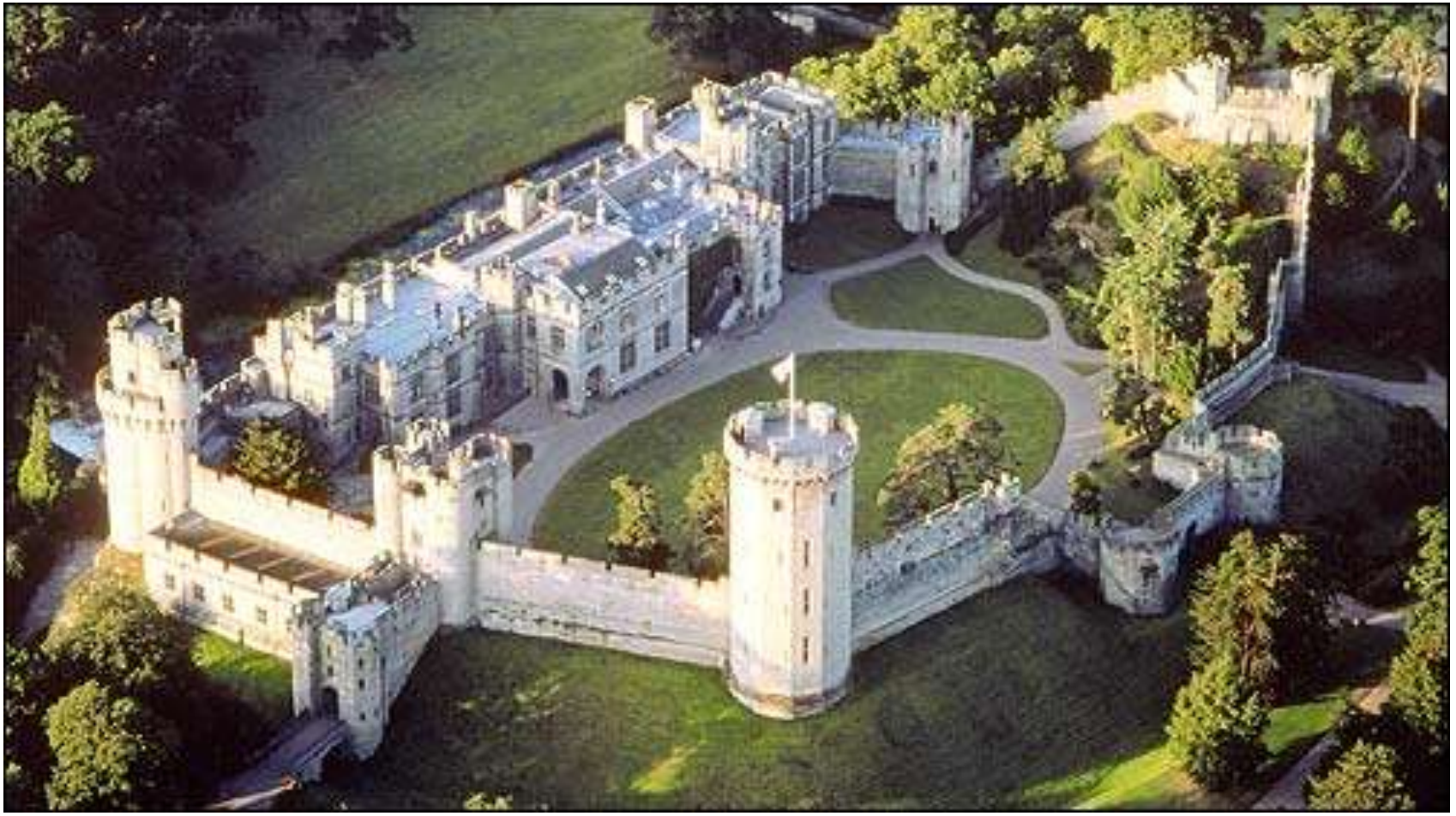
Interesting Cases from Referral Practice
Part 2: Adnexal Theme



