

# Cases

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Update in Dermatopathology

Liverpool, Nov 28 2017

# Case 1

# Clinical history

- 52 year old female with several year history of lesions of nose
- HIV positive on HAART
- Deceased donor kidney transplant 2015 with rejection renal failure on belatacept, cellcept



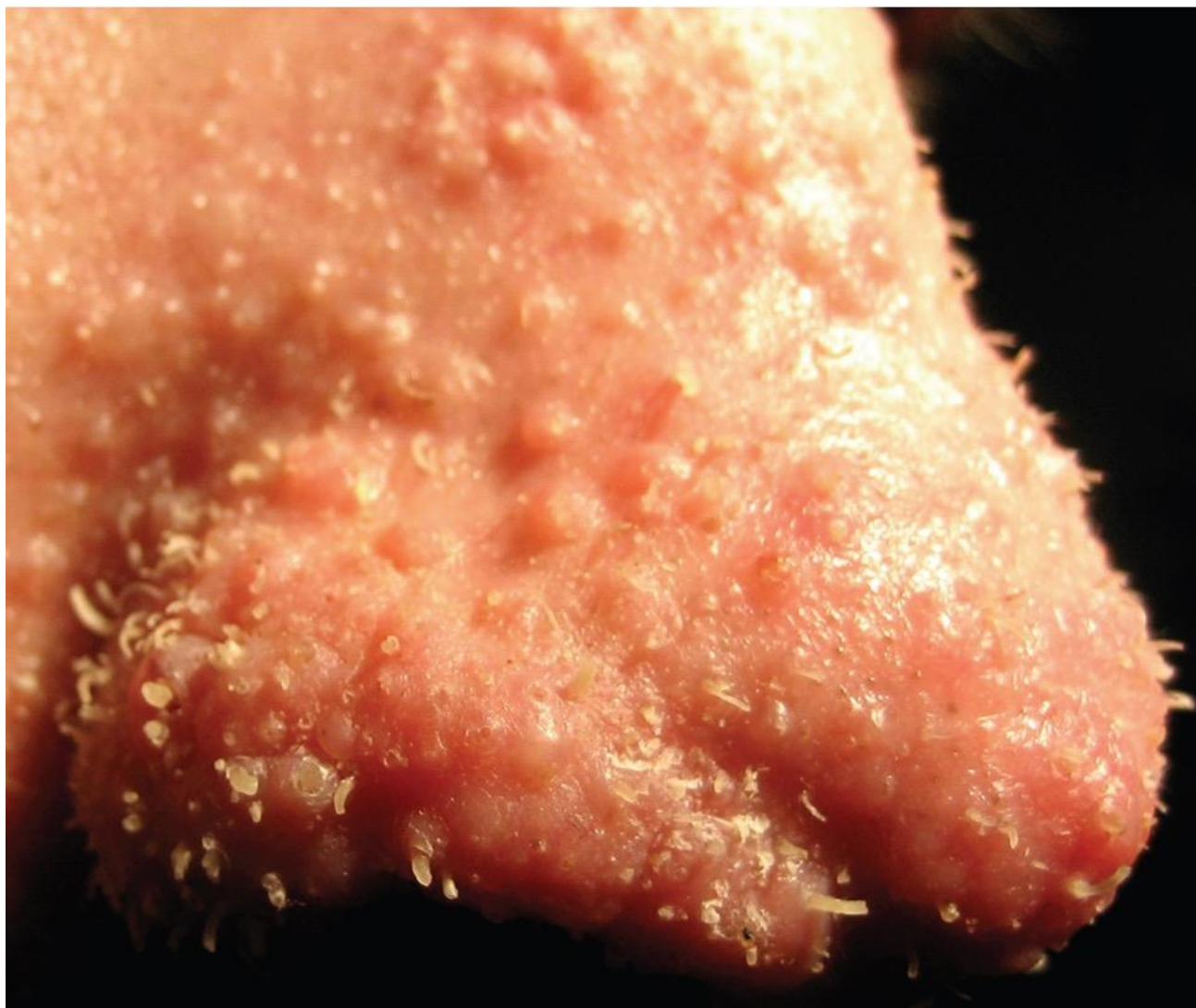




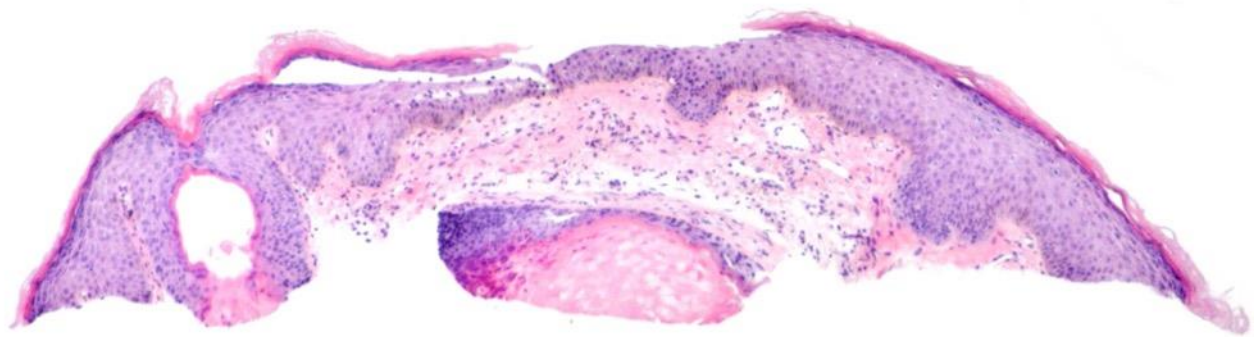


