

Diagnoses that matter, and not so much

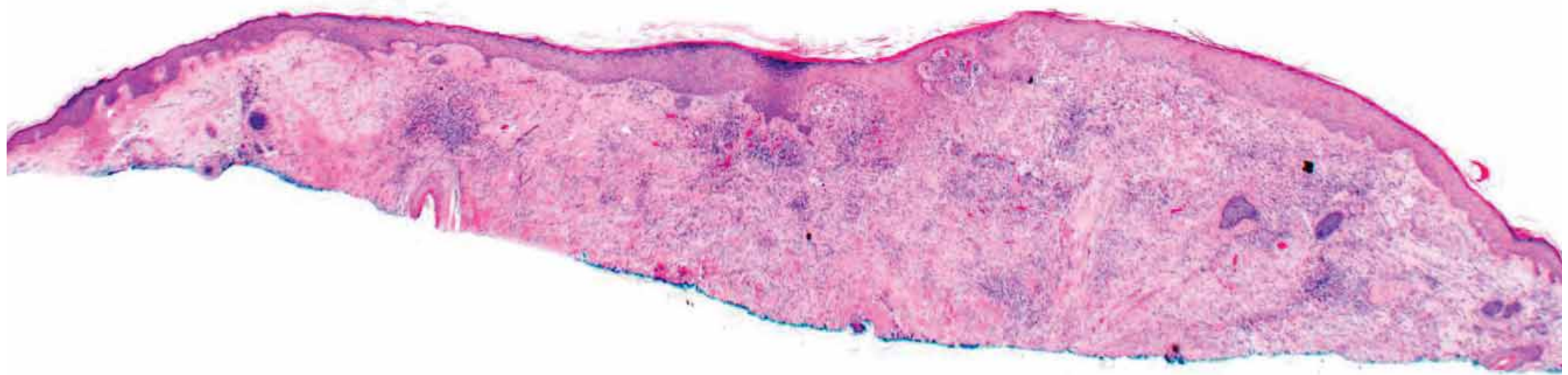
Philip LeBoit, M.D.

