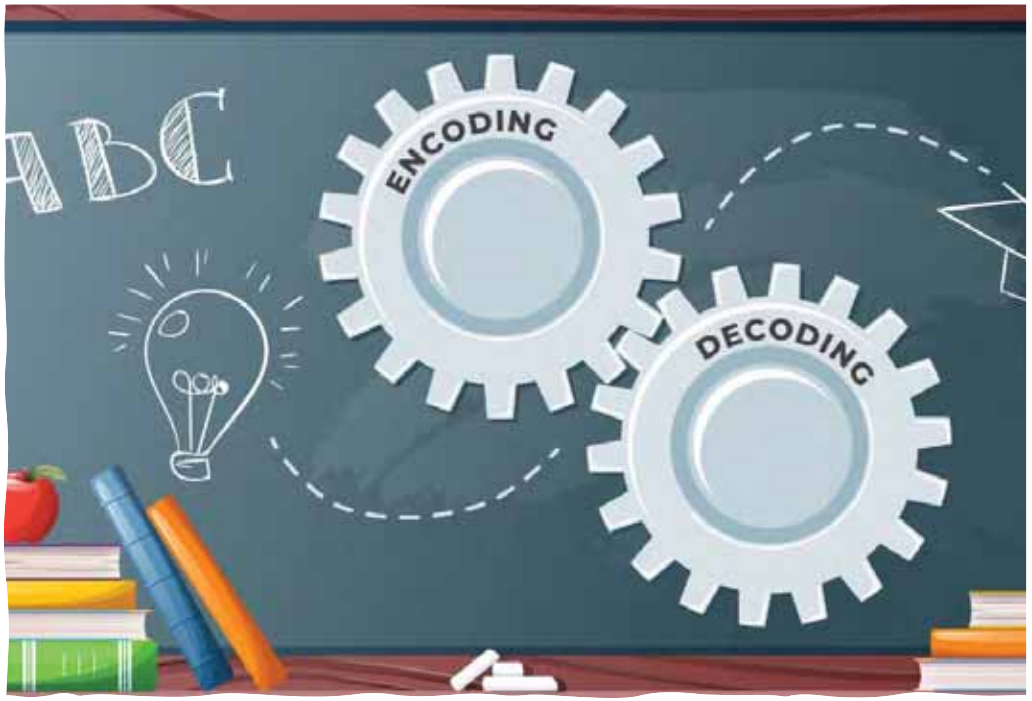
'Decoding' Scarring Alopecia

Catherine M. Stefanato, MD, FRCPath

Consultant Dermatopathologist

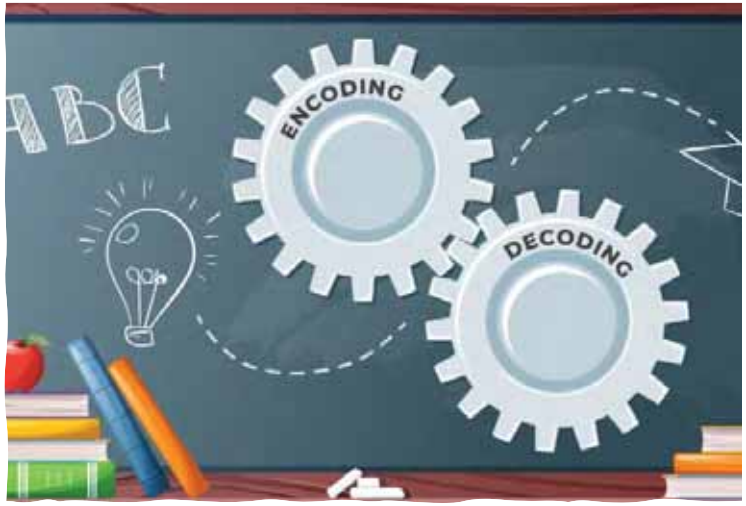
Honorary Senior Lecturer, King's College London

London, UK



‘DECODING’

‘Producing language”



‘Understanding it’

At first glance, these two skills—encoding and decoding—might seem to exist in separate silos. One could think of them as opposite ends of the literacy spectrum: one focused on producing language and the other on understanding it. However, a closer look reveals that these two skills are not just linked but deeply symbiotic. The proficiency in one often informs and enhances the proficiency in the other, creating a continuous loop of literacy development.

PRIMARY SCARRING ALOPECIA

Group of disorders characterized by a common final pathway of replacement of the follicular structures by fibrous tissue

LYMPHOCYTIC
DLE, LPP Brocq, CCCA

NEUTROPHILIC
Folliculitis decalvans
Dissecting folliculitis

TRAUMA
Chronic traction

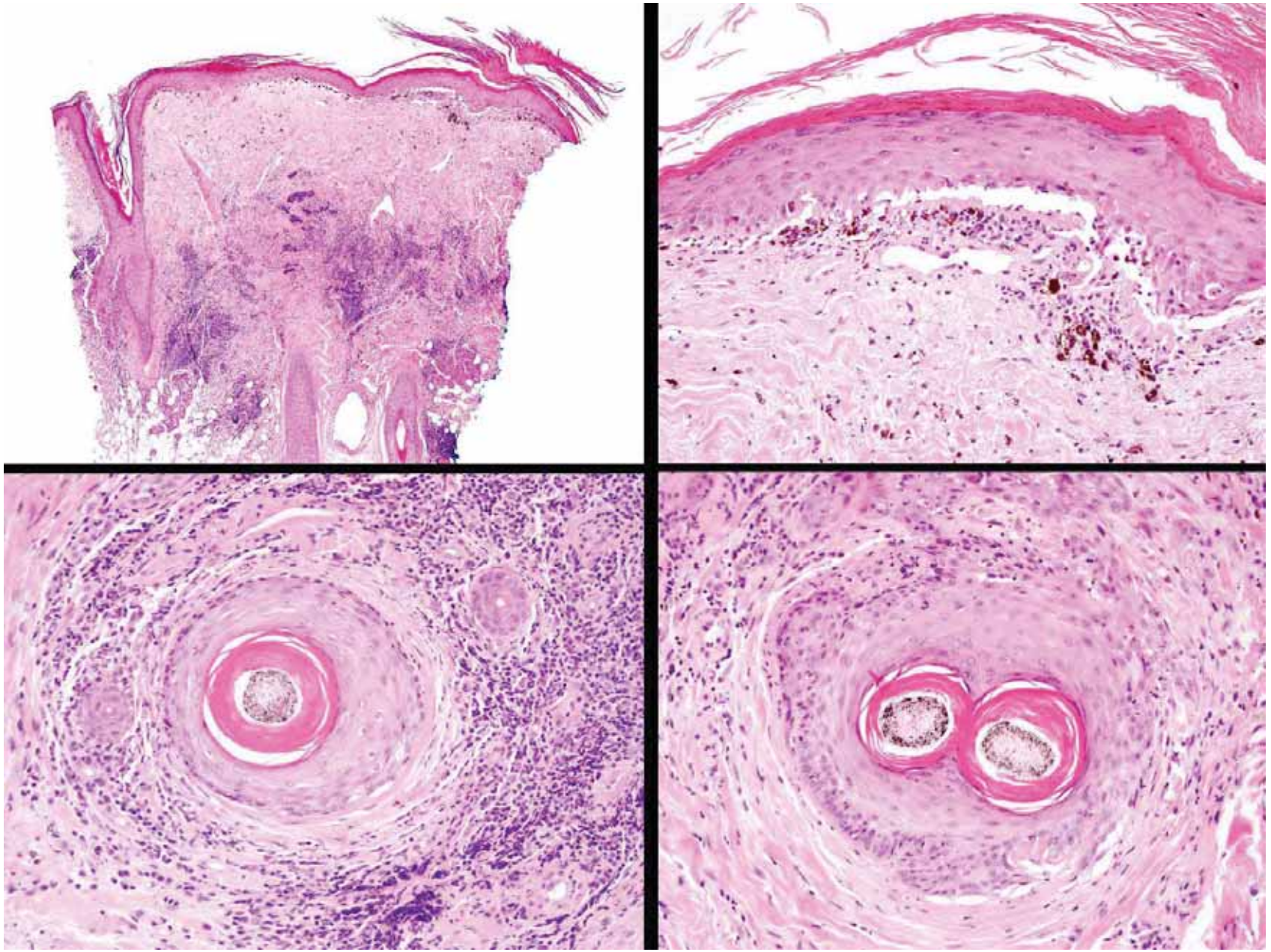
MIXED
Acne keloidalis
nuchae

Table I. Proposed NAHRS working classification of primary cicatricial alopecia*

Lymphocytic
Keratosis follicularis spinulosa decalvans
Chronic CLE
LPP
Classic LPP
Frontal fibrosing alopecia
Graham-Little syndrome
Alopecia mucinosa
Classic pseudopelade (Brocq)
Central centrifugal cicatricial alopecia
Neutrophilic
Folliculitis decalvans
Dissecting cellulitis/folliculitis
Mixed
Folliculitis (acne) keloidalis
Folliculitis (acne) necrotica
Erosive pustular dermatosis

CLE, Cutaneous lupus erythematosus; *LPP*, lichen planopilaris; *NAHRS*, North American Hair Research Society.
*From Olsen et al.[5]

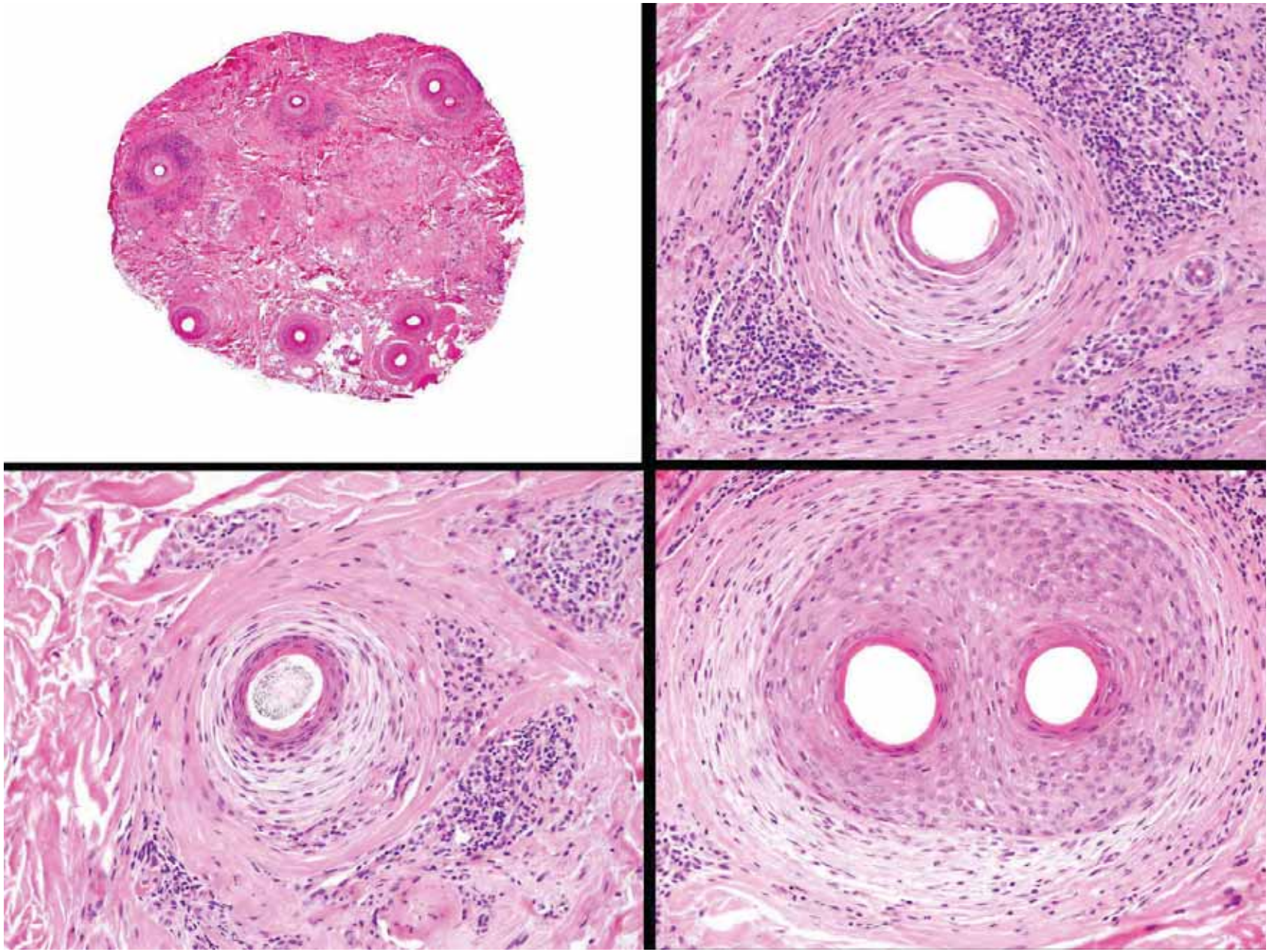
‘CLASSIC’ DISCOID LUPUS ERYTHEMATOSUS

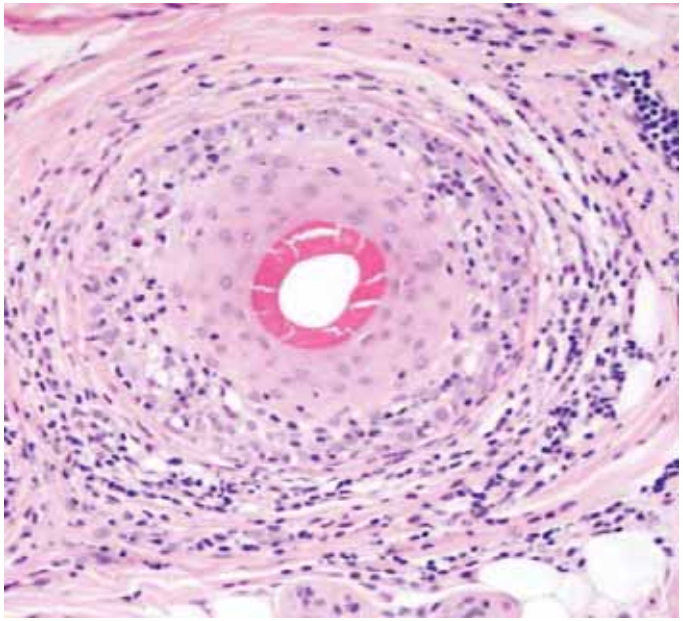


Histopathologic Variants of Alopecia in Lupus

- Discoid lupus: Scarring Alopecia
- Systemic lupus: Non-scarring alopecia: AA-like: Peribulbar lymphocytes, plasma cells, mucin
- Systemic lupus: Non-scarring alopecia: telogen effluvium
- Mixed connective tissue disease: Mixed scarring & non-scarring alopecia

‘CLASSIC’ LICHEN PLANOPILARIS





Terminal

- Patchy: Classic LPP

Terminal & vellus

- Patterned: Frontal fibrosing alopecia

Vellus

- Facial papules (with or w/o FFA)

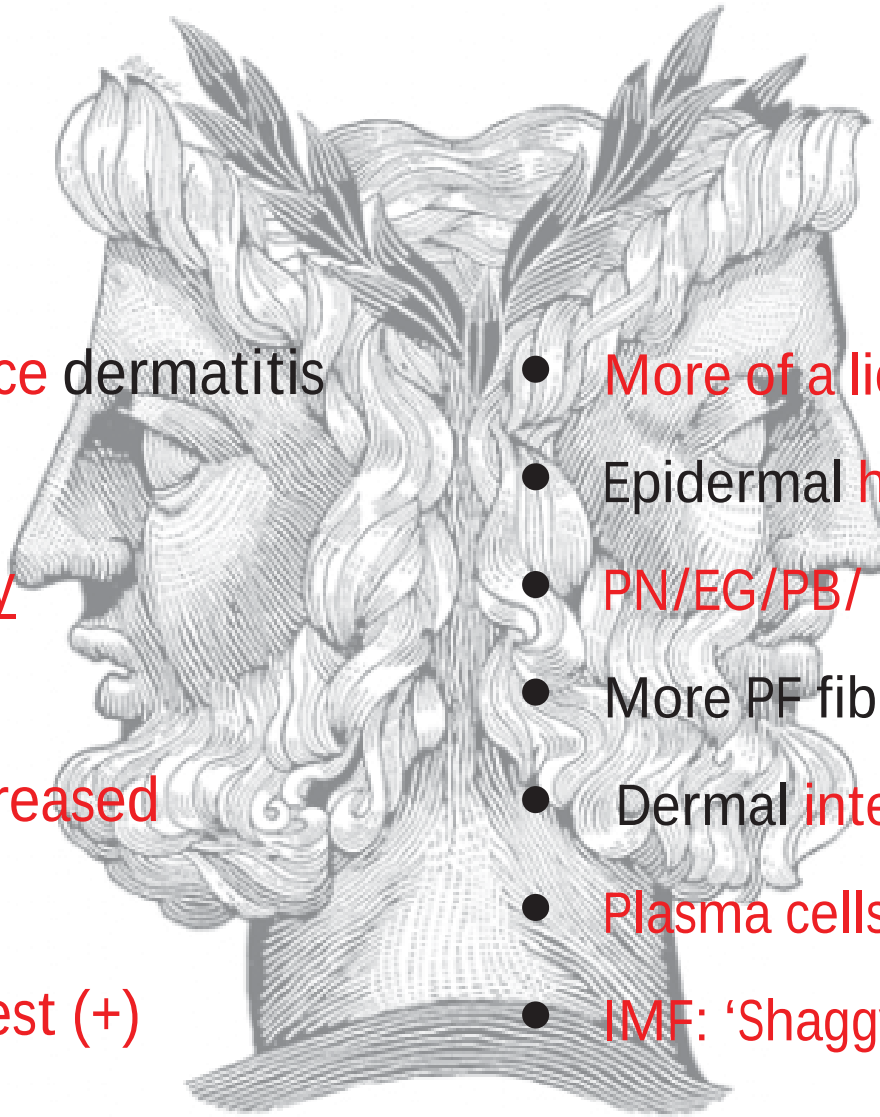
One histopathologic pattern, multiple clinical presentations

LUPUS AND LICHEN PLANUS SIMILARITIES

- Lymphocytic scarring alopecias
- Perifollicular lymphoid cell infiltrate
- Loss of sebaceous glands
- Perifollicular fibrosis
- Tufting

LUPUS

- More of an interface dermatitis
- Epidermal atrophy
- PV/PN/EG/PB likely
- Less PF fibrosis
- Dermal mucin: increased
- Plasma cells (+++)
- IMF: Lupus band test (+)

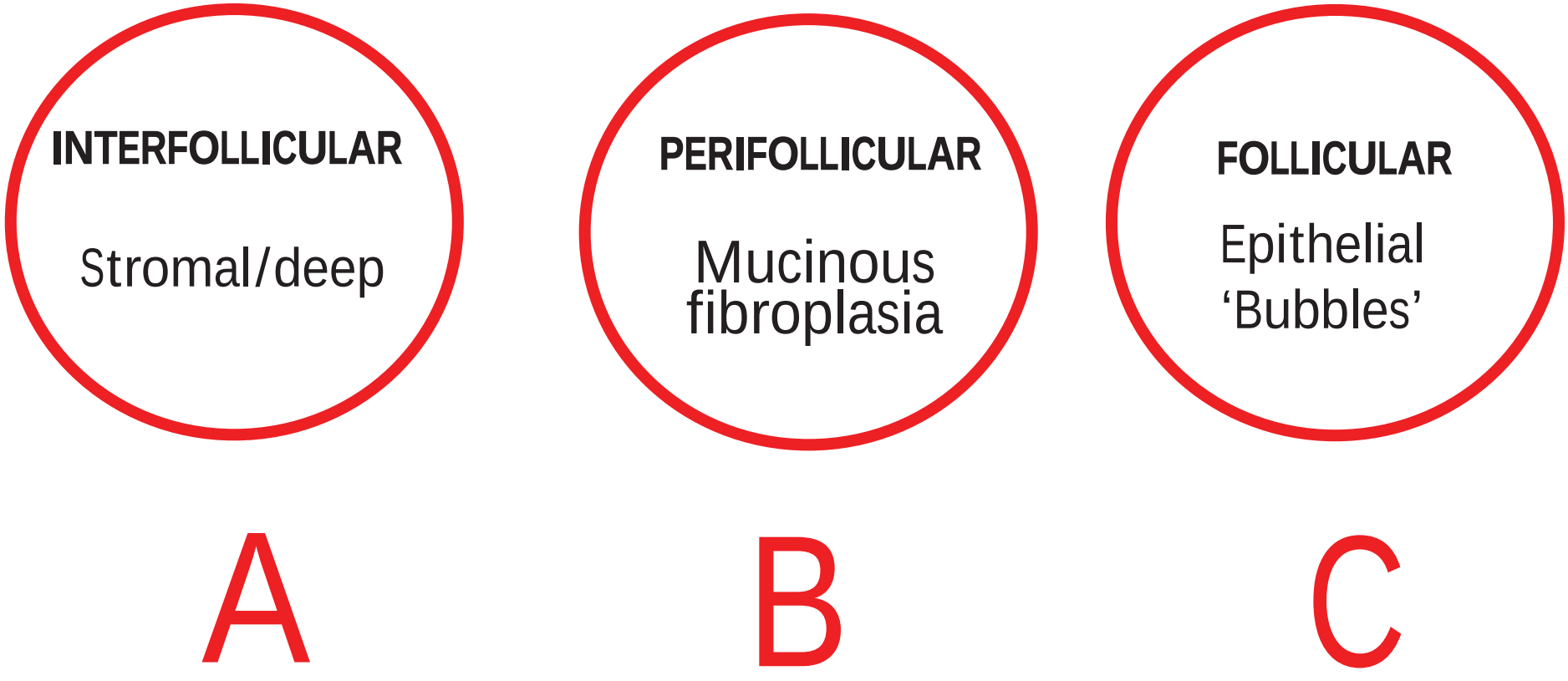


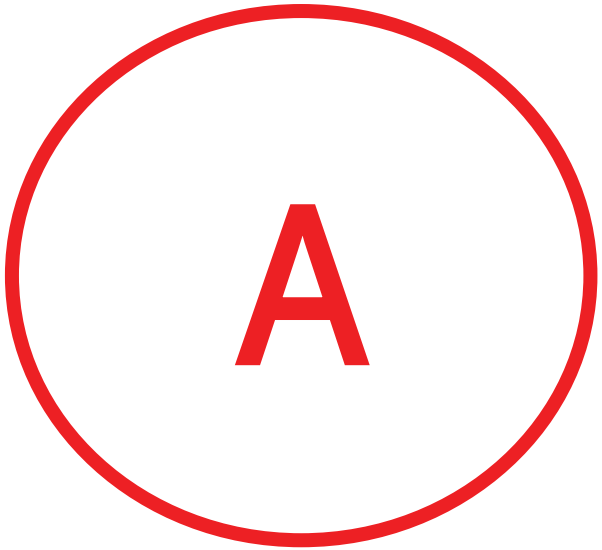
LICHEN planopilaris

- More of a lichenoid dermatitis
- Epidermal hyperplasia
- PN/EG/PB/ PV less likely
- More PF fibrosis
- Dermal interstitial mucin: not a feature
- Plasma cells (+/-)
- IMF: ‘Shaggy’ linear band of FB at BMZ

Differential diagnoses

MUCIN patterns IN SCARRING ALOPECIA



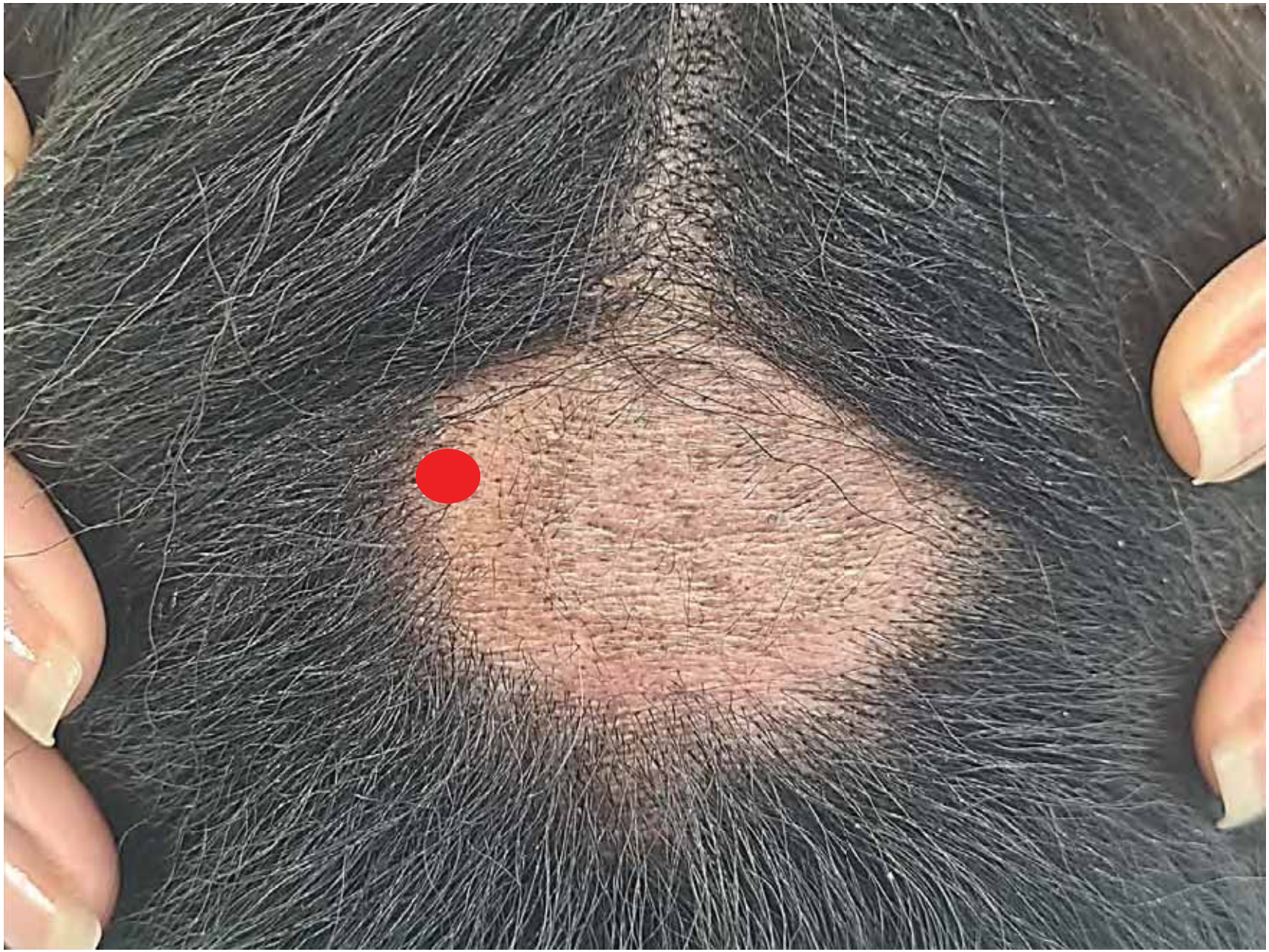


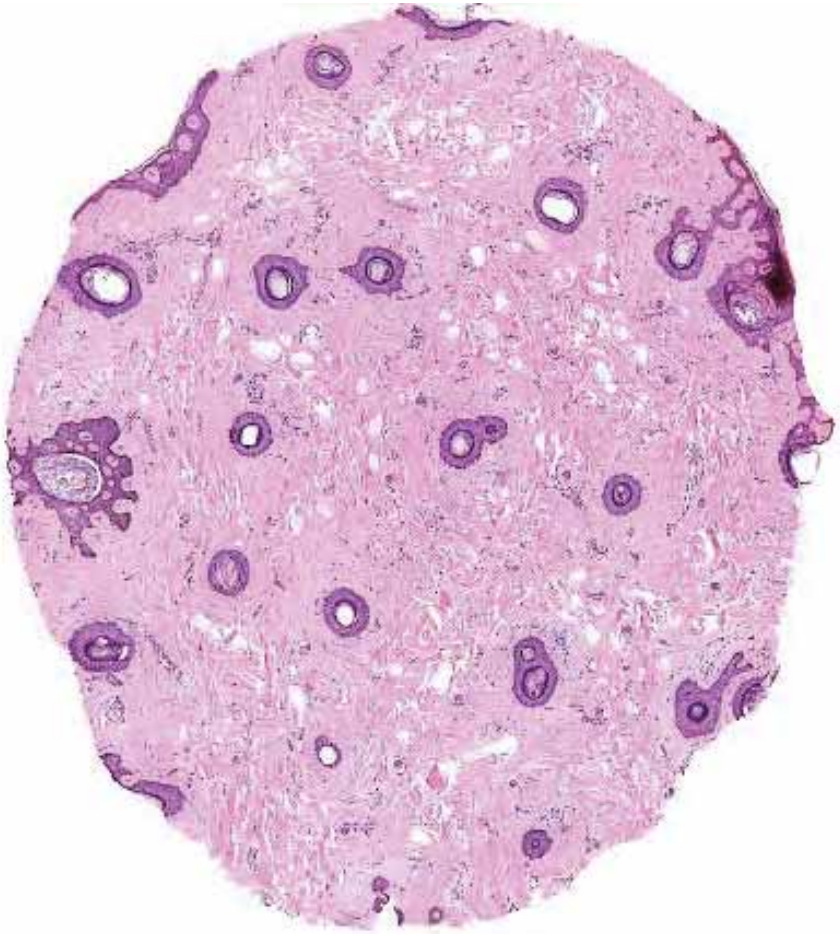
INTERFOLLICULAR STROMAL/DEEP

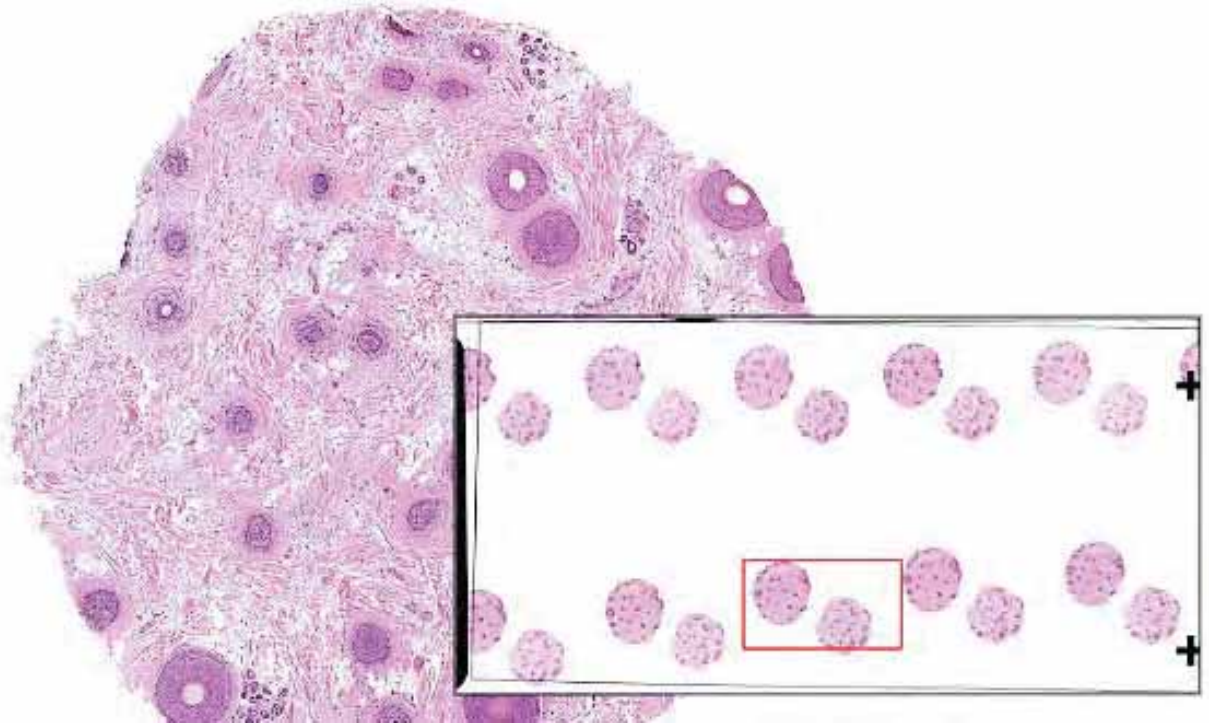
CLINICAL HISTORY

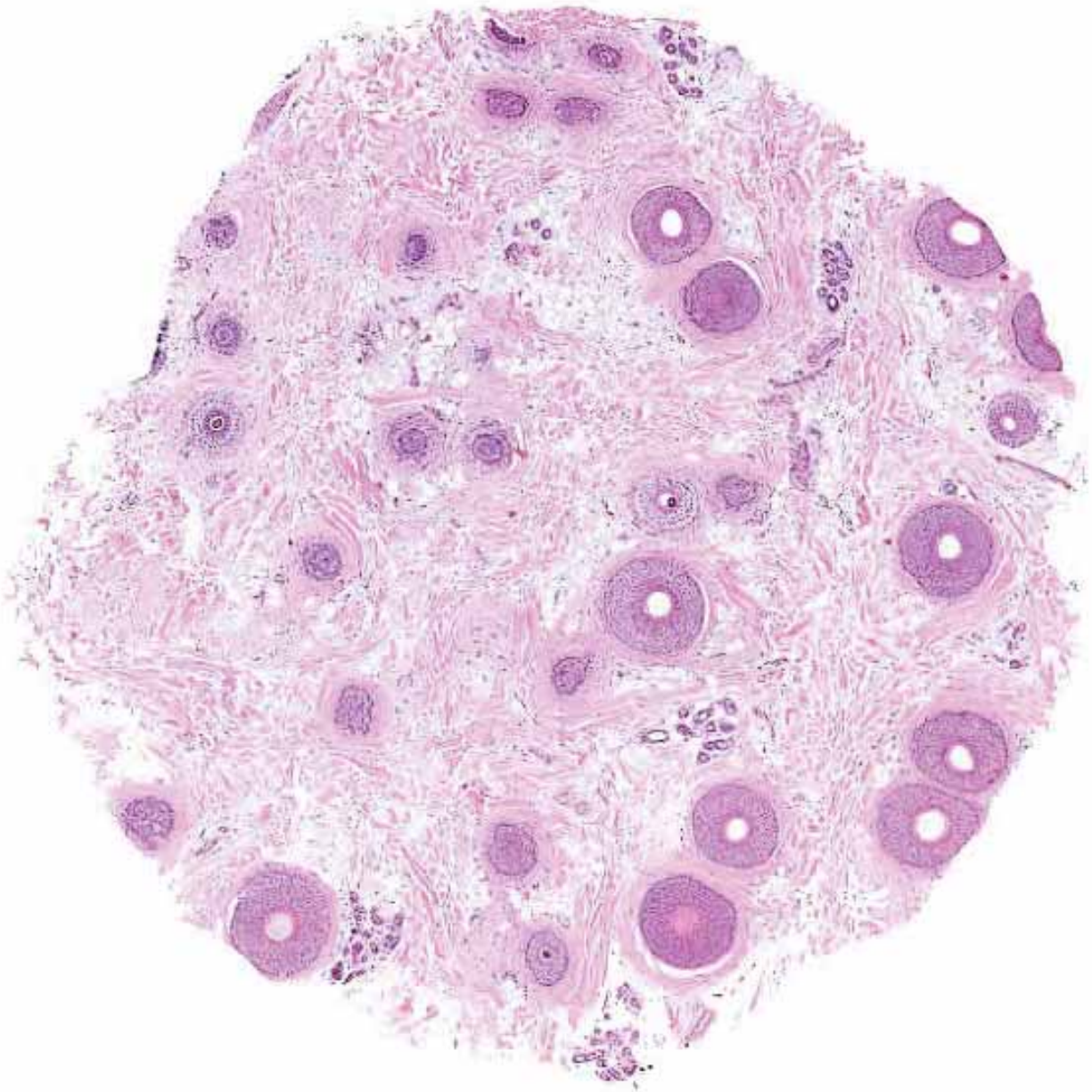
A 43-year-old female presented with a one-year's history of a solitary, asymptomatic patch of hair loss affecting the vertex of her scalp. Rule out alopecia areata versus scarring alopecia.