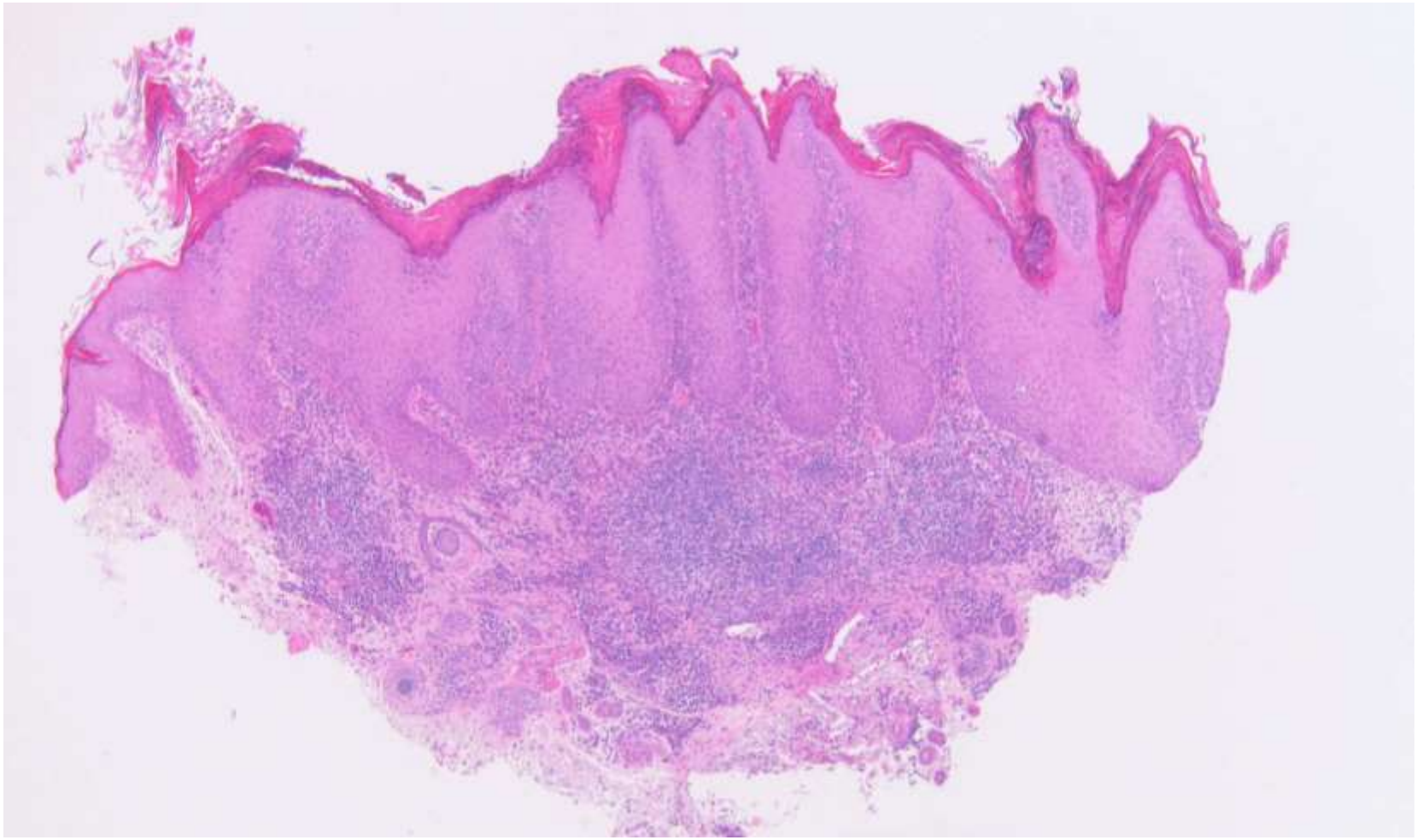
Richard Carr

Warwick Hospital

Rec'd: 20/06/2019

F88. A. Right Temple: keratotic nodule, red papular area, ?SCC

A



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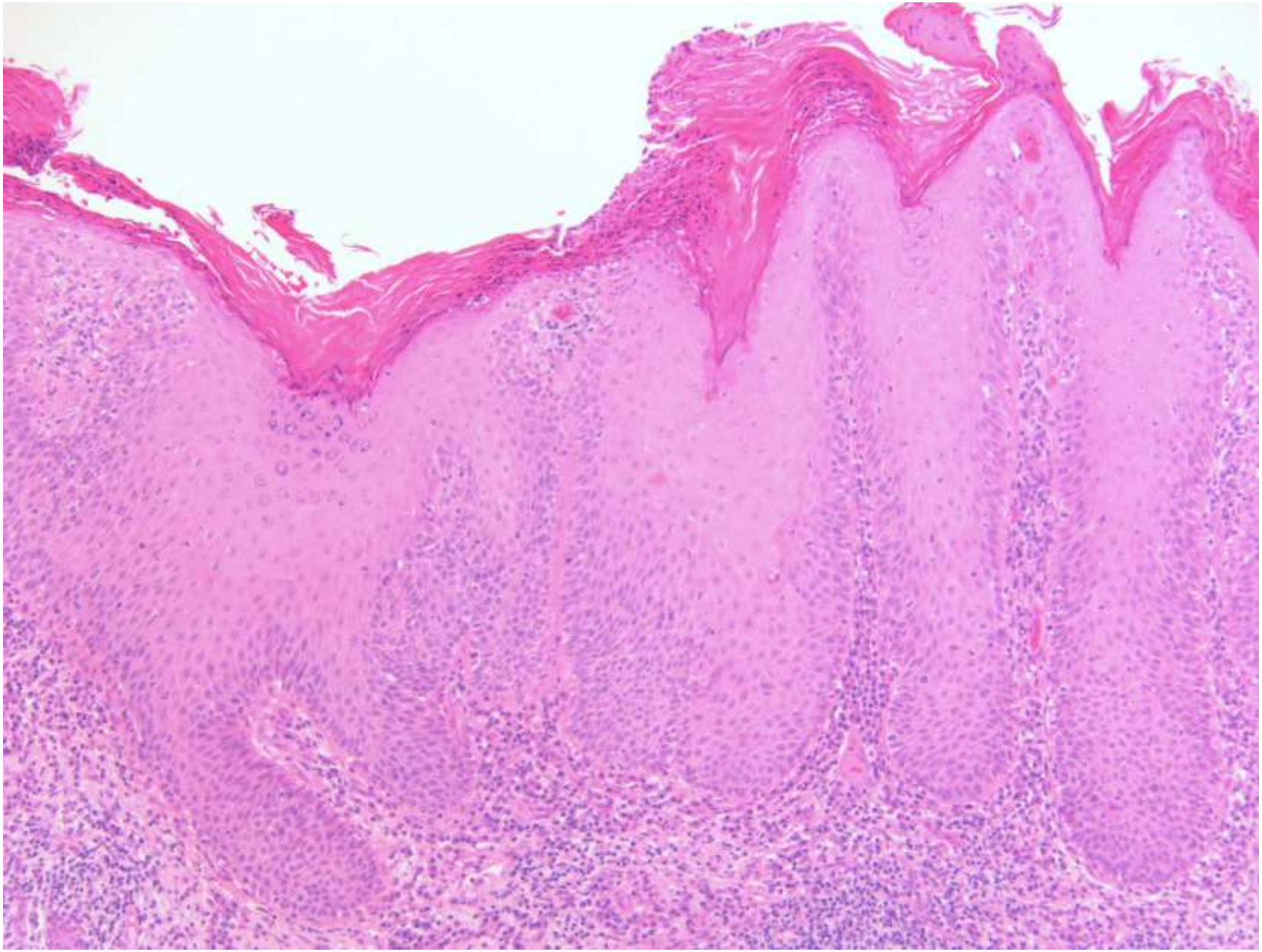
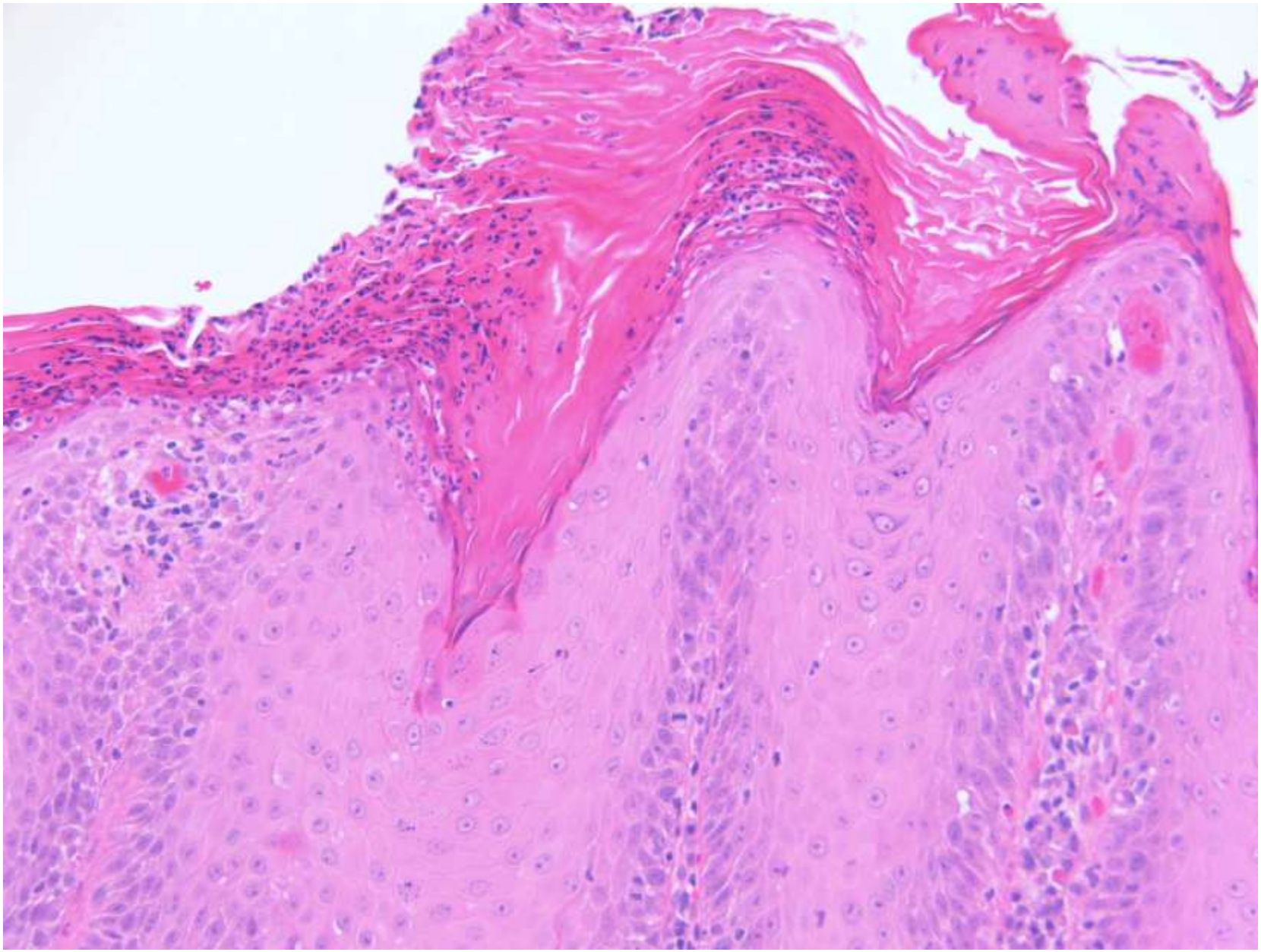
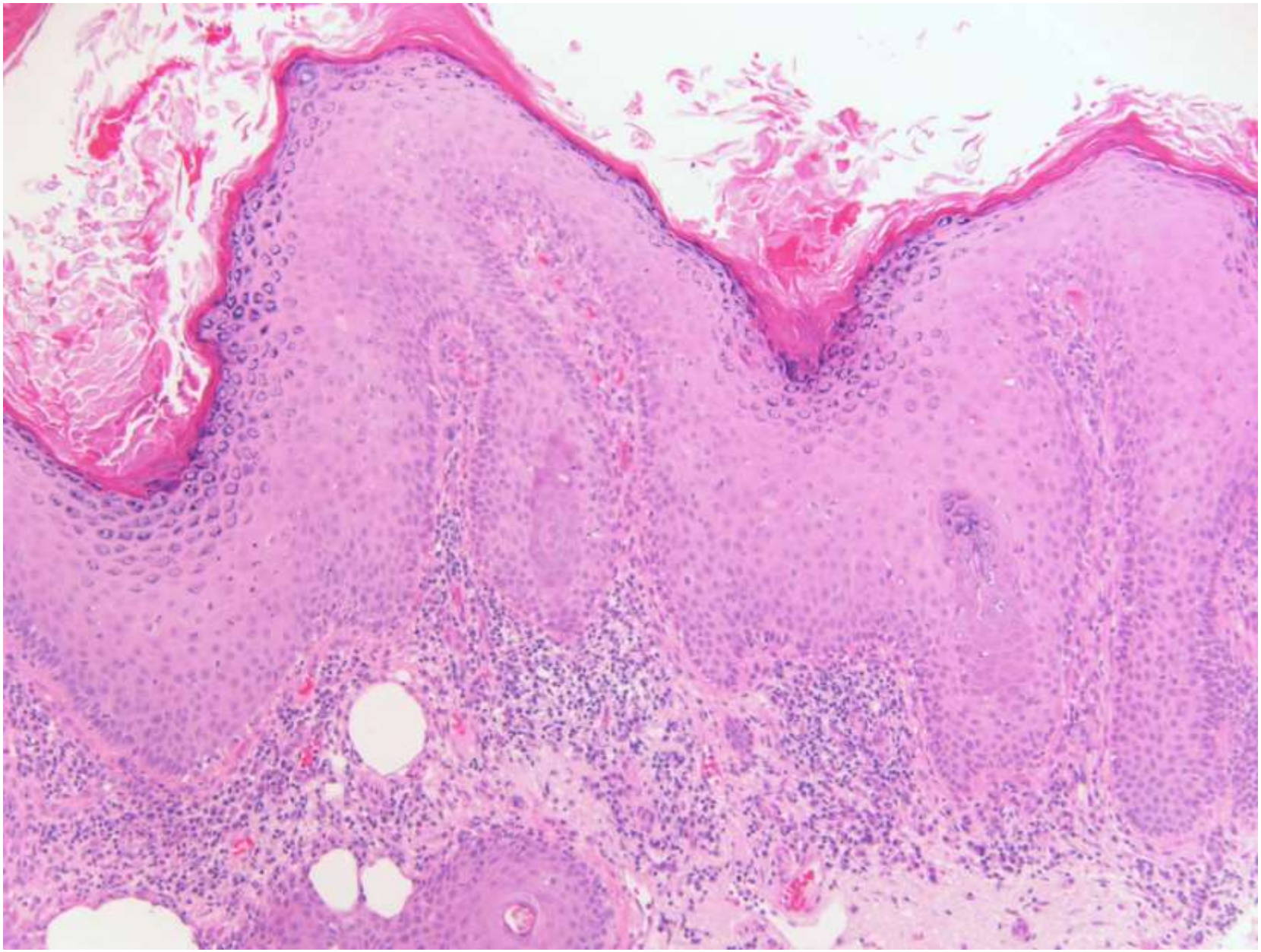
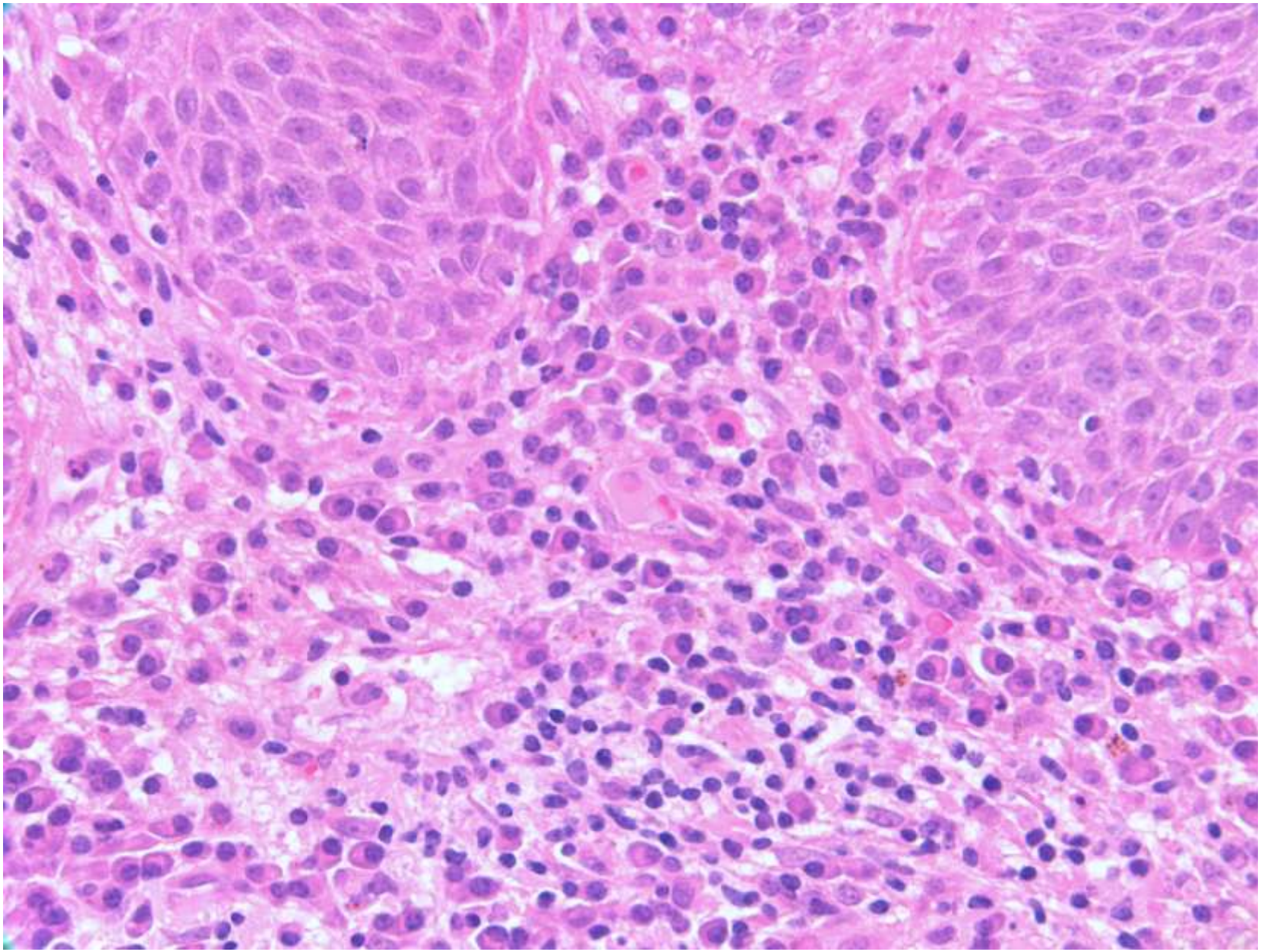
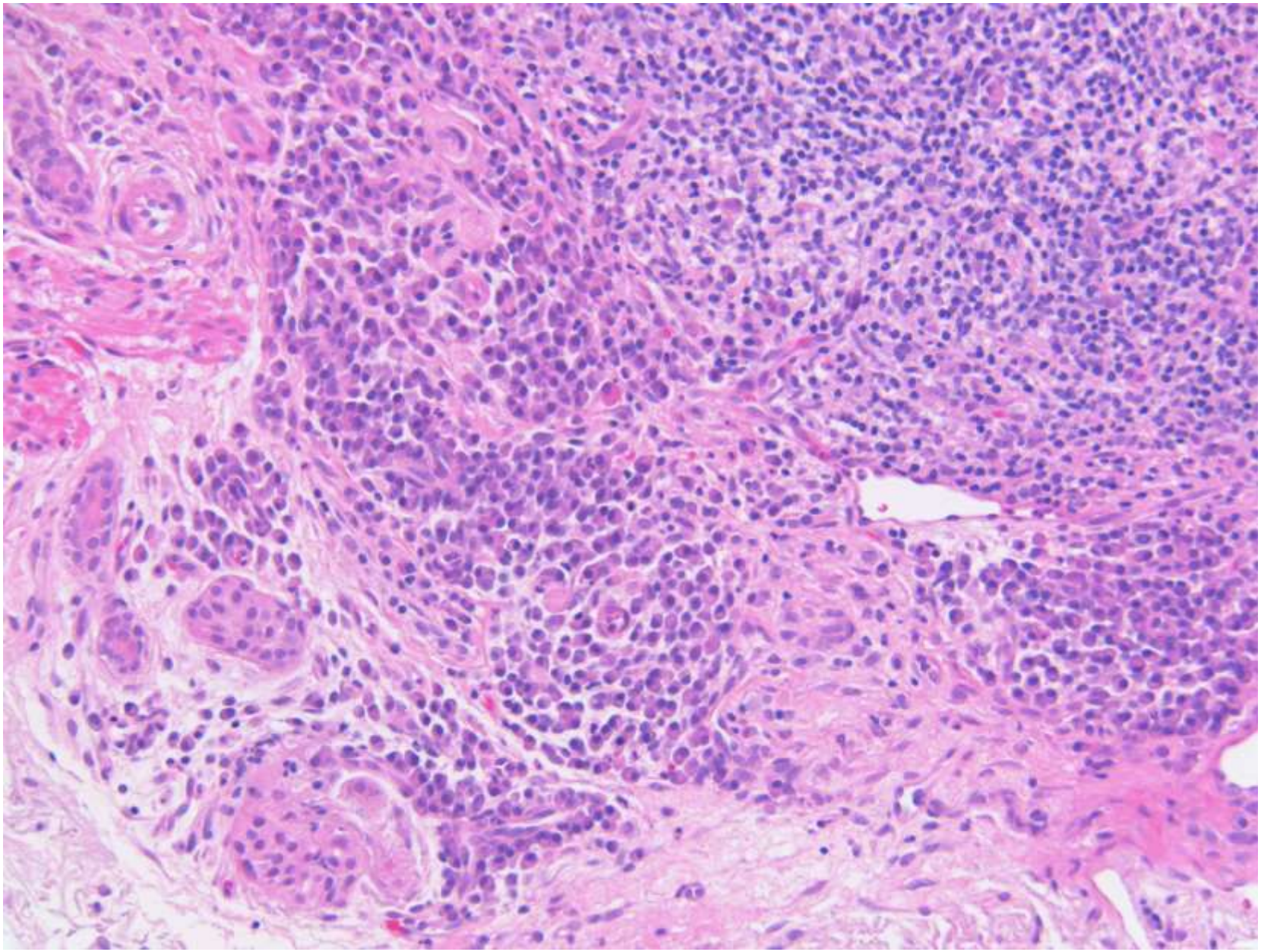
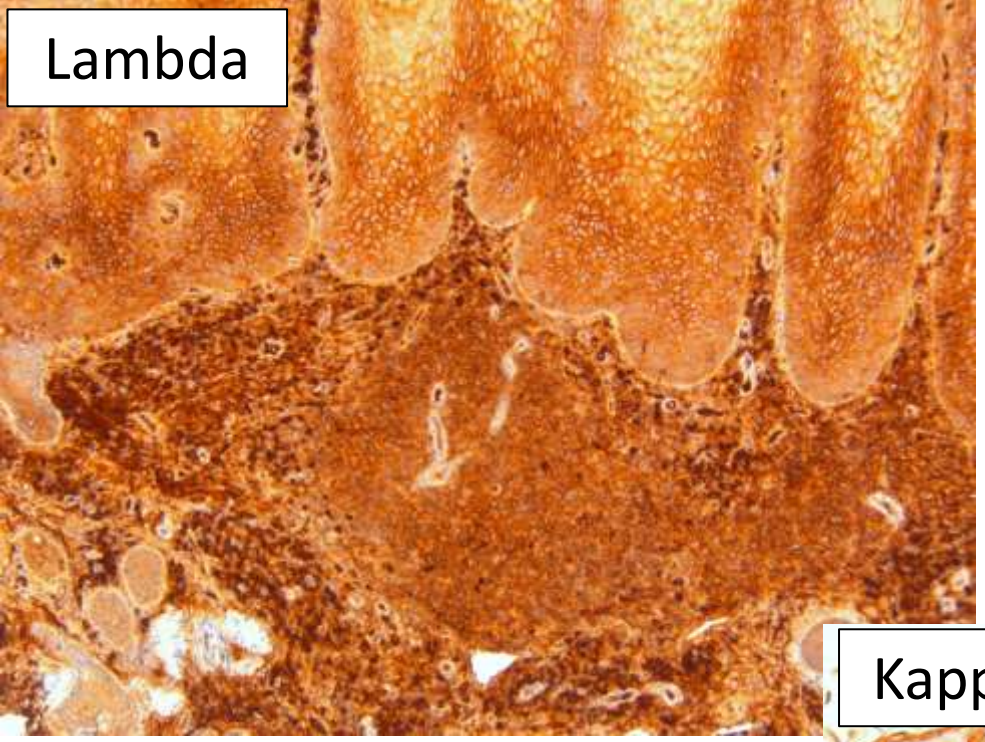


