

# Case Review

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# Case 1

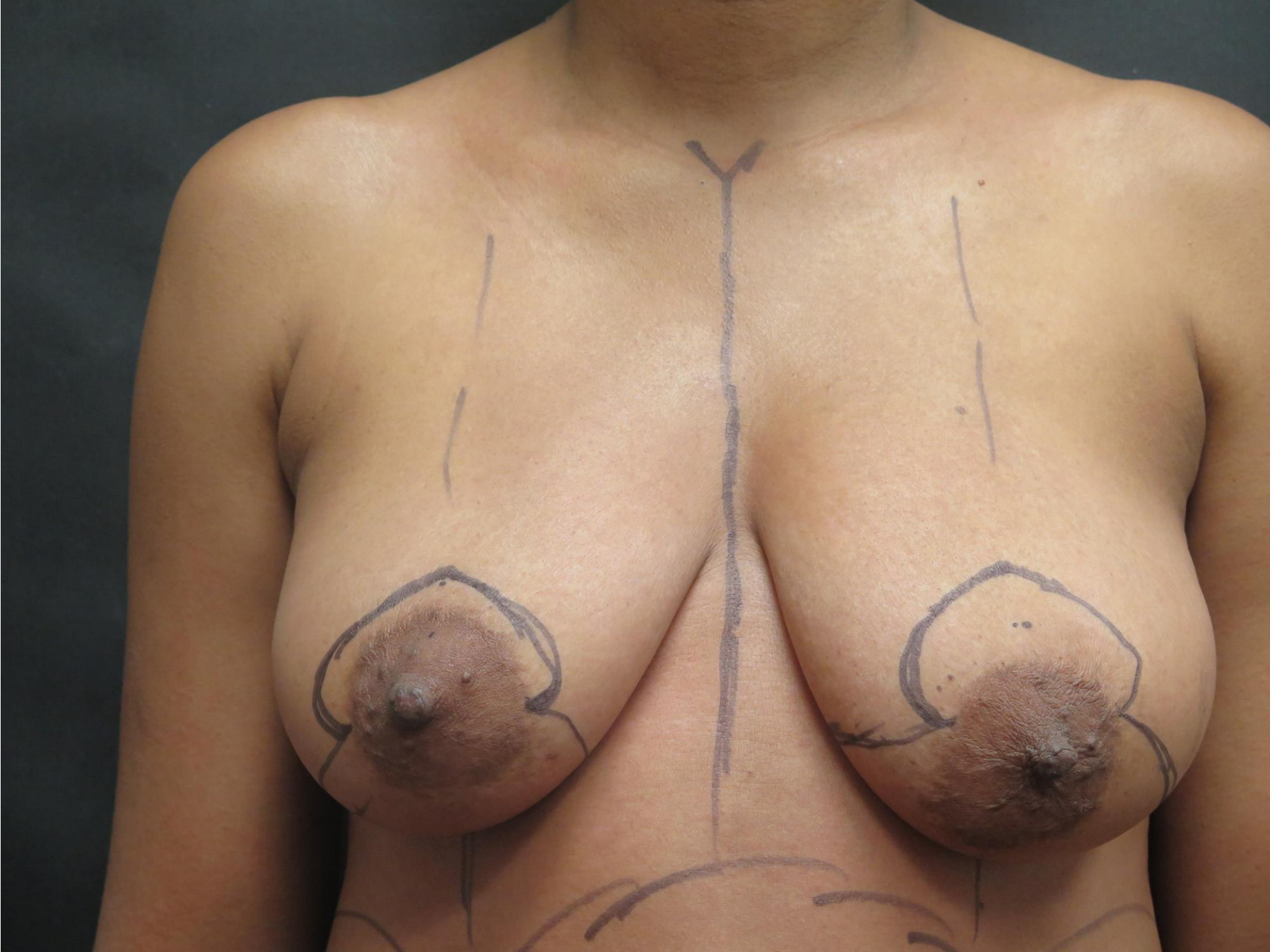
# Clinical history

- 44-year-old woman with history of hypertension treated with HCTZ and amlodipine
- Allergic to latex
- BMI of 26.1
- No other clinical abnormalities, no symptoms, everything WNL
- Prestented for liposuction of the abdomen, back and flanks, gluteal fat transfer, and a vertical pattern breast lift and small reduction

Physical examination







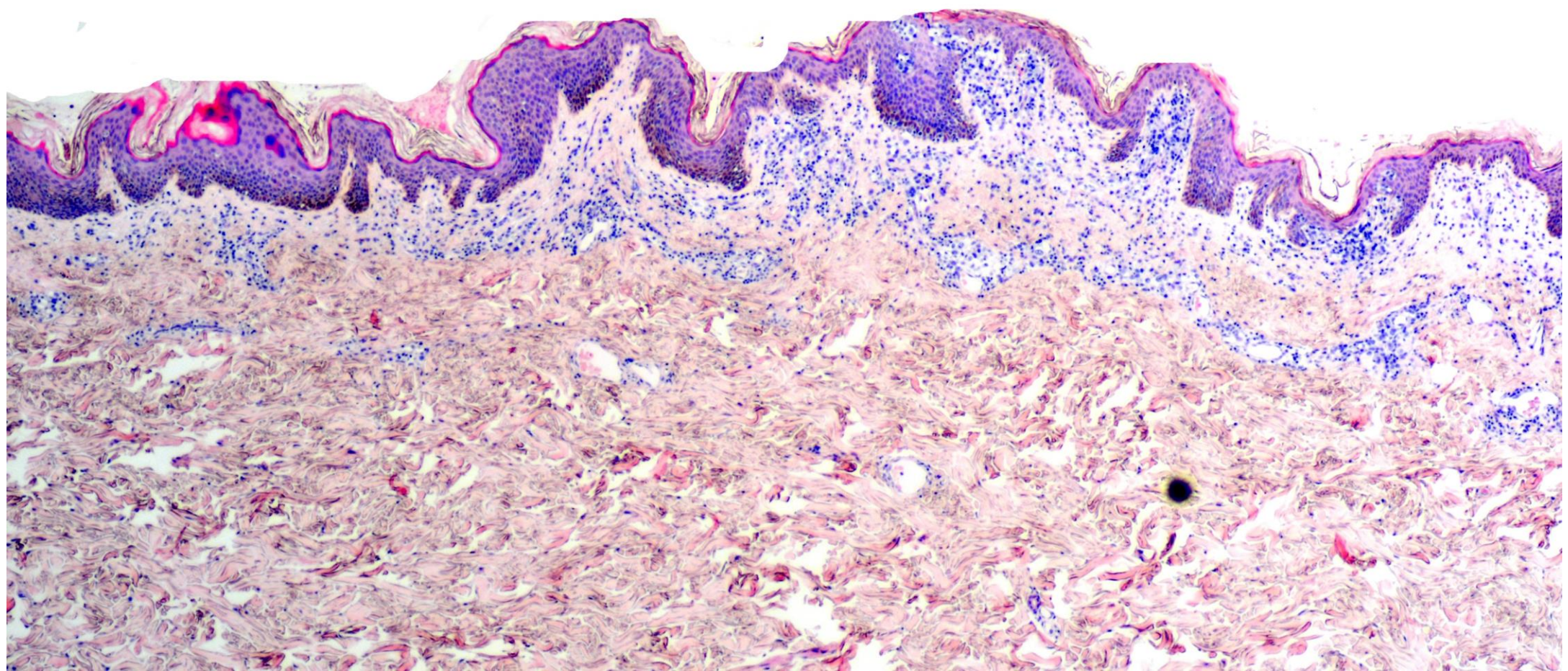




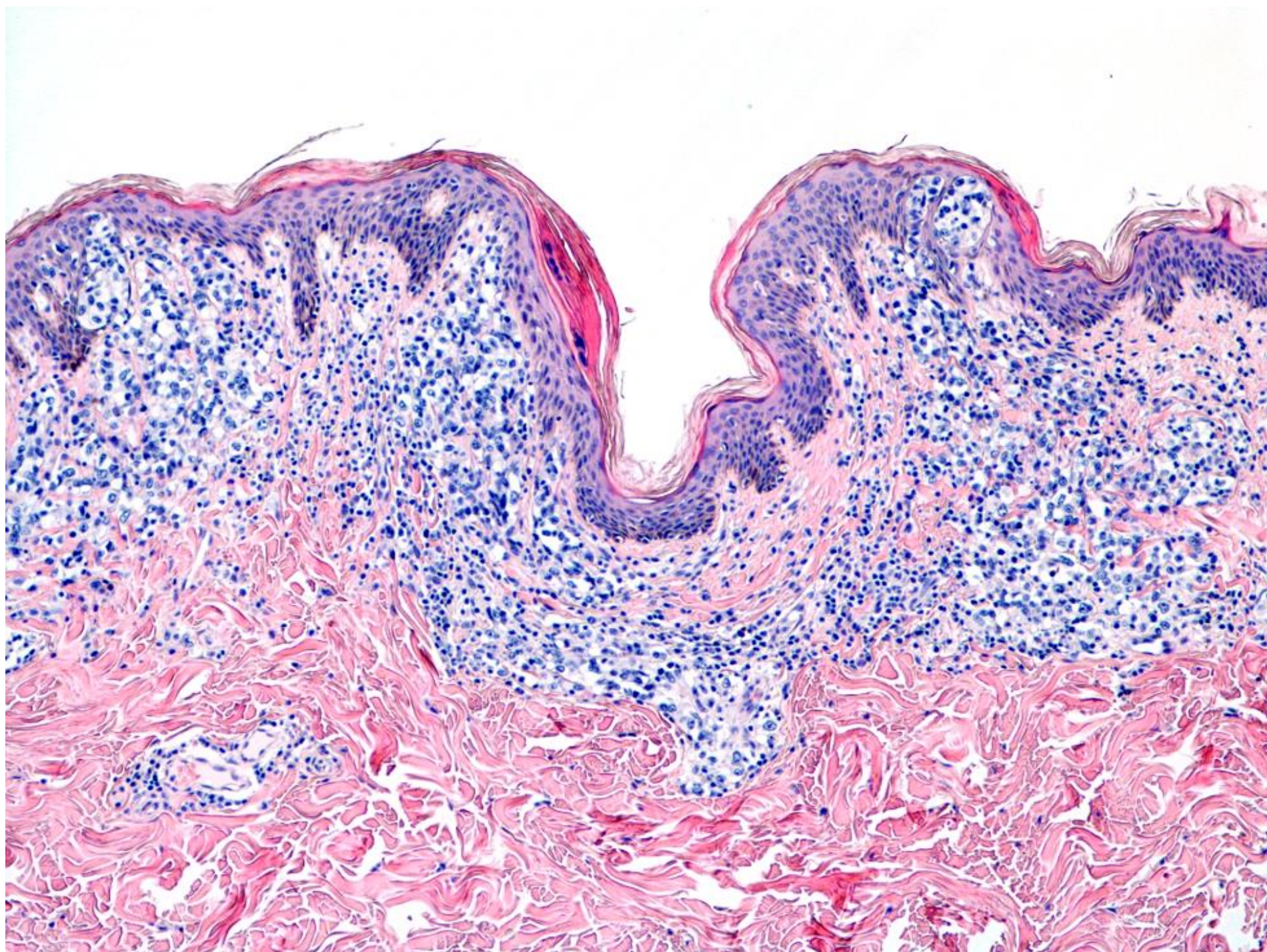
Removed 220 , 45 grams left and right breast

Clinical diagnosis?