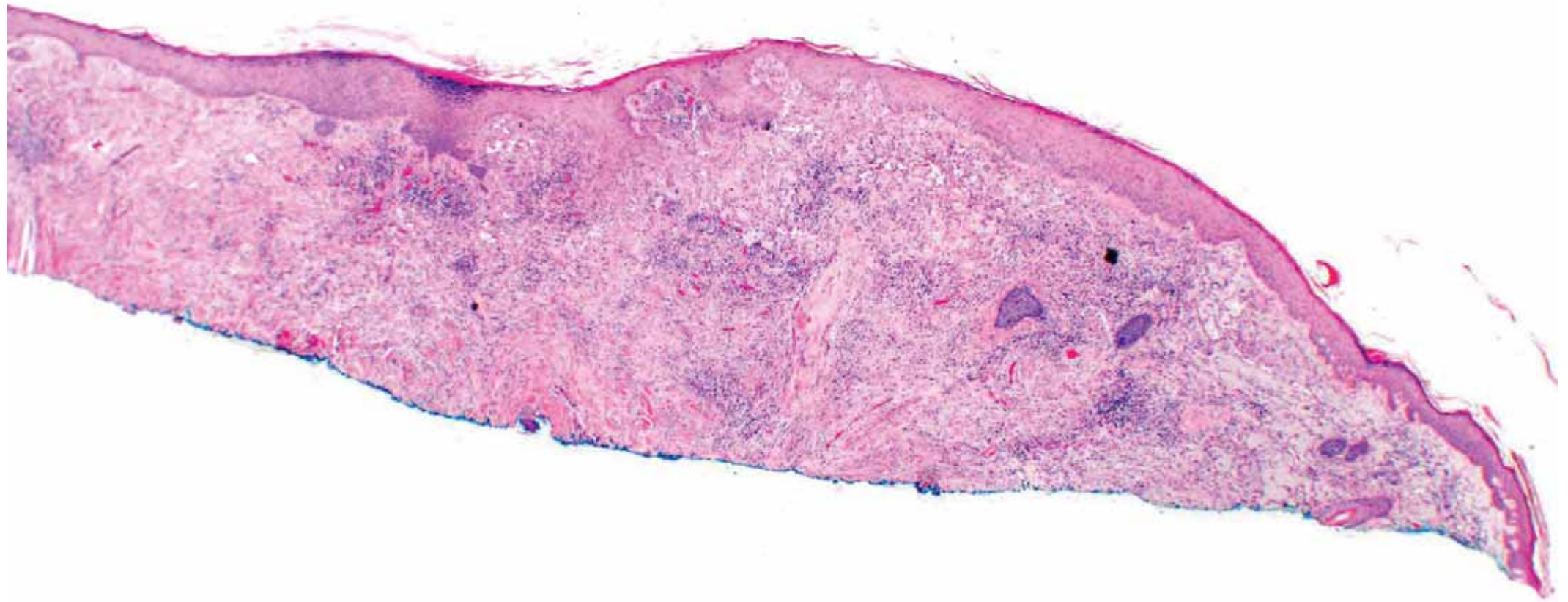
Depts. of Pathology and Dermatology

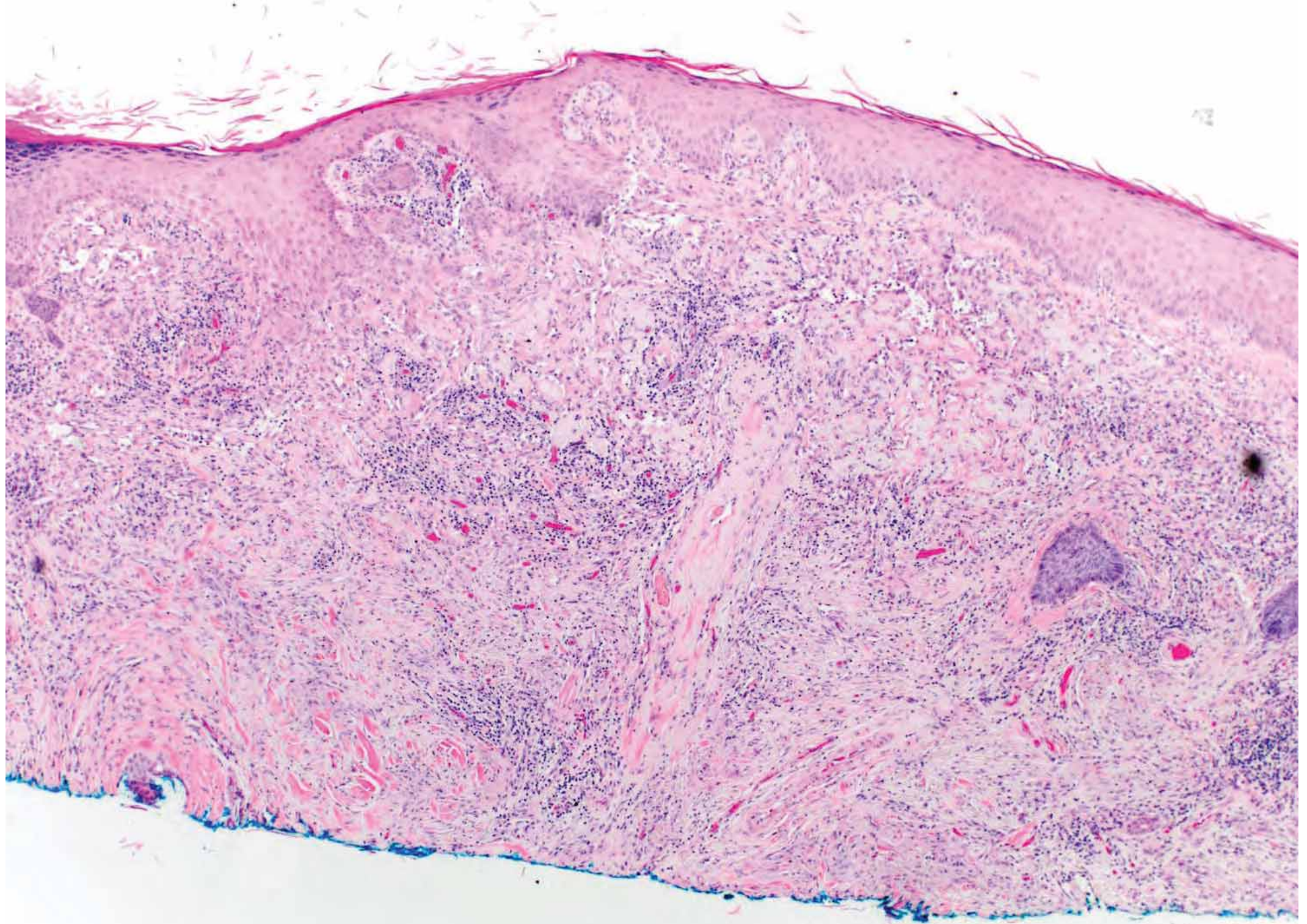
Univ. of California, San Francisco

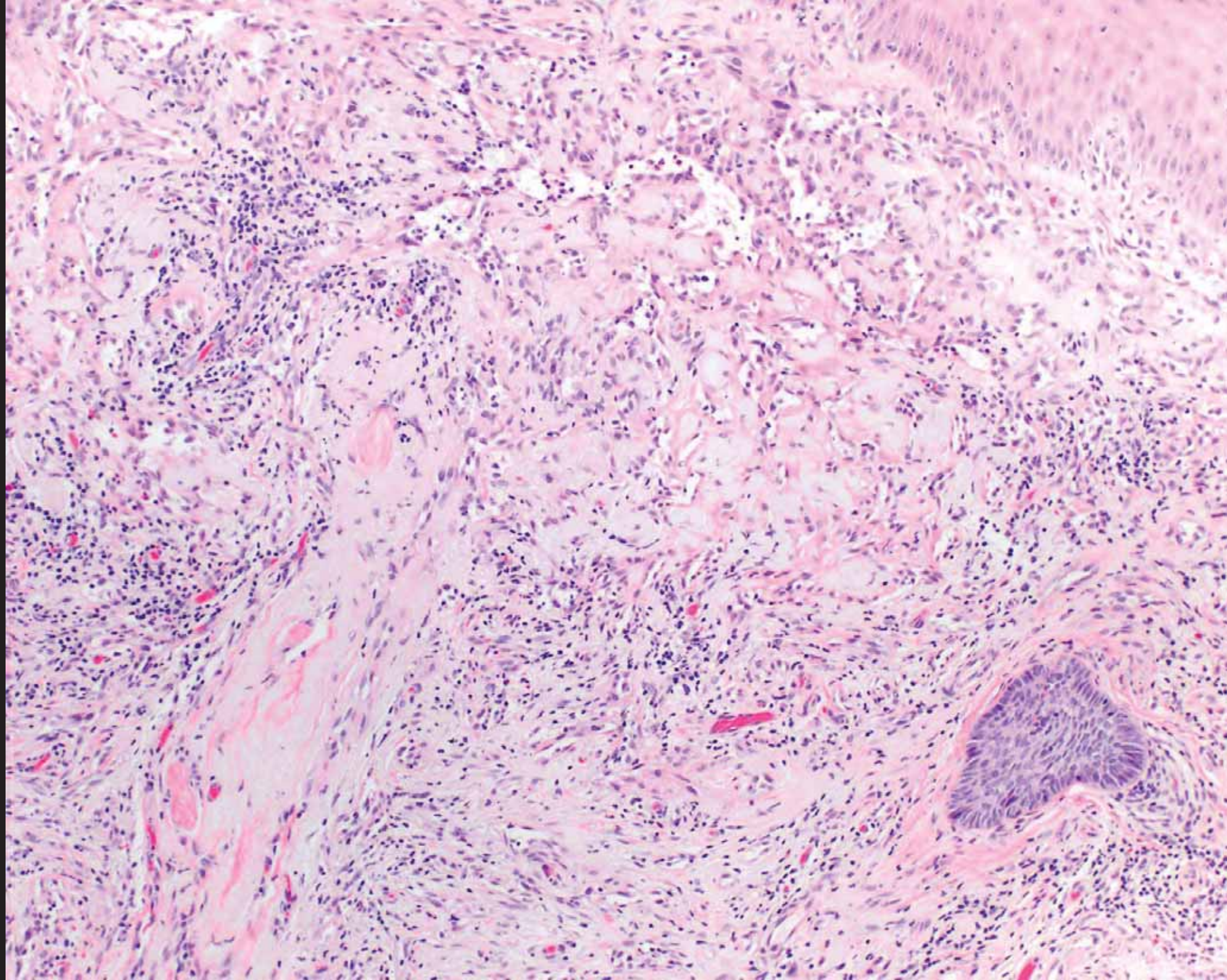
Case 1

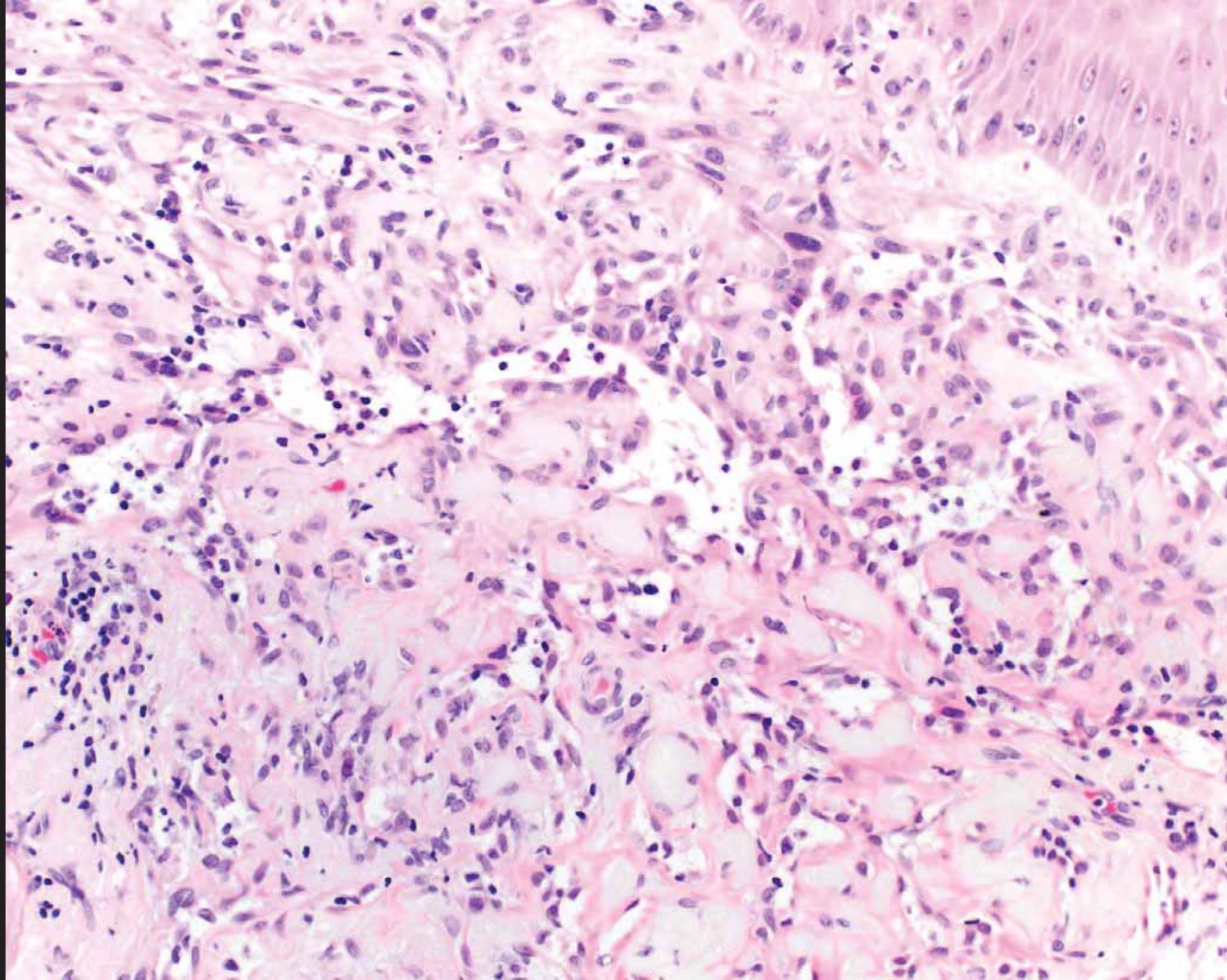
86 year old man, biopsy

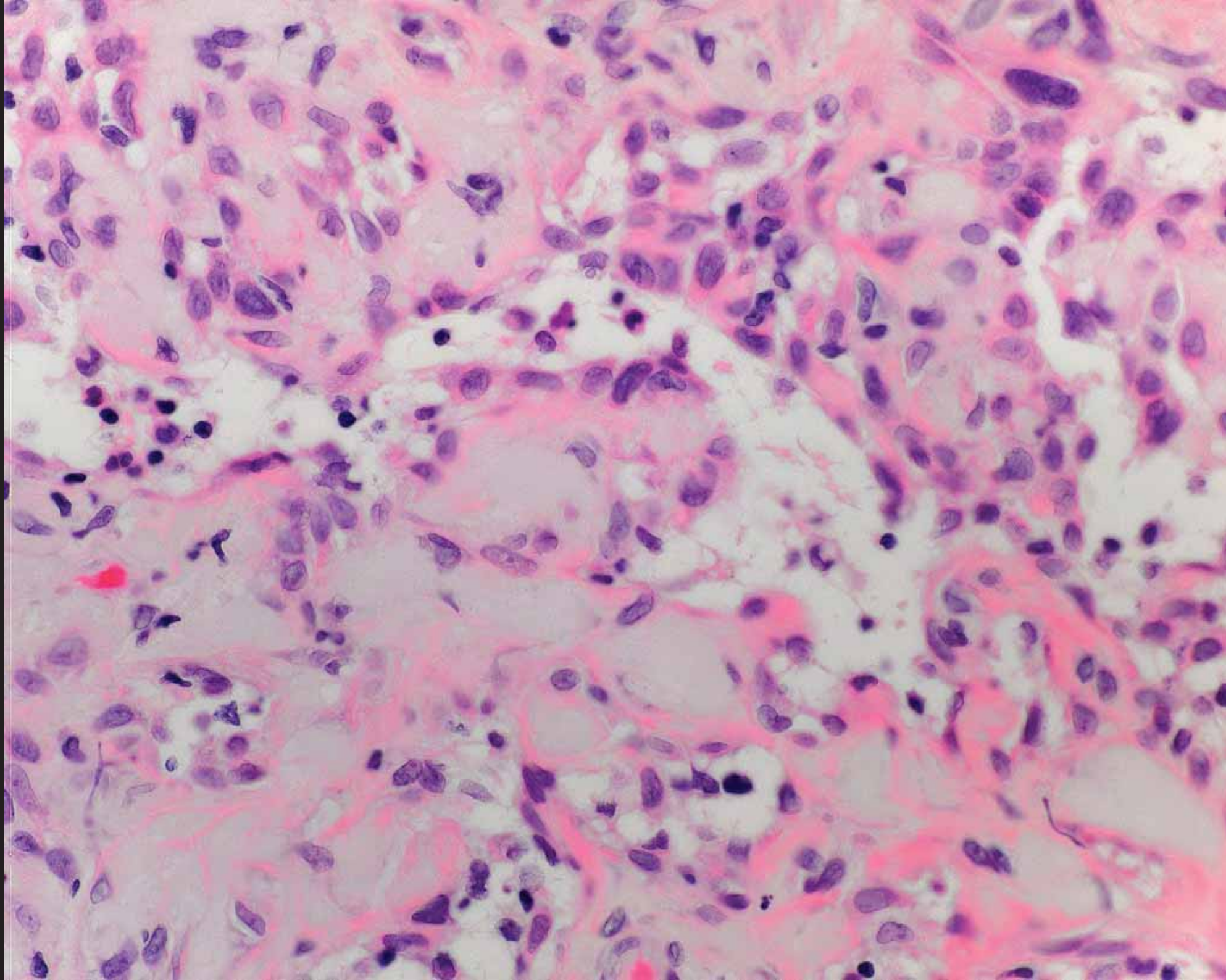


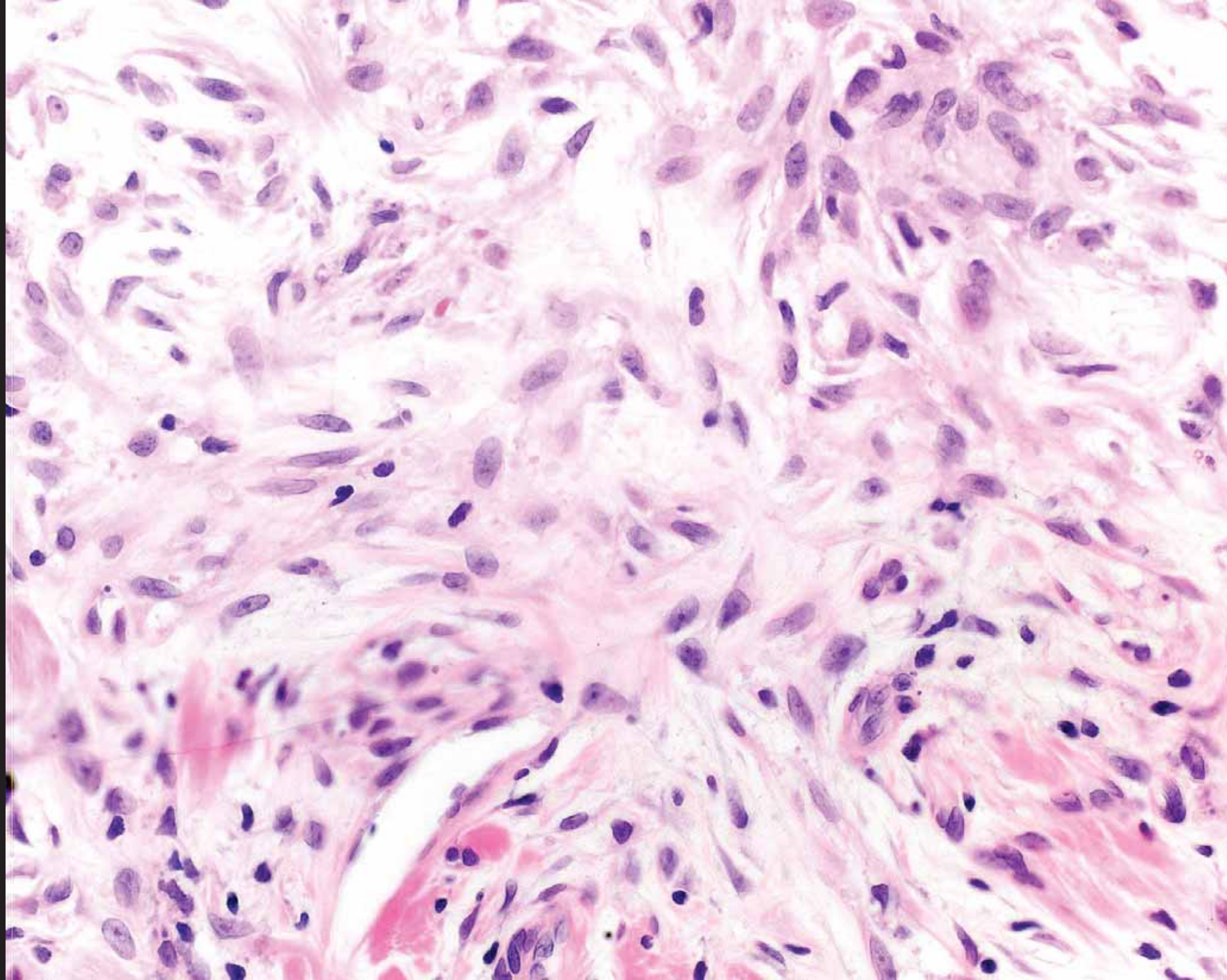




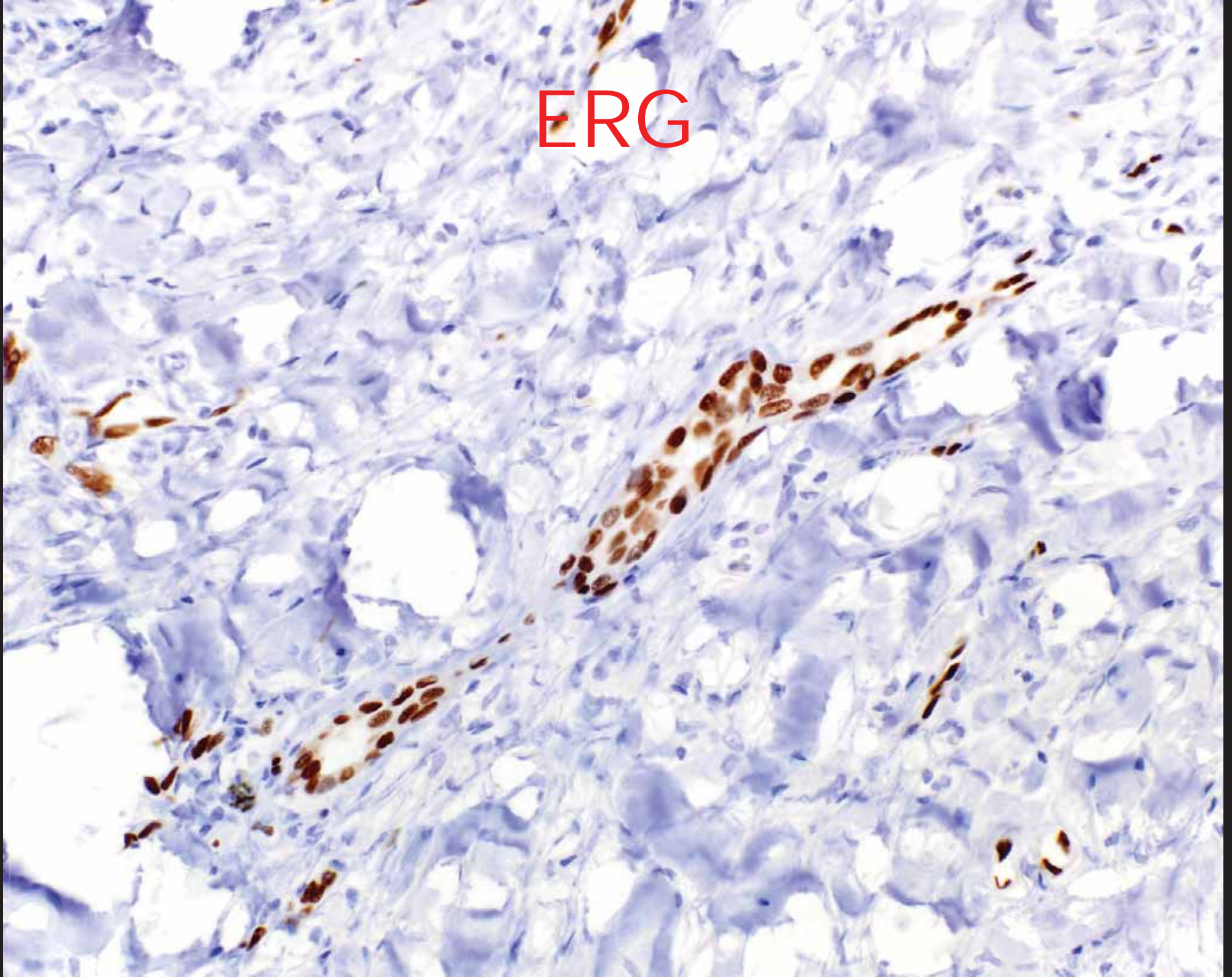




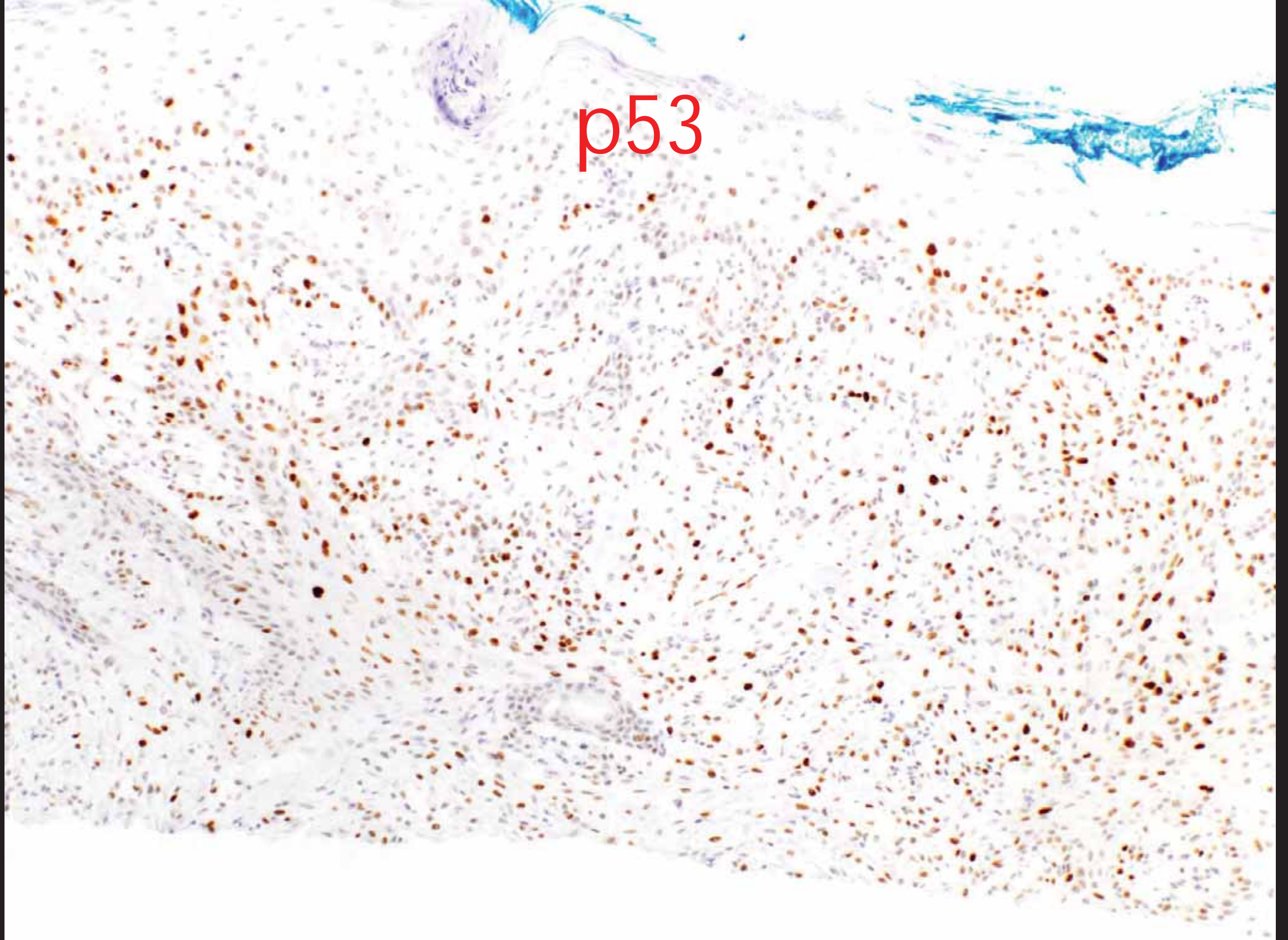




ERG



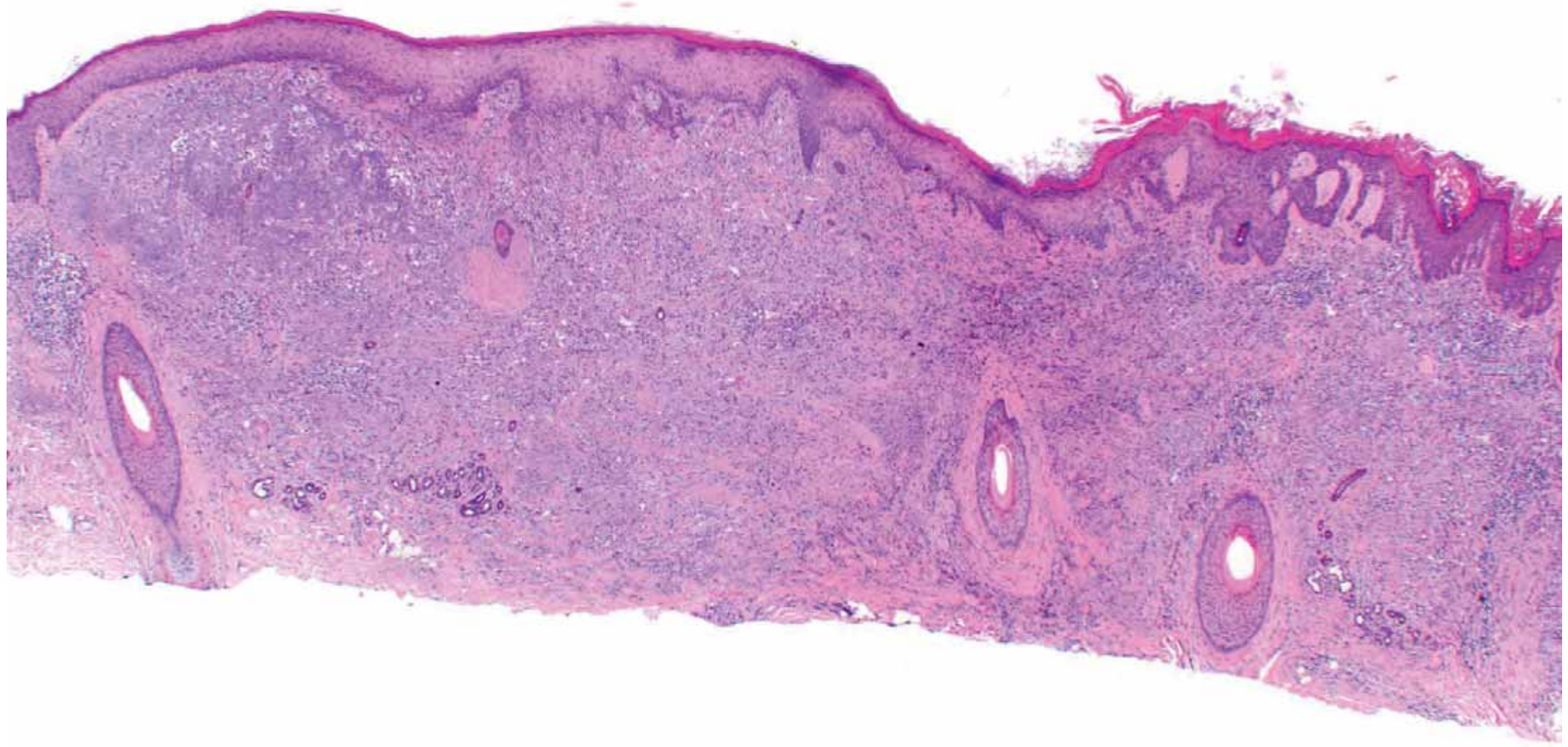
p53

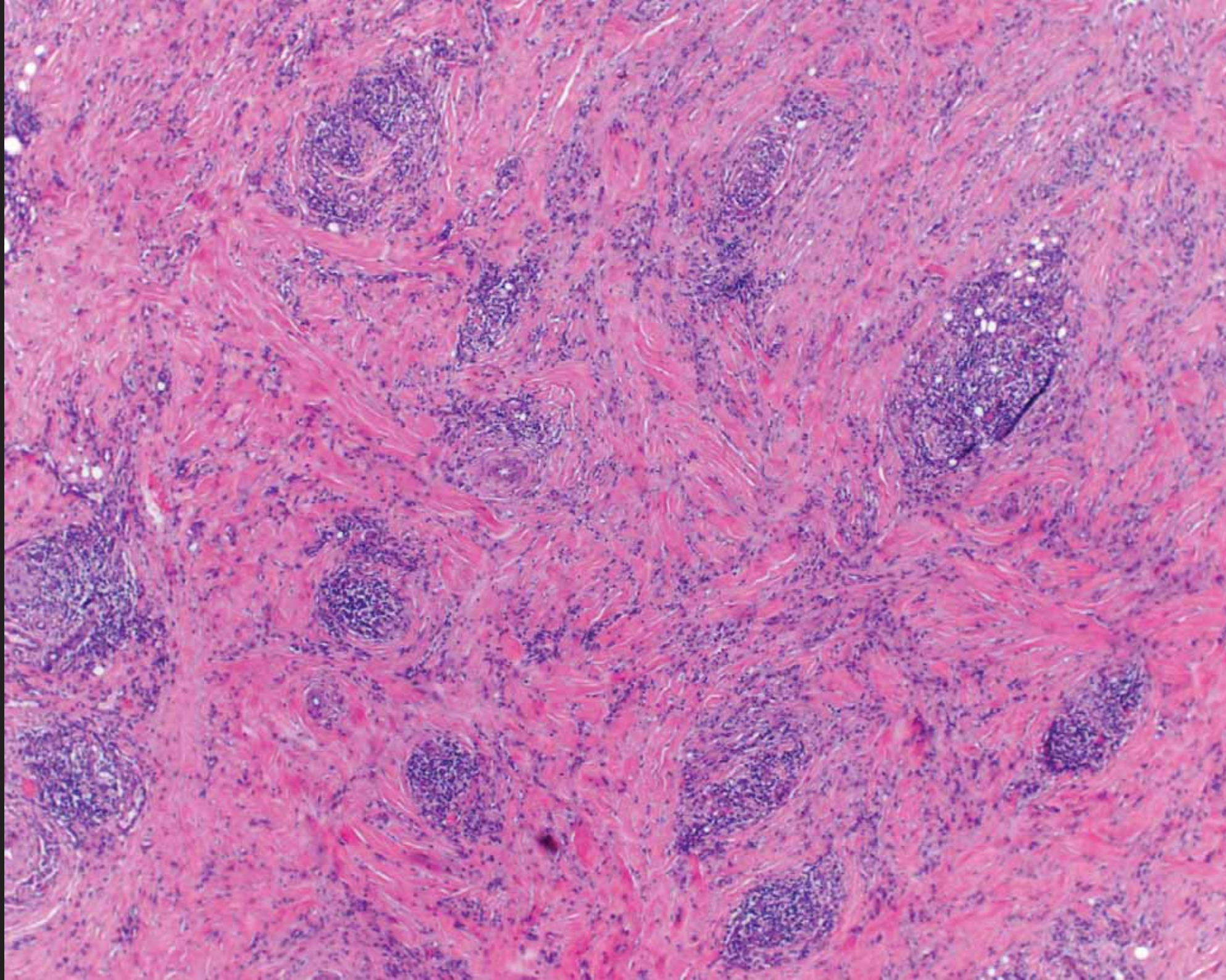


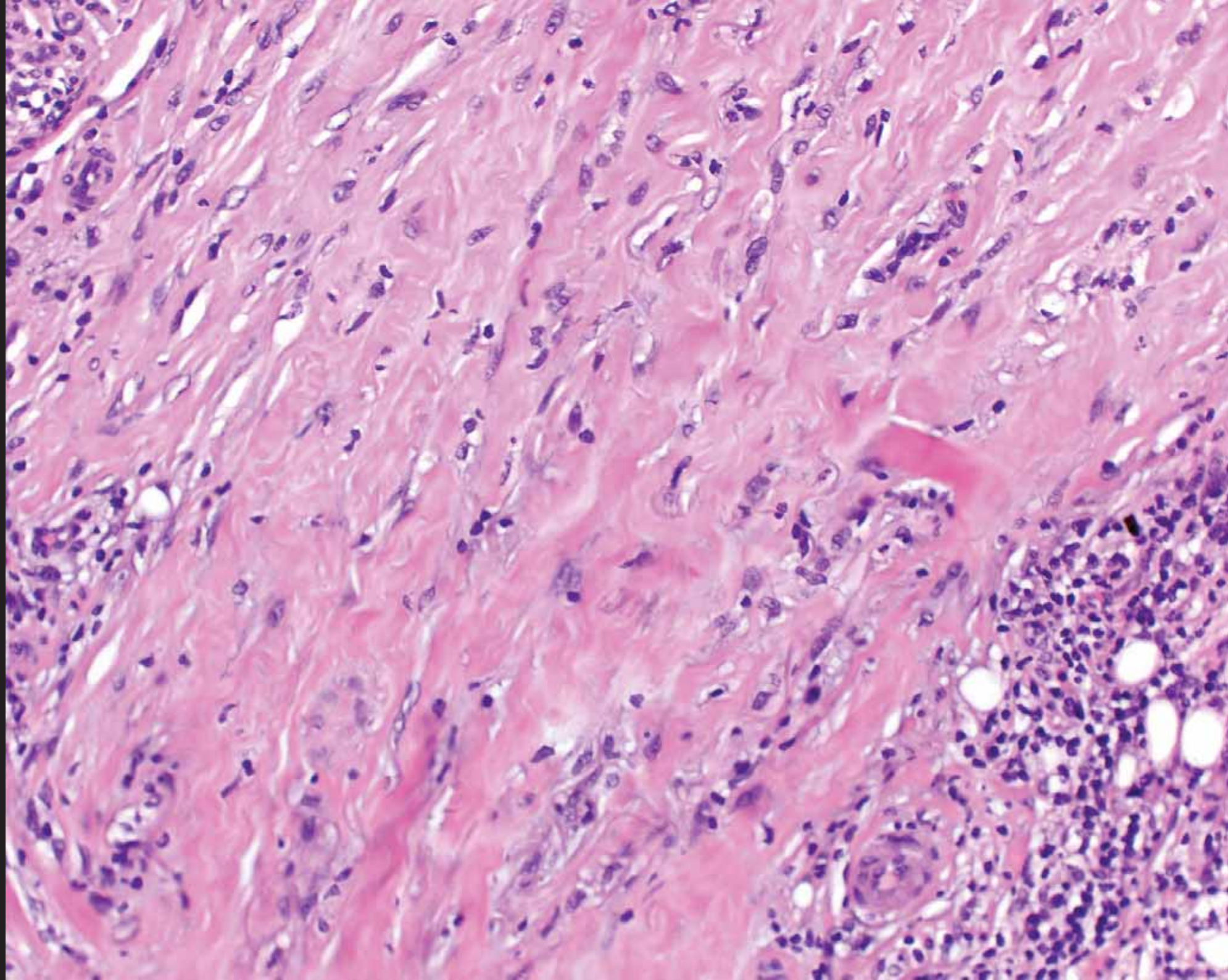
Diagnosis- angiosarcoma

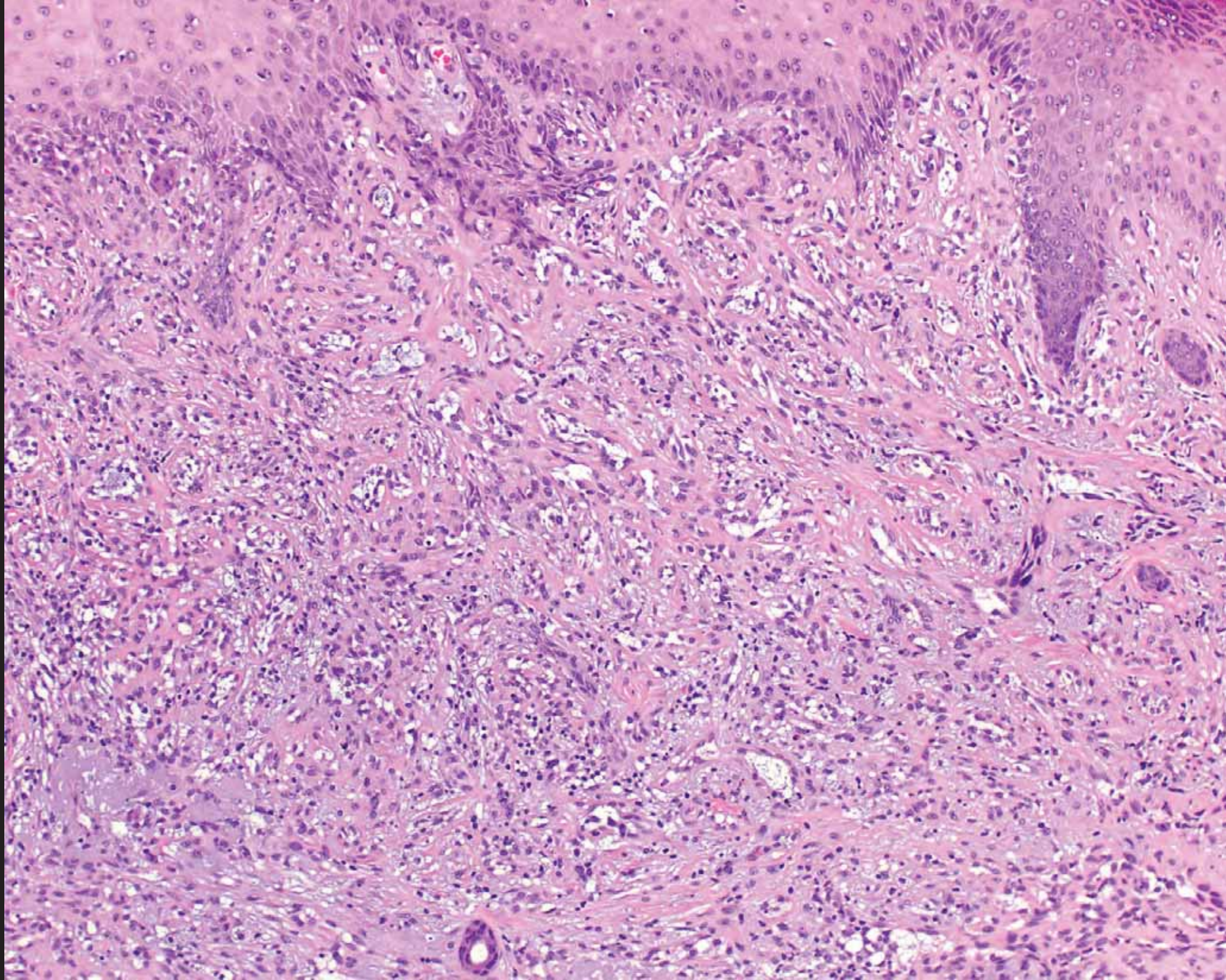
- Infiltrative growth pattern
- Jagged clefts lined by protuberant cells
- Positive for ERG, the most specific endothelial cell marker
- Positive for p53; TP53 mutations are the most common in angiosarcomas of the head in elderly patients

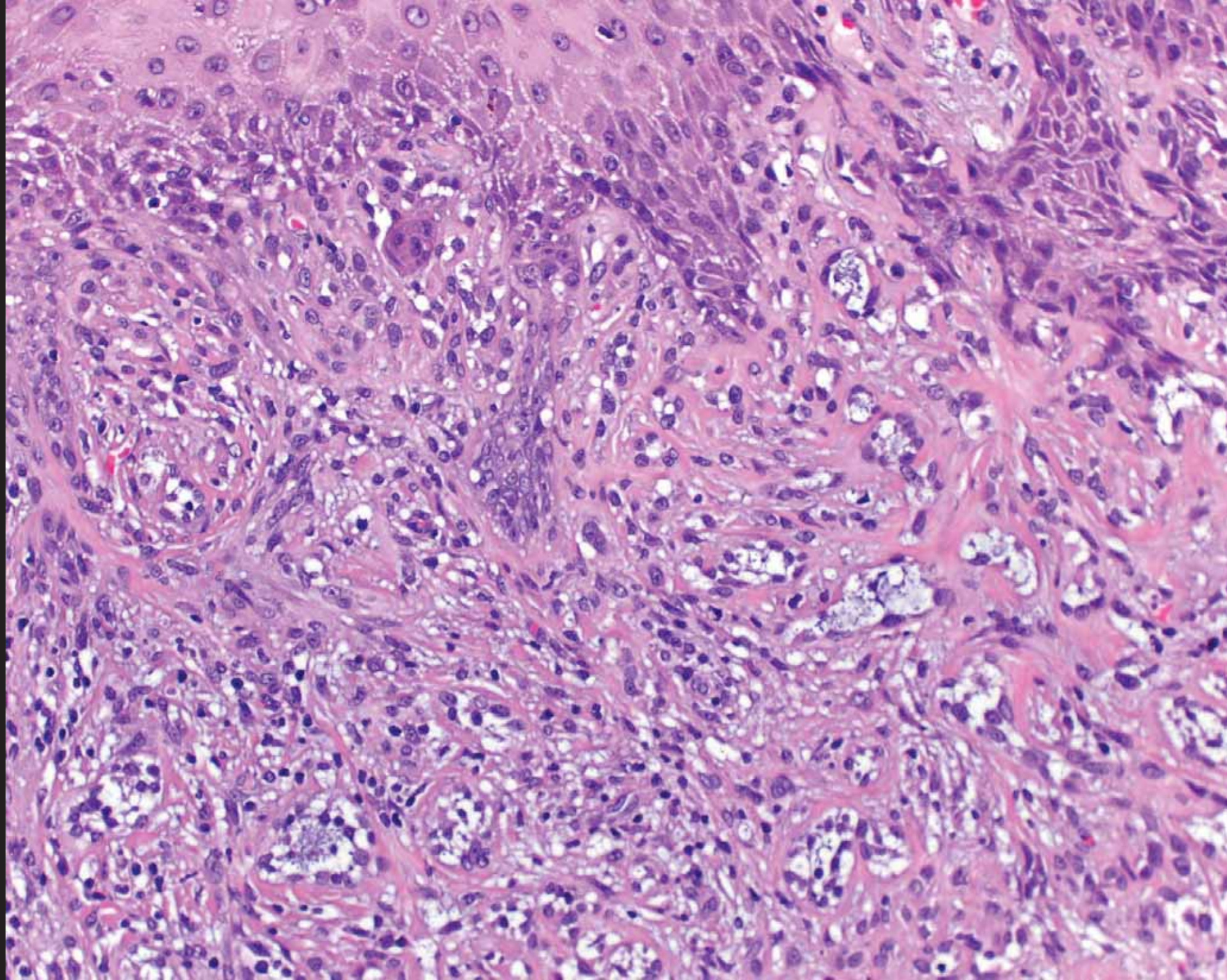
Debulking performed prior to
micrographic surgery