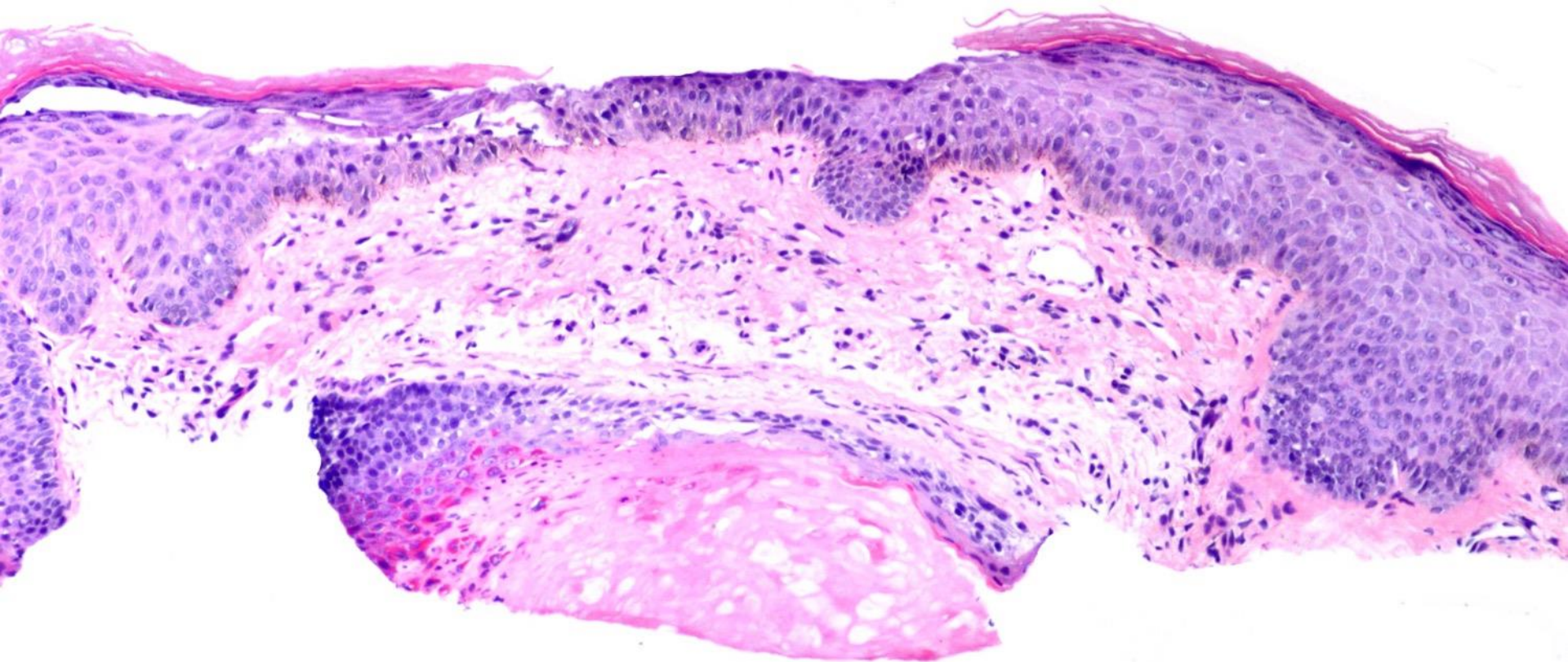


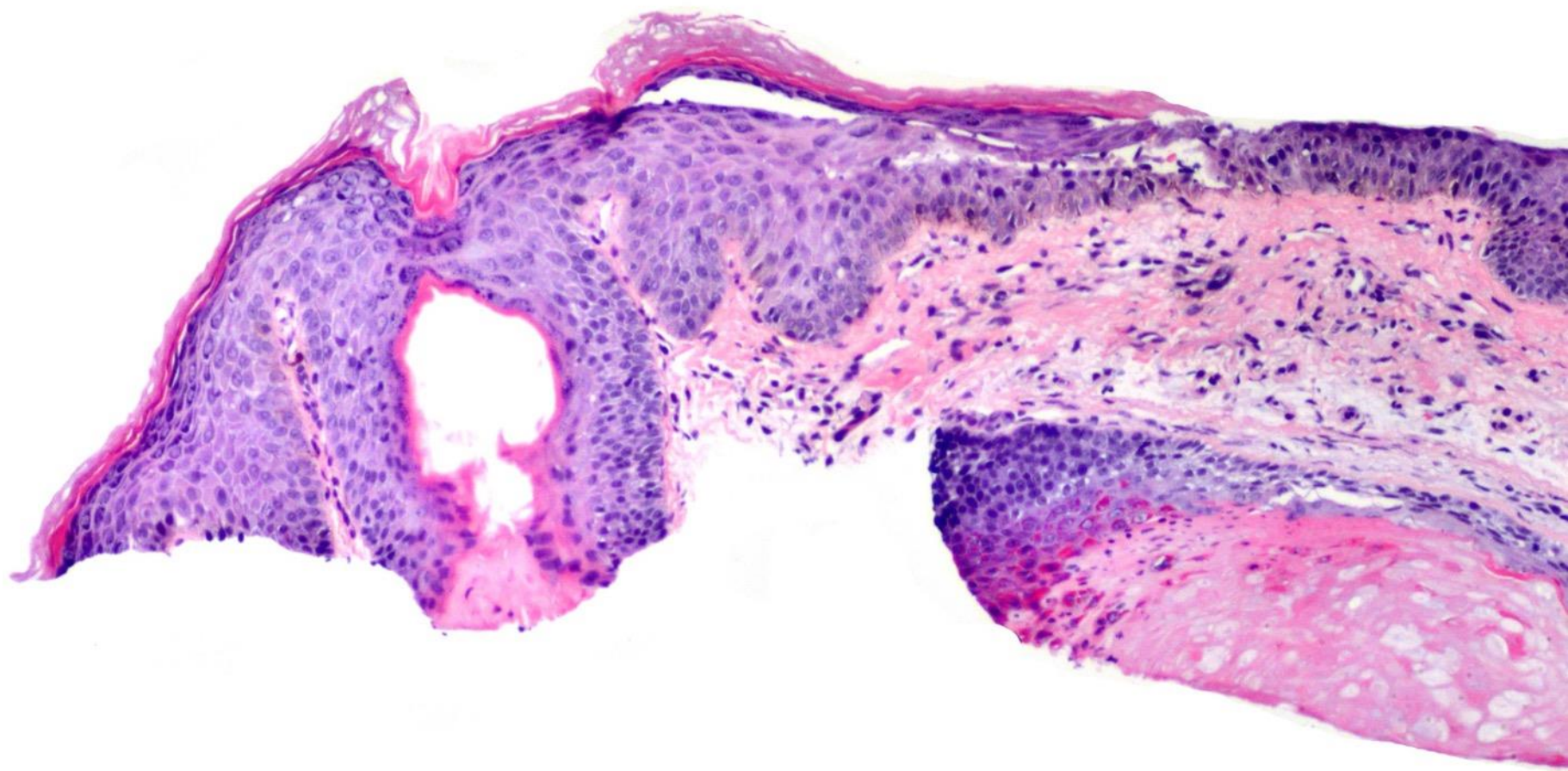




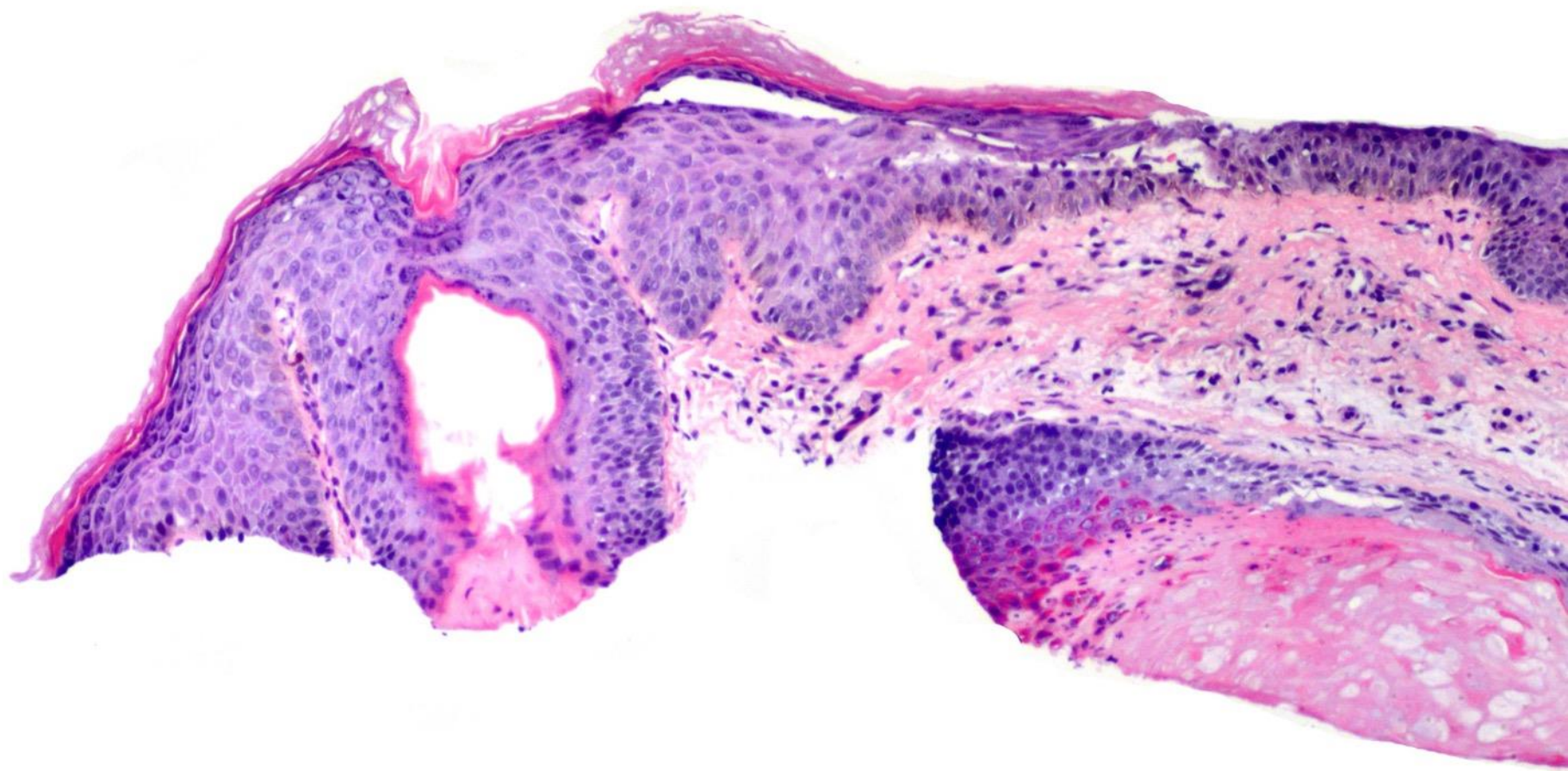




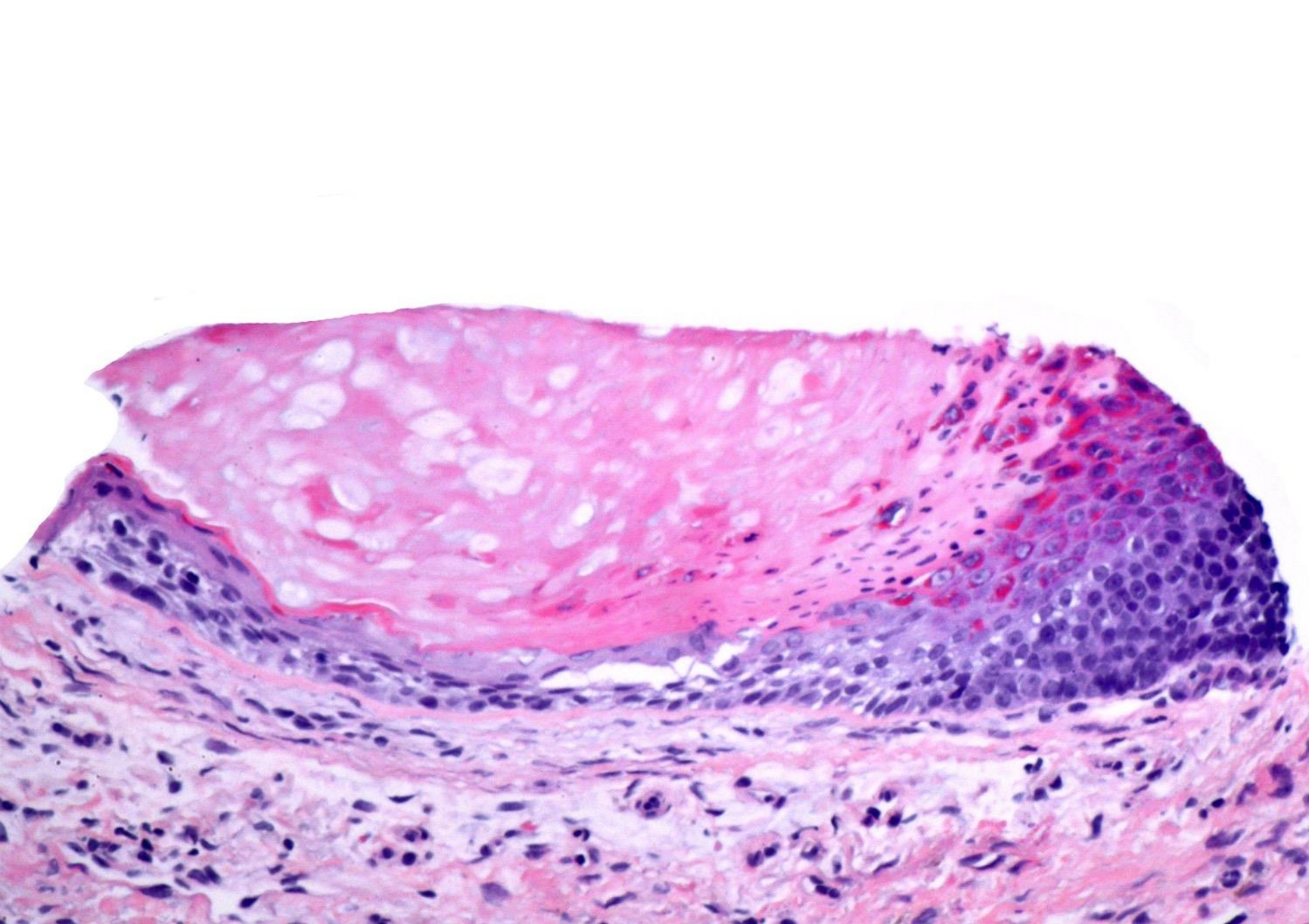




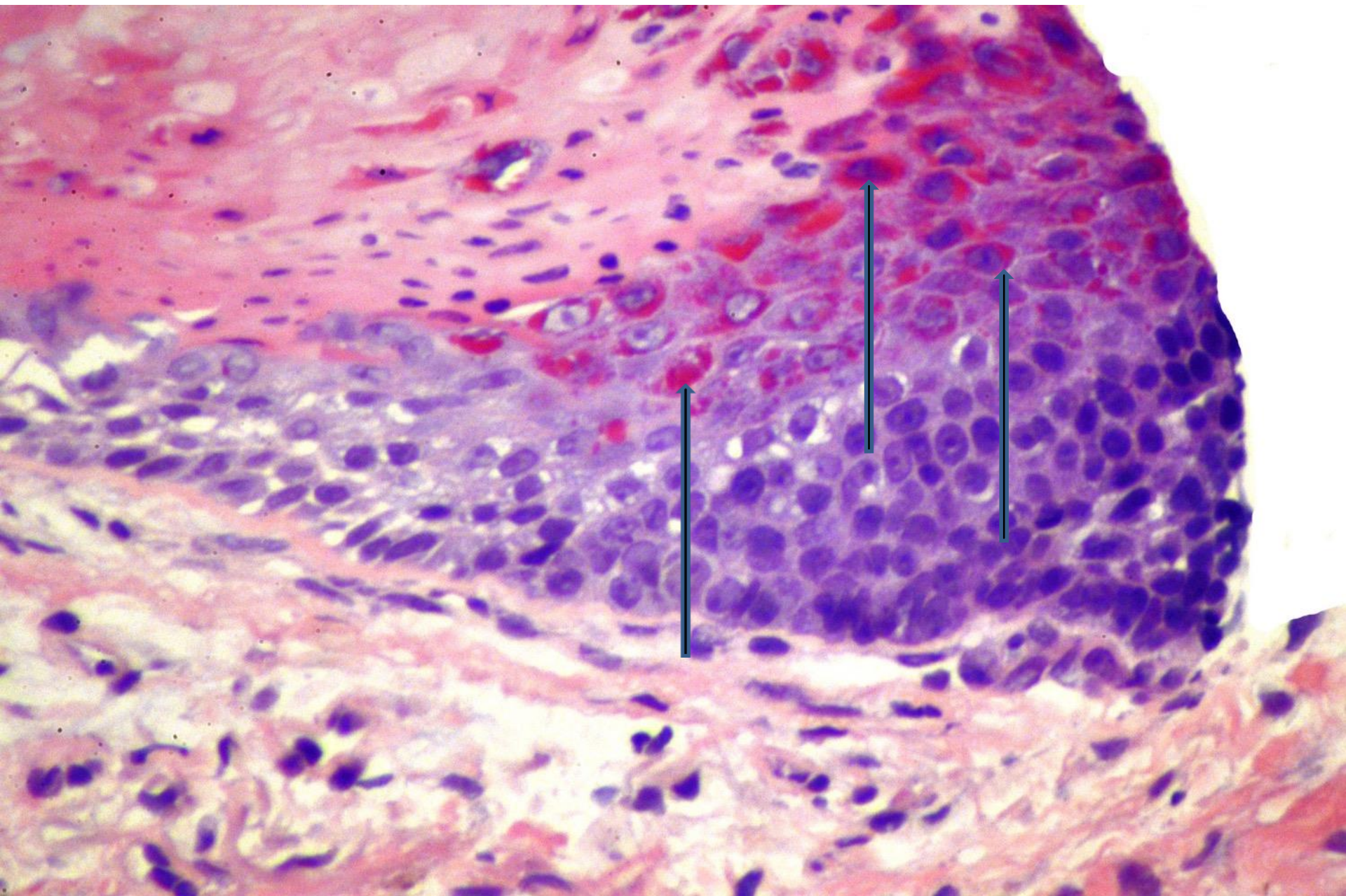


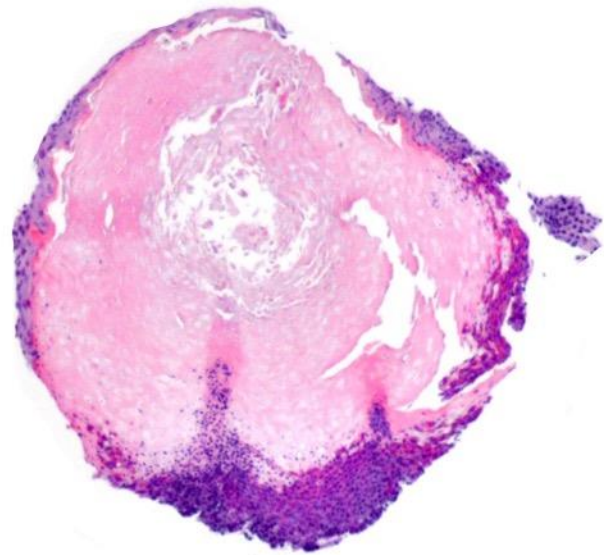




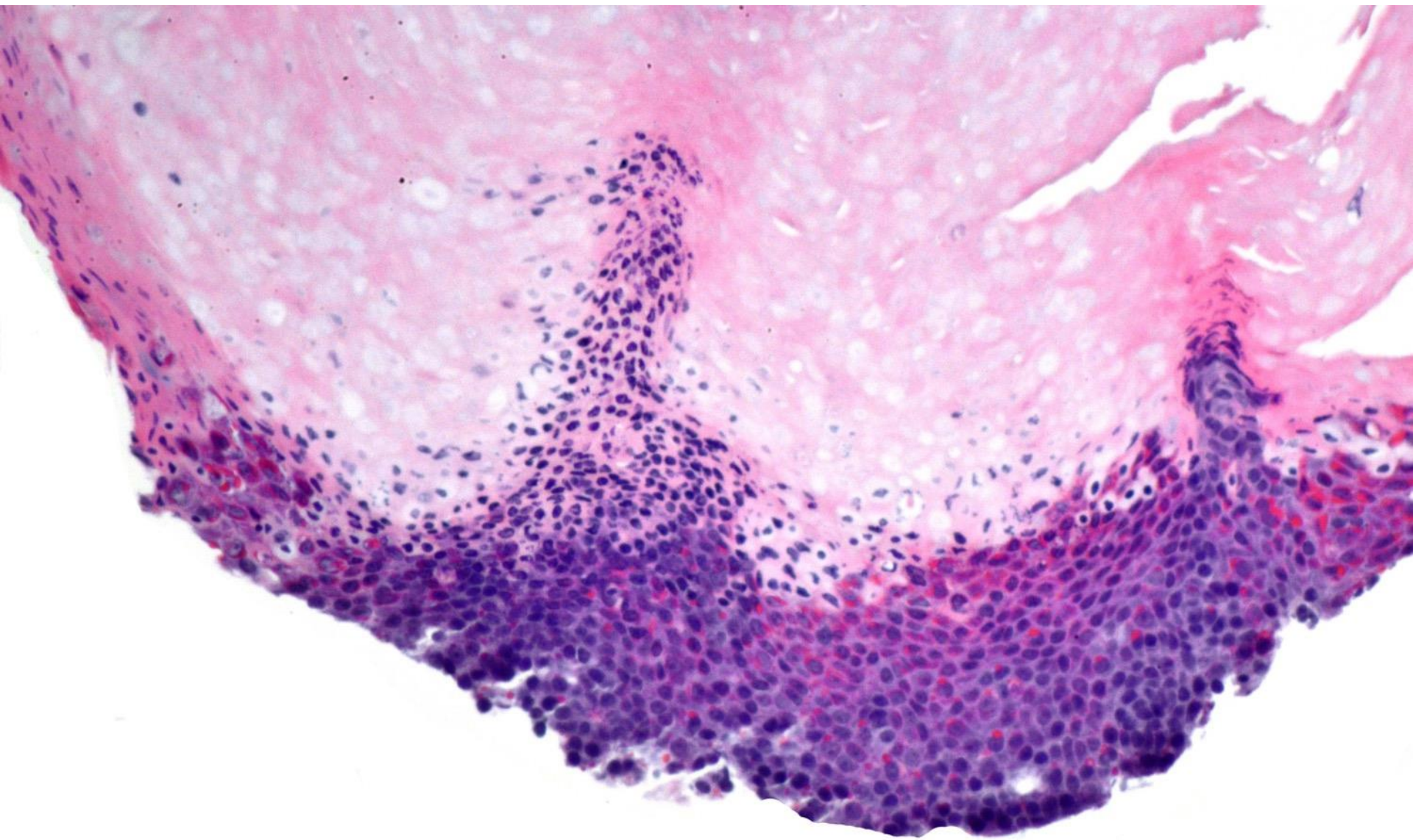




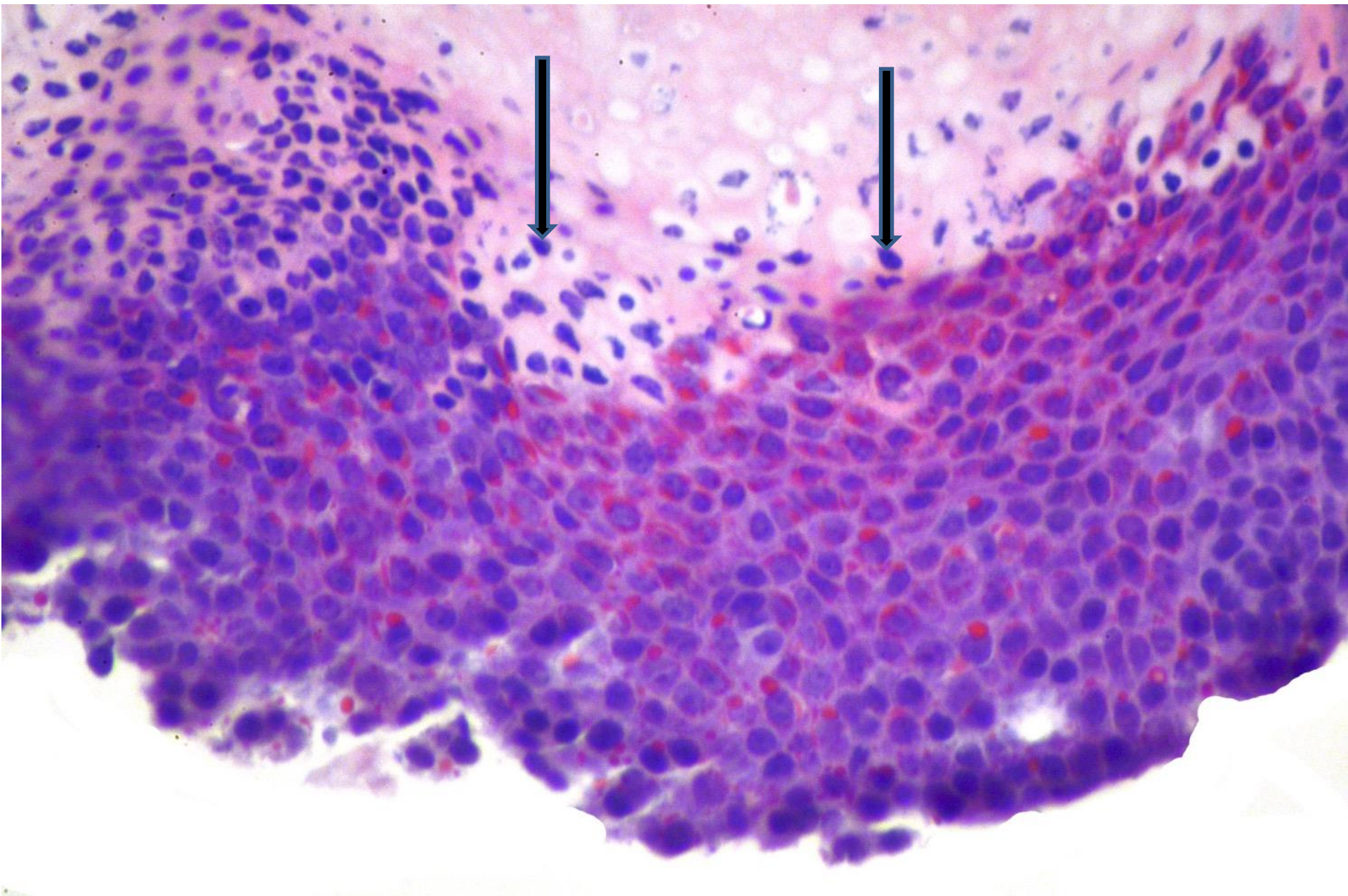












# Trichodysplasia spinulosa

- Usually occurs in immunosuppressed individuals, often transplant recipients
- Follicular papules in the face
- Whitish spicules on face and nose
- Thickened skin, leonine facies
- Alopecia
- Caused by trichodysplasia spinulosa-associated polyomavirus (TSPyV),
- Very rare (20 cases in 2013)

# Physical exam

- Flesh colored follicular papules
- Central part of face, trunk and extremities
- Infiltrated appearance, dysmorphic, of nose
- Alopecia, eyebrows and eyelashes