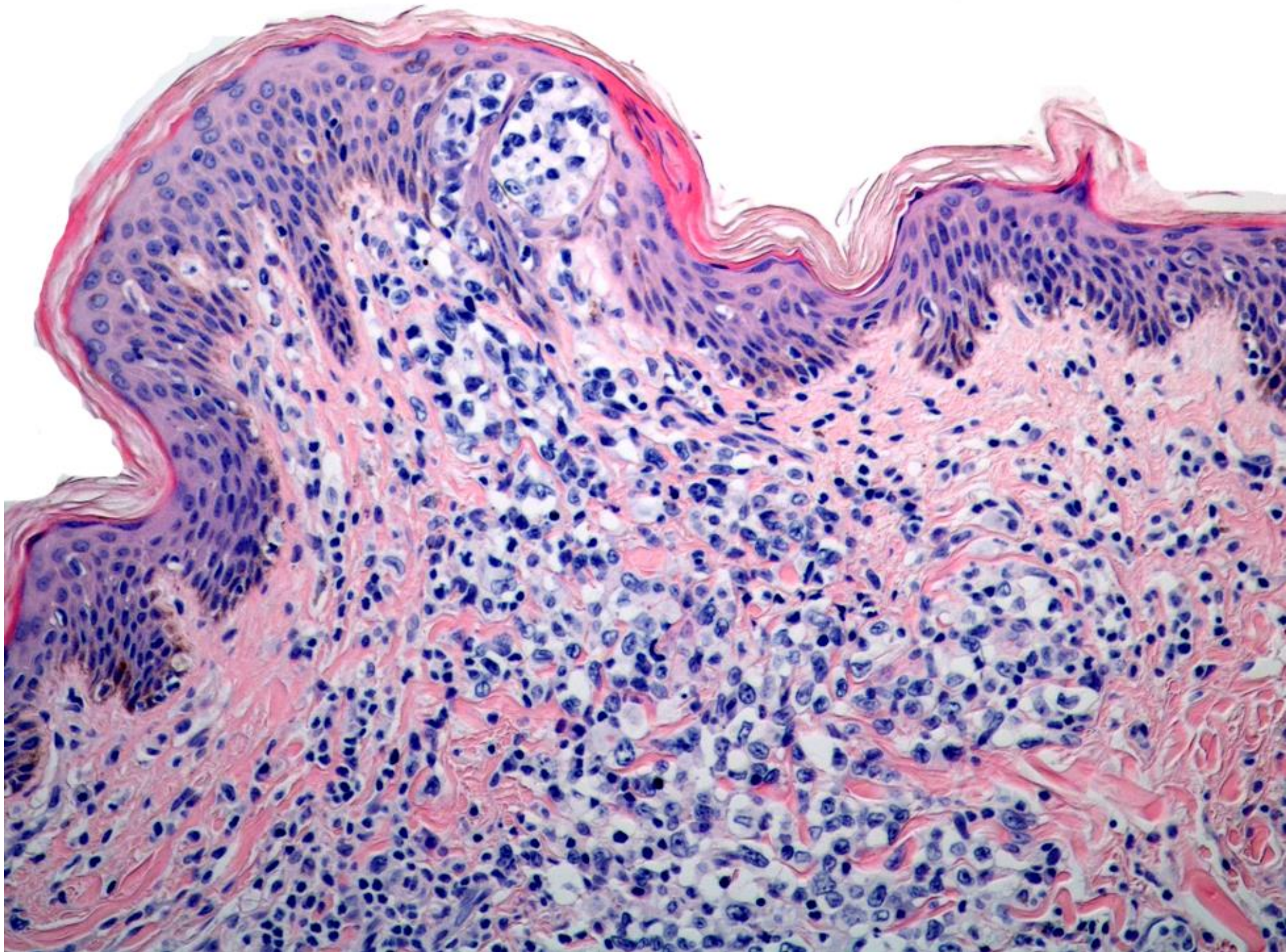




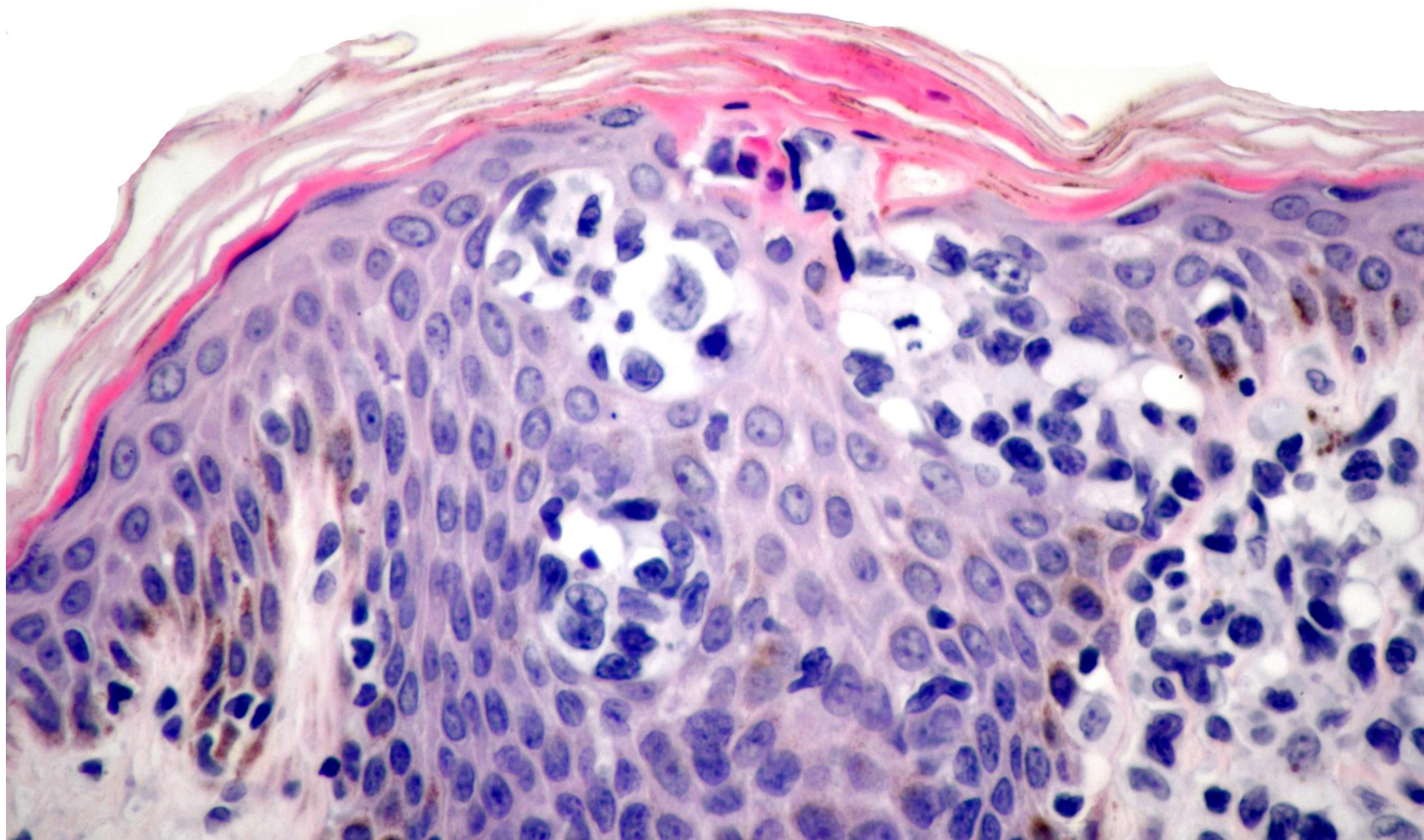




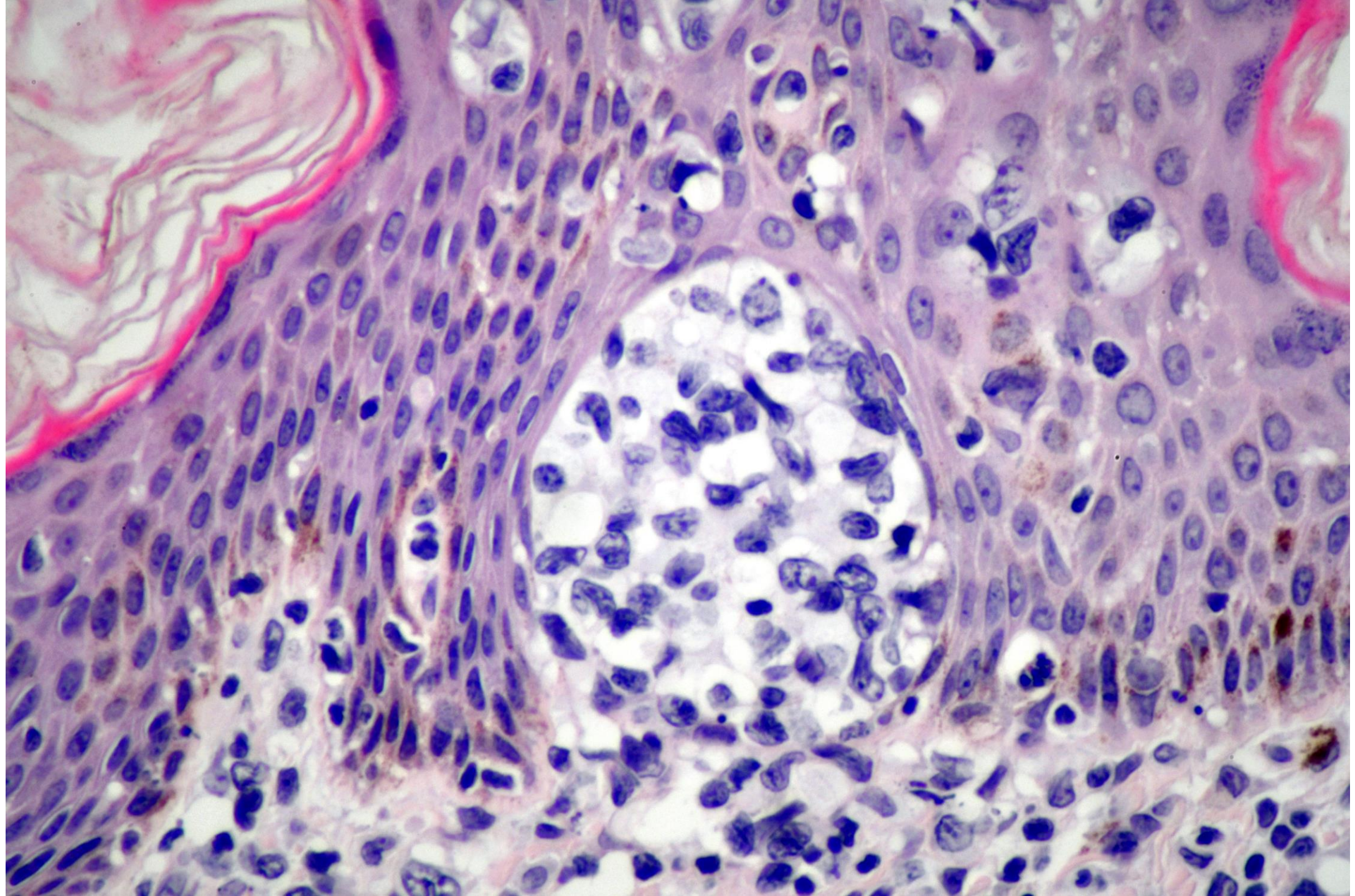




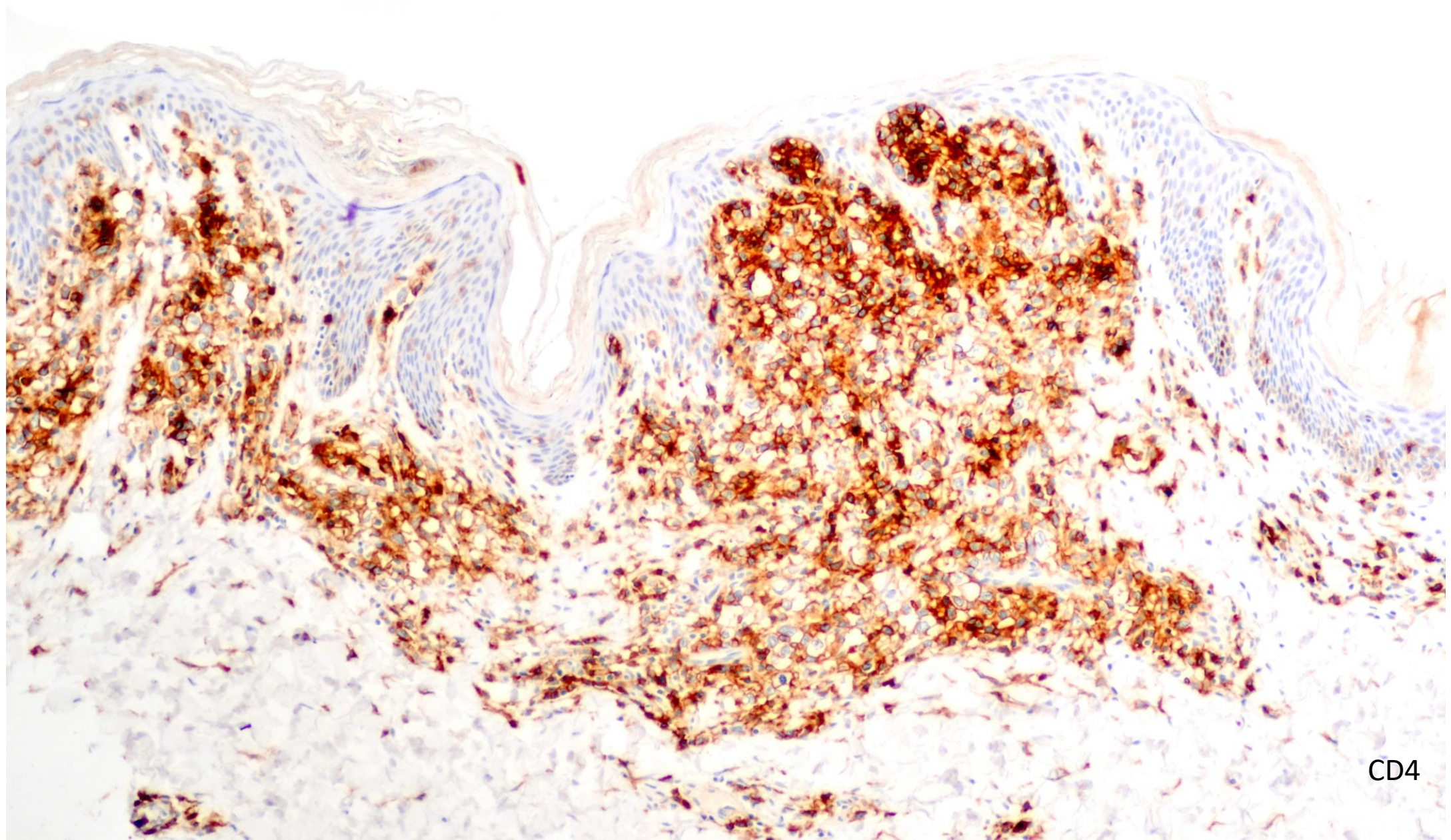






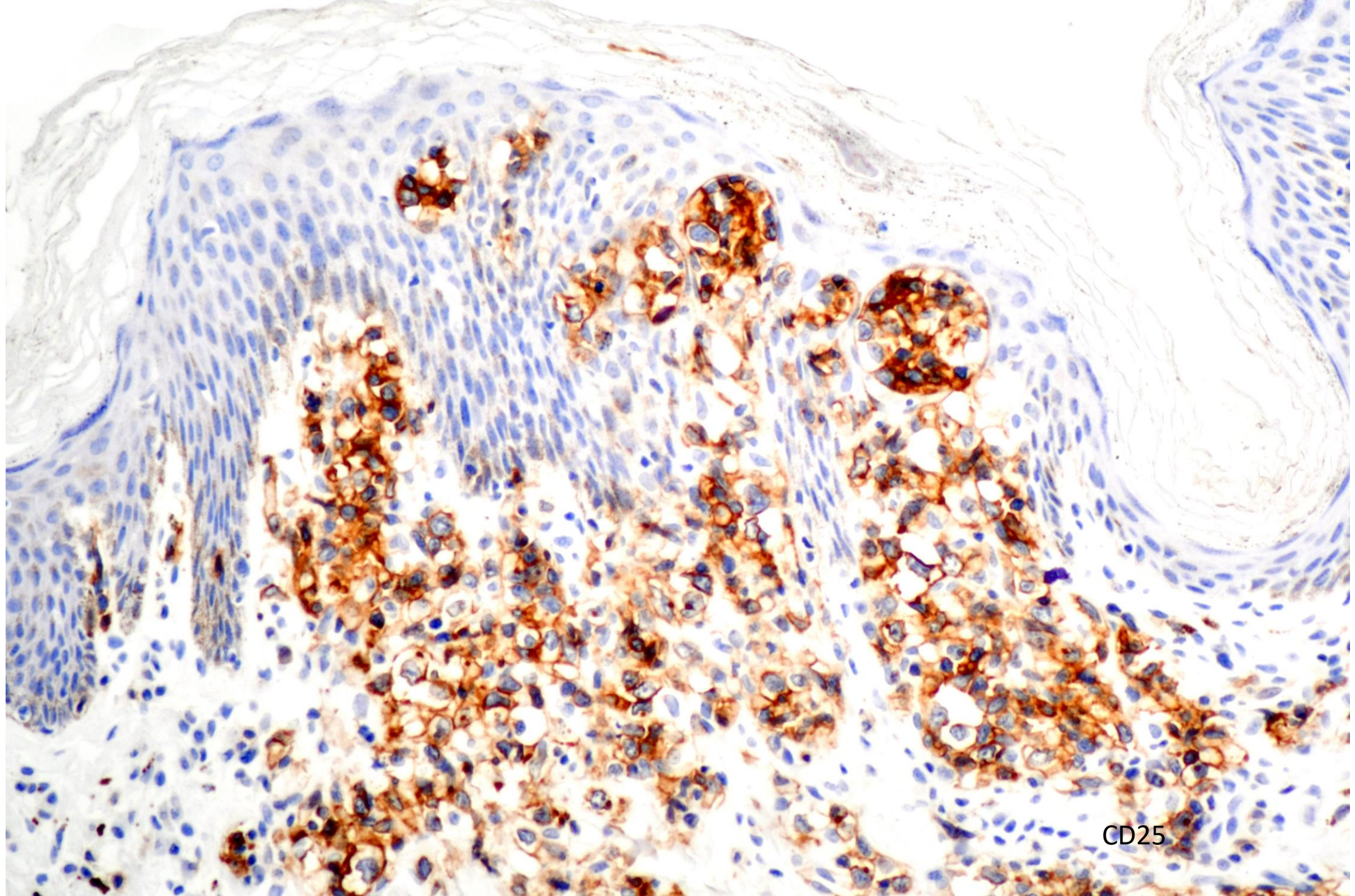






CD4





CD25

Diagnosis: ATLL

# Lab results

- PET: Cutaneous and subcutaneous small hypermetabolic nodules, hypermetabolic pelvic nodes
- Positive serology for circulating anti HTLV-1/2 antibodies
- Positive for Clonal T Cell Receptor Beta Gene Rearrangement (V and J regions) skin biopsy and blood
- IL-2 R  $\alpha$  elevated
- No circulating atypical cells in the blood
- LDH negative

# Clinical presentation of HTLV-1

- Older patients : Japan and Caribbean, other
- Four types: acute and chronic leukemic, lymphomatous and **smoldering\*** – skin involvement only may be another subset worse than smoldering but better than others