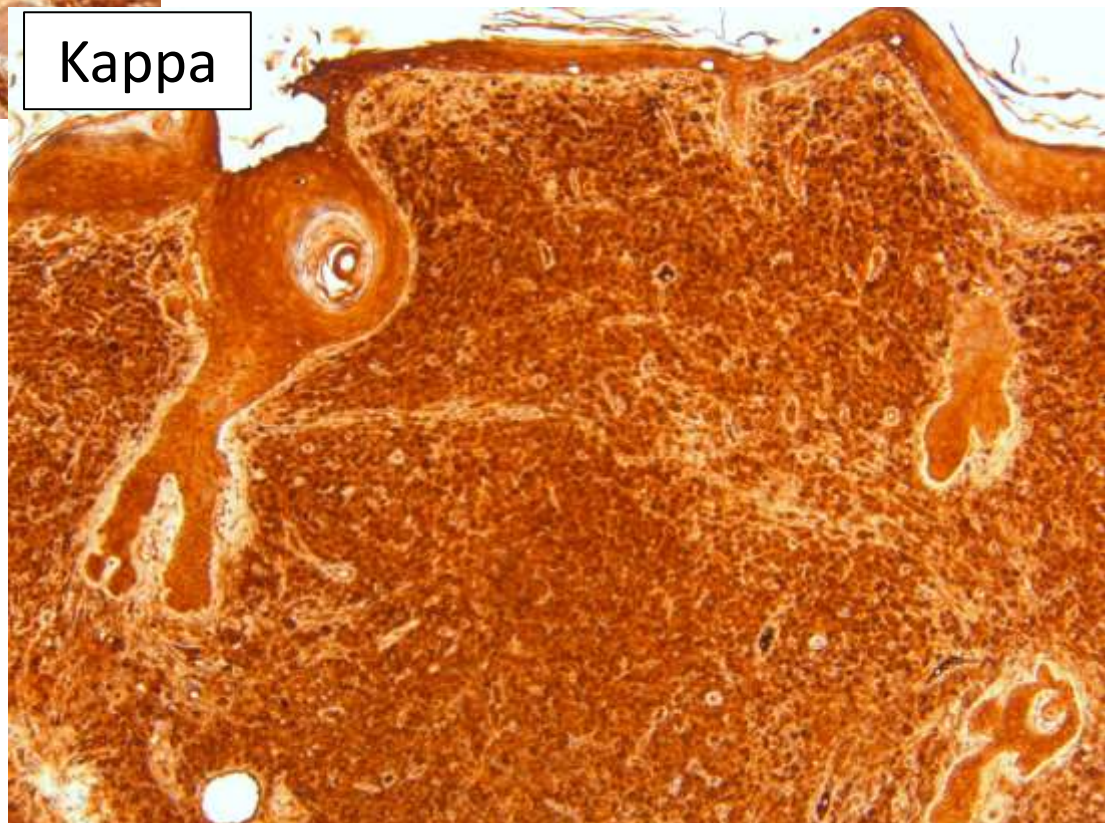


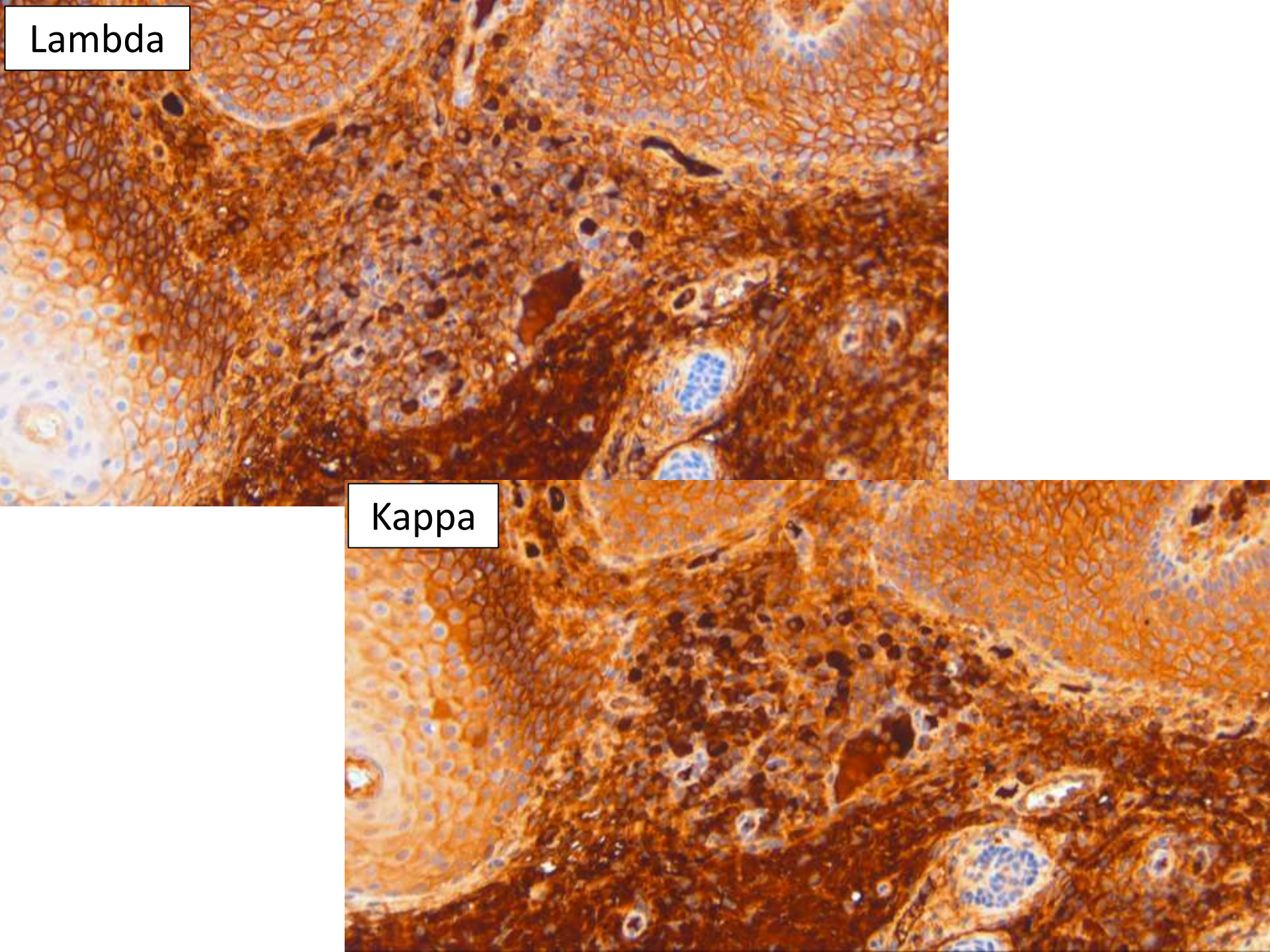




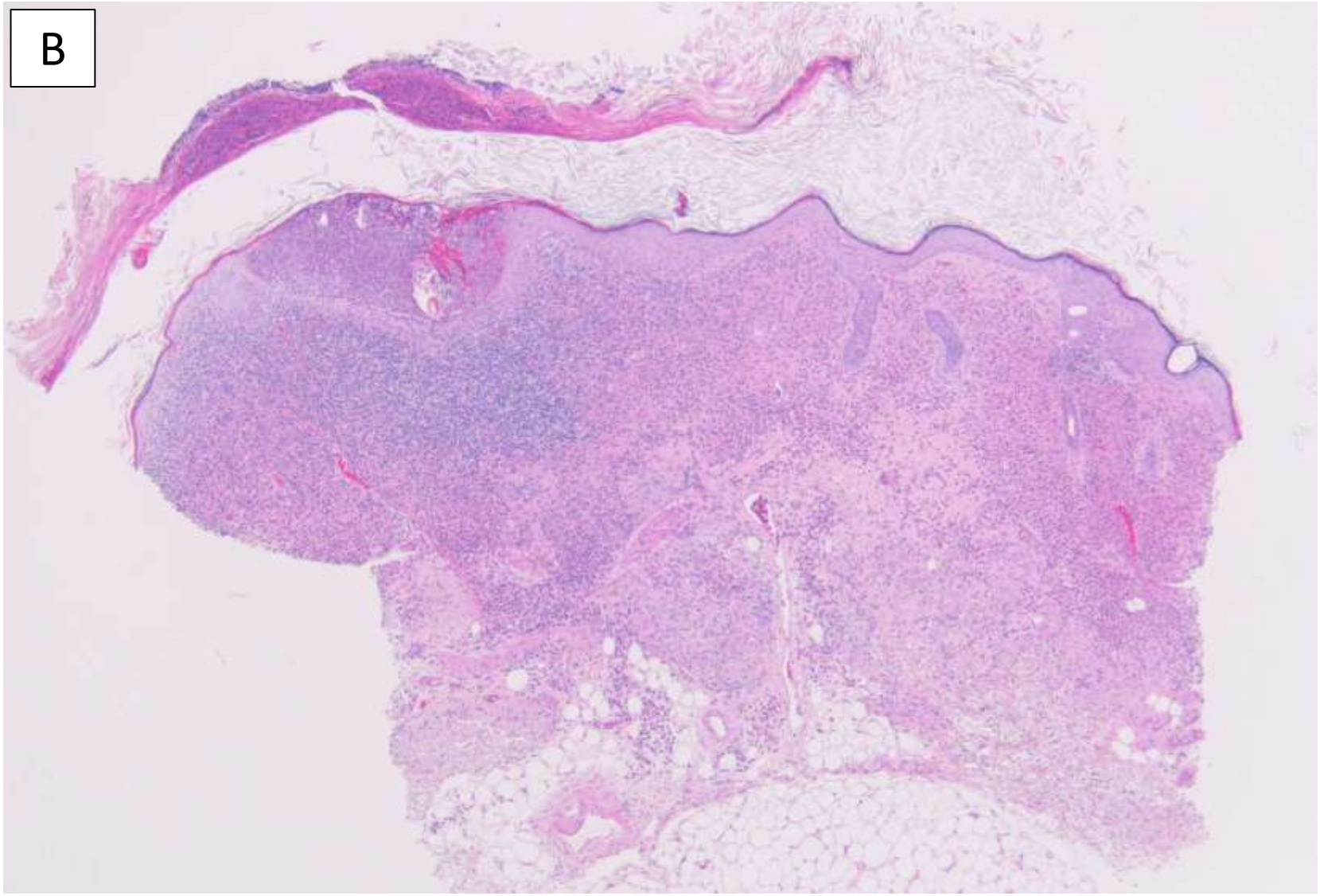


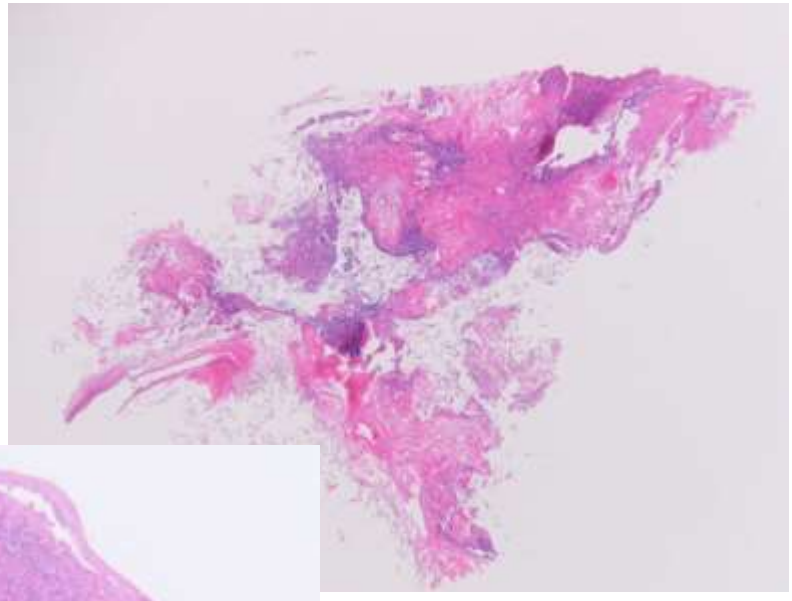

Lambda

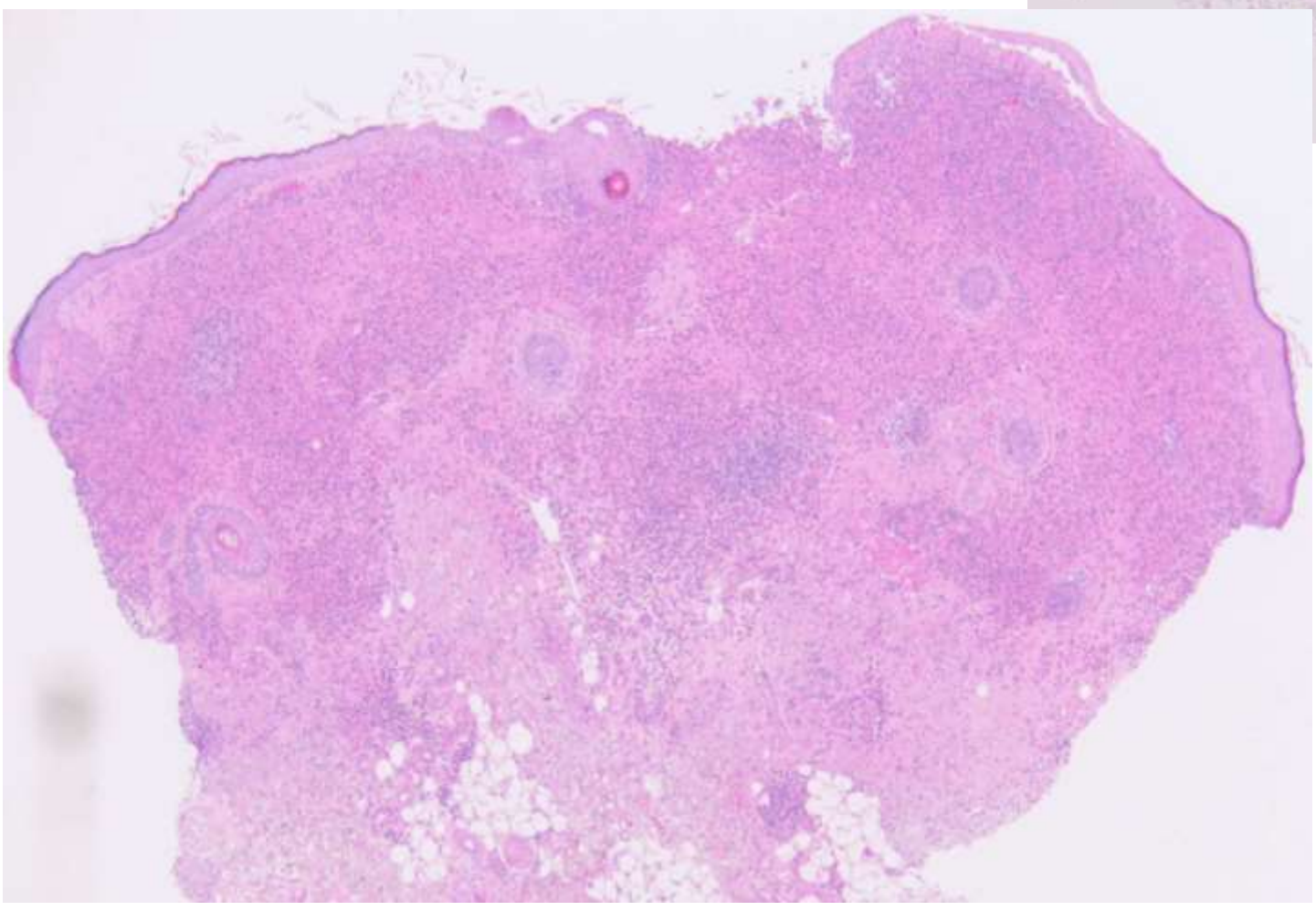

Kappa

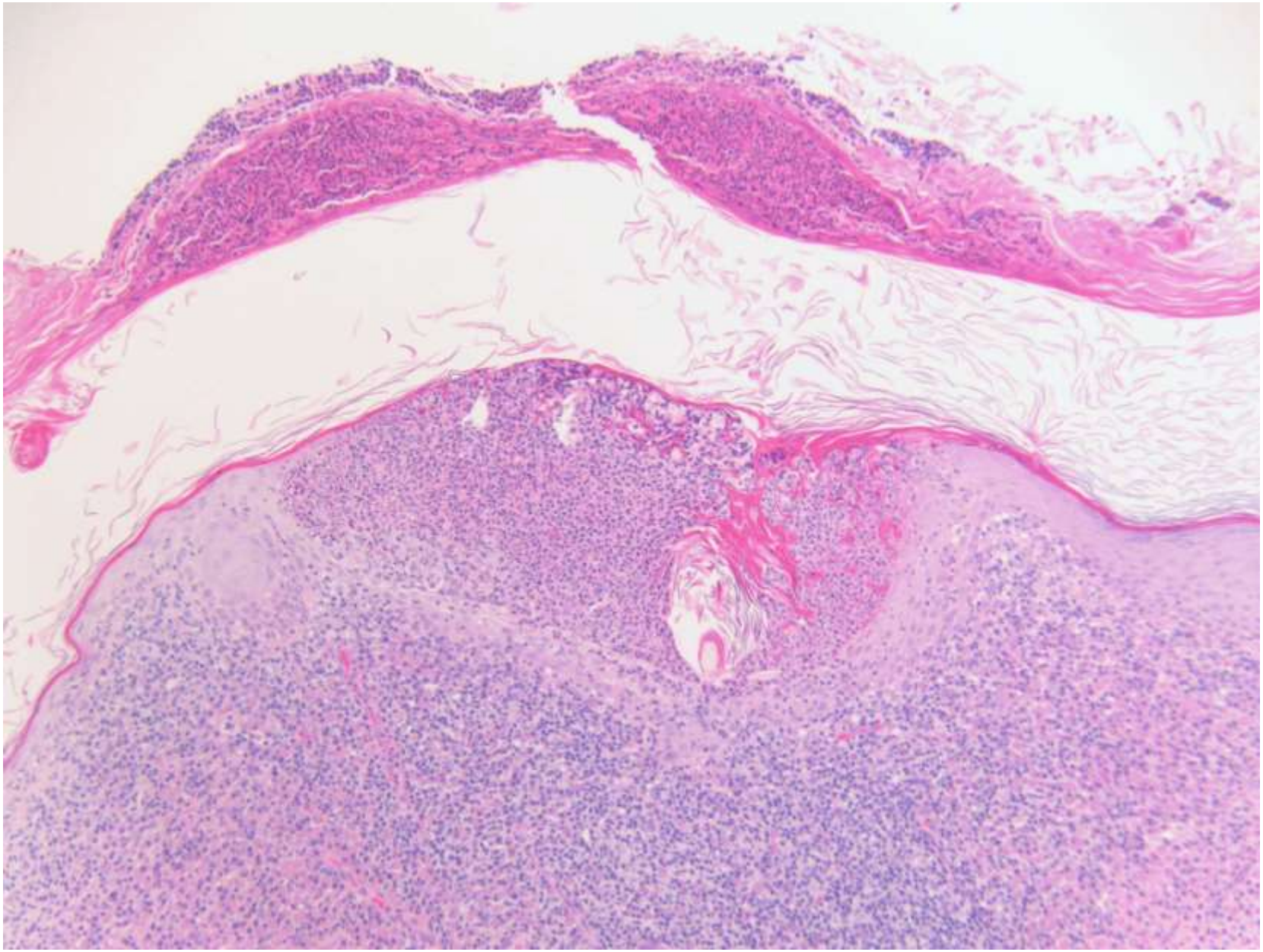

Lambda
Kappa

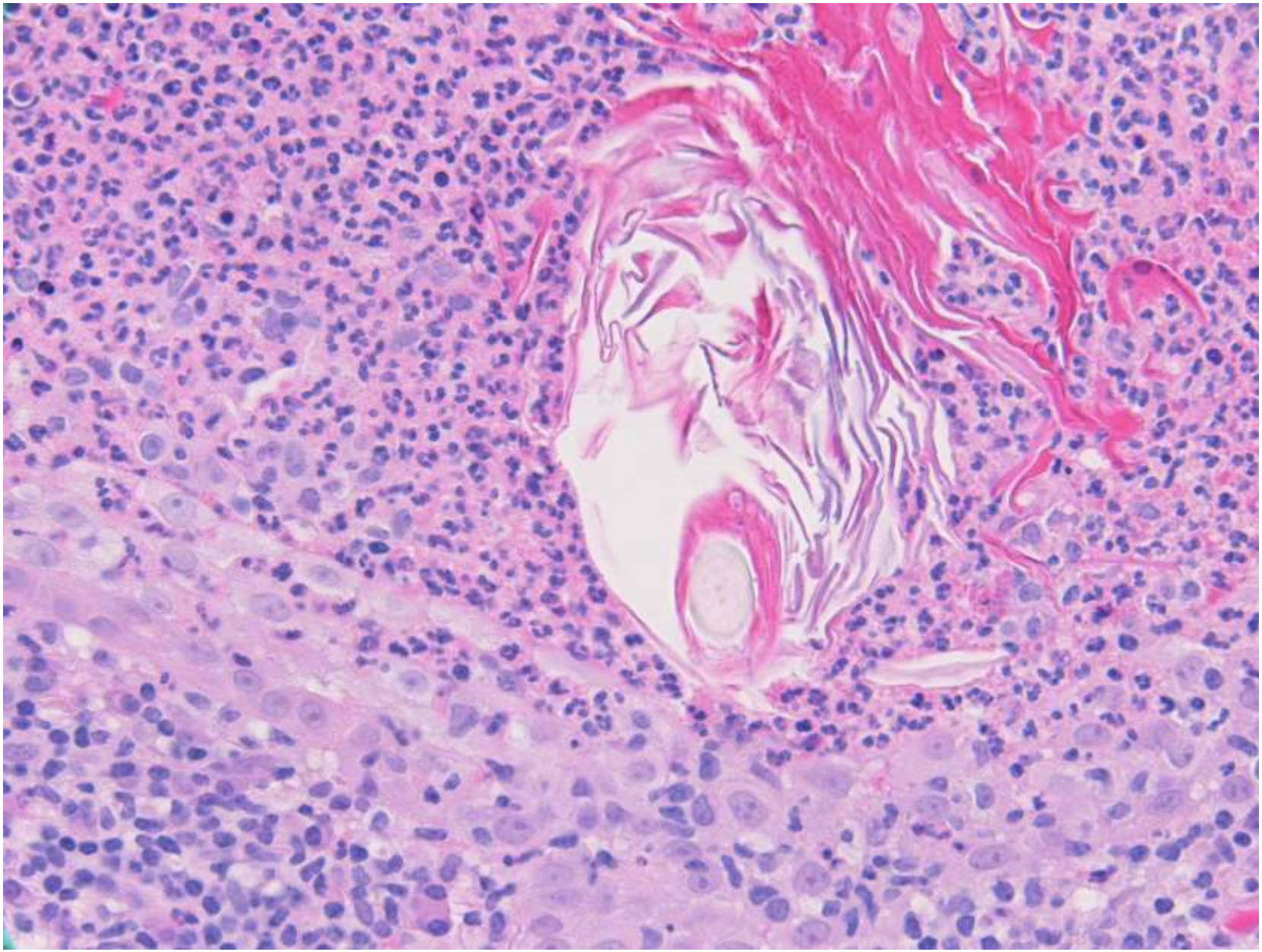
F88. B. Forehead. Crusty, red skin, 1 year AK

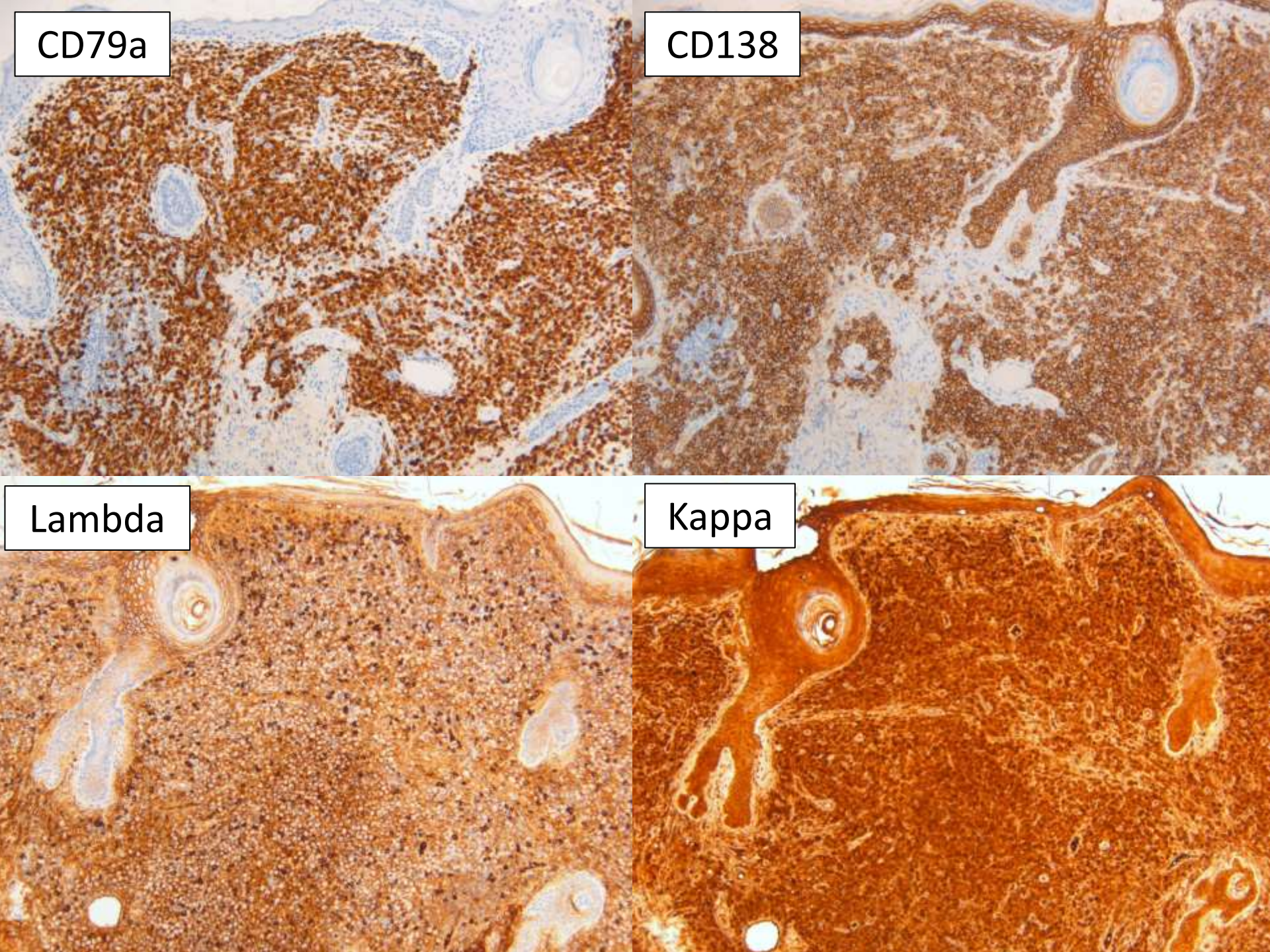


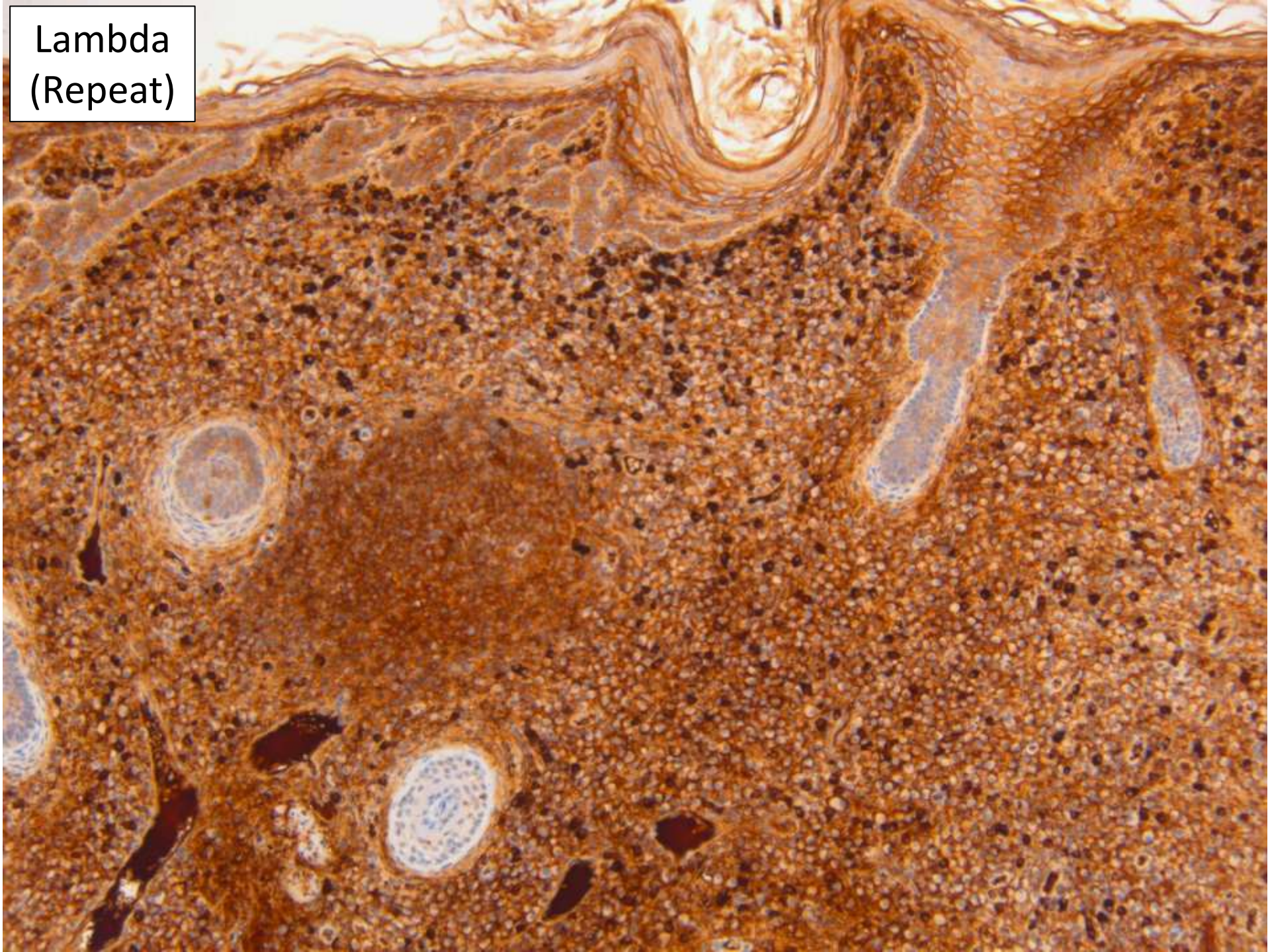


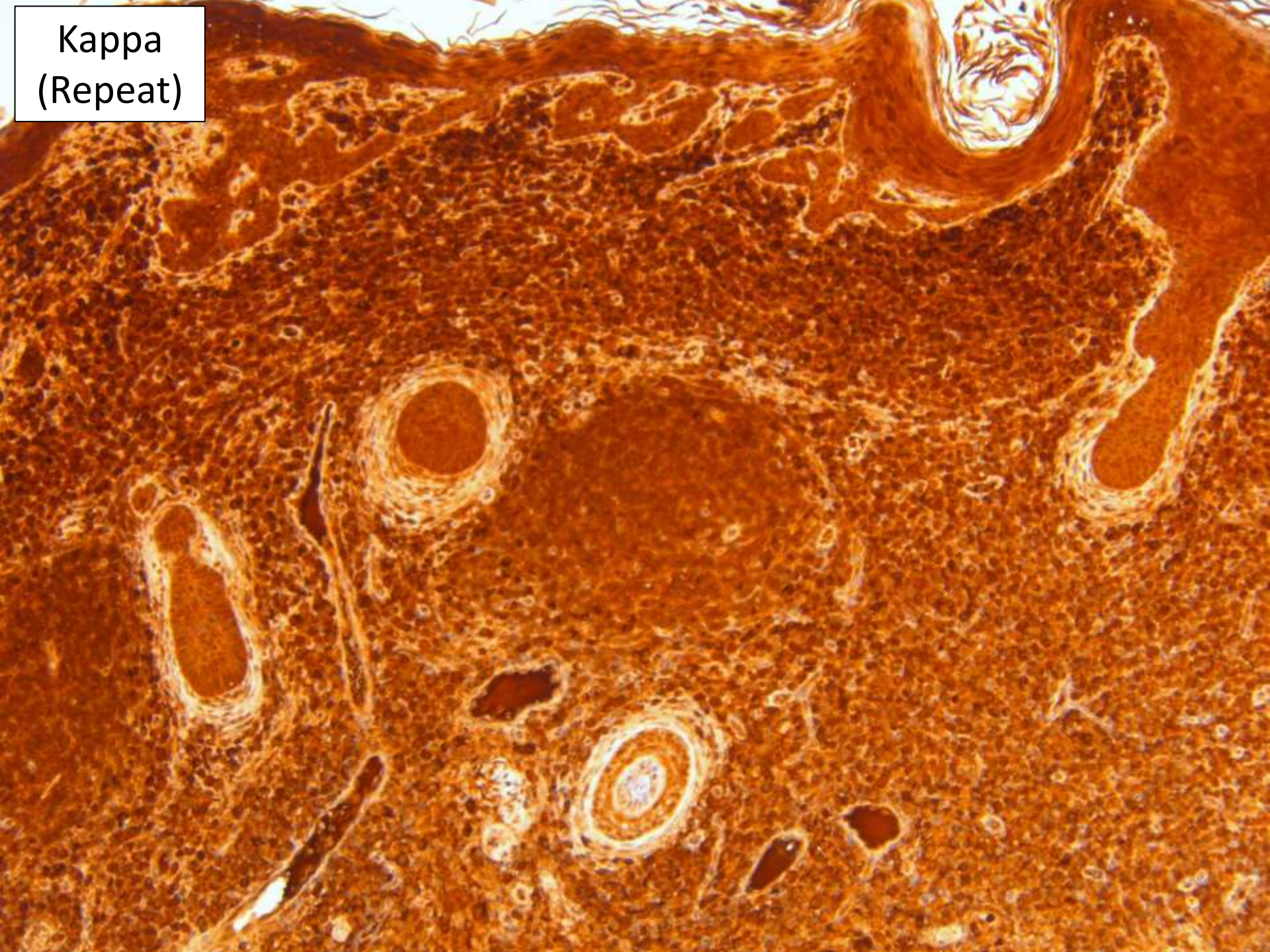







CD79a
CD138
Lambda
Kappa


Lambda
(Repeat)


Kappa
(Repeat)

A: There is no evidence of epithelial dysplasia or malignancy.
B: This sample shows hyperkeratotic skin with superficial acute inflammation. **The dermal infiltrate is almost purely plasma cells with relatively few background small B and T cells. A first attempt at light chain immunochemistry shows possible kappa exccess but the background** staining is heavy and this stain is being repeated. No epithelial malignancy is seen.
DIAGNOSIS:
The findings are unusual**. If the light chain immunochemistry again shows kappa restriction, the possibility of skin involvement by a plasma cell neoplasm would be raised**. Please await additional immunochemistry.
Reporting Pathologist:
XXXX

SUPPLEMENTARY REPORT 10/07/2019
Repeat light chain immunochemistry on **specimen B again shows an excess of Kappa restricted** forms. The possibility of a plasma cell neoplasm cannot be excluded. I will discuss the case with my Haematopathology colleague Dr YYYY and consider molecular tests. Does the patient have a paraprotein?