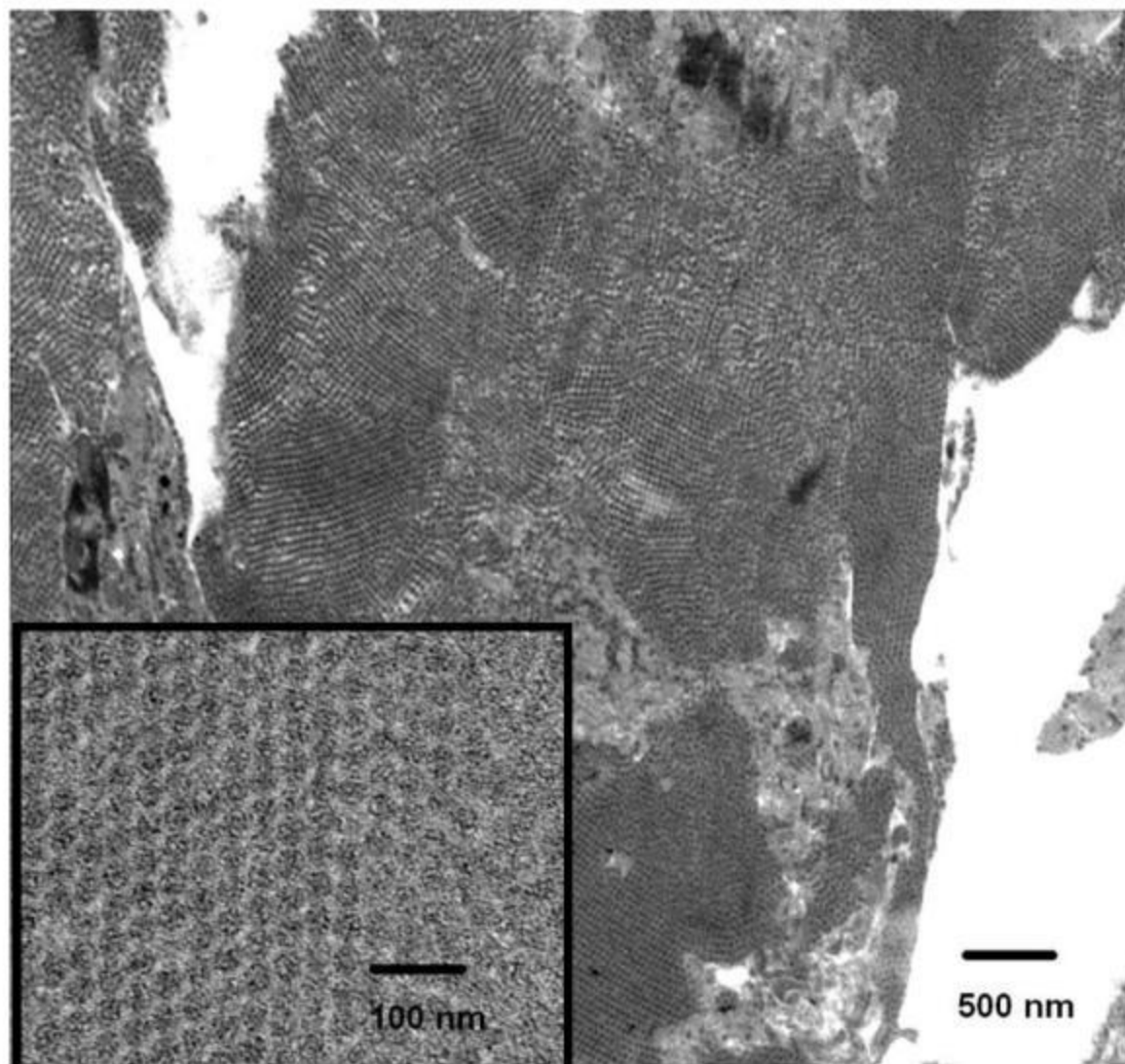
# Histopathology

- Abnormal bulbous hair follicle
- Excessive inner root sheath production
- Nucleated eosinophilic cells with excess trichohyaline
- Intraepithelial intranuclear viral inclusions (sometimes extracellular)
- No formed hair shaft



# Polyoma virus

- Double-stranded DNA viruses 42nm icosahedral
- Affects mammals and birds
- 13 types infect human
- 5 types cause disease
  - MCPyV, TSPyV, BKPyV, JCPyV, HPyV6, HPyV7
- Most are asymptomatic



# Therapy

- Patient currently on isotretinoin 30 mg BID
- Some antivirals are also effective cidofovir cream, oral valganciclovir

# Case 2



# History

## HISTORY OF PRESENT ILLNESS

45 y/o M presented with red bumps on lower extremities, present for 3 years. First started on right thigh around the knee with new lesions appearing over time on right ankle, perhaps a few lesions on the left. Lesions never resolved. Asymptomatic. No clear triggers or associations. Patient has history on ankle fracture on right side

ROS negative. No systemic complaints.

## Medical HISTORY

- IBS, DM2

## MEDICATIONS

- None

## FAMILY HISTORY

- Non-contributory

## Social HISTORY

- Married with children
- Occupation: Insurance broker
- Tobacco: none
- EtOH: occasionally
- Illicits: none

# Clinical Images





# Laboratory Evaluation

9.4      16  
          48  
247

AST 21  
ALT 48  
ALP 101  
Tbili 0.4

142	98	17	< 131
4.2	21	0.87	

Total cholesterol 174  
TG 135

HbA1c 7.5%

TSH 1.8

# Differential Diagnoses

- **Leukocytoclastic Vasculitis**

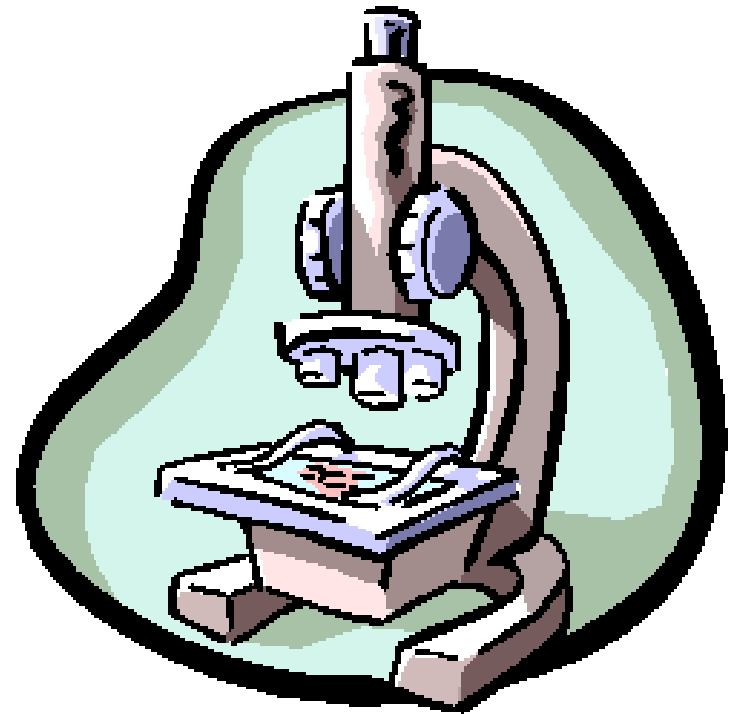
- **Drug-induced**
- **Infectious:** Strep, RMSF, GC, TB, syphilis, hepatitis
- **Malignancy:** Lymphoma, Leukemia, Hodgkin's, multiple myeloma
- **Collagen Vascular Disease**
- **Abnormalities in blood viscosity:** cryoglobulinemia, cryofibrinogen, cold agglutinins, hypergammaglobulinemic purpura, other coagulable disorders
- **Small vessel vasculitis i.e.**

HSP

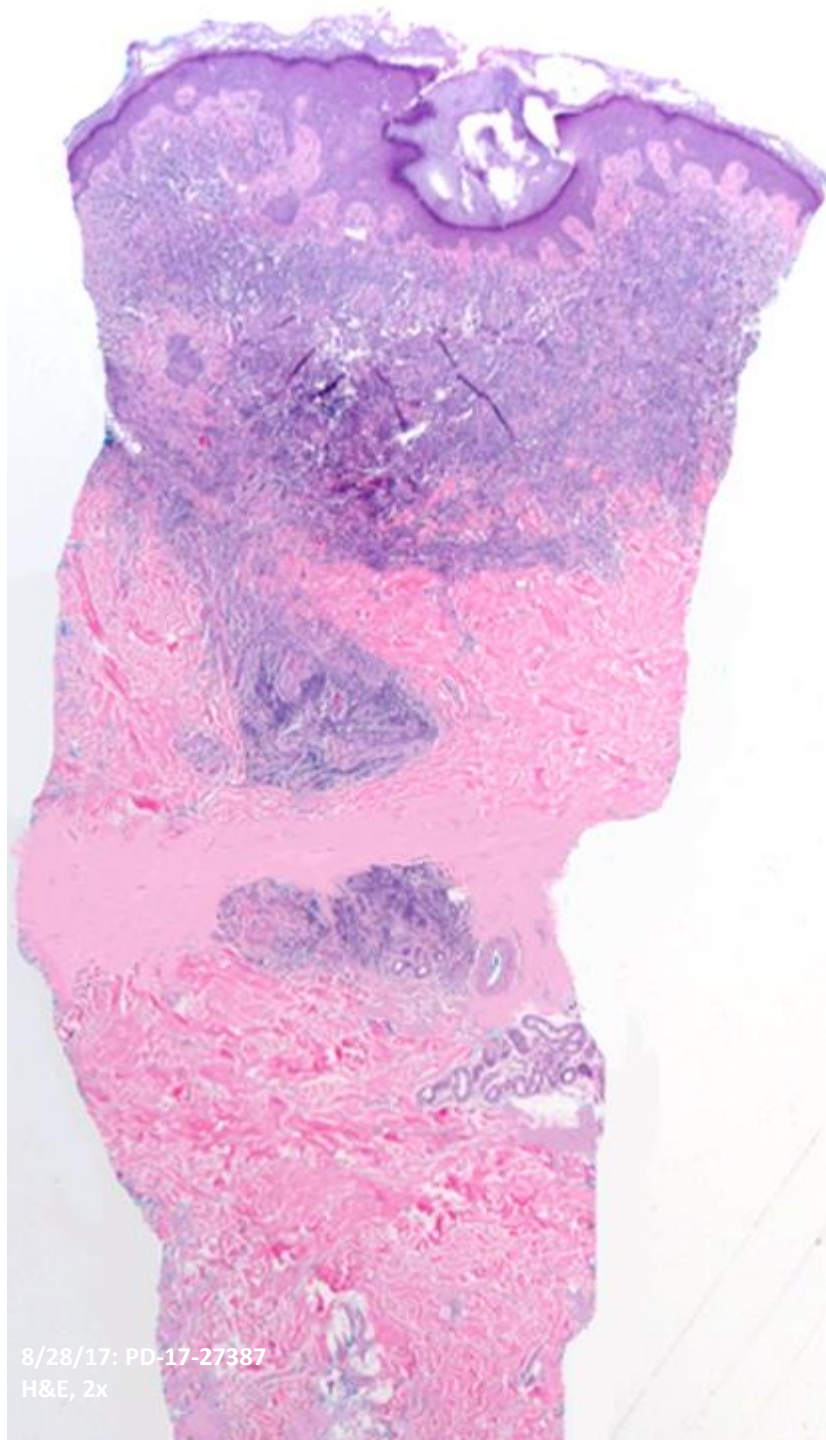
- Medium vessel vasculitis i.e. PAN, Wegener's

- Pityriasis lichenoides et varioliformis acuta (PLEVA)
- Sweet's syndrome
- Urticarial vasculitis
- Kaposi's sarcoma
- Pigmented purpura (pigmented purpuric lichenoid dermatitis of Gougerot-Blum)
- Cutaneous T-cell Lymphoma
- Pseudolymphoma
- Lymphomatoid Papulosis