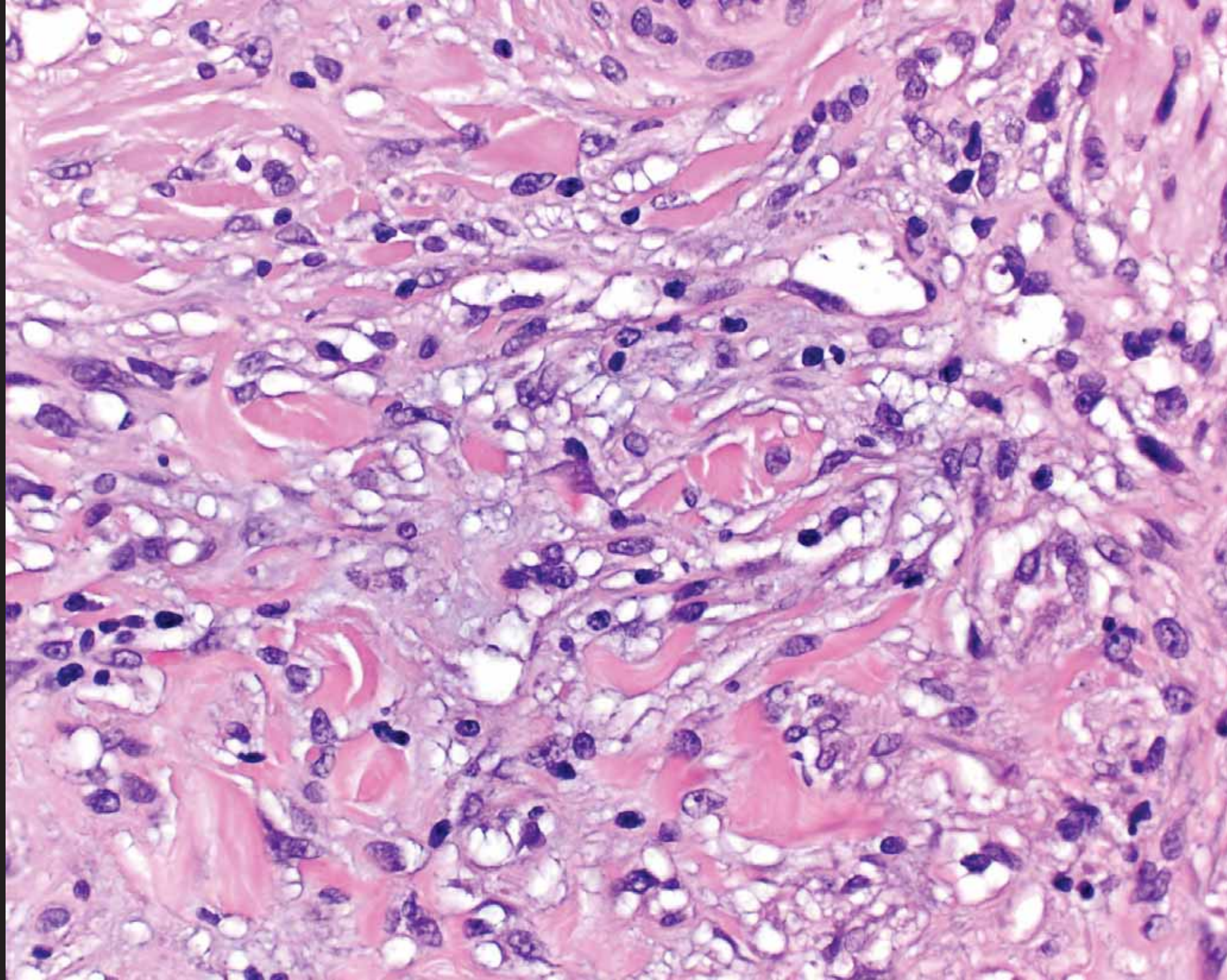


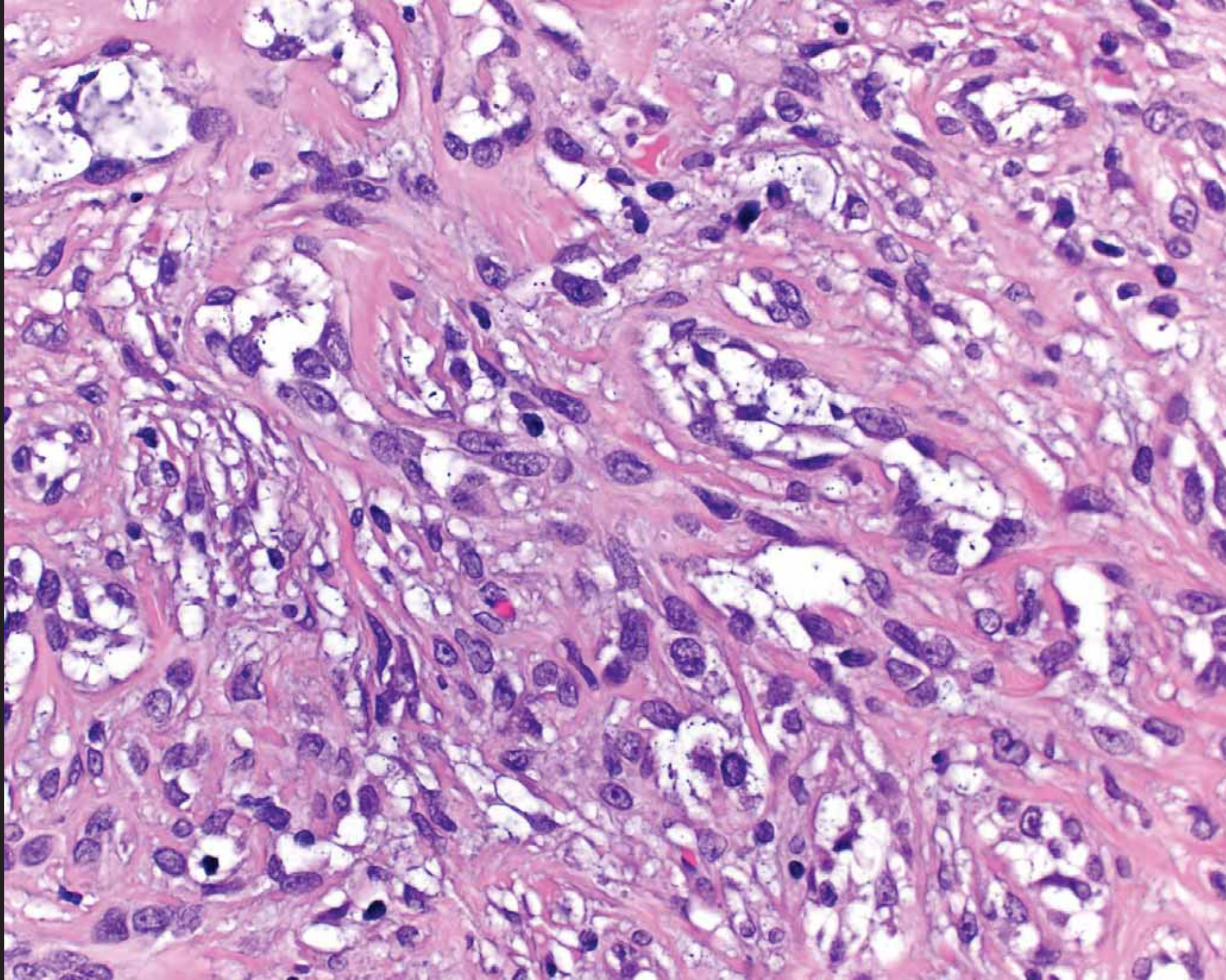








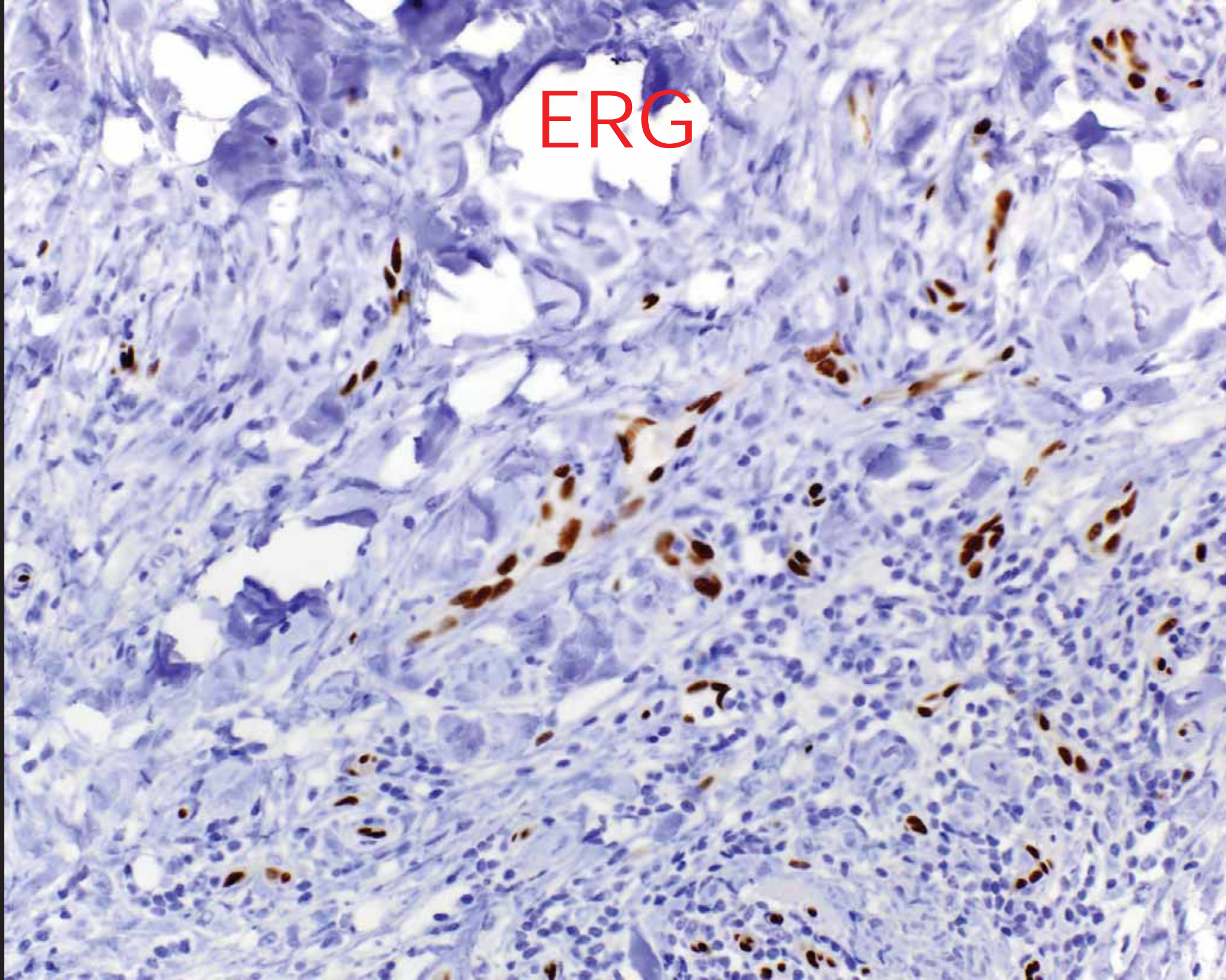




We know that mucin is overproduced due to

- hyaluronic acid synthase gene (HAS2) mutation in Shar Pei dogs
- Can the cells of angiosarcoma mutate to produce mucin?
- Is this Shar Pei angiosarcoma?
- Let's repeat the ERG stain!

ERG



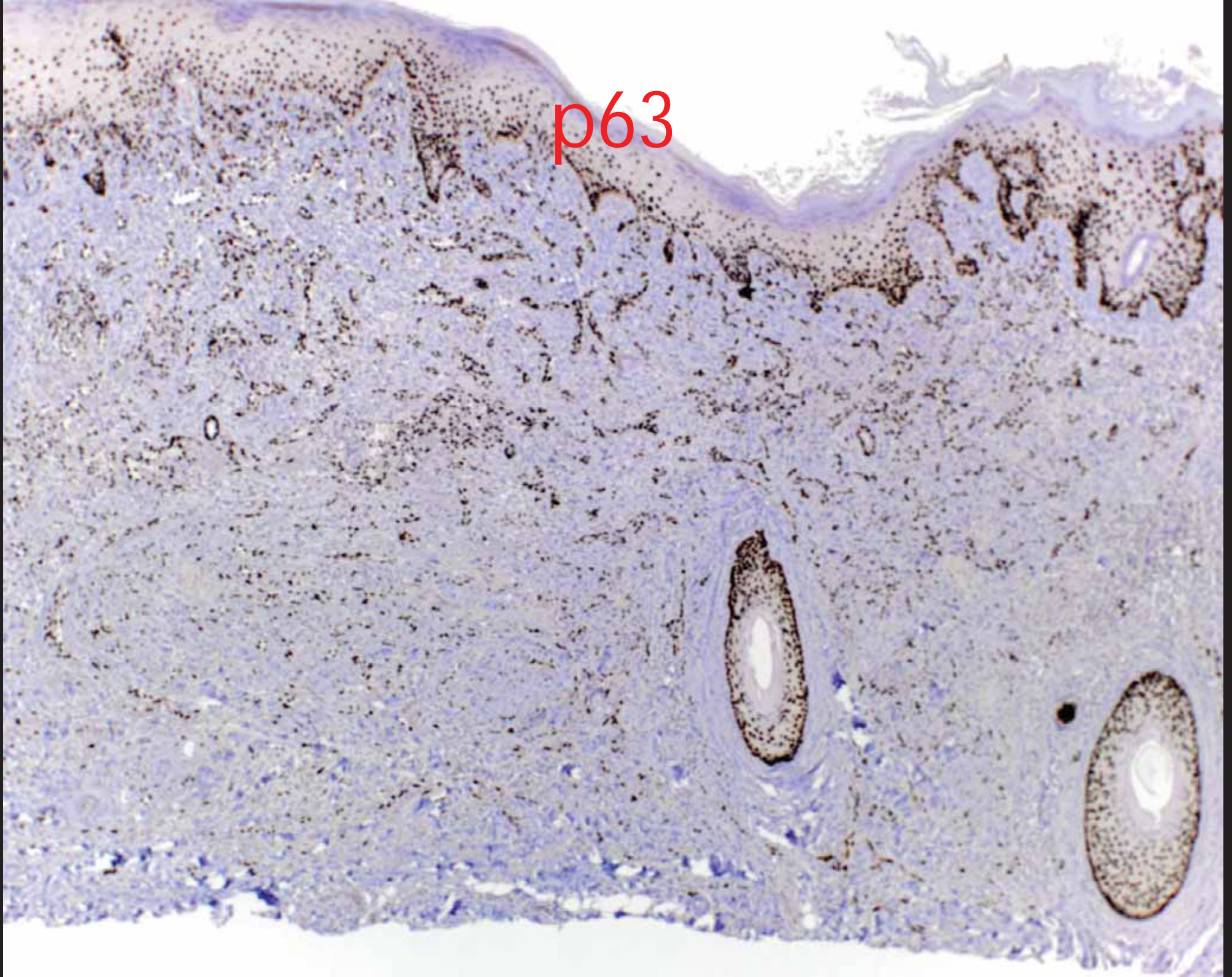
But ERG is so specific!

Essential features

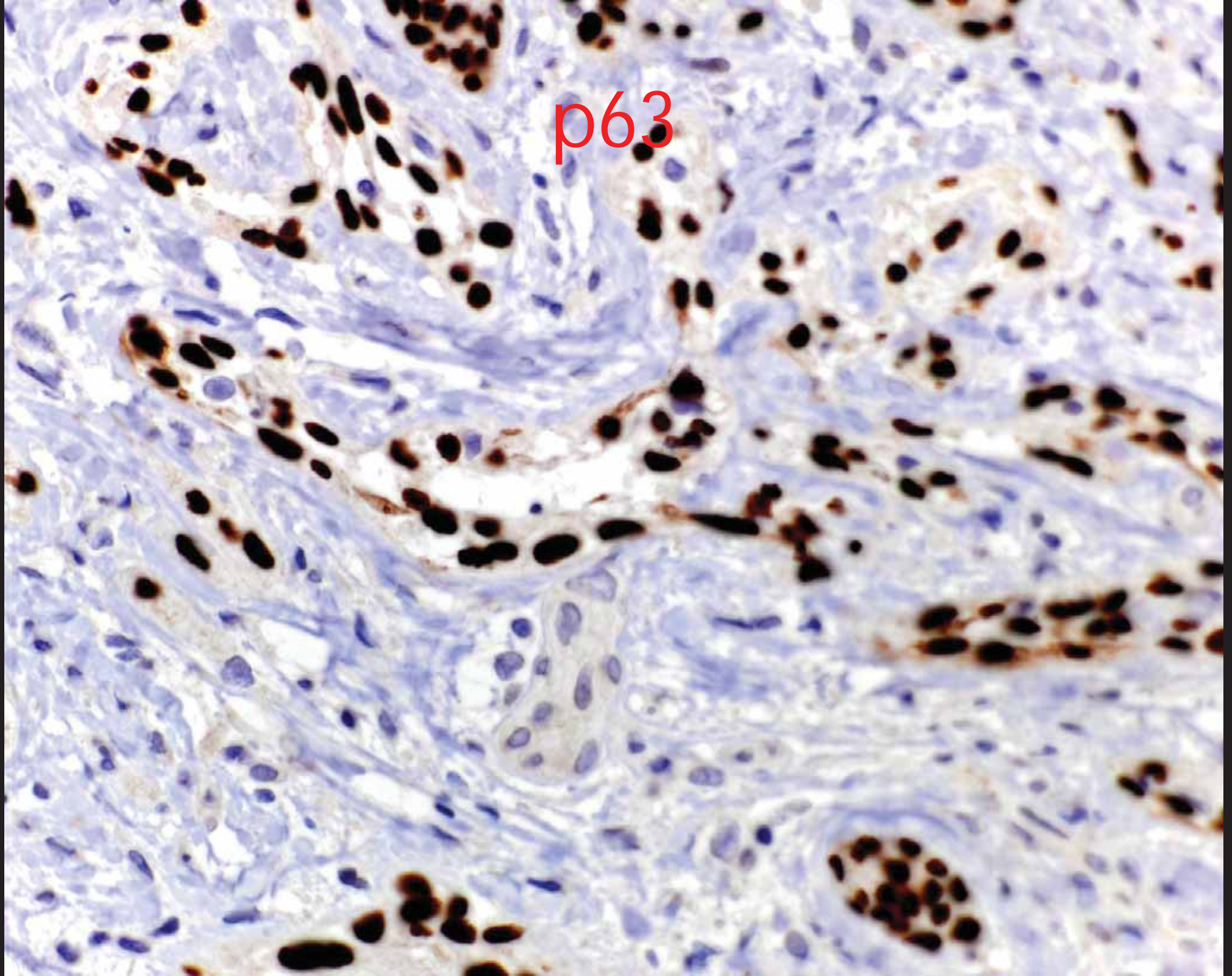
- *ETS* family transcription factor ERG has nuclear staining
- Highly sensitive and quite specific immunohistochemical marker for vascular differentiation, including benign and malignant vascular tumors
- Anti-N terminus ERG monoclonal antibody is a useful marker for certain chondrogenic tumors
- Also expressed in tumors harboring *ERG* rearrangements: 50% of prostatic adenocarcinomas and 5% of Ewing sarcomas
- Marker for immature myeloid differentiation including cases of acute myeloid leukemia / myeloid sarcoma



p63



p63



Histopathology 1992, 21, 13-23

Pseudoangiosarcomatous carcinoma: a clinicopathological study of seven cases

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Date of submission 10 December 1991

Accepted for publication 27 January 1992

BANERJEE S.S., EYDEN B.P., WELLS S., MCWILLIAM L.J. & HARRIS M.

(1992) *Histopathology* 21, 13-23



Figure 1. Case 1. Islands of neoplastic squamous epithelium merging with the pseudoangiosarcomatous component. H & E.

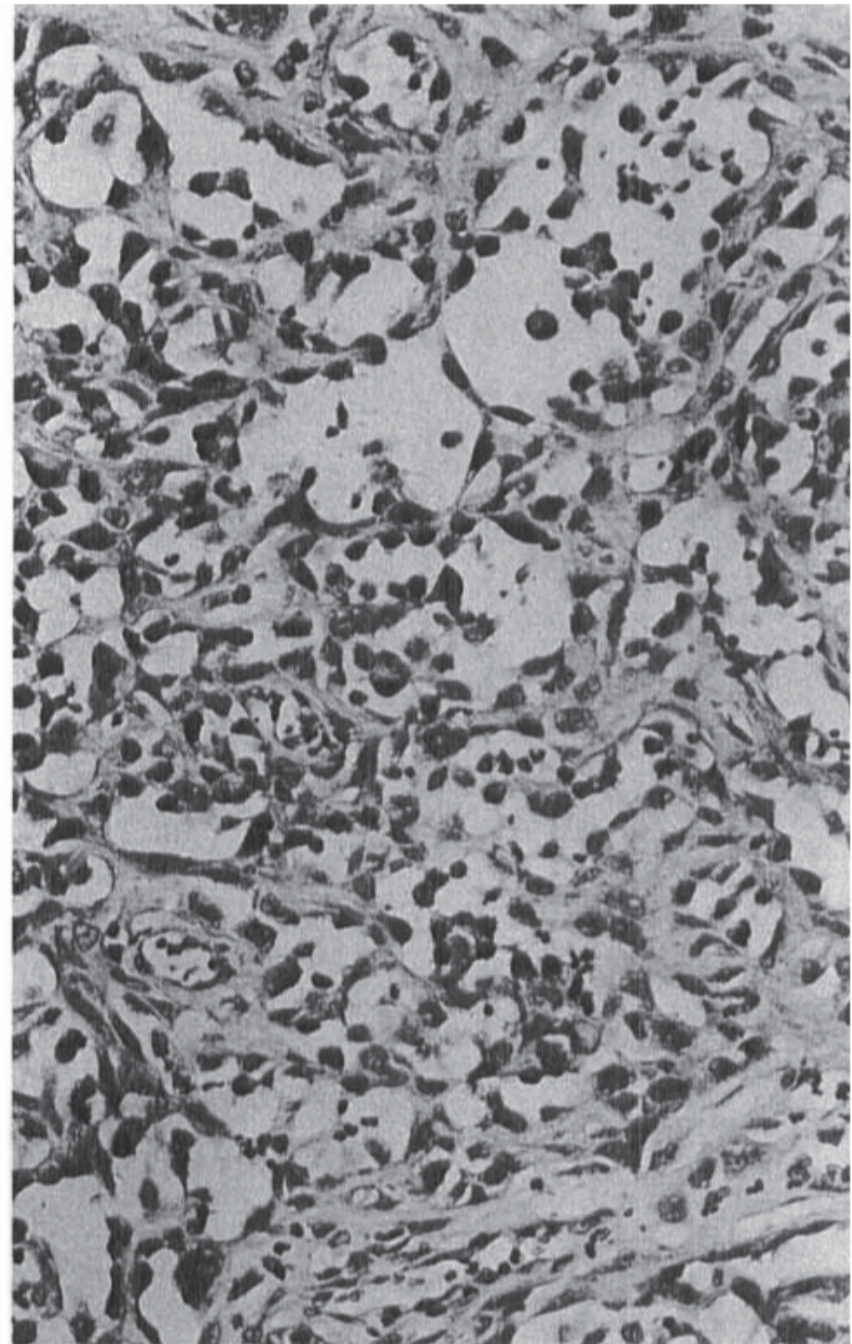


Figure 2. Case 2. Detail of the pseudoangiosarcomatous pattern. Small cystic spaces and complex anastomosing channels lined by neoplastic cells. H & E.