SUPPLEMENTARY REPORT 11/07/2019
I have reviewed this case with my colleague Dr YYYY. In specimen B the monotypic plasma cell proliferation **could either represent a plasma cell neoplasm or alternatively a low grade B-cell lymphoma of marginal zone type with extreme plasmacytic differentiation**. Please correlate closely with other findings.

CLINICAL DETAILS

Possible plasma cell neoplasm. Low grade B cell lymphoma diagnosed from forehead biopsy. Staging bone marrow biopsy.

A) : BONE MARROW TREPHINE

Two cores 7 and 10mm.

MICROSCOPY

This is an adequate specimen.

Cellularity: 20% (high normal).

Erythroid: Well-formed normoblastic islands seen

Myeloid: Present with good maturation. M:E ratio 2:1.

Megakaryocytes Normal number distribution and morphology.

Reticulin: Very focal fine fibre increase (Grade 1).

Infiltrates: Only 5% of cells are plasma cells with no overt light chain restriction. They are negative for CD56and Cylin D1. No increase in B cells is seen.

Bone trabeculae: Within normal limits.

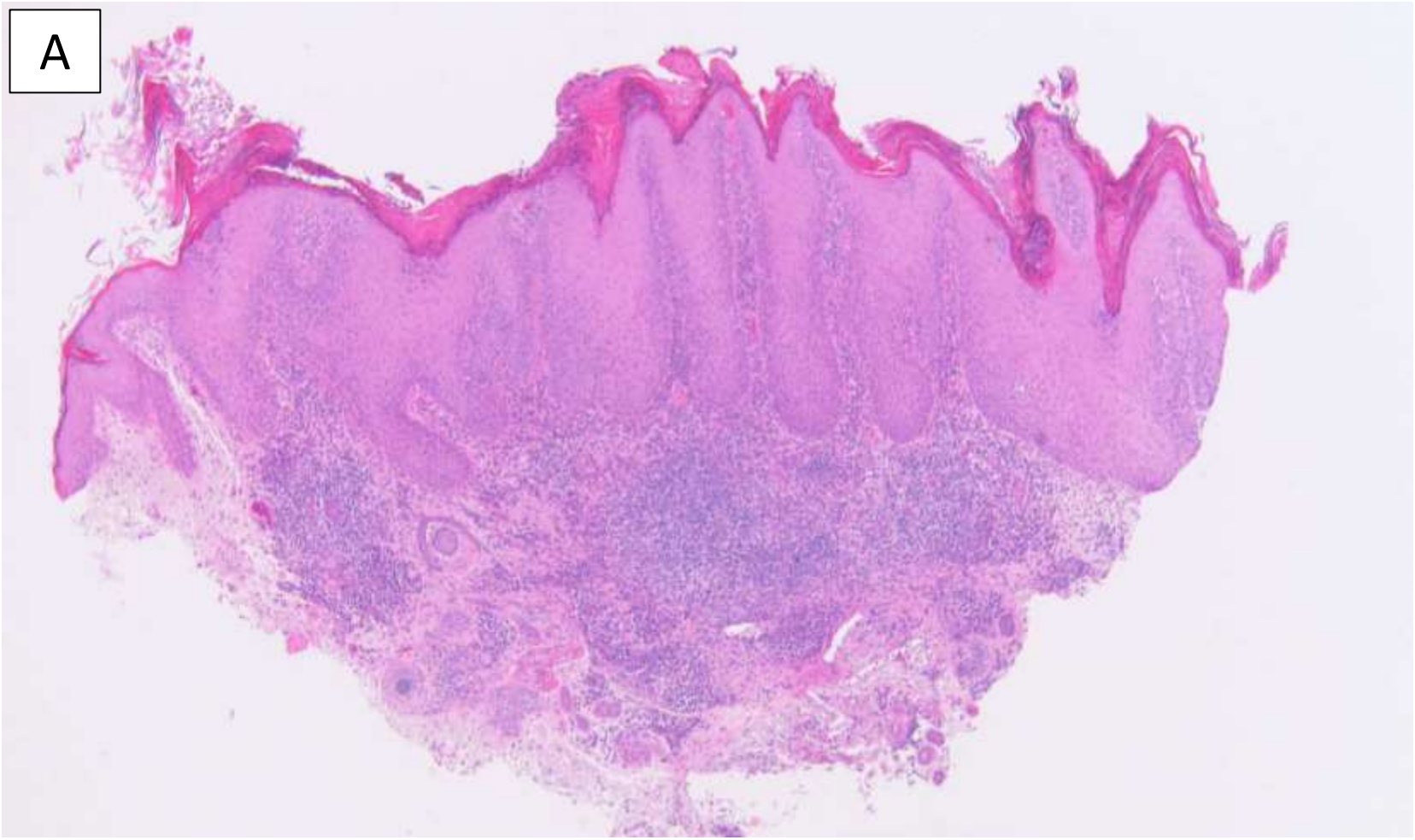
DIAGNOSIS:

Bone marrow: No evidence of a plasma cell neoplasm or lymphoma.

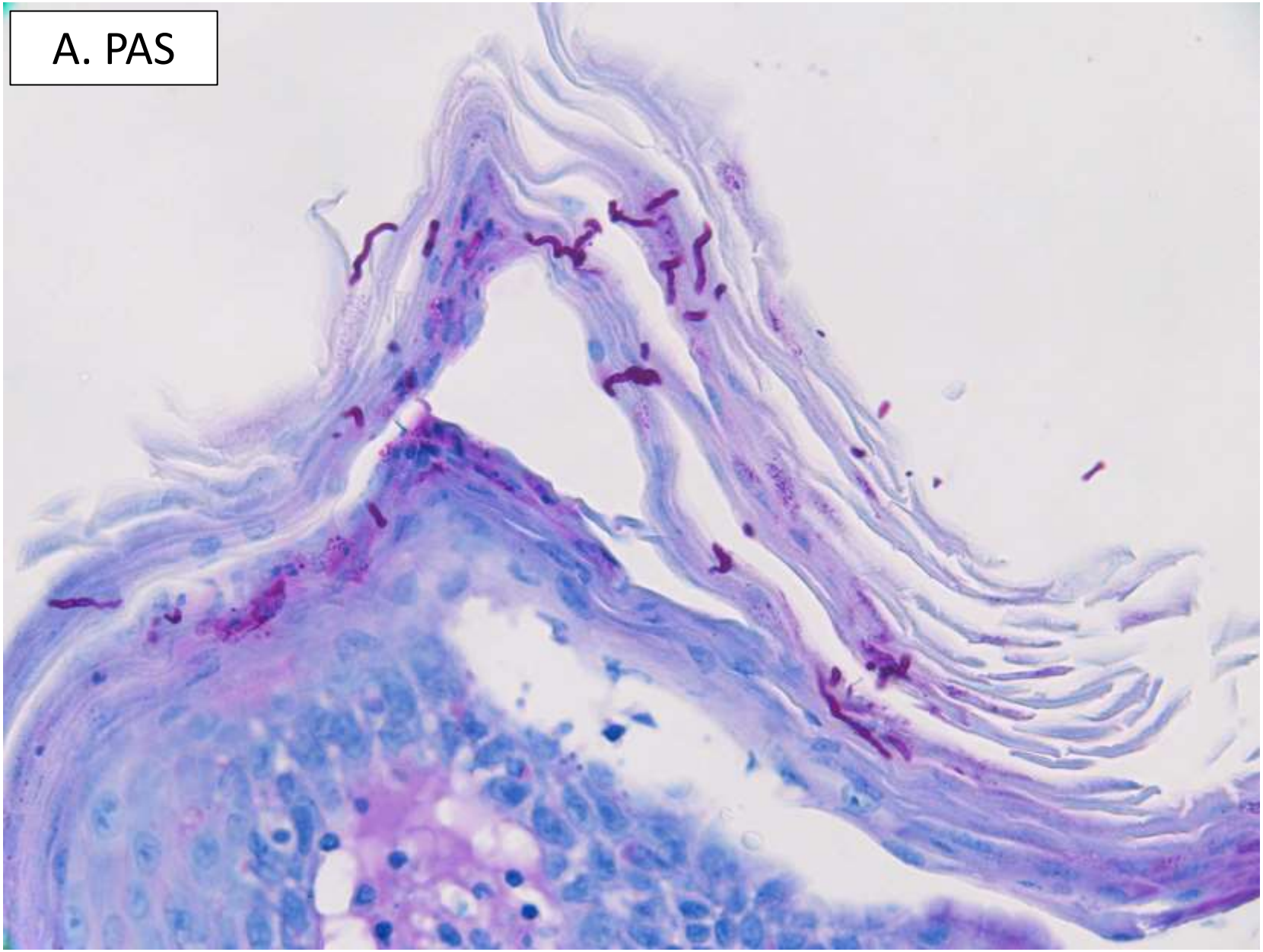
Reporting Pathologist: XXXX

AT HAEMPATH MDT THE HAEMATOLOGISTS SUGGESTED GETTNG A DERMATOPATHOLGOY OPINION

F88. A. Right Temple; A: keratotic nodule, red papular area, ?SCC

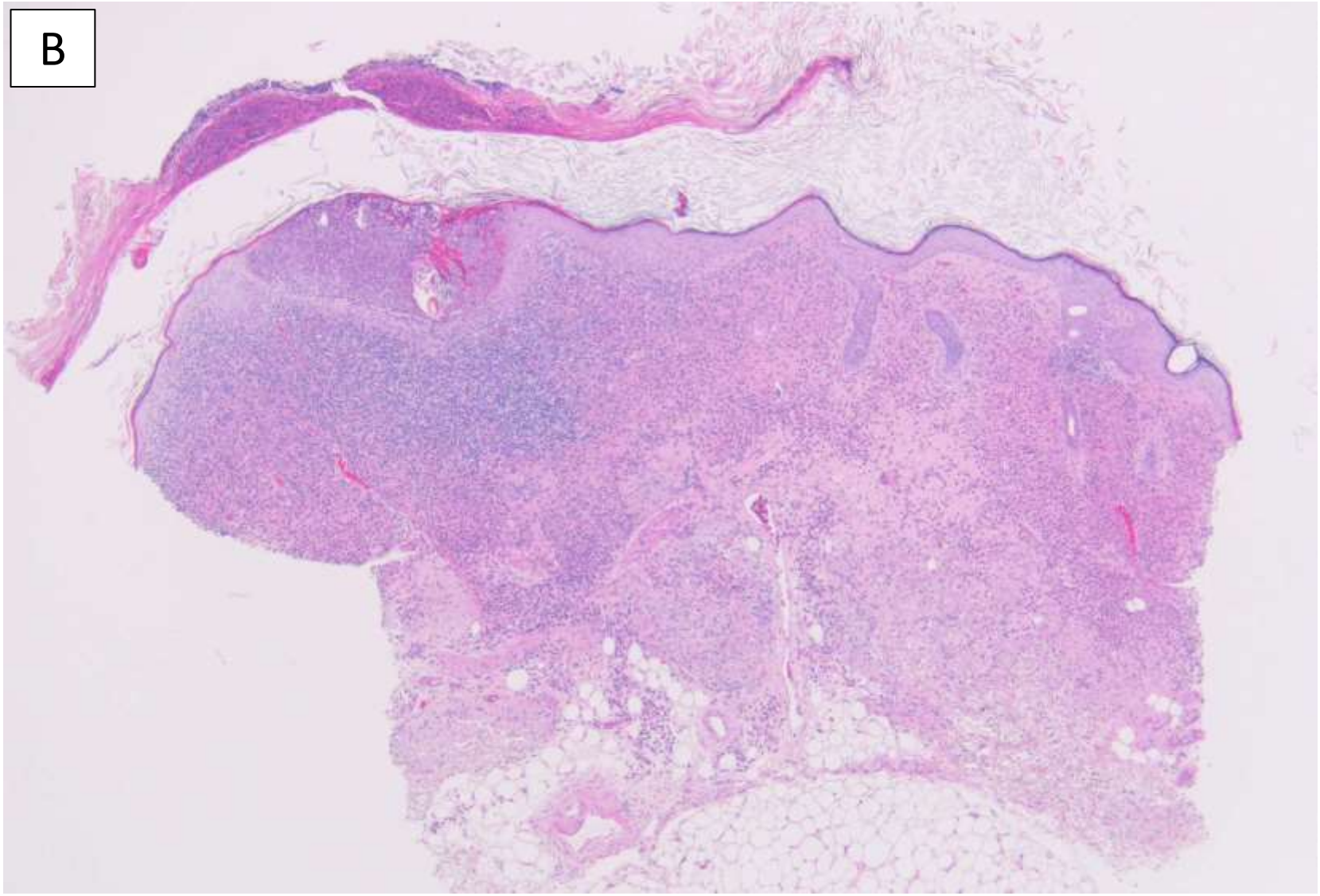


Psoriasiform, spotty parakeratosis, Munro-type microabscesses, dense upper dermal chronic inflammation rich in plasma cells



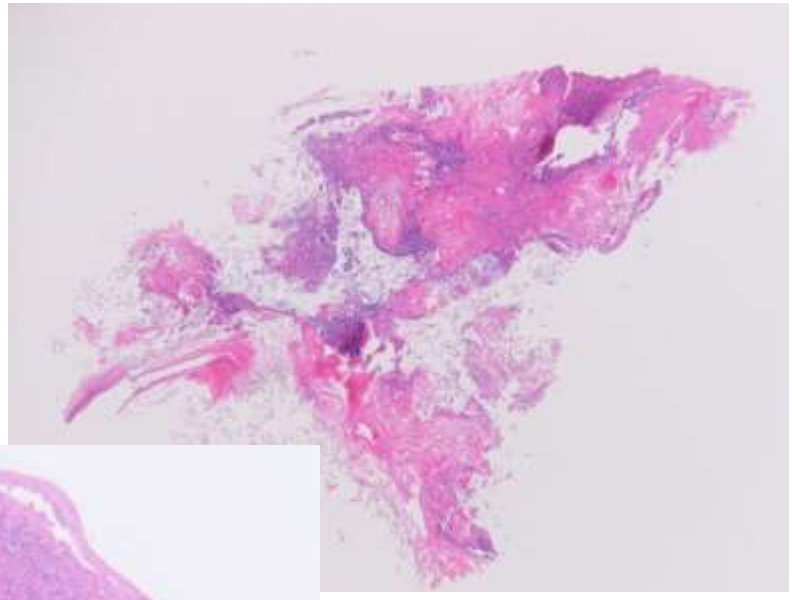
Fungal hyphae, some cross-sectoned and some fungal spores