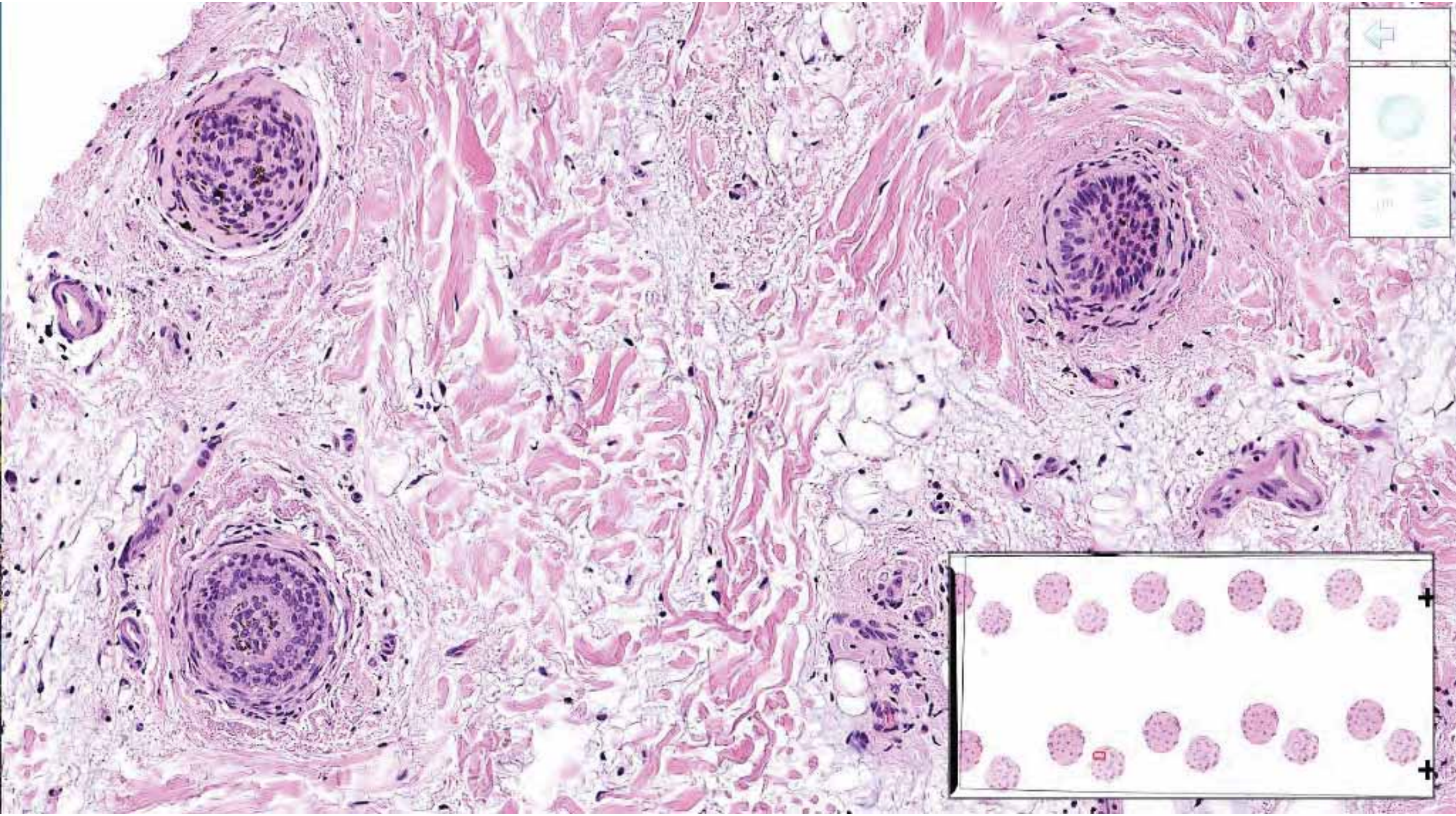
Presented in GRAZ in 2022

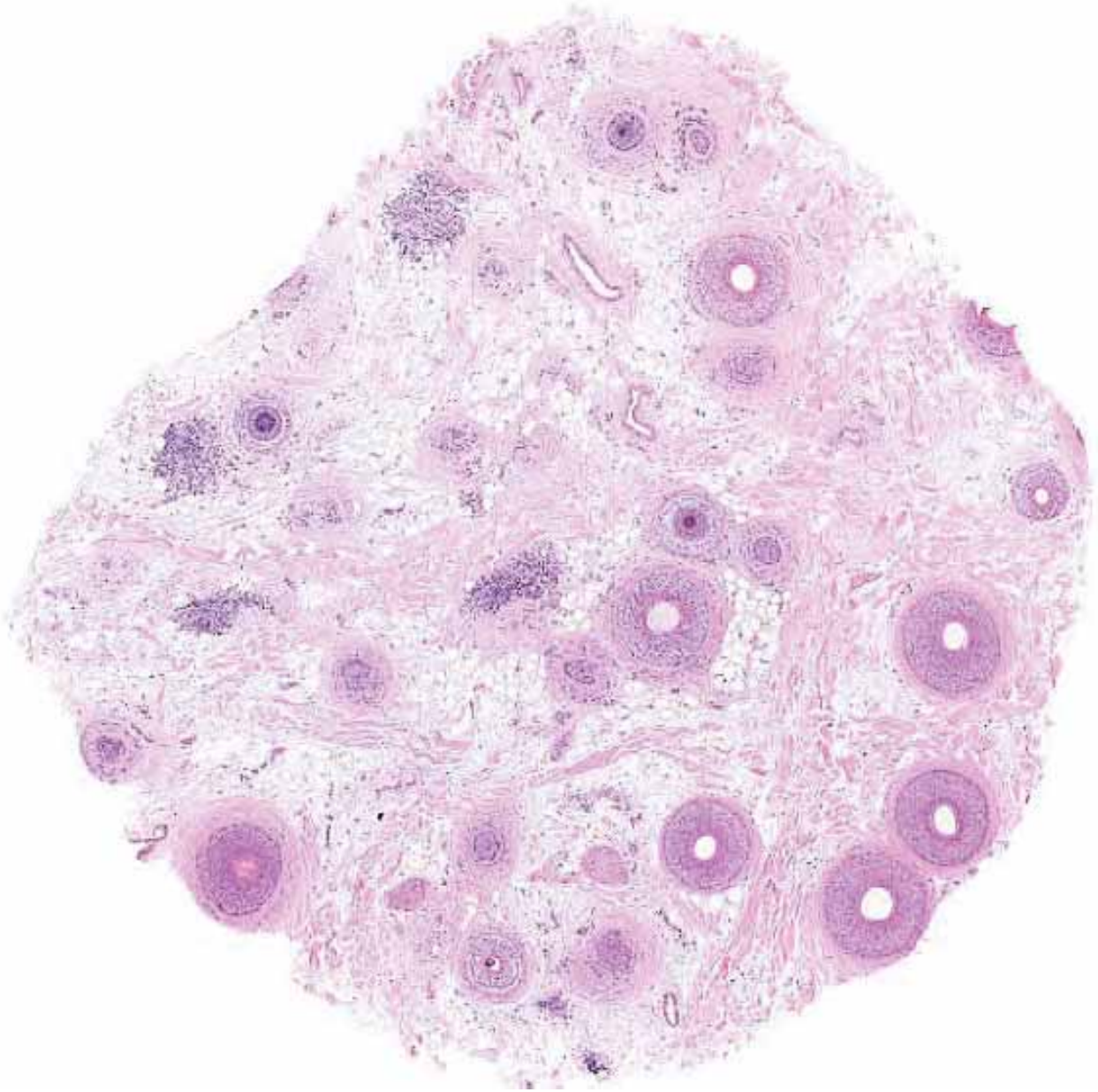


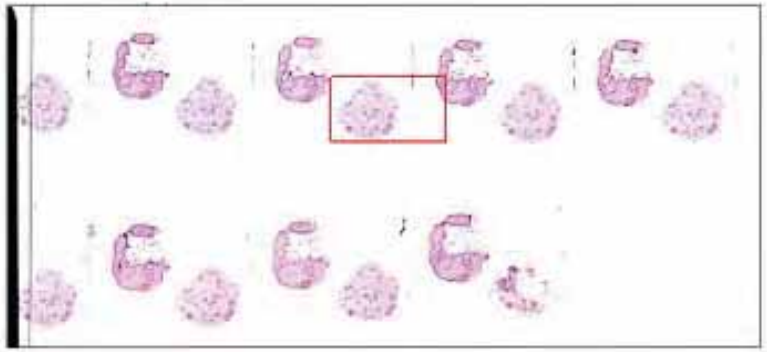


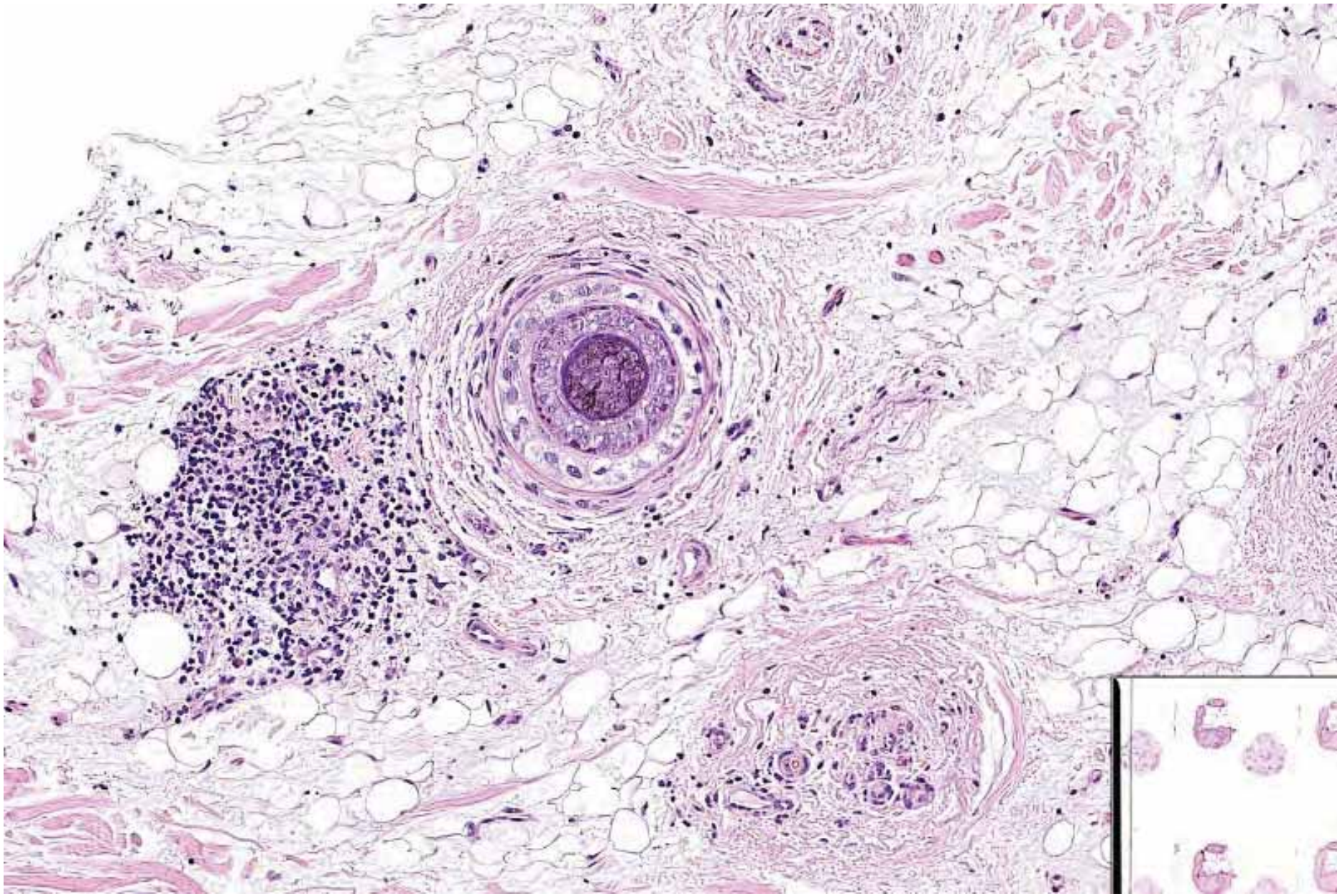




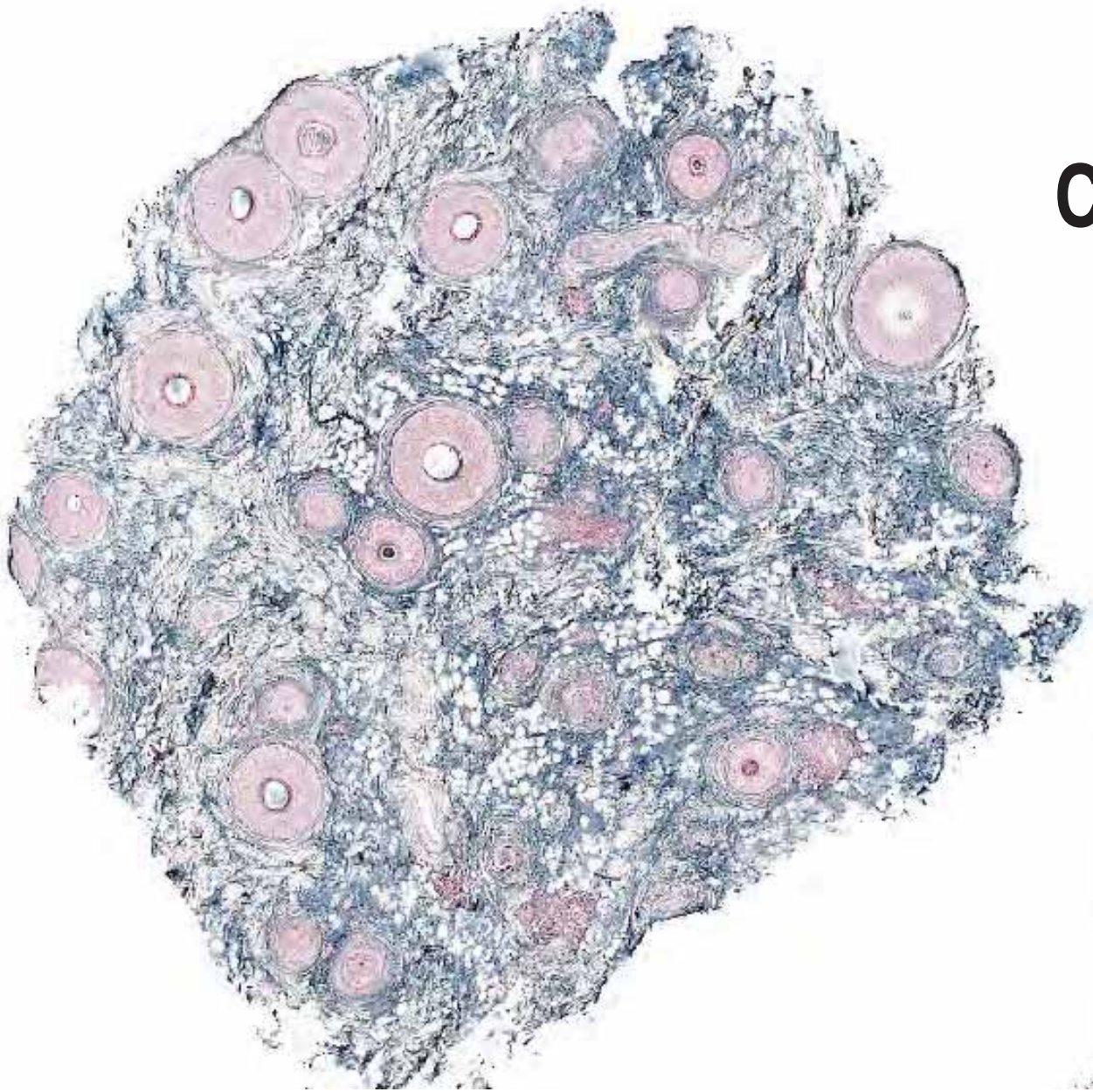


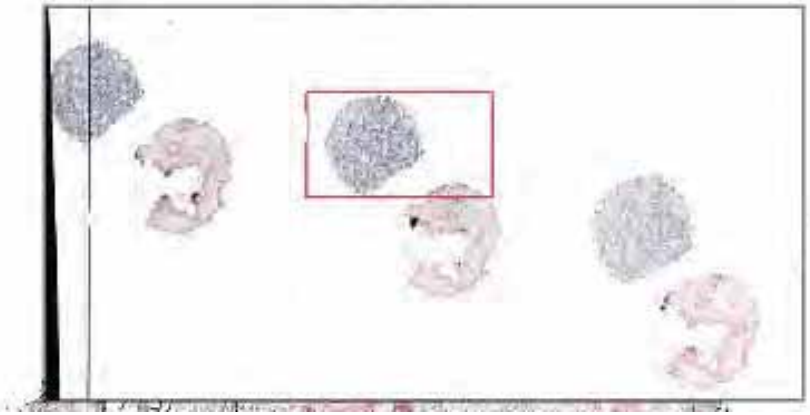






COLLOIDAL IRON





J Cutan Pathol 2008: 35: 757–760
doi: 10.1111/j.1600-0560.2007.00875.x
Blackwell Munksgaard. Printed in Singapore

Journal of
Cutaneous Pathology

Dermal mucin in alopecia areata – tell tale sign or incidental finding?

Alopecia areata is an autoimmune disease causing patchy hair loss, which occurs with an increased incidence in patients with lupus erythematosus. We report a 27-year-old African-American female with systemic lupus erythematosus and alopecia areata, whose biopsy showed a marked increase in mucin in the deep dermis and subcutis. Archival biopsies of alopecia areata were then reviewed to see if this finding occurs in patients without systemic lupus. Of 13 recent biopsies diagnostic of alopecia areata, we detected deposition of mucin in 3 (23%), but all mild in degree and in a superficial location. We speculate that the marked deposition of mucin in this patient's biopsy of alopecia areata may be related to her underlying systemic lupus, and that the presence of marked, deep dermal deposition of mucin might serve as a diagnostic clue for the presence of underlying systemic lupus in patients with alopecia areata.

Goldberg LJ, Sekhri V, Bhawan J. Dermal mucin in alopecia areata – tell tale sign or incidental finding?
J Cutan Pathol 2008; 35: 757–760. © Blackwell Munksgaard 2008.

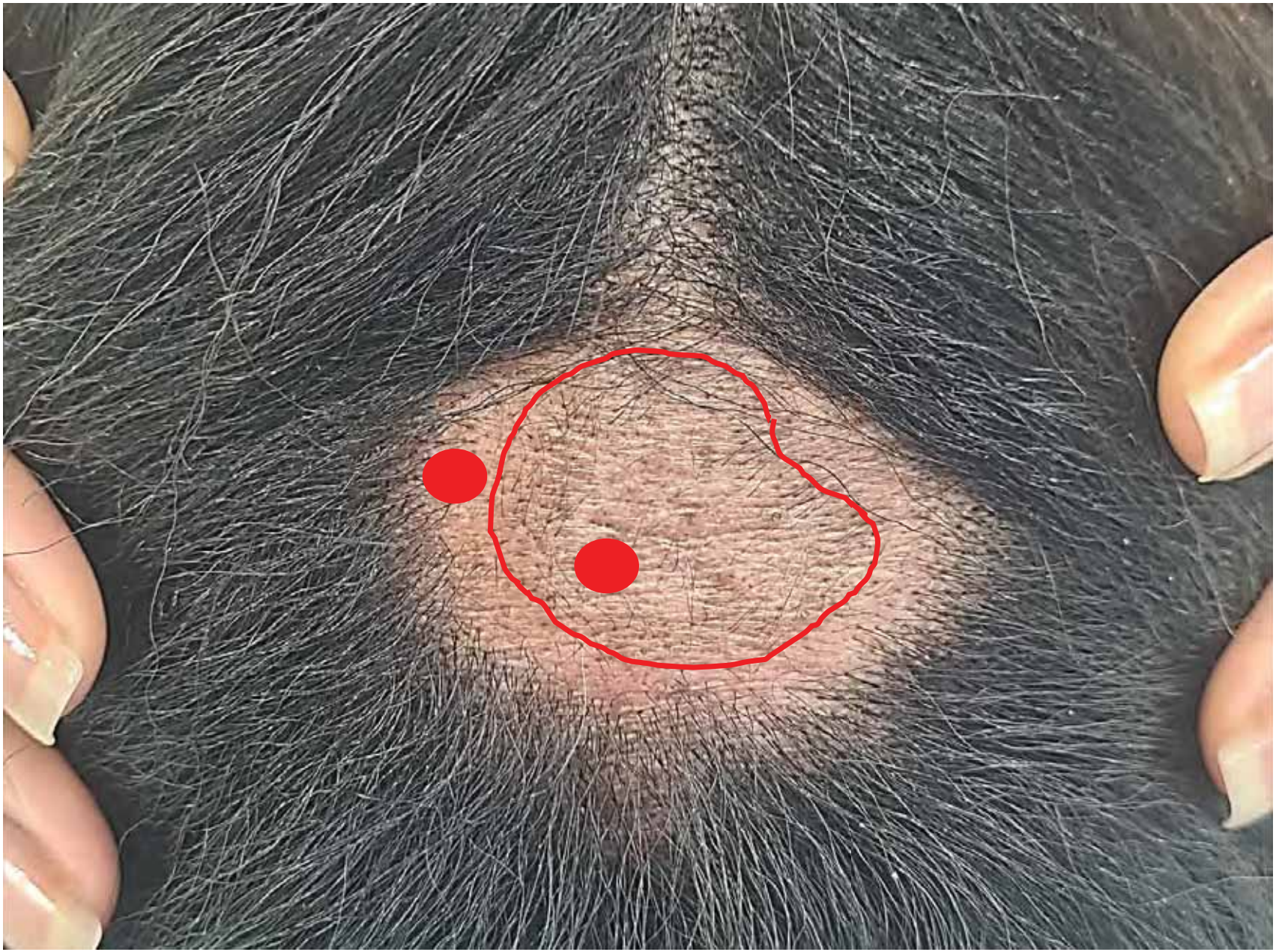
Lynne J. Goldberg, Vinoo Sekhri and Jag Bhawan

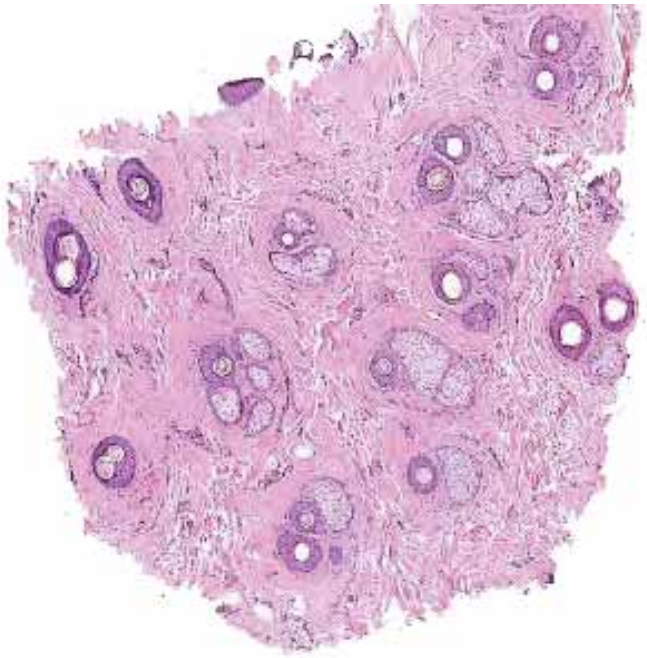
Section of Dermatopathology, Department of Dermatology, Boston University School of Medicine, Boston, MA, USA

Lynne J. Goldberg, MD, Section of Dermatopathology, Department of Dermatology, Boston University School of Medicine, 609 Albany Street, Boston, MA 02118, USA
Tel: +617 638 5562
Fax: +617 638 5515
e-mail: lynngold@bu.edu

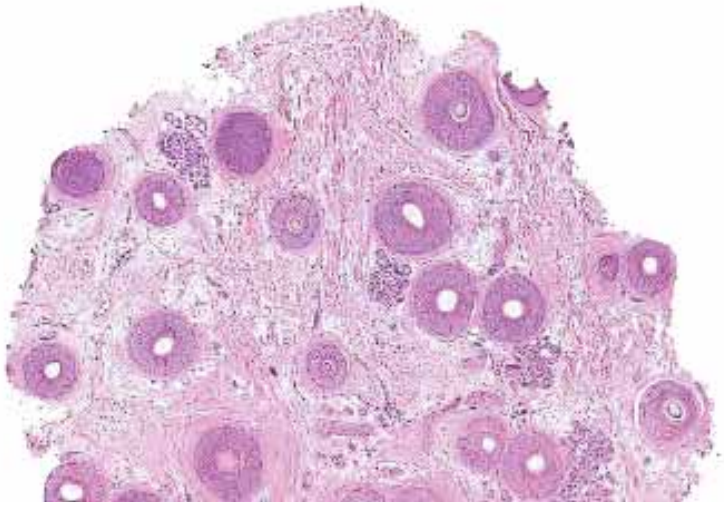
Accepted for publication July 24, 2007

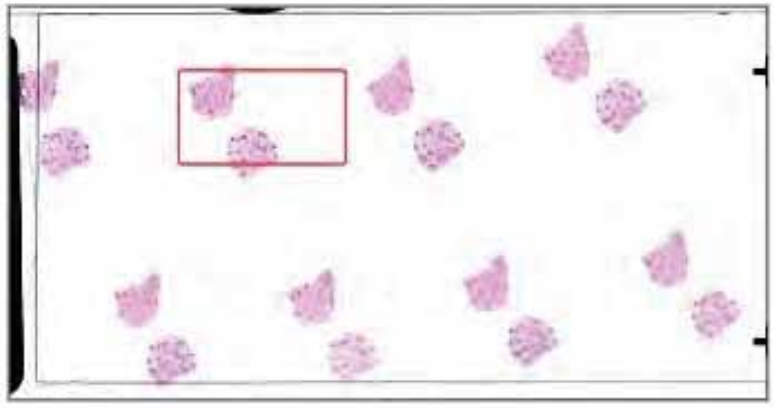
Serology: ANA and ENA Abs positive

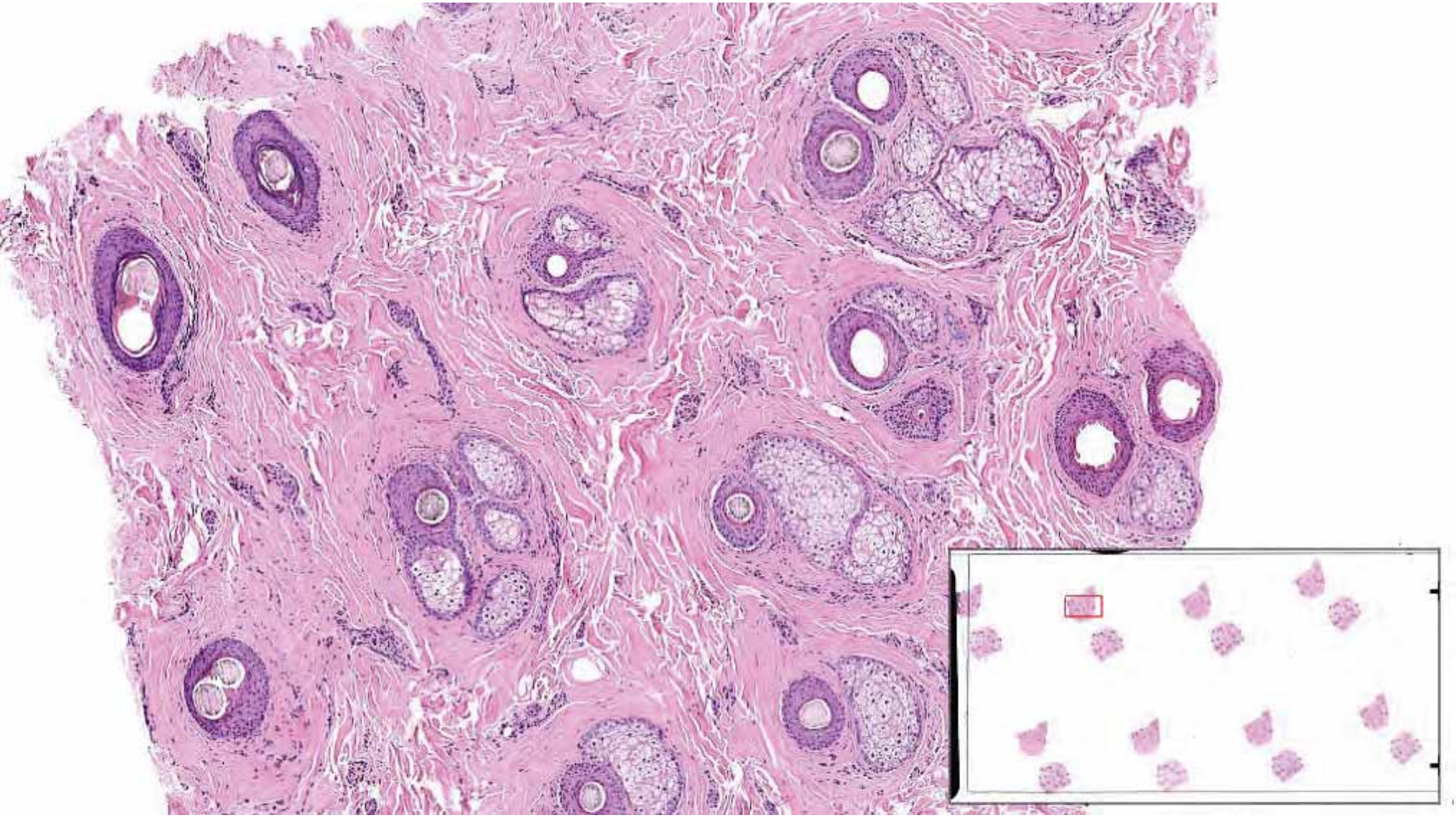


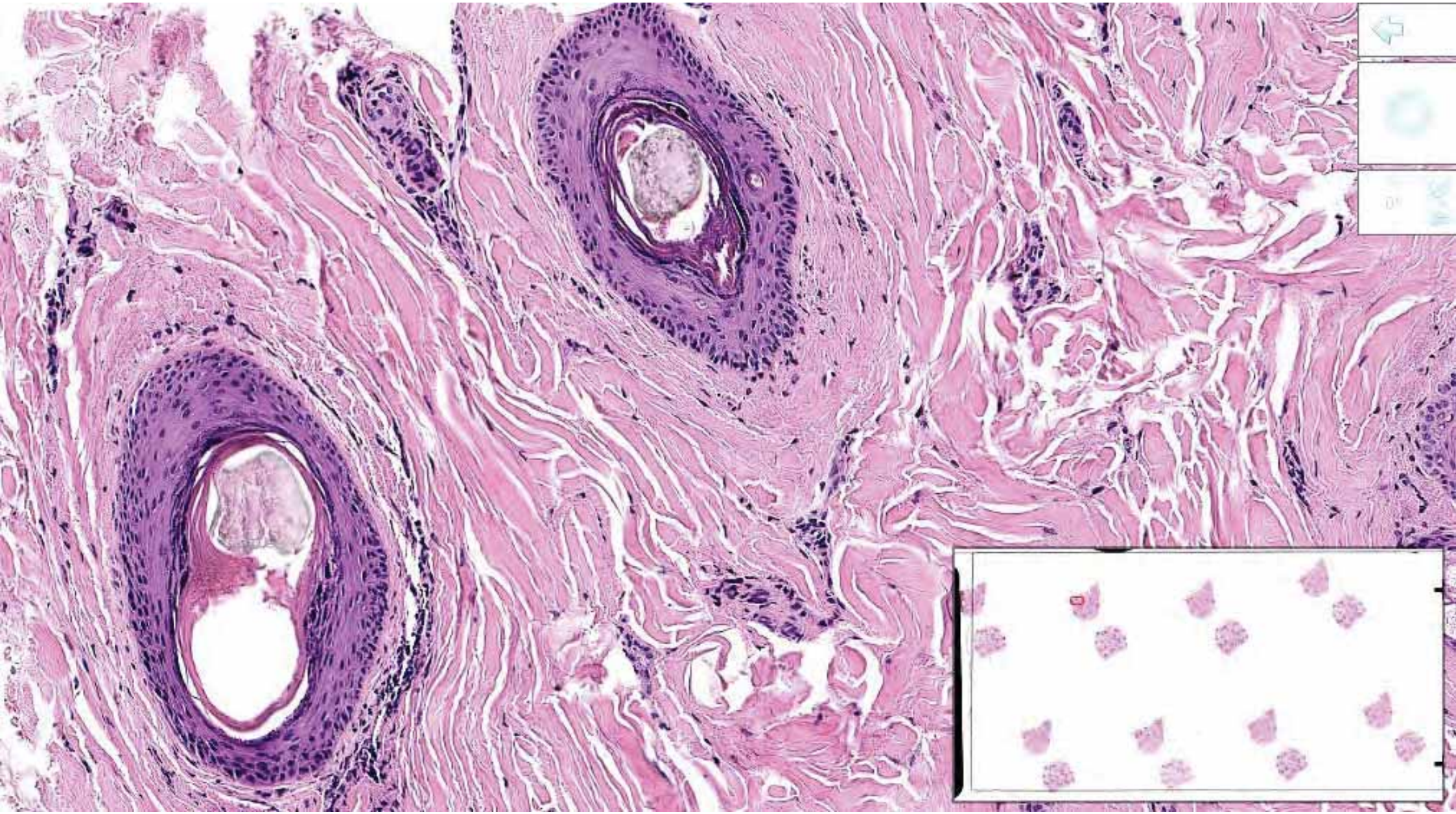


2nd biopsy

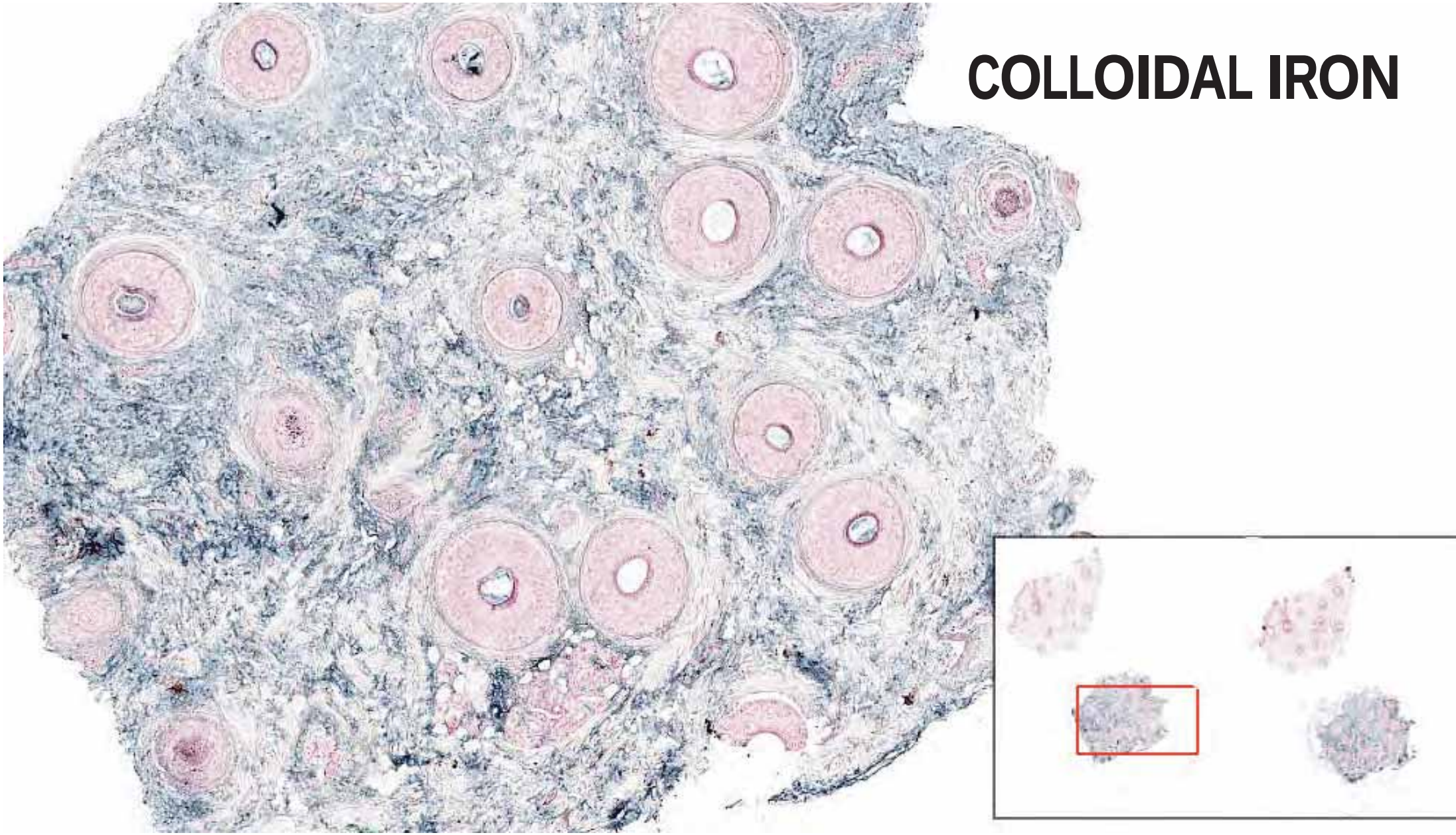


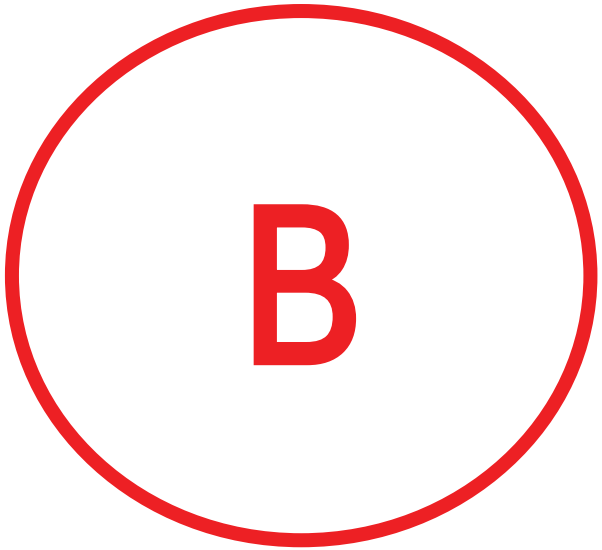






COLLOIDAL IRON





PERI-FOLLICULAR/MUCINOUS FIBROPLASIA

A histologic review of 27 patients with lichen planopilaris

Yasmeen K. Tandon,[a,*] Najwa Somani, MD,[b,c,*] Nathaniel C. Cevasco, MD,[b] and Wilma F. Bergfeld, MD[b,c]
Rootstown and Cleveland, Ohio

Background: Lichen planopilaris (LPP) is a potential trichologic emergency that can result in permanent scarring alopecia. Histopathology is a key component of the diagnostic work-up.

Objective: To identify the key histologic features that characterize LPP in order to facilitate diagnosis, ultimately leading to improved patient outcomes.

Methods: Scalp biopsy specimens from 27 confirmed cases of LPP were reviewed in a blinded fashion to determine diagnostically helpful histologic features.

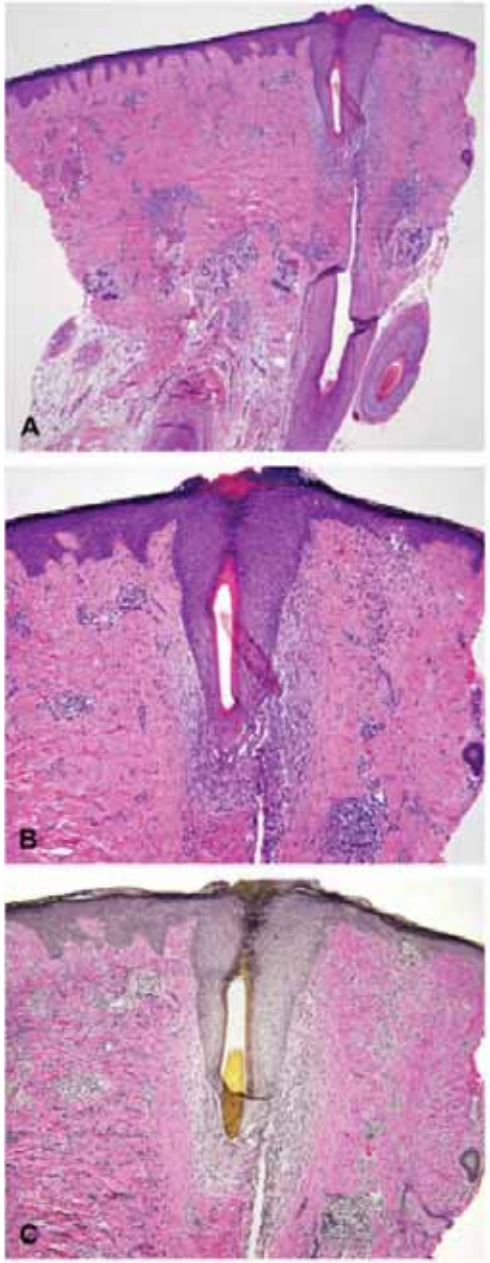
Results: Absence of arrector pili muscles and sebaceous glands, a perivascular and perifollicular lymphocytic infiltrate in the reticular dermis and mucinous perifollicular fibroplasia within the upper dermis with absence of interfollicular mucin, and superficial perifollicular wedge-shaped scarring were characterizing features.

Limitations: Sample size was limited, given that biopsy specimens were taken from lesions at varying stages of evolution and findings vary with disease stage.

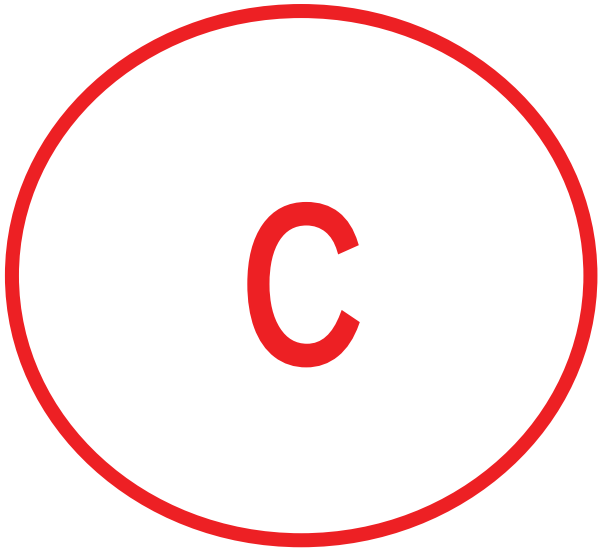
Conclusions: This study confirms many previously reported histologic features and highlights new characterizing features of mucinous perifollicular fibroplasia. (J Am Acad Dermatol 2008;59:91-8.)