- Clinical: macules, papules, plaques, tumors; can be similar to MF

\*Smoldering ATLL is defined as having 5% or more abnormal T lymphocytes and lactate dehydrogenase (LDH) levels up to 1.5× the upper limit of normal, with normal lymphocyte count and calcium levels, and no lymph node or visceral disease other than skin or pulmonary disease.

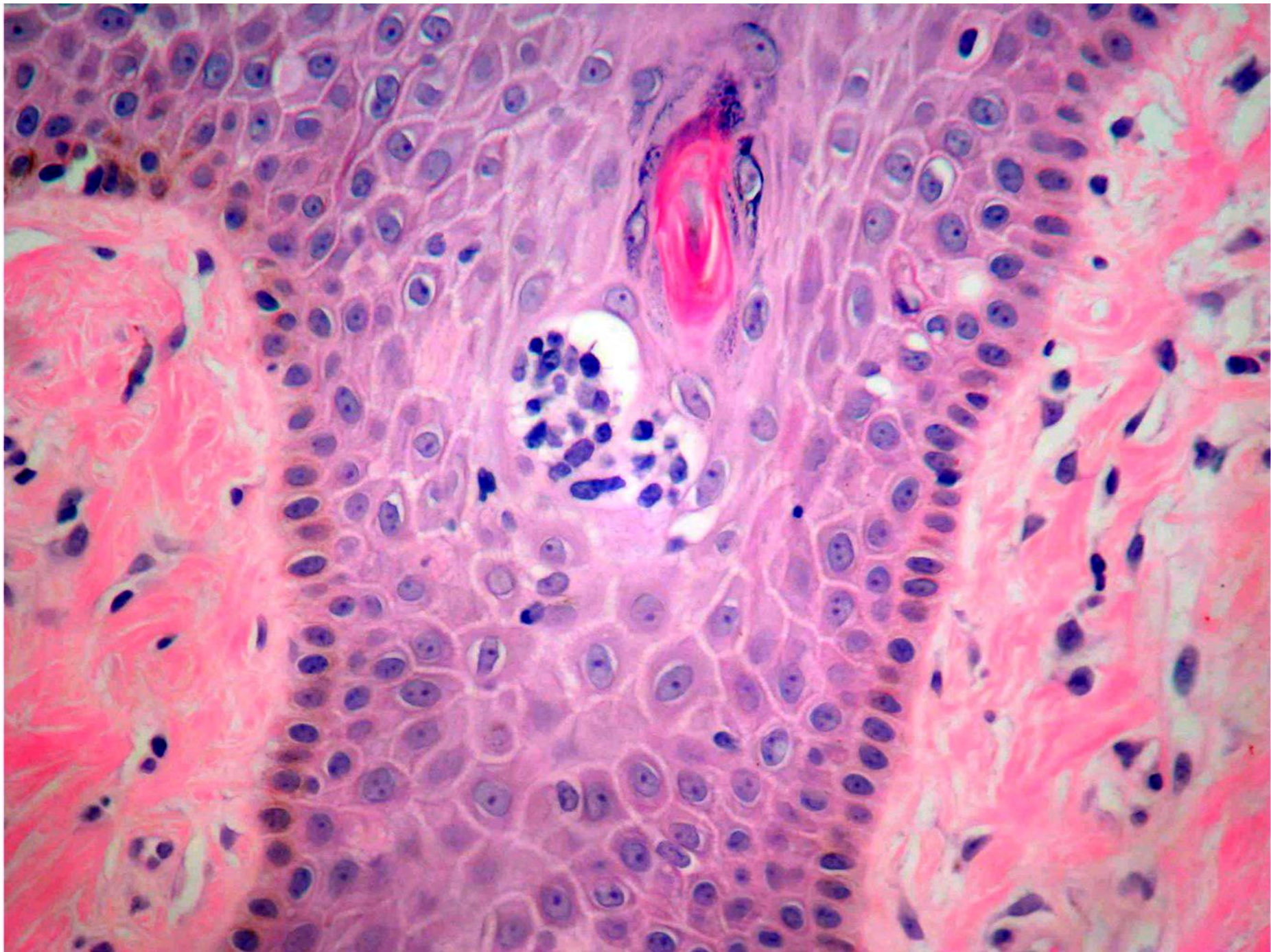
# Pathology

- Bandlike infiltrate but can be nodular or diffuse
- Epidermotropism with large nests (Pautrier-like)
- Varying size of lymphocytes, most often large and pleomorphic
- Angiocentrism and angiodestruction possible
- CD4, CD25, and PD-1 expression possibly; FOXP3, CD30
- TCR +
- Integrated genomic material of HTLV-1



Differential diagnosis





Nevoid hyperkeratosis

Patient lost to follow up: No known therapy given. Better survival but with progression.