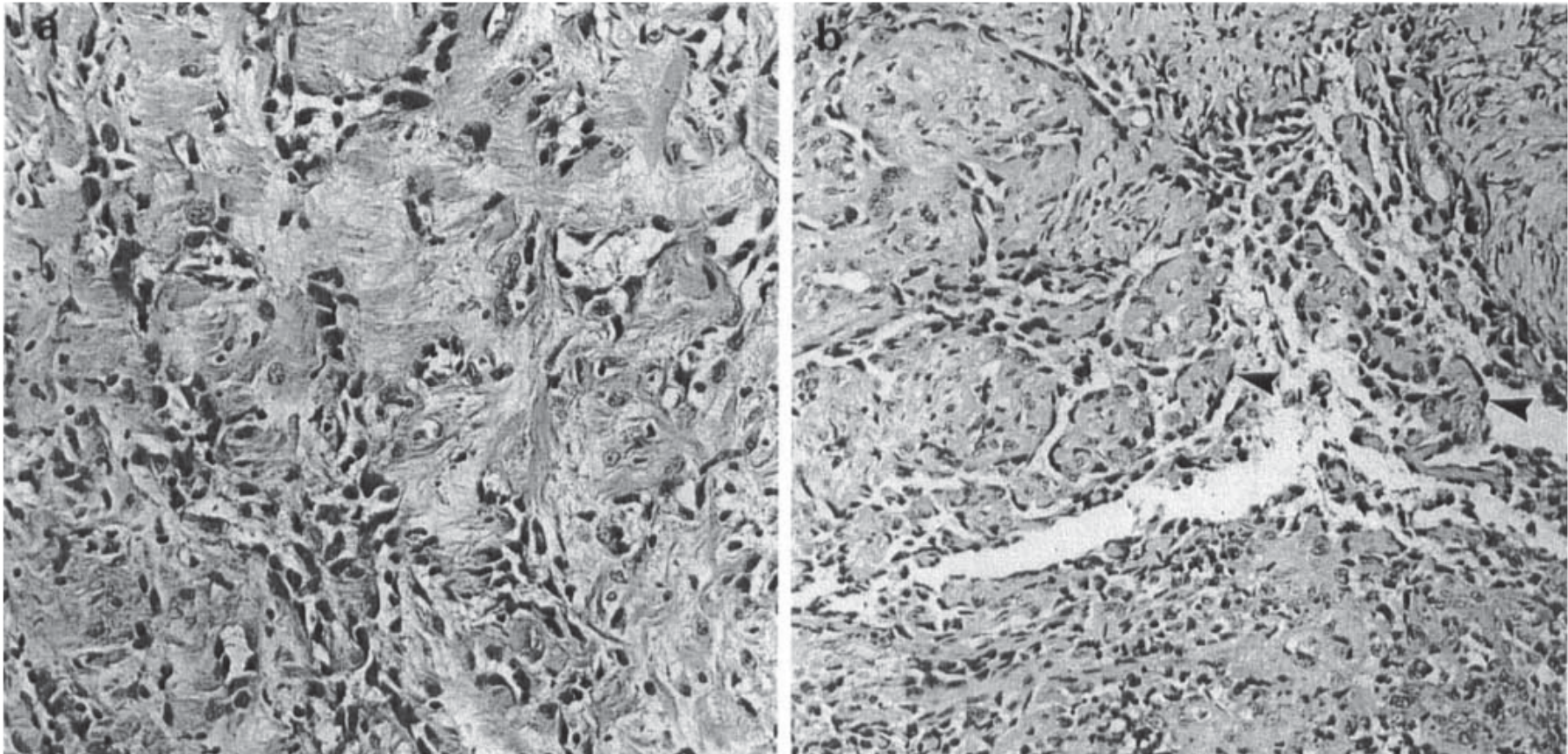


Figure 3. Case 1. a Pseudovascular structures dissecting dense collagen. H & E. **b** Small tufts of fibrous tissue covered by neoplastic cells (arrowheads) within an irregular space. Also note the collagen dissection pattern. H & E.

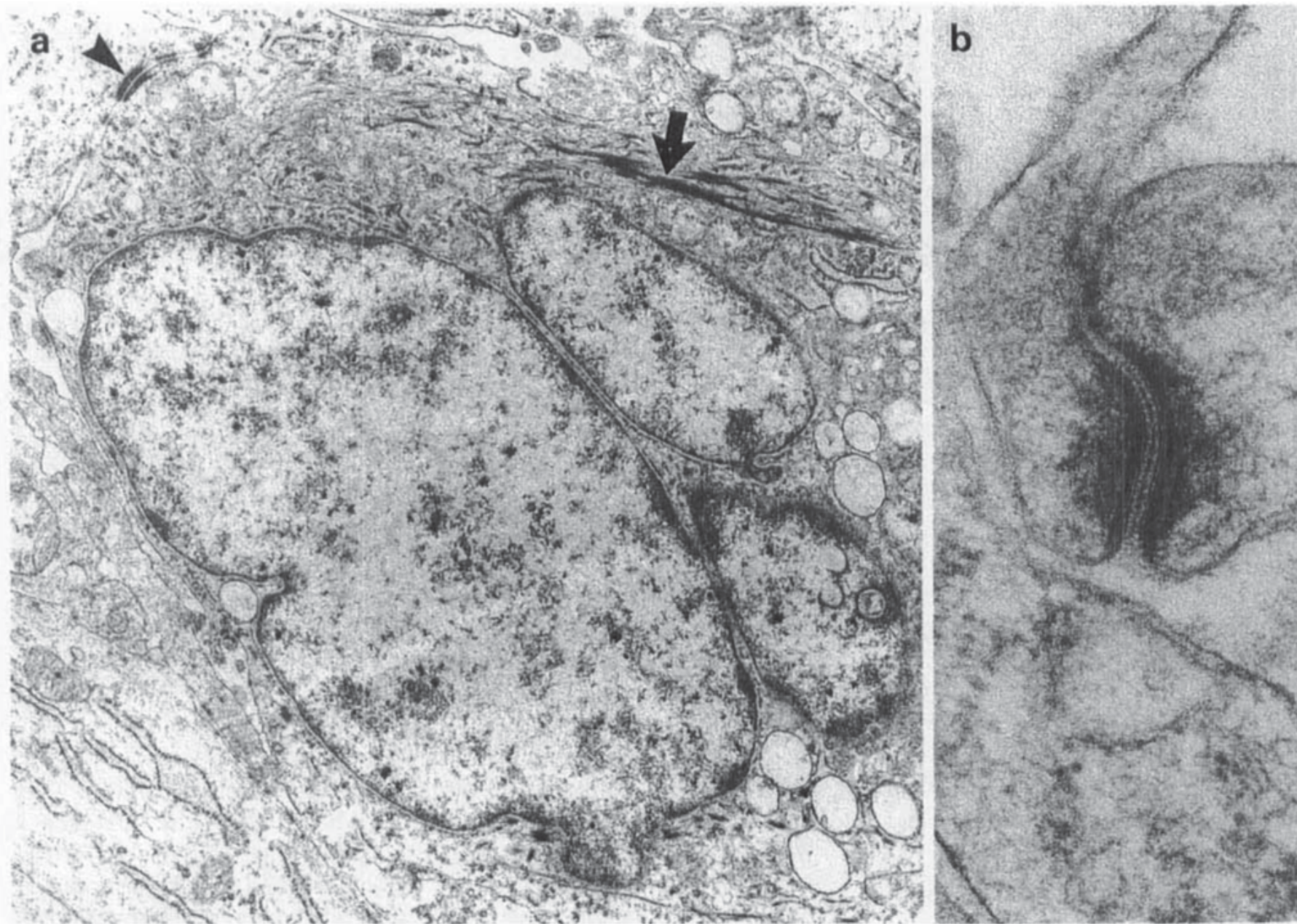


Figure 9. Case 1. Electronmicrographs. a Tumour cell showing tonofibrils (arrow) and desmosome (arrowhead). **b** Detail of desmosome. **a** $\times 13\,130$, **b** $\times 92\,920$.

CONCISE COMMUNICATION

Pseudovascular squamous cell carcinoma: A review of the published work and reassessment of prognosis

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ABSTRACT

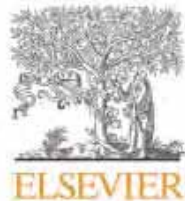
A 90-year-old Japanese woman presented with a dome-shaped, dark-red, ulcerated nodule measuring 23 mm × 19 mm × 9 mm on the right side of the nasal root. Histologically, anastomosing cord-like arrays of atypical polygonal keratinocytes exhibiting internal pseudolumina containing detached cells and erythrocytes were observed. Although acantholytic and cohesive areas overlapped, cancer pearls were not detected. The lower epidermis partially demonstrated scattered dyskeratotic and acantholytic keratinocytes with loss of polarity, continuous with an underlying tumor mass. The tumor cells were positive for a variety of cytokeratins, p40 and vimentin. The Ki-67 proliferation index was 50–60%. Both CD31 and CD34 were expressed in reactive blood vessels of the tumor. A local excision margined by 1 mm was performed, followed by X rays and electron beam irradiation. Neither lymph node nor distant metastasis has appeared over the 14 months since the excision. We performed a review of the published work and identified 24 previously reported patients with pseudovascular squamous cell carcinoma of the skin, oral mucosa and vulva to reassess the prognosis of this tumor. In 12 of these patients (50%), sites other than the head and neck were involved. Eight (33%) tumor-associated deaths occurred. It is believed that pseudovascular squamous cell carcinoma has a tendency to develop at morbid skin and mucous membranes sites in organs other than the face and neck and to possess an aggressive clinical behavior.

Key words: acantholytic, aggressive, pseudoangiosarcomatous, pseudovascular, squamous cell carcinoma.

Table 1. Summary of pseudovascular SCC cases

No. (reference)	Age, years/sex	Site	Treatment	Prognosis	Possible genesis
1-6 ¹	46-80/ M (5) F (1)	Face and neck (3) Hand (2) Leg (1)	EX and LND, Chemo	Died (3)	Cadaveric renal transplant (1) Cadaveric renal allograft (1)
7 ²	57/F	Vulva	RV and bilateral inguinal femoral LND, Chemo	Died (PM)	HPV(-)
8 ³	59/M	Buccal mucosa	Radical EX and ND	Alive for 12 months	Heavy smoker
9 ³	77/F	Rt. floor of the mouth	EX and radical ND	Alive for 16 months	Non-smoker
10 ⁴	86/F	Rt. cheek	EX	Alive for 36 months	NS
11 ⁵	78/M	Rt. ear	Radical EX, RT	Died (mediastinal LNM)	NS
12 ⁵	64/M	Perineum	EX	Died (PM)	Urethral stricture and fistula
13 ⁵	82/M	Lt. cheek	Biopsy, RT	Alive	NS
14 ⁶	96/F	Lt. leg	EX	Alive for 4 months	Chronic venous insufficiency
15 ⁸	65/F	Rt. ankle	EX	Alive for 8 months	Burn scar
16 ⁹	75/F	Lt. leg	Biopsy	NS	Lymphedema from surgical wound
17 ¹⁰	43/M	Rt. cheek	EX, RT	NS (extensive local recurrence)	NS
18 ¹⁰	82/F	Rt. leg	Biopsy, amputation	NS	Venous ulcer
19 ¹¹	79/F	Sacrum	Biopsy	Died	Decubitus
20 ¹²	69/M	Lt. ear	EX and radical ND, RT, Chemo	Died (PM)	NS
21 ¹³	84/F	Vulva	RV, inguinal LND	Died (perineal abscess)	NS
22 ¹⁴	74/M	Face	EX, RT	Died (PM and preauricular LNM)	NS
23 ¹⁵	78/F	Vulva	RV and bilateral inguinal LND, RT	Alive for 5 months	NS
24 (present case)	90/F	Root of nose	EX, RT	Alive for 12 months	NS

Chemo, chemotherapy; EX, excision; HPV, human papillomavirus; LND, lymph node dissection; LNM, lymph node metastasis; Lt., left; ND, neck dissection; NS, not stated; PM, pulmonary metastasis; RT, radiotherapy; Rt., right; RV, radical vulvectomy.



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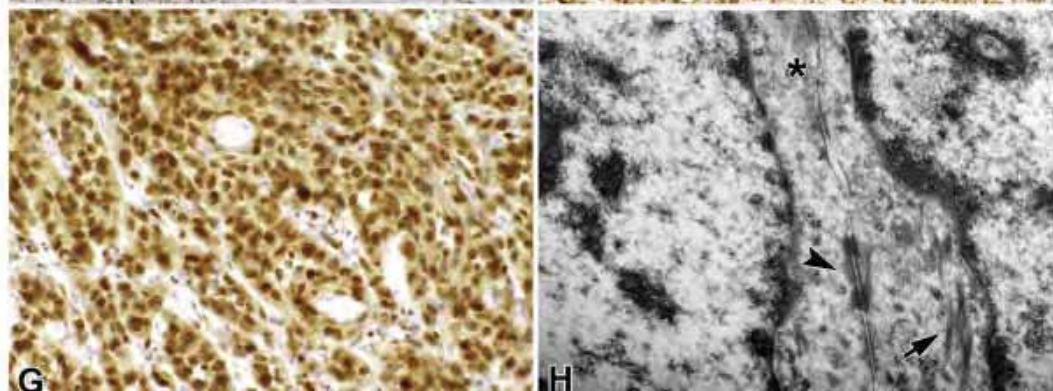
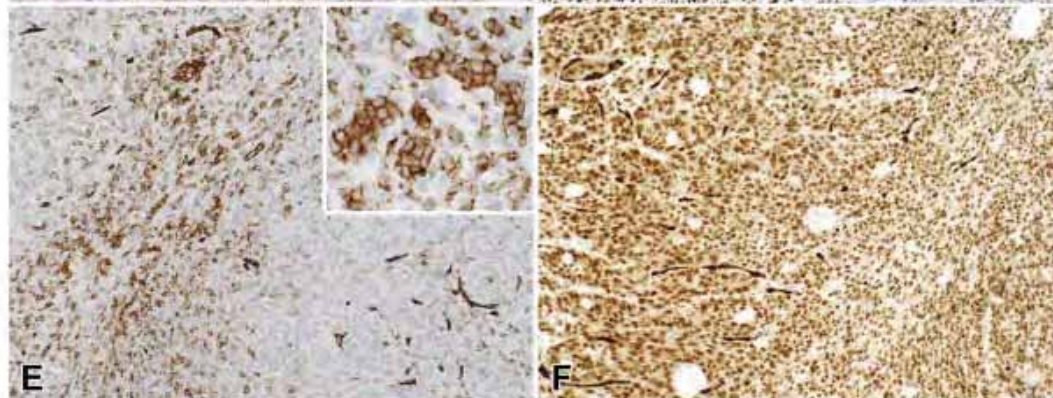
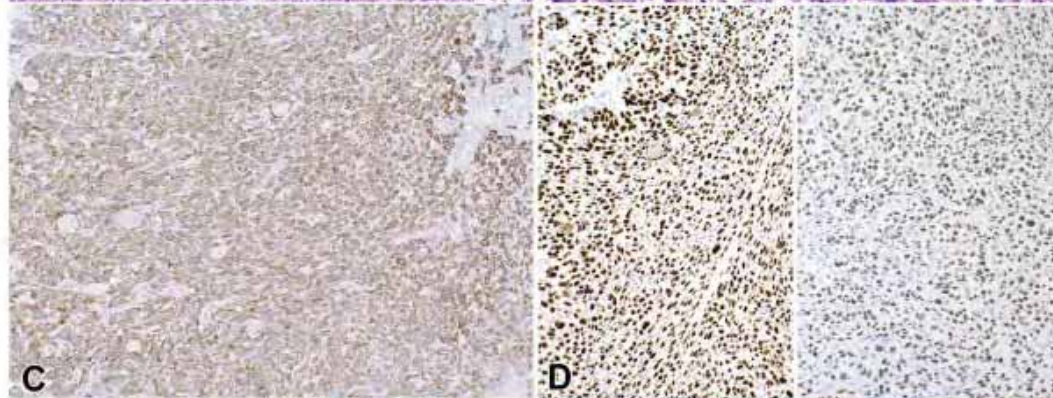
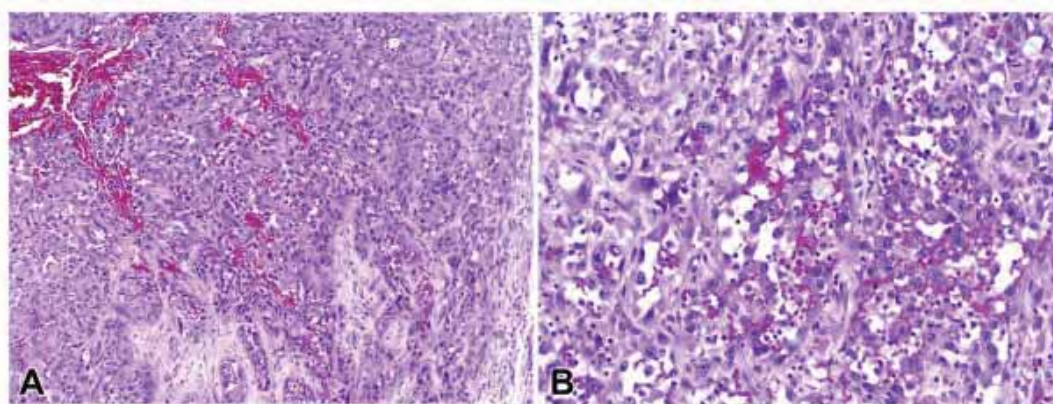
Metastatic squamous cell carcinoma with pseudoangiosarcomatous features and aberrant expression of vascular markers



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Pseudoangiosarcomatous SCC

- Rare
- About half on head and neck, skin and mucosa
- Death from disease in about a third
- Aberrant vascular markers can occur
- Mucin deposition

Why this ddx matters

- Both are malignant
- Pseudoangiosarcomatous SCC can be cleared surgically, true angiosarcoma is more difficult to excise
- Post-op radiotherapy often given for true angiosarcoma

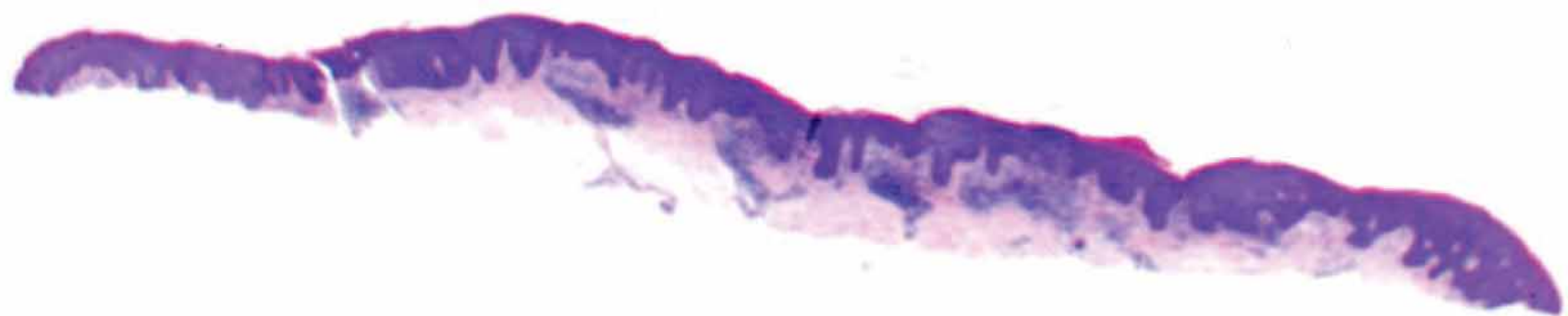
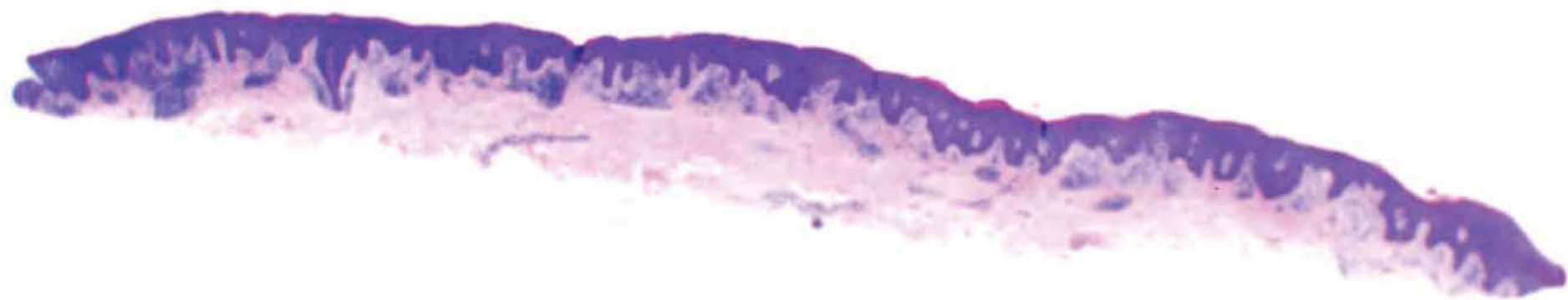
Case 2

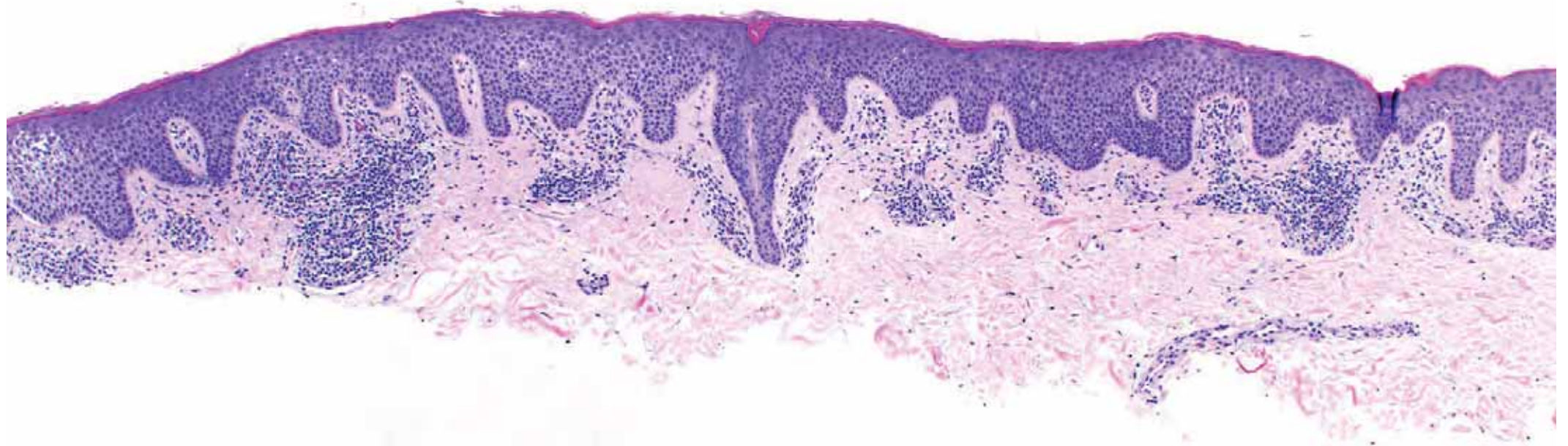
49 year old man, biopsy from inner arm. Many orange-pink digitate and oval lesions on trunk and proximal limbs, reportedly told in London at age 44 that she had mycosis fungoides