Table VIII. Histologic changes to the fibrous and interstitial tissue in LPP

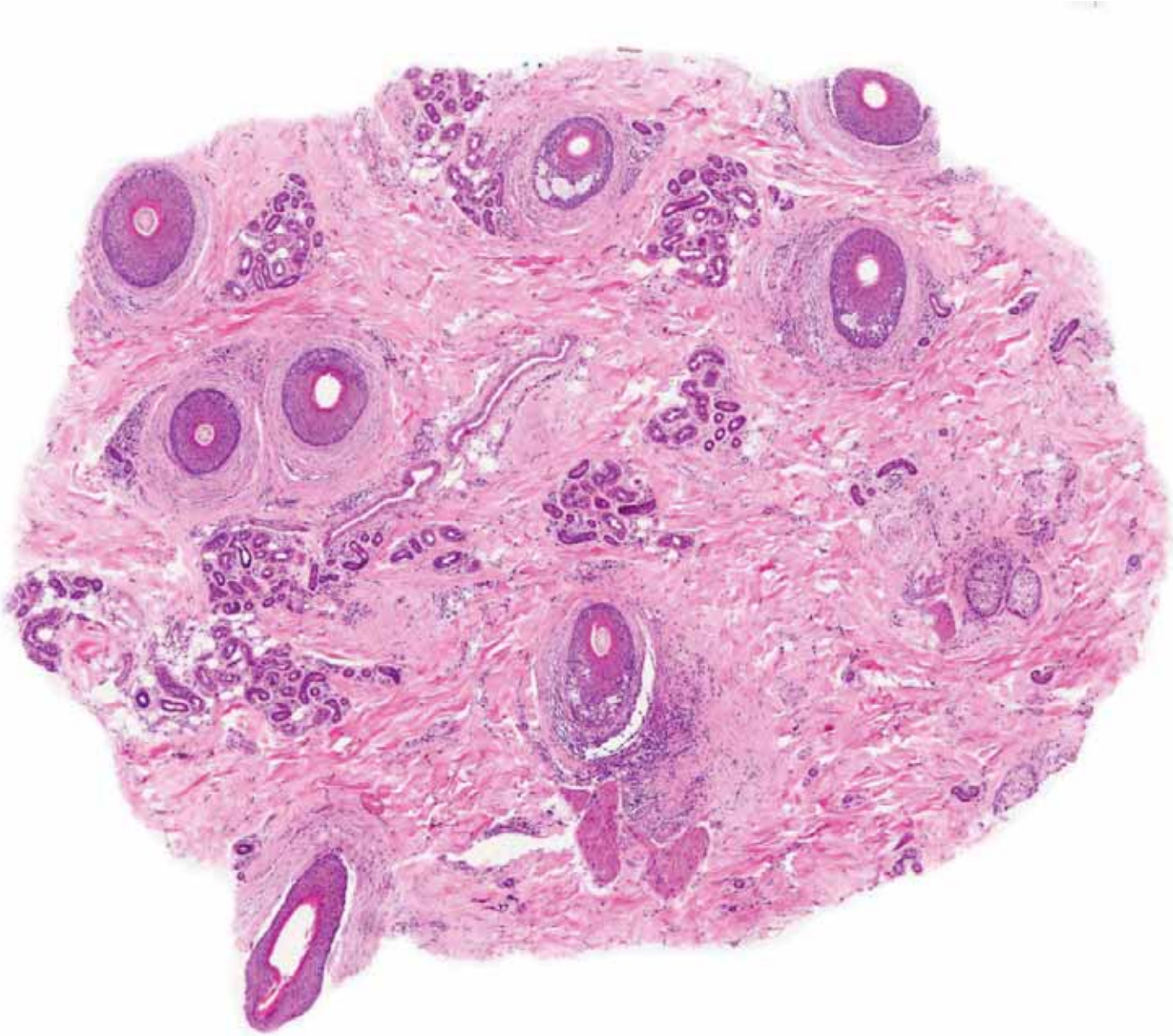
Findings	No. (%) (N = 27)
Fibrous tissue	
Perifollicular fibrosis	
Upper dermis (above bulge)	
Absent	1 (4)
Concentric lamellar fibroplasia	3 (11)
Mucinous fibroplasia	10 (37)
Hyalinization	12 (44)
Lower dermis (below bulge)	
Absent	6 (22)
Concentric lamellar fibroplasia	0 (0)
Mucinous fibroplasia	1 (4)
Hyalinization	19 (70)
Follicular tract	
Absent	2 (7)
Fibrovascular	7 (26)
Hyalinized	20 (74)
Mucinous/elastotic fibroplasia	2 (7)
Interstitial tissue	
Interfollicular mucin	
Absent	20 (74)
Mild	4 (15)
Moderate	1 (4)
Possible mucin	2 (7)
Elastic fiber pattern	
Normal	1 (4)
Perifollicular scar	11 (41)
Superficial perifollicular wedge shaped	16 (60)
Diffuse scar (involving interfollicular dermis)	6 (22)

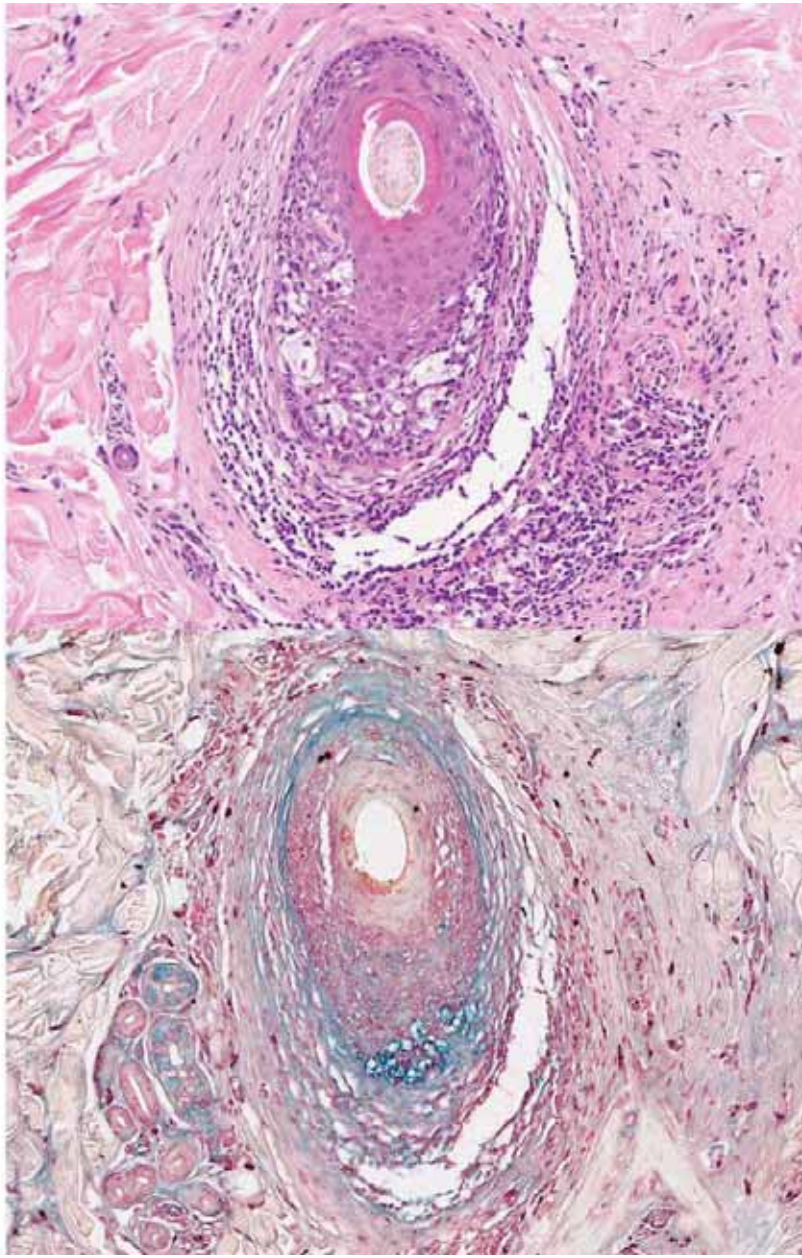


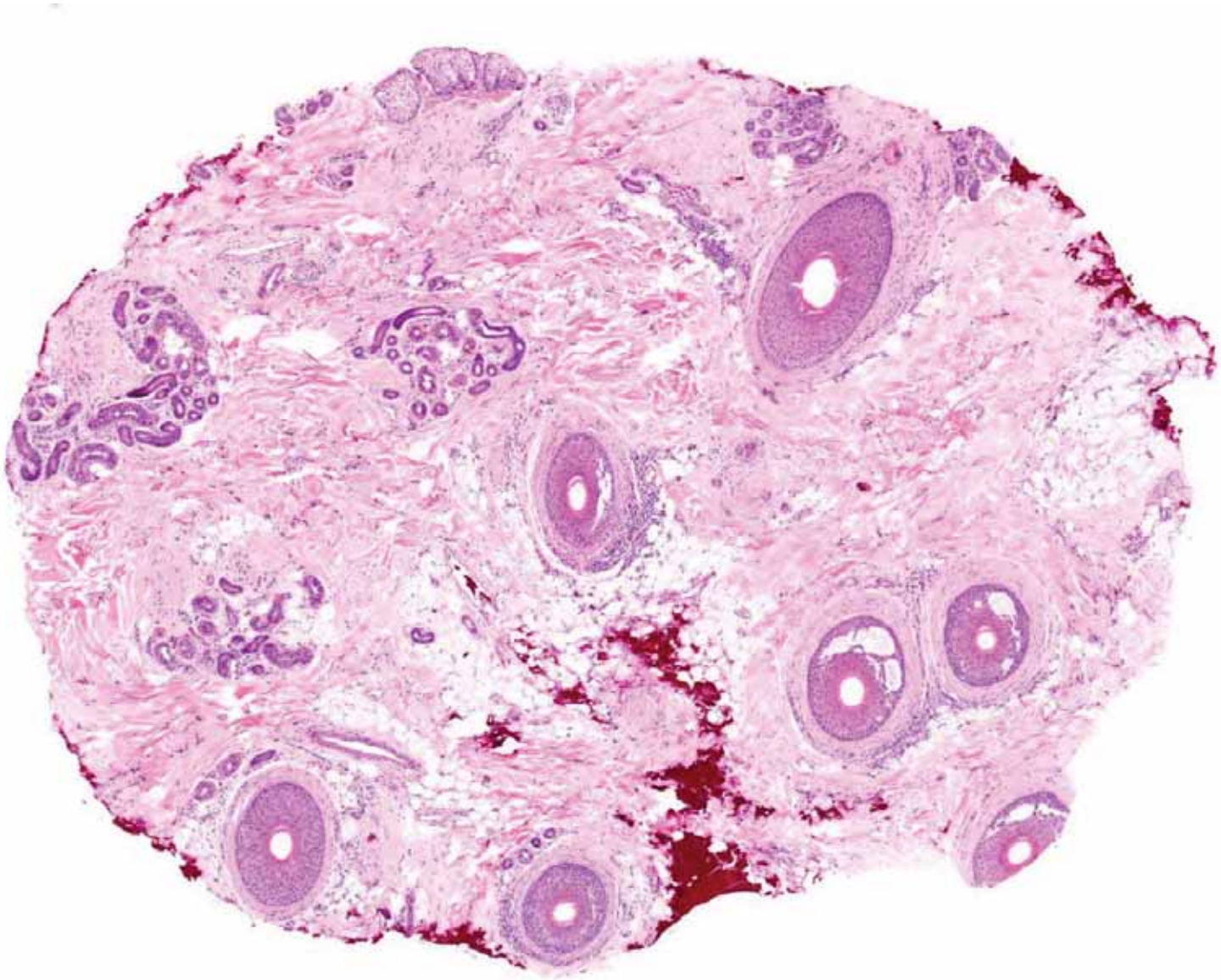
Tandon et al. JAAD 2008;59:91-98

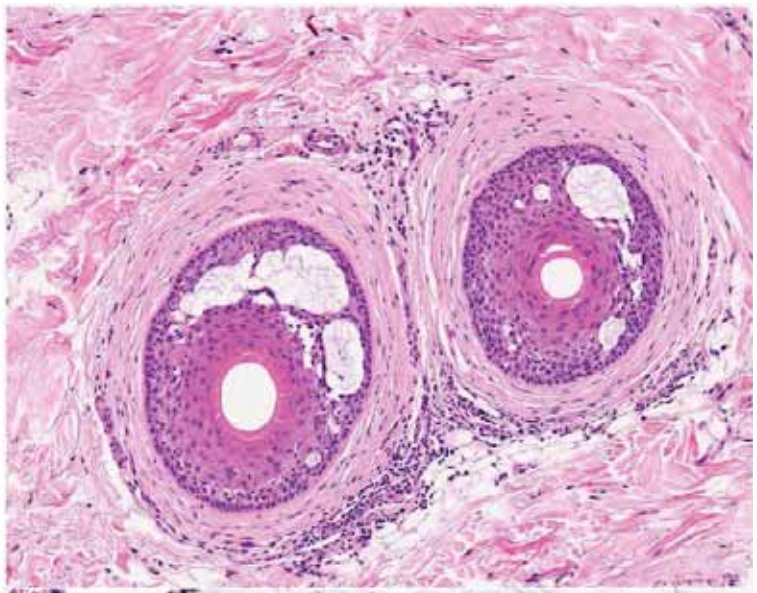


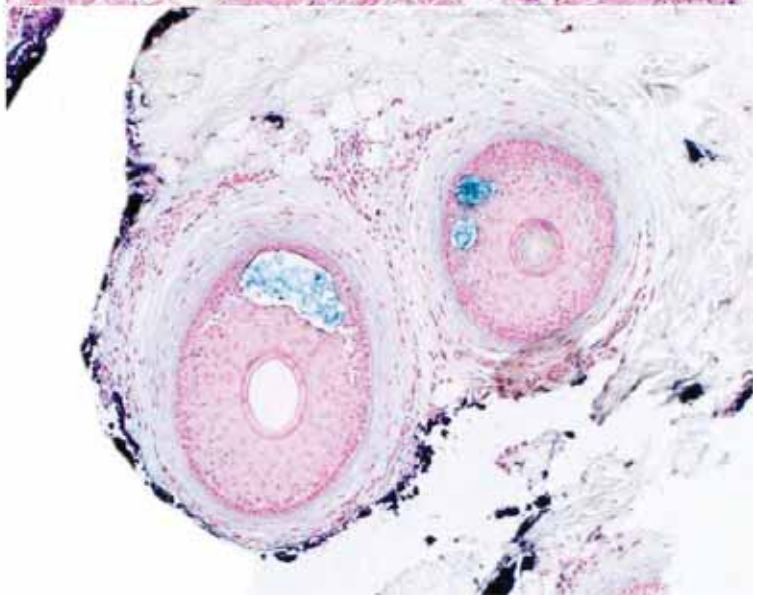
FOLLICULAR/EPITHELIAL ‘BUBBLES’

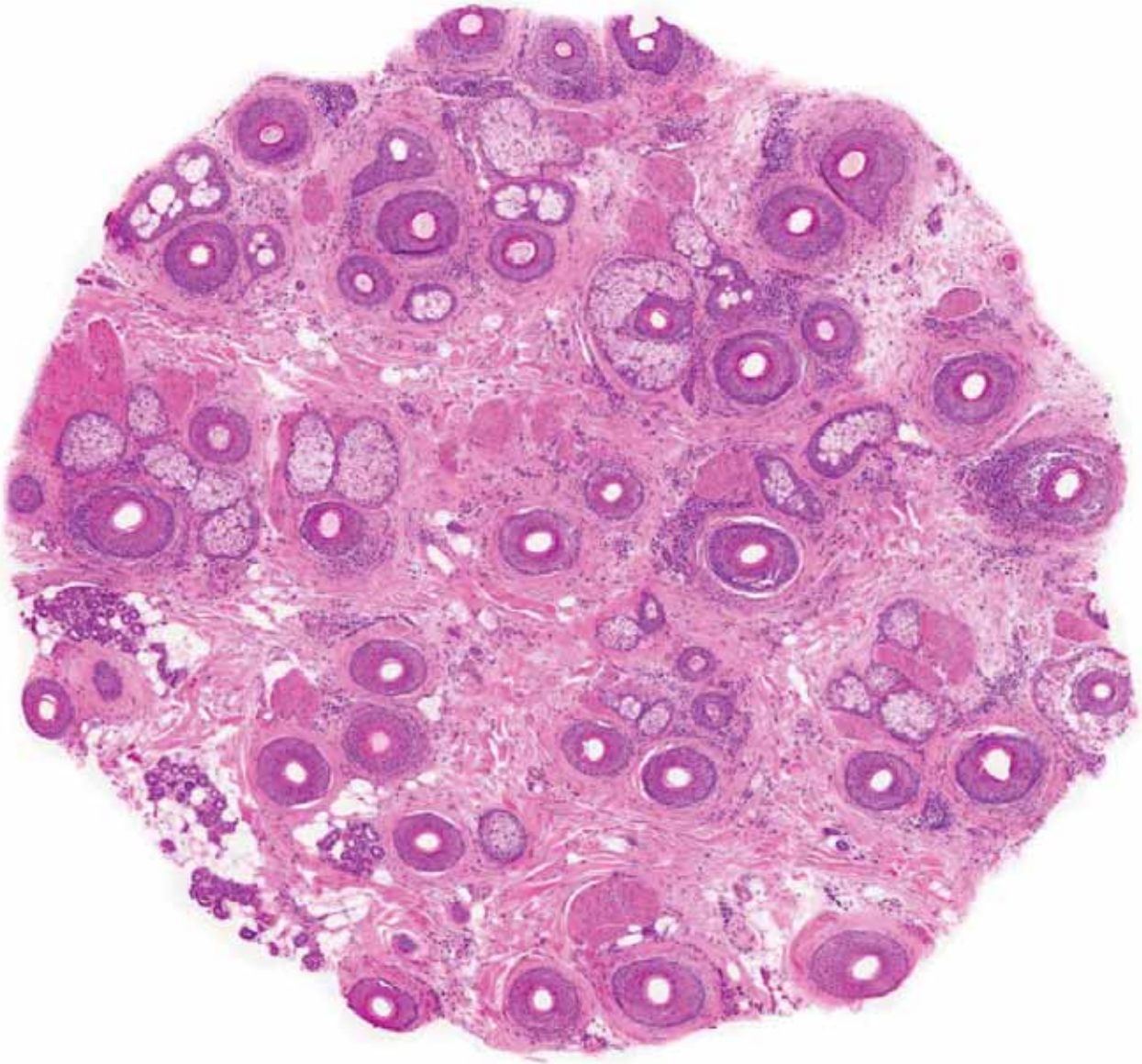


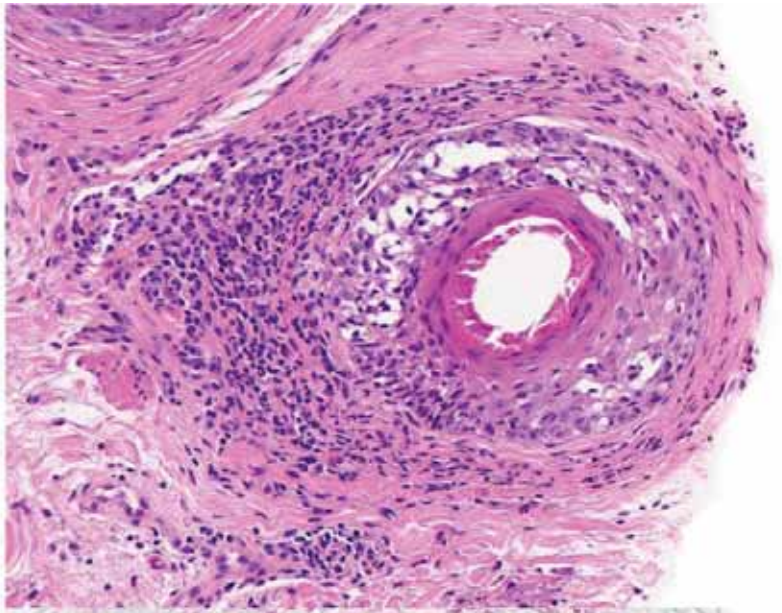


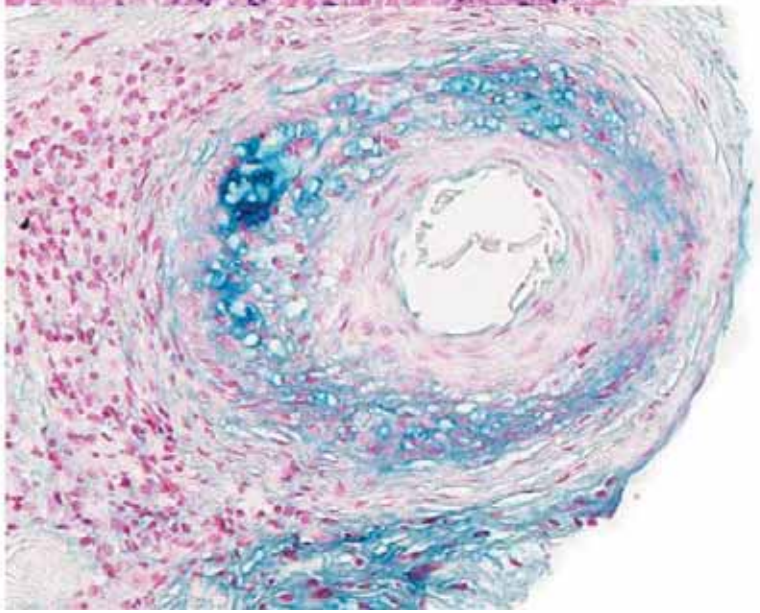


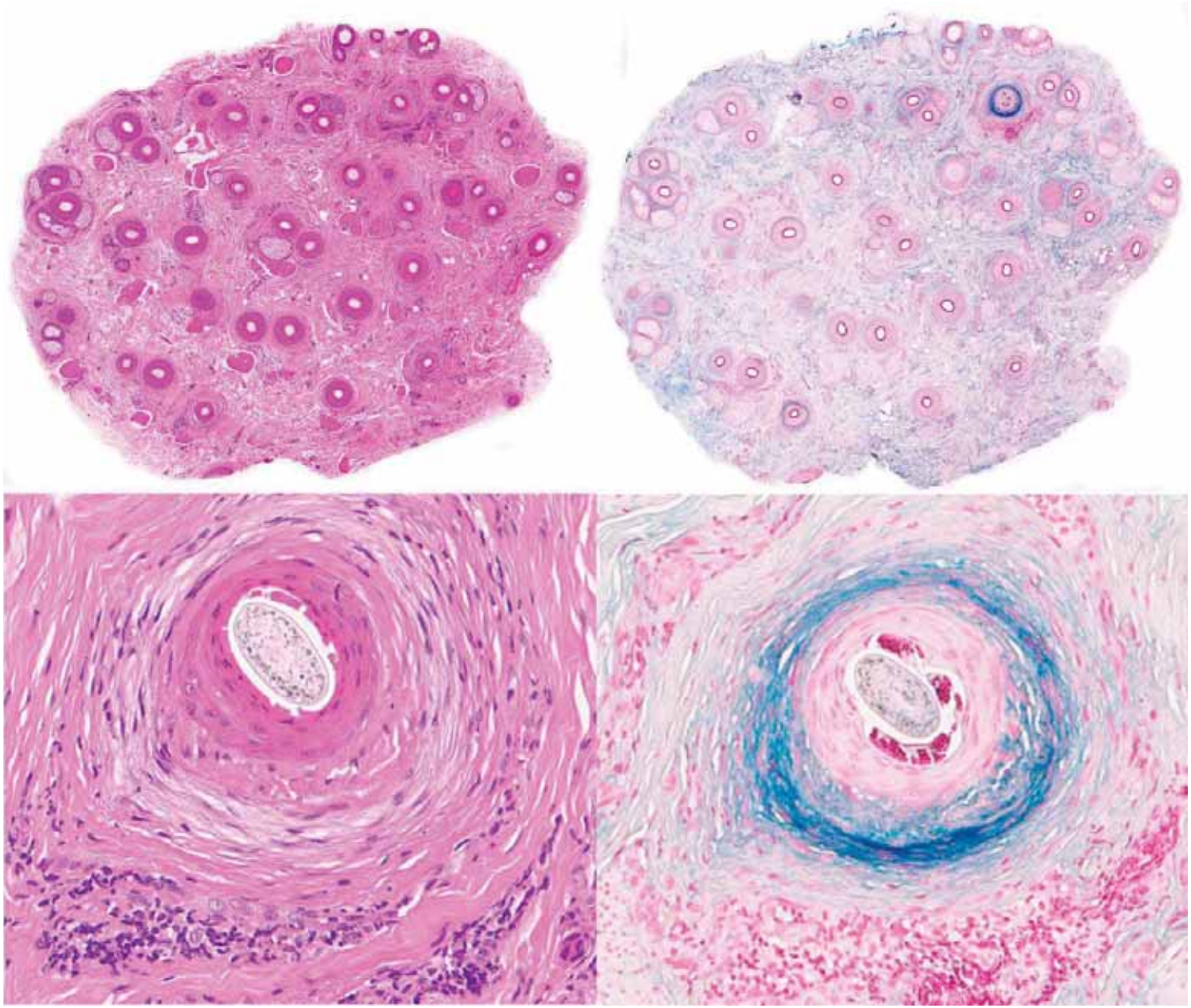












Patterns of Mucin Deposition in Lichen Planopilaris: A Journey From Follicular "Bubbles" to Perifollicular Fibroplasia

Maged Daruish, MSc, Eleni Ieremia, MD, Dip RCPath,† and Catherine M. Stefanato, MD, FRCPath**

Abstract: Lichen Planopiolaris (LPP) is a scarring alopecia characterised by a perifollicular lymphoid cell infiltrate at the level of the infundibulum and isthmus. While perifollicular mucinous fibroplasia is an established finding in LPP, intrafollicular mucin deposition has not been previously reported. We describe two cases with this histopathology and suggest it may represent a helpful clue to the diagnosis of LPP, in the appropriate clinical setting.

Key Words: lichen planopilaris, scarring alopecia, follicular mucinosis, mucin

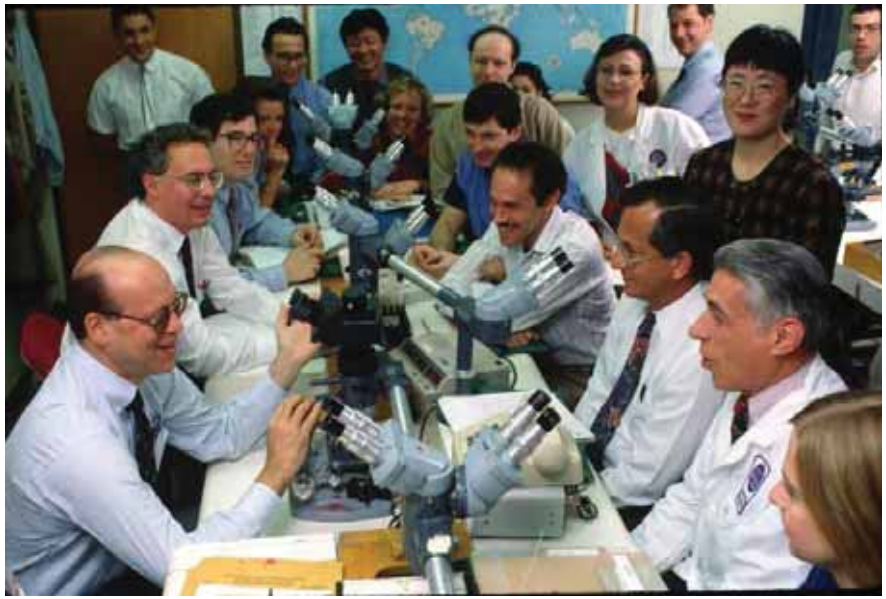
(*Am J Dermatopathol* 2023;45:635–638)

fibroplasia develops in which mucin deposition has been described.[4]

Typically, no intraepithelial or interfollicular dermal mucin is present.[4] We report 2 cases in which mucin accumulation was observed within the hair follicle epithelium in LPP.

CASE 1

A 36-year-old woman of Latin-American origin presented with a long-term history of hair thinning that started postpartum and a new area of hair loss involving the left temple only. Mild itching of the scalp was present. Otherwise, she was fit and well. On examination, there were pilar casts, mild scale and postinflammatory hyperpigmentation on the left temple, and an overall diffuse hair thinning. Laboratory workup



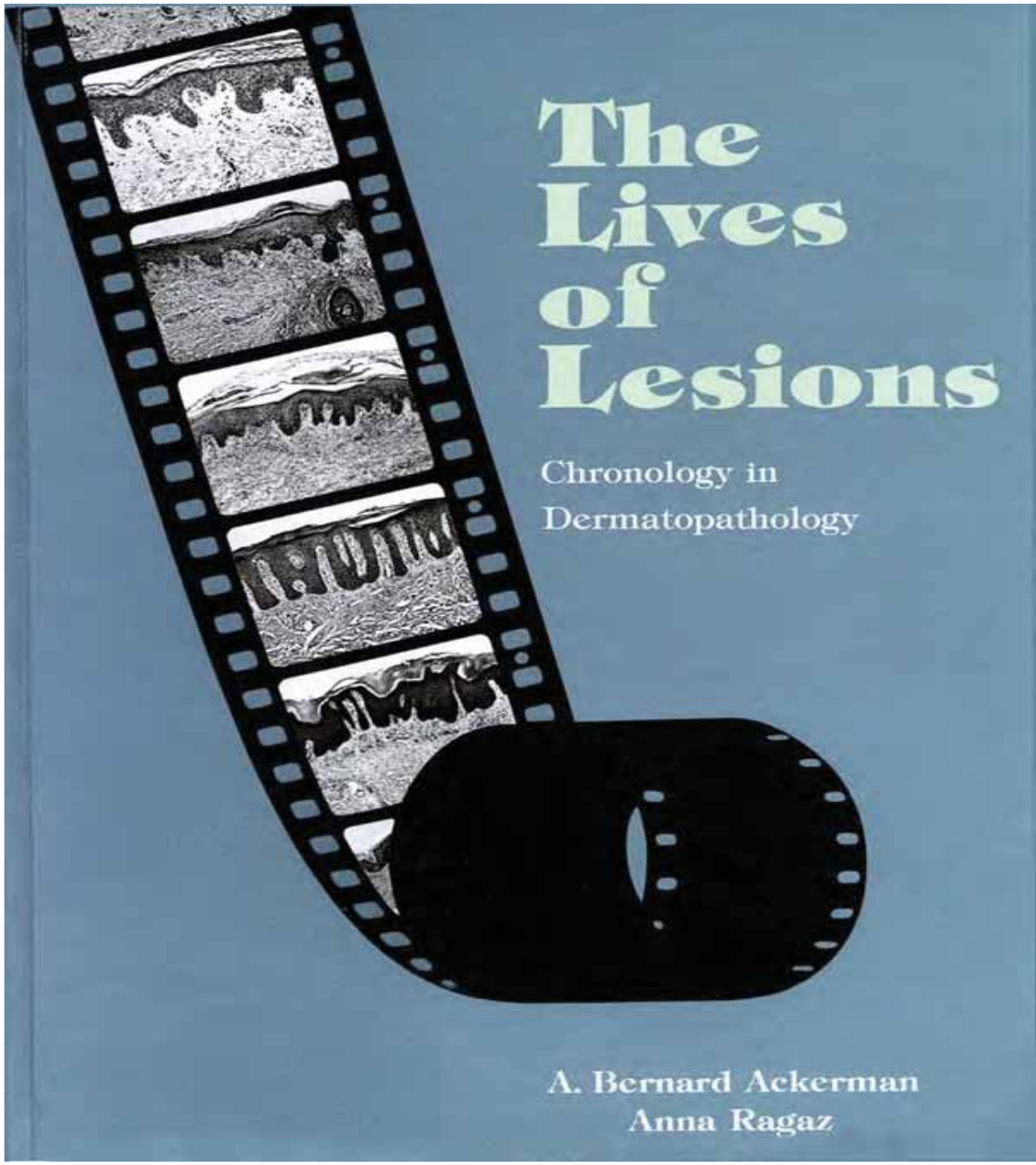

The Lives of Lesions
Chronology in Dermatopathology
A. Bernard Ackerman
Anna Ragaz

TABLE 1. Patterns of Follicular Mucin Deposition in Our Cases of Lichen Planopilaris

	Active LPP	Late LPP
Intraepithelial follicular mucin	+/++	—
Vacuolar-interface changes of the follicular basal cell layer	+/++	—
Perifollicular lymphoid cell infiltrate	+/++	+/—
Perifollicular mucinous fibroplasia	—	++

"+", present; "—", absent.

DIFFERENTIAL DIAGNOSIS

TABLE 2. Differential Diagnosis of Follicular Mucin Deposition in Alopecia

	Lichen Planopilaris	Idiopathic Follicular Mucinosis	Folliculoptropic Mycosis Fungoides
Atypia of lymphocytes	—	—	+
Vacuolar-interface changes of the follicular basal cell layer	+	—	—
Perifollicular mucinous fibroplasia	+	—	—

"+", present; "—", absent.

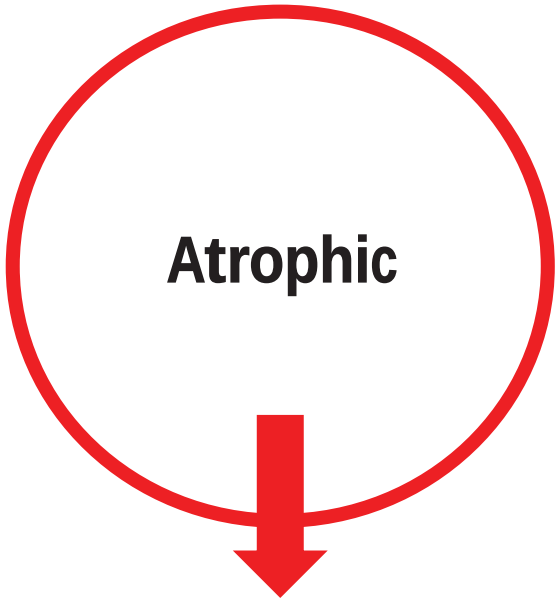
‘DECODING’ MUCIN patterns IN SCARRING ALOPECIA

INTERFOLLICULAR	FOLLICULAR	PERI-FOLLICULAR
Stromal/deep	Epithelial bubbles	Mucinous fibroplasia
		
LUPUS	**LPP (EARLY)**	**LPP(LATE)**

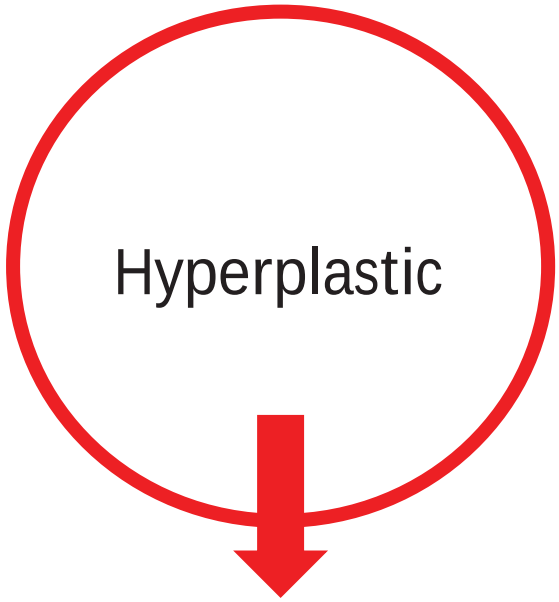
‘DECODING’ EPIDERMAL PATTERNS IN SCARRING ALOPECIA

Differential diagnoses

EPIDERMAL PATTERNS IN SCARRING ALOPECIA



Lupus



FOLLICULITIS DECALVANS

‘DECODING’ EPIDERMAL HYPERPLASIA IN FOLLICULITIS DECALVANS

Received: 28 August 2016 | Revised: 25 December 2016 | Accepted: 3 January 2017

DOI 10.1111/cup.12892

WILEY JCP

ORIGINAL ARTICLE

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

Bruno Matard[1] | **Benedicte Cavelier-Balloy**[2] | **Pascal Reygagne**[1]

[1]Hôpital Saint-Louis, Centre Sabouraud, Paris, France

[2]Department of Dermatopathology, Policlinique Hôpital Saint Louis, Paris, France

Correspondence
Bruno Matard, Centre Sabouraud,
Hôpital Saint-Louis,
1 Avenue Claude Vellefaux,
75010 Paris,
France.
Email: bruno.matard@wanadoo.fr

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.

KEYWORDS

dysbiosis, epidermal hyperplasia, follicular microbiota, folliculitis decalvans, hidradenitis suppurativa

92%

TABLE 1 Histological findings in 26 FD patients

Histological criteria	Severity	n, (%)
Follicular hyperkeratosis[1]	No	6/26 (23)
	Mild	19/26 (73)
	Pronounced	1/26 (4)
Epidermal hyperplasia	Yes	24/26 (92)
	Not assessable	2 /26 (8)
	Hyperkeratosis of the stratum Corneum	0/26
	Orthokeratosis	21/24 (88)
	Parakeratosis	3 /24 (12)
	Psoriasiform	21/24 (88)
	No psoriasiform	3/24 (12)
Hyperplasia of the follicular epithelia	Yes	0/26
Atrophy of the follicular epithelia	Yes	22/26 (85)
	No	4/26 (15)
Perifollicular cellular inflammatory infiltrate[3]		
Polymorphonuclear neutrophils	No	3/26 (11)
	Mild	10/26 (39)
	Pronounced	13/26 (50)
Lymphocytic cells	No	0/26
	Mild	14/26 (54)
	Pronounced	12/26 (46)
Plasma cells	No	2/26 (8)
	Mild	13/26 (50)
	Pronounced	11/26 (42)
Inflammatory granuloma	No	9/26 (35)
	Yes	17/26 (65)
Follicle rupture	Not assessable	3/26 (11)
	No	6/23 (26)
	Yes	17/23 (74)
Follicular microcyst	Yes	15/26 (60)
	No	11/26 (40)
Polytrichia	Yes	14/26 (54)
	No	12/26 (46)
Presence of sebaceous glands	Yes	4/26 (15)
	No	22/26 (85)
Dermal fibrosis[2]	No	0/26
	Mild	12/26 (46)
	Pronounced	14/26 (54)
Ostial pustules	Yes	16/26 (62)
	No	10/26 (38)

characterized by a folliculitis whose perifollicular inflammatory infiltrate is composed of lymphocytes[1,26,27] and neutrophils.[1,28,29] Such neutrophils are most commonly situated on the upper part of the hair follicle in the case of FD. The progression of both diseases leads to the destruction of the hair follicle with an inflammatory granuloma formation, which contains histiocytes, giant cells and keratin fragments. The indefinite chronicity of FD and HS is a significant common fact as well as the very frequent but always temporary efficiency of antibiotics on potential causative bacteria (essentially *Staphylococcus aureus* in FD[5,15] and *S. aureus*, *Staphylococcus epidermidis* and anaerobic bacteria in HS[30-32]) even though the majority of patients do not have any deficit of the adaptive immune system nor any known genetic abnormality. Moreover, both diseases share a surprising and almost constant absence of inflammatory draining lymph nodes in spite of suppurating lesions which suggest that the immune system is involved in a common and specific manner. The fact that the hair follicle is the privileged place for inflammation in the form of folliculitis with neutrophils (superficial in FD, deep in HS) and that one of the only ways to stop it definitively is the destruction or removal of all hair follicles in the affected regions[16,26-28,33,34] suggests that the hair follicle is the place to look for the cause of chronicity in both diseases. The presence of follicular hyperkeratosis in FD with a histological aspect of follicular plugging as described in both FD and HS[7,35] raises the question of the role of follicular occlusion in its pathogenesis, like in HS, where it is currently considered as a crucial event.[36-39]

The psoriasiform aspect of the epidermal hyperplasia in both FD and HS suggests a common pathological mechanism of this hyperplasia. Indeed, by analogy with HS, for which the presence of interleukine-17 (IL-17) and of a T17 cells infiltrate in lesional and normal-appearing skin[35,40,41] was shown, the presence of epidermal hyperplasia in FD suggests the hypothesis of an IL-17 cytokine profile and/or the role of other cytokines produced by Th17 cells, such as IL-22, known for its role in inducing hyperplasia.[40,42] This hypothesis is consistent with the only immunohistological study that had been previously conducted in FD,[9] which showed that transforming growth factor beta, a cytokine implicated in Th-17 cells differentiation, is strongly expressed in the perifollicular and interfollicular dermis as well as, to a lesser extent, in the follicular epithelia. Moreover, IL-17A and IL-17 F, produced by the Th-17 lymphocytes, are key cytokines for the recruitment and activation of neutrophils. They can target numerous cell types including keratinocytes, endothelial cells, monocytes and fibroblasts to induce

Matard B et al. JCP2017;44:352-357

Epidermal thickness is useful in distinguishing lichen planopilaris from neutrophil-poor/lymphocyte-predominant folliculitis decalvans

26 cases of LPP

Lichen planopilaris (LPP) and folliculitis decalvans (FD) can, at times, be difficult to distinguish clinically as well as histopathologically from each other.[1] There are a small number of reported cases of FD-LPP overlap[2]; however, these cases are limited in number, and the most commonly used working classification of primary cicatricial alopecia remains the North American Hair Research Society (NAHRS) in which FD is categorized as a "neutrophilic" primary cicatricial alopecia along with dissecting folliculitis/cellulitis. Likewise, LPP is classified as "lymphocytic." However, advanced lesions of FD may be neutrophil-poor, featuring mostly lymphocytes and plasma cells.[3] Neutrophil-poor FD may also be termed "lymphocyte-predominant." In cases of neutrophil-poor FD, a distinction from LPP may be challenging if not impossible.[1,4,5] One useful clue in distinguishing FD from LPP is the number of fused follicular infundibula in a compound follicle. FD may have six or more fused follicles, colloquially referred to as "six-packs," while LPP never has more than two or three fused follicular infundibula in a compound follicle.[6] Still, this clue may be absent, and there is a need for additional tools to help make a diagnostic distinction between LPP and neutrophil-poor FD. Recently, previously unrecognized epidermal hyperplasia in FD was described and measured, thereby providing a new histopathologic criterion to diagnose FD.[1] In that report, however, no comparison of the epidermal thickness in FD was made to LPP. Here, we report a distinct difference in epidermal thickness between FD and LPP, thereby confirming the utility of an assessment of epidermal thickness in distinguishing neutrophil-poor FD from LPP (Figure 1).