## Case 2



# Clinical history

- 2-day old newborn with widespread rash
- Clinical diagnosis epidermal nevus

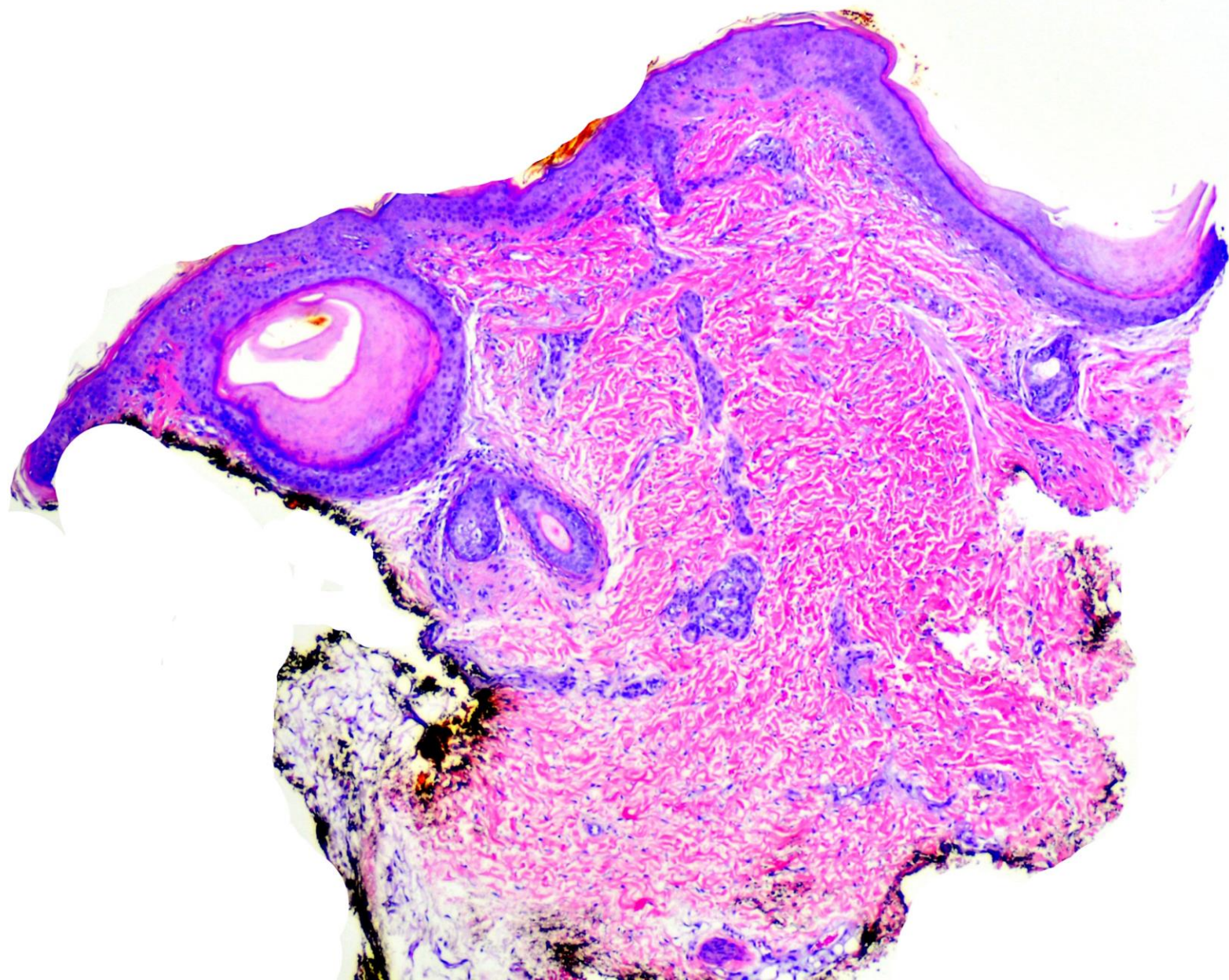




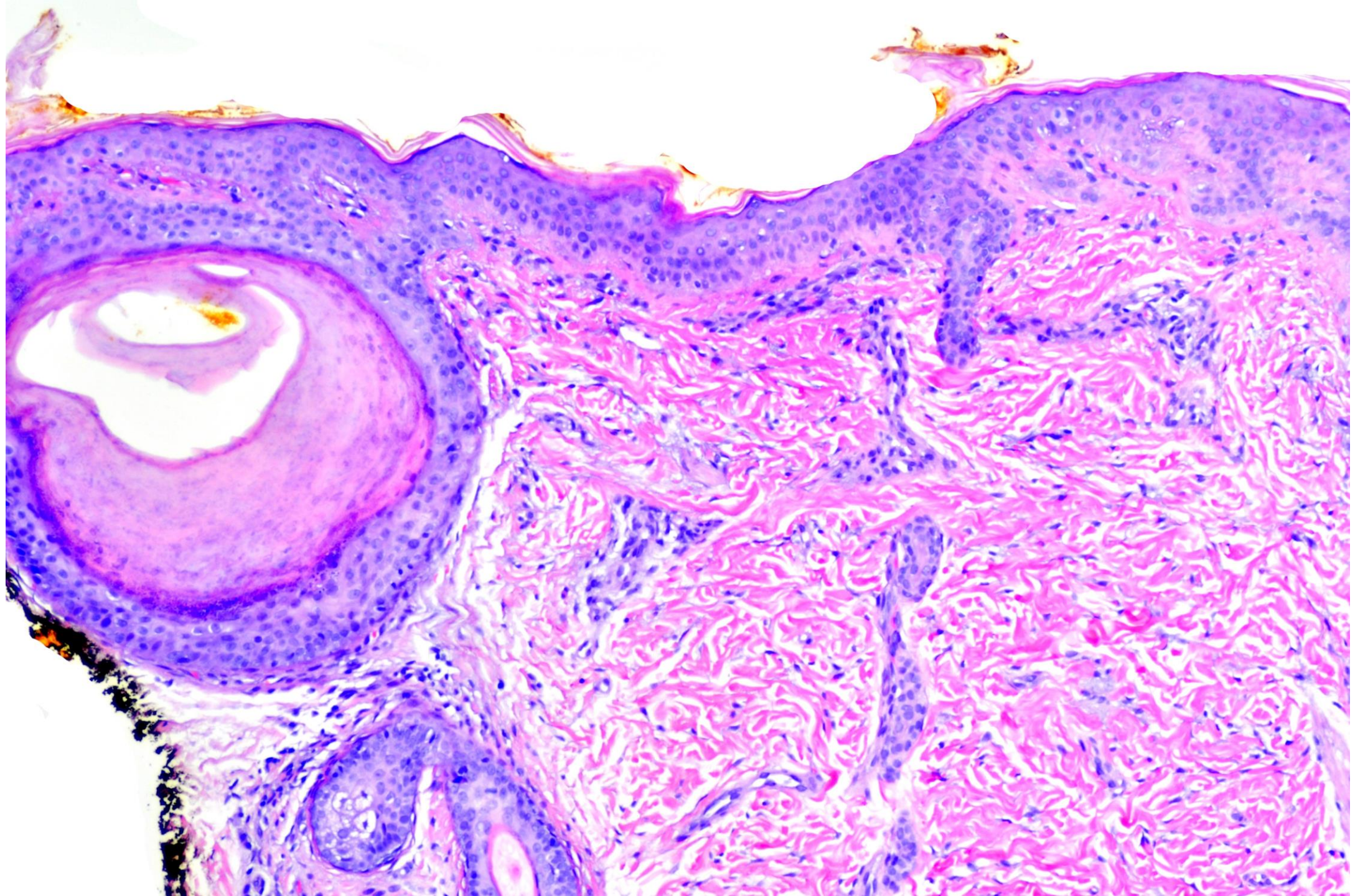
# Physical examination

- Linear hyperpigmented patches left flank
- Shortly after birth
- No other clinical manifestations