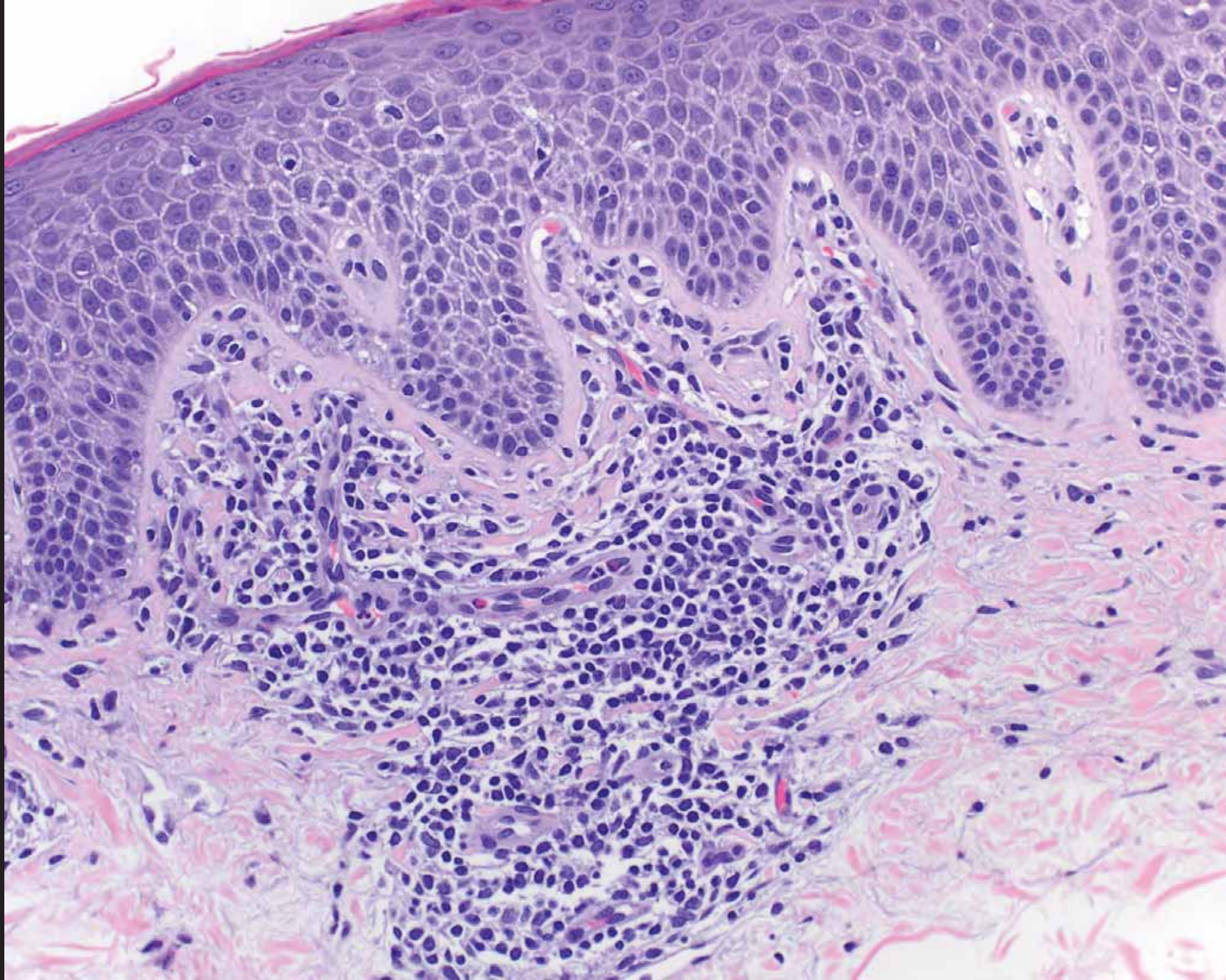


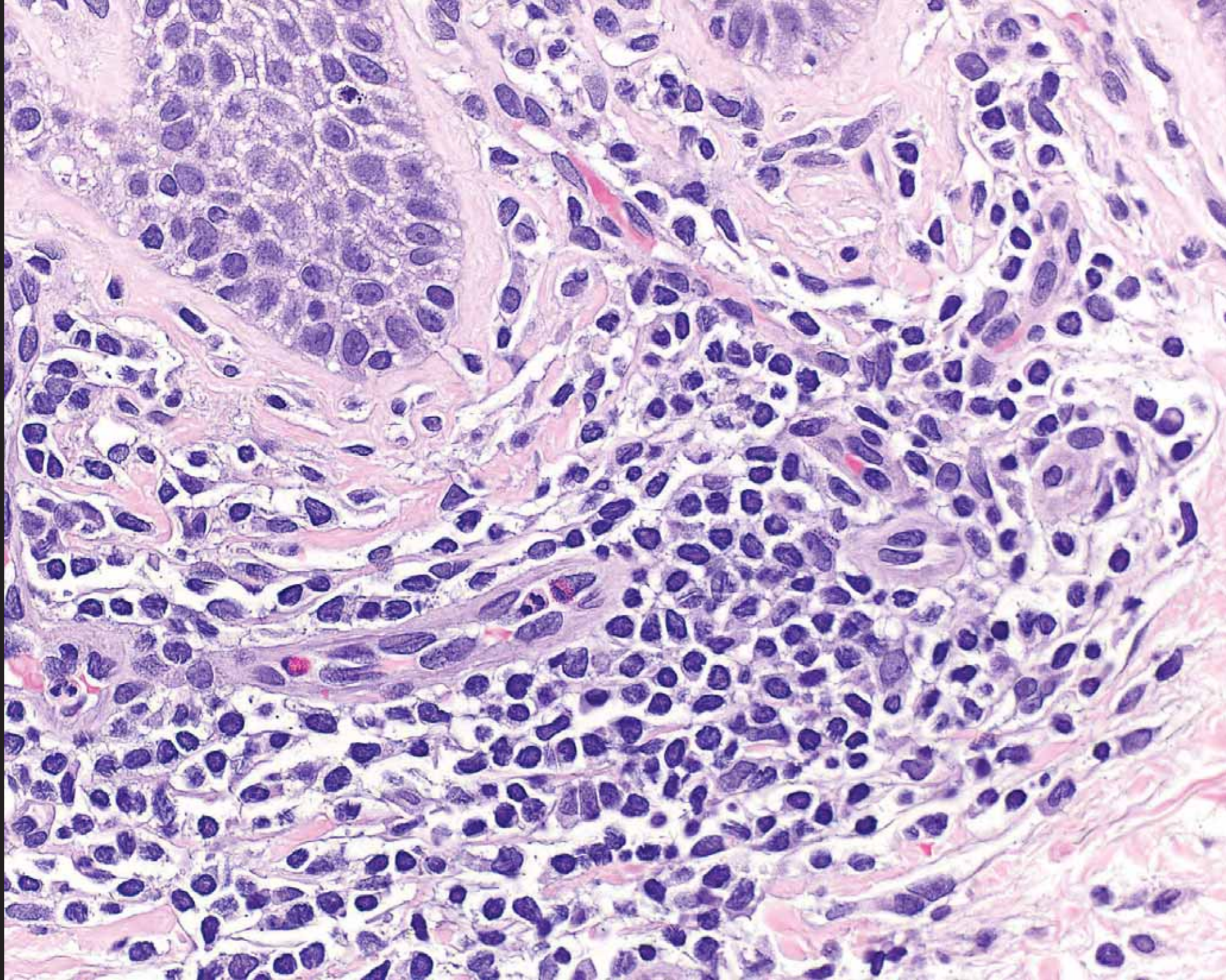


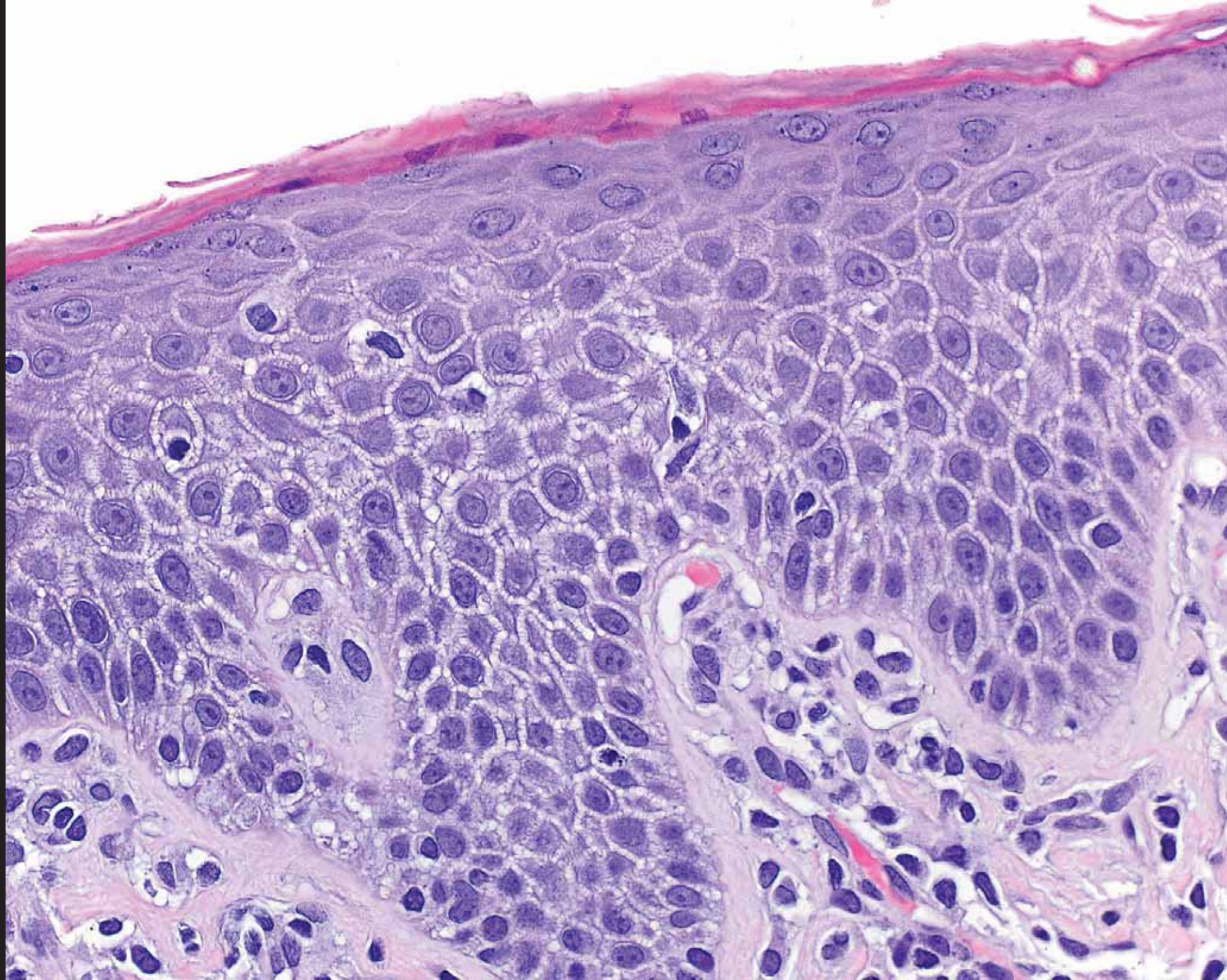


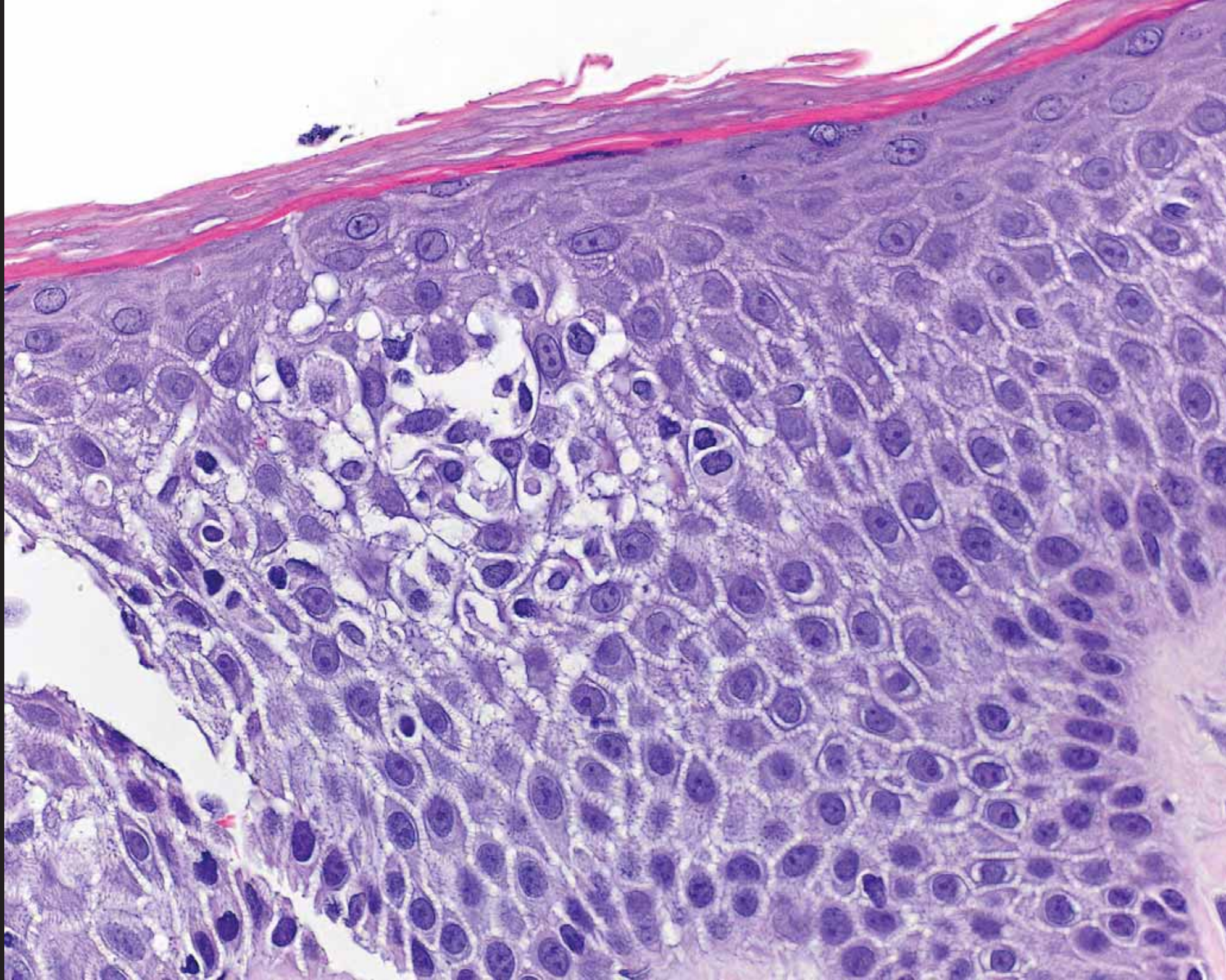


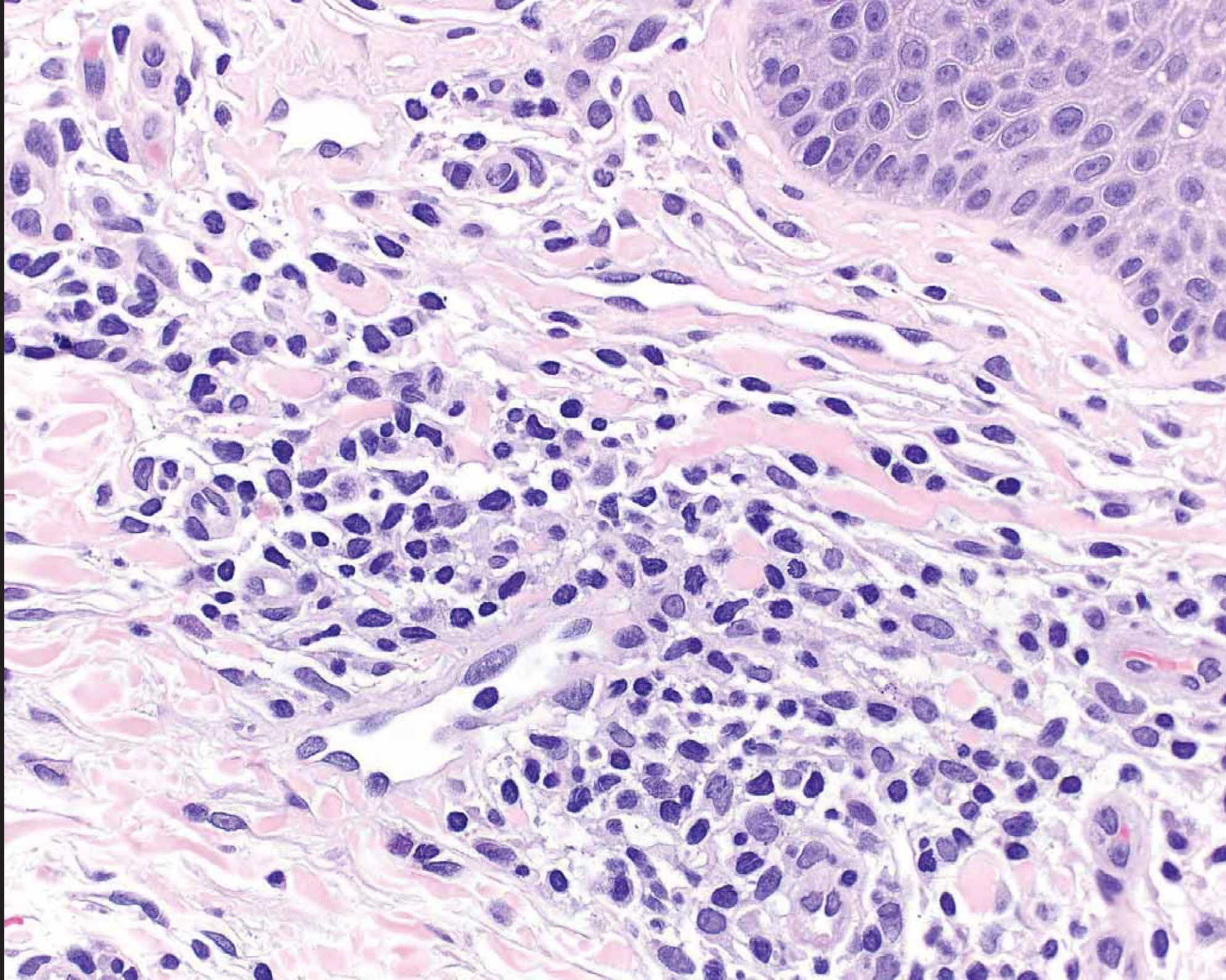


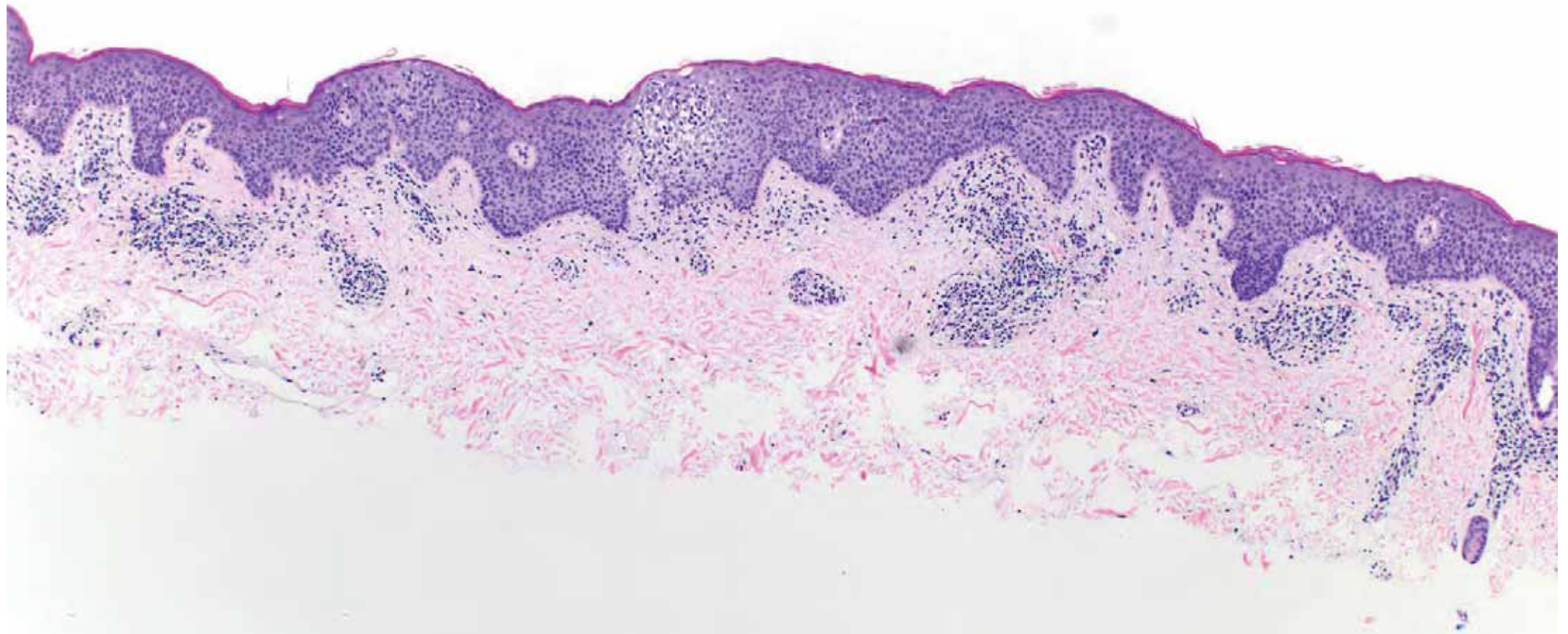


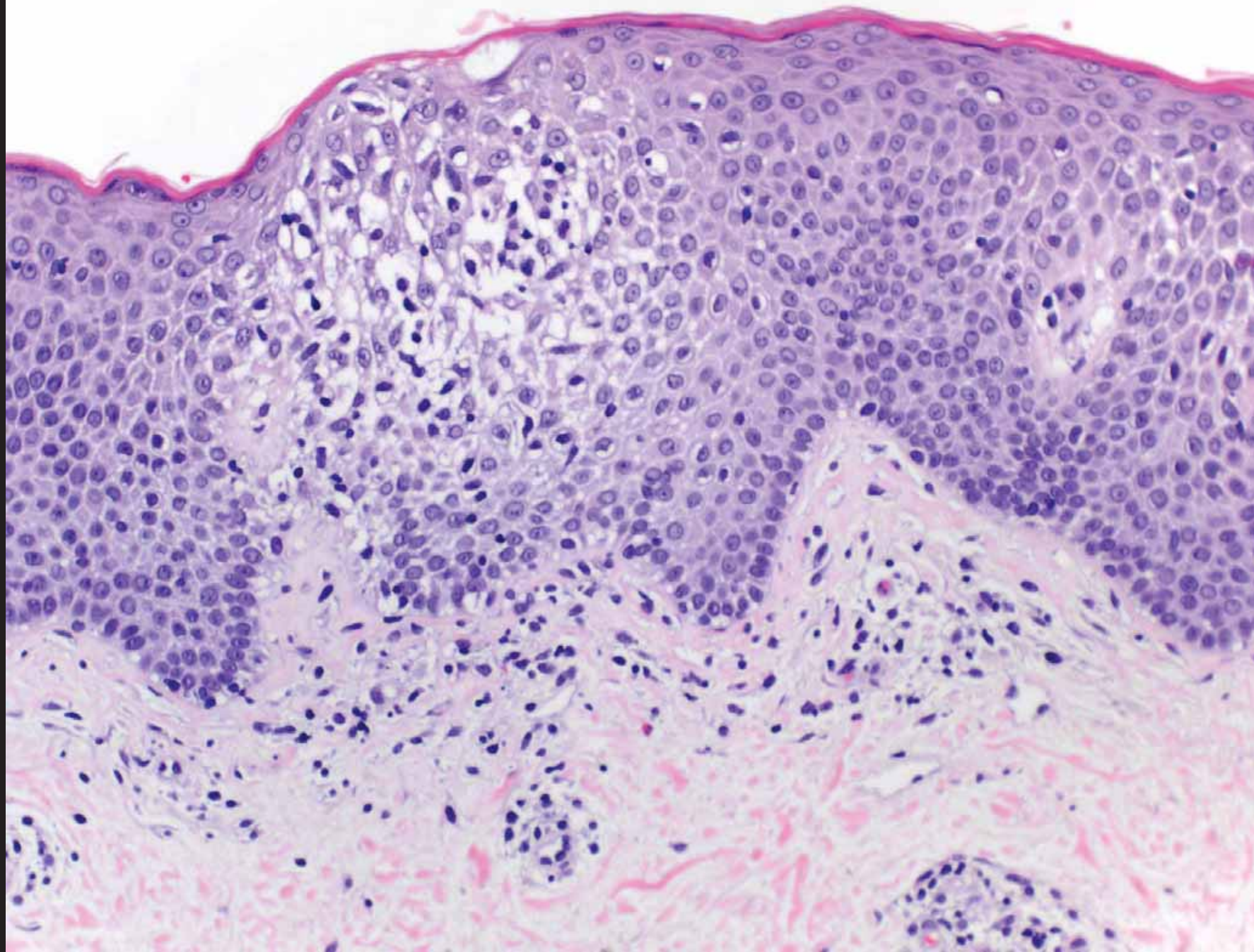


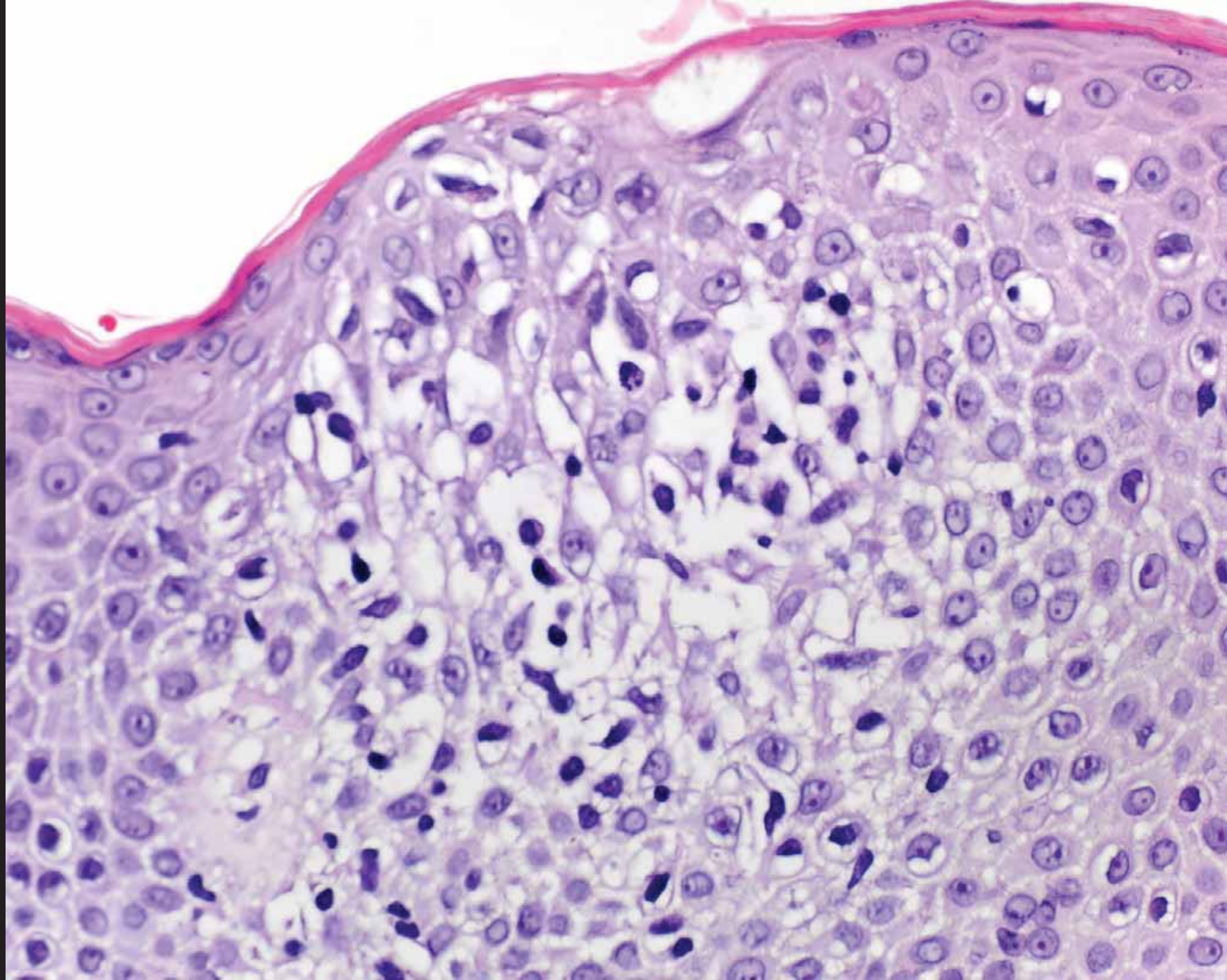












Digitate Dermatosis

A New Look at Symmetrical,
Small Plaque Parapsoriasis

*Chung-Hong Hu, MD, and
Richard K. Winkelmann, MD, Rochester, Minn*

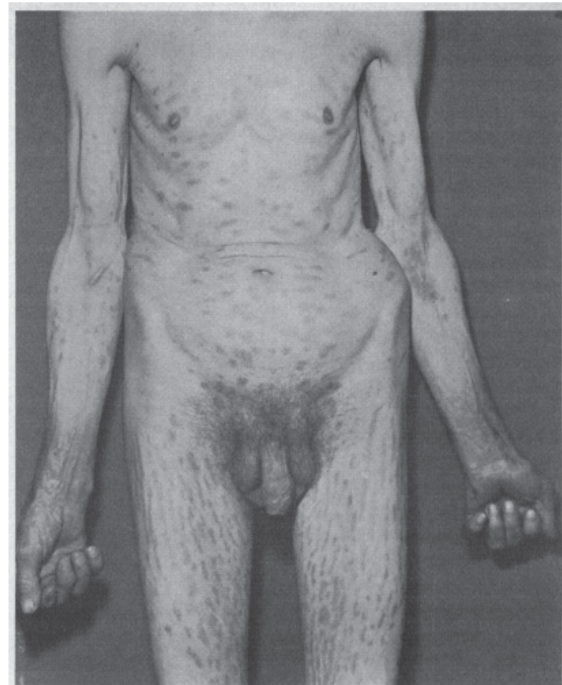


Fig 1.—Anterior of trunk and limbs showing bilateral symmetrical fingerprint-like eruption (case 1)

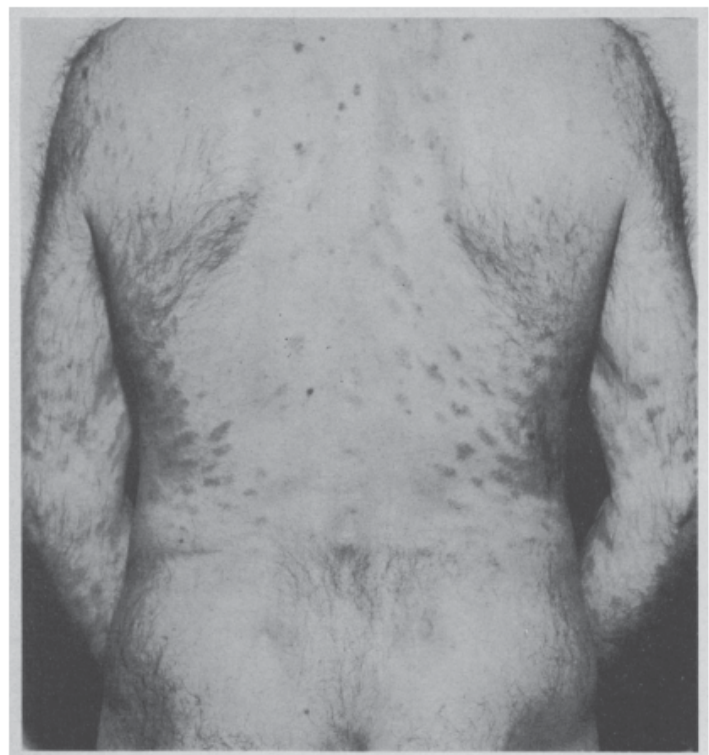


Fig 2.—Symmetrical oval and fingerprint-like patches showing pityriasis rosea-like distribution (case 2).