We performed a retrospective descriptive study using 30 FD and 26 LPP cases. Included cases had a single diagnosis of either LPP or FD, without qualifiers, made by a board-certified dermatopathologist with expertise in alopecic diseases, using histopathologic features and clinical information. Only cases of classic-type LPP with high histopathologic architectural integrity (no torn specimens), that had been initially reviewed within the previous five years, were selected. All cases had at least one 4-mm punch biopsy. The specimens were processed through the horizontal & vertical (HoVert) technique.[7] The slides were scanned into the Philips IntelliSite digital pathology

Bohnett MC et al. J Cutan Pathol 2021;48:816-818

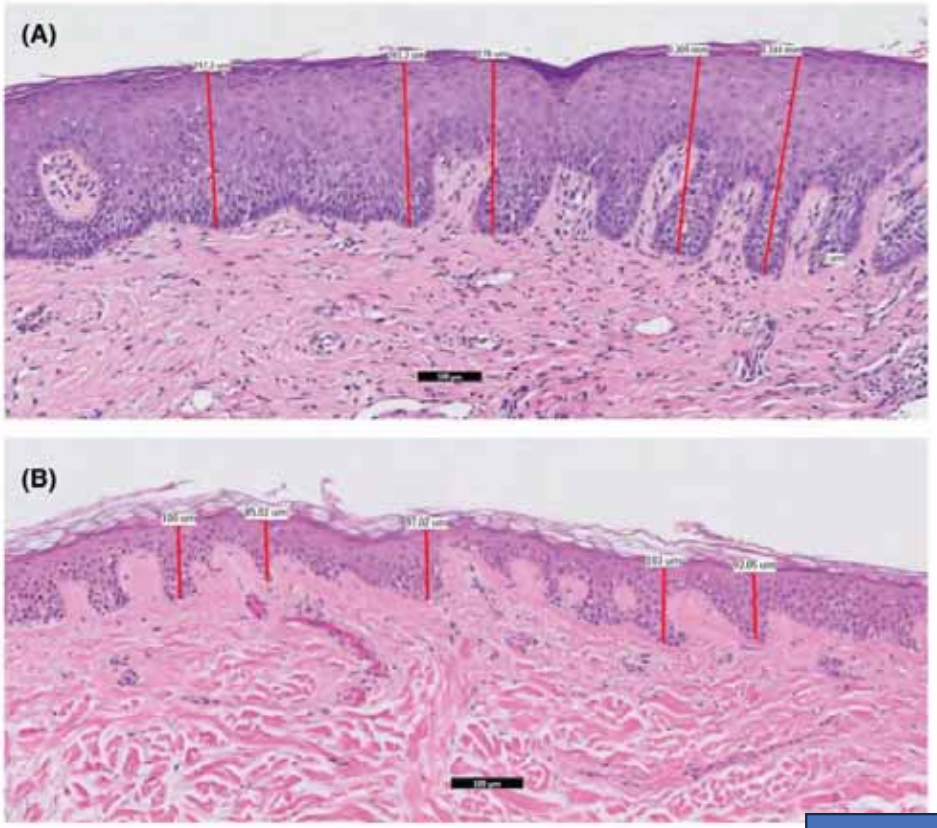


FIGURE 1 Comparison of epidermal thickness measurements in folliculitis decalvans (top), lichen planopilaris (bottom). The FD sample shows markedly acanthotic and thickened epithelium compared to the thin epithelium of LPP. The measurements (red lines and digital boxes) are made using a Philips IntelliSite digital pathology system. FD, folliculitis decalvans; LPP, lichen planopilaris

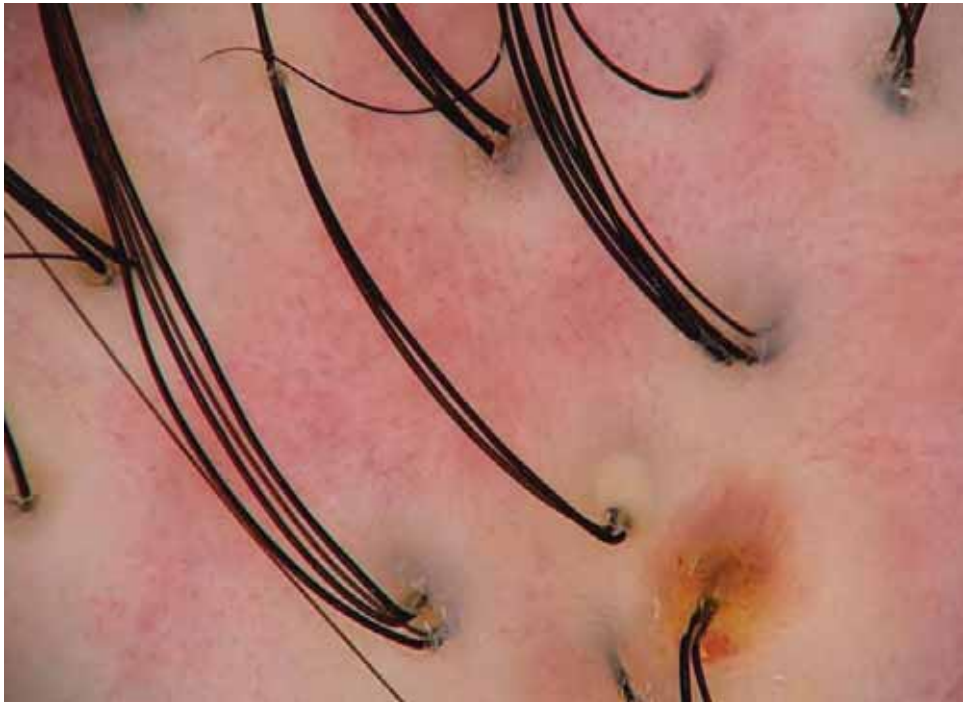
TABLE 1 Mean epidermal thickness of folliculitis decalvans (FD) vs lichen planopilaris (LPP) showing that the epidermis in FD is significantly thicker than in LPP

	Folliculitis decalvans	Lichen planopilaris
Number of cases reviewed (N)	30	26
Mean epidermal thickness (μm)	264.2	133.3
95% confidence interval	246.0-282.4	126.9-139.7
SD	±17.7	±6.6
SD of variance (σ)	105.1	39.4
P value	$P < 0.0001$	

50-year old man. Confluent scarring alopecia at the vertex with peripheral active erythema with crusting, pustules and follicular tufting

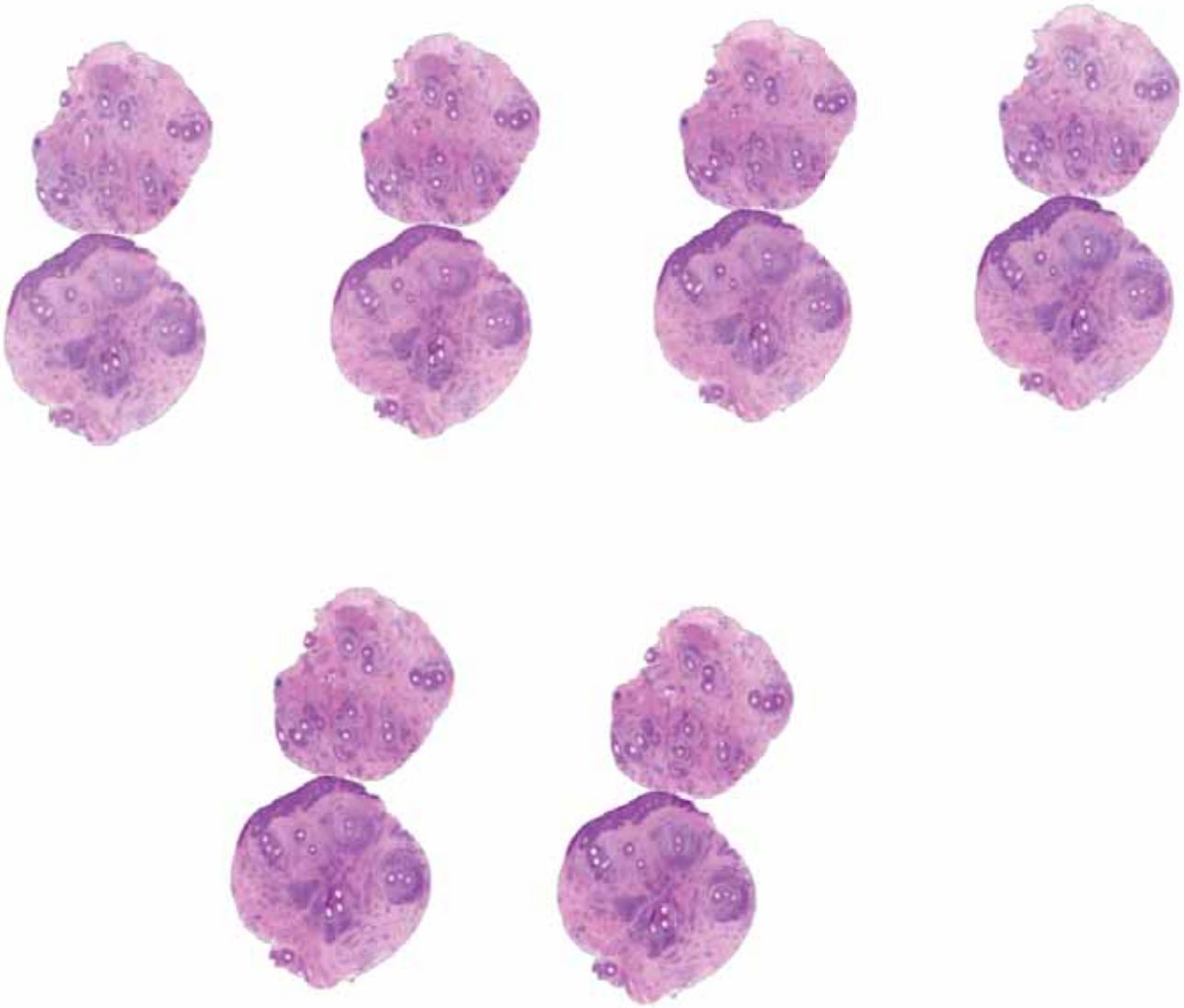
ISDP 2025 ORLANDO

TRICHOSCOPY

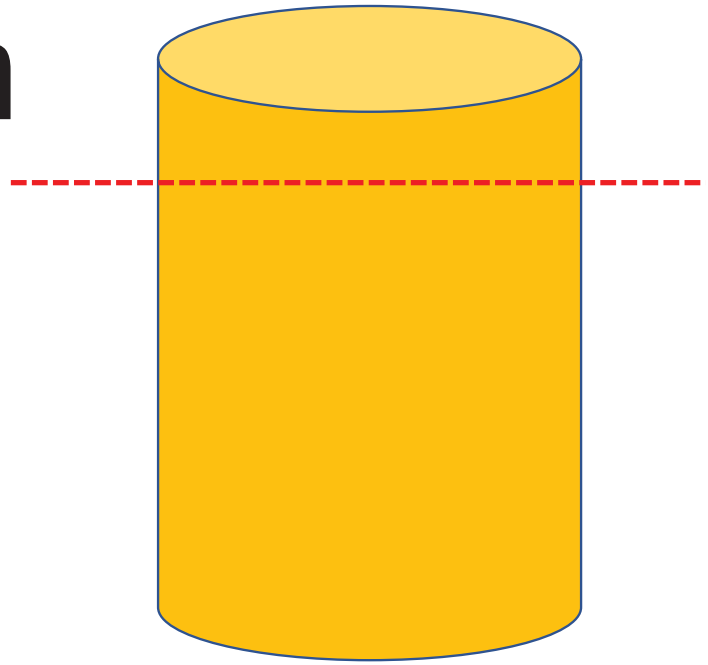


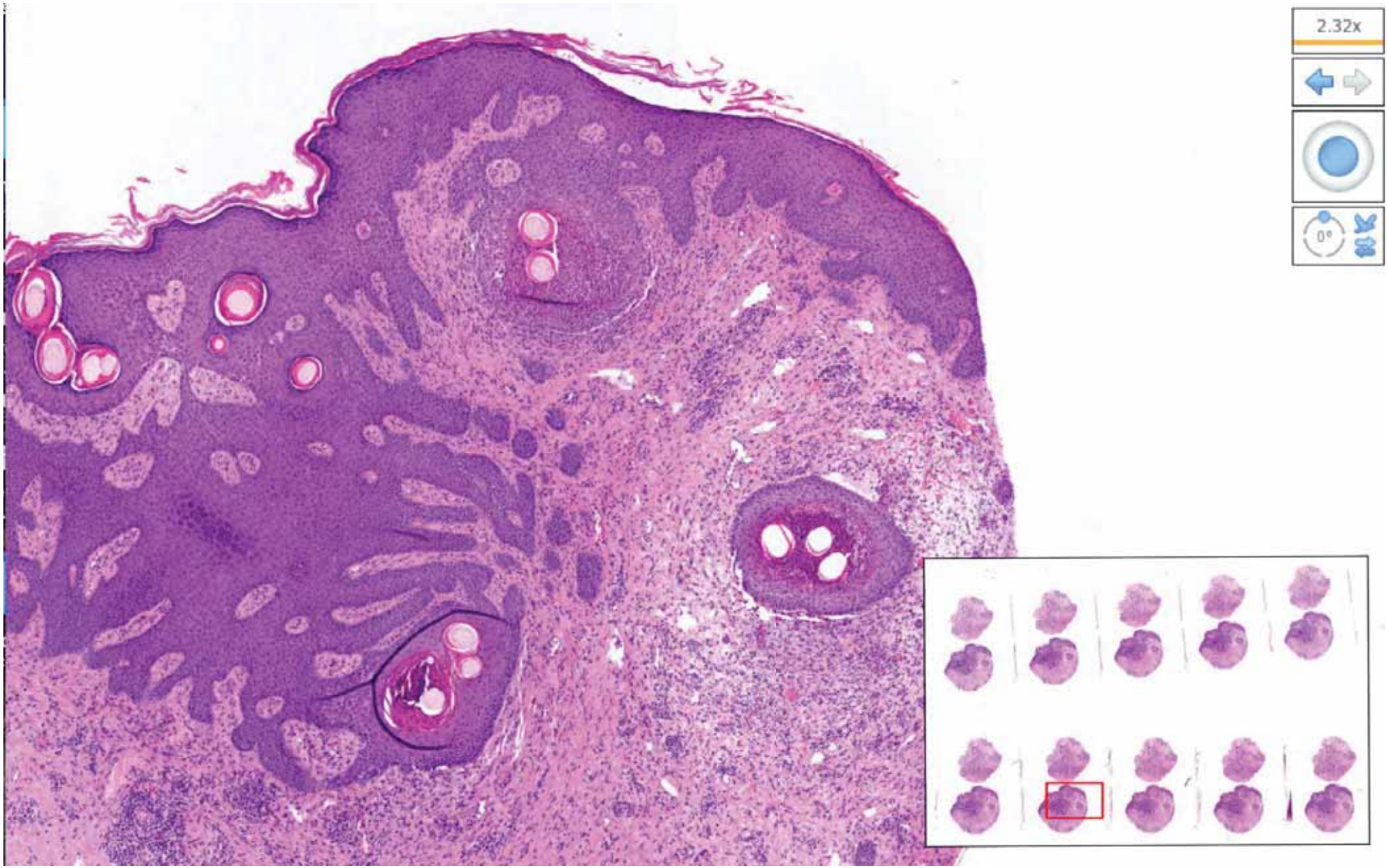
Pilar casts, pustules dilated blood vessels

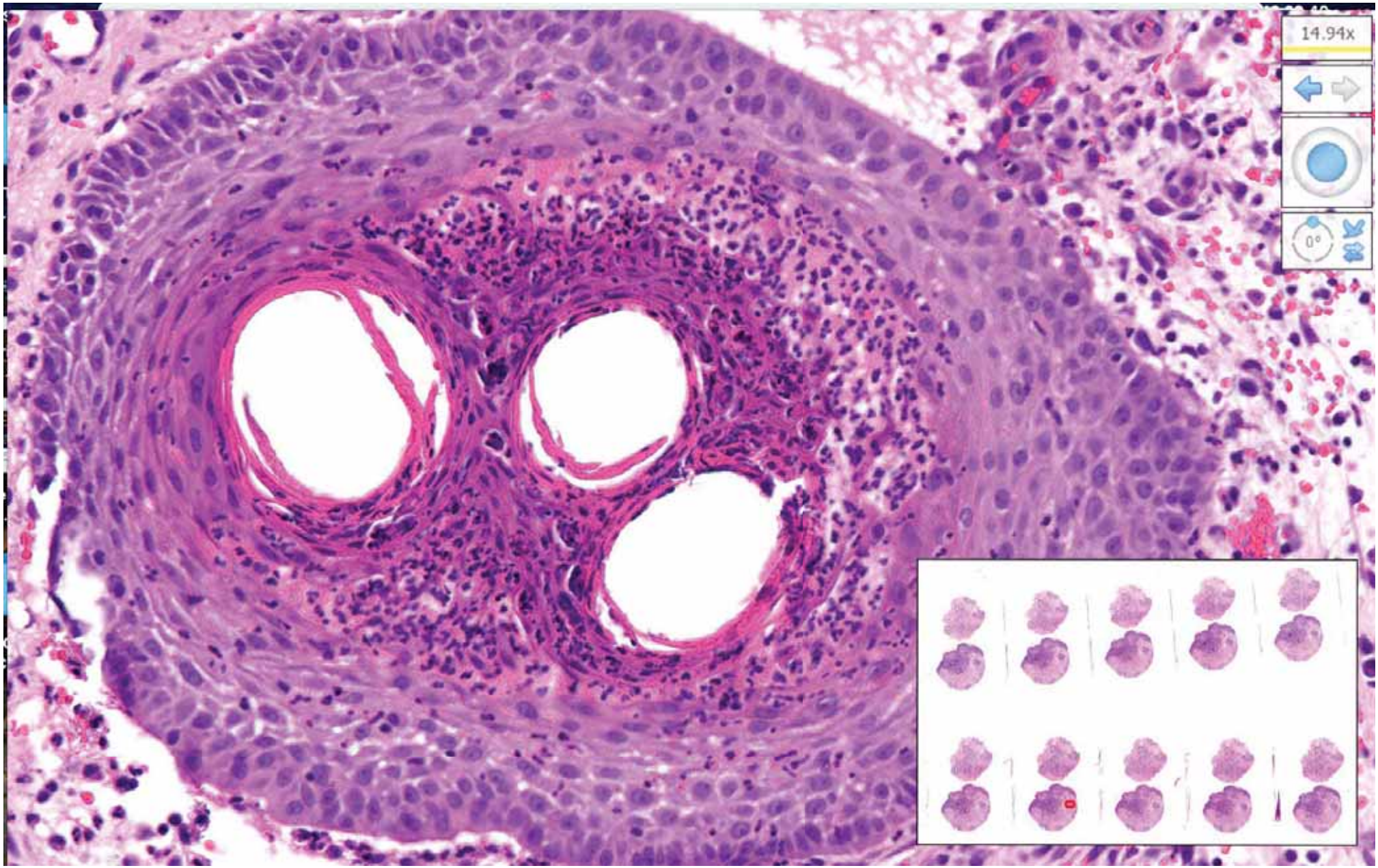
Zhang et al. Clin Cosmet and Investig Dermatol 2022:15 993–996