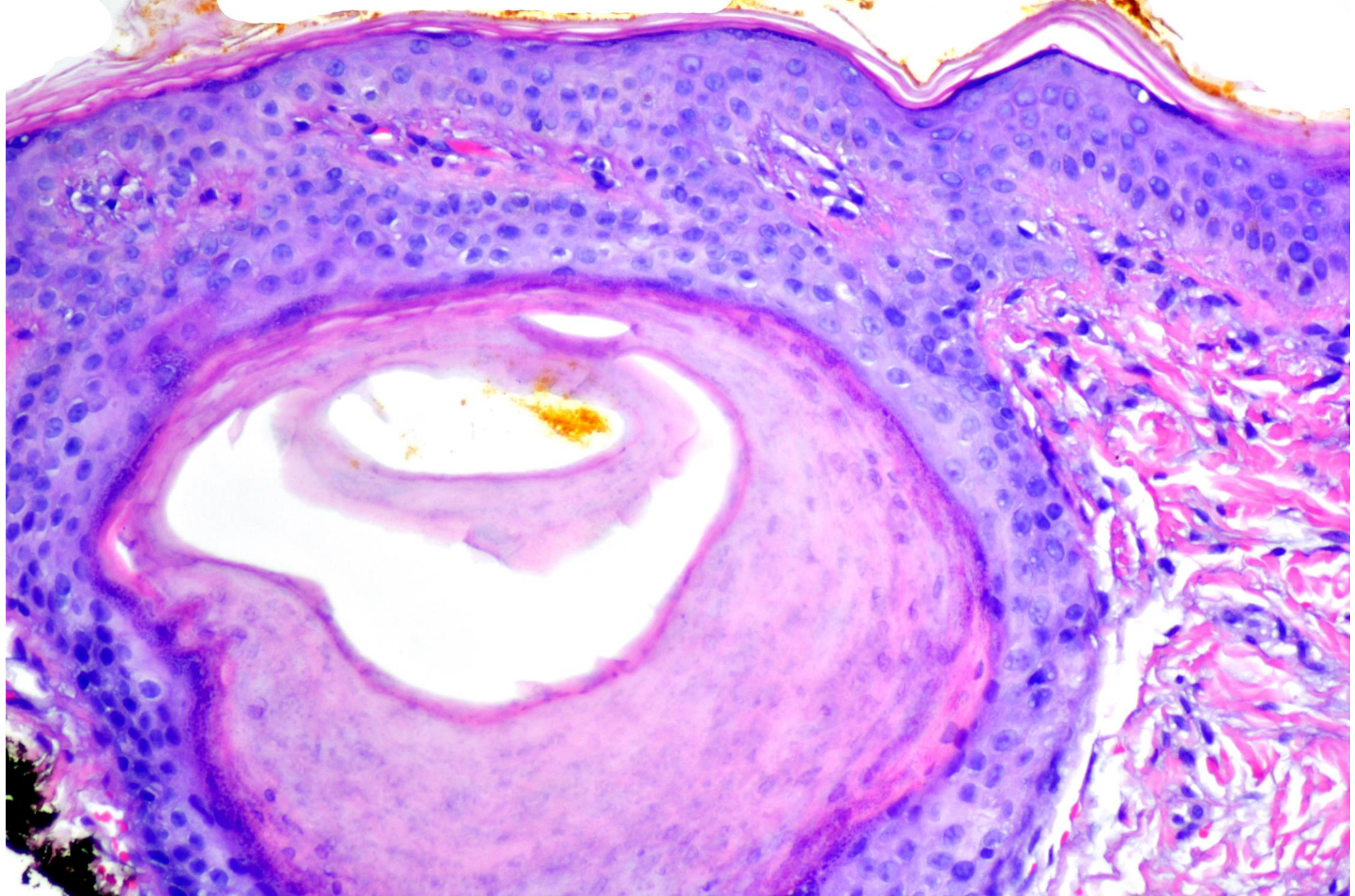
Pathology



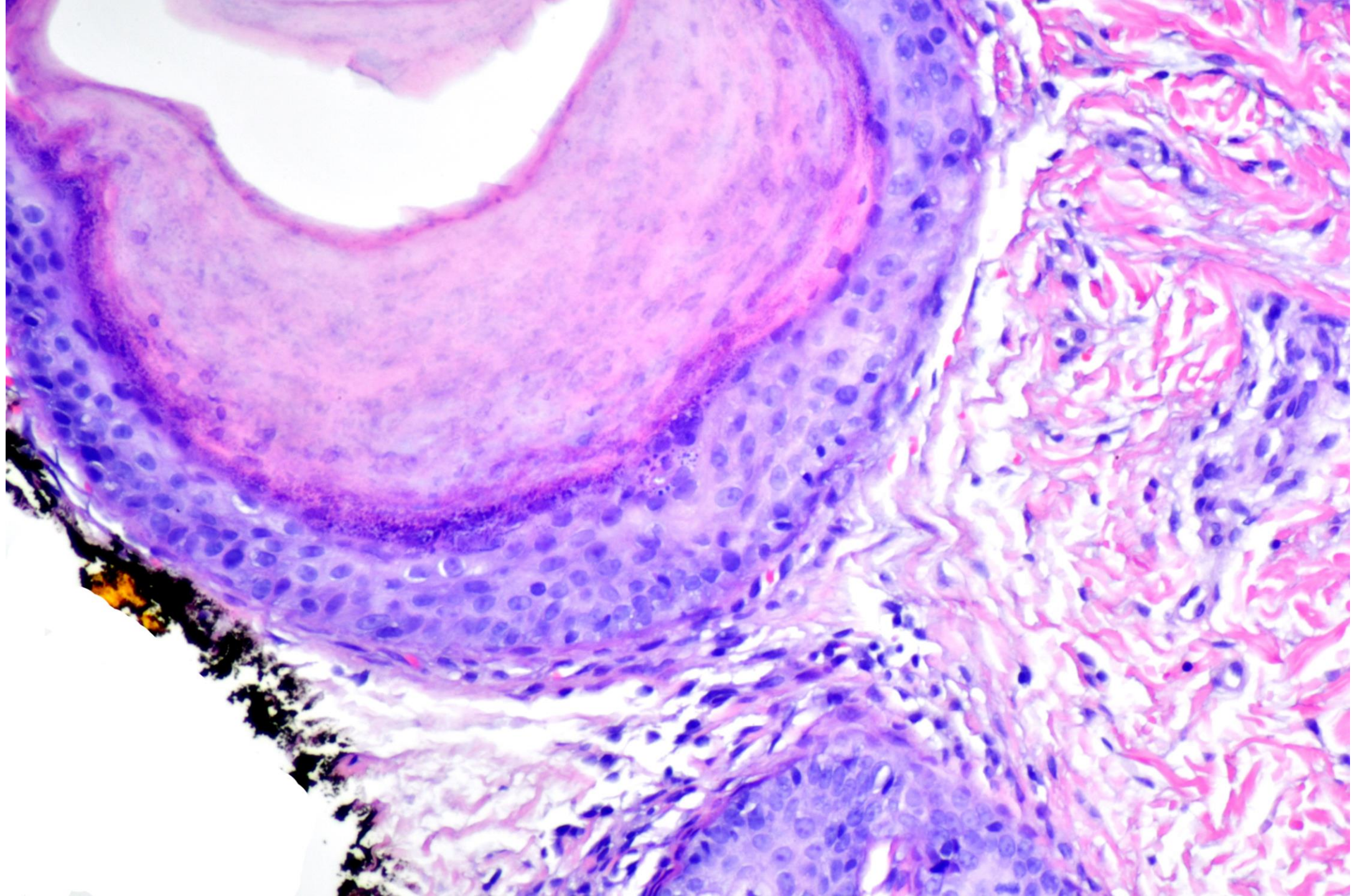




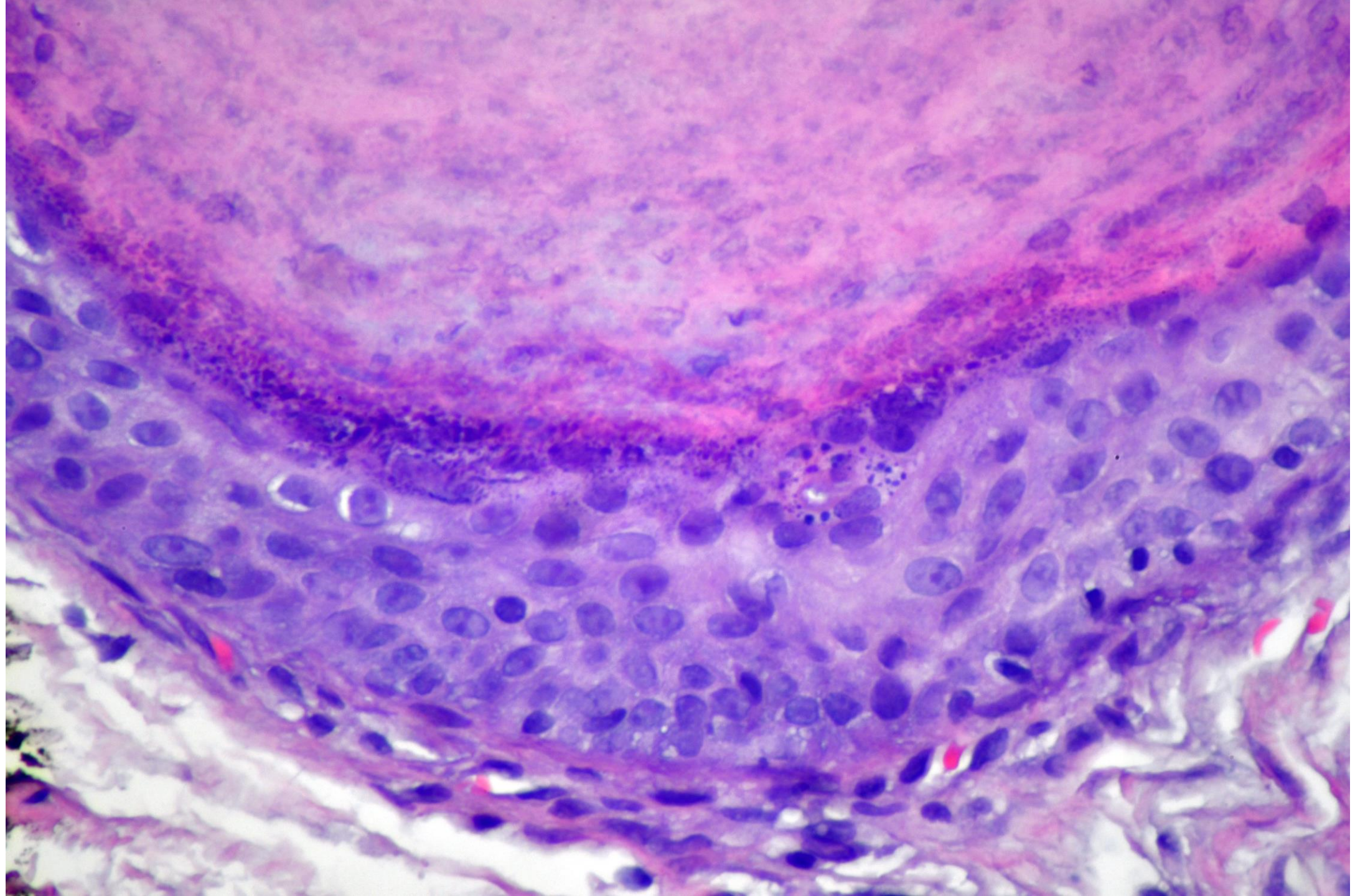




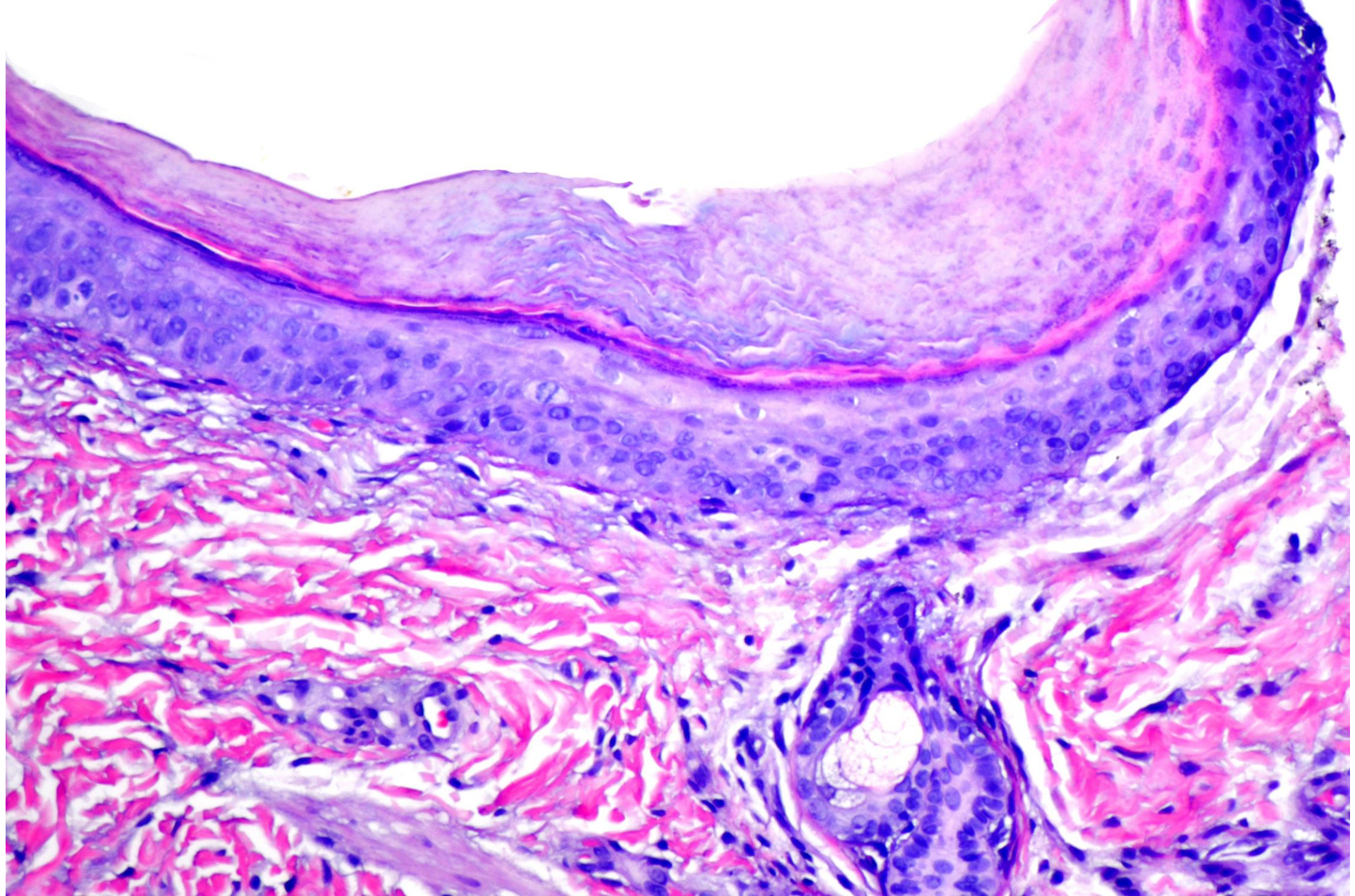




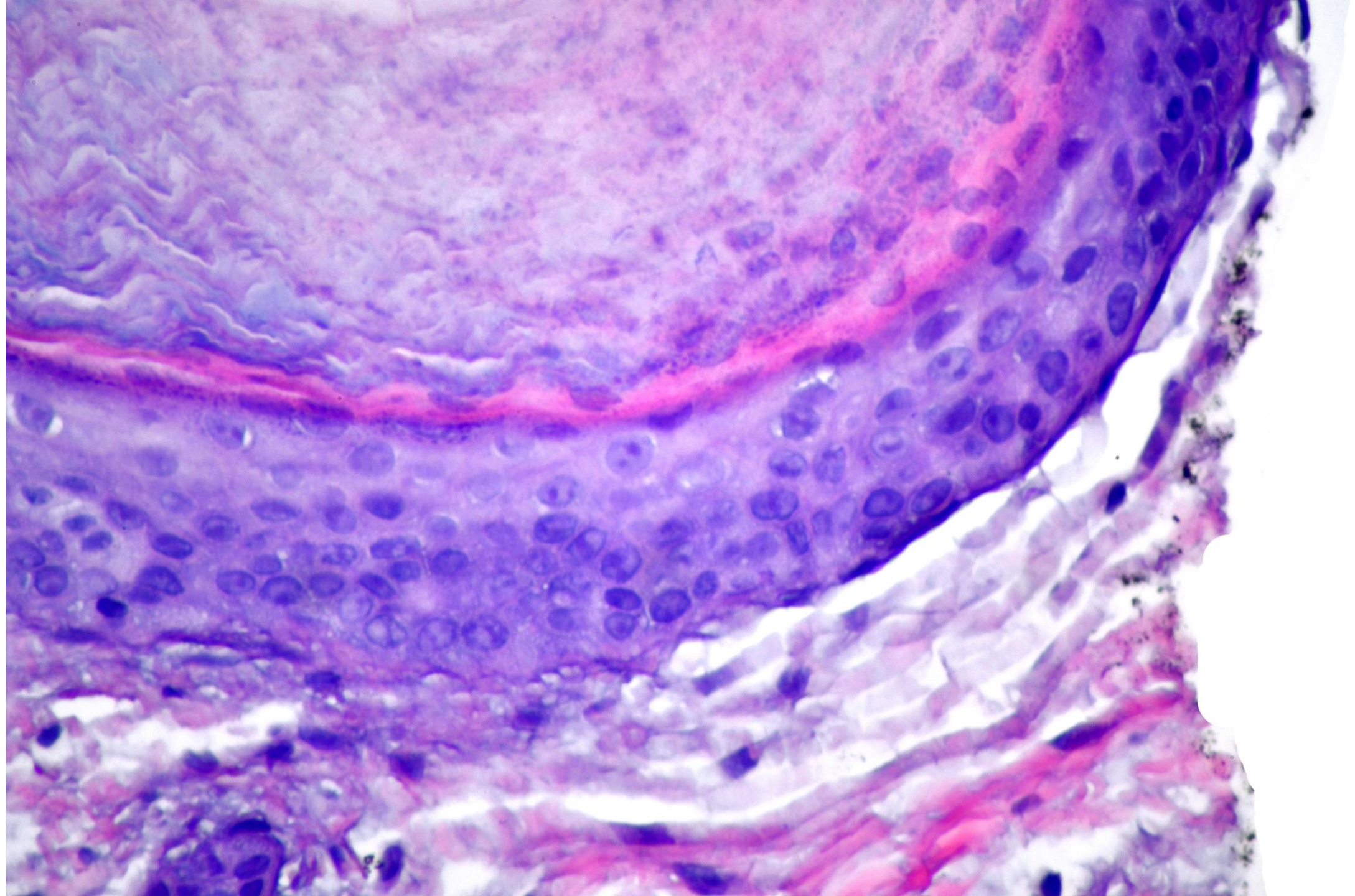












Granular parakeratosis: mimicking an epidermal nevus

# Clinical features

- Often in intertriginous areas
- Reticulated, hyperpigmented, hyperkeratotic
- Can be erosive
- In infants can occur as linear plaques inguinal folds or geometric areas
- Related to diapers/zinc oxide pastes
- Theories in general: antiperspirants/deodorants/moisture/friction