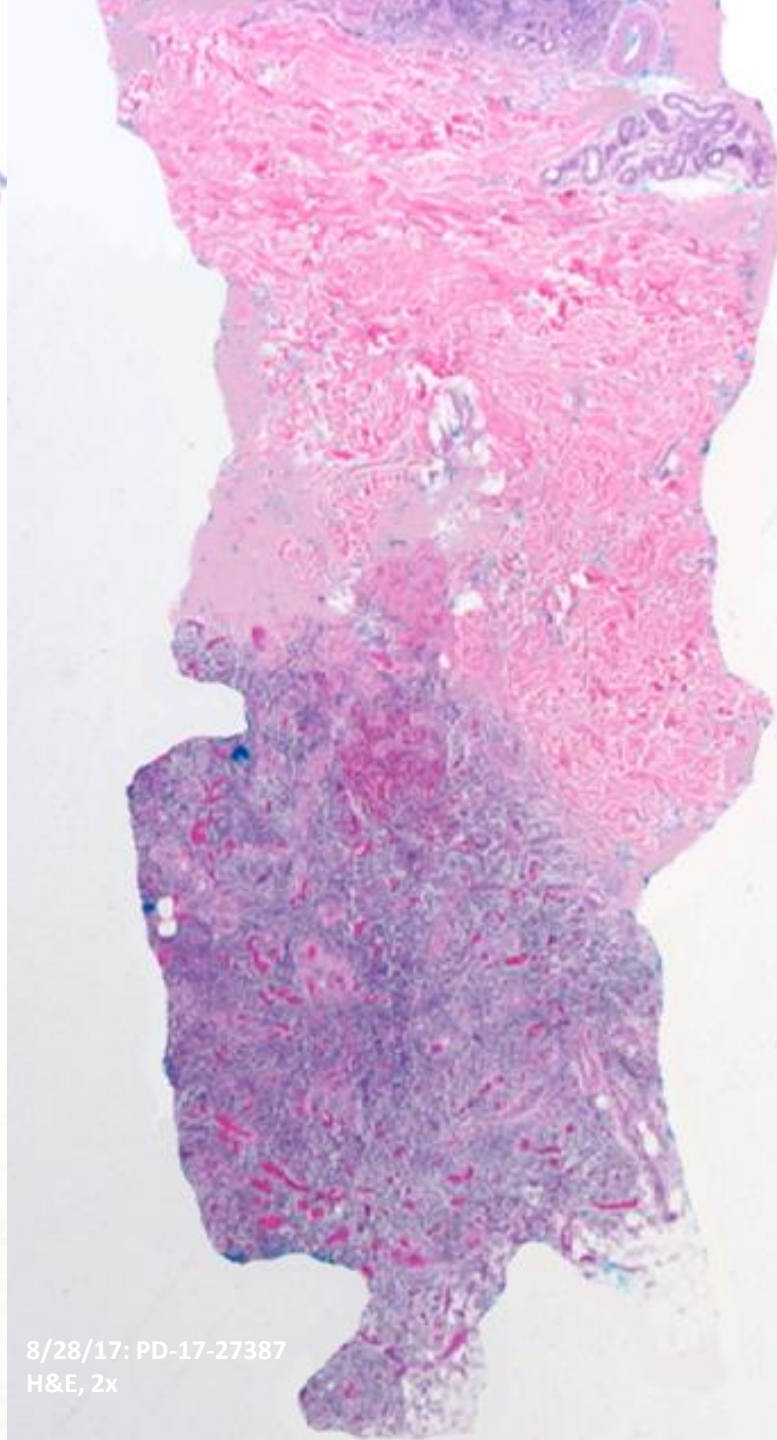
# DERMATOPATHOLOGY





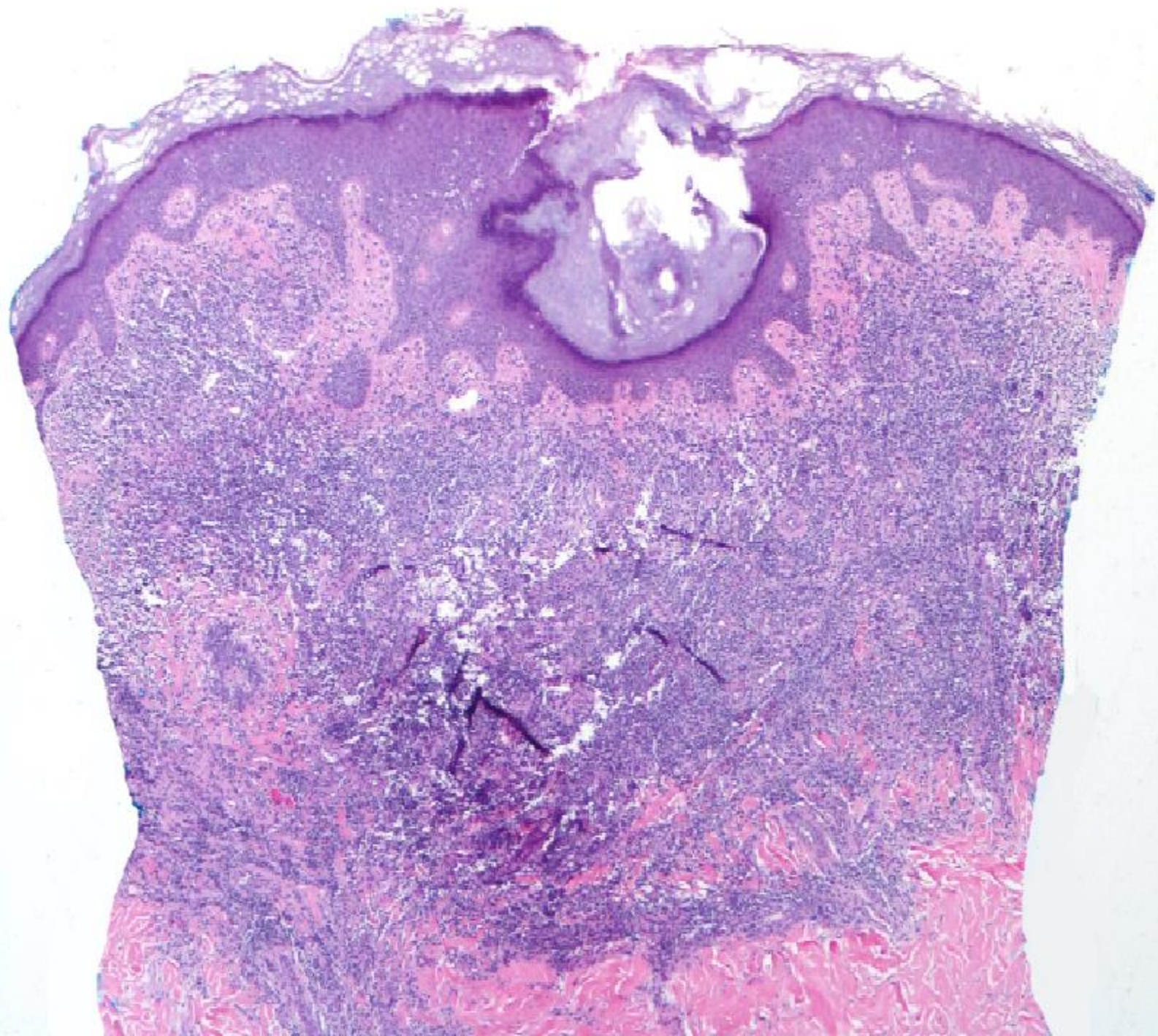


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H&E, 2x



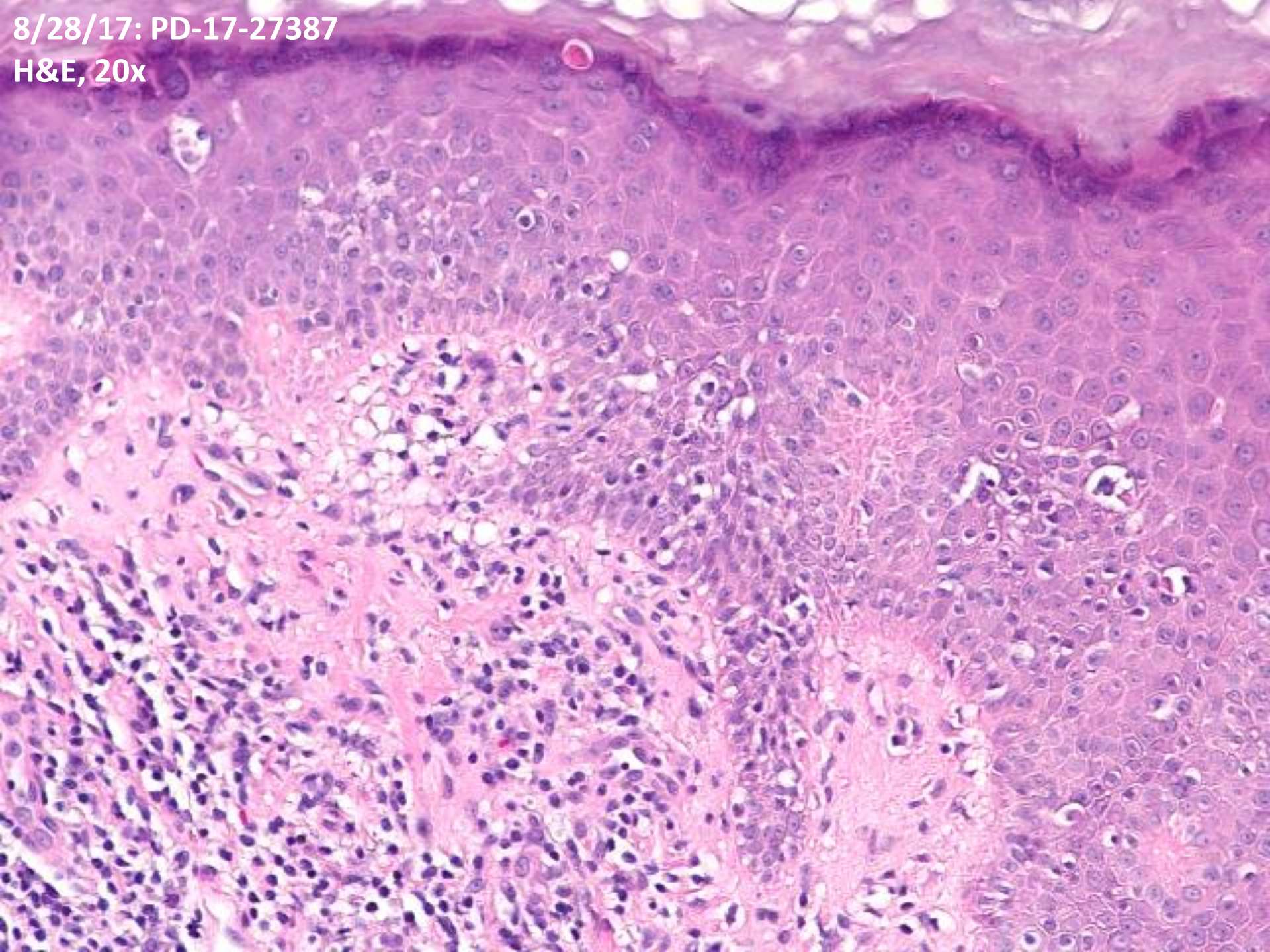
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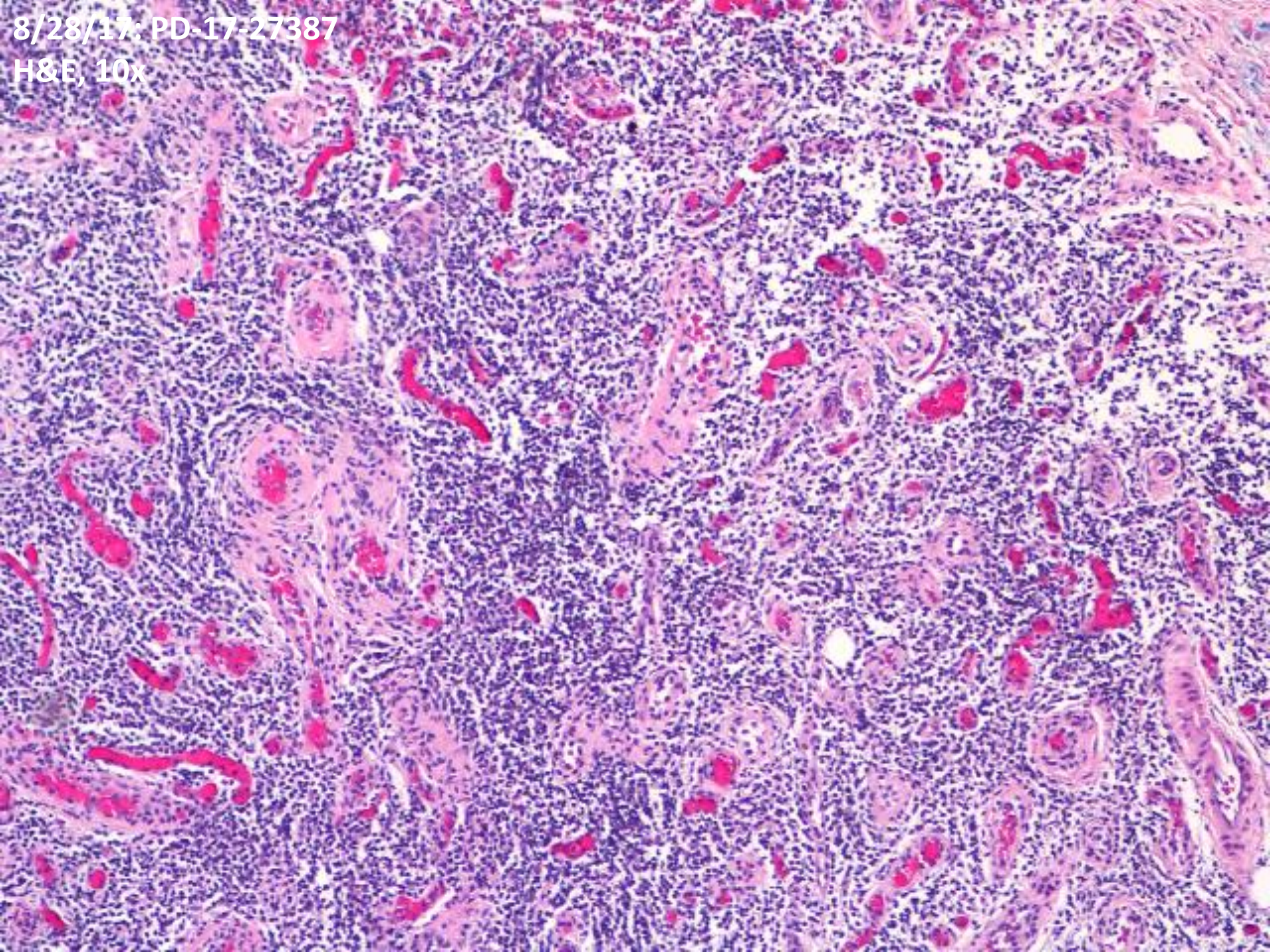