Fig 3.—Discrete oval and fingerprint-like patches follow lines of cleavage (case 5).

Controversies in Dermatopathology

Guttate Parapsoriasis/Digitate Dermatitis (Small Plaque Parapsoriasis) Is Mycosis Fungoides

Daisy King-Ismael, M.D., and A. Bernard Ackerman, M.D.

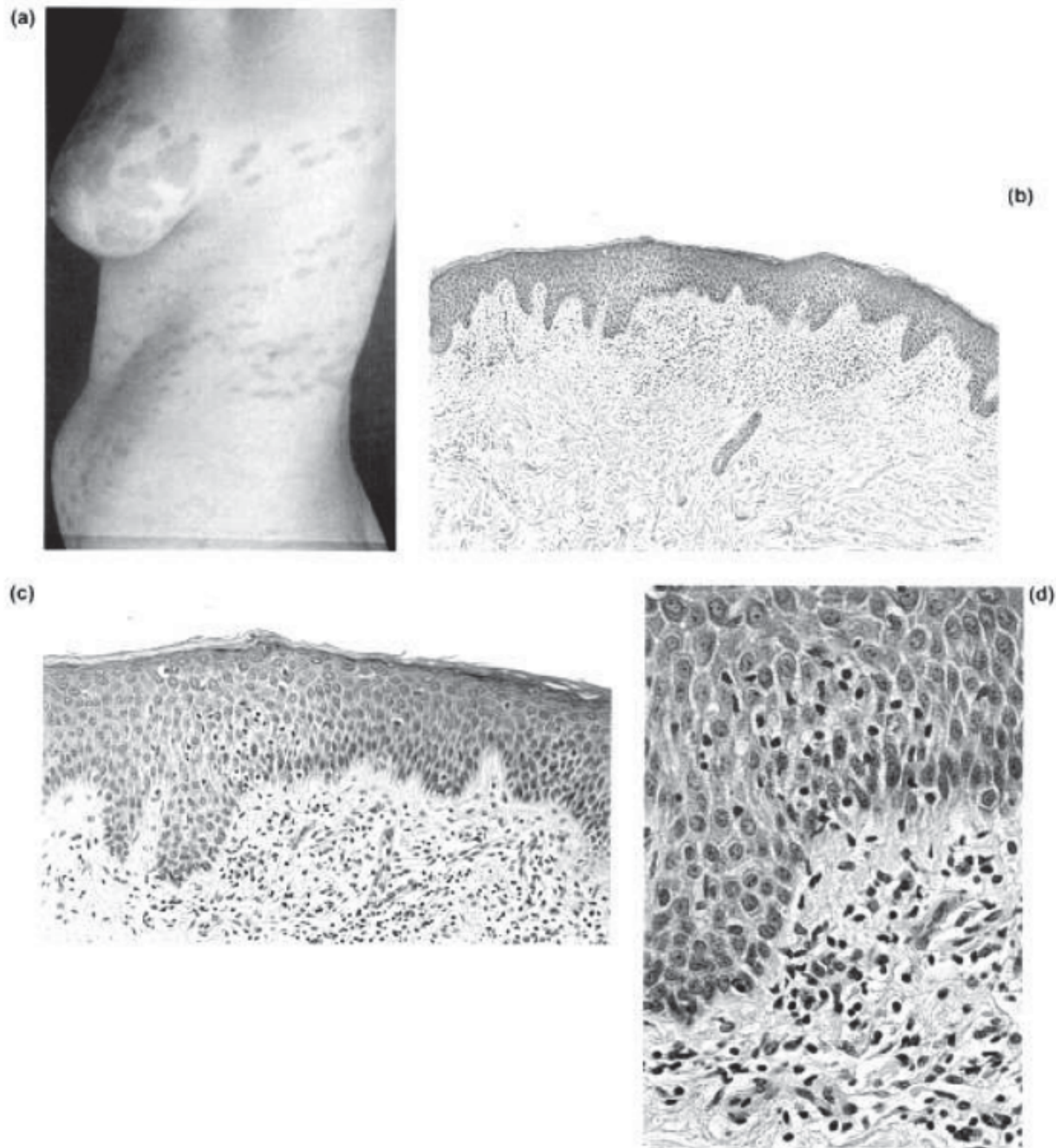


FIG. 3. (a) Mycosis fungoides. Several types of lesions are seen in this photograph of a 29-year-old woman. On the breast there are yellow-red patches and slightly elevated plaques—i.e., lesions of "parapsoriasis en plaques." On the side of the torso, toward the back, there are ovoid and elliptical red macules, patches, papules, and subtle plaques that resemble lesions of pityriasis rosea but, in some foci, have the appearance of fingers pressed against the skin—i.e., lesions of "digitate dermatosis." On the

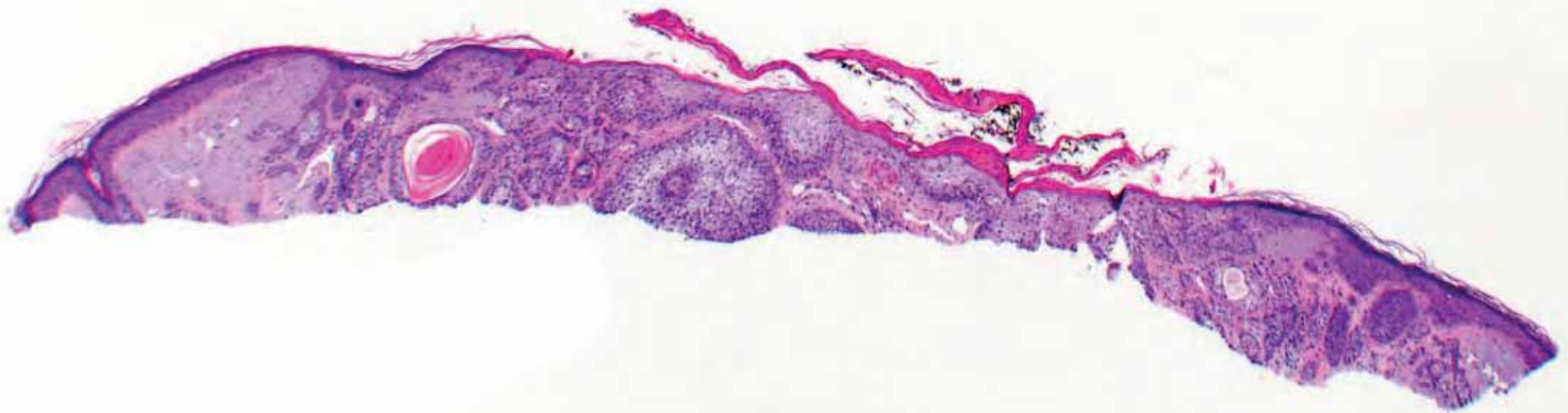
Does the ddx between DigDerm and patch MF matter?

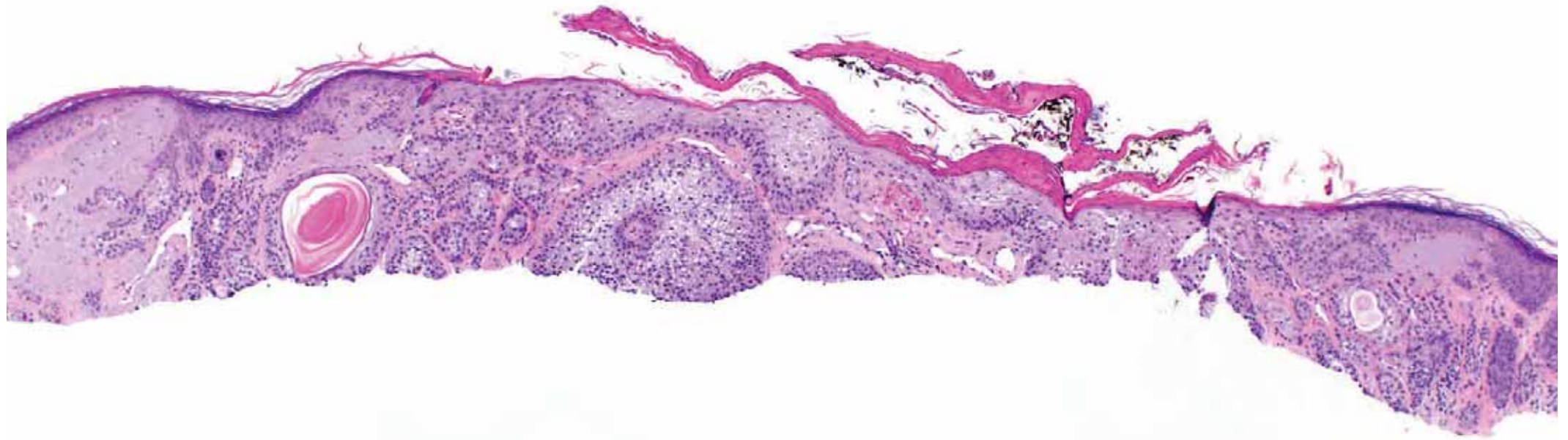
- Dr. Chung Hong-Hu, asked at SFO if any of his patients from Mayo developed MF, said "no"
- Both could be treated with topical CS and UVB
- Seemingly, "not so much", but....

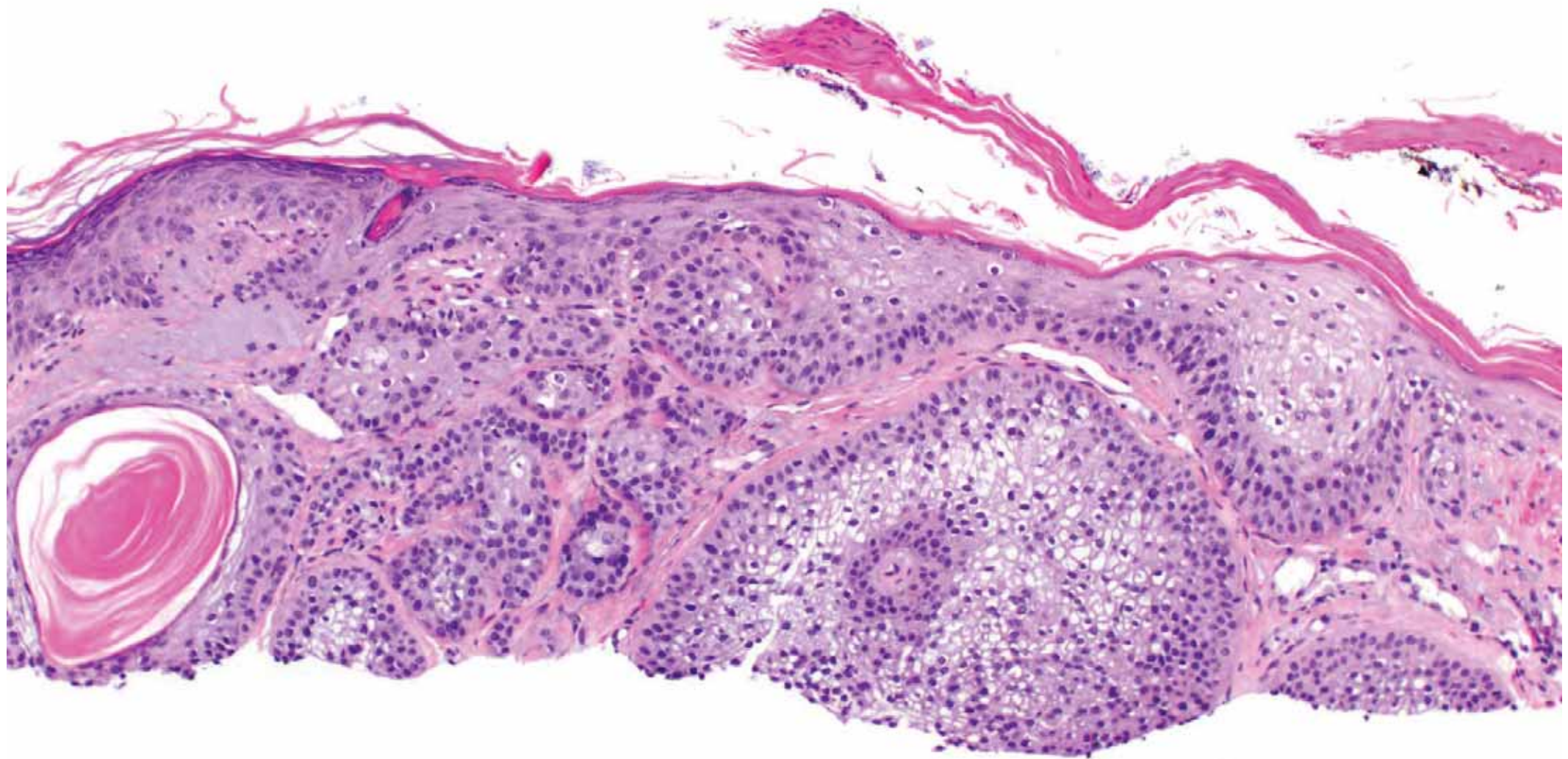
Case 3

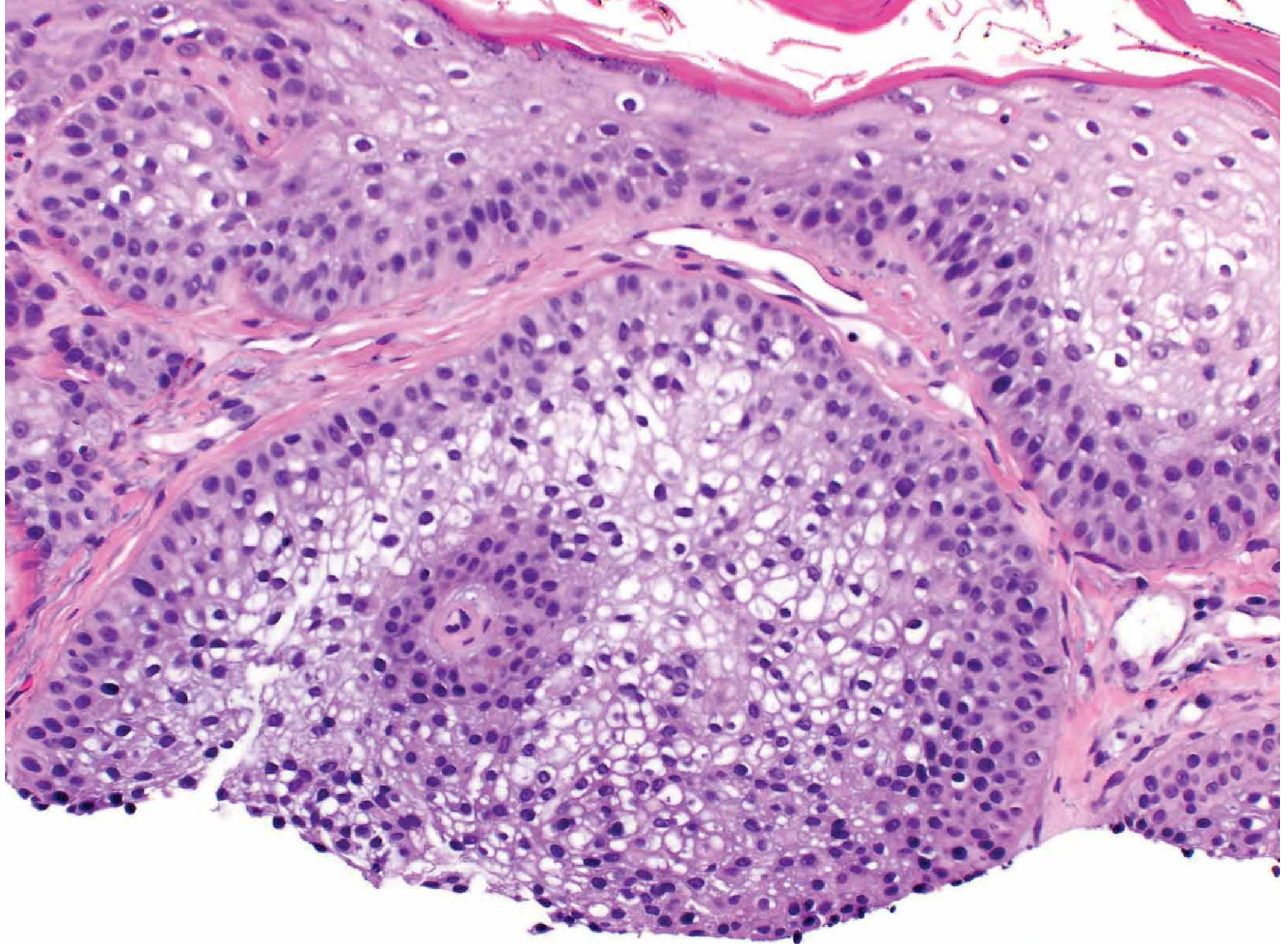
55 year old woman, 8 mm eroded papule,
bridge of the nose