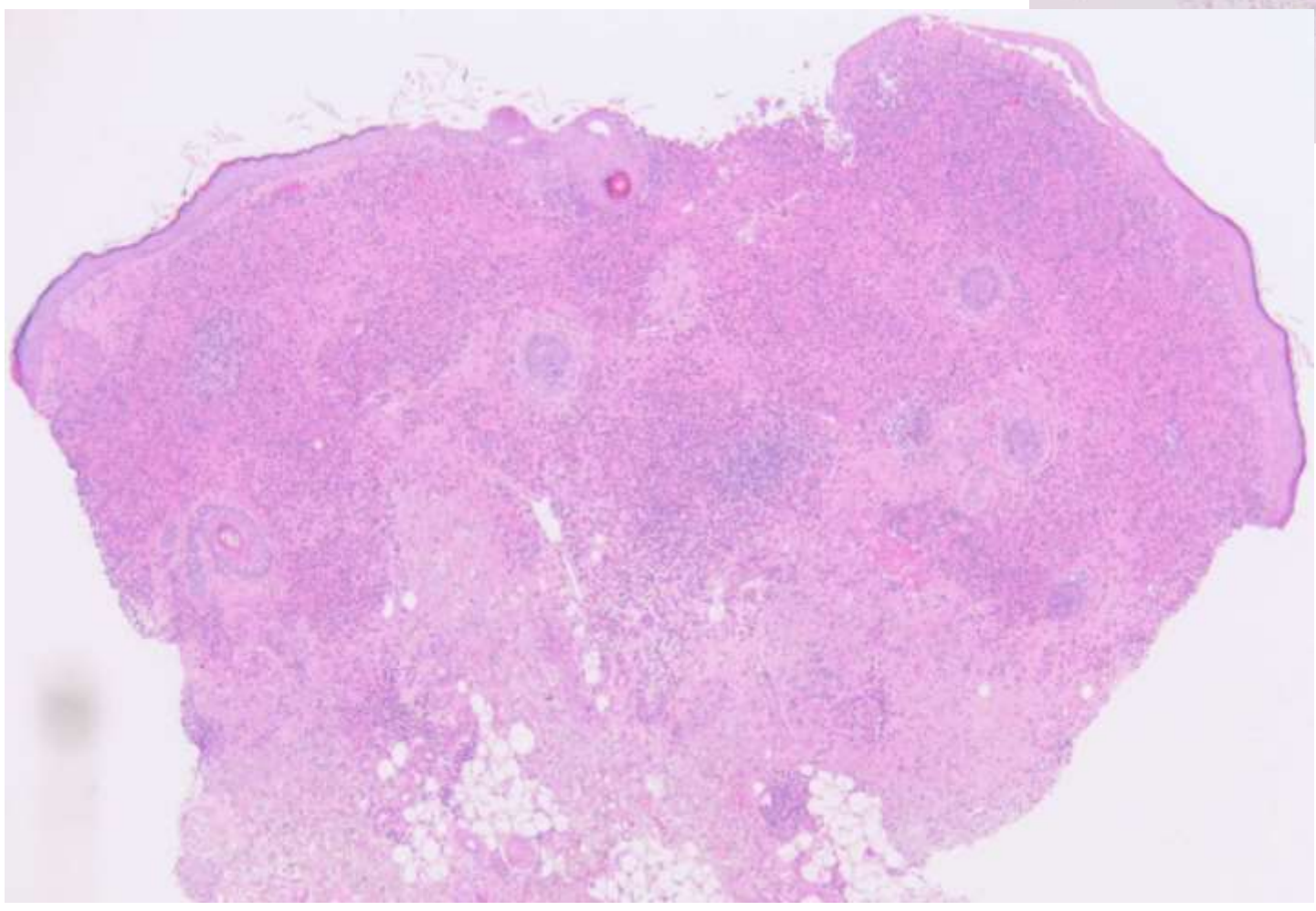
F88. B. Forehead. Crusty, red skin, 1 year AK

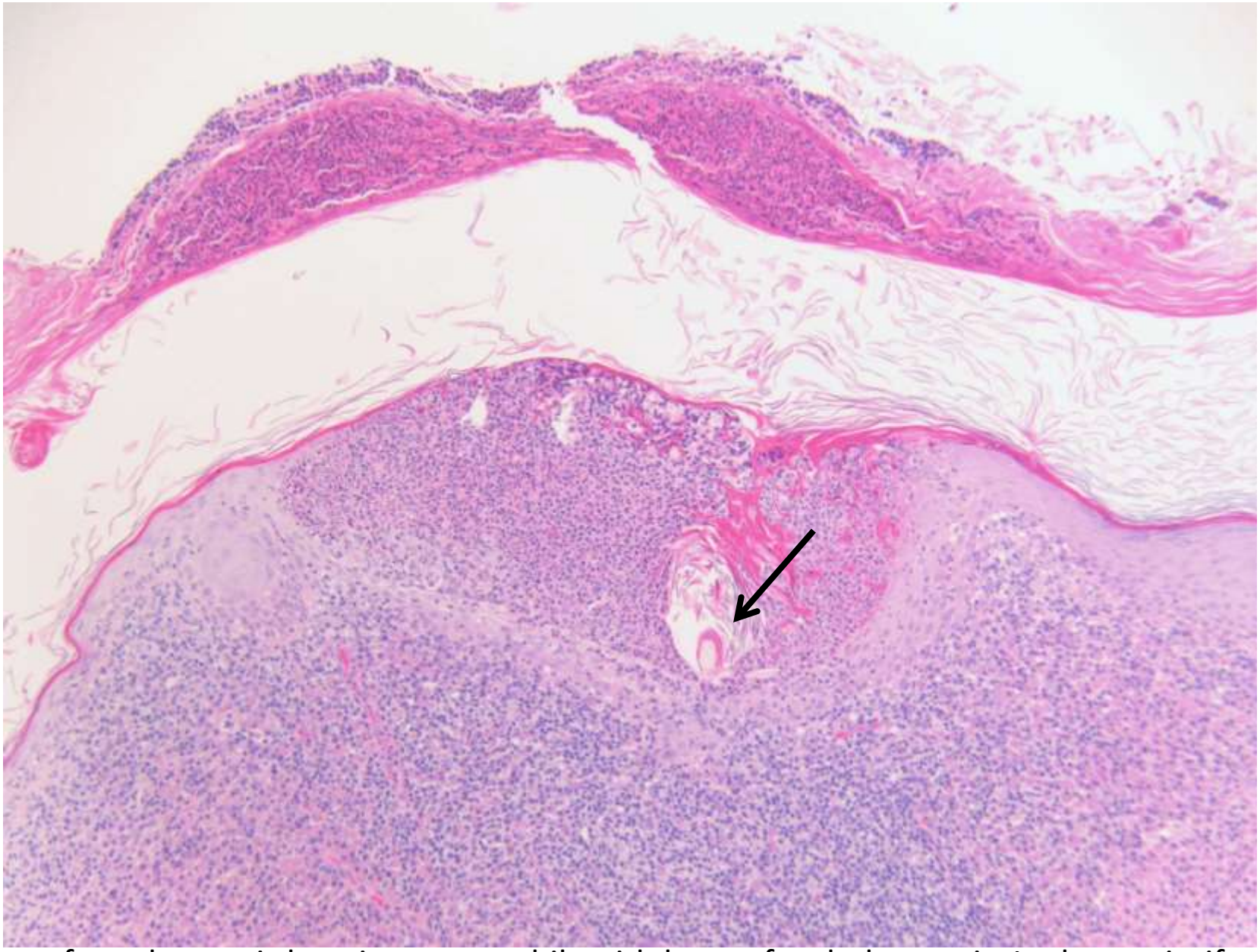


Layer of parakeratosis housing neutrophils with layer of orthokeratosis. Lacks psoriasiform pattern. Dense chronic inflammation in the upper dermis with innumerable plasma cells

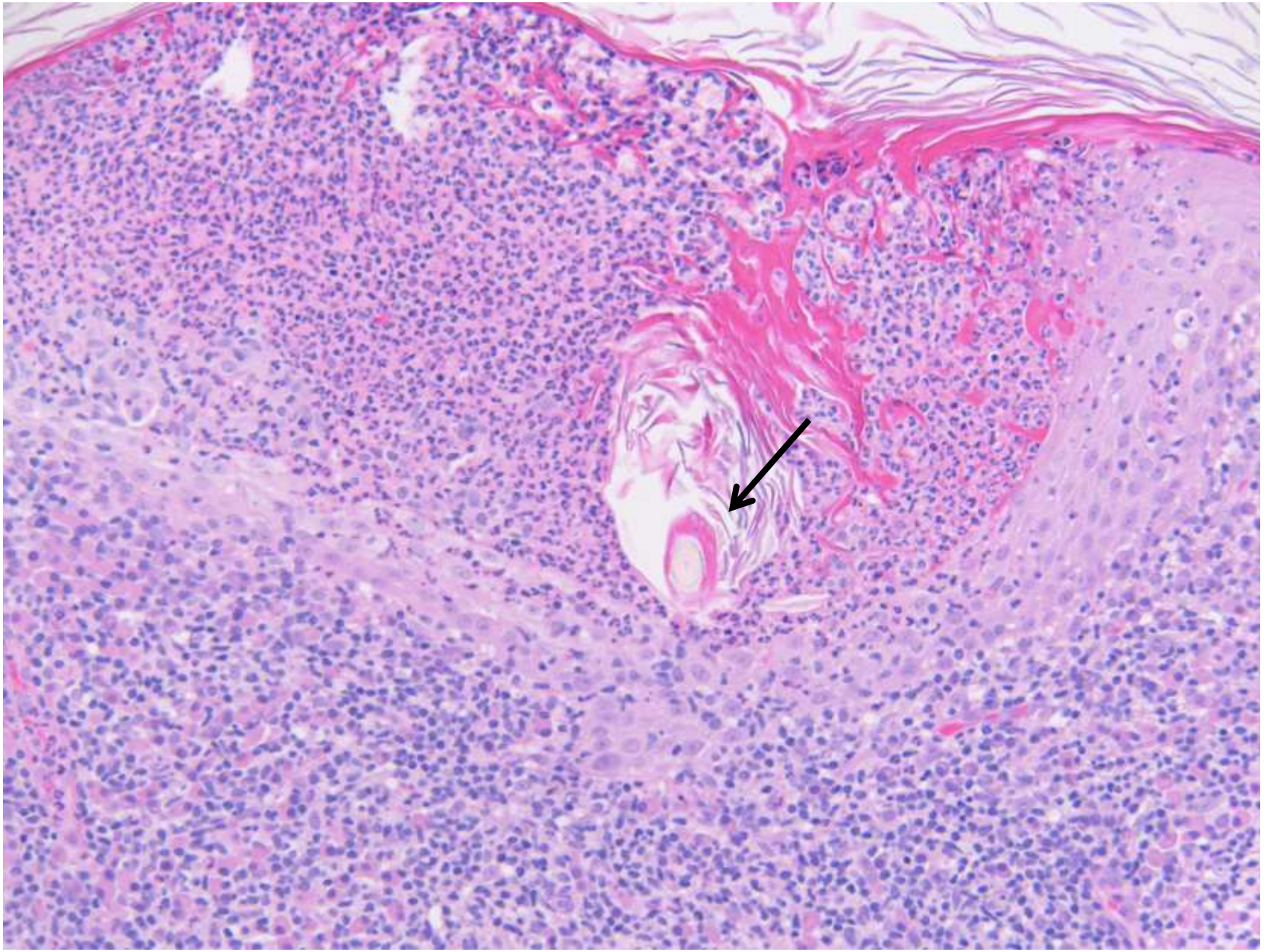
The other half of the biopsy has lost the surface keratin and parakeratin (“floating” on slide – upper right)