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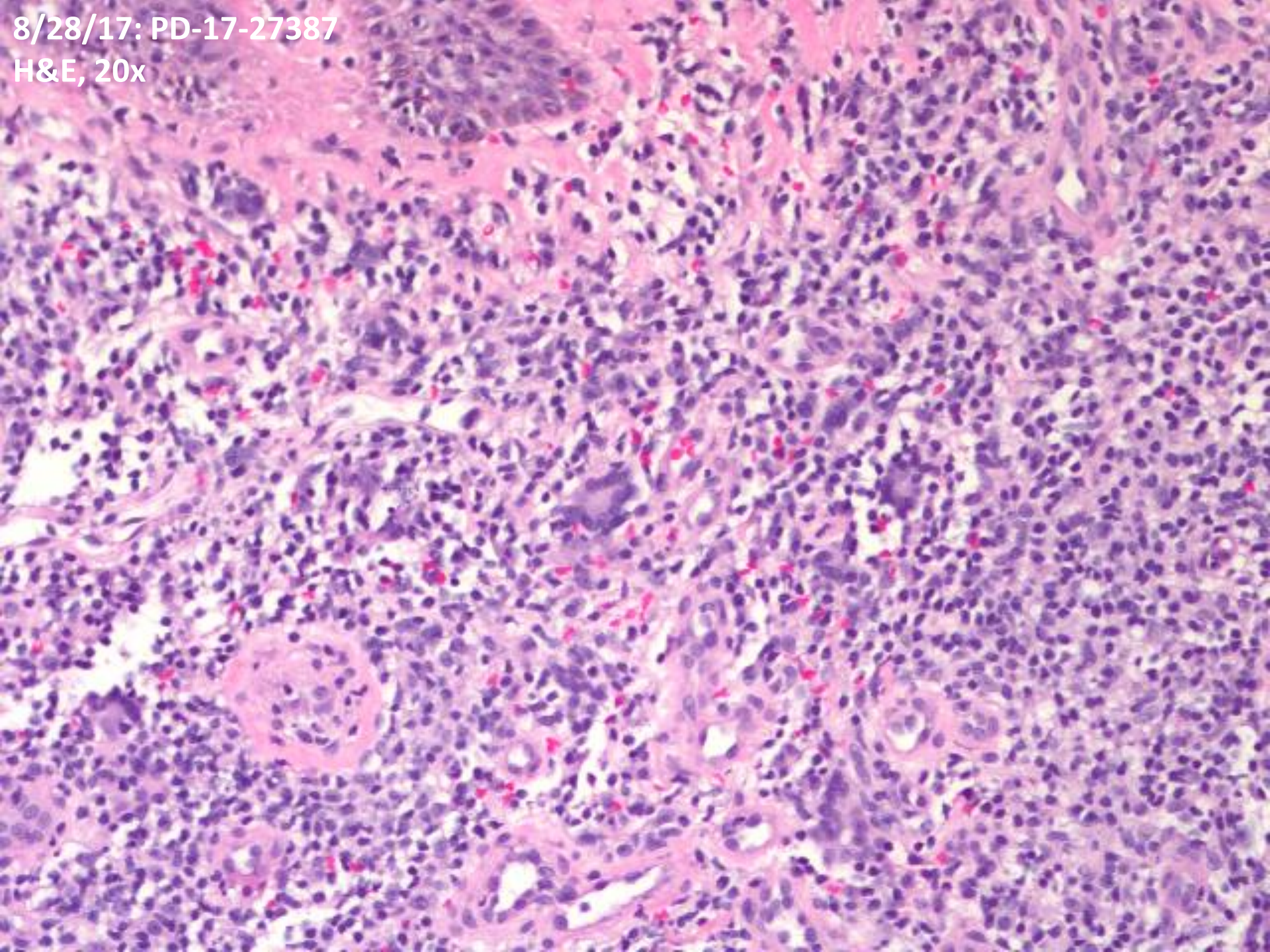


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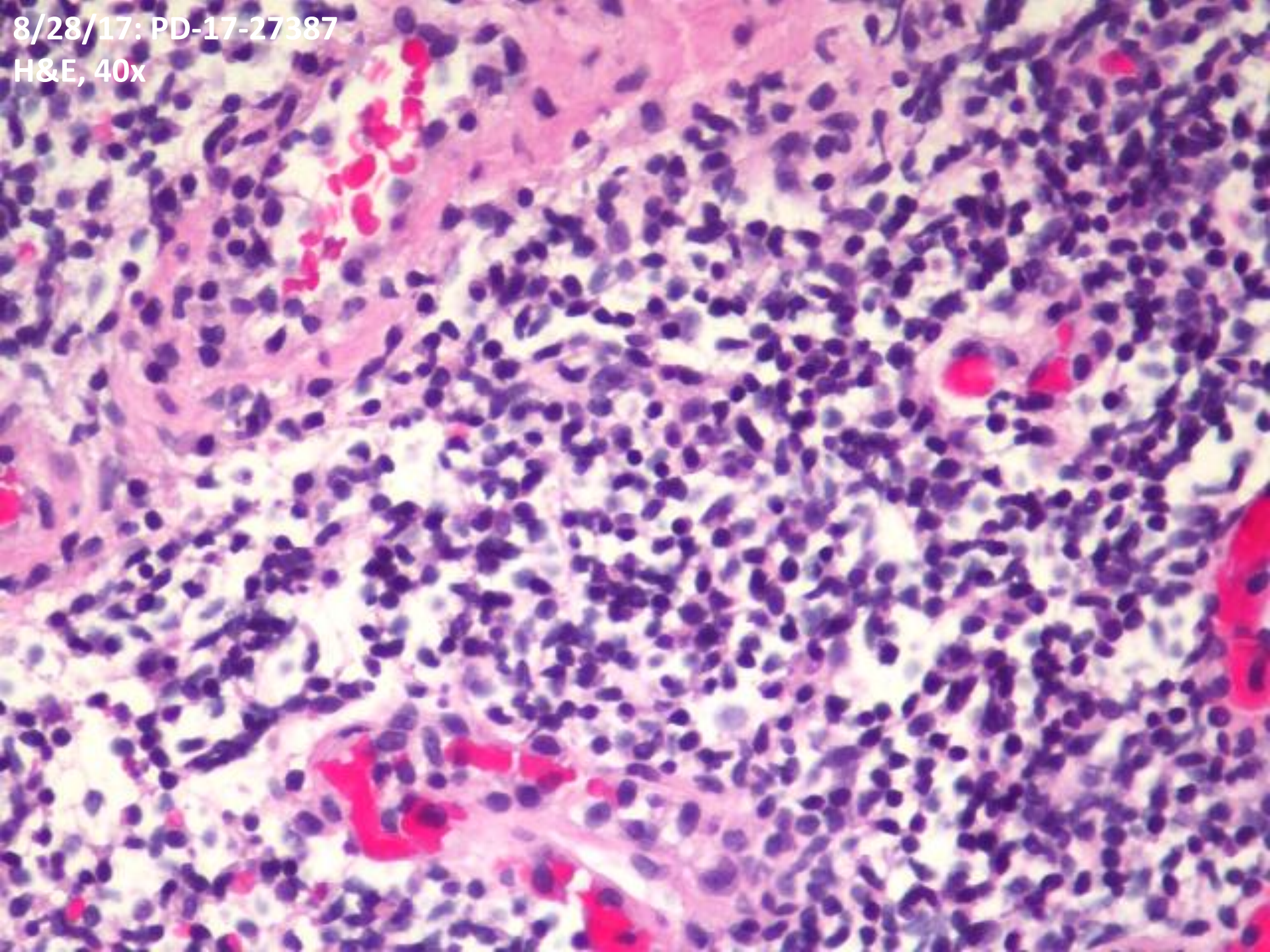