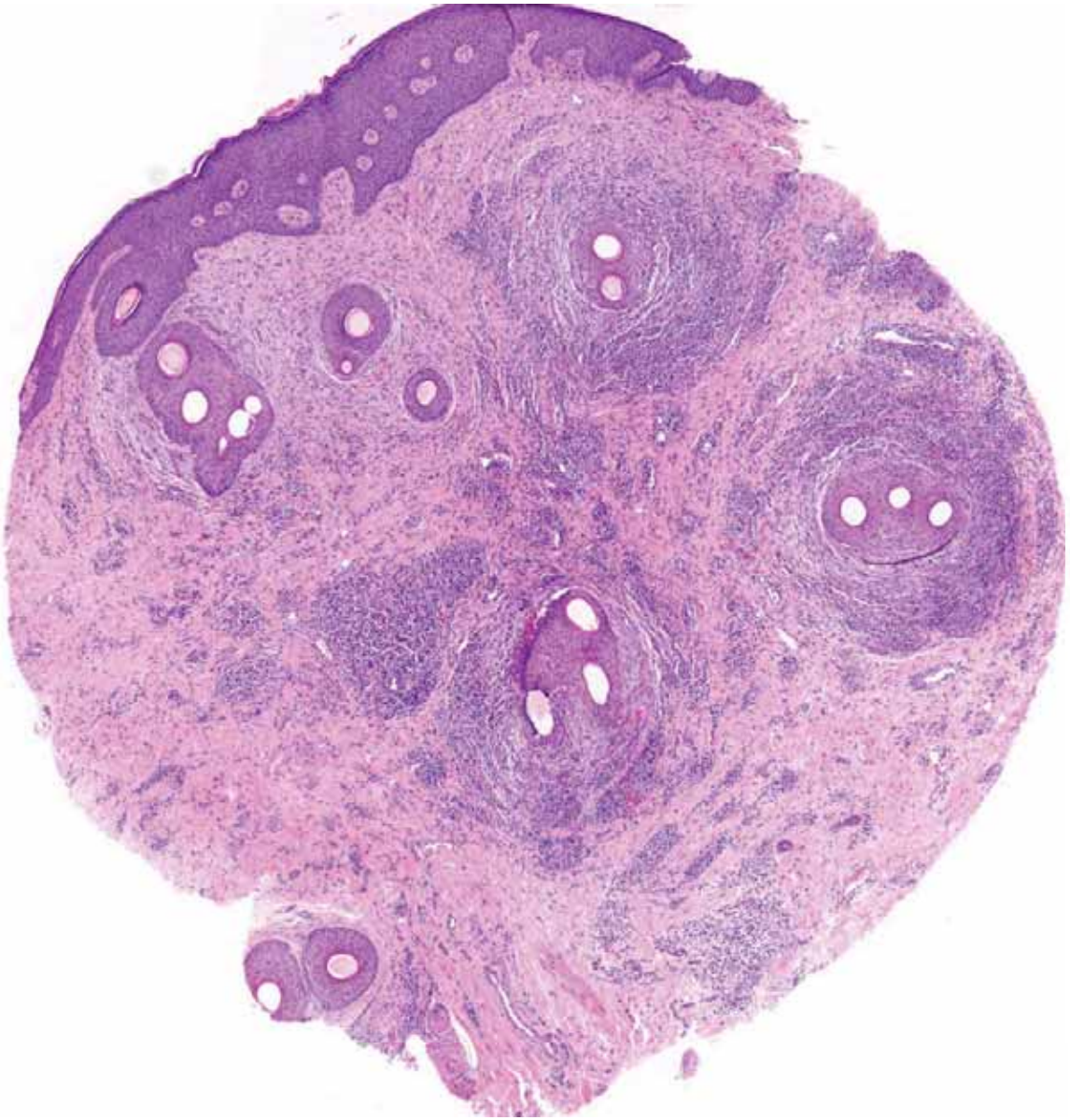


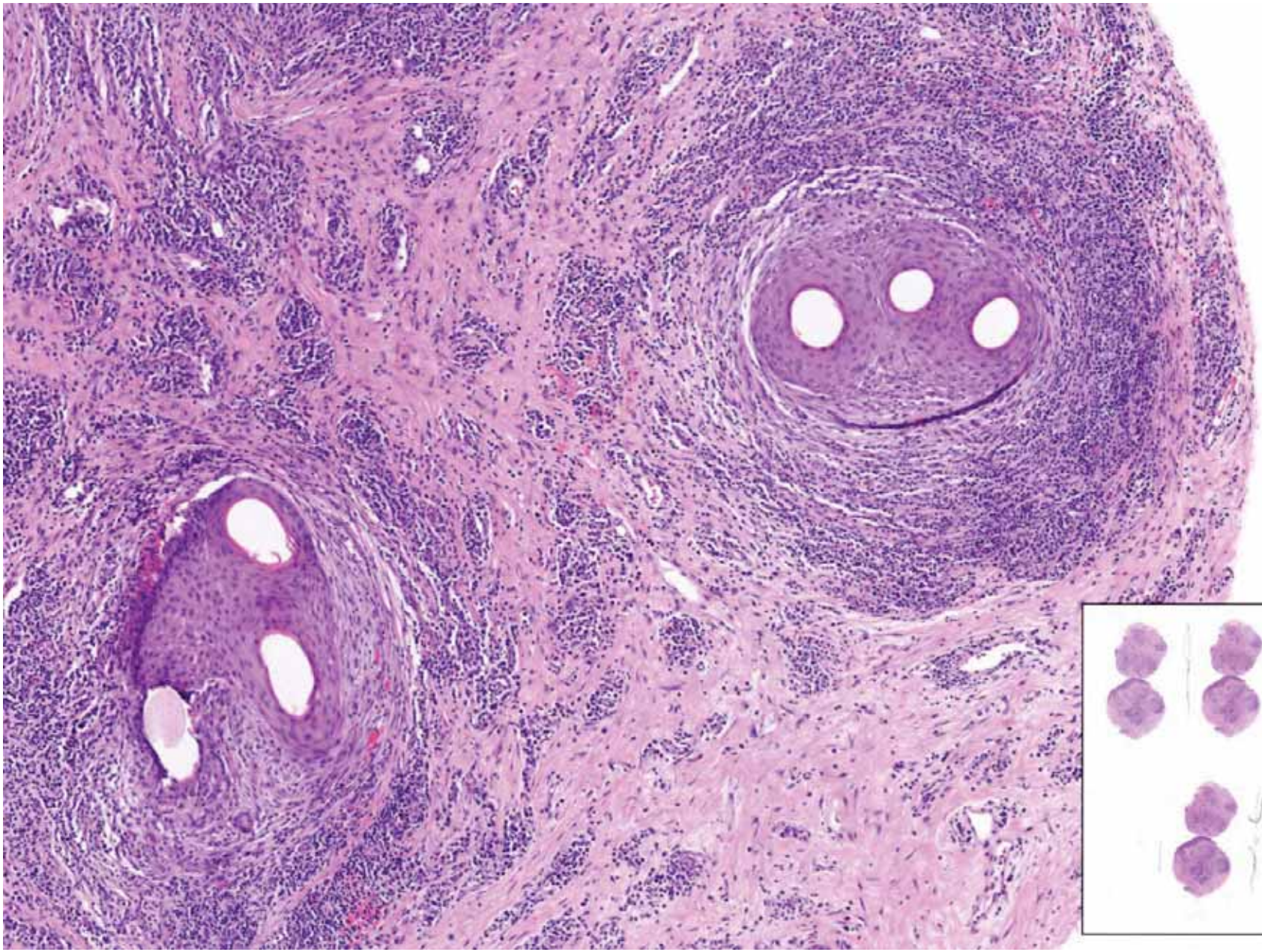
Supra-isthmus/ infundibulum

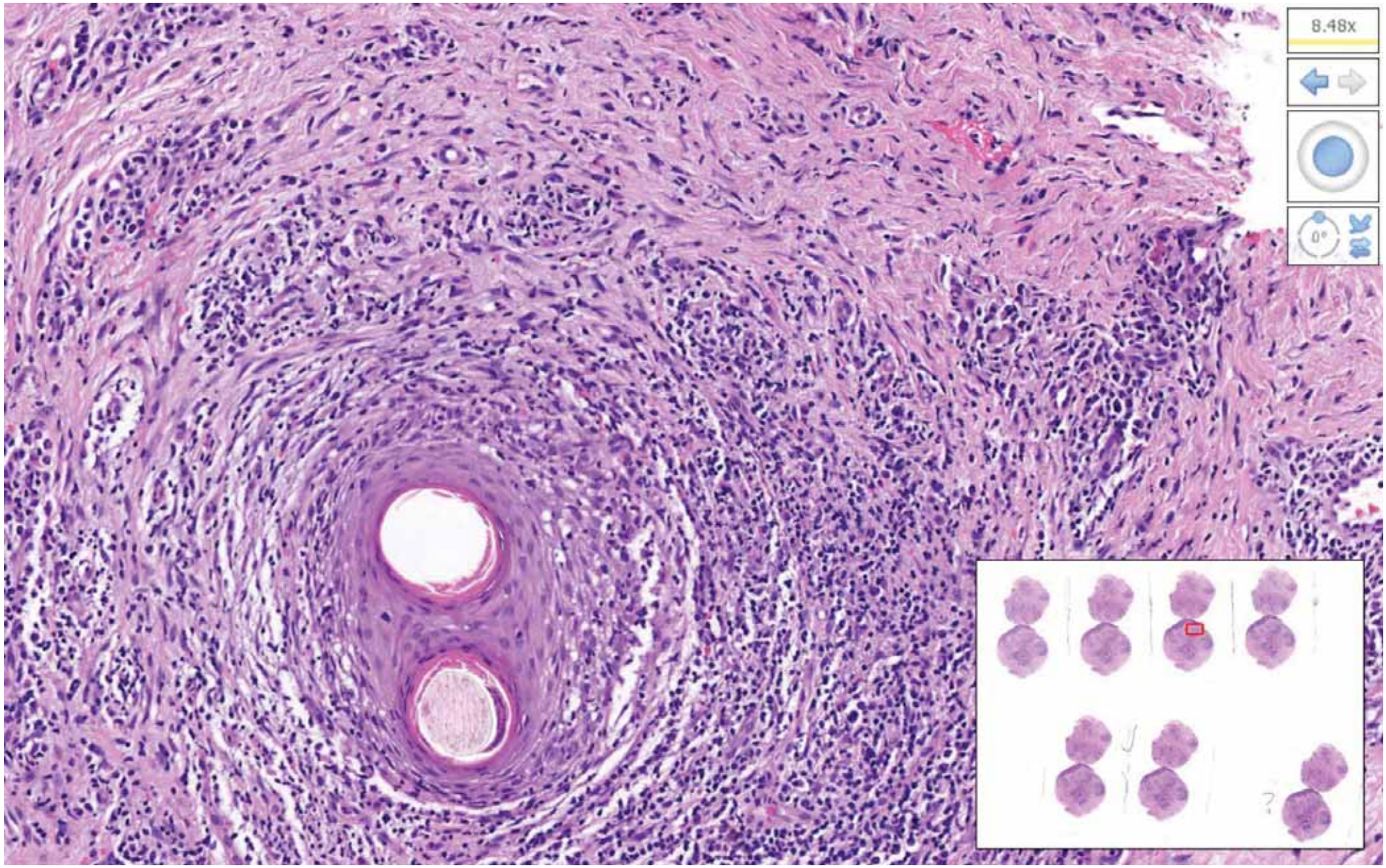


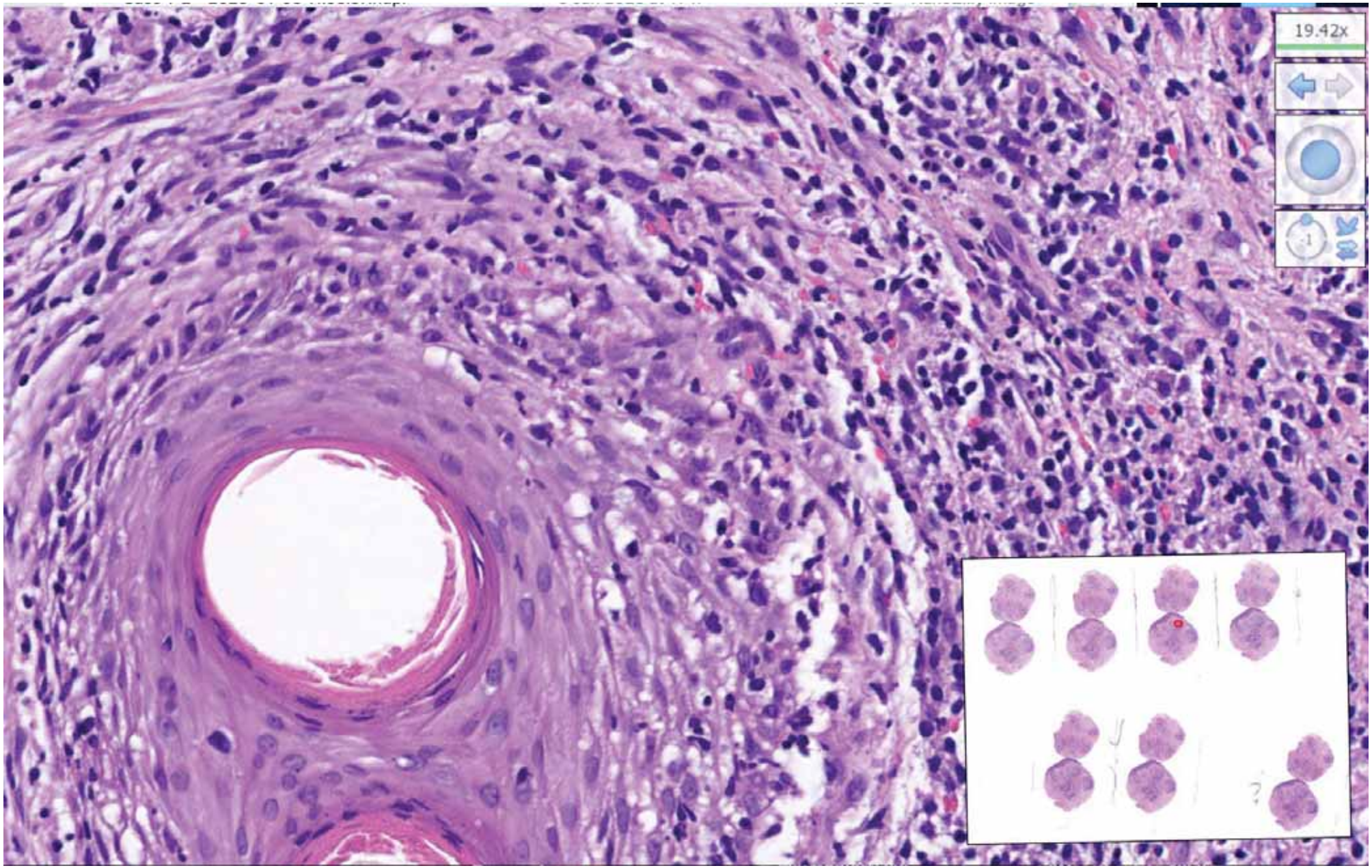

2.32x
0°


14.94x
0°

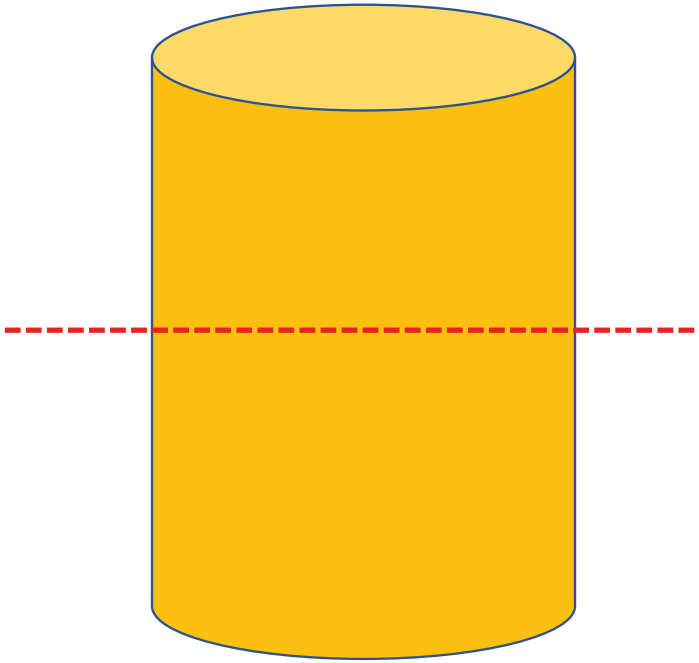


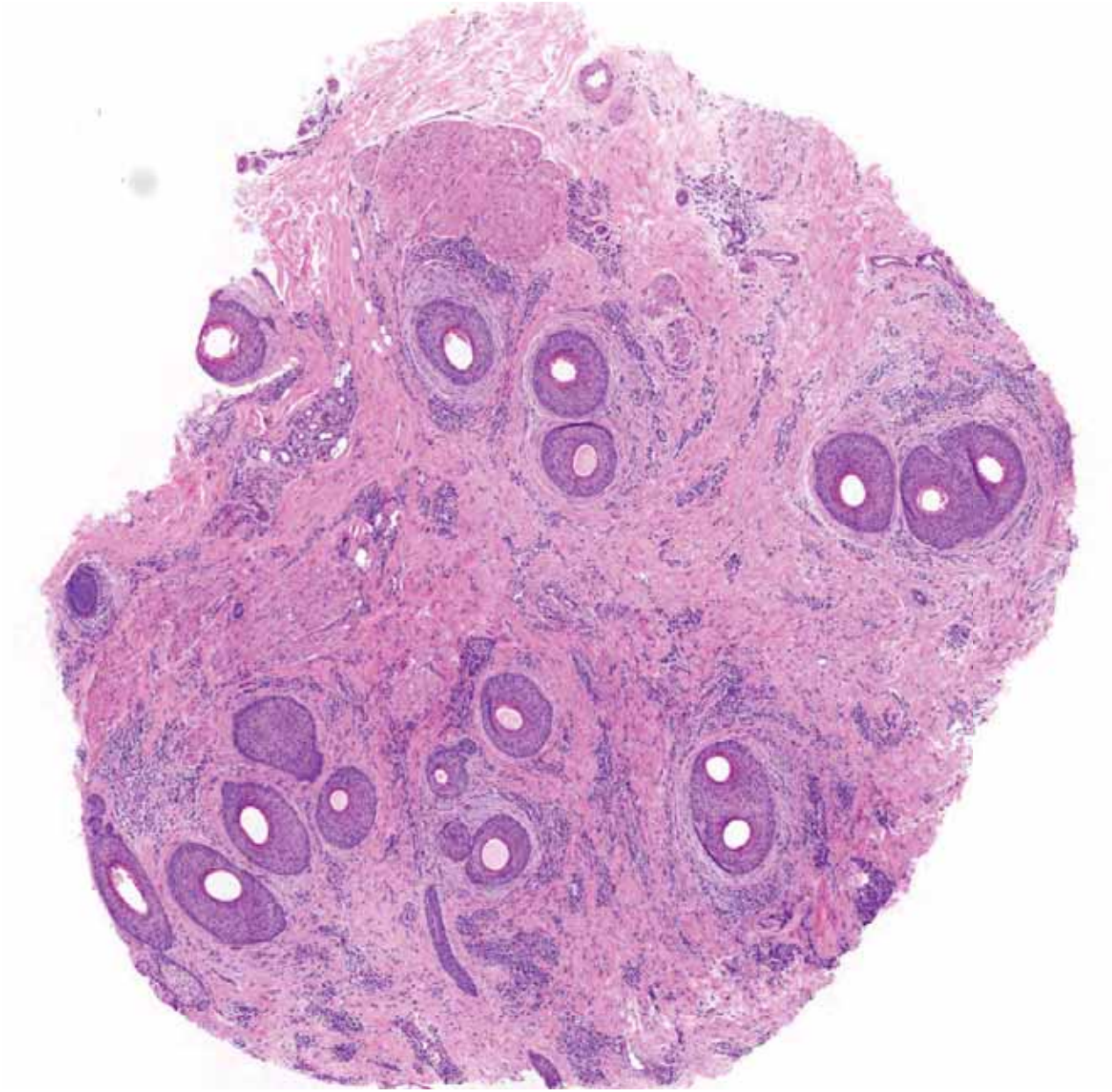


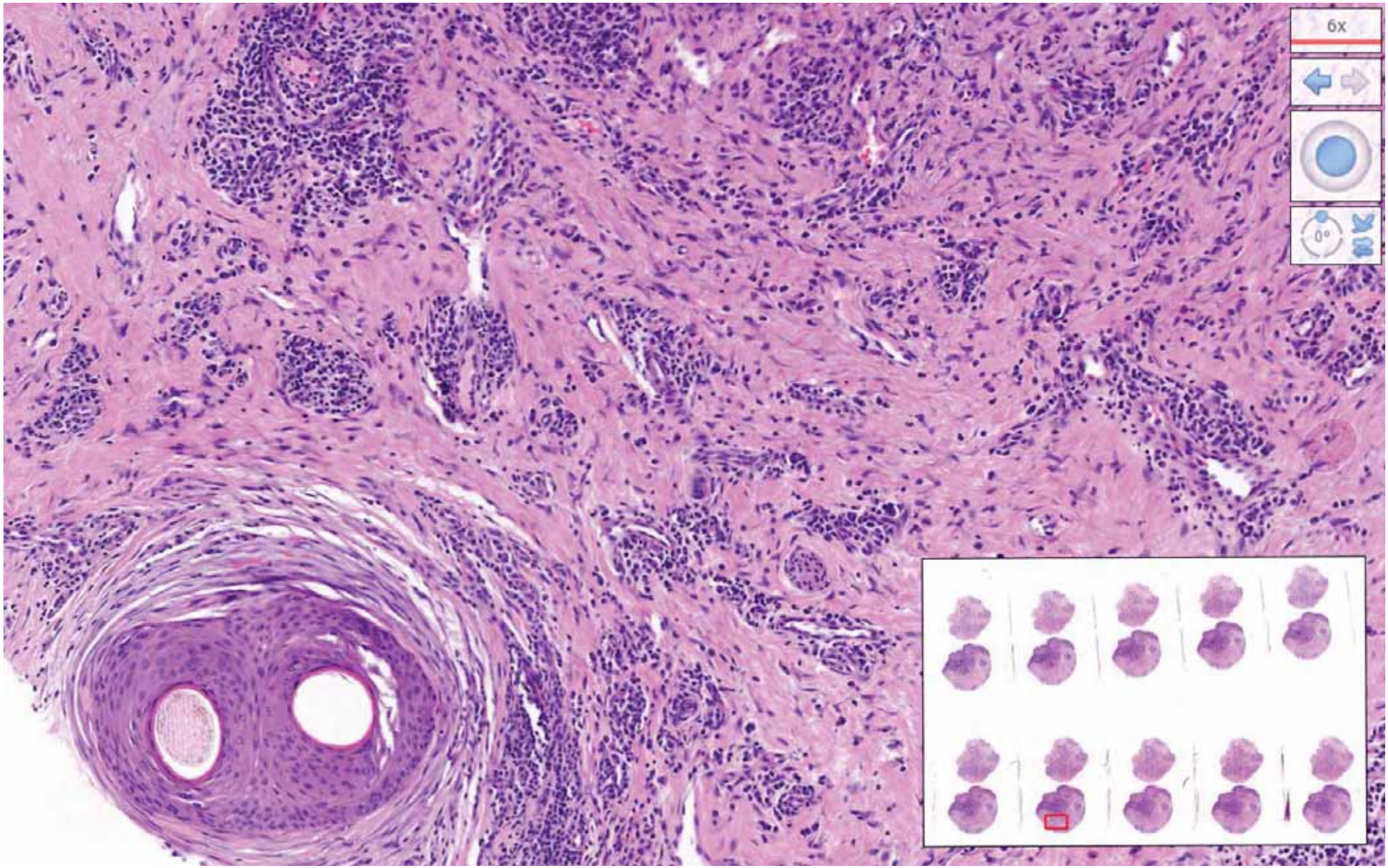

8.48x
0°

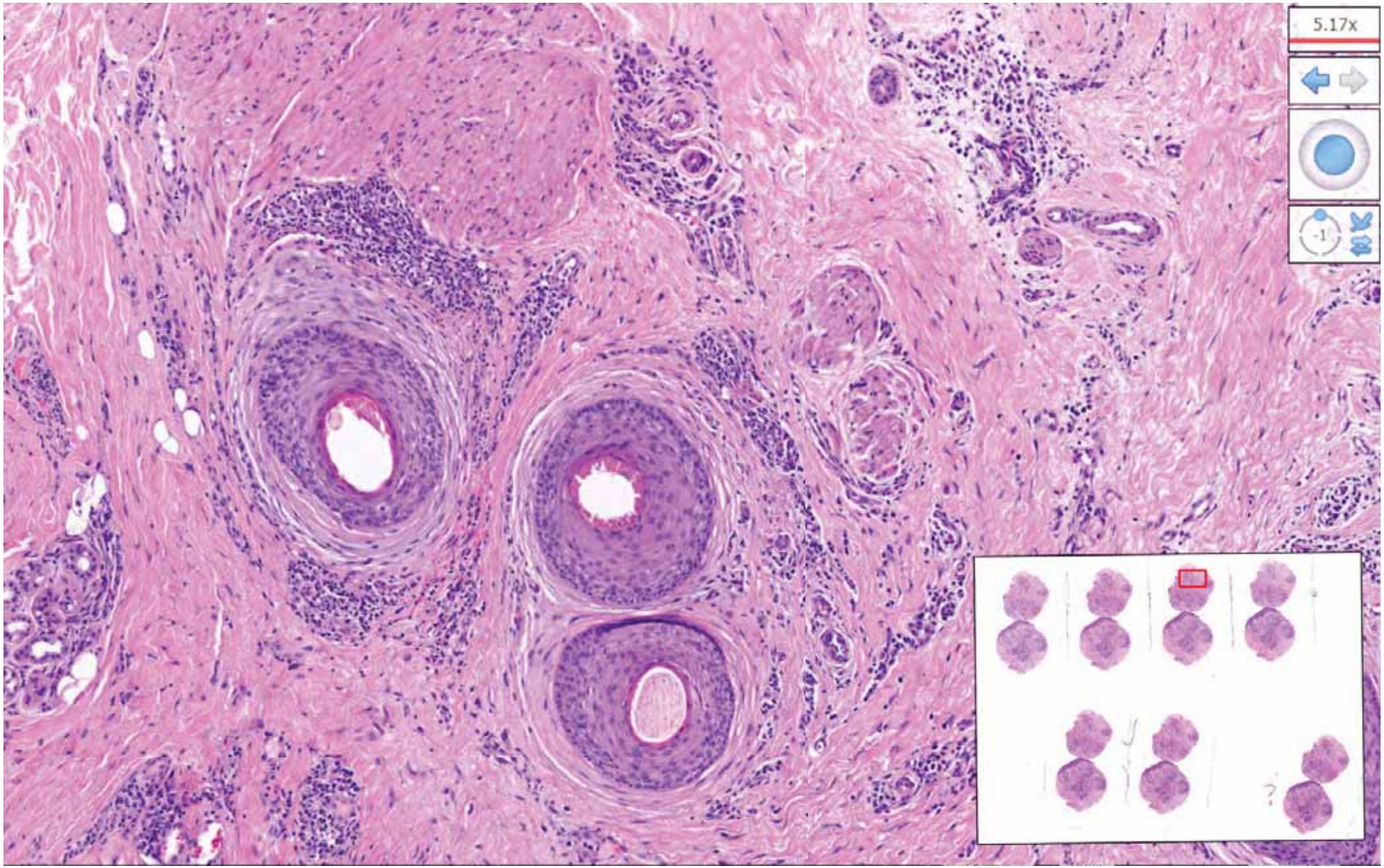

19.42x

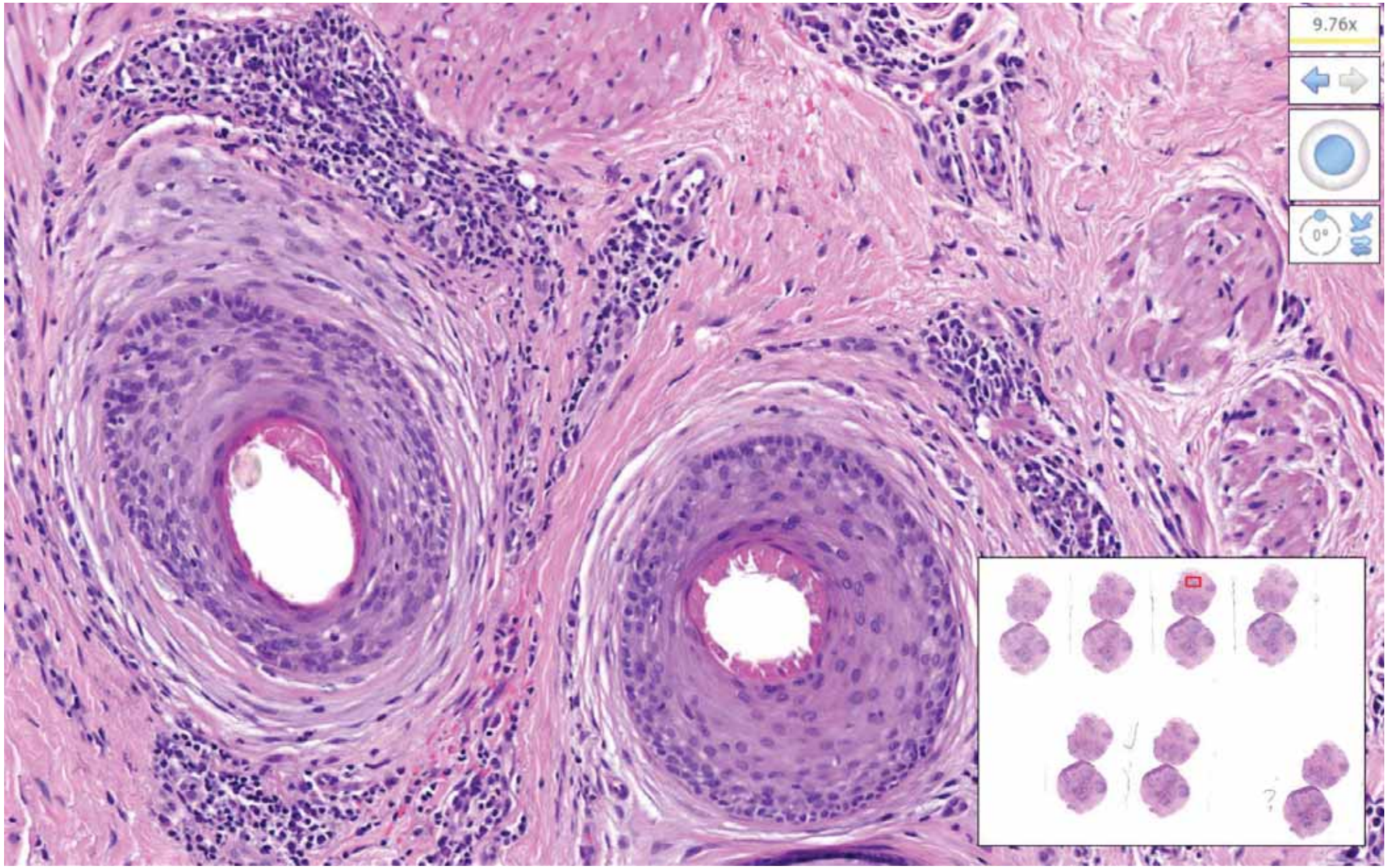
Isthmus



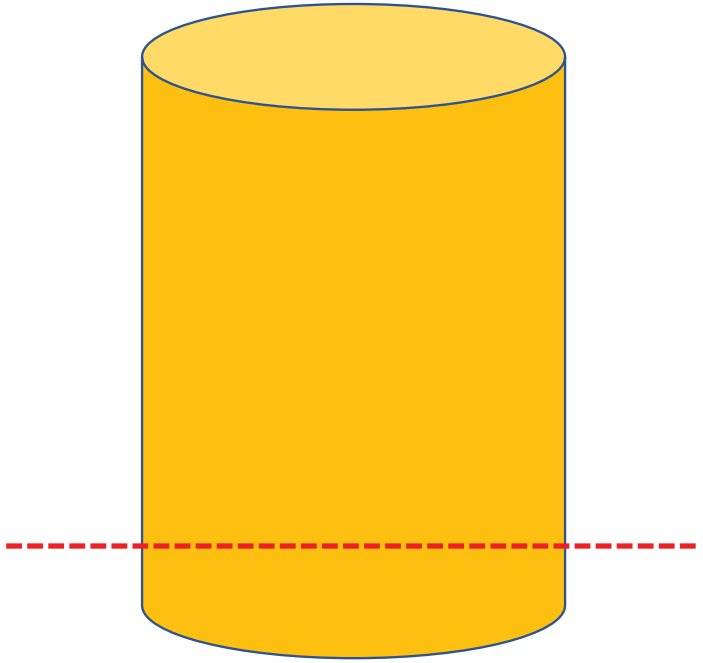


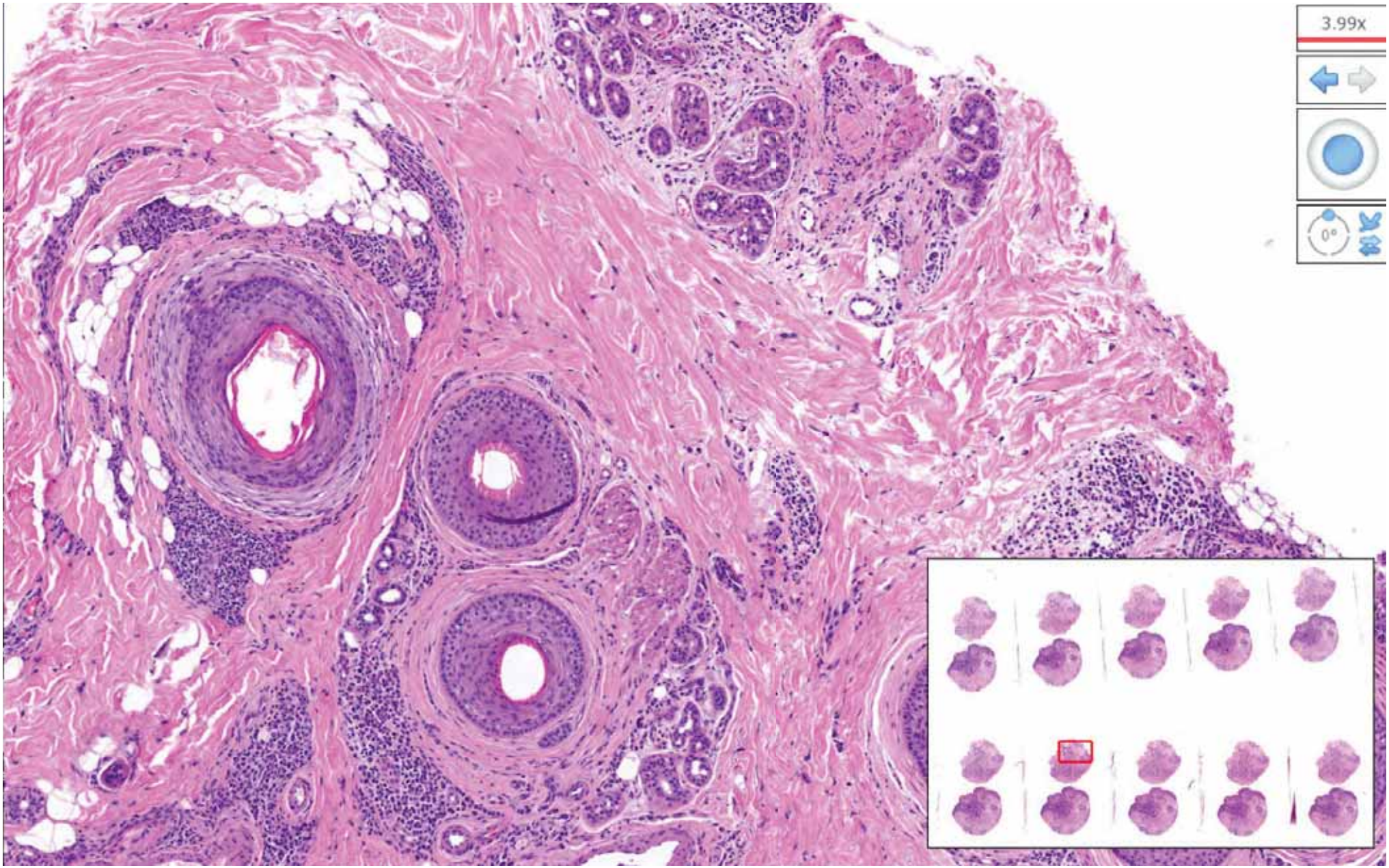

6x
0°

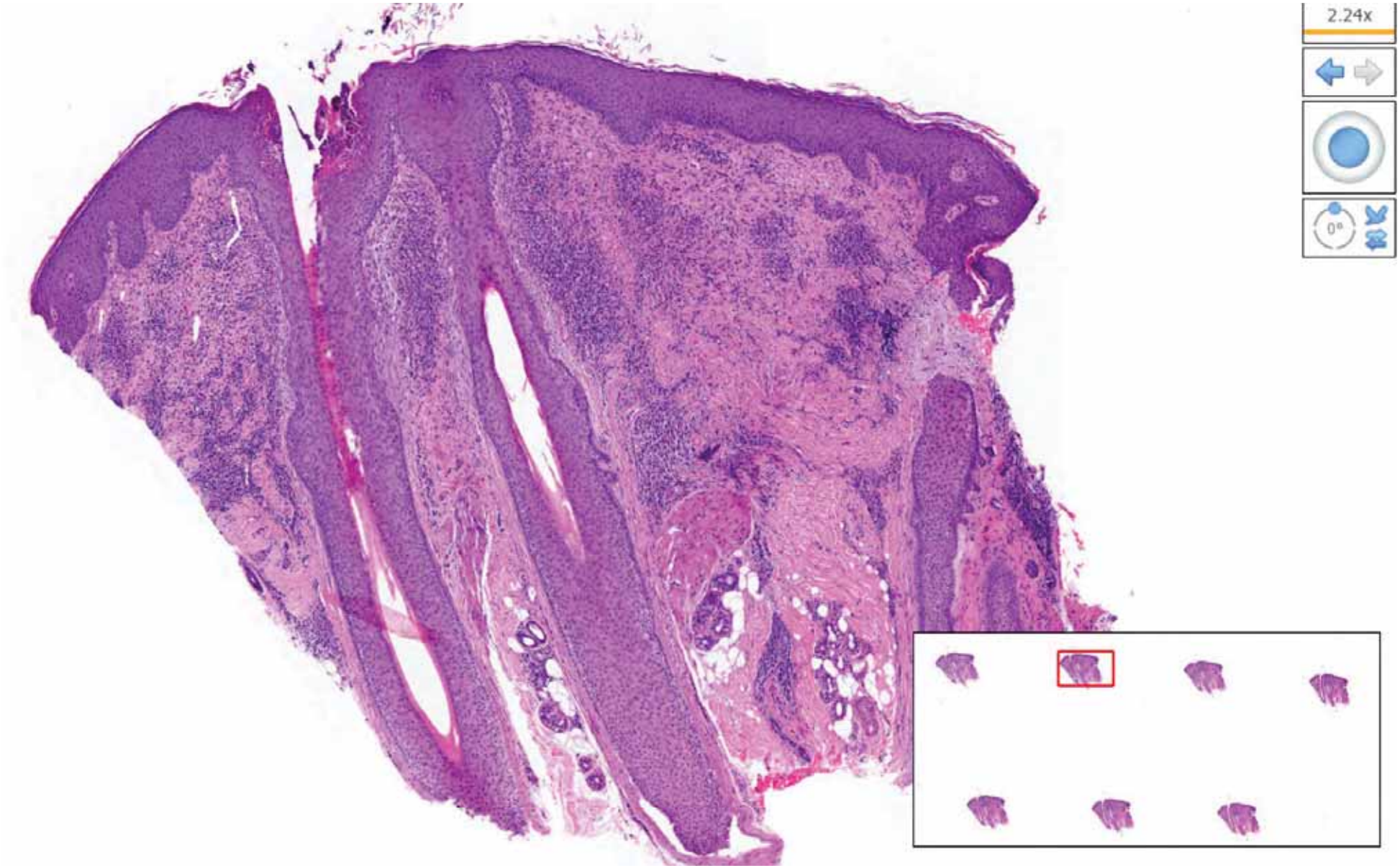

5.17x
-1

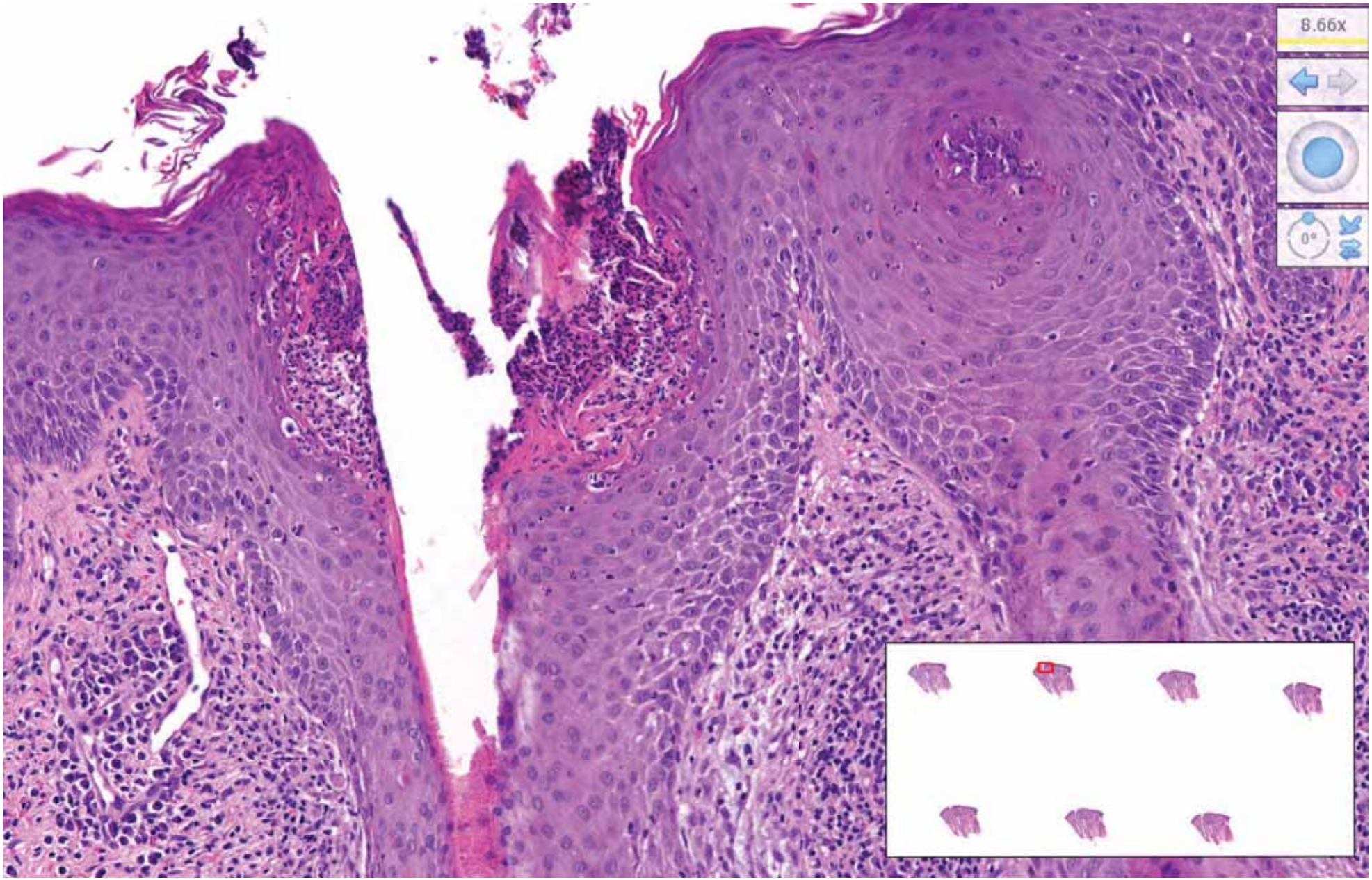

9.76x
0°

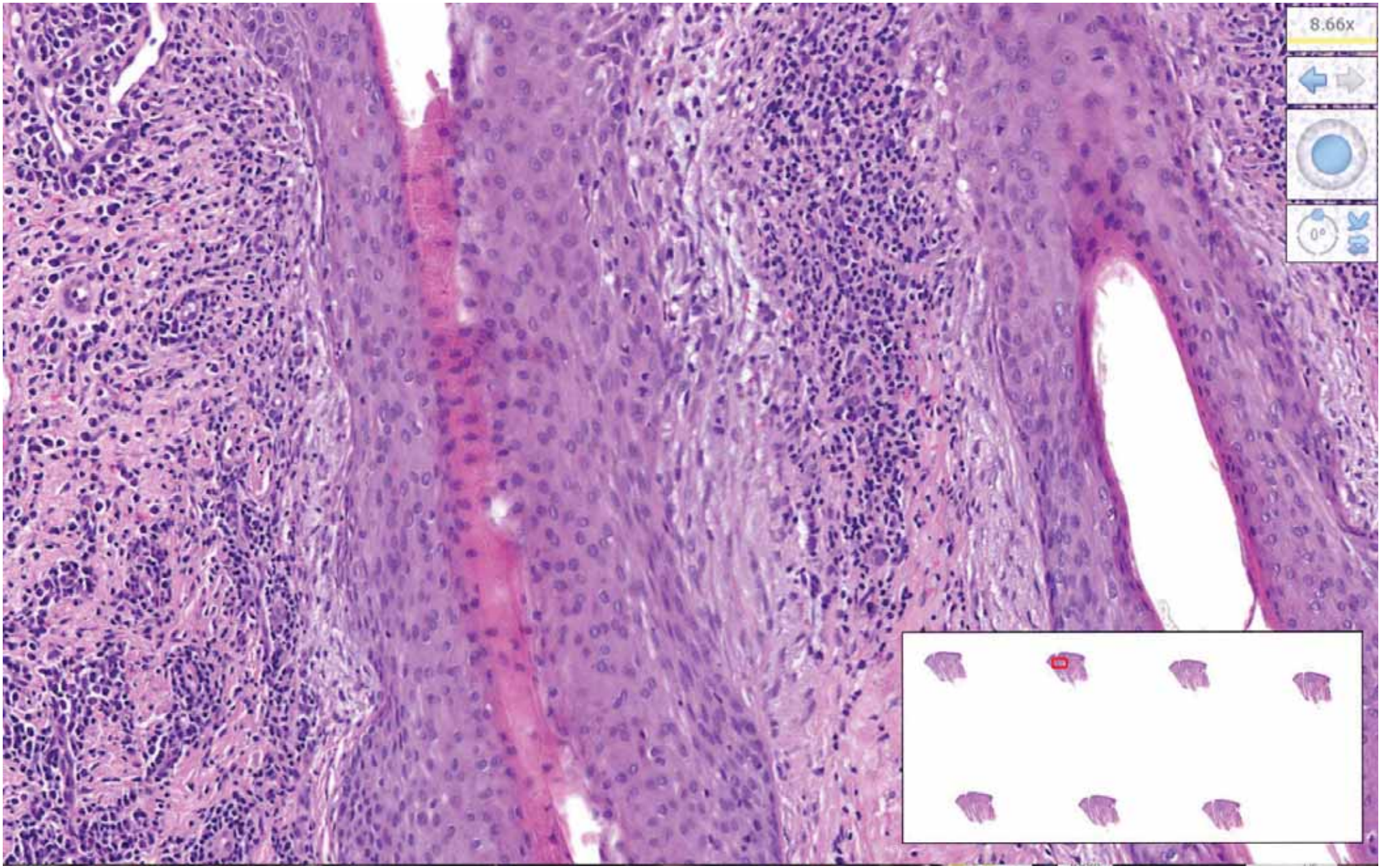
Sub-isthmus

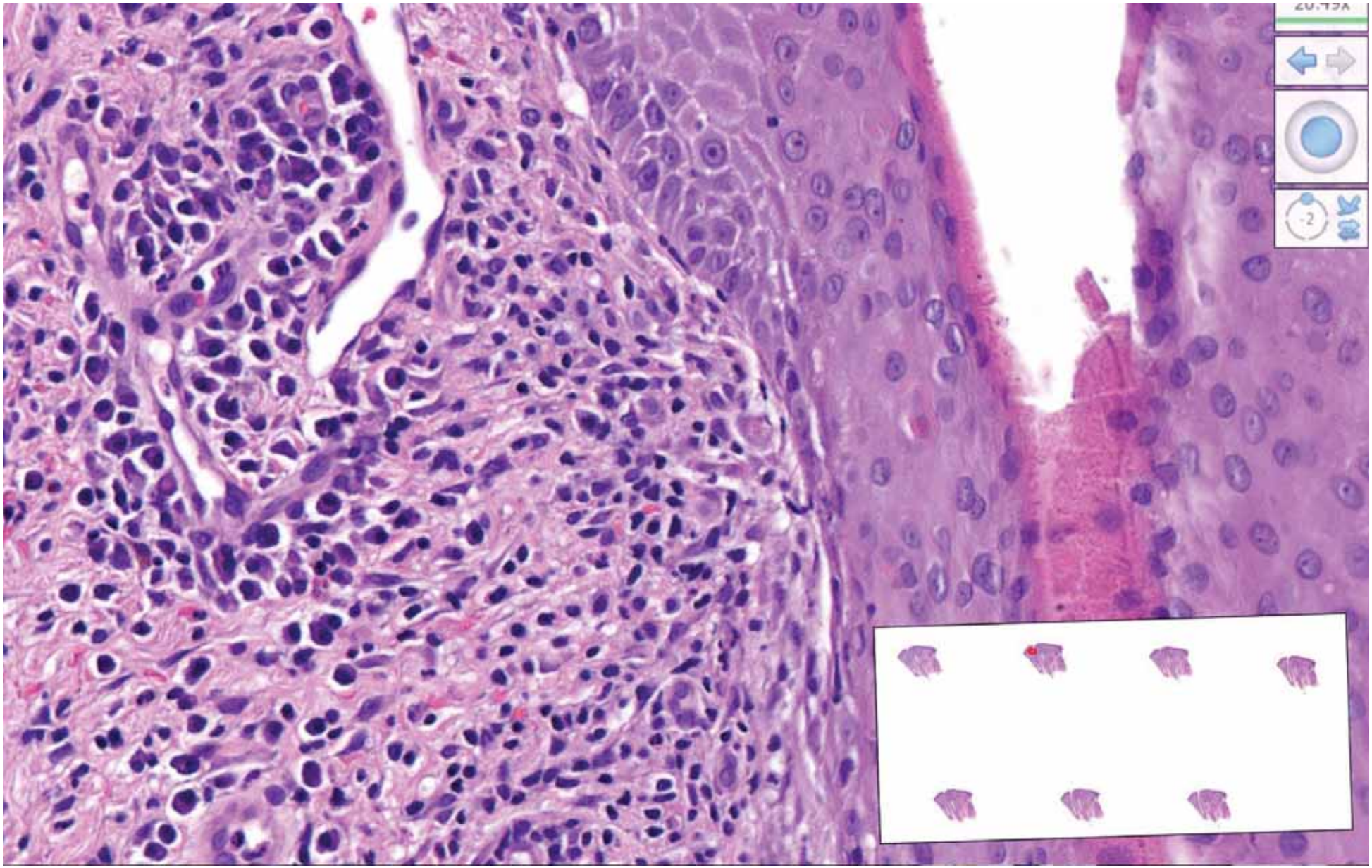



3.99x
0°

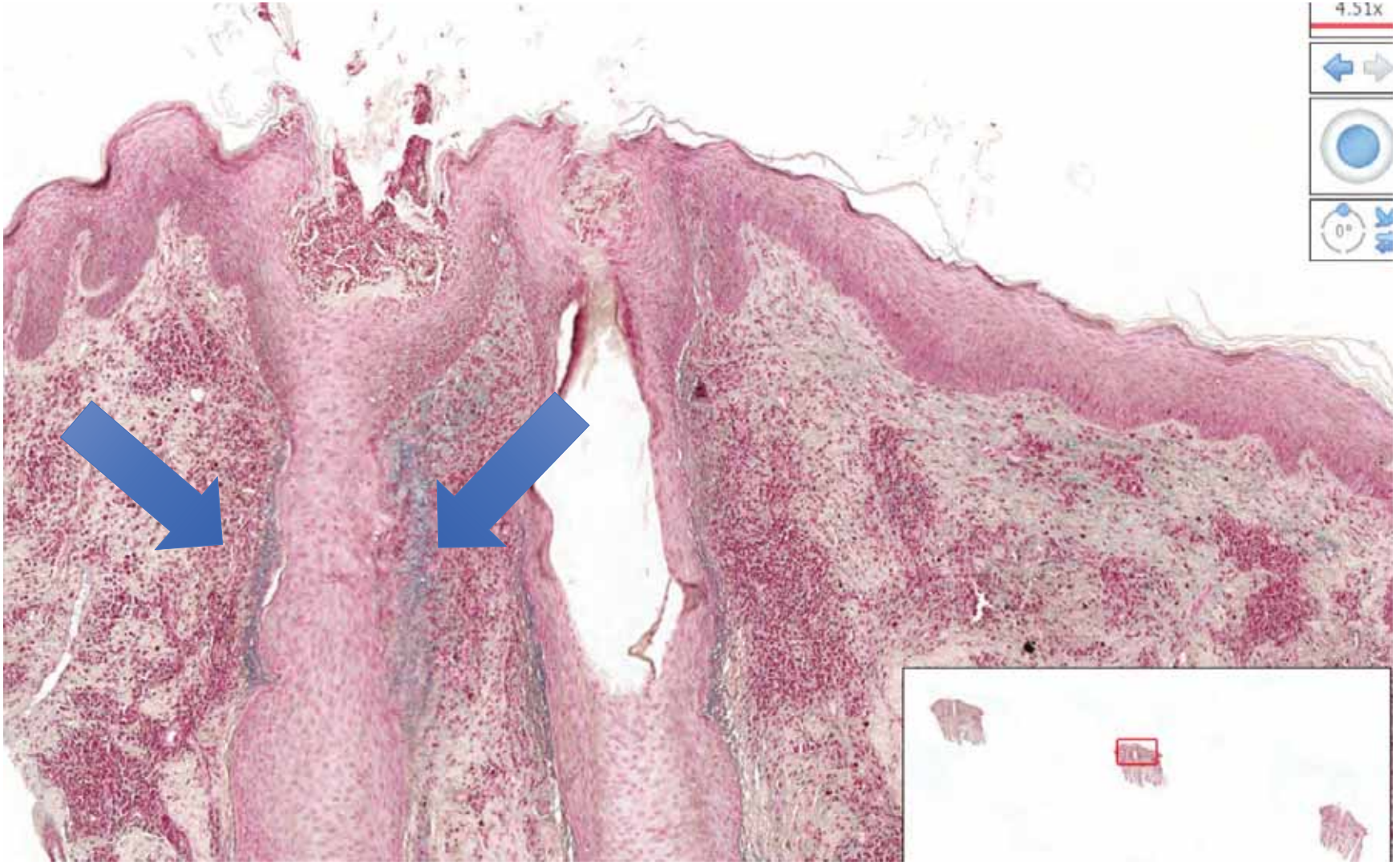

2.24x
0°


8.66x
0°


8.66x
0°



Colloidal iron



Folliculitis Decalvans and Lichen Planopilaris Phenotypic Spectrum—A Series of 7 New Cases With Focus on Histopathology

Andjela Egger, BS, Olivera Stojadinovic, MD, and Mariya Miteva, MD

Background: Folliculitis decalvans (FD) and lichen planopilaris (LPP) are classified as neutrophilic and lymphocytic cicatricial alopecias according to the North American Hair Research Society. Recently, a clinical phenotype combining concomitant or sequential features for both was described as a FD LPP phenotypic spectrum (FDLPPPS).

Objectives: To review the most common phenotypic presentation of FDLPPPS with a main focus on histopathology.

Methods: We reviewed retrospectively series of 7 patients with a similar phenotypic presentation with special focus on the histologic pattern. All patients presented with concomitant features for FD and LPP and recalcitrant course unresponsive to topical and systemic immunomodulatory/anti-inflammatory agents.

Results: The most common clinical phenotype was that of hairless patches on the vertex with lost follicular ostia and perifollicular scale and the following diagnostic findings: (1) polytrichia; (2) positive bacterial culture for *Staphylococcus* in over 50% of the samples isolated from pustules and hemorrhagic crusts; (3) "mixed" histologic features for primary cicatricial alopecia including multicompound follicular structures of average 2–5 follicles (follicular packs), atrophy of the follicular epithelium, lymphohistiocytic infiltrate with granulomas, and prominent plasma cells, but absence of neutrophilic infiltrate in all cases except scarce neutrophils in one; and (4) clinical improvement with adjuvant systemic antimicrobials.

Conclusions: The FDLPPPS may be underreported and should be considered in all cases of LPP recalcitrant to treatment. Dermatologists and dermatopathologists should recognize this phenotypic spectrum to guide optimal clinical management consisting of immunomodulatory and anti-inflammatory agents along with systemic antimicrobials.

Key Words: scarring alopecia, hair loss, hair pathology, scalp biopsy, trichoscopy, alopecia

(*Am J Dermatopathol* 2019;00:1–5)

INTRODUCTION

The North American Hair Research Society's classification of the primary cicatricial alopecias (PCA) is based on predominance of the primary cell-type infiltrate on histopathology.[1] According to this classification, folliculitis decalvans (FD) and lichen planopilaris (LPP) are characterized by neutrophilic and lymphocytic infiltrates, respectively, and are listed under 2 distinct categories—"neutrophilic" and "lymphocytic" PCAs. Neither is mentioned under the third category—"mixed."

However, Morais et al described a series of 13 patients diagnosed with "LPP with pustules" based on the clinical presentation of perifollicular scaling (12/13), erythema, crusts (11/13), follicular tufts (10/13), and pustules (13/13). On histology, however, only 7/13 cases showed lichenoid perifolliculitis, but 10/13 showed neutrophilic folliculitis and polytrichia (8/13). Yip et al[2] recently reported 13 patients with FD and LPP phenotypic spectrum (PS) (FDLPPPS). The clinical presentation was consistent either with (1) initial FD features (such as tufting, pustules, and scarring) that transitioned into LPP features (perifollicular erythema, perifollicular casts, and scarring) over time or (2) concomitant features for both. Biopsies from the FD areas showed neutrophilic cicatricial alopecia, whereas the concomitant/sequential LPP areas showed lymphocytic inflammation with perifollicular fibrosis.