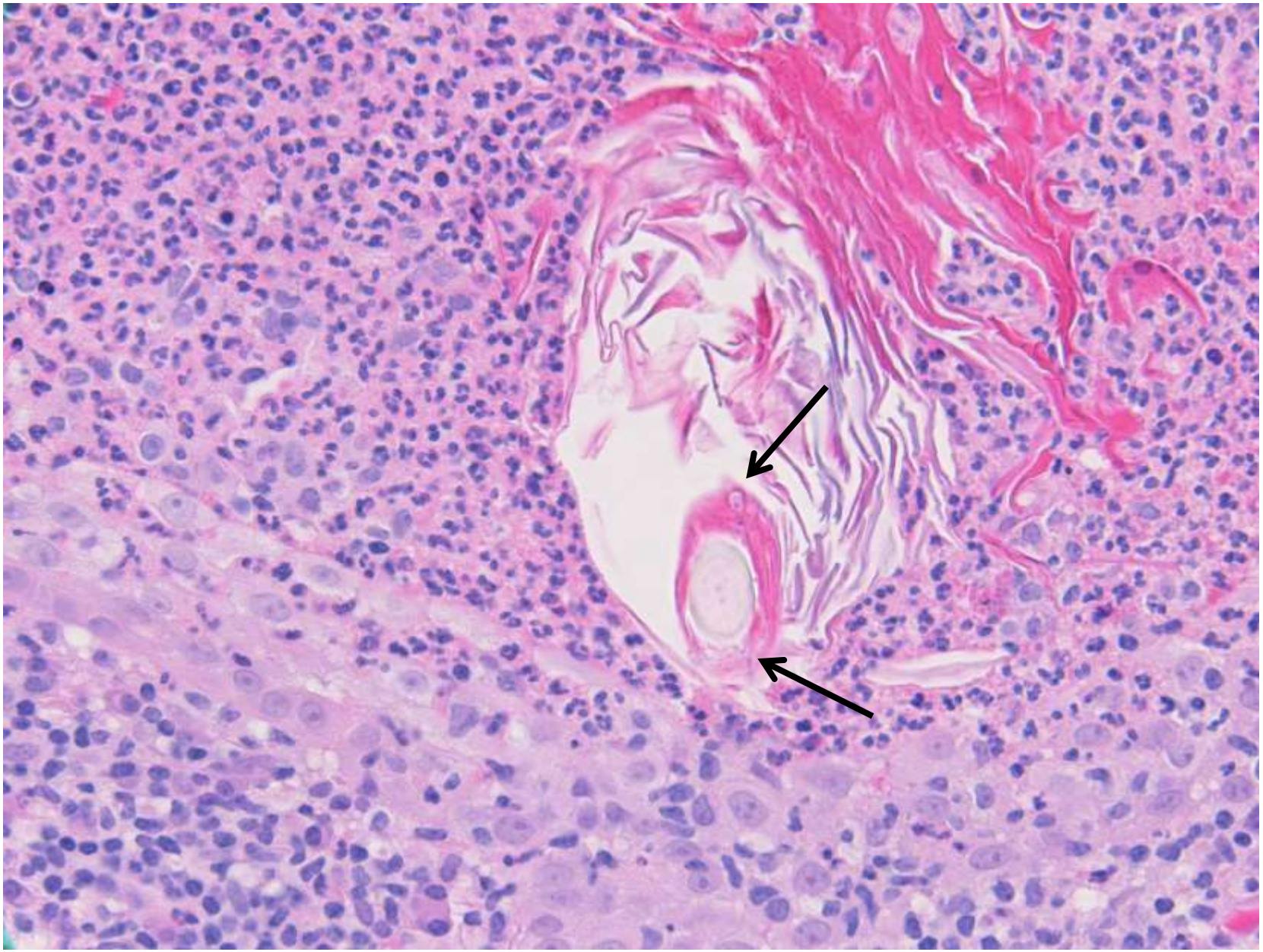




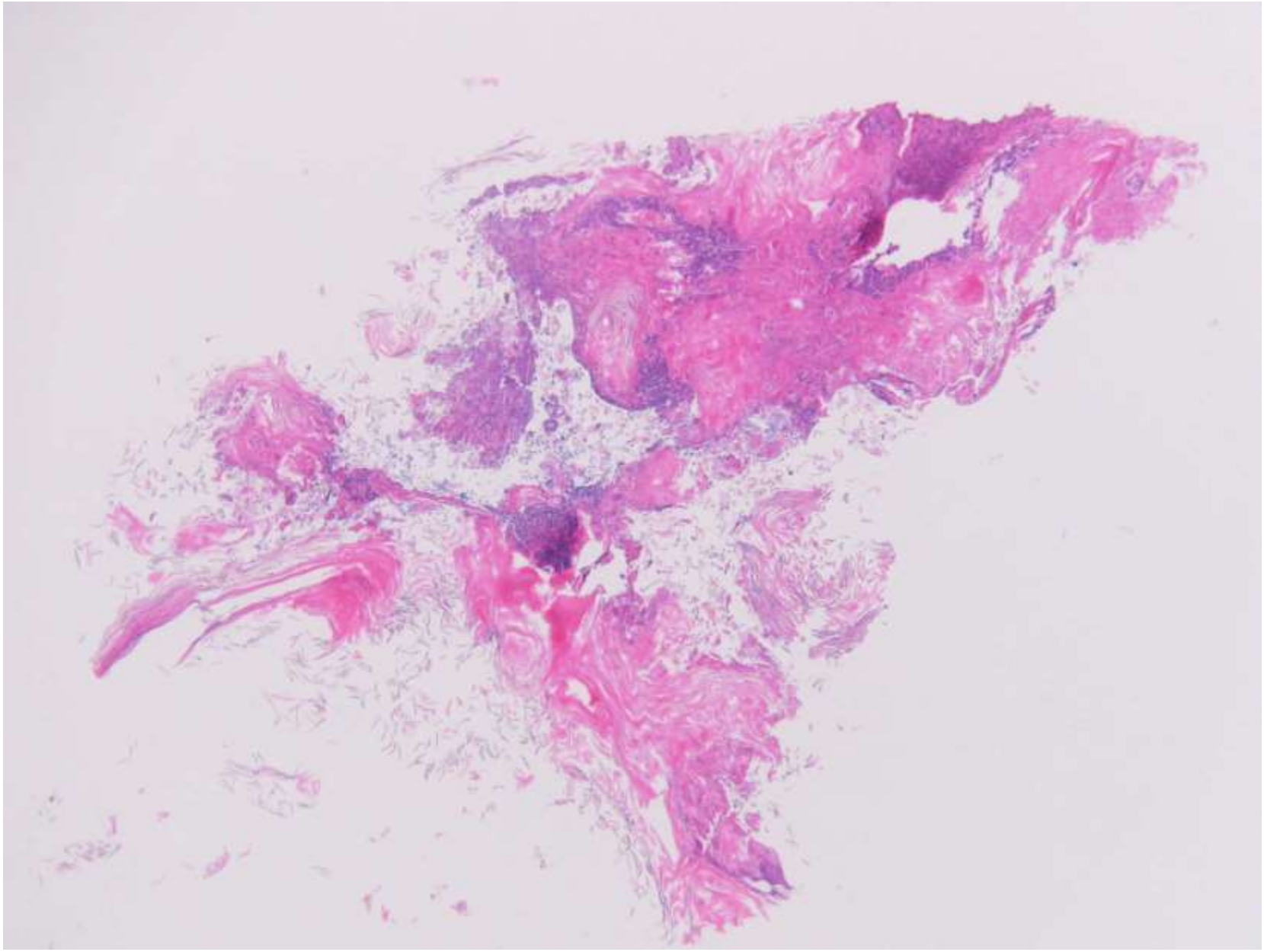
Layer of parakeratosis housing neutrophils with layer of orthokeratosis. Lacks psoriasiform pattern. Dense chronic inflammation in the upper dermis with innumerable plasma cells. Note hair shaft (arrow).



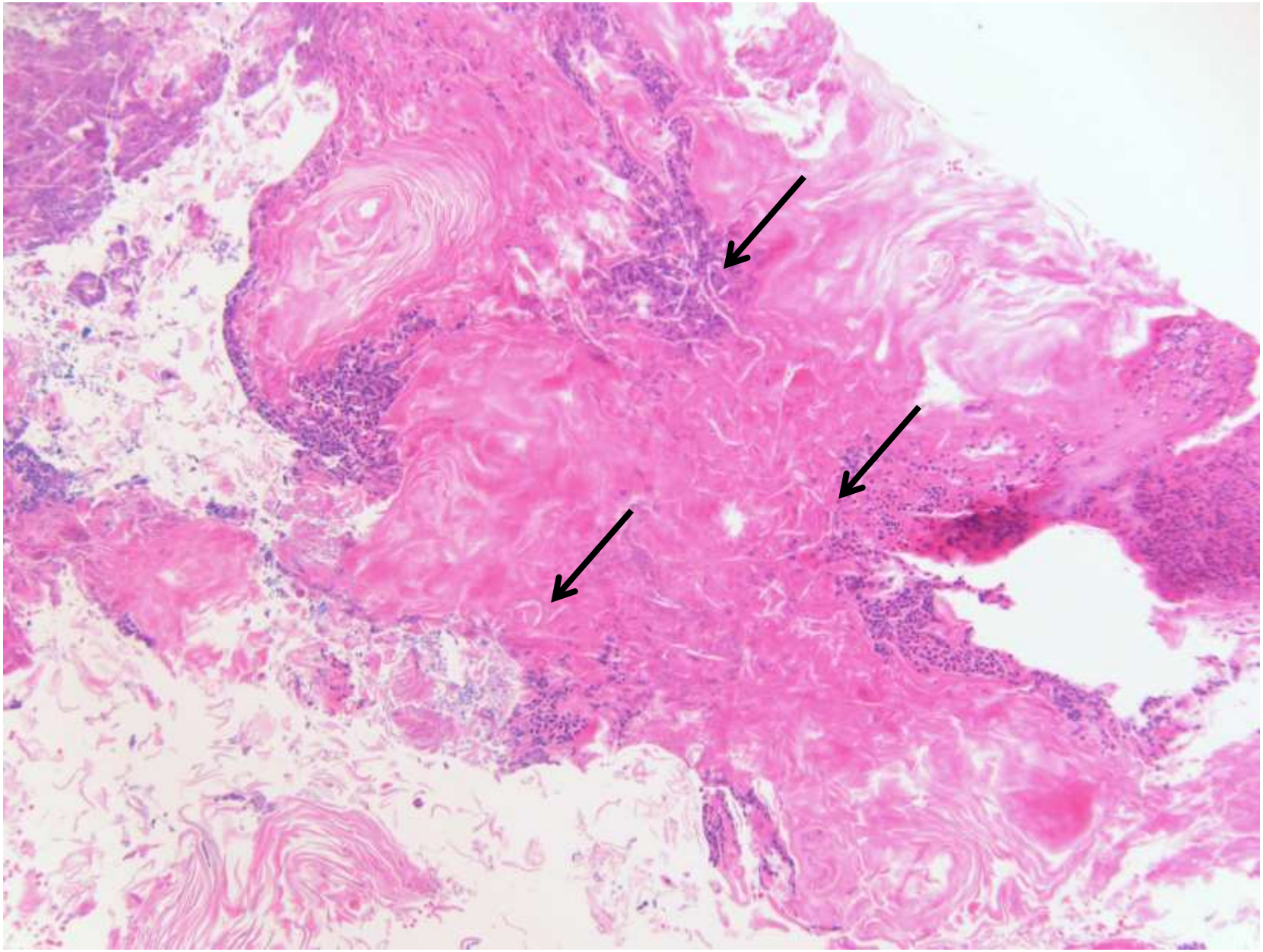
Layer of parakeratosis housing neutrophils with layer of orthokeratosis. Lacks psoriasiform pattern. Dense chronic inflammation in the upper dermis with innumerable plasma cells. Note hair shaft (arrow).



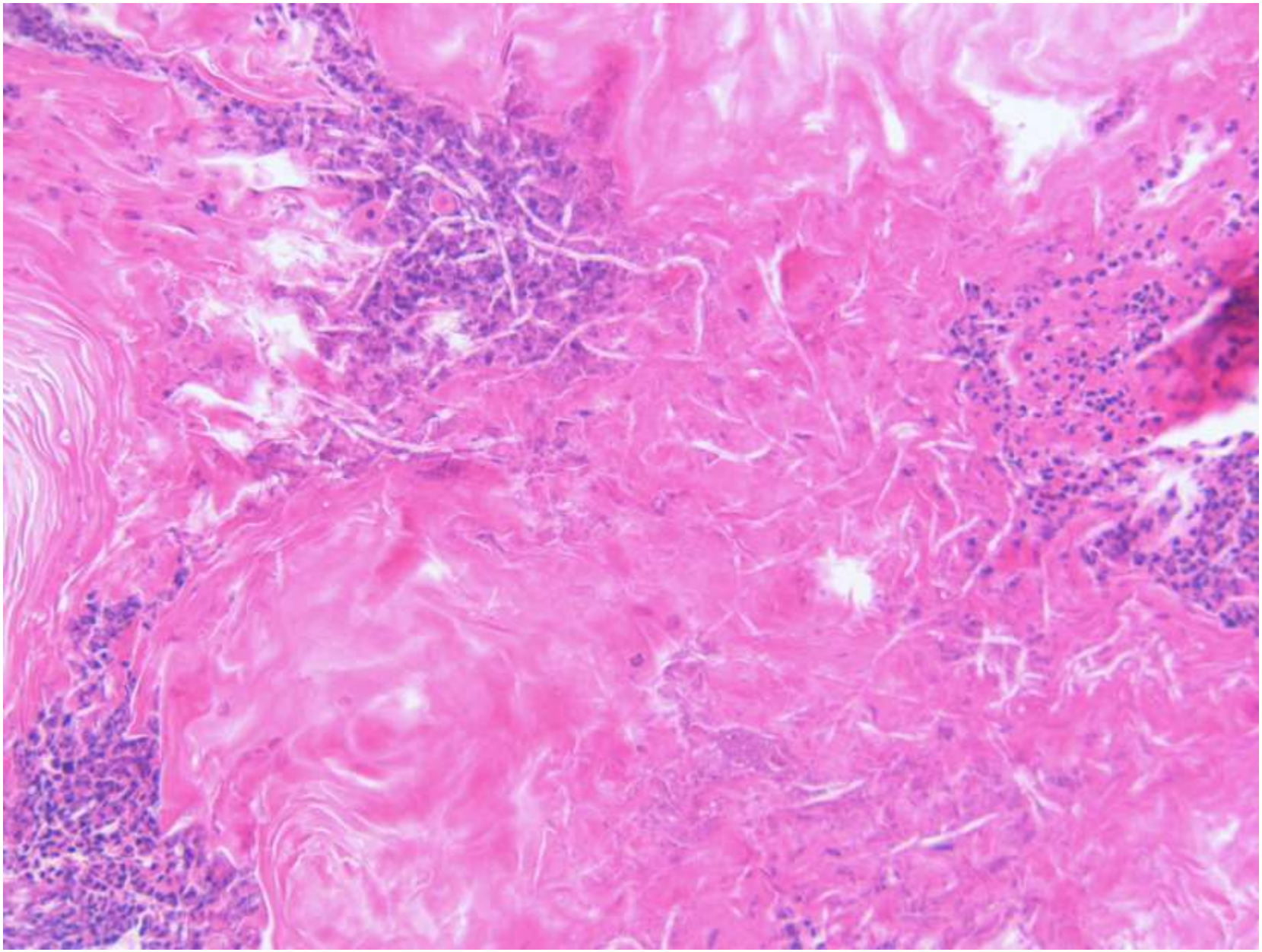
Always look for hyphae in the infundibula of follicles (they can hide here).



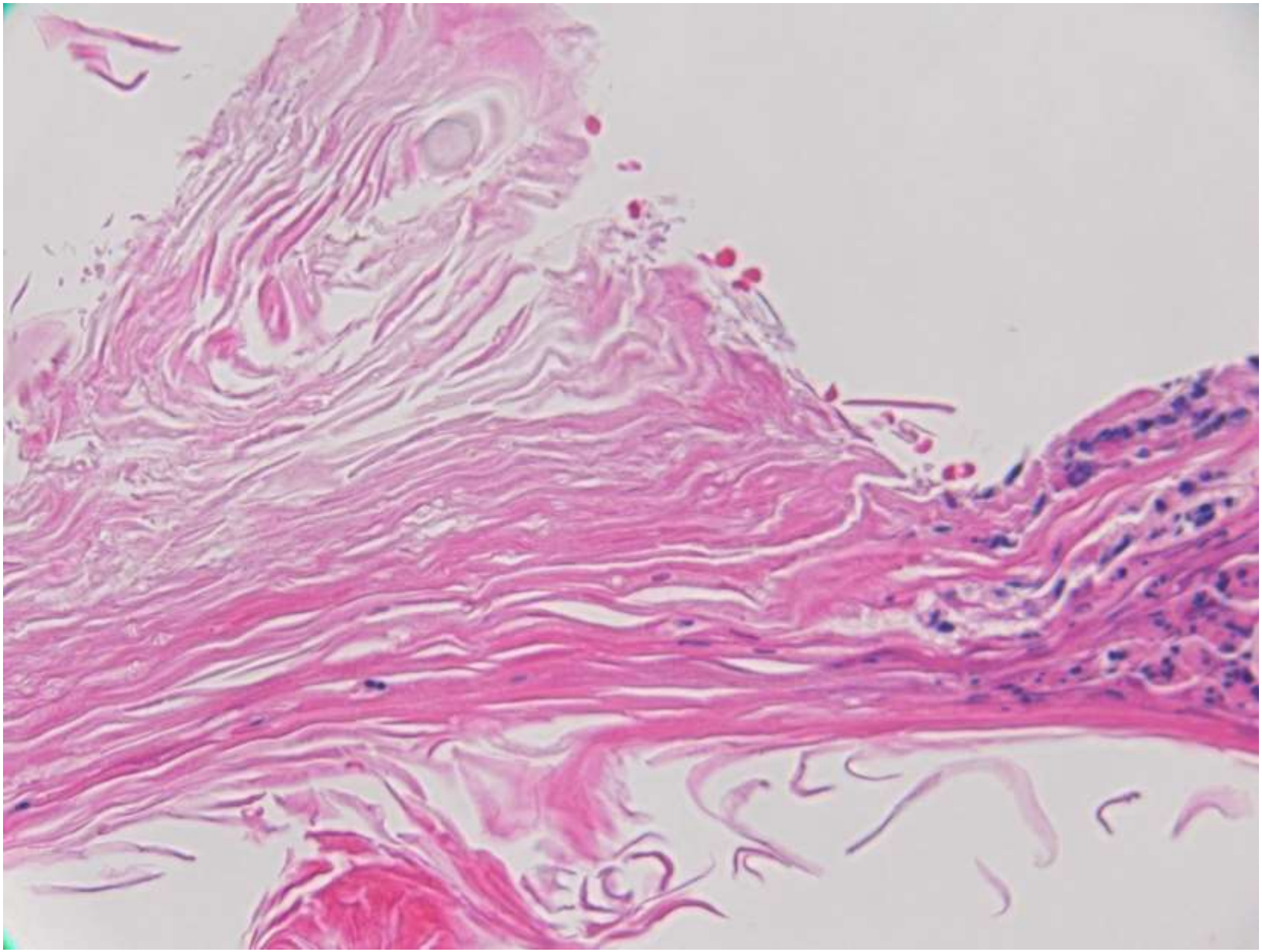
The detached inflammatory crust contains abundant fungal hyphae