# Pathology

- Hyperkeratosis
- Retention of keratohyaline granules in stratum corneum
- Epidermis acanthotic or thinned
- Involvement of adnexa is common

# Case 3

# Clinical history

- 62-year-old with well controlled HIV
- Presents with pruritic facial rash



# Clinical images

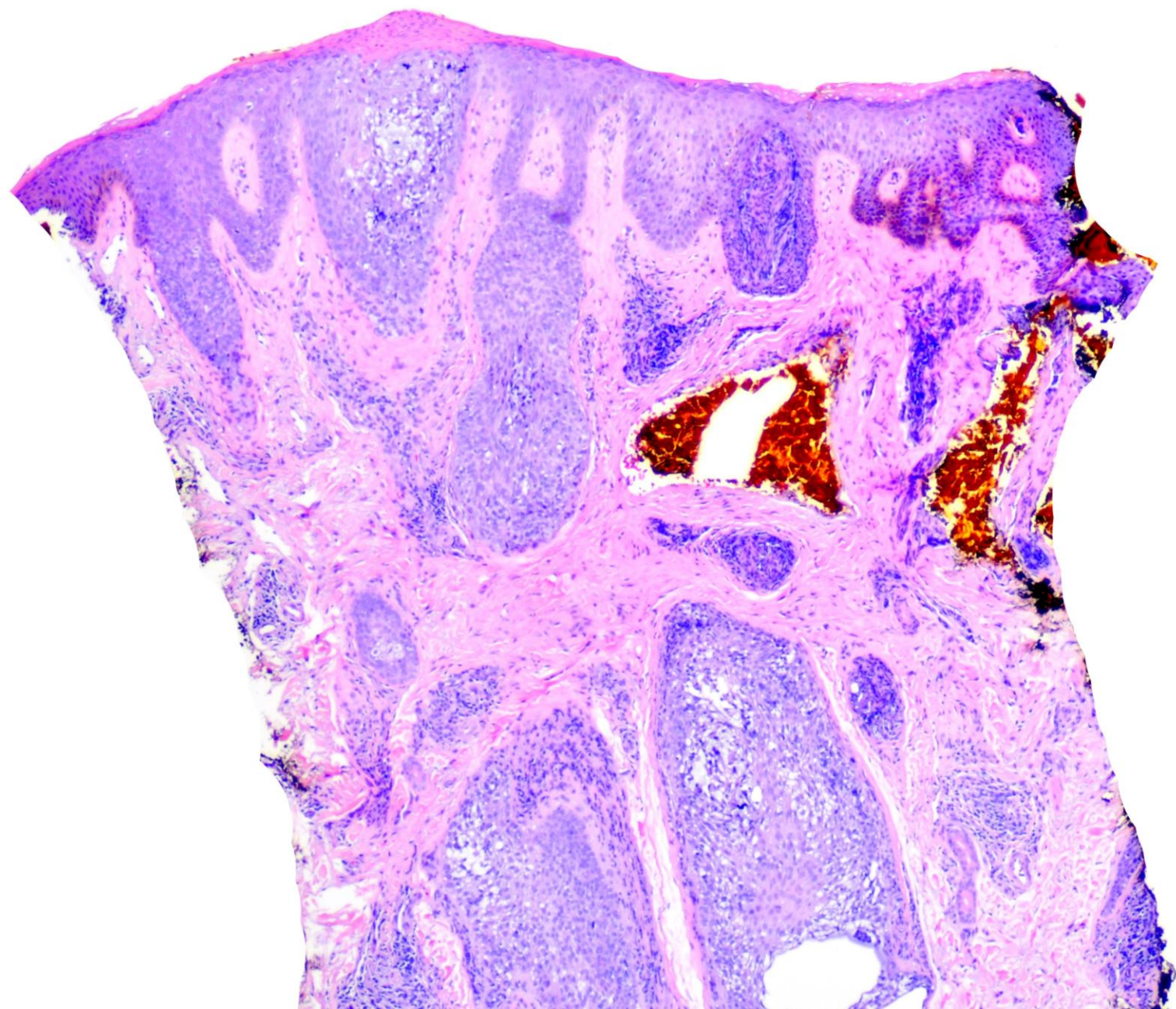


# Clinical images

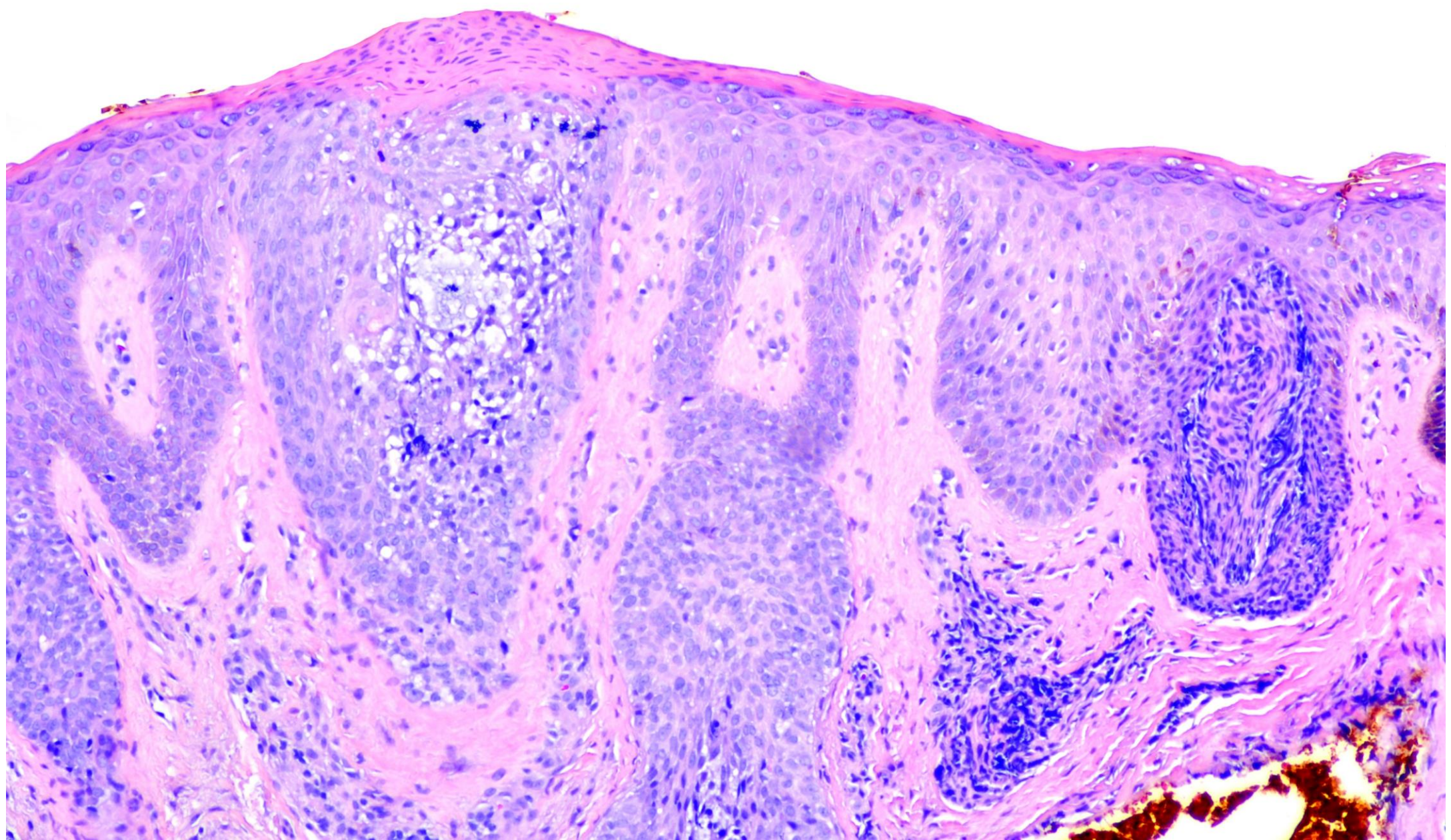




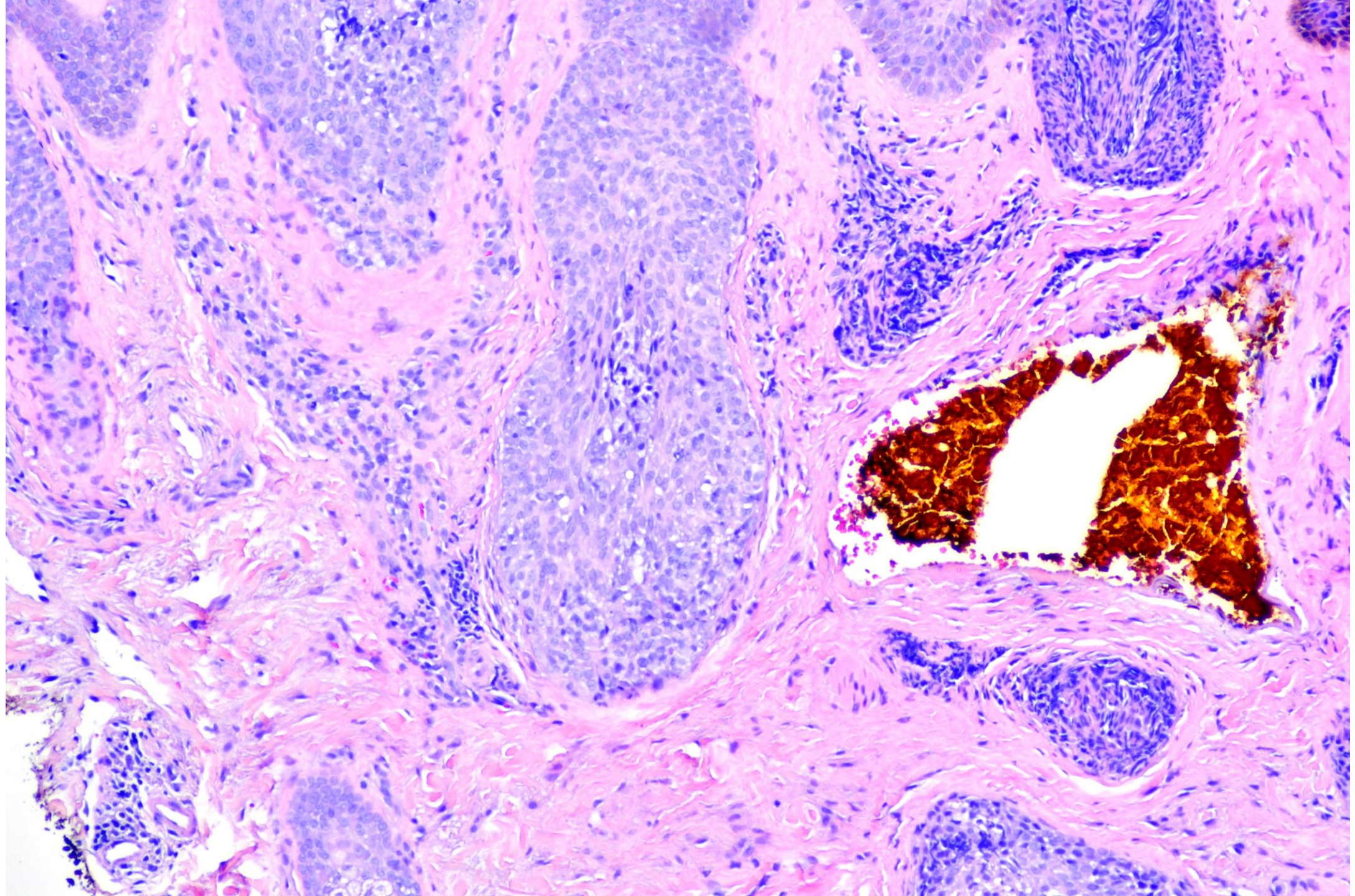




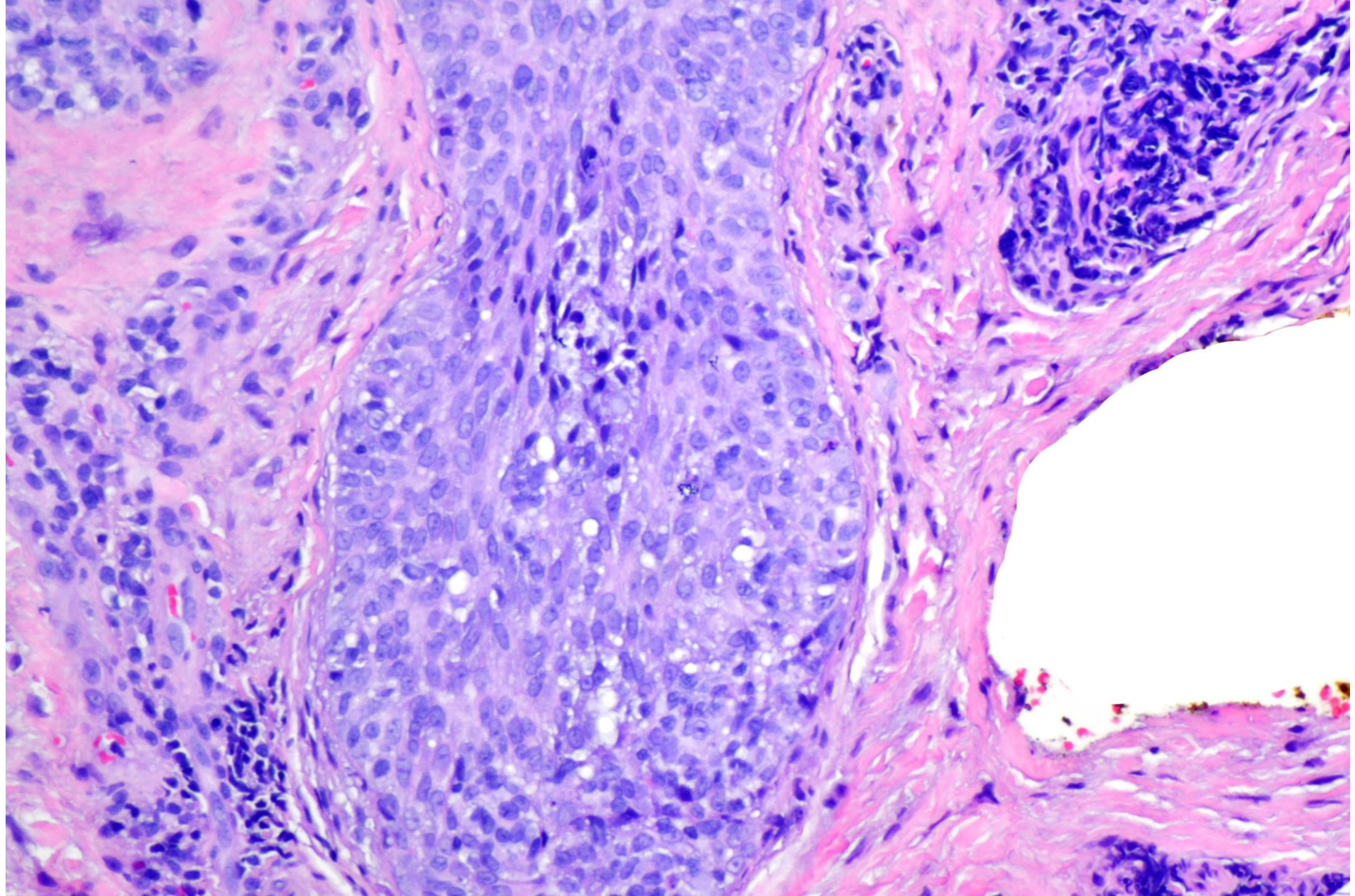




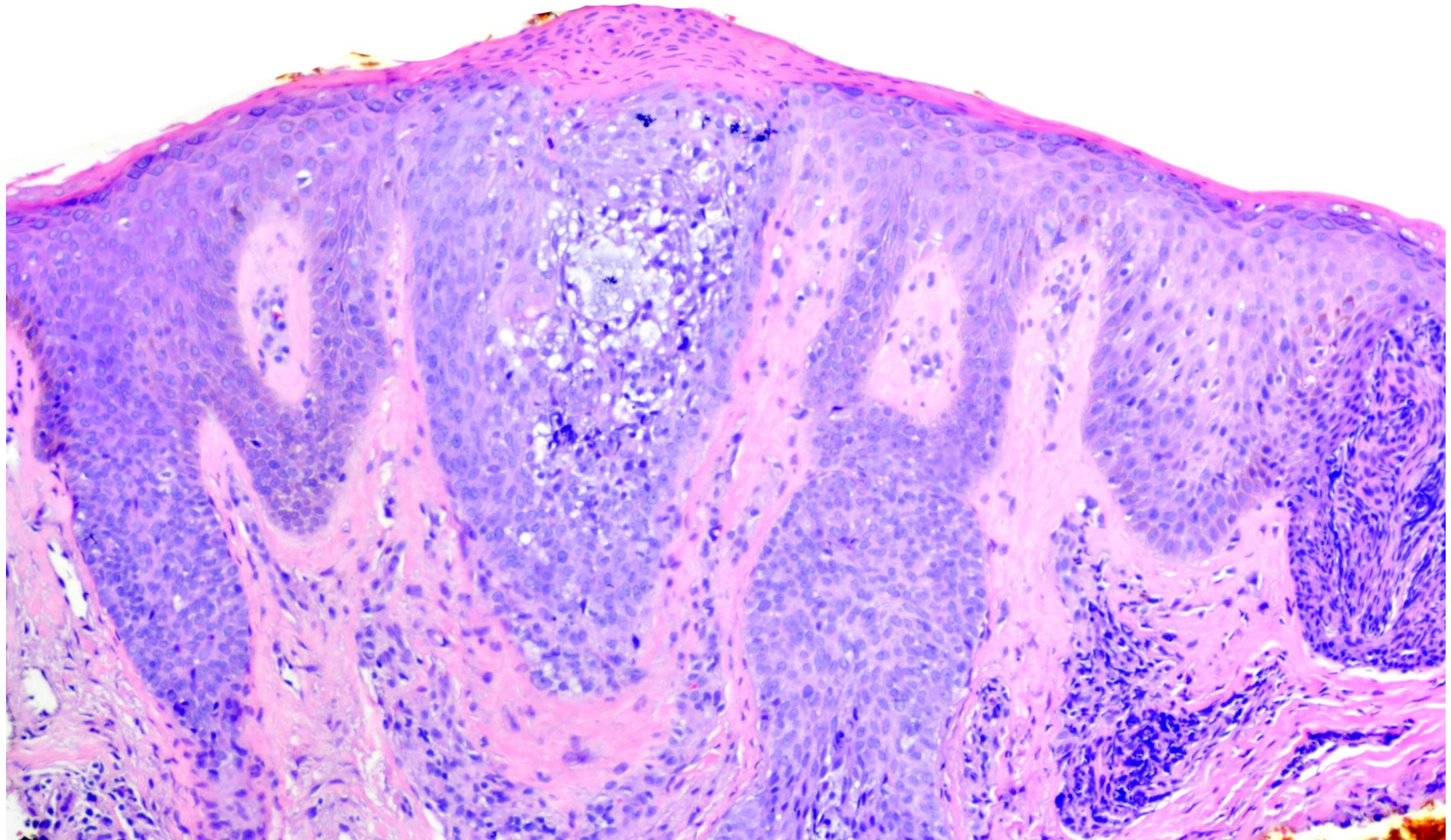




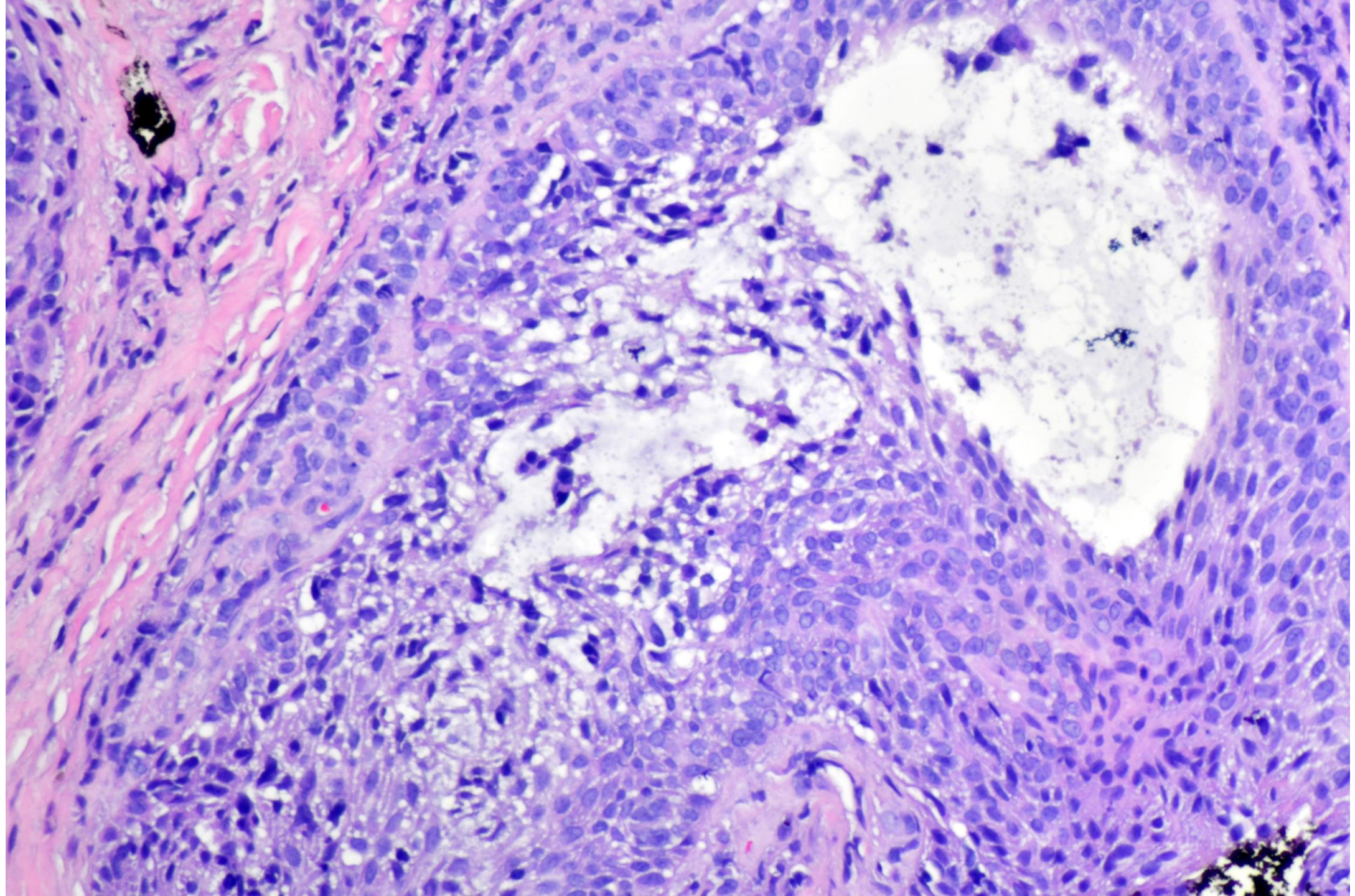




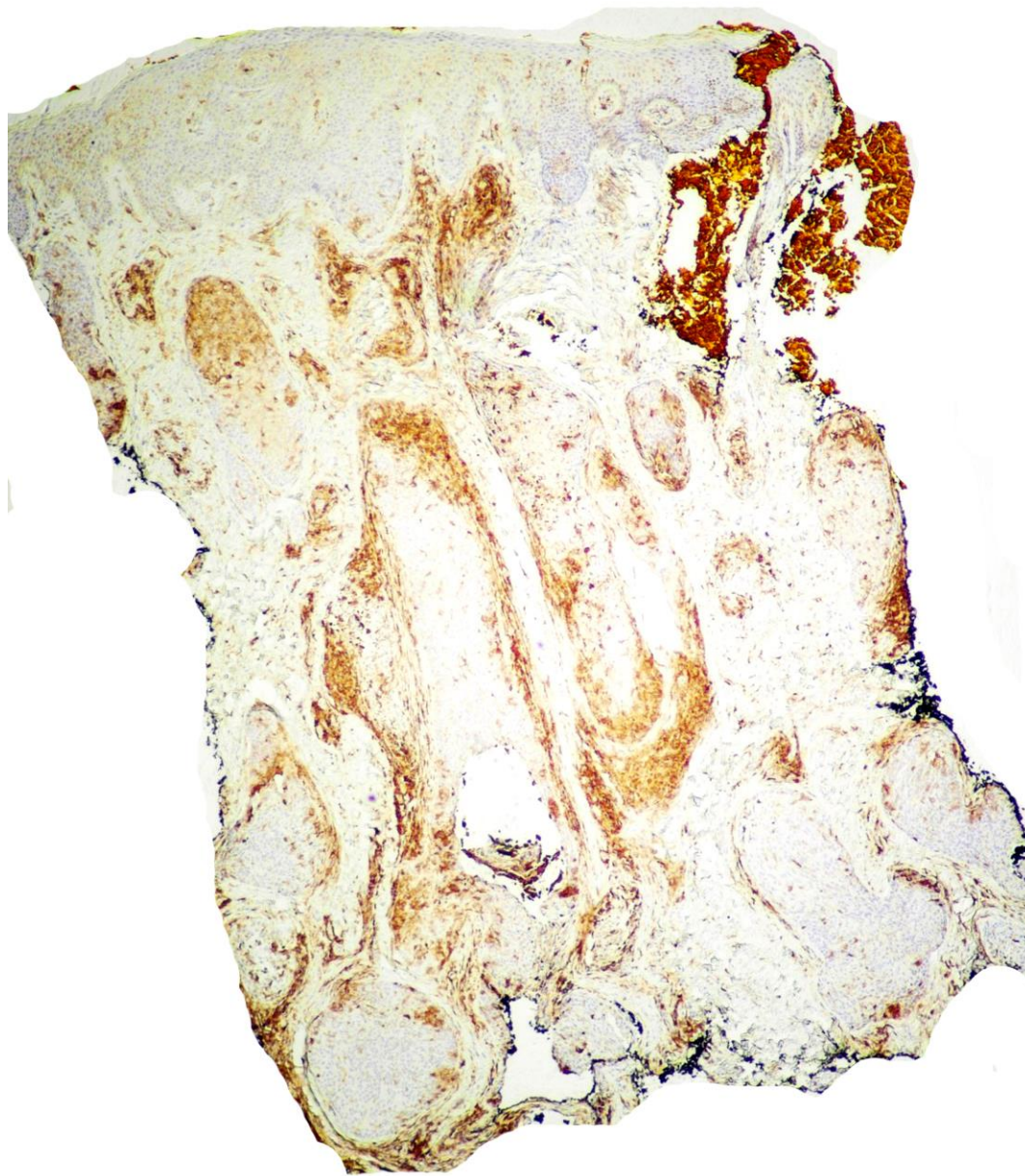






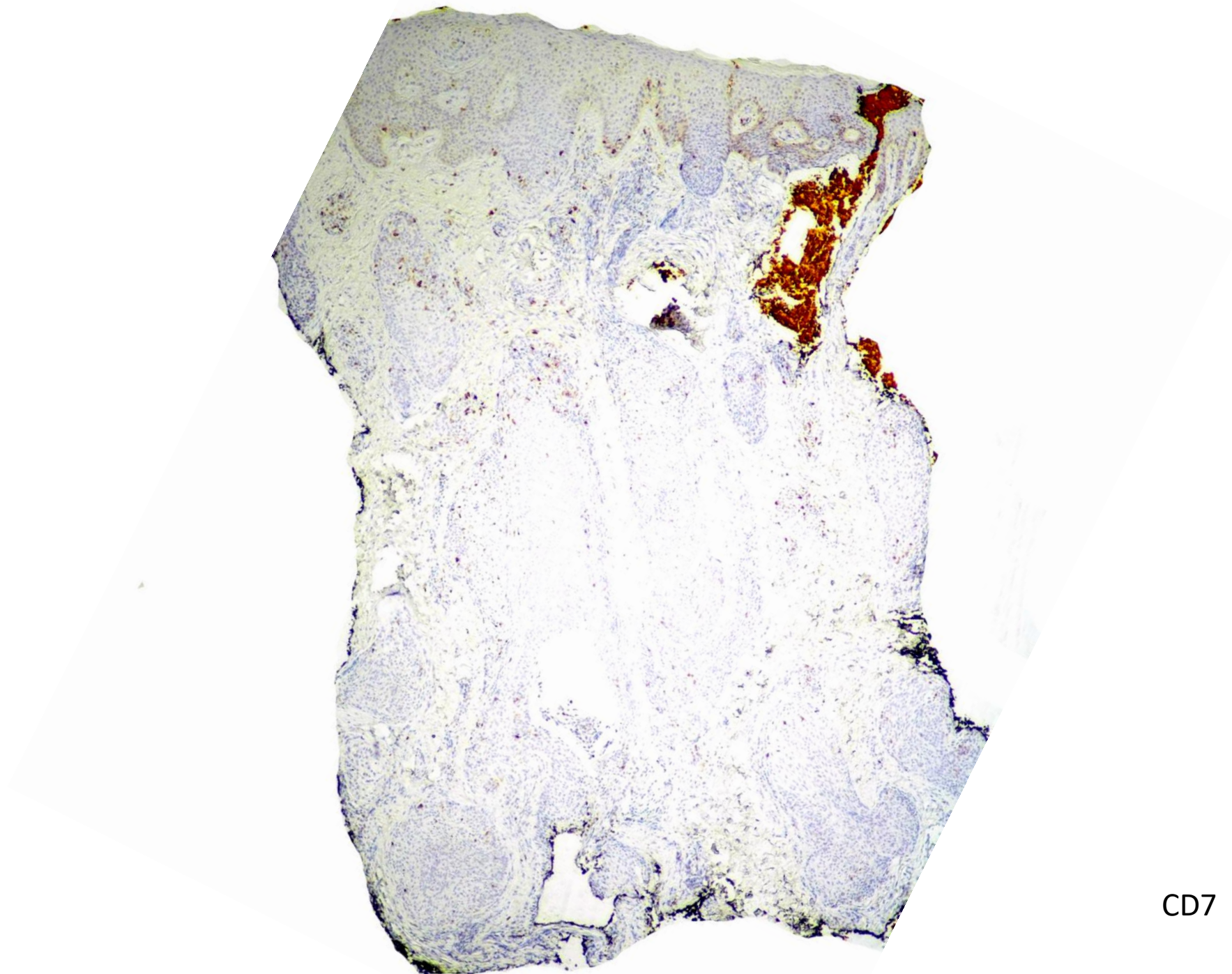




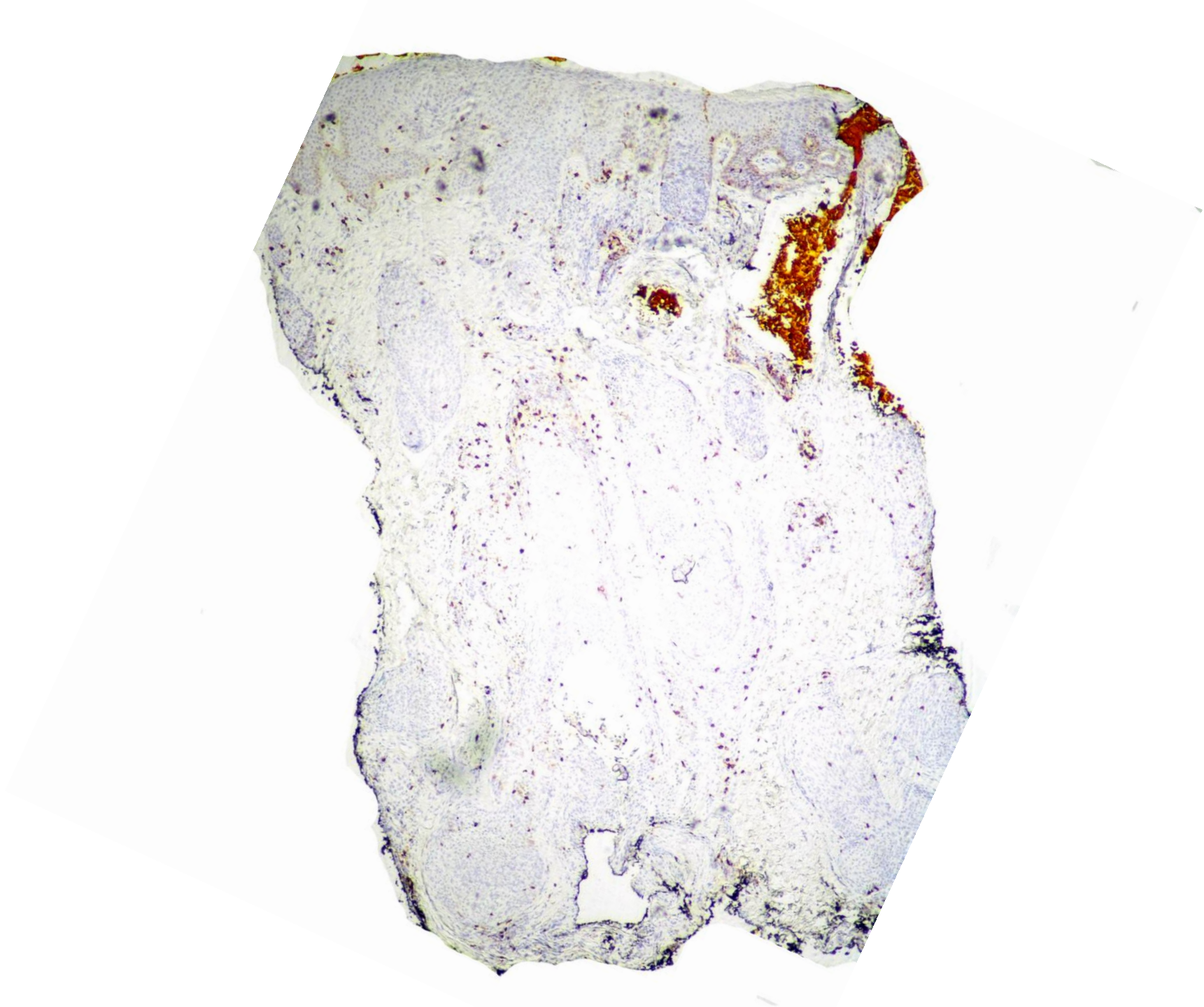


CD4





CD7



CD8



# Molecular Pathology Report

## **SPECIMEN SOURCE:**

A. MOLECULAR TESTIN ON PD-19-26125 A1

## **CLINICAL HISTORY AND CLINICAL DIAGNOSIS:**

Follicular mucinosis with atypical lymphoid infiltrate, left glabella