We have also encountered the difficulty of stratifying the diagnosis in patients with mixed FD and LPP features, and particularly in those showing resistance to treatment with topical and systemic steroids, and/or other immunomodulatory and anti-inflammatory regimens. The recent report by Yip et al prompted us to review the clinical and histologic features from 7 patients with a similar phenotypic presentation focusing specifically on the histologic findings.

MATERIALS AND METHODS

Folliculitis Decalvans and Lichen planopilaris phenotypic spectrum (FDLPPPS)

- **Mixed scarring alopecia**, combining neutrophilic-mediated FD and immune-lymphocytic mediated LPP
- **Clinically**: Initial FD-like (crusts, follicular tufts, pustules, followed by LPP-like features (erythema, perifollicular casts, scarring). Overtime: concomitant features of both
- **Histopathology**: Initially neutrophilic cicatricial alopecia with compound hair follicles and interfollicular scarring, followed by perifollicular lymphocytic infiltrate with perifollicular fibrosis
- **Microbiological signature**: *Staphilococcus aureus* (+) cultures in pustules in over 50% of patients in typical FD and less 20% in FDLPPPS.

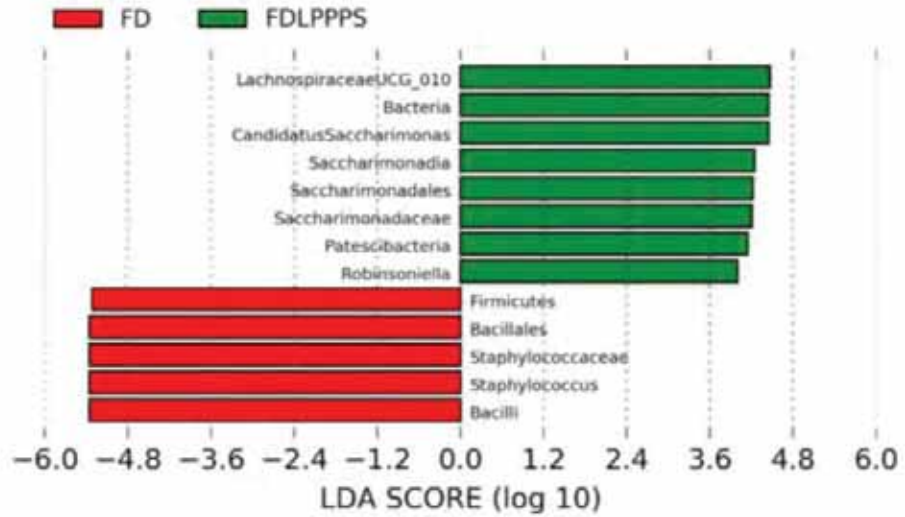


Fig 1. Relative abundance of bacterial taxa between typical folliculitis decalvans (*FD*) and FD and lichen planopilaris phenotypic spectrum (*FDLPPPS*) calculated by the linear discriminant analysis (*LDA*) effect size tool. Each bar represents the $\log_{10}$ effect size (LDA score) for a specific taxon. A longer bar represents a higher LDA score. The *red bars* represent bacterial taxa found to be more abundant in typical neutrophilic FD compared with FDLPPPS and vice versa. For instance, *Staphylococcus* has a high LDA score (more than 5 orders of magnitude), reflecting marked abundance in FD compared with FDLPPPS.

Moreno-Arrones: JAAD 2021;85:1355-1357

significant levels of this bacteria.[1] Moreover, because the *S aureus* bacteria colonizing patients with FD are not more virulent than those present in the general population, it is reasonable to hypothesize that the bacteria could take advantage of an immunologic vulnerability or follicular structural alteration.

Taking into consideration the findings of this study and recent publications, the real impact of *S aureus* on the development of FD may not be as significant as previously thought, and treatment could be changed accordingly: in the more neutrophilic and acute stages of FD, antibiotics should be prioritized, whereas in FDLPPPS, anti-inflammatory drugs could be the first choice.

17). All participants signed an informed consent before the initiation of study procedures.

Correspondence and reprint requests to: Oscar M. Moreno-Arrones, Dermatology Department, Ramon y Cajal Hospital, Carretera Colmenar Viejo km 9.100, 28034 Madrid, Spain

E-mail: o.m.m.arrones@gmail.com

REFERENCES

1. Saceda-Corralo D, Moreno-Arrones OM, Rodrigues-Barata R, et al. Trichoscopy activity scale for folliculitis decalvans. *J Eur Acad Dermatol Venereol*. 2020;34(2):e55-e57.
2. Matard B, Donay JL, Resche-Rigon M, et al. Folliculitis decalvans is characterized by a persistent, abnormal subepidermal microbiota. *Exp Dermatol*. 2020;29(3):295-298.
3. Samrao A, Mirmirani P. Gram-negative infections in patients

Dermatology

Skin Infections – Research Article

Dermatology 2023;239:454–461
DOI: 10.1159/000529301

Received: March 29, 2022
Accepted: January 8, 2023
Published online: January 30, 2023

Folliculitis Decalvans Has a Heterogeneous Microbiological Signature and Impaired Immunological Response

Oscar M. Moreno-Arrones[a, b] Carlota Garcia-Hoz[c] Rosa del Campo[d, e] Garbiñe Roy[f] David Saceda-Corralo[a, b] Juan Jimenez-Cauhe[g] Manuel Ponce-Alonso[d, e] Sergio Serrano-Villar[h] Pedro Jaen[a, b] John Paoli[i, j] Sergio Vano-Galvan[a, b]

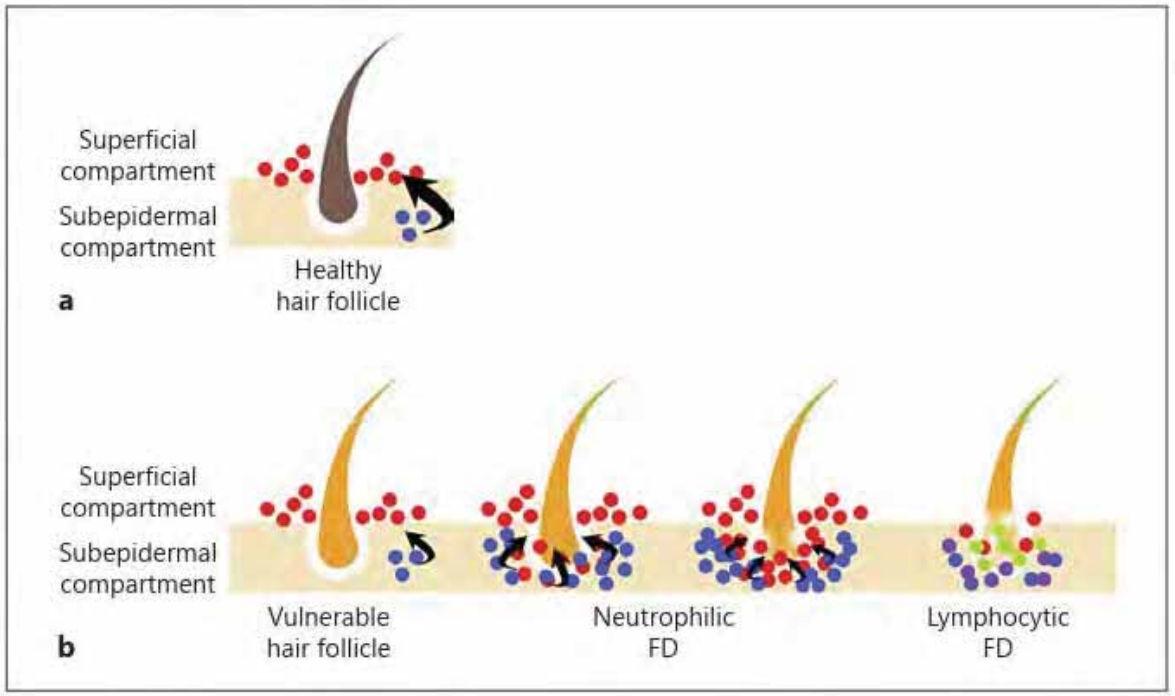
[a]Dermatology Department, Trichology Unit, Ramon y Cajal University Hospital, University of Alcala, Instituto Ramón y Cajal de Investigación Sanitaria (IRYCIS), Madrid, Spain; [b]Trichology and Hair Transplantation Unit, Grupo Pedro Jaen Clinic, Madrid, Spain; [c]Immunology Department, Hospital Clínic de Barcelona, Barcelona, Spain; [d]Microbiology Department, Ramon y Cajal University Hospital, University of Alcala, Instituto Ramón y Cajal de Investigación Sanitaria (IRYCIS), Madrid, Spain; [e]Red Española de Investigación en Patología Infecciosa (REIPI), Madrid, Spain; [f]Immunology Department, Ramon y Cajal University Hospital, Madrid, Spain; [g]Dermatology Department, Trichology Unit, Ramon y Cajal University Hospital, University of Alcala, Instituto Ramón y Cajal de Investigación Sanitaria (IRYCIS), Madrid, Spain; [h]Department of Infectious Diseases Ramon y Cajal University Hospital, University of Alcala, Instituto Ramón y Cajal de Investigación Sanitaria (IRYCIS), Madrid, Spain; [i]Department of Dermatology and Venereology, Institute of Clinical Sciences, Sahlgrenska Academy, University of Gothenburg, Gothenburg, Sweden; [j]Department of Dermatology and Venereology, Region Västra Götaland, Sahlgrenska University Hospital, Gothenburg, Sweden

PATHOGENESIS of FDLPPPS

1. Persistent microbiota dysbiosis of *Staph aureus* burden after anti-staphylococcal antibiotic treatment

2. Collapse hair follicles immune privilege triggering Th1-mediated inflammation

3. Features of neutrophilic FD (early) or lymphocytic LPP (late) at different stages



Dermatology 2023;239:454–461

‘DECODING’ SCARRING ALOPECIA CLINICALLY MIMICKING ALOPECIA TOTALIS/ UNIVERSALIS

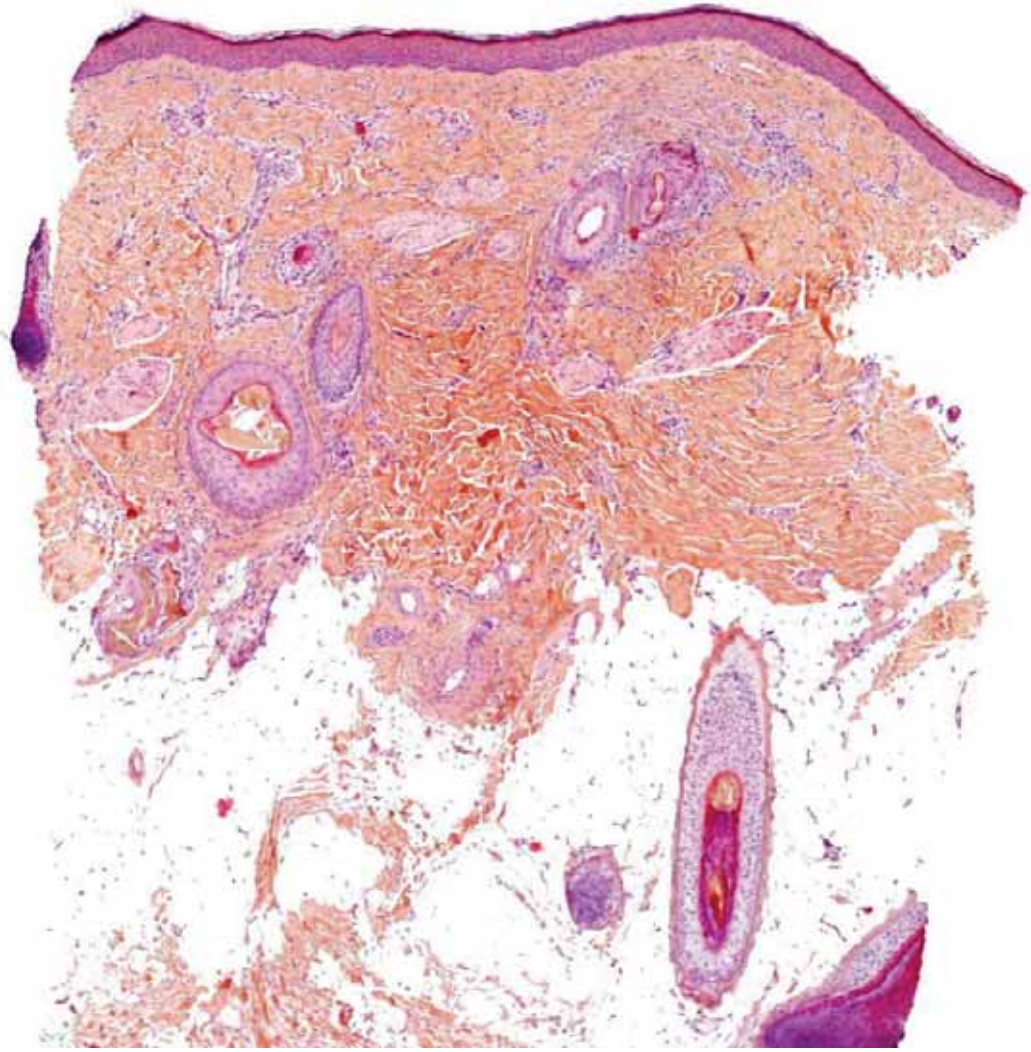
Consultation from Dr Stephanie Leclerc-Mercier, PARIS

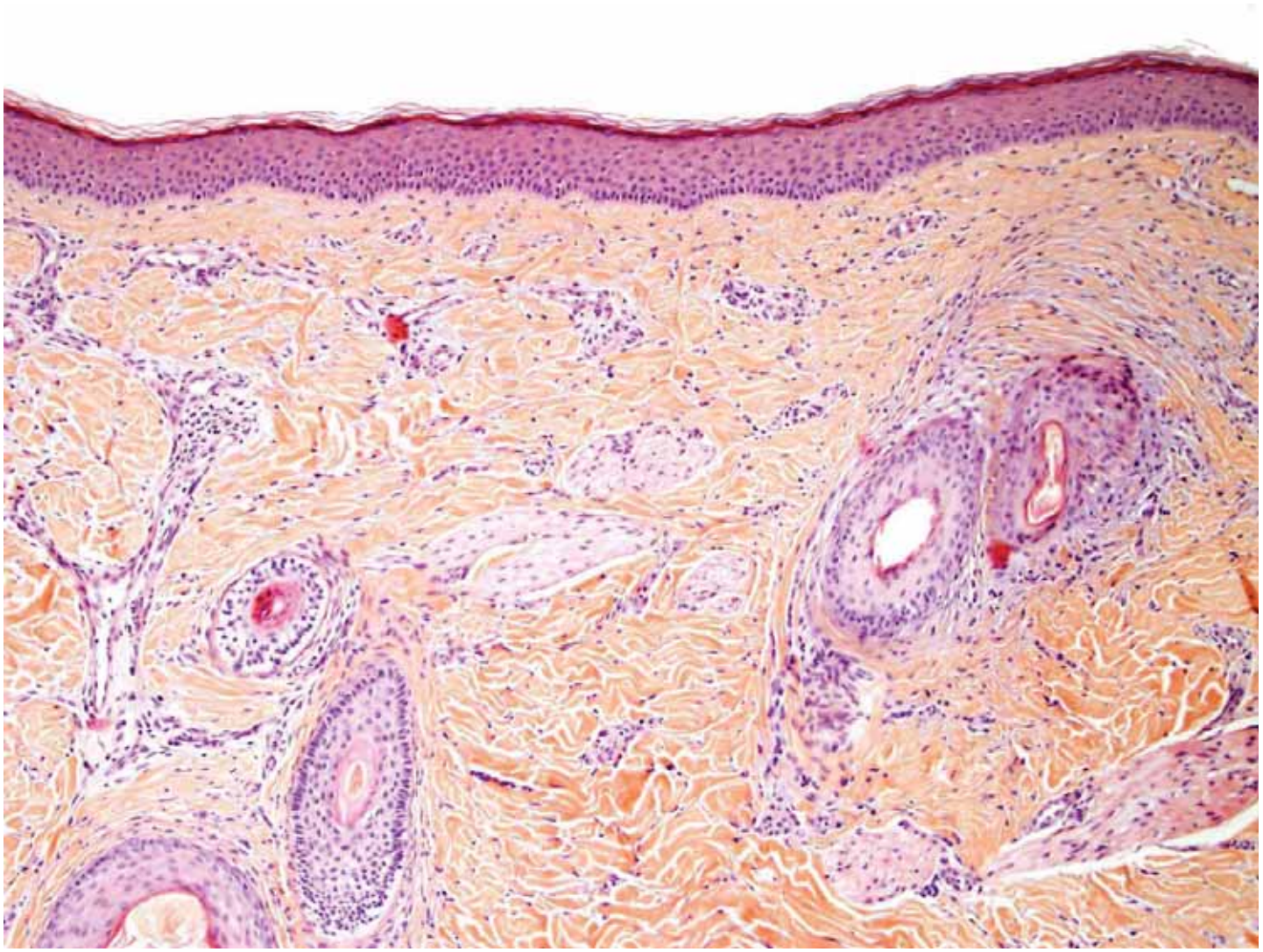
Necker
ENFANTS MALADES
HÔPITAL UNIVERSITAIRE

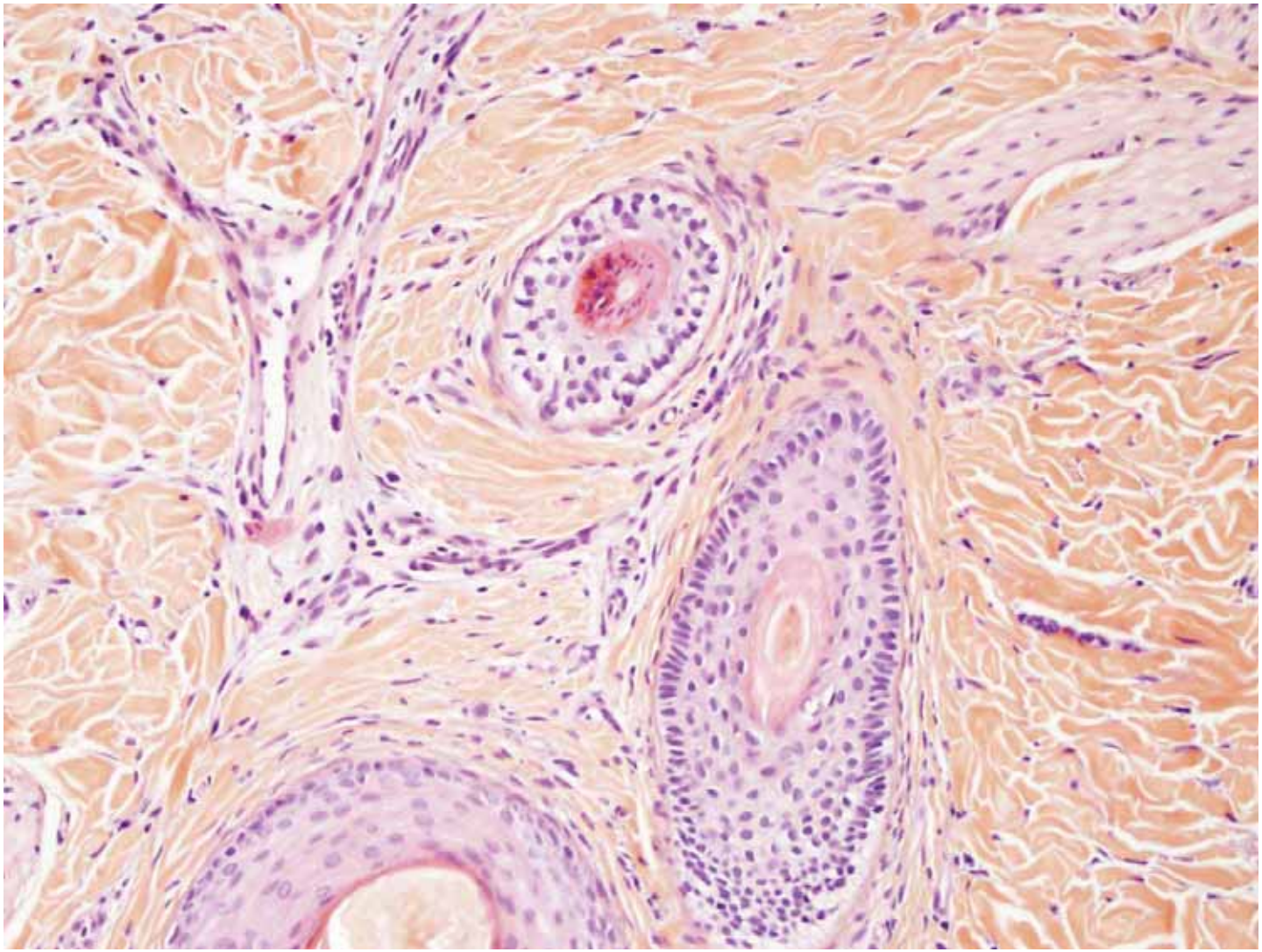
UNNA DARIER CLUB MAY 2025
MUNSTER

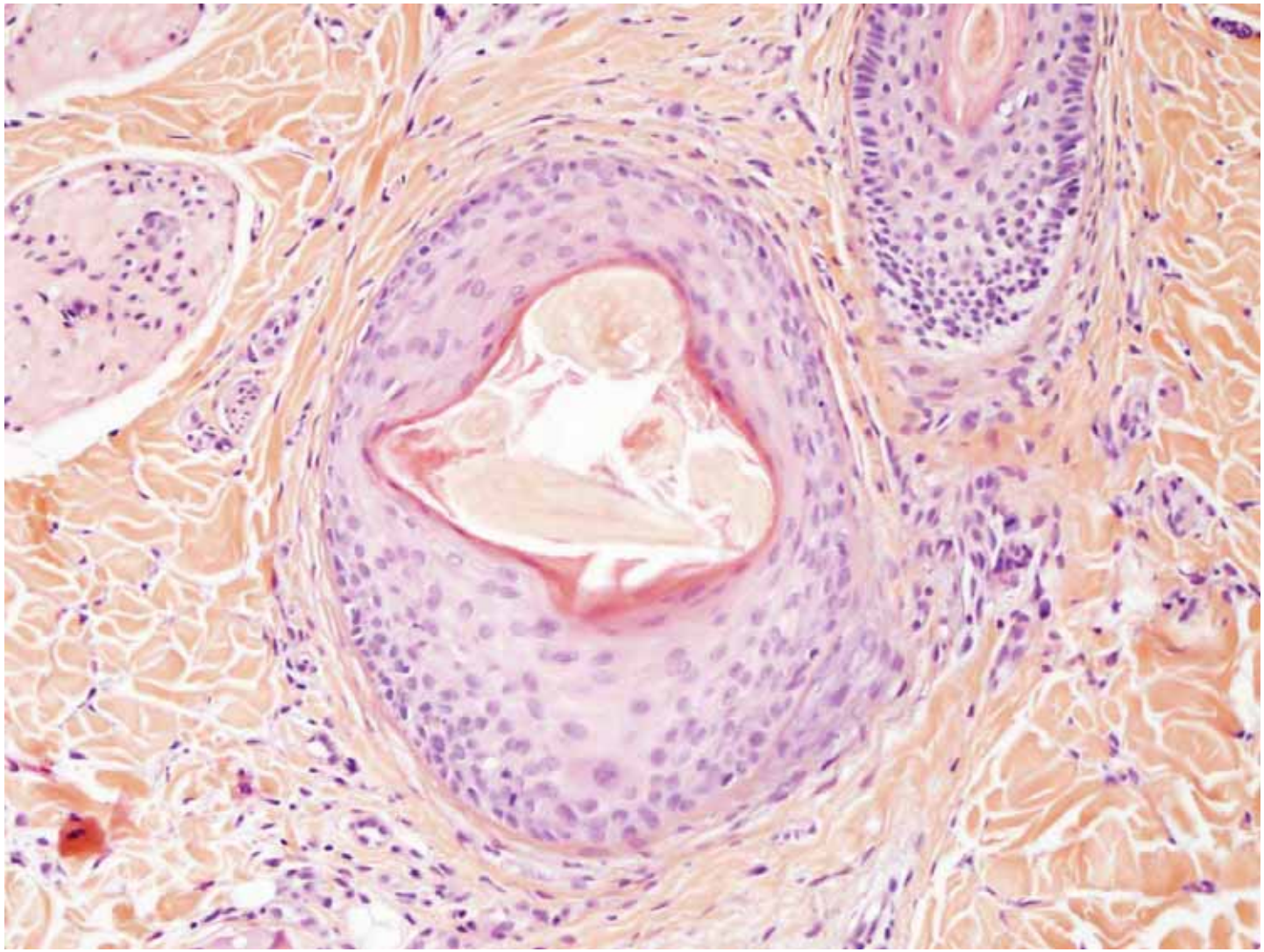
One year old boy with loss of eyebrows and diffuse alopecia

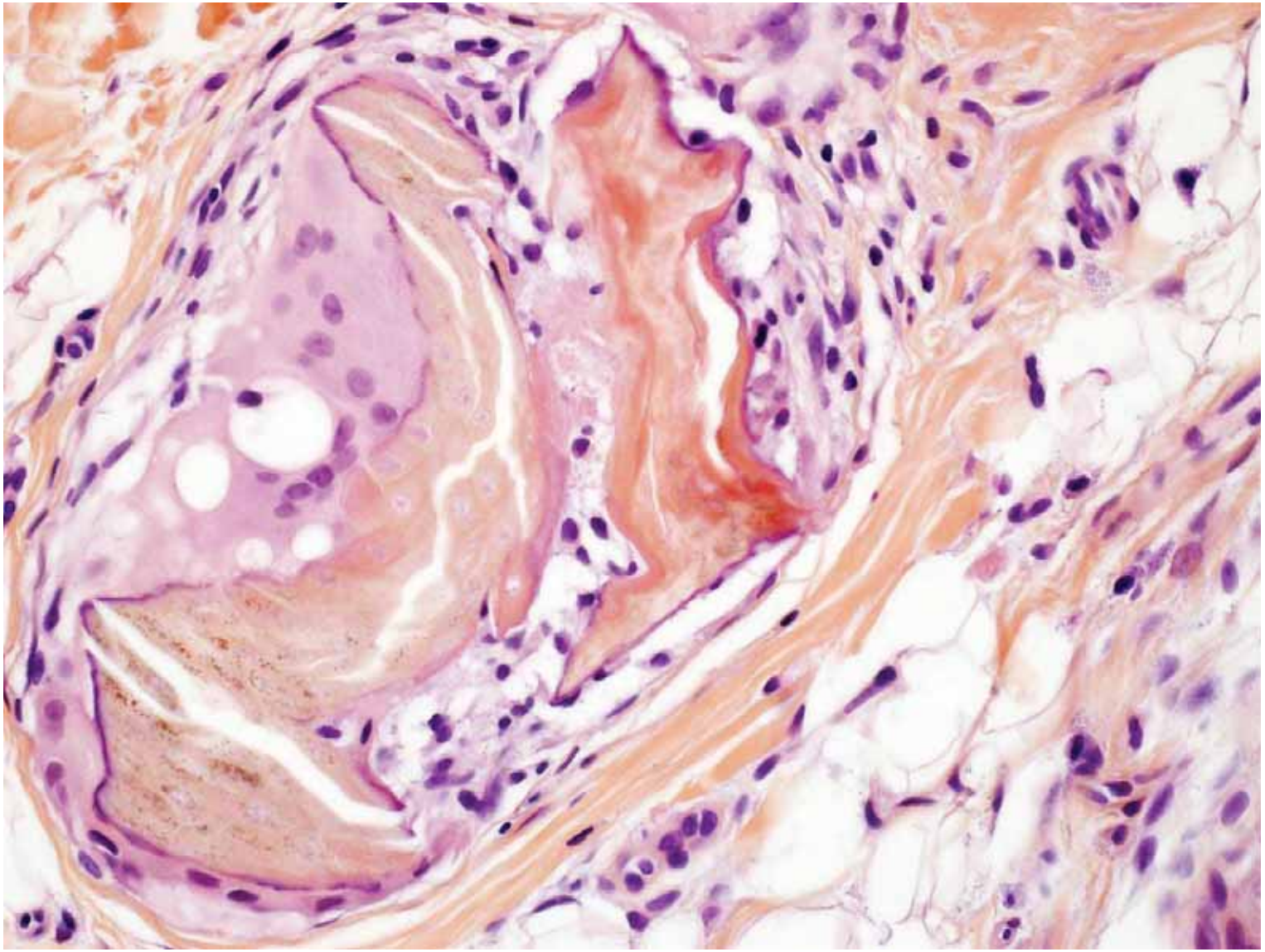
On examination











ATRICHIA WITH PAPULAR LESIONS

- Rare genetic condition
- Early-onset irreversible hair loss (within first few months of life)
- Development of keratin-filled papules on face/trunk/extremities
- Rarely may present without the characteristic papular lesions early in the disease, or due to variability of expression
- Some patients may have only alopecia and develop papules later, or not at all
- DDX: Alopecia universalis
- Final diagnosis: genetic testing (HR or DSG4 gene mutations)

Acta Derm Venereol 2008; 88: 346–349

INVESTIGATIVE REPORT

Atrichia with Papular Lesions: A Report of Three Novel Human Hairless Gene Mutations and a Revision of Diagnostic Criteria

Leona YIP[1], Liran HOREV[2], Rodney SINCLAIR[1] and Abraham ZLOTOGORSKI[2]

[1]Skin and Cancer Foundation of Victoria and the Department of Dermatology, St Vincent's Hospital Melbourne, Victoria, Australia, and [2]Department of Dermatology, Hadassah - Hebrew University Medical Center and the Center for Genetic Diseases of the Skin and Hair, Jerusalem, Israel

Atrichia with papular lesions is a rare autosomal recessive condition characterized by complete irreversible hair loss during the first months of life and papules that appear during early childhood. Atrichia with papular lesions is frequently misdiagnosed as alopecia universalis, despite increasing reports of its prevalence and the presence of well-defined diagnostic criteria. Most cases of atrichia with papular lesions have been reported in consanguineous families residing in small geographical regions, but the increasing number of sporadic cases of unrelated individuals suggests that atrichia with papular lesions is more common than previously thought. Mutations in the human hairless gene on chromosome 8p12 have been implicated in this disease. Here, we report two novel heterozygous mutations in an Australian family and a novel homozygous mutation in 2 Arab siblings. We also revise the diagnostic criteria for atrichia with papular lesions in order to clarify its uniqueness and distinguishing features from alopecia universalis. ***Key words: hair loss; alopecia universalis; mutation; atrichia with papular lesions.***

(Accepted February 5, 2008.)

Acta Derm Venereol 2008; 88: 346–349.

Abraham Zlotogorski, Department of Dermatology, Hadassah - Hebrew University Medical Center, Jerusalem IL-91120, Israel. E-mail: zloto@cc.huji.ac.il

permanently immature hair follicle structures, resulting in papules that are made up of follicular cysts filled with cornified material (9).

Zlotogorski et al. (9) described 12 clinical and laboratory criteria for diagnosing APL, which included atrichia, papules, and a family history of consanguinity. APL is still under-diagnosed, despite established diagnostic criteria and increasing reports of its sporadic prevalence. It is important to recognize APL and differentiate it from alopecia areata, as treatments for APL are futile, and treatments for refractive alopecia areata can have significant side-effects. Previous reports of APL have concentrated on certain Mediterranean and Pakistani populations due to the high incidence of consanguinity in these regions. However, the increasing reports of novel compound mutations outside these geographic boundaries suggest that APL is not exclusive to consanguineous families and is more prevalent than previously thought. The first report of a compound heterozygous mutation in a non-consanguineous individual was in 2002 (10), thereafter followed by another 5 reports (4, 11, 12). Here, we report 2 novel heterozygous mutations in an Australian family and a novel homozygous mutation in 2 Arab siblings. We also revise the diagnostic criteria for APL, with the aim of further clarifying and simplifying its diagnosis and distinguishing features from alopecia universalis.

CASE REPORTS

Table I. *Revision of diagnostic criteria for atrichia with papular lesions*

MAJOR CRITERIA
(*4 out of 5 required for diagnosis*)
• Permanent and complete absence of scalp hairs by the first few months of life.
• Few to widespread smooth, whitish, or milia-like papules on the face, scalp, arms, elbows, thighs or knees from infancy or childhood.
• Replacement of mature hair follicle structures by follicular cysts filled with cornified material in scalp histology.
• Mutation(s) in the human hairless gene through genetic testing.
• Clinical and/or molecular exclusion of vitamin-D-dependent rickets.
MINOR CRITERIA
(*supplementary criteria*)
• Family history of consanguinity.
• Absence of secondary axillary, pubic, or body hair growth and/or sparse eyebrows and eyelashes.
• Normal growth and development, including normal bones, teeth, nails and sweating.
• Whitish-hypopigmented streaks on the scalp.
• Lack of response to any treatment modality.

Fig. 1. (a) Dermal papules on the extensor surface of the arm in case 1. These papules can be difficult to differentiate clinically from keratosis pilaris but, histologically, these are follicular cysts filled with cornified material. (b) Complete scalp atrichia with a few papules in case 2.

Atrichia with Papular Lesions: Dermoscopy to the Rescue

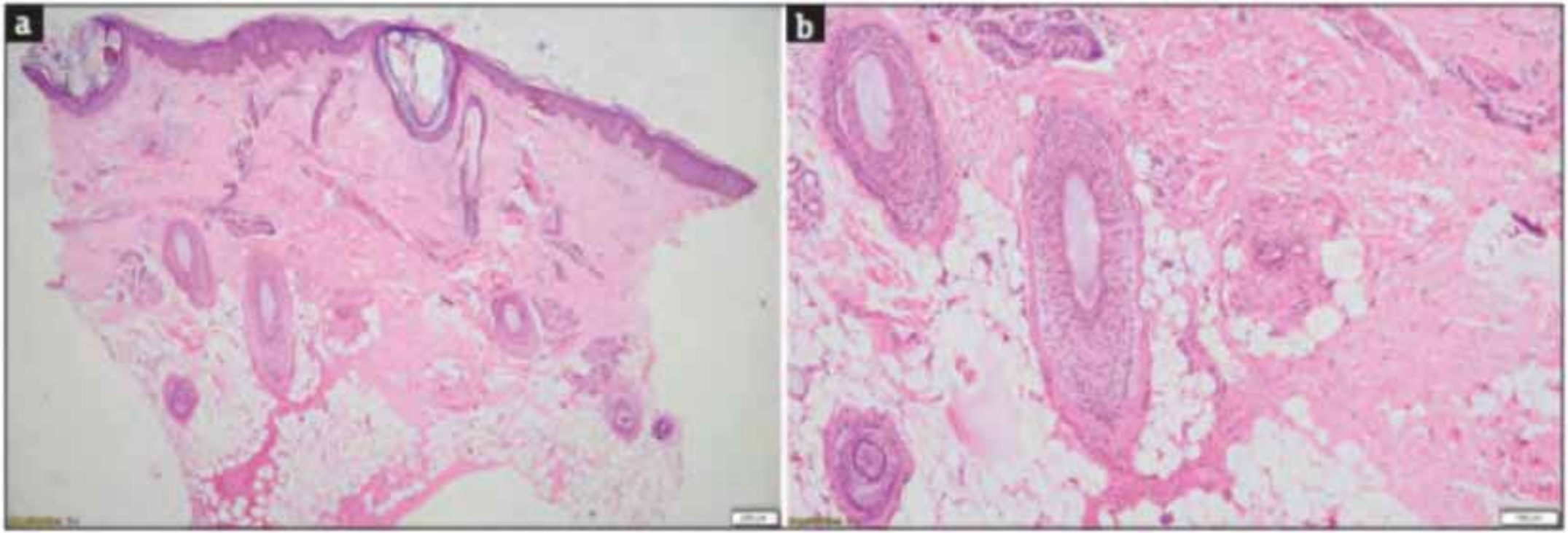
Neha Yadav, Molisha Bhandari, Vivek Sharma

From the Department of Dermatology and STD, Vardhaman Mahavir Medical College and Safdarjung Hospital, Ansari Nagar, New Delhi, India
E-mail: molishabhandari2905@gmail.com

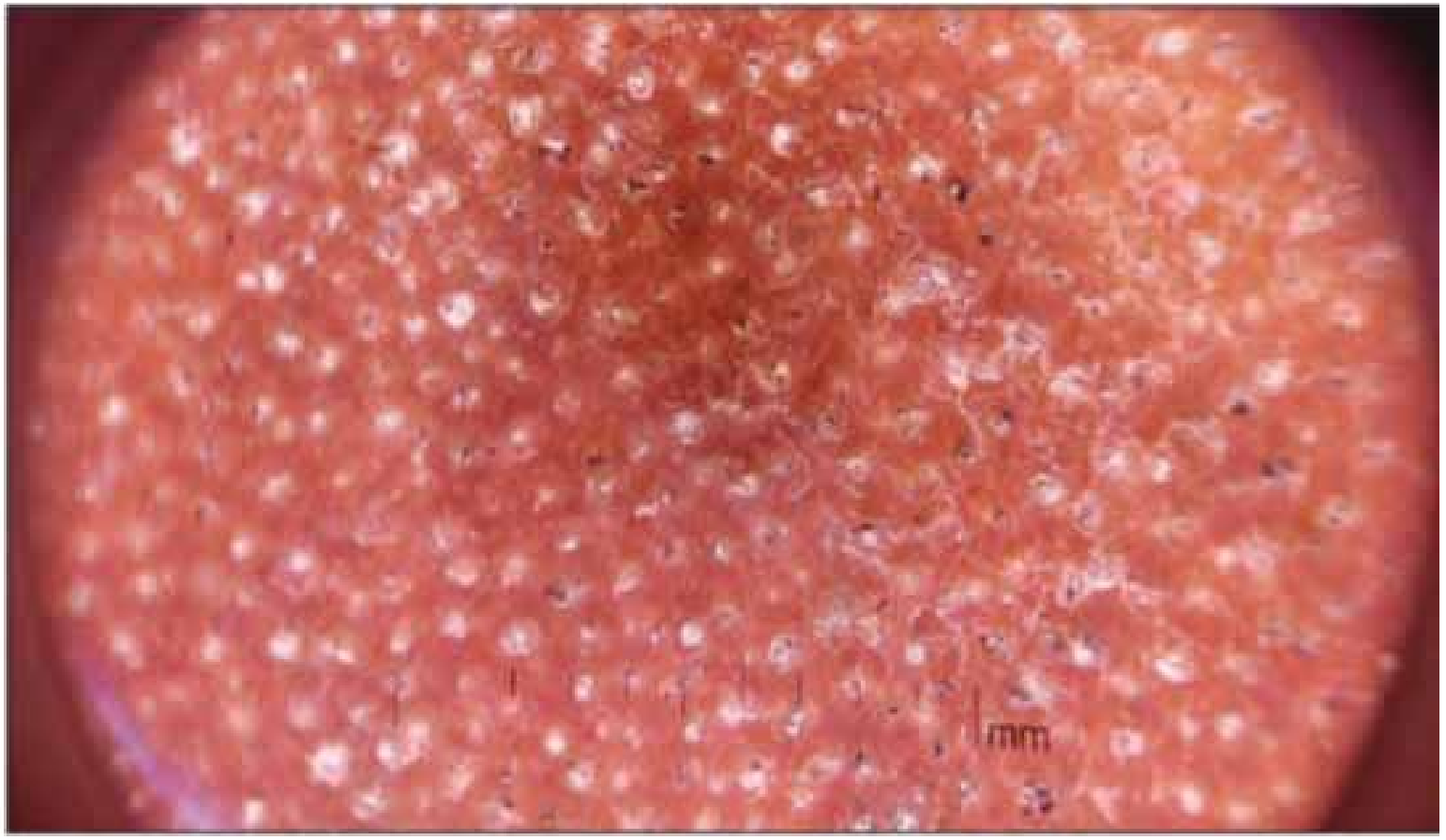
Indian J Dermatol 2025:70(2):106-7

Atrichia with papular lesions (APL) is a rare autosomal recessive (AR) follicular disorder caused by a mutation of the hairless (HR) gene.[1] The child is born with normal hair, but progressive hair loss is usually complete within the first few months of life, never to grow back. Here, we present a case of a 3-year-old boy presenting with alopecia universalis with papular lesions, where dermoscopy helped in clinching the diagnosis.