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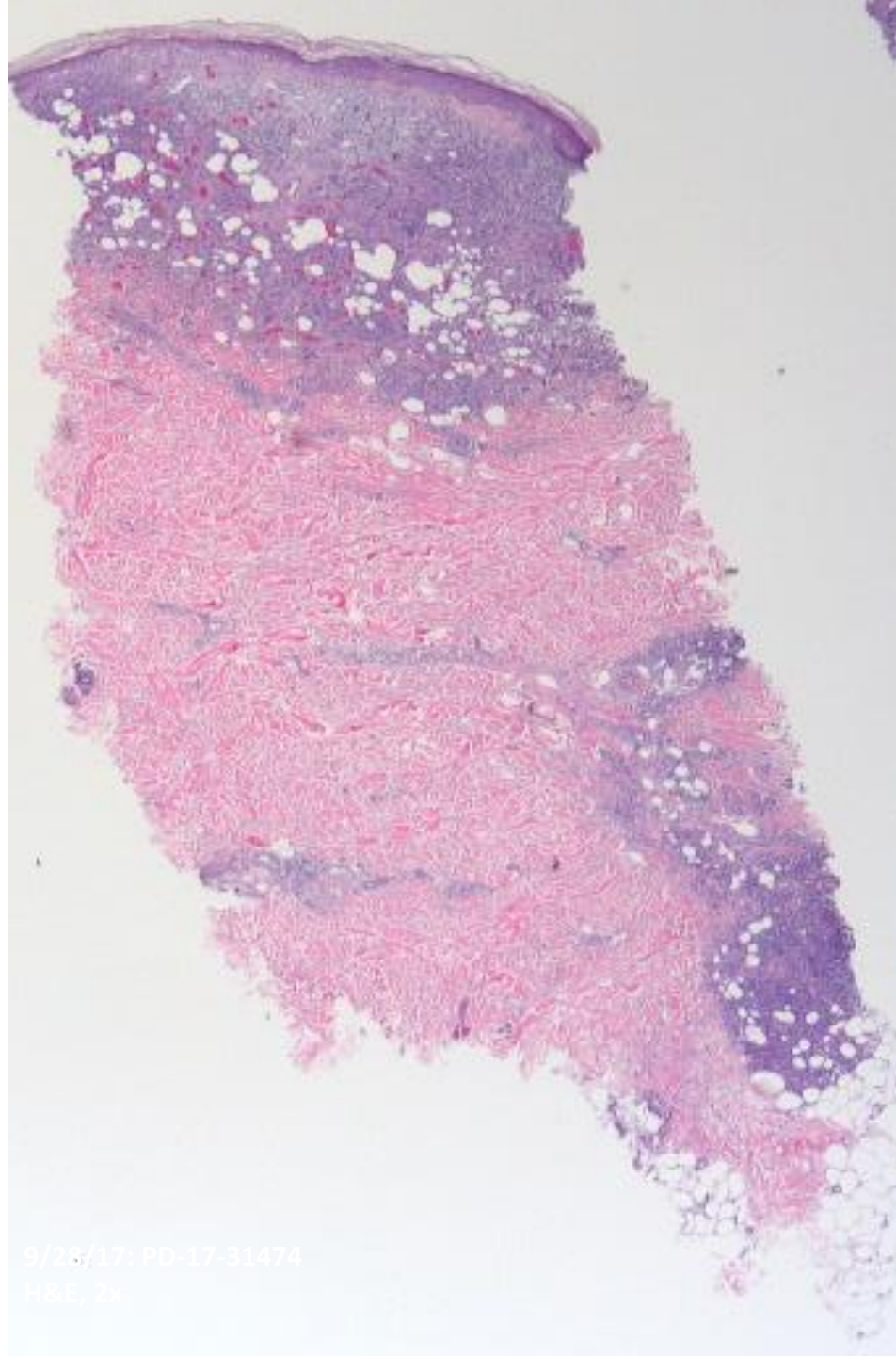




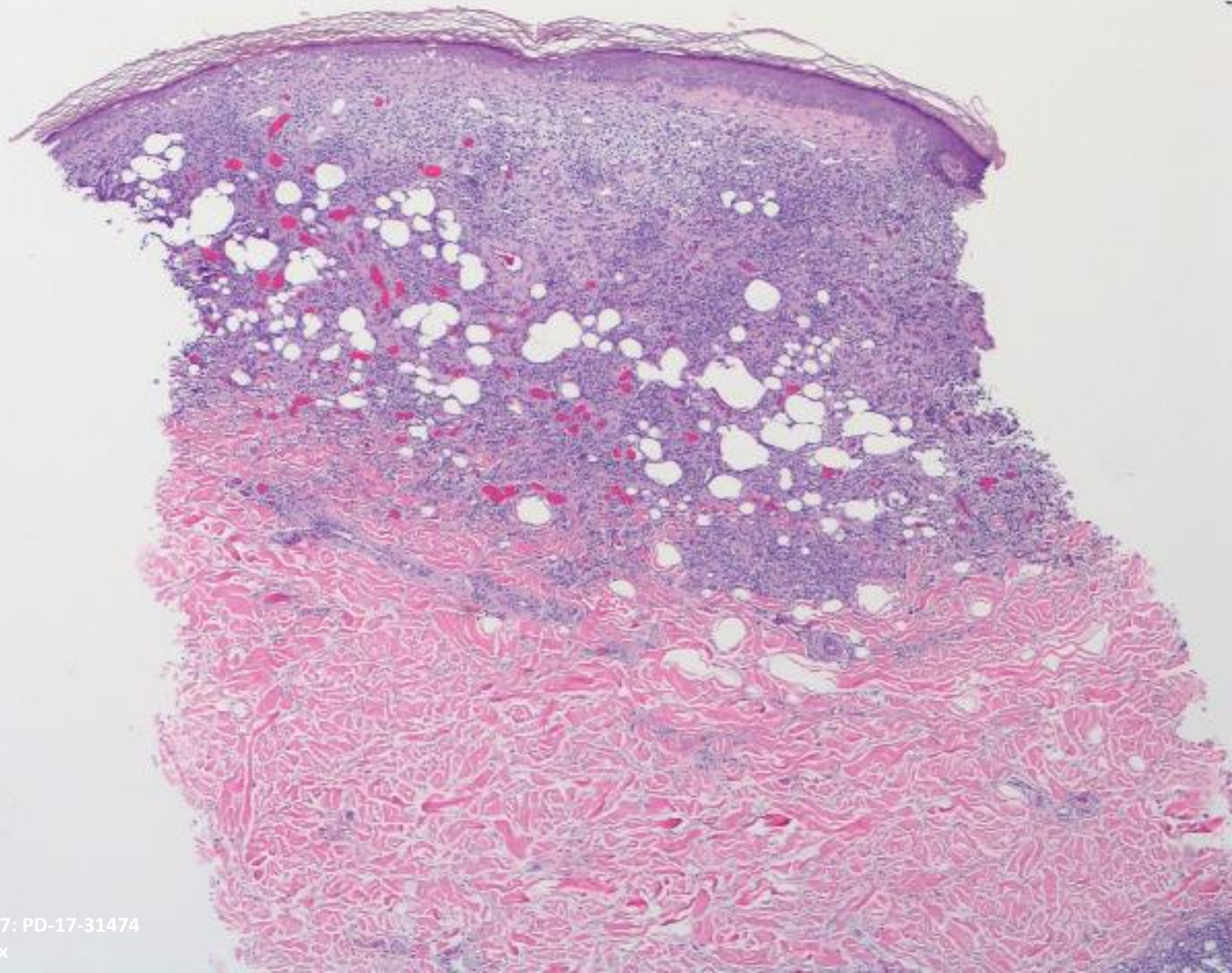
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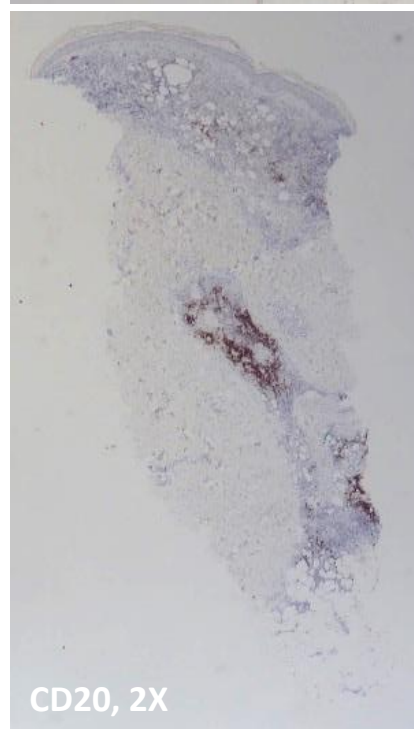
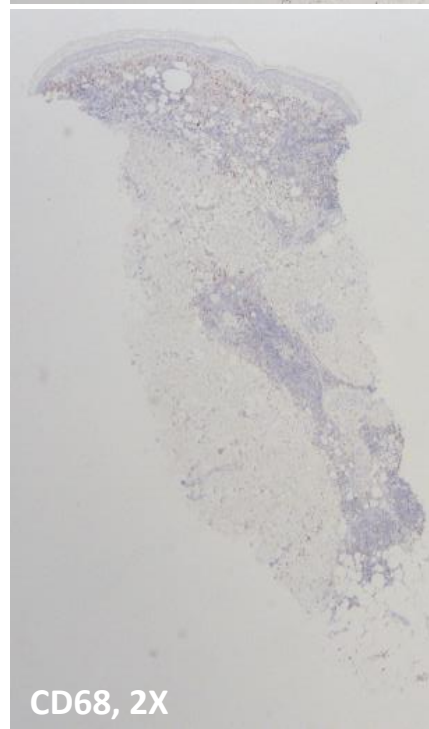
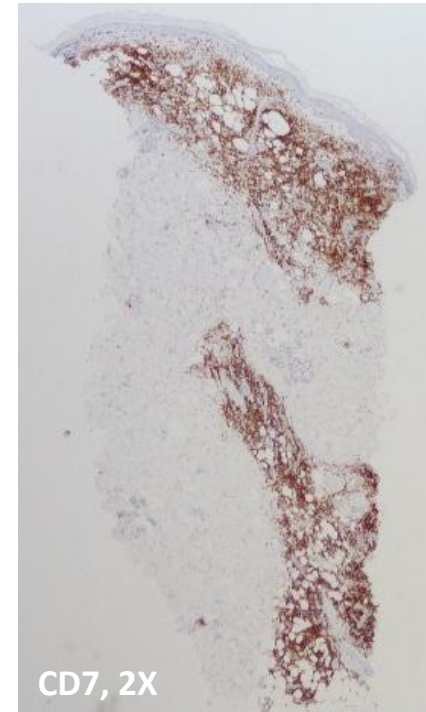
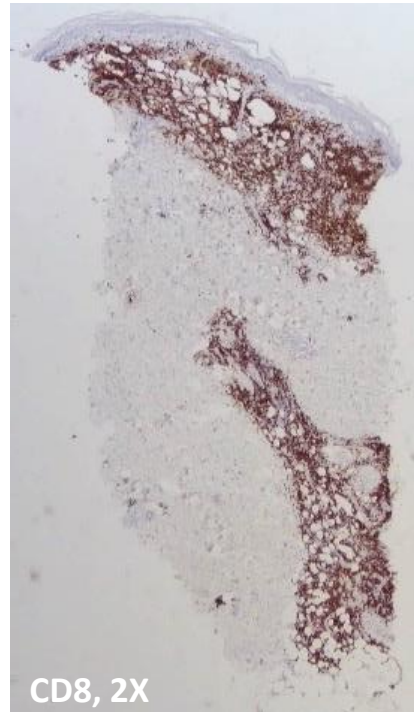
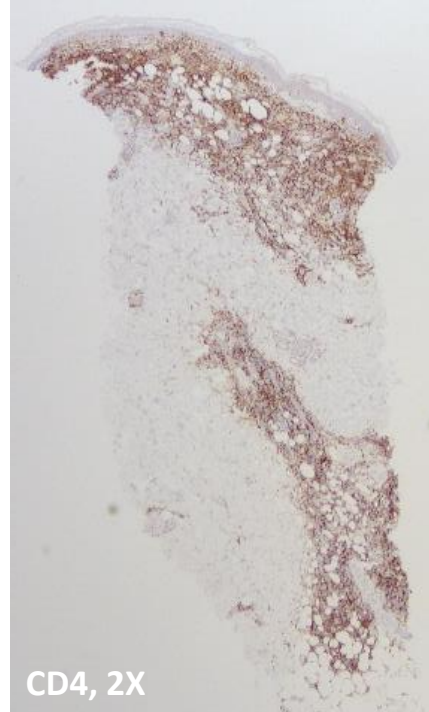
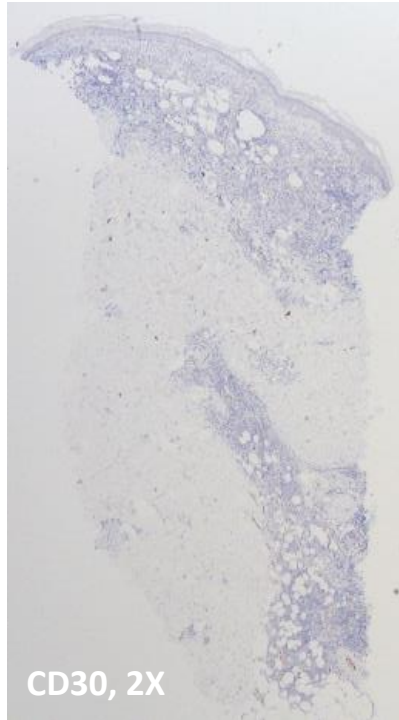