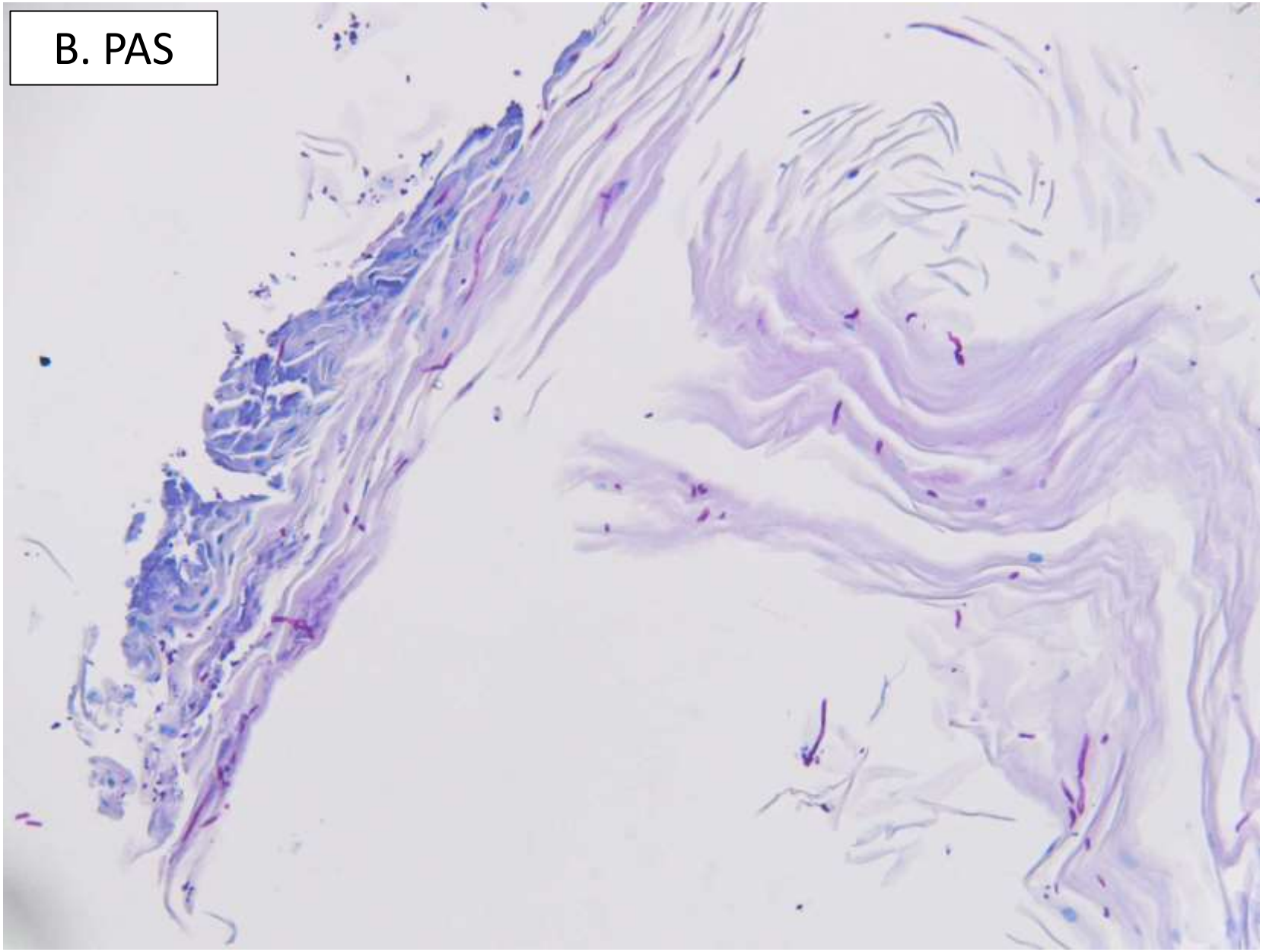
The detached inflammatory crust contains abundant fungal hyphae. Note the ghost lines of fungal hyphae H&Ex10, arrows.



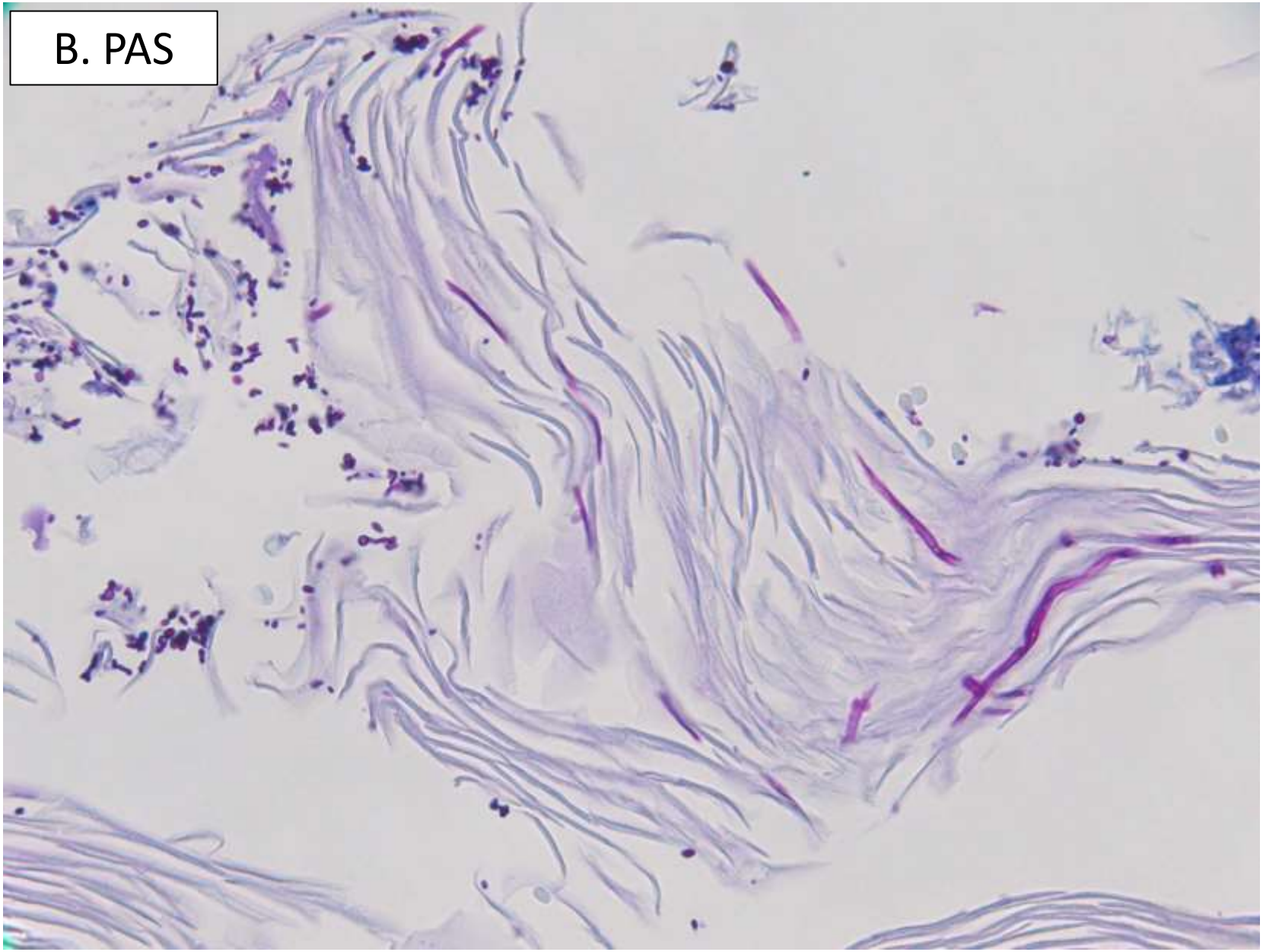
The detached inflammatory crust contains abundant fungal hyphae. Note the ghost lines of fungal hyphae, H&Ex20.



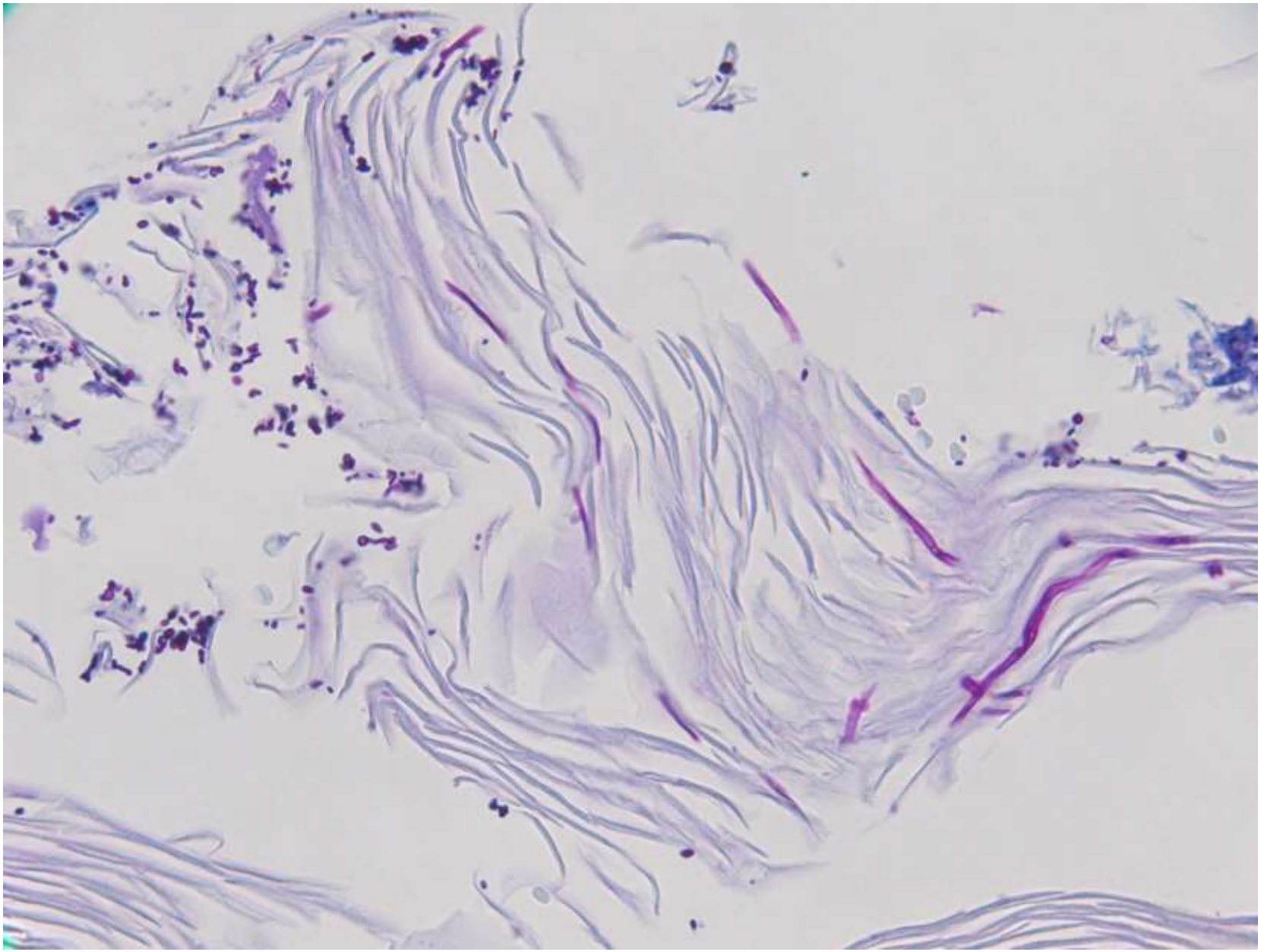
Subtle round holes (of fungal elements) in the orthokeratosis adjacent to the neutrophils in the stratum corneum are easily missed on H&E. Hyphae are much more difficult to see in the vertically orientated area but are no doubt present.



Fungal hyphae, many cross-sectoned



Fungal hyphae, some cross-sectoned and fungal spores



FURTHER SUPPLEMENTARY REPORT – 9/8/2019 – Dermatopathologist

Sections reviewed at the request of Dr XXXX and haematopathology team.

A and B show features in keeping with tinea faciei (abundant fungal hyphae on PAS), pustulation and psoriasiform pattern. The kappa stain appears rather capricious/uninterpretable.

I am not suspicious for a lymphoreticular disorder.

Suggest treatment for fungal infection and follow-up.

Supplementary report – 03/09/2019

Please see detail below. As discussed previously, while this may be a reactive population, given the morphology, immunochemistry and clonality studies, the possibility of a lymphoid neoplasm with plasma cell differentiation cannot be totally excluded and clinical follow up is advised.

This is a transcribed report. The original report (HMG19-01363) was issued by Dr ZZZZ,

Specimens: A Skin, MA6467. 1x green block "Q 19 56584 B". Specimen Details. A paraffin block of skin biopsy with an infiltrate of plasma cells.

IG gene rearrangement (clonality) result: DNA was extracted from the above referred specimen and its quality as shown by PCR of the control genes is adequate for the following IG gene rearrangement analysis using BIOMED-2 PCR assays and GeneScanning of PCR products.

Tests Results

IGH gene rearrangement

VH FR1-JH: No apparent amplification

VH FR2-JH: No apparent amplification

VH FR3-JH: **Weak clonal**

DH-JH: **Weak clonal**

IGK gene rearrangement

VK-JK: **Weak clonal**

VK-Kde/JKCK intron-Kde: No apparent amplification

IGL gene rearrangement

VL-JL: No apparent amplification

CONCLUSION

Clonality analysis using BIOMED-2 assays PCR showed weak clonal IGH and IGK gene rearrangements in the sample analysed. This result indicates that **a small population of clonal B-cells is present in this specimen**.

Verified by ZZZZ, ConScientist on 28/8/2019

MML Notes

Please note the above clonality assays require above 1%-5% of clonal B cells present in the specimen in order to demonstrate a clonal IG gene rearrangement. The clinical sensitivity and specificity of the assays are over 90% in mature B-cell neoplasms. Molecular results must be interpreted in the context of clinical, histological and immunophenotypic findings.

Learning Points: Quite a lot!!!

- **In dermatopathology the dermatologist is often right.**
- Clinical picture “keratotic nodule” and “crusty red skin”
- Try to think what might mimic these clinically.
- If you cannot find the diagnosis suggested by the dermatologist, and **you have lot’s of pathology**, suggest showing the case to a dermatopathologist **before doing any levels and special stains**
- They will often make a diagnosis based on CPC and can use appropriate targetted special stains.
- **A suspected cutaneous lymphoma should be seen in a team that includes a specialist dermatopthologist and discussed in a skin MDM before referral to haematopathology team**

Learning Points:

- Neutrophils in the stratum corneum and alternating para- and othrokeratosis are clues to psoriasis and fungal infection
- Tinea & Candidiasis can almost perfectly mimic psoriasis
- Fungal spores and bacterial colonisation are NOT typical features of psoriasis.
- Psoriasis is a hyper-proliferative condition and usually fungi cannot gain a hold due to constant shedding
- Psoriasis does not usually have a prominent chronic inflammatory cell infiltrate with abundant plasma cells
- Suspect fungal infection
  - **Clinical describes patches of crusty, erythematous skin**
  - **Especially if localised isolated or a few lesions in one anatomical area**
  - **Especially near hands, feet, moist areas and mucosae**
- **Generally if the clinician or you (pathologist) suspects fungal infection…**

Part 3: Learning Points Continued: Kappa & Lambda

- Generally I pay most attention to the hyper- positive plasma cells or plasmacytoid cells (rather than lymphoid cells)
- In skin a Lambda:Kappa of >5:1 and a Kappa:Lambda >10:1 would be considered significant
- The antibodies are difficult to interpret with confidence
- Even apparent clonality in a single lesion does not necessarily indicate a neoplasm
- Similarly a lack of clonality does not exclude a neoplasm
- **Clinicopathological correlation is essential to good dermatopathology**

LEARNING POINT

Haempath and Dermpath should be complementary and work in a close team but suspected skin lymphomas should be discussed at the skin MDM in the first instance

Dermatophytosis

- Invisible
- Spongiotic
- Psoriasiform
- Pustular
- Folliculitis
- Boggy masses (kerion)
- Nodular lesions
  - (Majochi granuloma)

- LEARNING POINTS
- Listen to clinical
- PAS strips or at 3 levels
- Look carefully

Learning Points

- Dermatophytosis may perfectly mimic eczematous dermatoses and psoriasis (of all sub-types)
- “Sandwich sign” (layered orthokeratosis and parakeratosis) clue to fungal infections & psoriasis
- Negative stains still does not r/o

Learning Points Cont…

- Do not rely wholly upon the clinical history or suggested diagnosis
- BUT be particularly wary of non-expert dermatologists!!!
- Lower your threshold for PAS stain on mucocutanous sites, localised areas such as hands & feet and moist areas
- Candida may colonise (no neutrophils) or superinfect (neutrophils) leucoplakia and lichenoid dermatoses at mucocutaneous sites (but the underlying condition should be present)

Acknowledgements: Part 3
Paul Matthews
Hesham El Daly
David Snead
Saleem Taibjee
David Slater
Alister Robson
Werner Kempf