A 3-year-old boy presented to the outpatient department with complete loss of hair on the scalp and body since the age of 1 year. The child was born with scanty hair at birth, but the hair was lost progressively. The child also developed multiple skin-colored lesions on the scalp and body from the age of 1 year, which gradually increased in number. The boy's physical and neurologic development were normal, as per his age. No history of decreased sweating, decreased hearing, seizures, or bone pains could be elicited. There was no history of consanguinity. There was no history of similar complaints in siblings or any of the family members. On examination, there was a complete absence of hair on the body and scalp. Multiple skin colored to whitish papules of size 0.2–0.5 cm were present on the entire body, most conspicuously on the scalp [Figure 1]. The rest of the cutaneous examination, including mucosae, nails, teeth, and sweat glands, was normal. He had no bony abnormalities. The rest of the systemic examination was within normal limits. The weight and height of the child were within the normal range as per

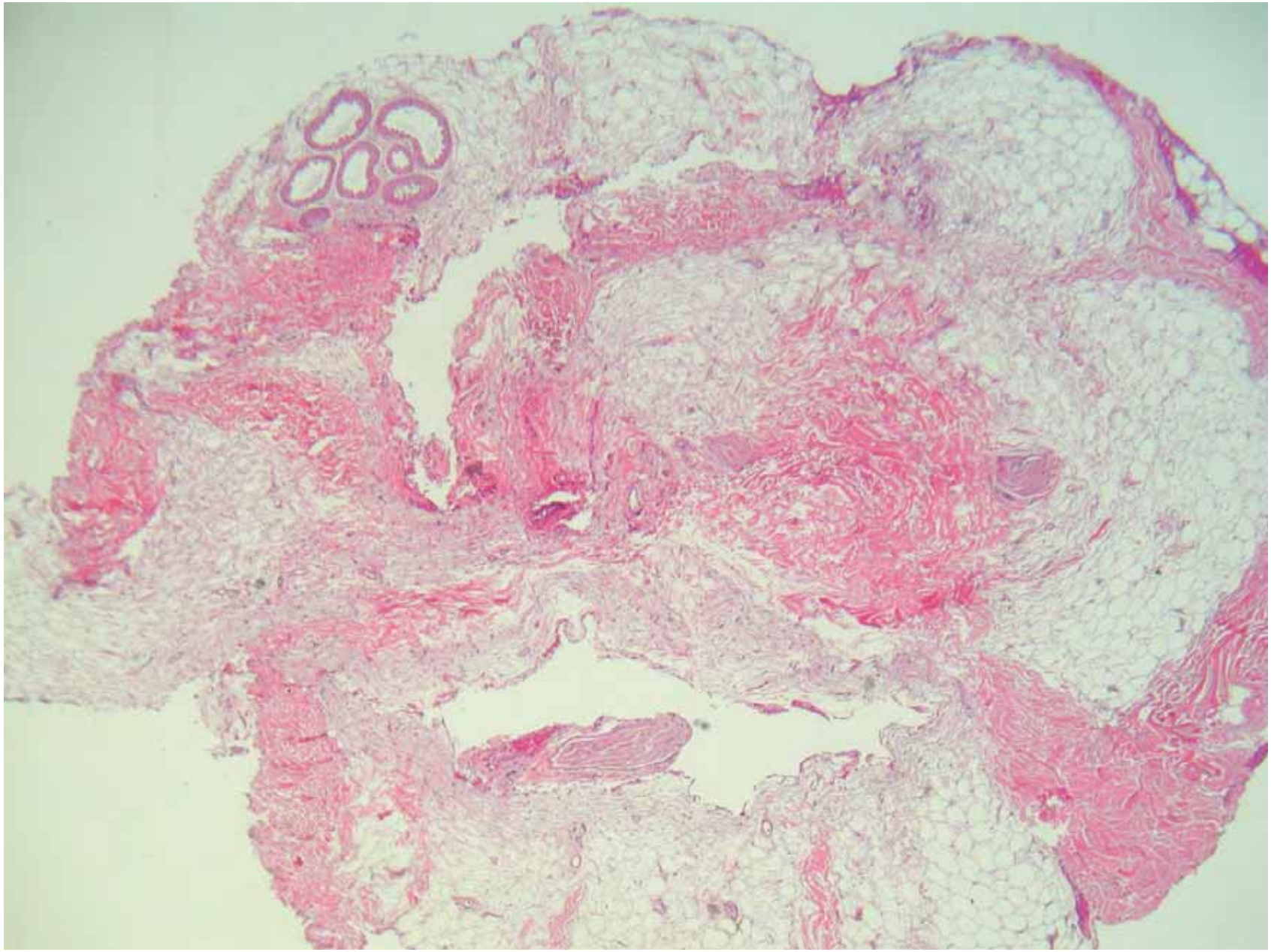

a
b

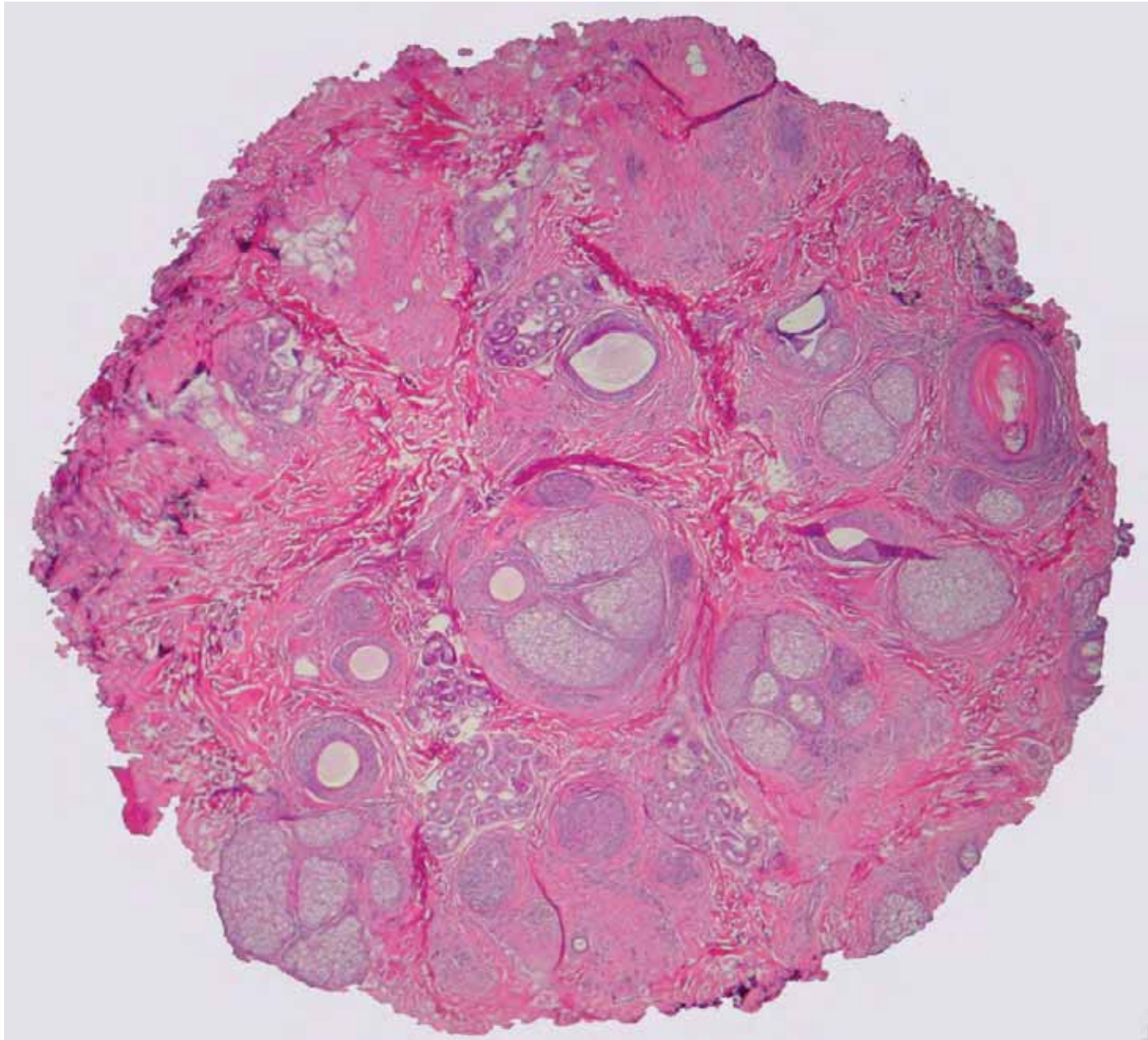
TRICHOSCOPY

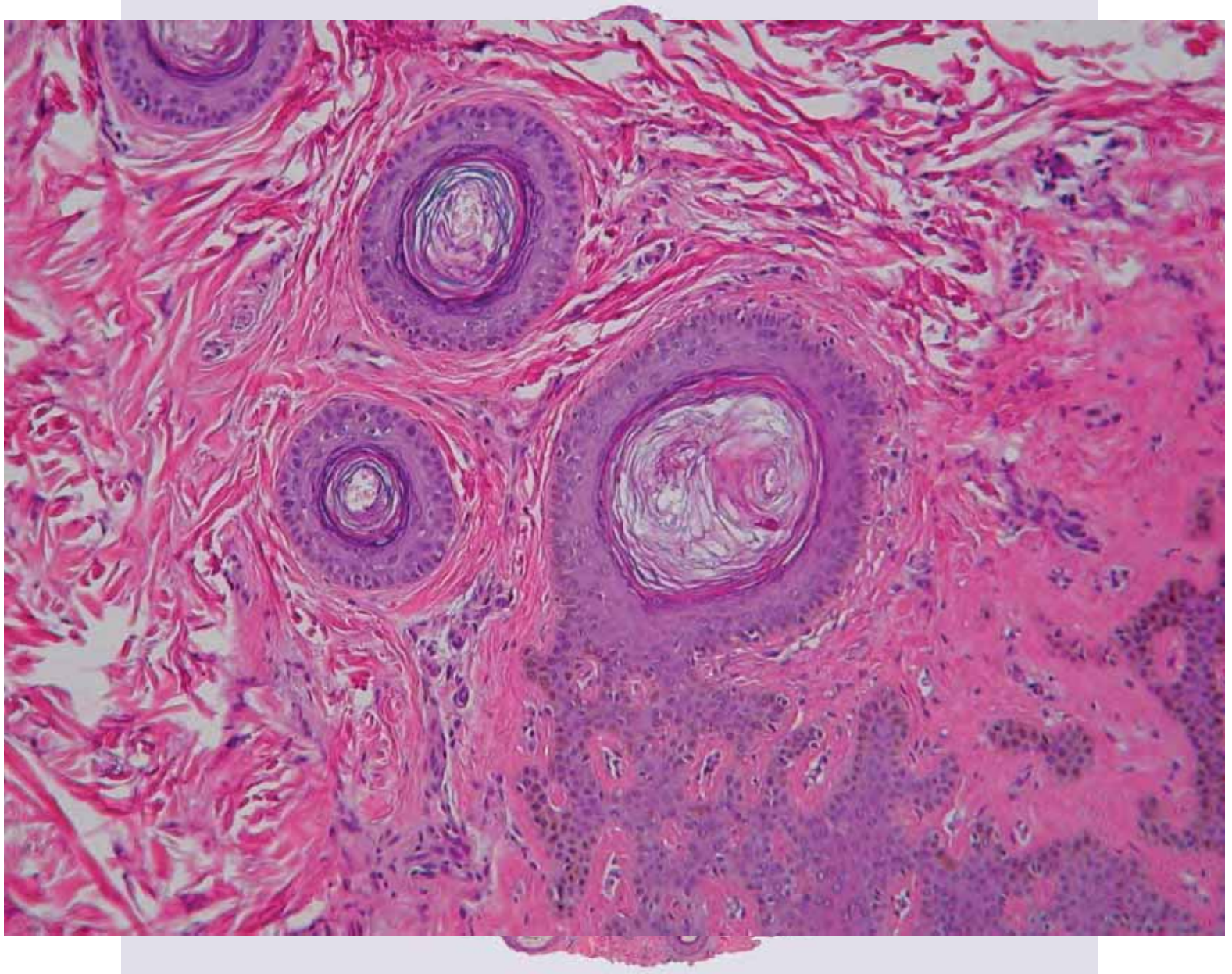


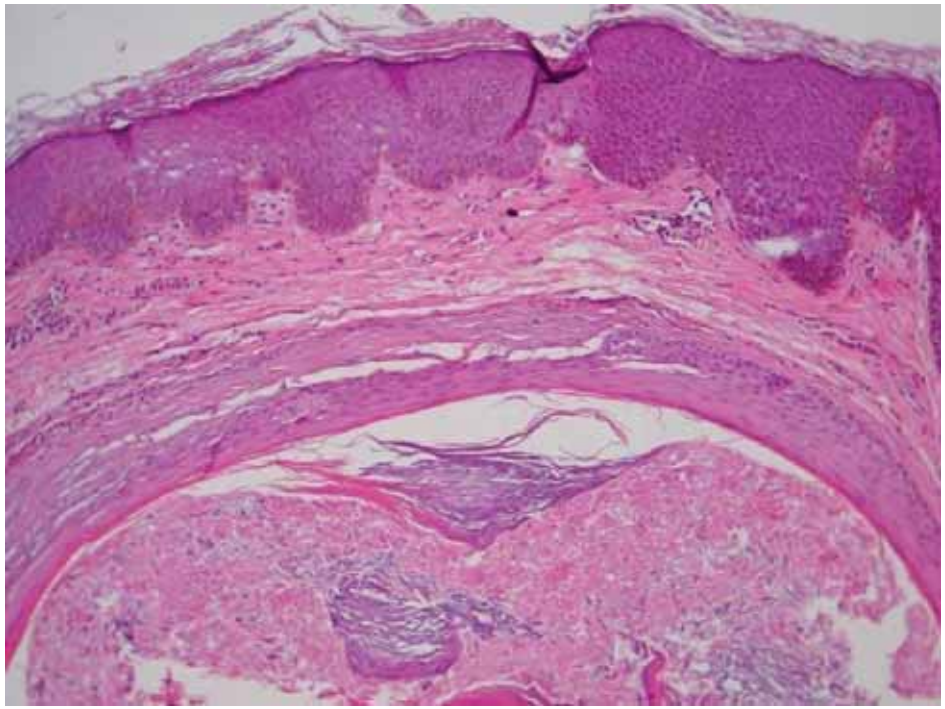
‘Clusters of stars’

ATRICHIA WITH PAPULAR LESIONS









Atrichia with papular lesions

- Total hair loss occurring shortly after birth
- Clinically confused with alopecia totalis/universalis
- Keratinous follicular cysts occurring several years later; face, scalp, extremities
- Histopathology: a scalp bx is diagnostic!
  - Abnormal development of the hair follicle: absence of a hair shaft
  - Epidermal inclusion cysts

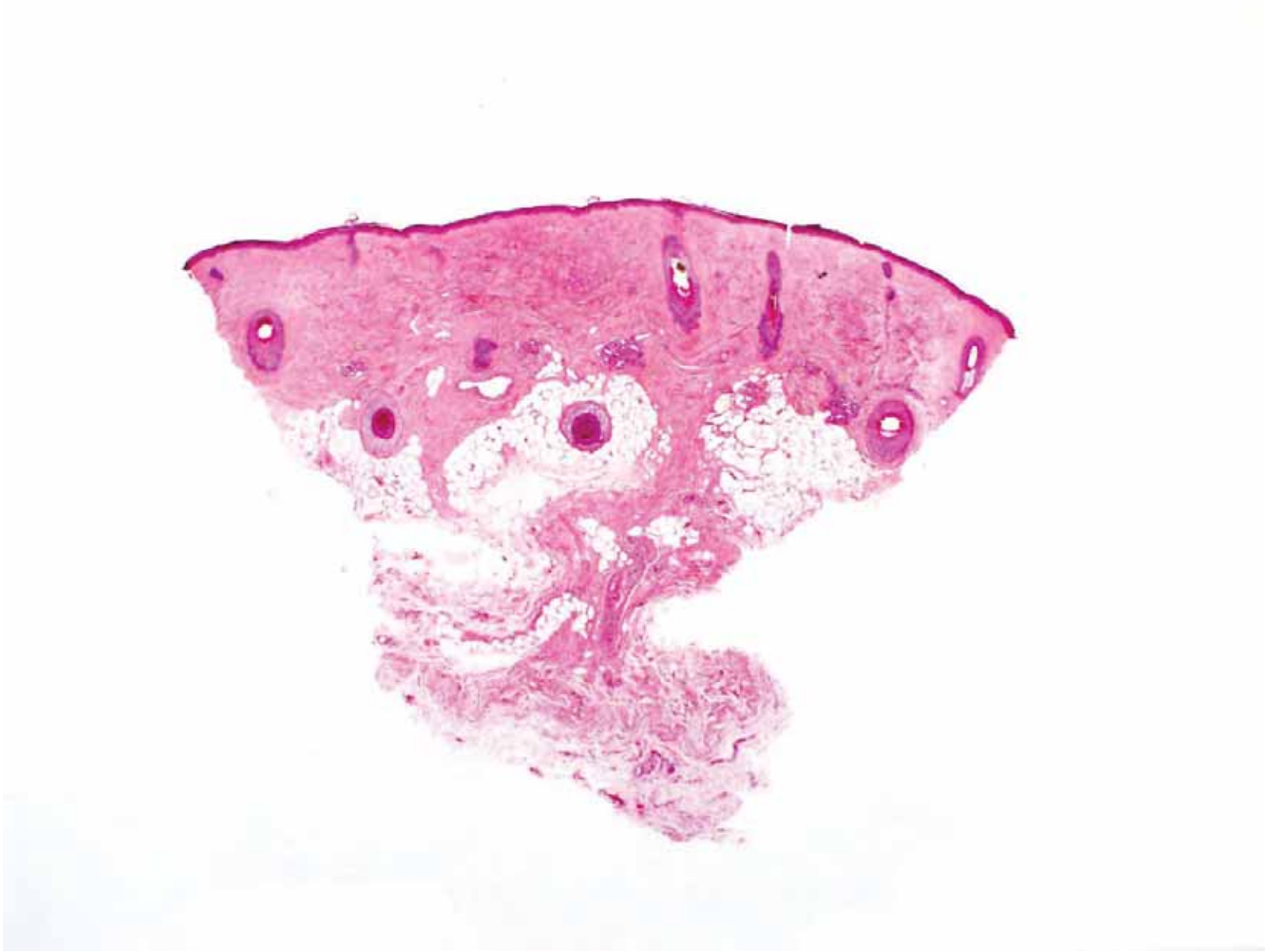
‘DECODING’ SCARRING ALOPECIA WITH STROMA hyalinization

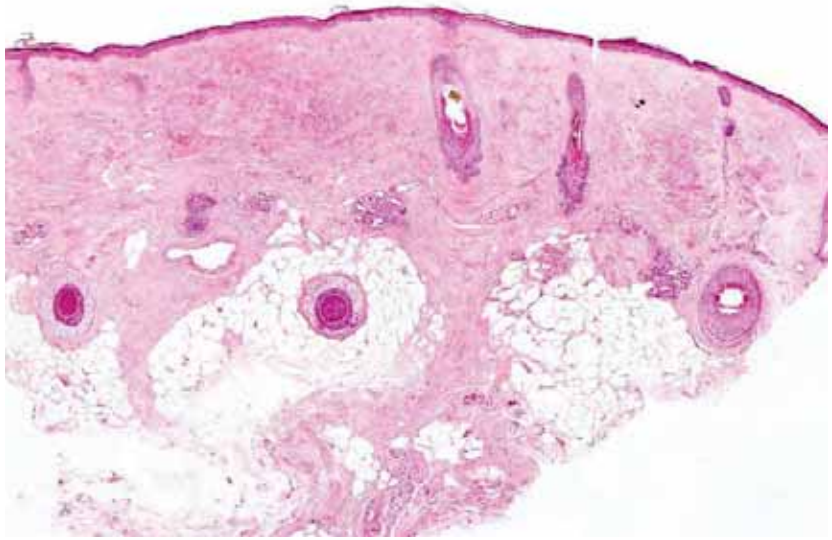
CASE ONE
CLINICAL HISTORY
F43. Scalp alopecia

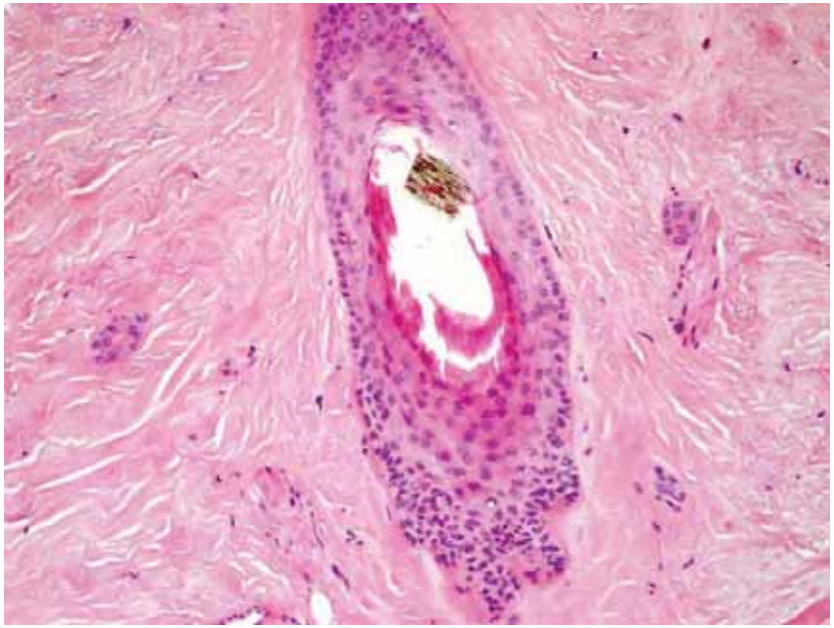
Additional History

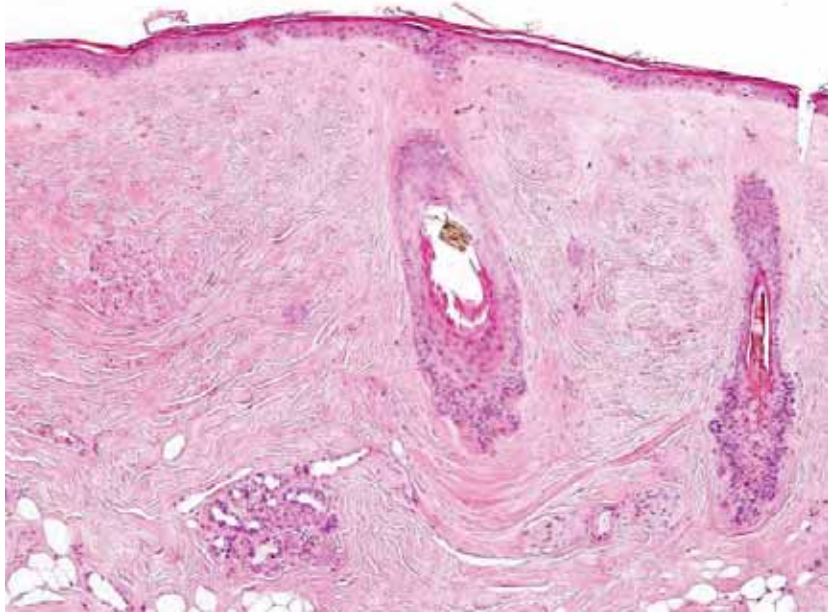
- Abnormal frontline with recession and a non-inflammatory patch of scarring alopecia
- Eyebrow thinning which recovered with use of topical corticosteroids
- Recent loss of axillary hair
- Query: R/O early frontal fibrosing alopecia
- Background hx of scleroderma
- No immunosuppressant Rx at the time of bx

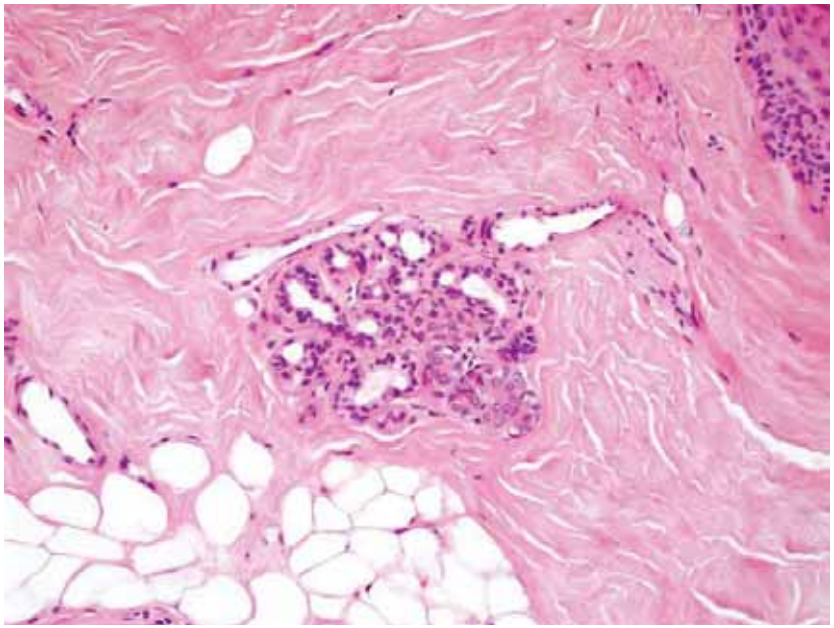
Courtesy Dr Raj Mallipeddi



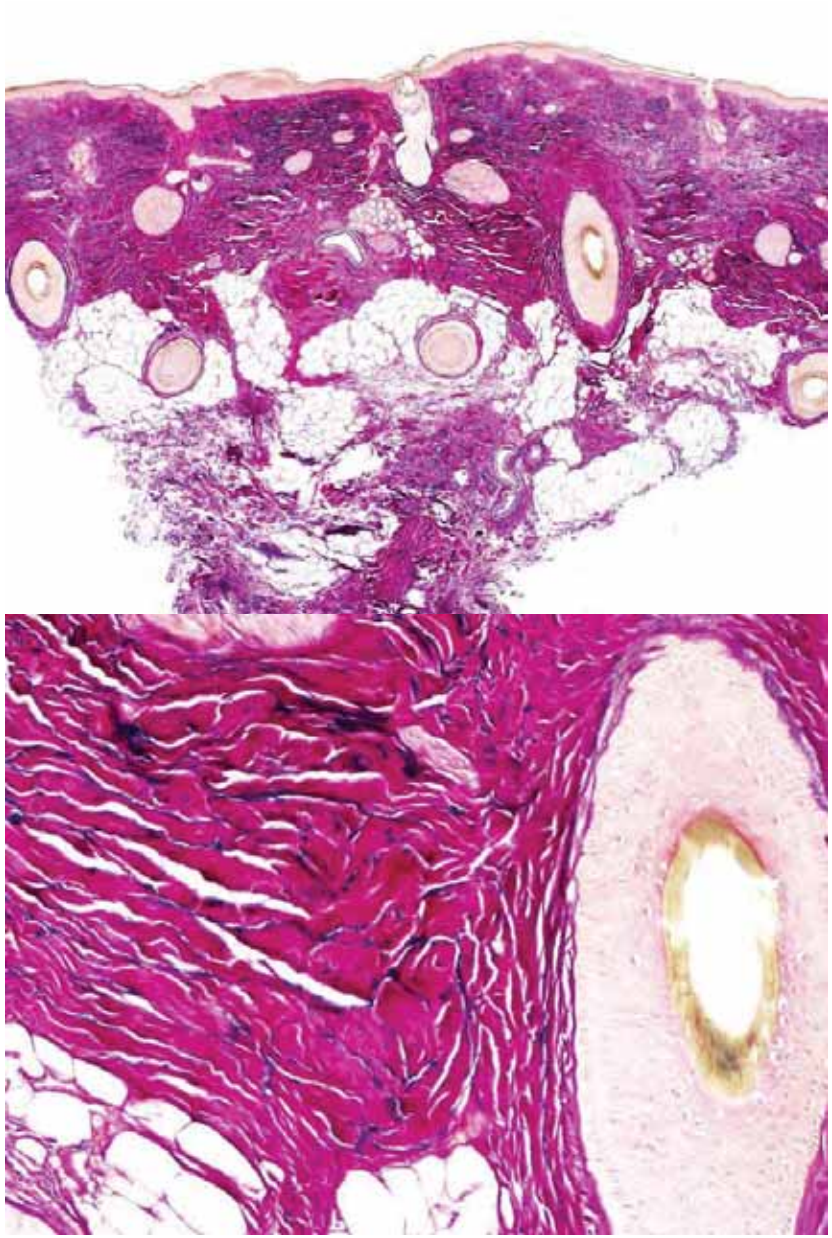


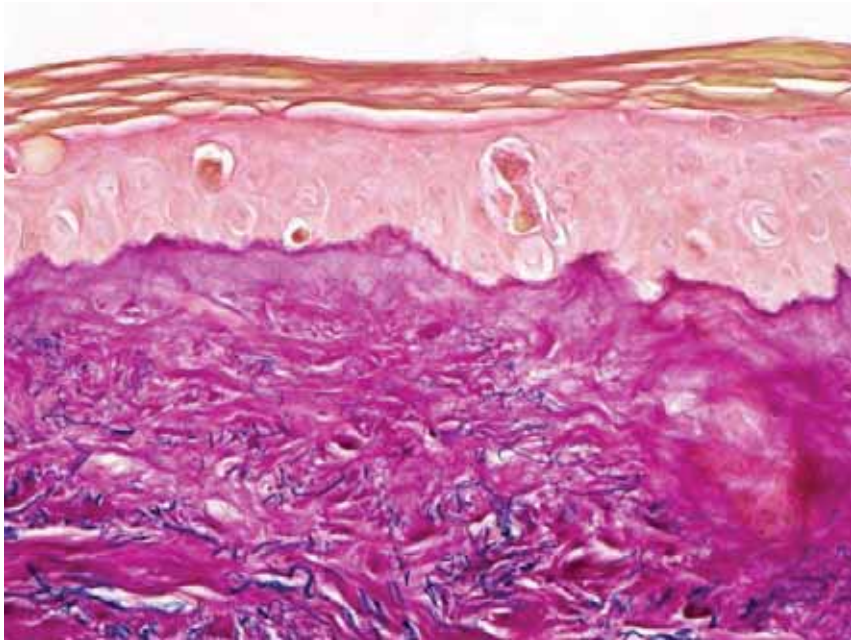


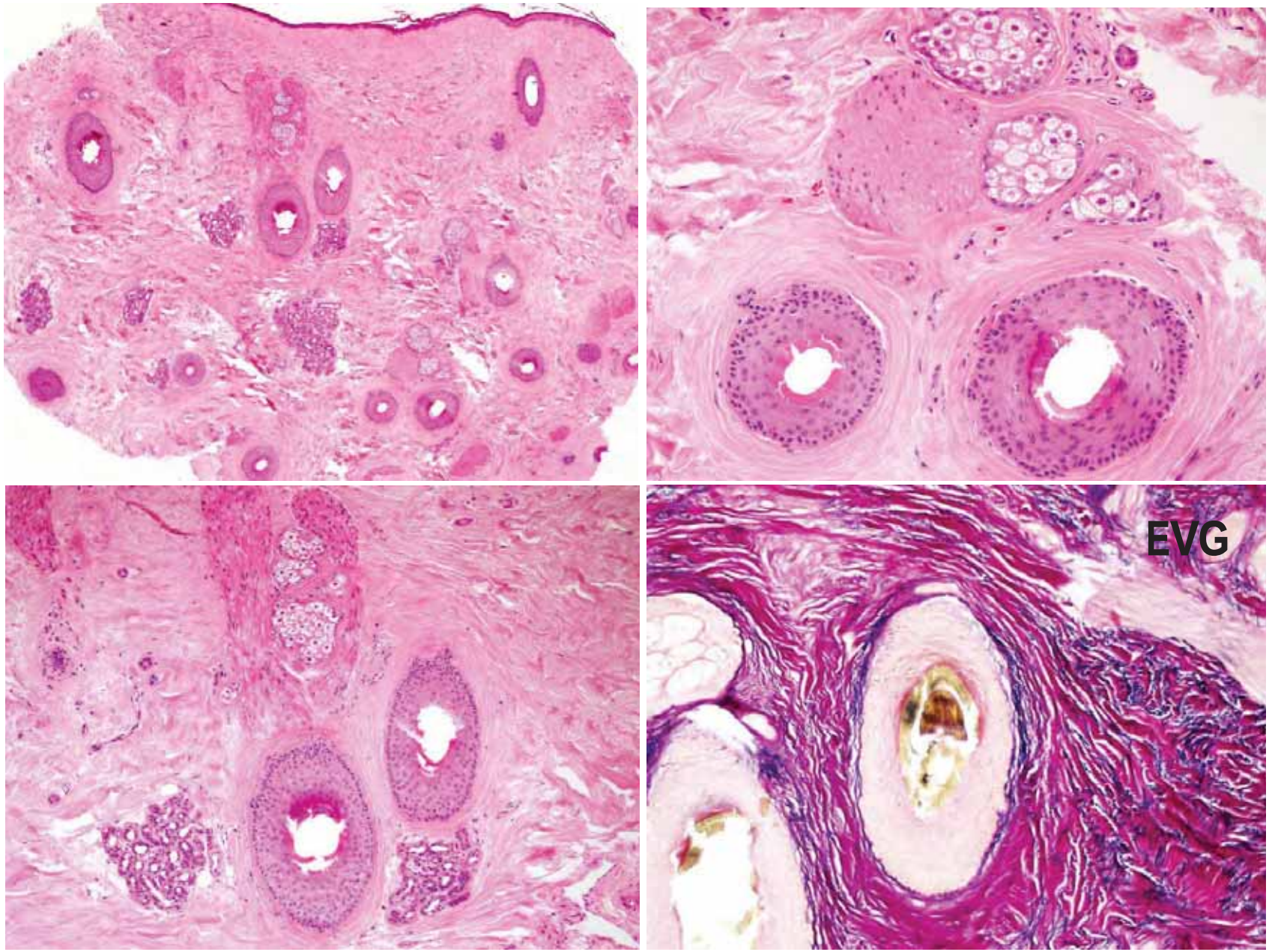




Elastic stain: EVG






EVG

CASE TWO
CLINICAL HISTORY
M13. Patch of scarring alopecia, left vertex

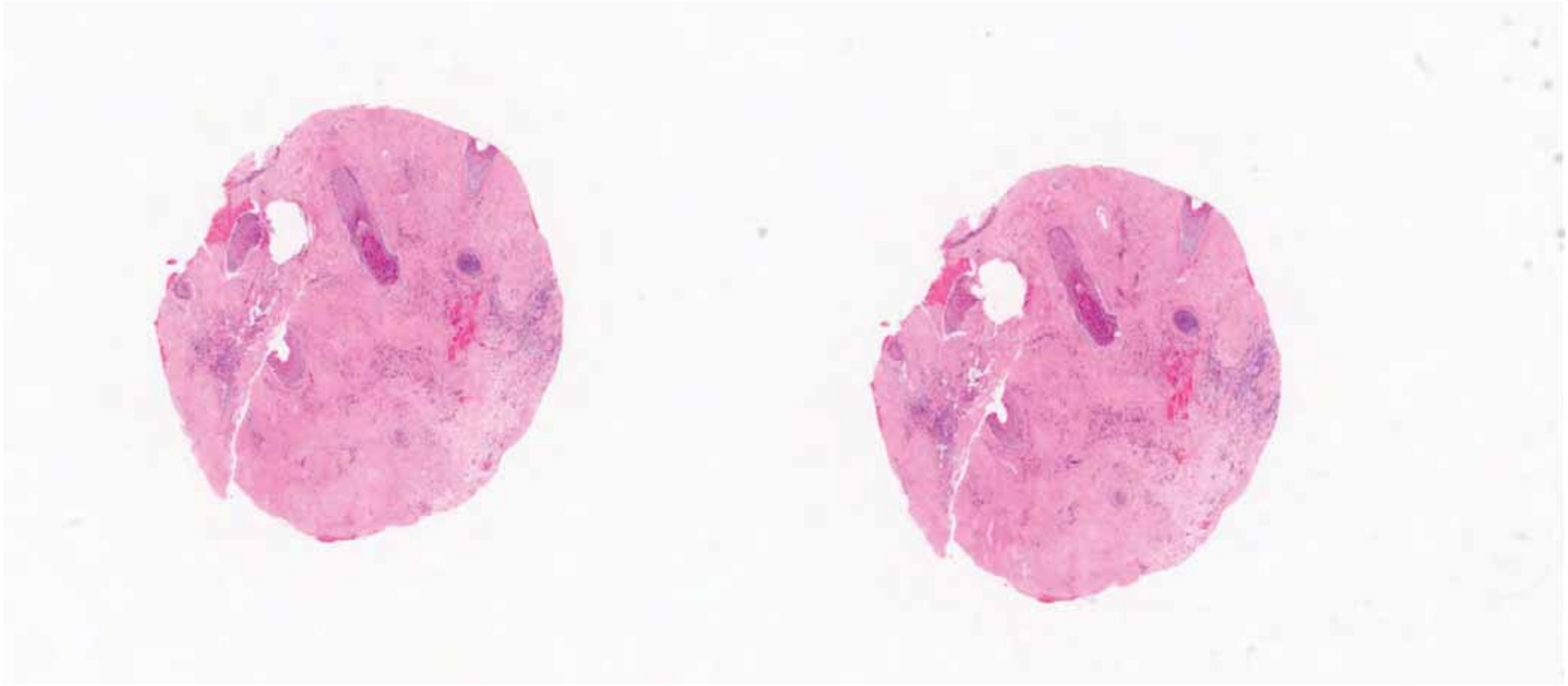
Clinical history

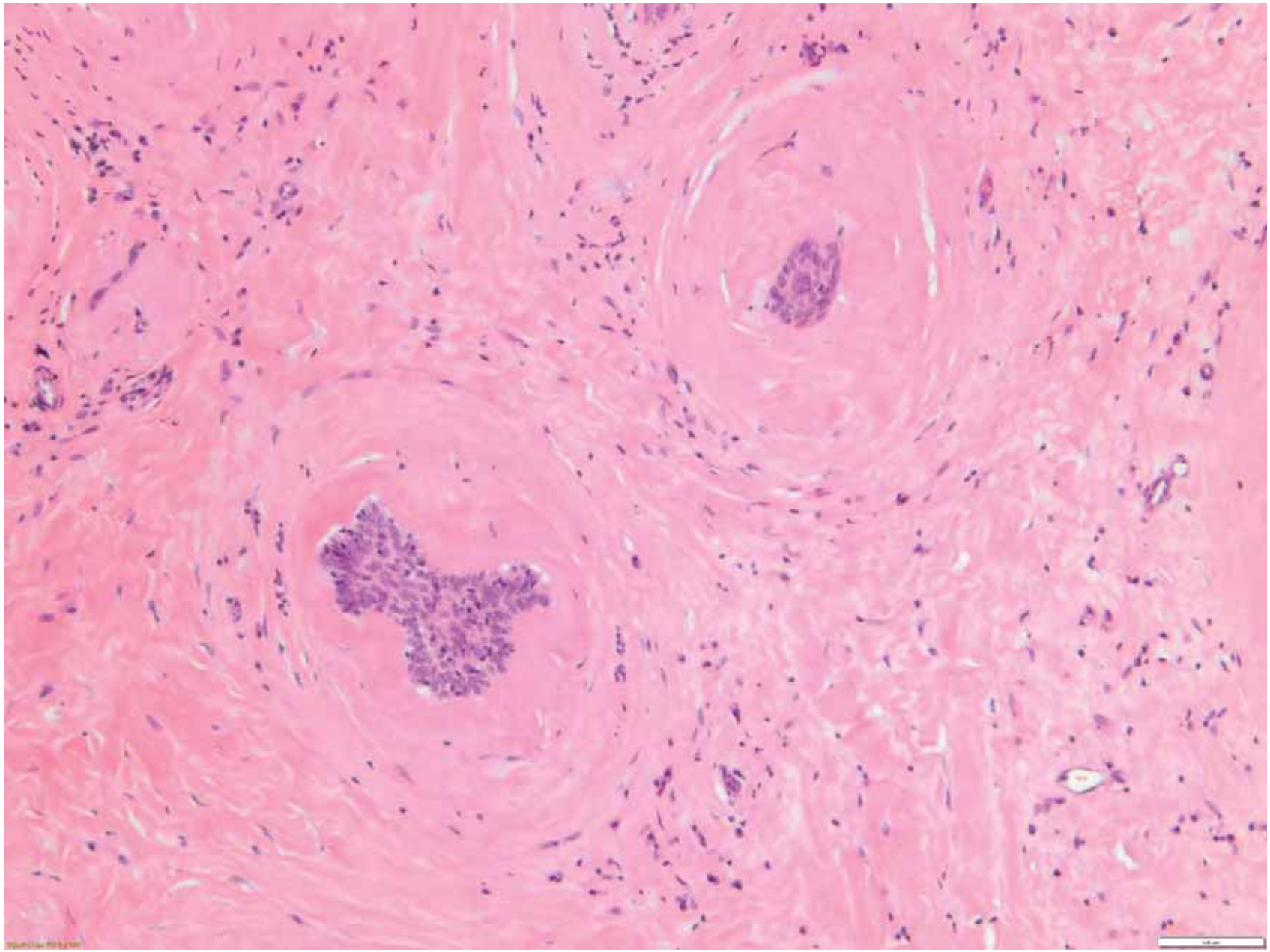
- 13-year-old boy
- Left vertex
- Single patch of scarring alopecia