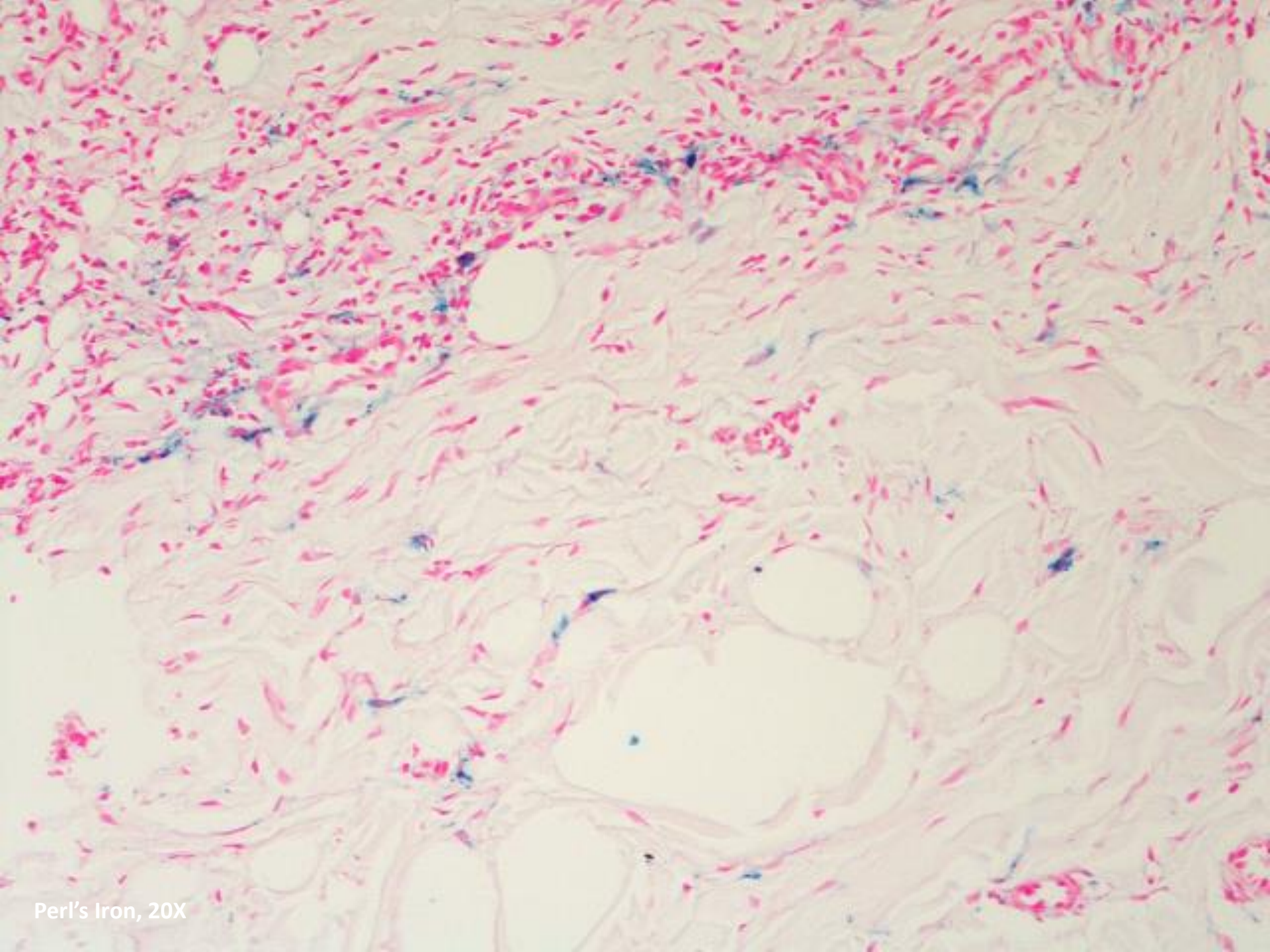
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H&E, 2x





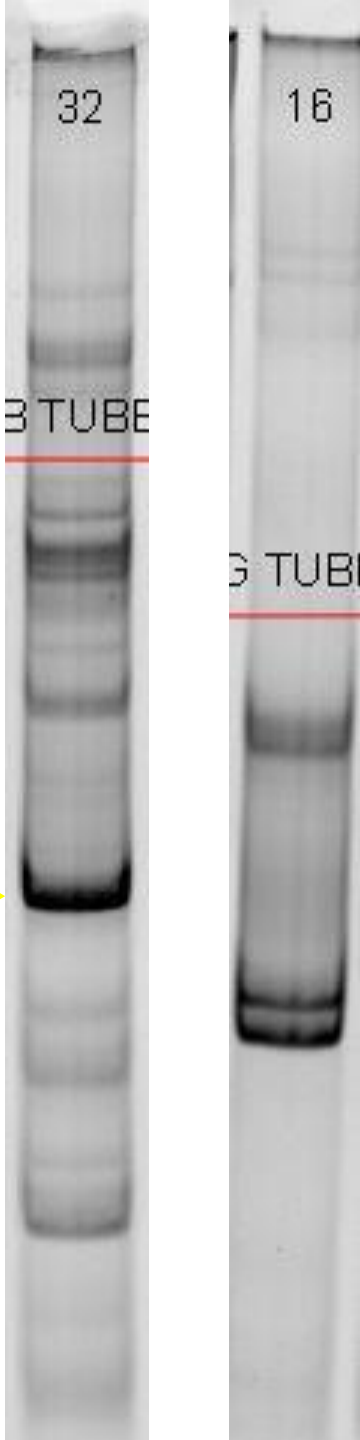






Perl's Iron, 20X

9/28/17: PM-17-00622



<b>TRB tube A and tube B</b>	Positive: 240-285 bp; 1 or 2 discrete bands (monoclonal)
	Negative: smear (polyclonal)

<b>TRG tube 1 and 2</b>	Positive: 170-300 bp; 1 or 2 discrete bands (monoclonal)
	Negative: smear (polyclonal)

# **DIAGNOSIS**

- **Pseudolymphomas**
- **Acral pseudolymphomatous angiokeratoma of children (APACHE)**
- **T-Cell rich angiomatoid polypoid pseudolymphoma (TRAPP)**
- **Giant cell lichenoid dermatitis**
- **Benign vs Malignant? Evaluation of Molecular Studies**

# APACHE

- Acral pseudolymphomatous angiokeratoma of children
- Children ages 2-13
- Extremities
- Single or multiple red papules in a linear configuration



# Pathology

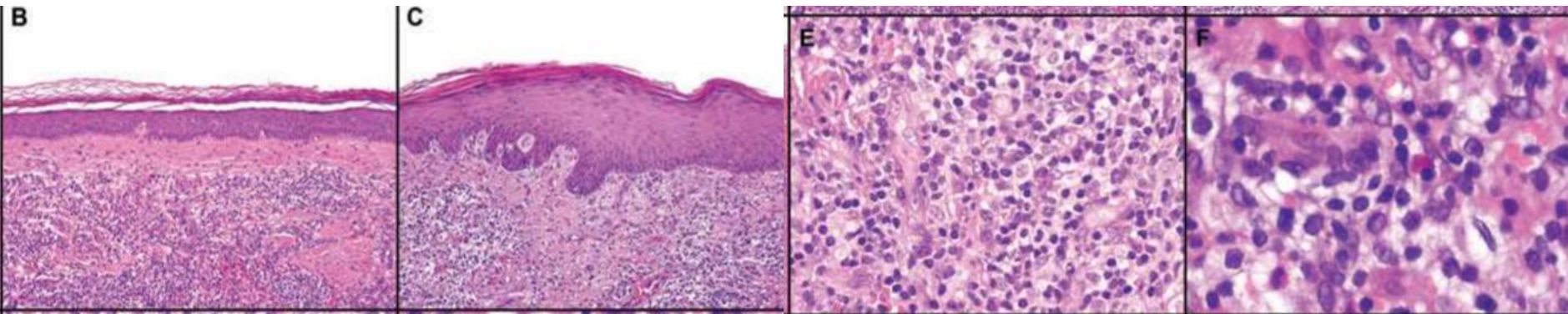
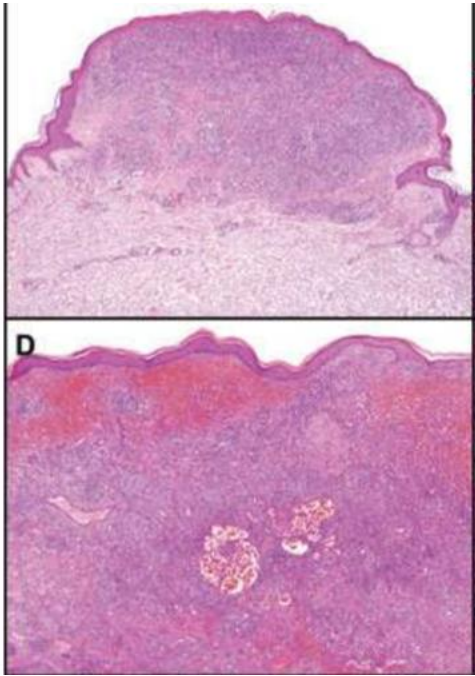
- Lymphocytes, histiocytes, plasma cells
- Sometimes giant cells
- Lichenoid reaction
- IHC: T and B cells
- MG: No clonality

# Misnomer?

- **Angiolymphoid hyperplasia with high endothelial venules** may be a more appropriate name rather than APACHE: ~~acral~~ pseudolymphomatous ~~angiokeratoma~~ of ~~children~~
- Not always acral, not always children

# TRAPP

- Demographics: ages 16 to 71, F:M = 14:3
- Clinical: solitary, polypoid, erythematous, papule ranging in size from 2.5 to 7.5 mm, mostly on head, neck, and trunk
- Path: dense dermal infiltrate of mildly atypical lymphocytes with plasma cells and histiocytes, vessels lined by plump endothelial cells +/- Grenz zone, +/- eosinophils
- IHC: Mixture of CD4, CD4 T cells
- 7 cases analyzed for clonality: all 7 suggested polyclonality



# Challenges in Interpreting Pseudolymphomas

## Clinical and laboratory studies

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Lymphoma versus pseudolymphoma of the skin:  
Gene rearrangement study of 21 cases with  
clinicopathologic correlation

Nerea G. Landa, MD, Brian D. Zelickson, MD, Margot S. Peters, MD,  
Sigfrid A. Muller, MD, and Mark R. Pittelkow, MD *Rochester, Minnesota*

- Retrospective study, 21 lymphoproliferative cases reviewed  
→ 7 with immunoglobulin and 2 with T cell receptor gene rearrangements
  - 6 of 9 positive: malignant histology → 1 with extracutaneous manifestations
  - 2 of 12 negative: malignant histology
- **Conclusion:** No correlation between clonality and aggressiveness.  
Recurrence seen with multiple lesions, not based on molecular studies



Atypical T-cell and angiomatous  
proliferation: does not fit into precise  
category