## **RESULTS:**

### ***TRG* GENE REARRANGEMENT:**

**Positive for clonal *TRG* Gene Rearrangement (V and J regions).**

**INTERPRETATION:** A discrete band was identified, consistent with the presence of a monoclonal or oligoclonal T cell population.

### ***TRB* GENE REARRANGEMENT:**

**Positive for clonal *TRB* Gene Rearrangement (V and J regions).**

**INTERPRETATION:** A discrete band was identified, consistent with the presence of a monoclonal or oligoclonal T cell population.

# **Surgical Pathology Report**

## **SPECIMEN SOURCE:**

A. LEFT INGUINAL LYMPH NODE BIOPSY

## **CLINICAL HISTORY:**

62 y/o HIV+ male with history of mycosis fungoides (positive TCR-gene rearrangement studies), elevated LDH (575) and elevated IgG (2,030) + IgA (479), now with inguinal adenopathy (SUV 7.9); rule out lymphoma.

## **DIAGNOSIS:**

A. LEFT INGUINAL LYMPH NODE, BIOPSY:

- T CELL LYMPHOMA, CONSISTENT WITH MYCOSIS FUNGOIDES.
- GRADE 3 LYMPH NODE INVOLVEMENT (DUTCH SYSTEM)



# Follicular mucinosis: clinical

- A histologic reaction pattern
- Can be benign or associated with T-cell lymphoma, particularly mycosis fungoides and Sezary syndrome
- In children and adolescent follicular papules, nodules often on facial skin and scalp associated with loss
- Older age group with more generalized

# Pathology

- Mucin – hyaluronic acid causes cystic spaces in follicular epithelium and sebaceous glands
- There is a surrounding perifollicular infiltrate of lymphocytes, histiocytes and eosinophils
- Distinction between the different types is difficult
- Clinical features, epidermotropism and atypical lymphocytes do help in distinguishing the benign and malignant types
- TCR and marker studies may not always be helpful



Differential diagnosis