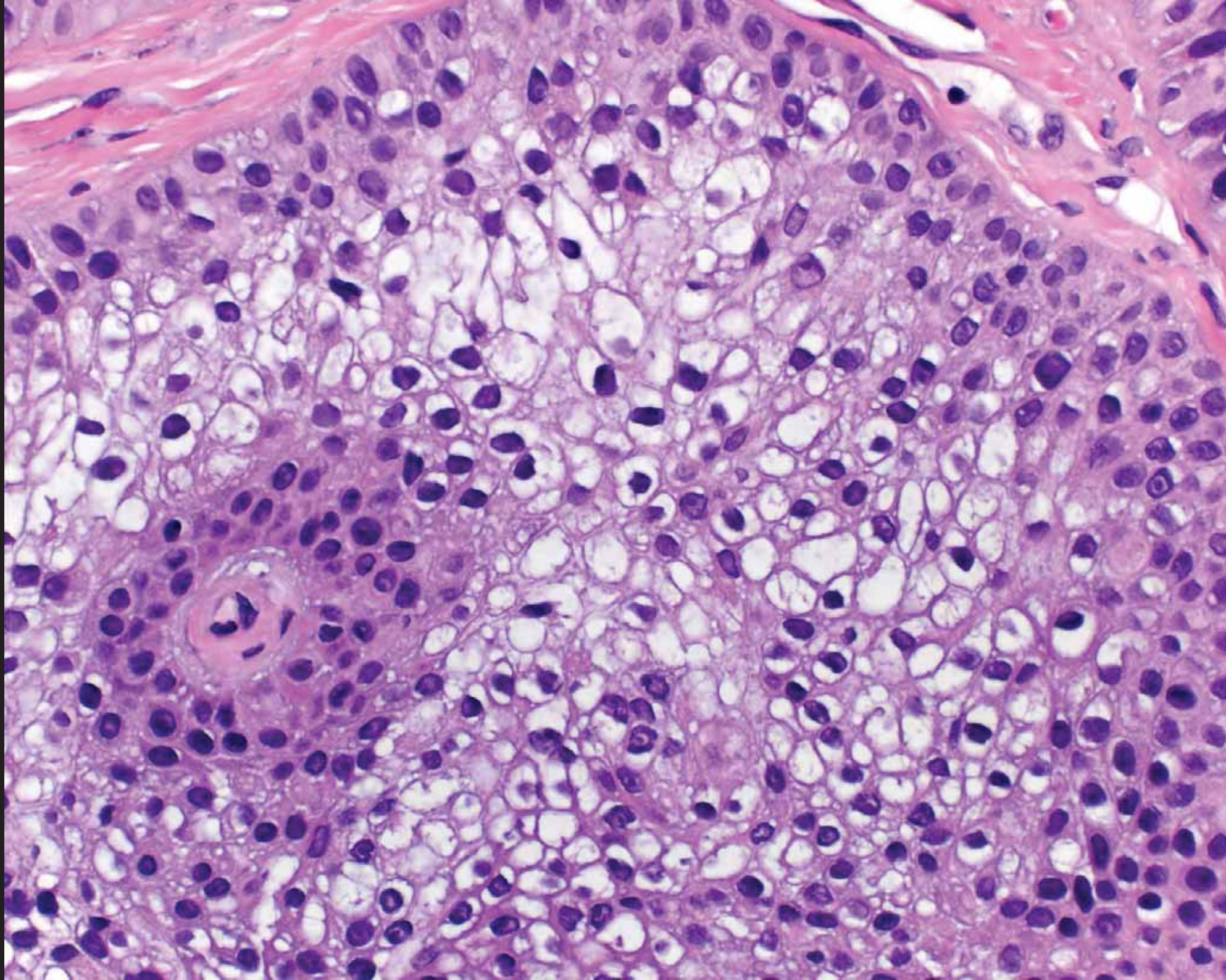
2020

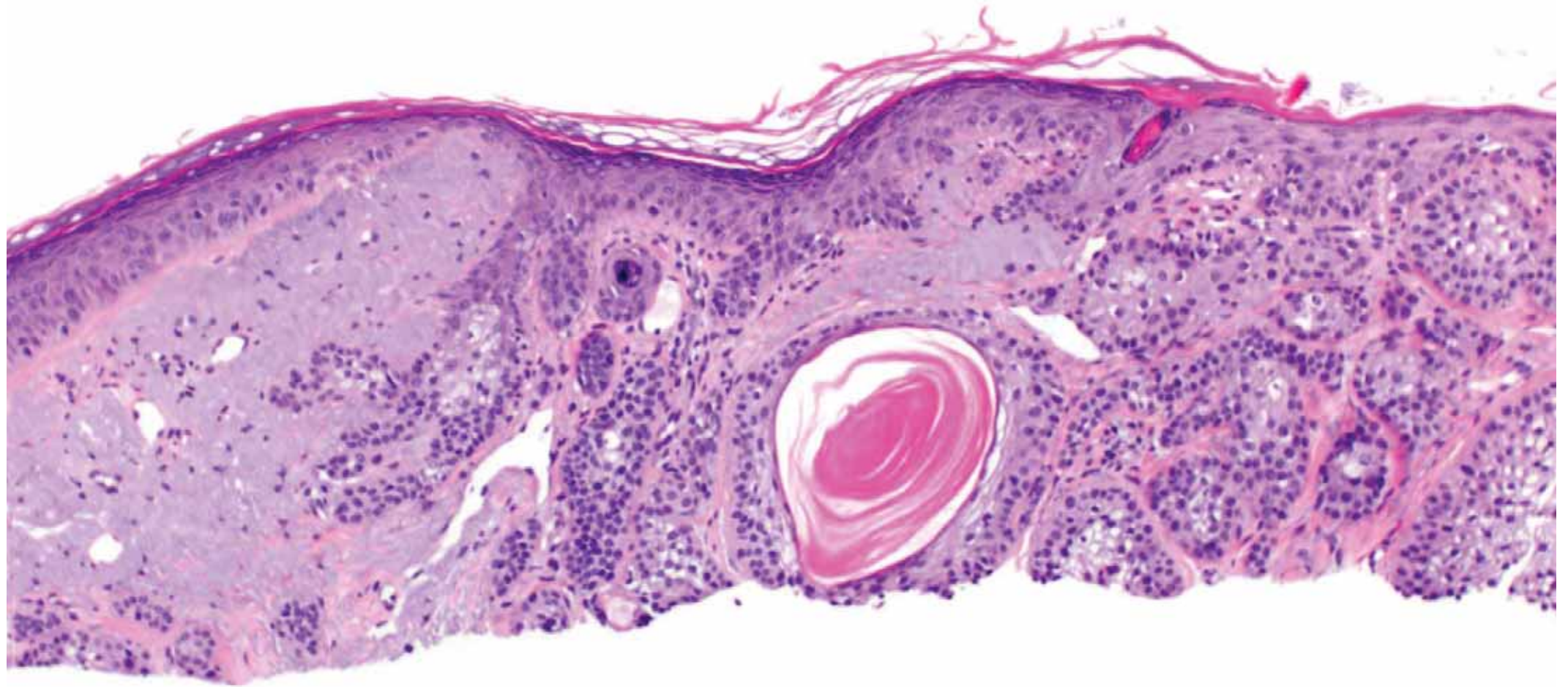


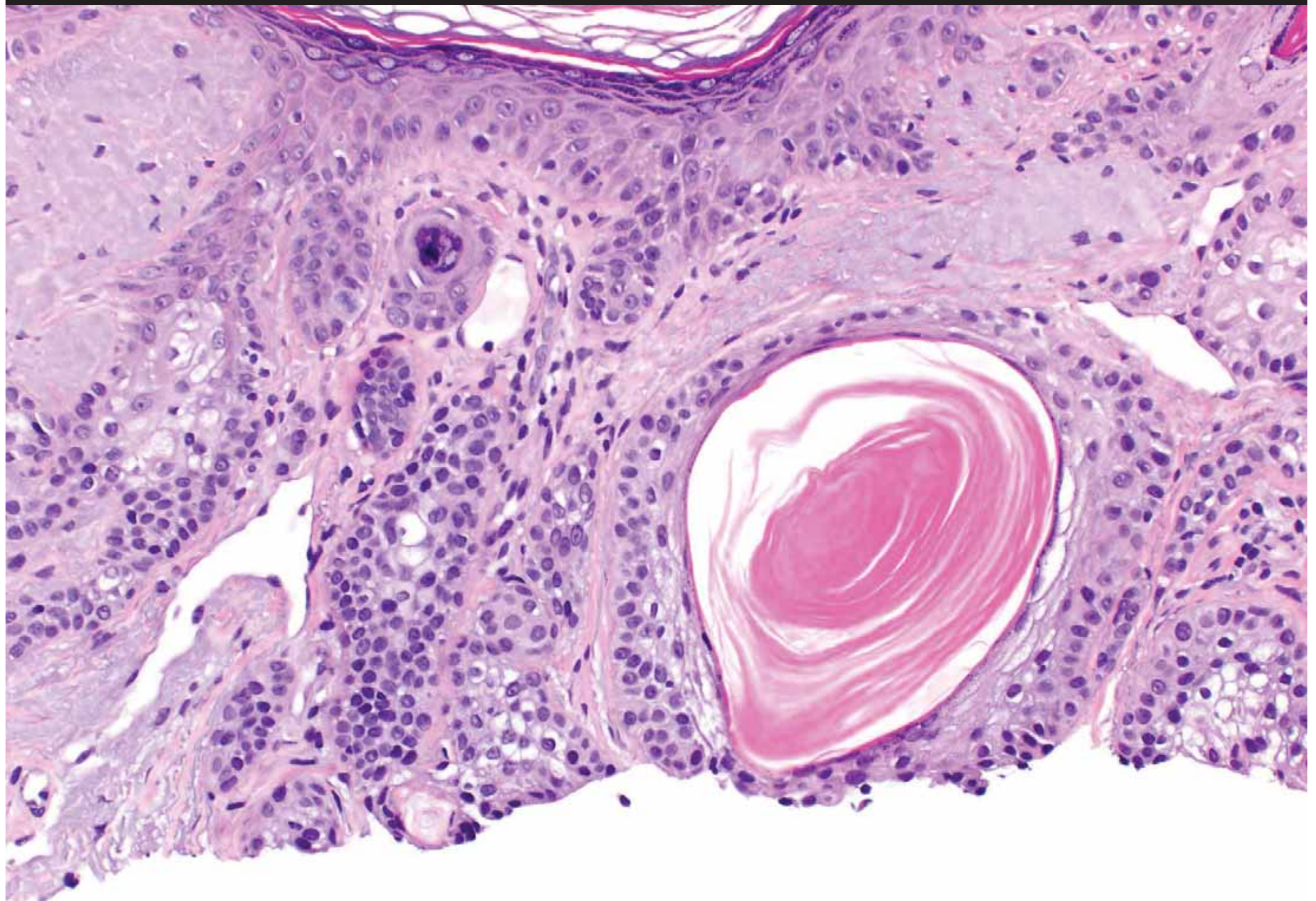


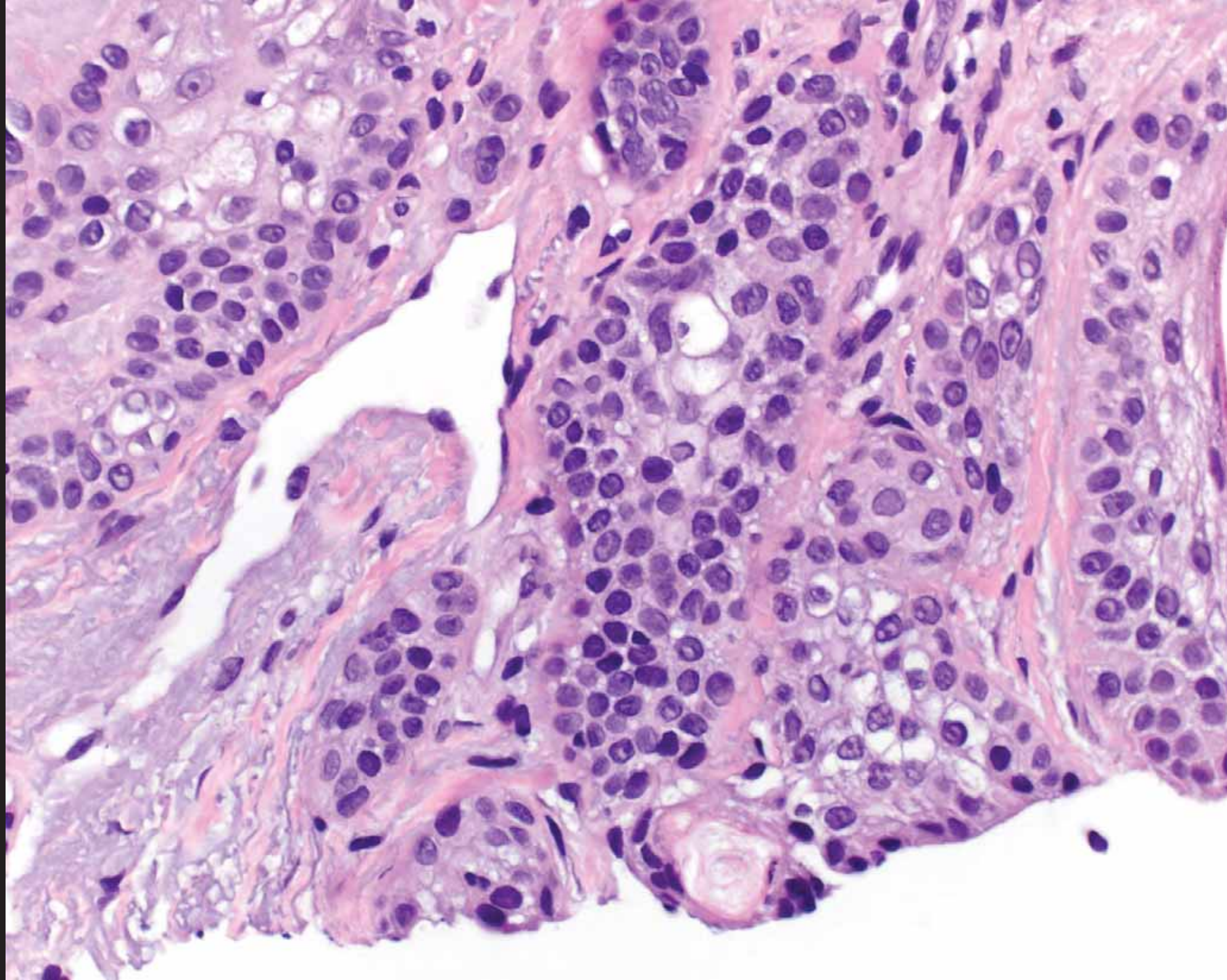












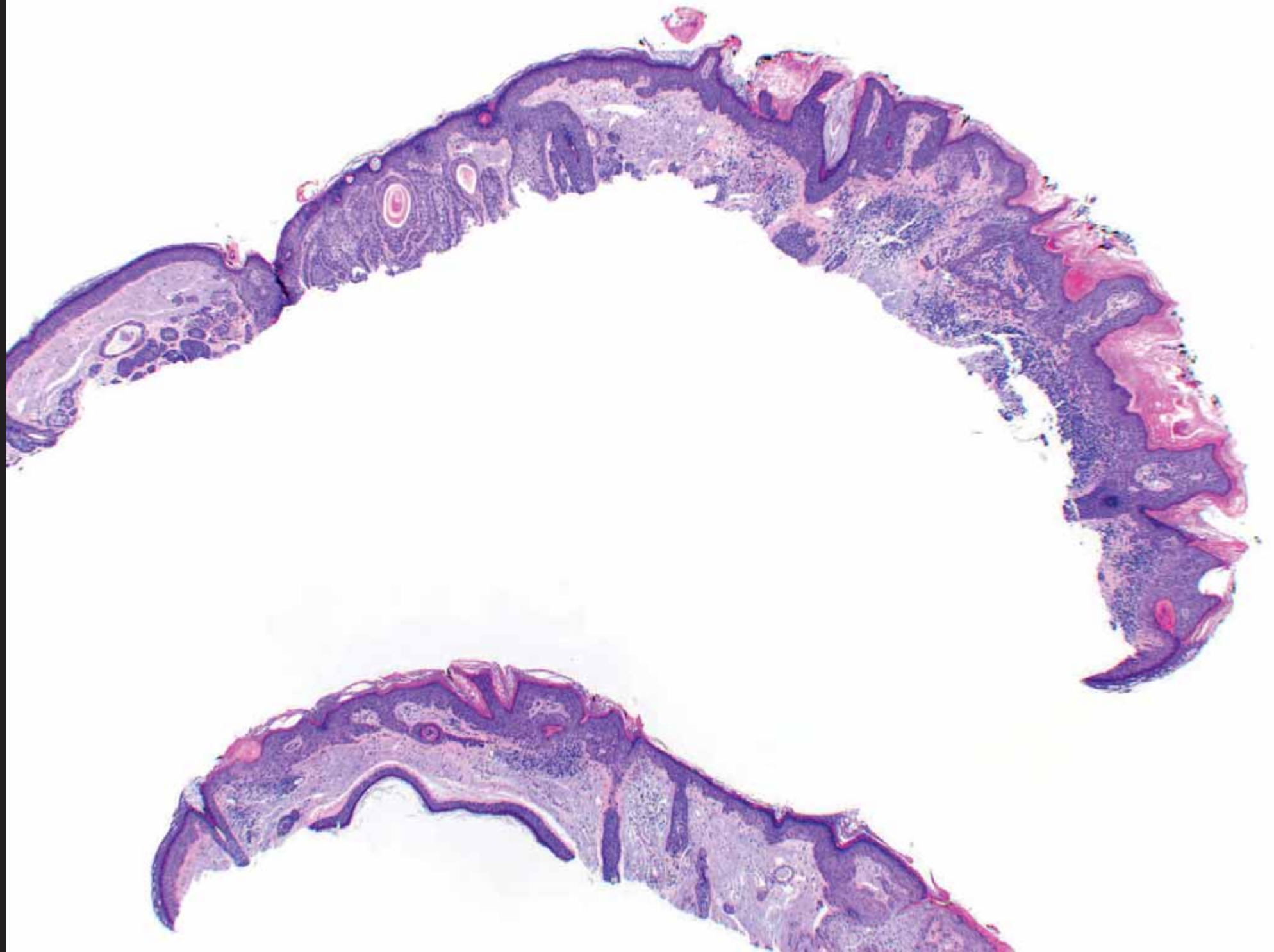
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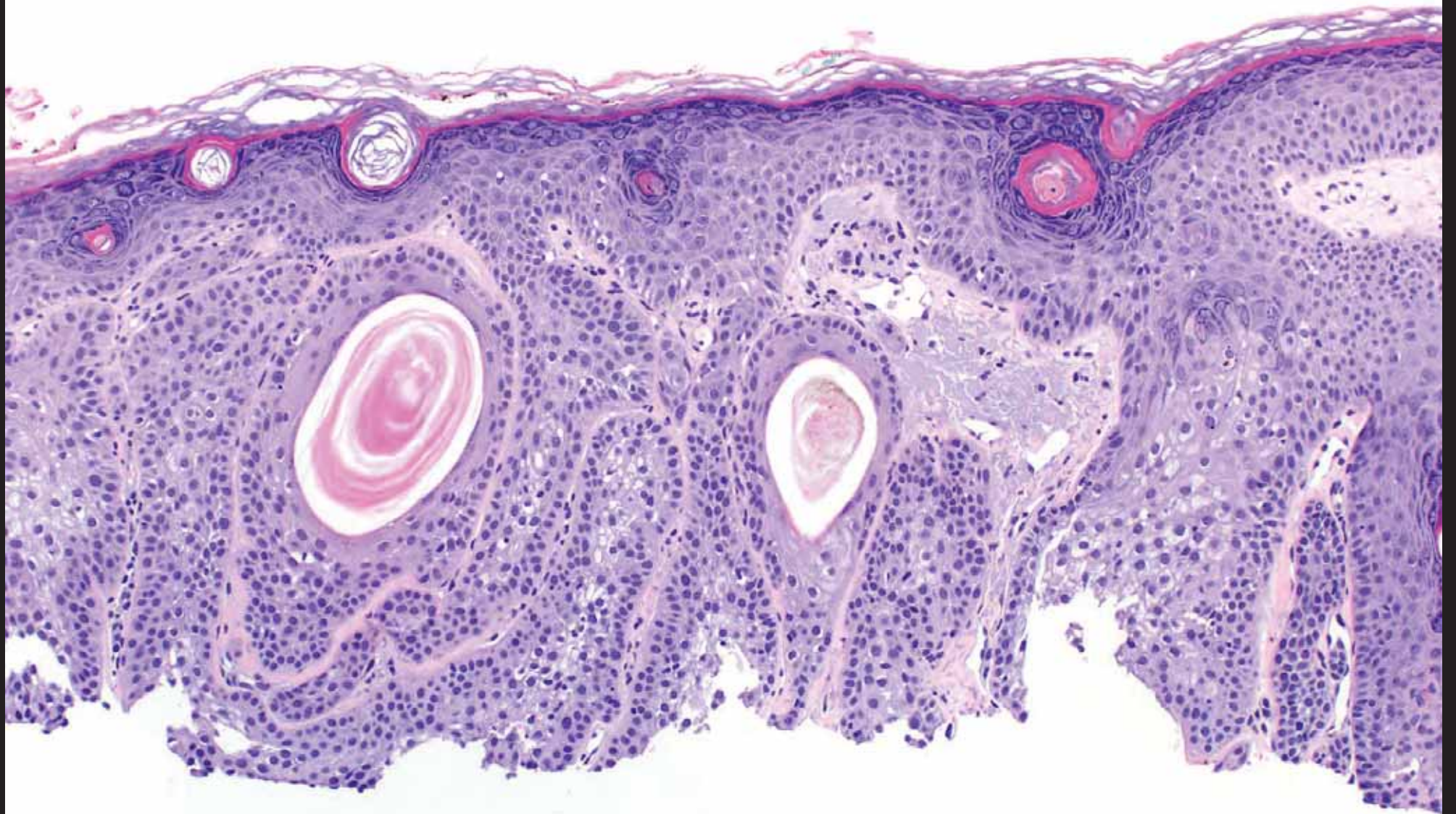
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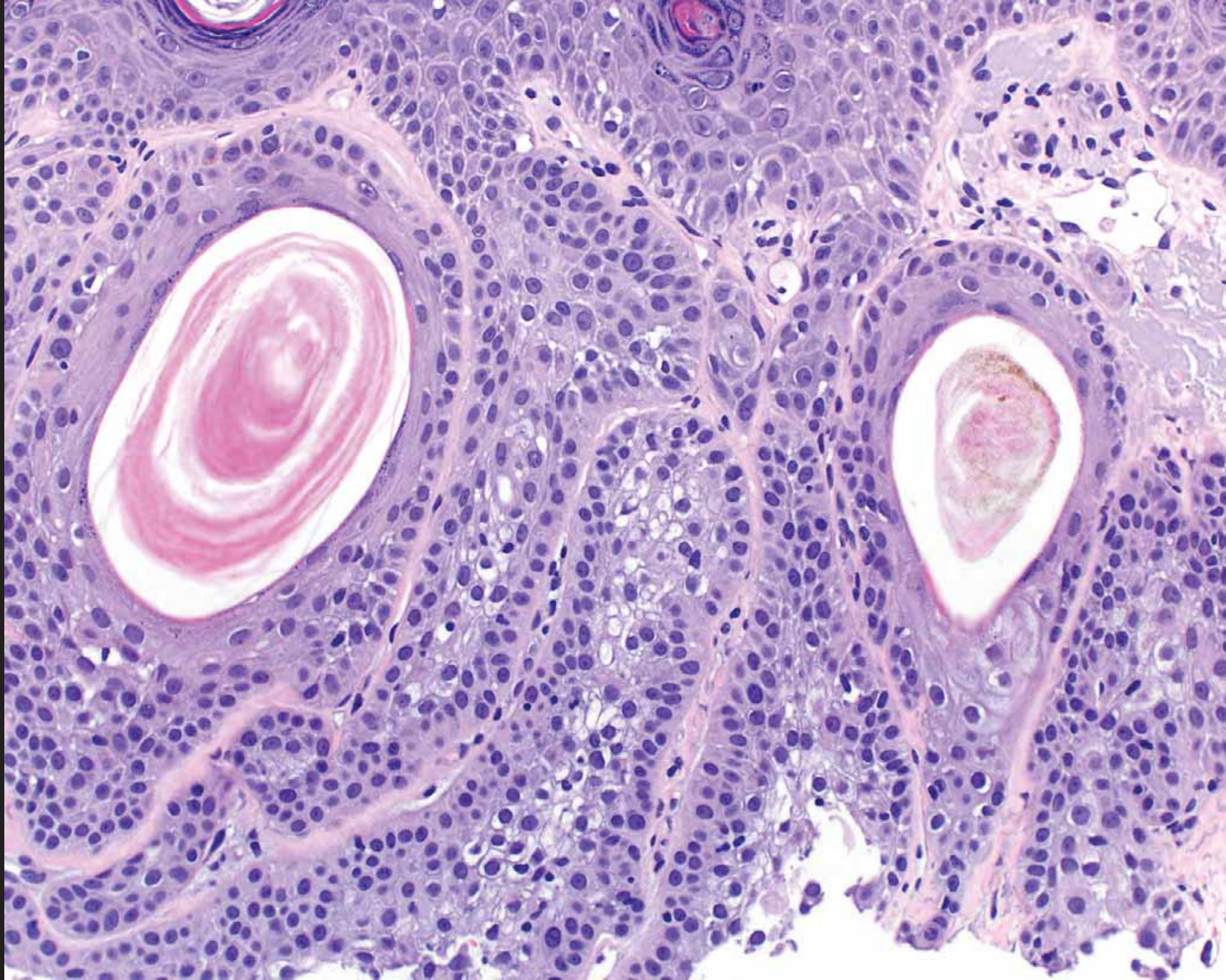
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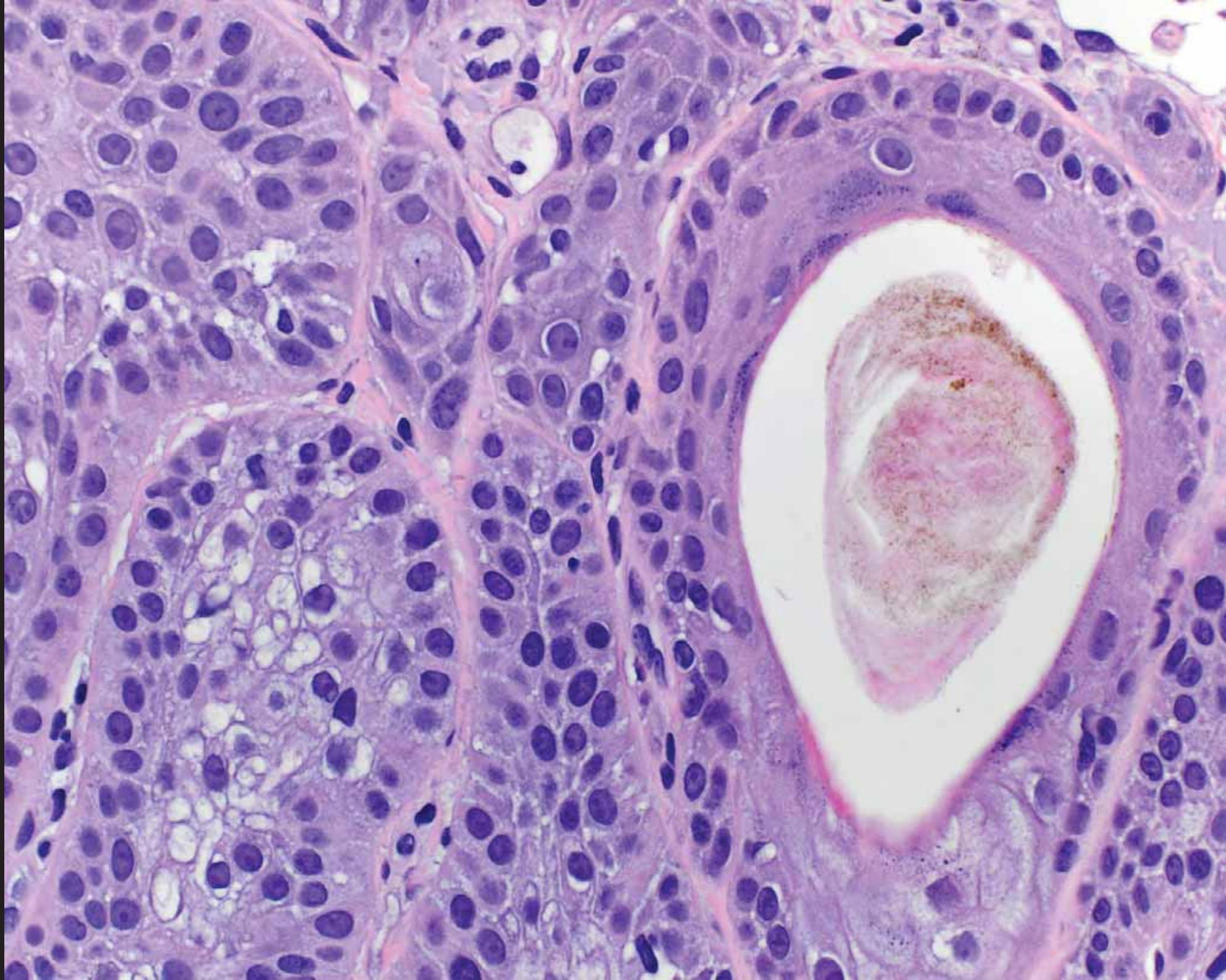
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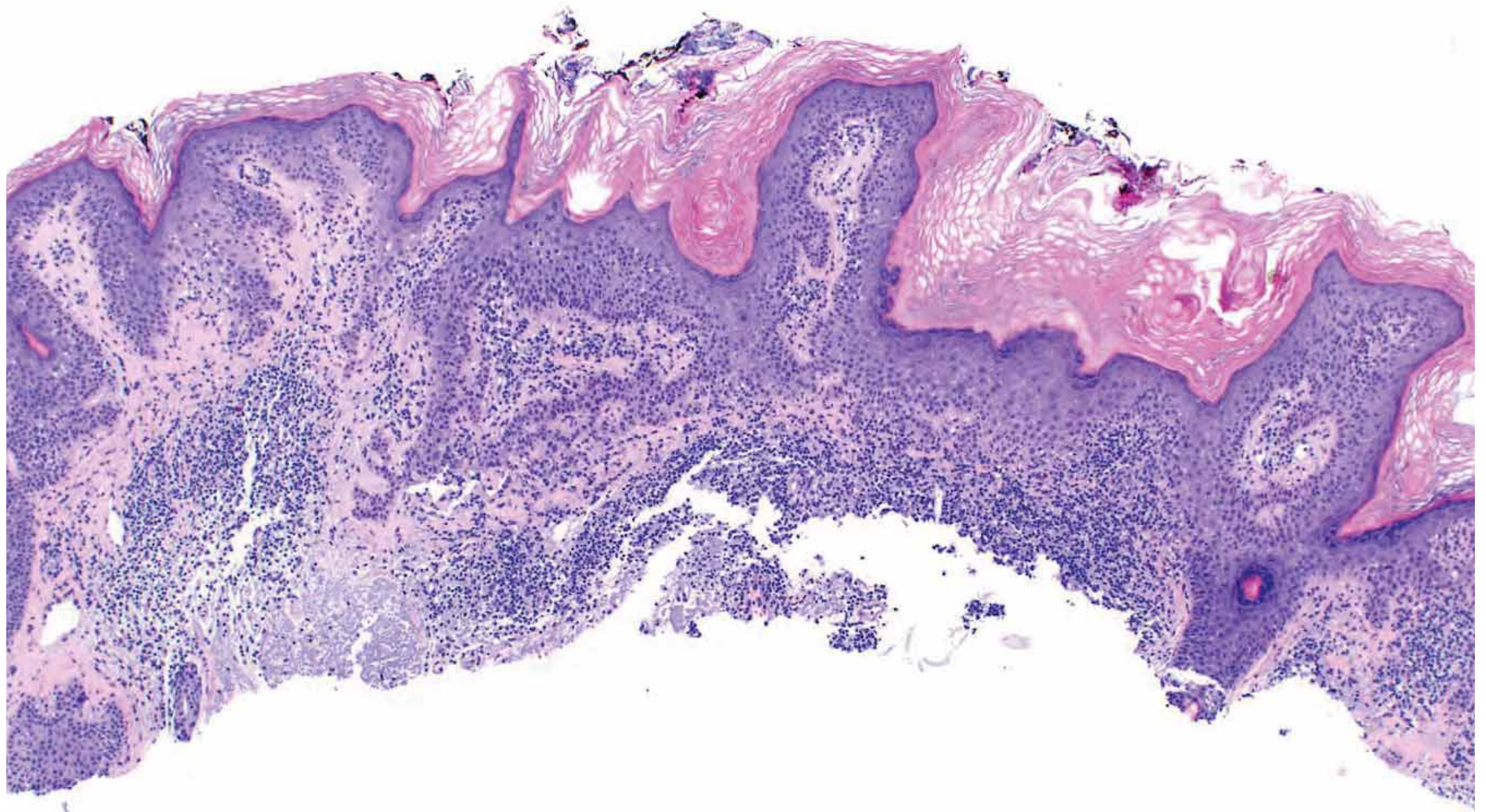