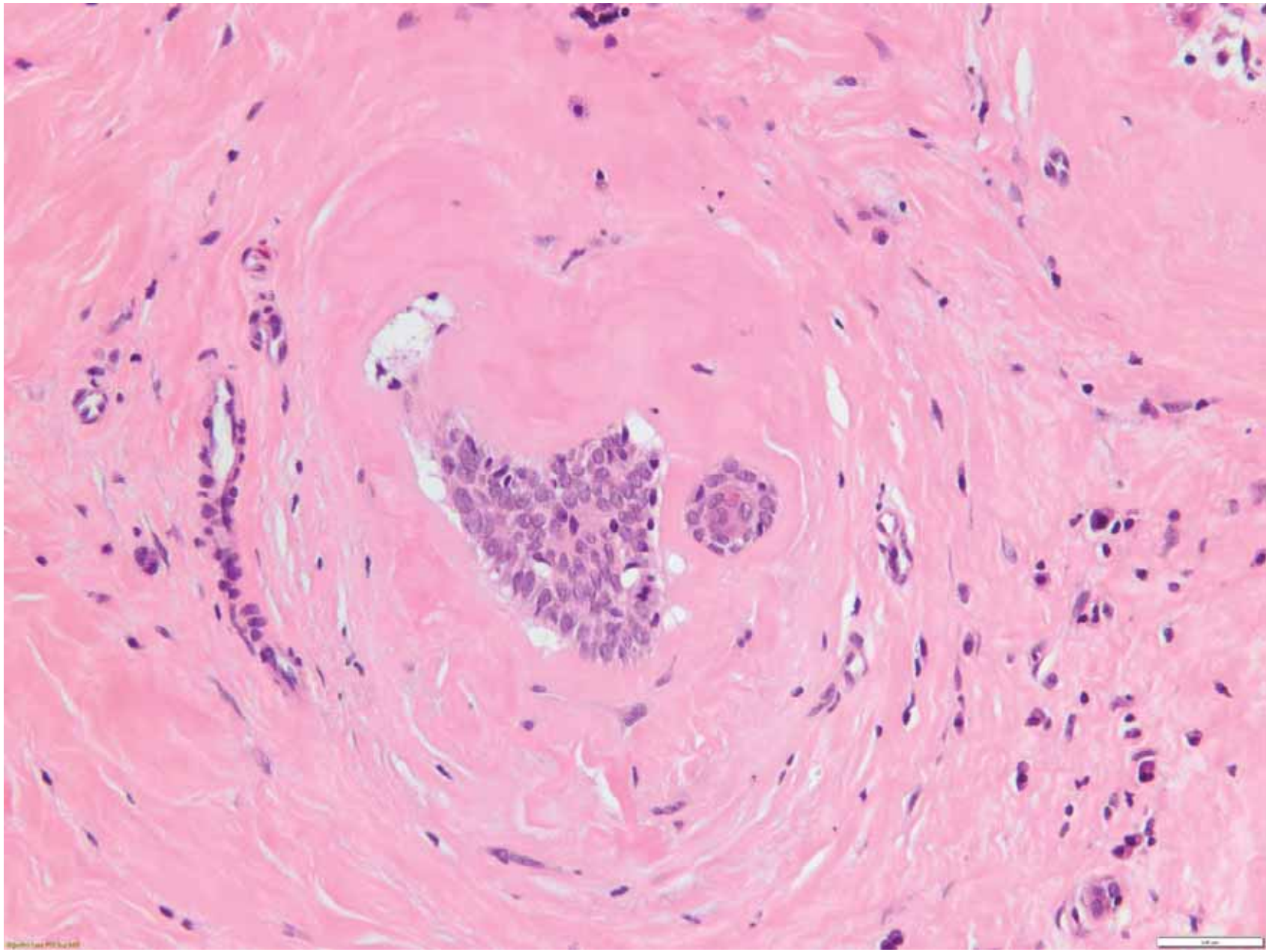
Clinical differential diagnosis;

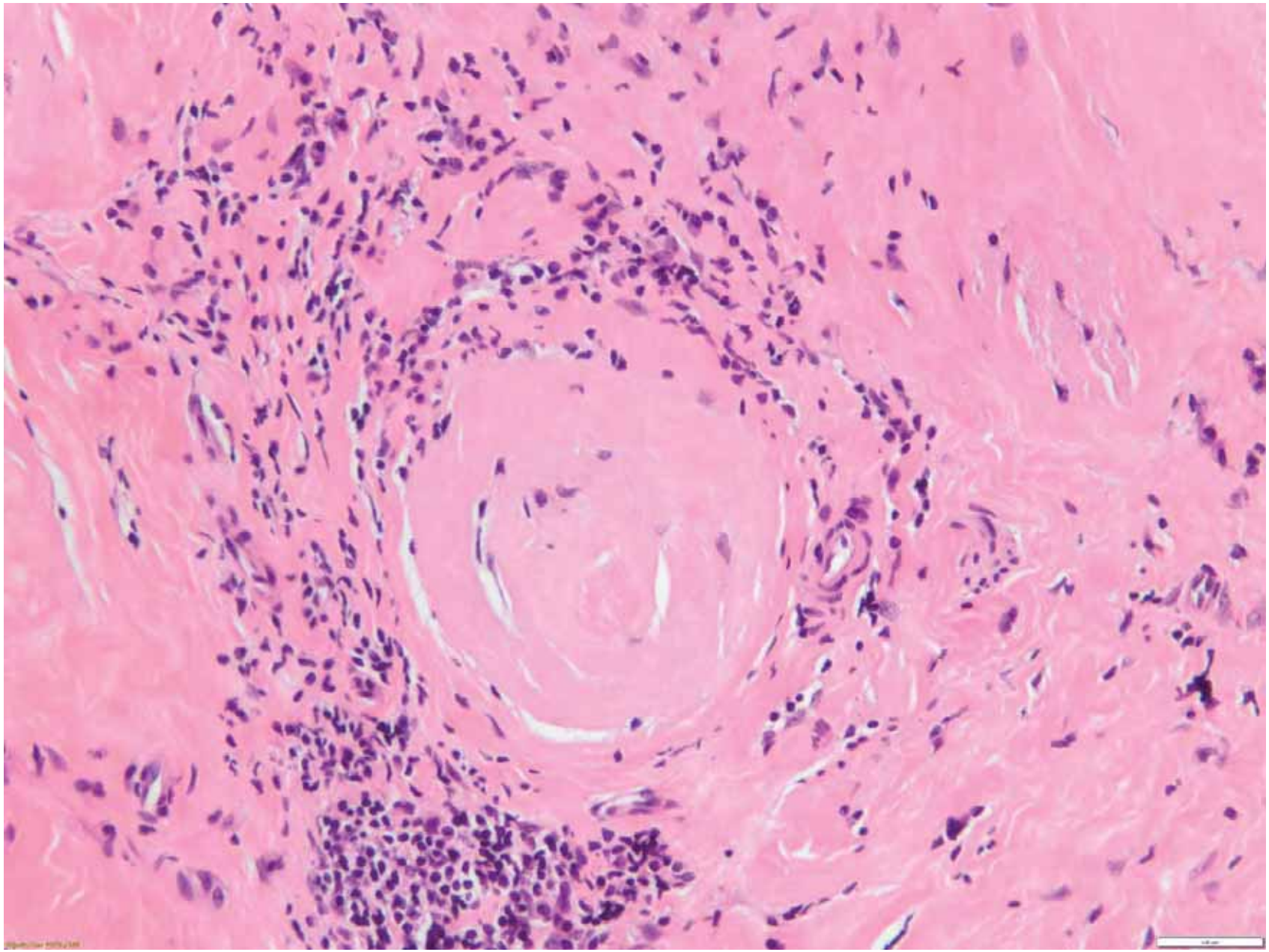
- 1. Lupus
- 2. Lichen planopilaris

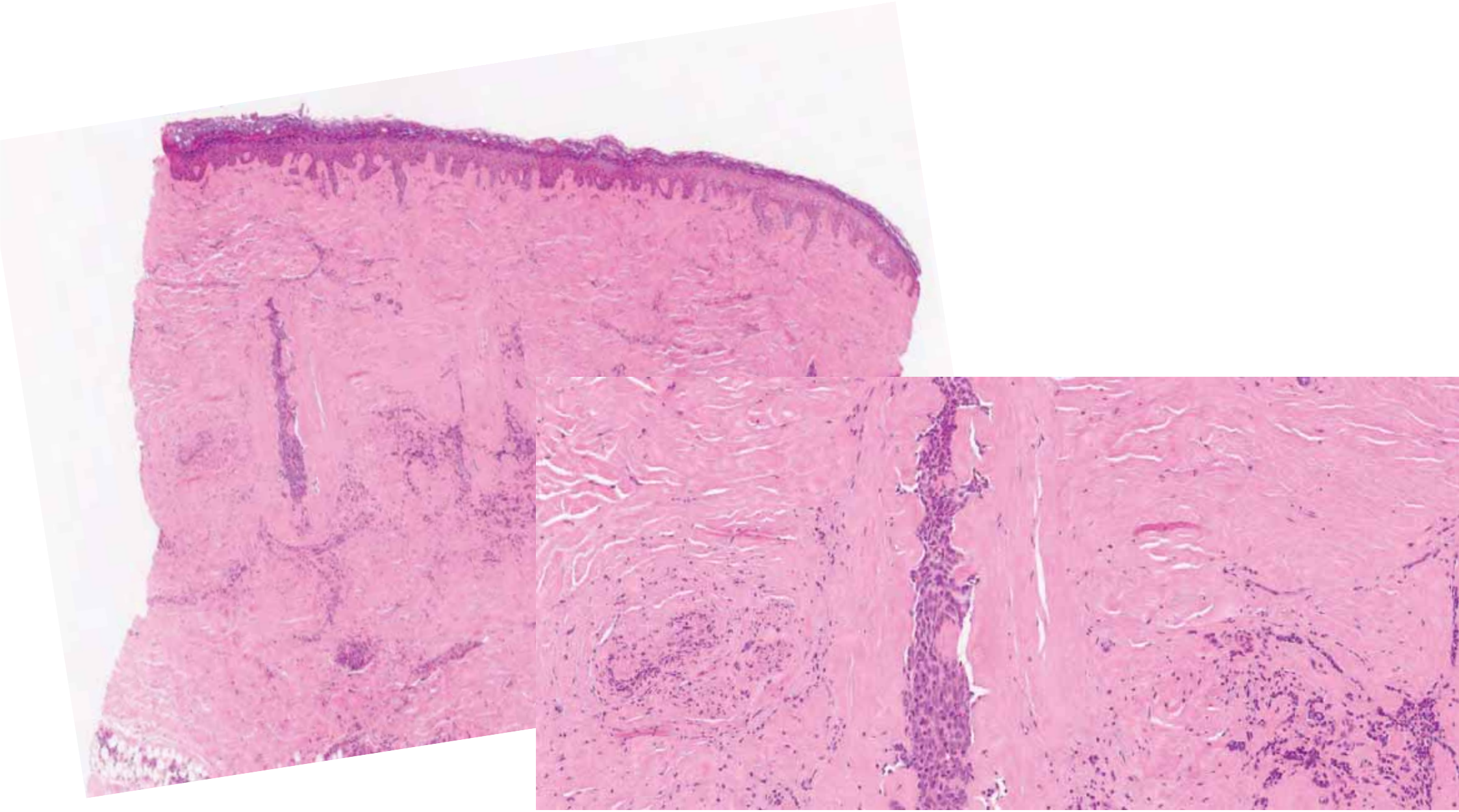
BSD 2025 CASE 40 Glasgow, Scotland: presented by Dr Anoud Zidan

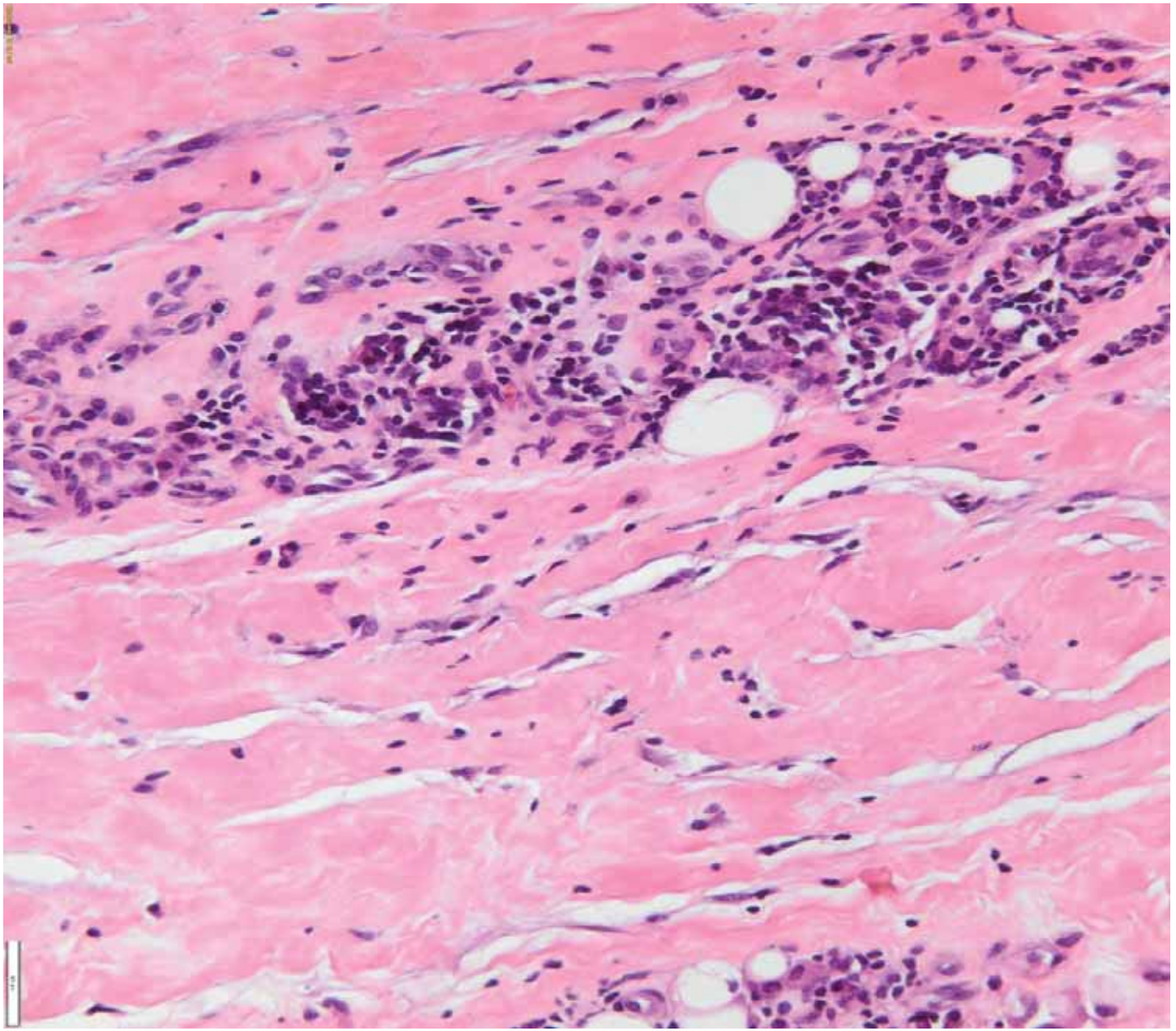












MORPHEA HISTOPATHOLOGY

- Reduction in the number of terminal hair follicles
- Loss of sebaceous glands and reduction in eccrine ducts
- Preservation arrectores pilorum
- Preservation of the elastic fibers with loss of the fine elastic fibers in the upper papillary dermis
- Extensive hyalinisation and sclerosis of dermal collagen
- Follicular remnants with geometric shape resembling telogen/catagen surrounded by rings of hyalinised collagen
- Perifollicular and perivascular infiltrate of lymphocytes and plasma cells

MORPHEA HISTOPATHOLOGY: STAGES

- Early lesions: more extensive inflammatory cell infiltrate (perineural lymphocytes and plasma cells)
- Well-developed lesions: dense dermal sclerosis with marked eccrine gland atrophy and replacement of much of the adipose layer by collagenous tissue.
- Late-stage lesions: increased dermal thickness and broad sclerotic bundles extending into the subcutis

J Cutan Pathol 2012: 39: 518–520
doi: 10.1111/j.1600-0560.2012.01899.x
John Wiley & Sons, Ltd

Journal of
Cutaneous Pathology

Alopecia with perineural lymphocytes: a clue to linear scleroderma *en coup de sabre*

Linear scleroderma *en coup de sabre* ('the stroke of the sword') is an uncommon form of morphea with onset typically in childhood or adolescence. Involvement is usually located on the paramedian forehead and is associated with alopecia. It is microscopically indistinguishable from other forms of scleroderma. We present a 51-year-old woman who presented with alopecia and subsequently developed linear scleroderma *en coup de sabre* on her adjacent forehead. Histopathology revealed a strikingly perineural lymphocytic and plasmacytic infiltrate, extending deeply into the subcutis and fascia. To our knowledge, this is the first report of alopecia with perineural lymphocytic inflammation as a presenting sign of linear scleroderma *en coup de sabre*.

Keywords: alopecia, lymphocytes, morphea, nerves, scleroderma

Goh C, Biswas A, Goldberg LJ. Alopecia with perineural lymphocytes: a clue to linear scleroderma *en coup de sabre*.
J Cutan Pathol 2012; 39: 518–520. © 2012 John Wiley & Sons A/S.

Carolyn Goh[1], Asok Biswas[2] and Lynne J. Goldberg[2]

[1]Division of Dermatology, David Geffen School of Medicine, University of California, Los Angeles, CA, USA and
[2]Dermatopathology Section, Department of Dermatology, Boston University School of Medicine, Boston, MA, USA

Lynne J. Goldberg, MD
Department of Dermatology
Boston University School of Medicine
609 Albany Street
Boston, MA 02118
Tel: +1 617 638 5574
Fax: +1 617 638 5515
e-mail: lynngold@bu.edu

Accepted for publication February 2, 2012

Morphea Alopecia *en coup de sabre*:
A.Early/Inflammatory: perineural inflammation clue as a precursor
B. Late/non-inflammatory/sclerotic

Fig. 1. Ill defined area of hair loss on the right scalp

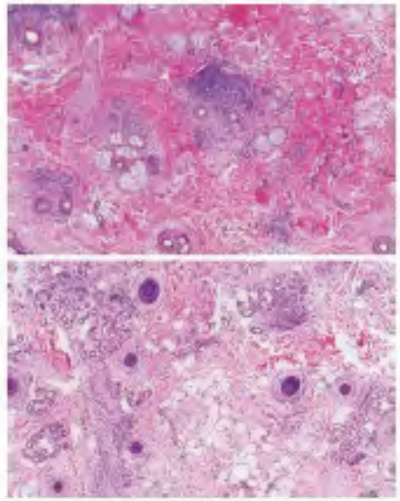


Fig. 3. Transverse sections at the level of the isthmus (top) and dermal/subcutaneous border (bottom). Note the decrease in follicular size, the presence of fibrous tracts deep, and deep inflammation around a nerve (bottom, lower left).

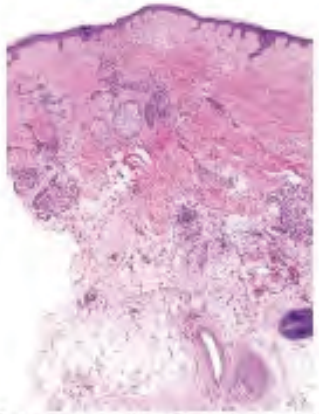


Fig. 2. Low power view revealing a superficial and deep perivascular and perineural (lower right) infiltrate and a decreased number of hair follicles

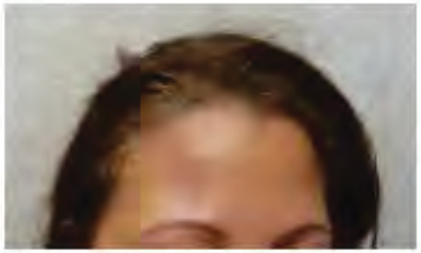


Fig. 4. Linear depression on the right forehead anterior to the alopecia, noticed at the follow-up visit

subdivided into linear lesions of the limbs and trunk, *en coup de sabre* typically involving the forehead

J Cutan Pathol 2013: 40: 580–584
doi: 10.1111/cup.12124
John Wiley & Sons, Printed in Singapore

Journal of
Cutaneous Pathology

Distinctive histopathologic findings in linear morphea (*en coup de sabre*) alopecia

Margareth Pierre-Louis[1], Leonard C. Sperling[2], Mark S. Wilke[1] and Maria K. Hordinsky[1]

[1]Department of Dermatology, University of Minnesota Medical School, Minneapolis, MN, USA and
[2]HCT Dermatopathology Services, Baltimore, MD, USA

Linear morphea *en coup de sabre* is a localized form of morphea that presents as paramedian face or frontal scalp depression. The histopathology of alopecia in linear morphea is typically characterized by sclerosis and a reduction in the number of follicular units. We present a 26-year-old Caucasian female with a 1.5-year history of linear morphea and alopecia with unique atrophic follicular remnants on scalp biopsy. Transverse and vertical sections of biopsy specimens showed dense, dermal sclerosis with marked eccrine gland atrophy and replacement of much of the adipose by collagenous tissue. All sebaceous glands had disappeared, but erector pili muscles persisted. Numerous vertical, columnar and epithelial structures were present at the sites of formerly viable hair follicles. Transverse sections of these atrophic follicular remnants had a resemblance to telogen follicles but were microscopically different. The morphology of these follicular remnants indicates an end-stage process or permanent alopecia. Similar follicular remnants have been reported in chemotherapy-induced permanent alopecia but not in alopecia secondary to morphea or other cicatricial alopecias. We discuss the significance of these findings and their relationship to other forms of cicatricial or permanent alopecia based on the literature and case review.

Keywords: alopecia, atypical features, linear morphea, sclerosis

Pierre-Louis M, Sperling LC, Wilke MS, Hordinsky MK. Distinctive histopathologic findings in linear morphea (*en coup de sabre*) alopecia. J Cutan Pathol 2013; 40: 580–584. © 2013 John Wiley & Sons A/S. Published by John Wiley & Sons Ltd

Maria K. Hordinsky, MD,
Department of Dermatology, University of Minnesota Medical School, Minneapolis, MN, USA
Tel: 612-624-6678
Fax: 612-624-7484
e-mail: hordi001@umn.edu

Accepted for publication February 18, 2013

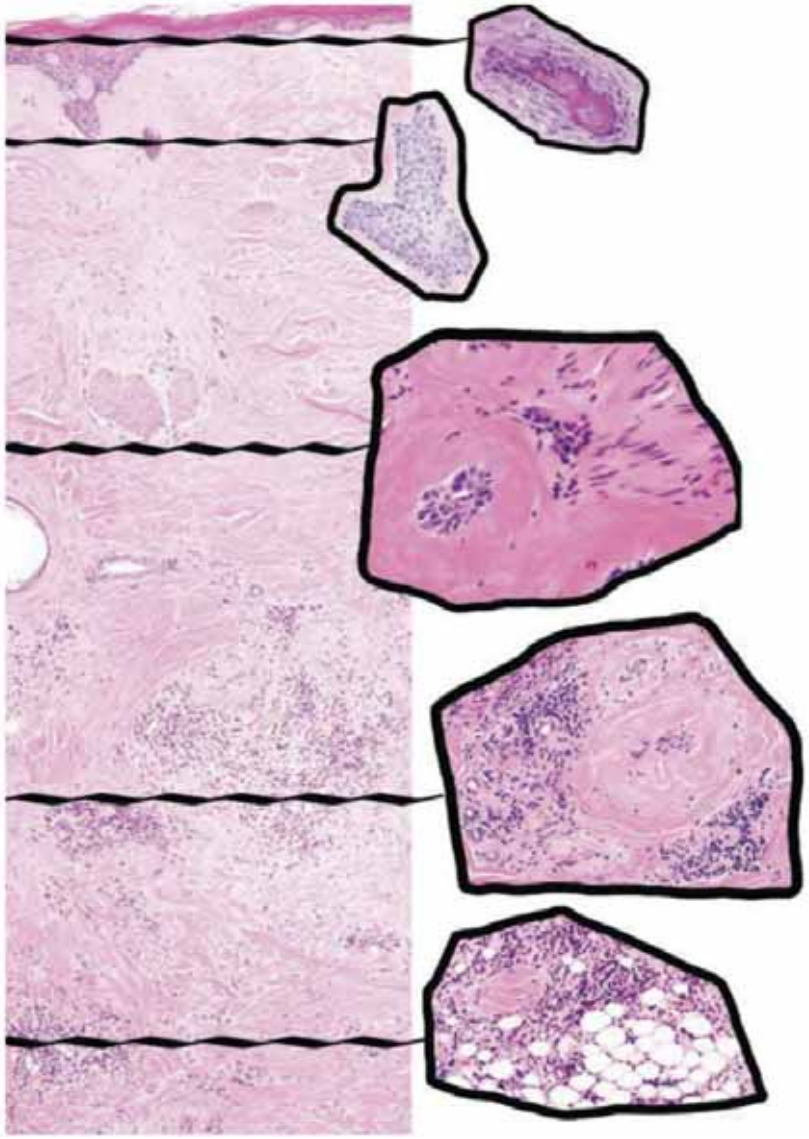
Fig. 1. There is an atrophic plaque with alopecia in linear distribution with hypopigmentation, erythema and superficial erosions at left frontal scalp to vertex.

adipose by collagenous tissue. Sebaceous glands were absent, eccrine glands were markedly atrophic and erector pili muscles were present (Fig. 2). Several vertically orientated columnar epithelial structures were identified at the sites of formerly viable hair follicles on transverse sections (Figs. 3 and 4). Foci of chronic (lymphocytes and plasma cells) inflammation were scattered at the level of the mid to lower dermis.

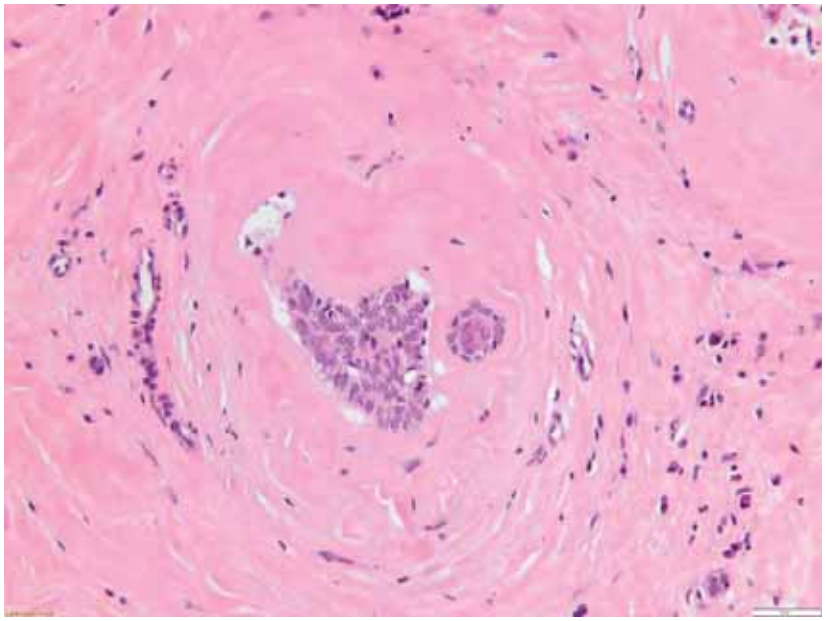
Extensive laboratory work-up was completed and was overall unremarkable. Magnetic resonance imaging of the head was completed to assess for

Fig. 2. Conventional vertical sections exhibit dense dermal sclerosis with marked eccrine gland atrophy and replacement of much of the adipose layer by collagenous tissue. All sebaceous glands are absent, but erector pili muscles persist.

Distinctive histopathologic findings in linear morphea alopecia

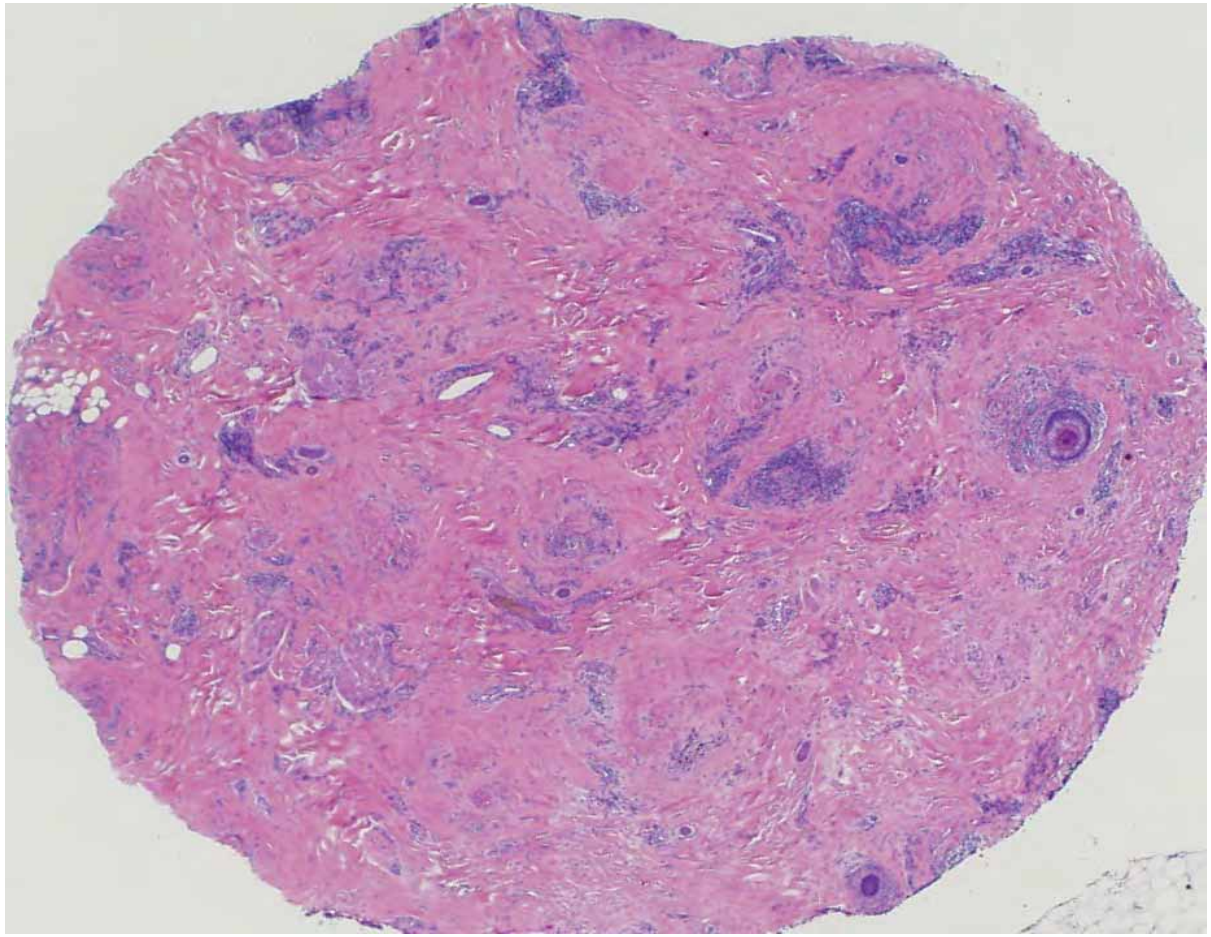


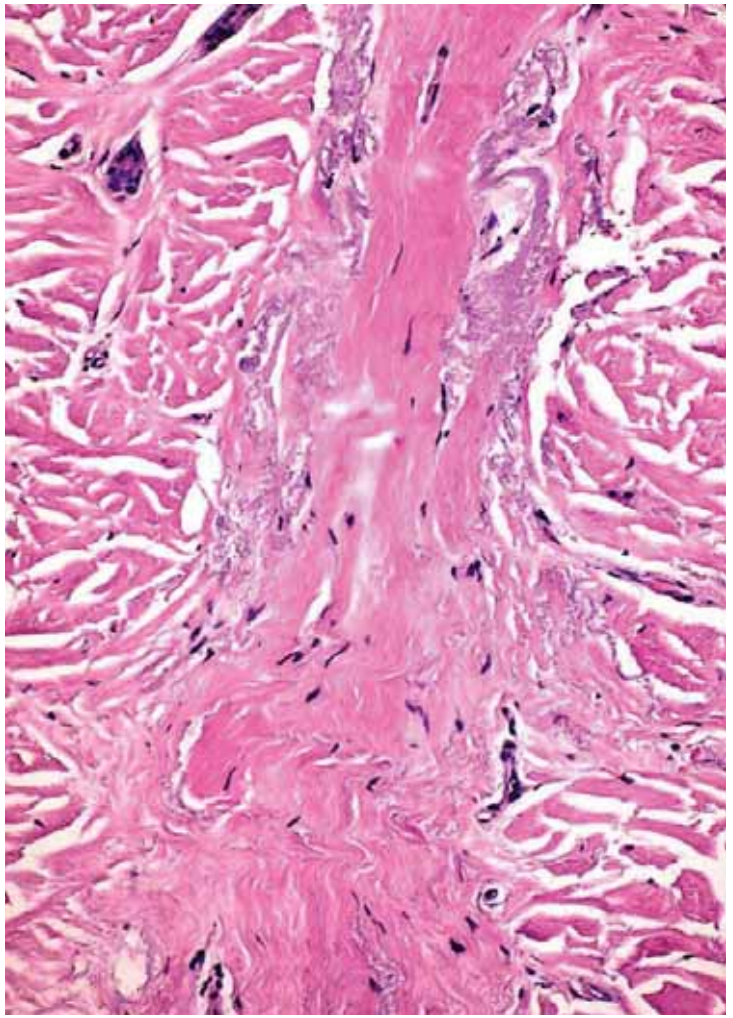
Histopathology



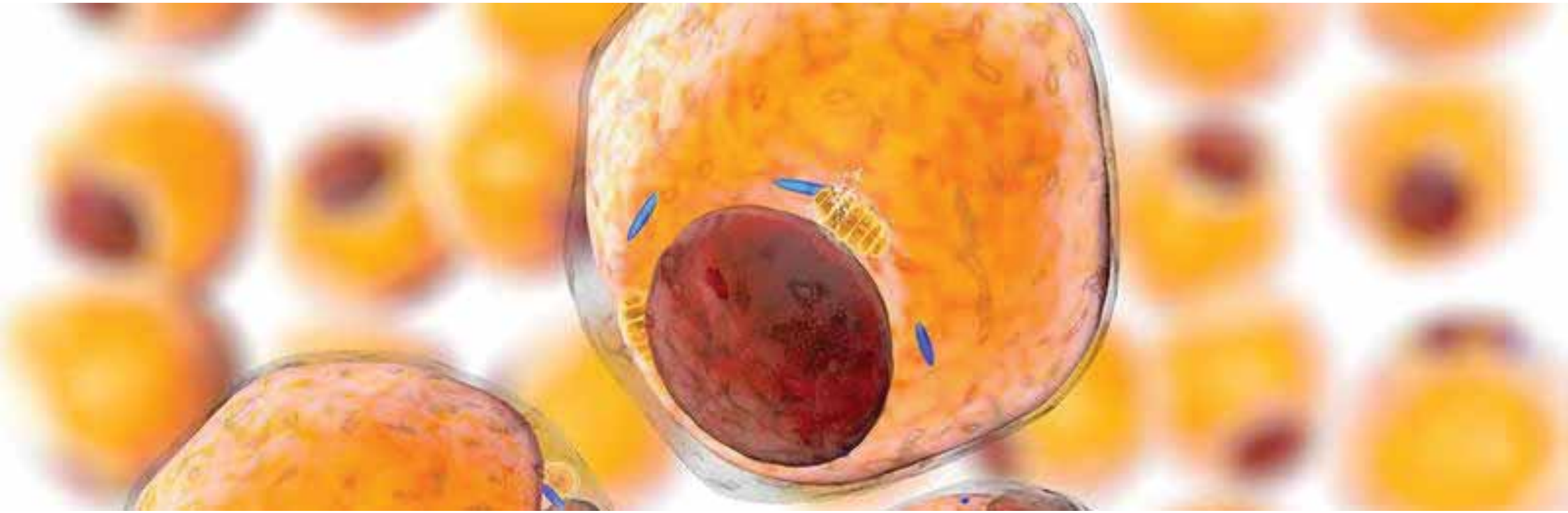
- Distinctive atrophic follicular structures first reported in cicatricial alopecia secondary to morphea in an adult female in 2013
- The unique structures resemble telogen follicles but have a distinctive morphology (lack of terminal hair differentiation) indicating non-viable follicular remnants)
- These structures most likely represent an end-stage process resulting in non-viable follicles compatible with a permanent alopecia
- Initially reported in permanent chemotherapy-induced alopecia in 2010

.....WHAT'S NEXT?





ADIPOCYTIC STAMINAL CELLS



s.i.tri.

Società Italiana di Tricologia e Chirurgia della Calvizie

Terapia autologa rigenerativa in tricologia

Dott. Piero Tesauro

Tecnica S&V: Innesto

Iniezione frazione cellulare

Espansione tessuto cicatriziale

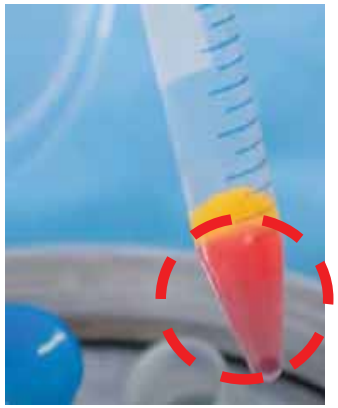
Distribuzione multistrato

Uniformità della frazione cellulare

Iniezione frazione volumetrica

Nel tessuto pre-espanso

Stem



Tecnica SeffiHair - Stem & Vol (S&V)

1. **Preparazione**
   Miglioramento qualità letto ricevente
2. **Rigenerazione**
   Utilizzo di componente vascolostromale
3. **Volumizzazione**
   Impiego di tessuti adiposi microfragmentati
4. **Trapianto**
   Miglior attecchimento dei follicoli