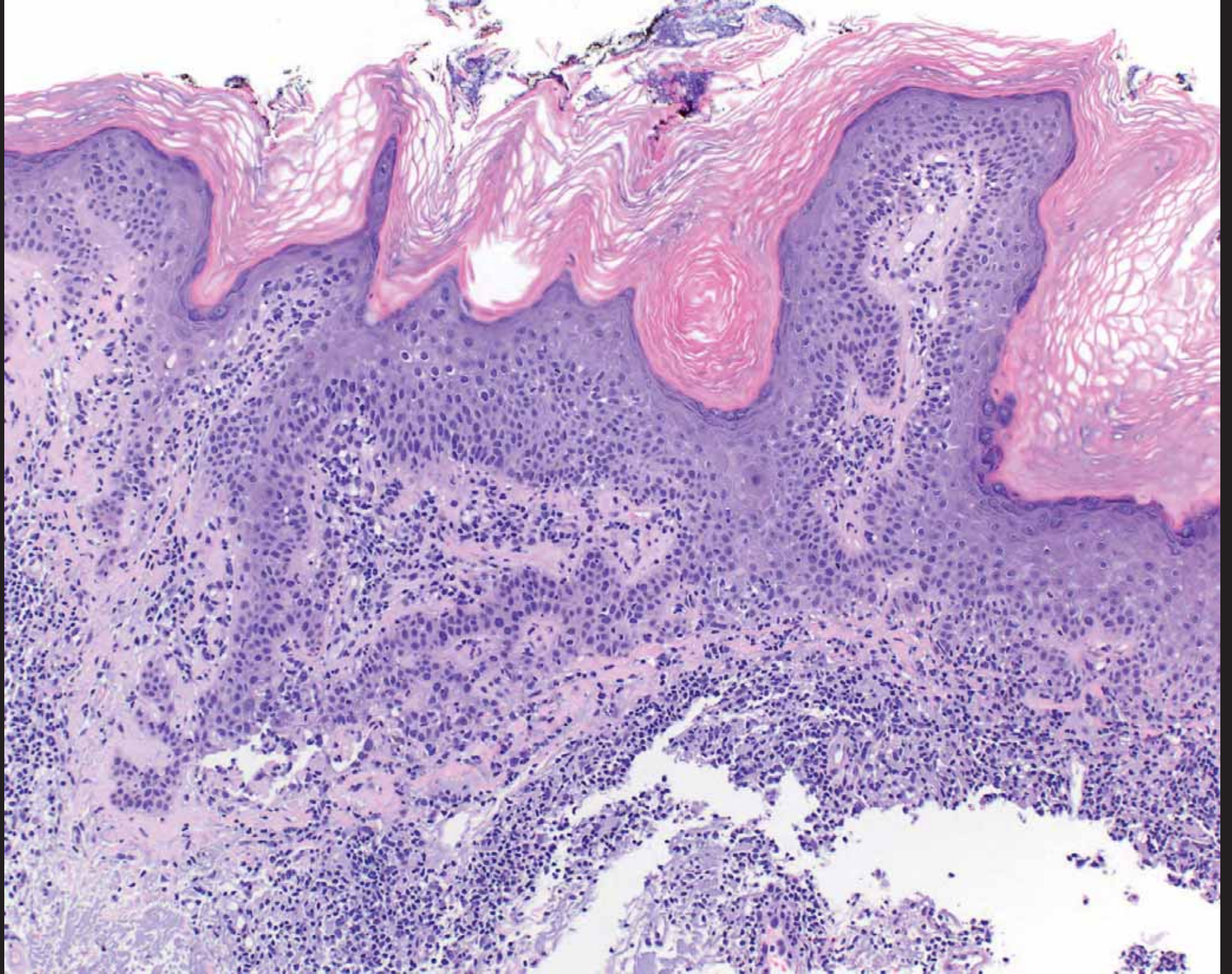


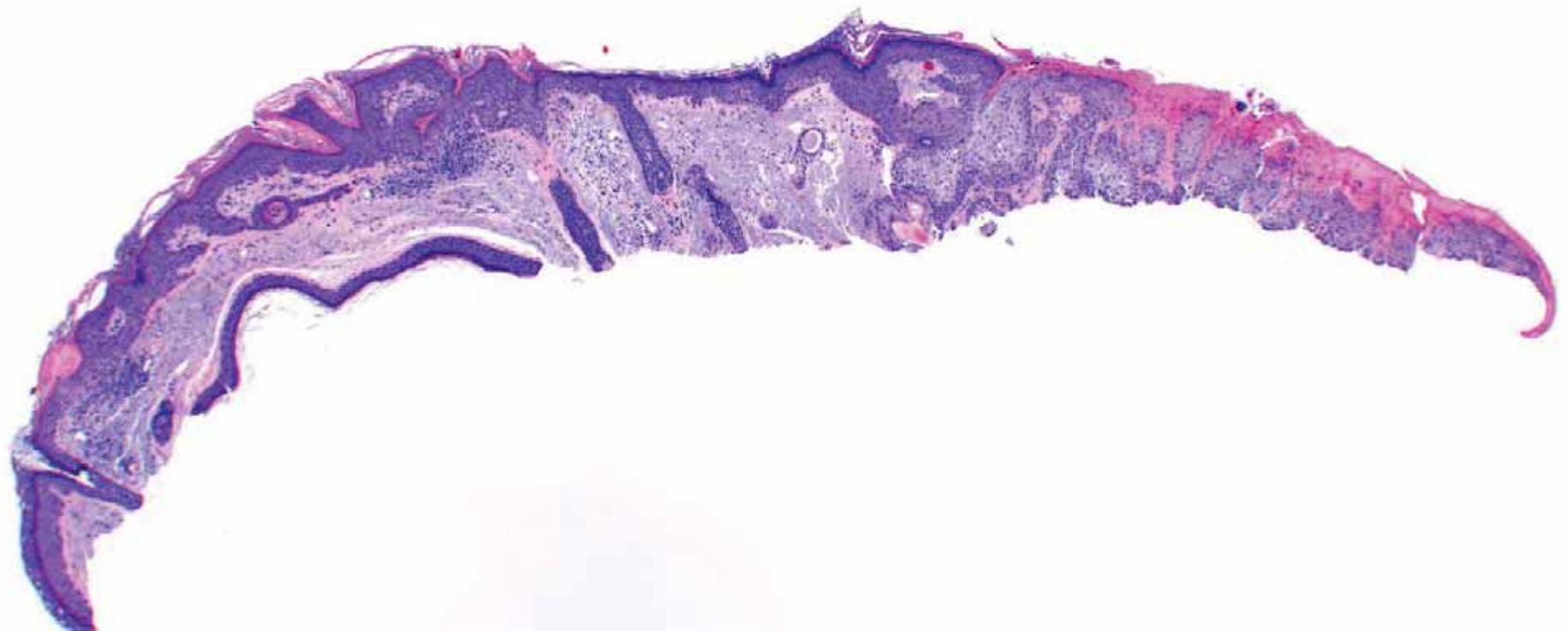


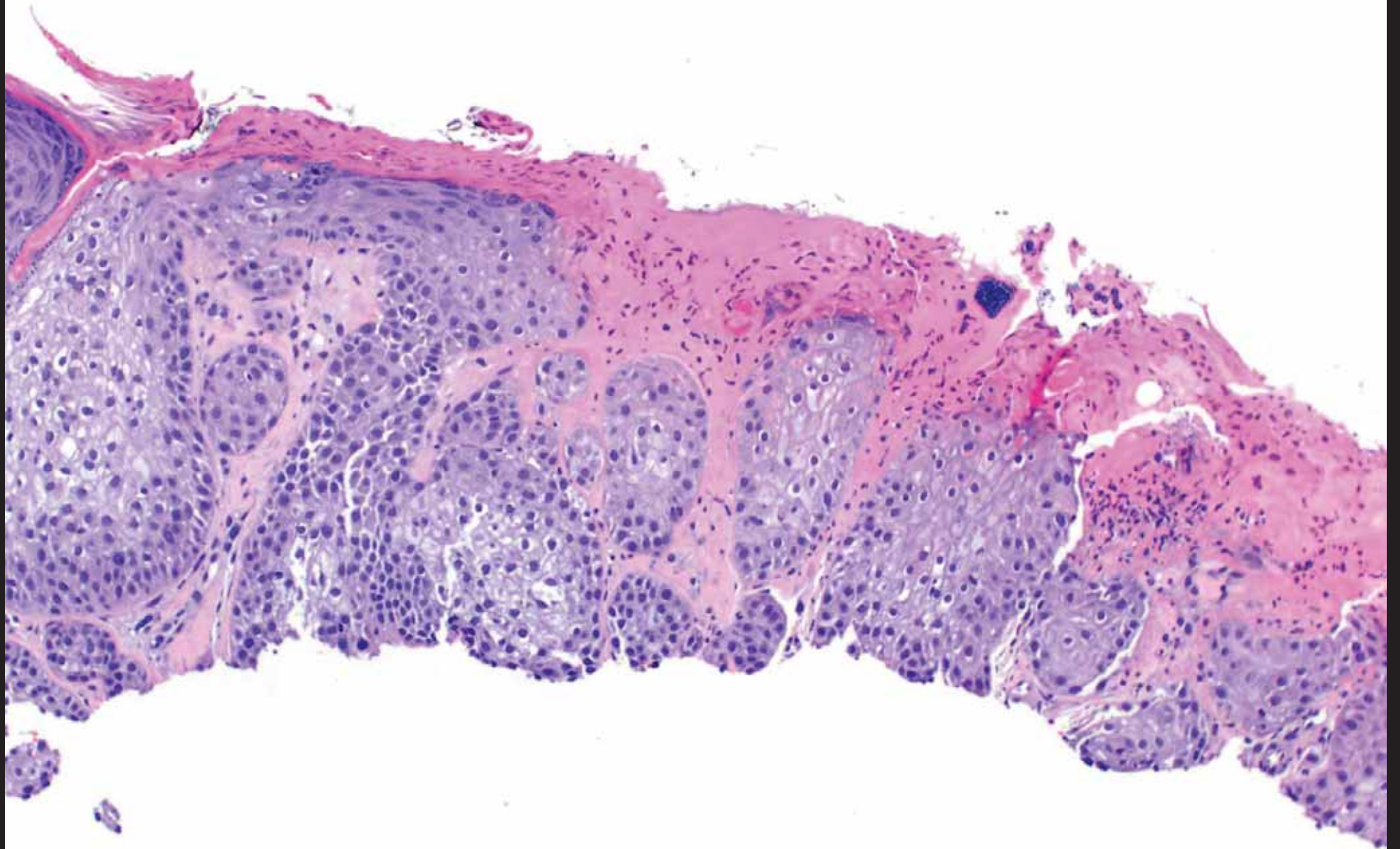


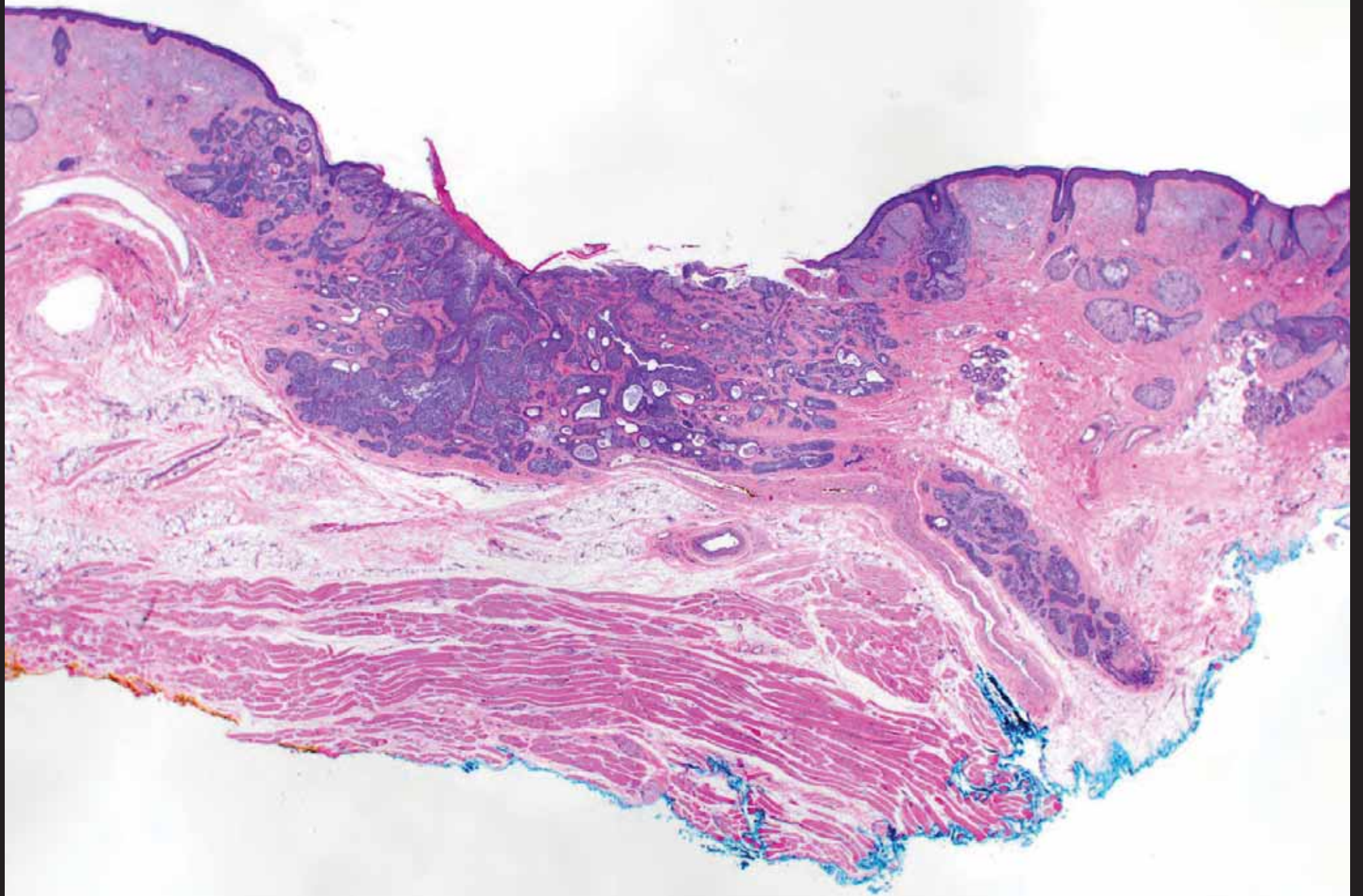


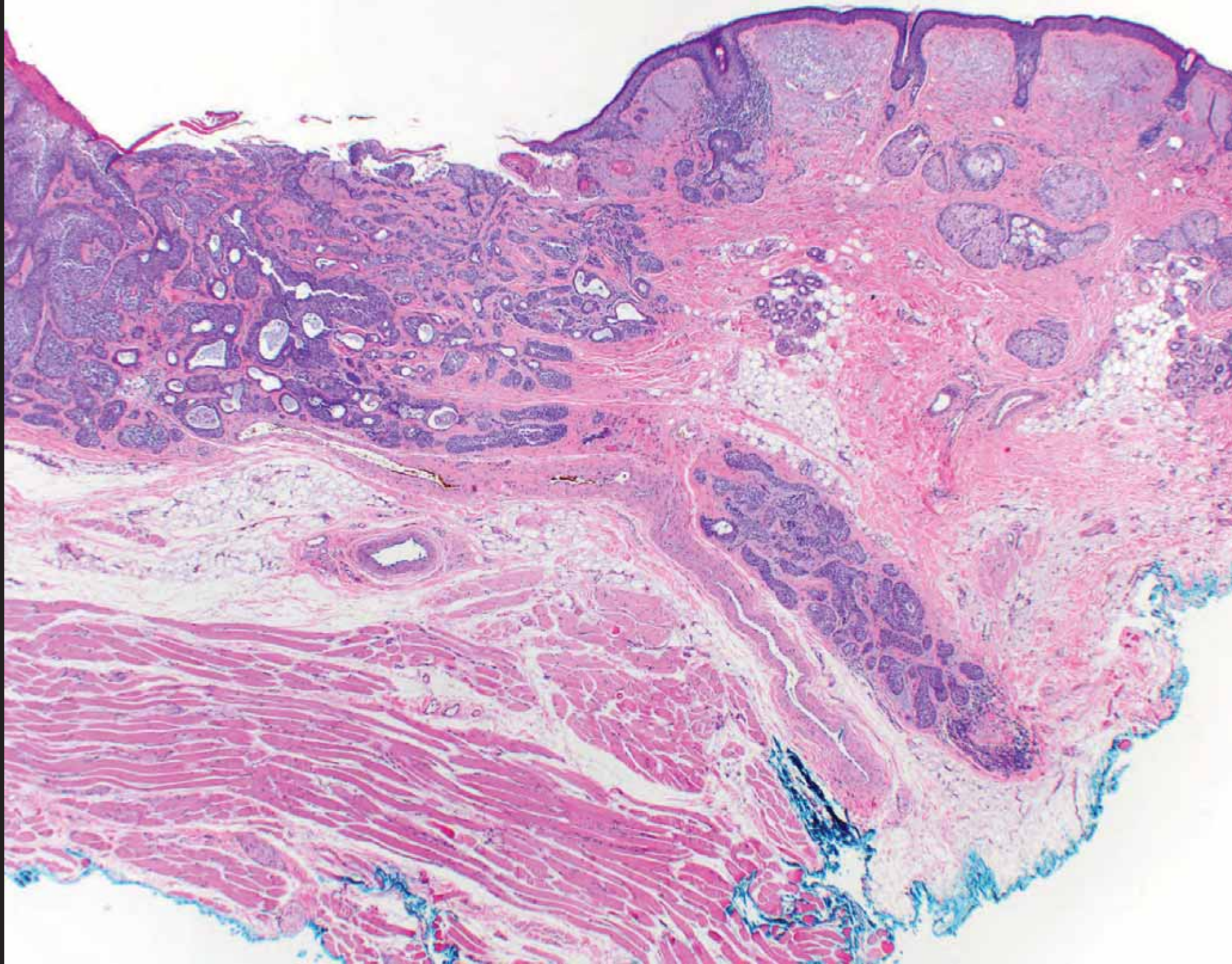


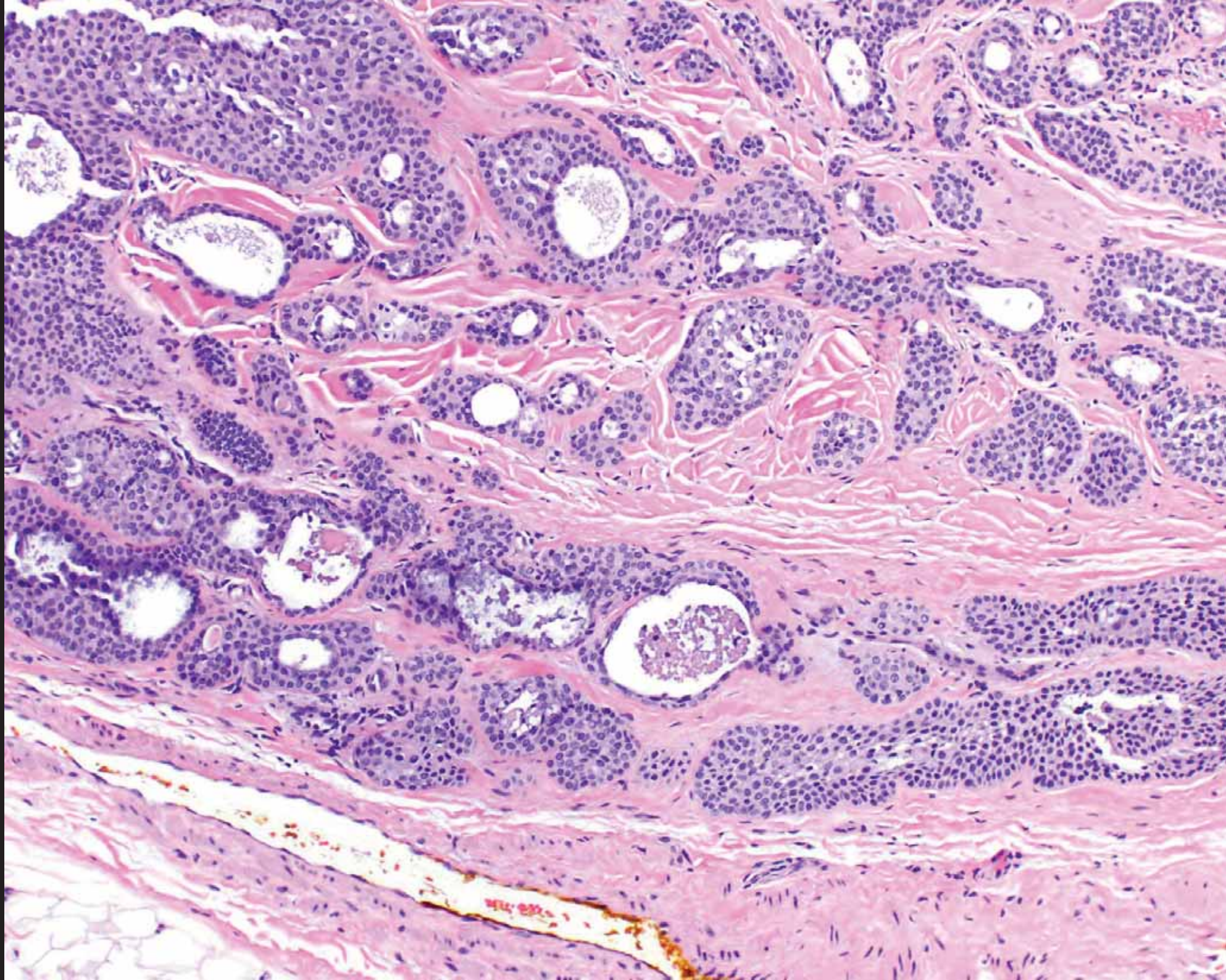


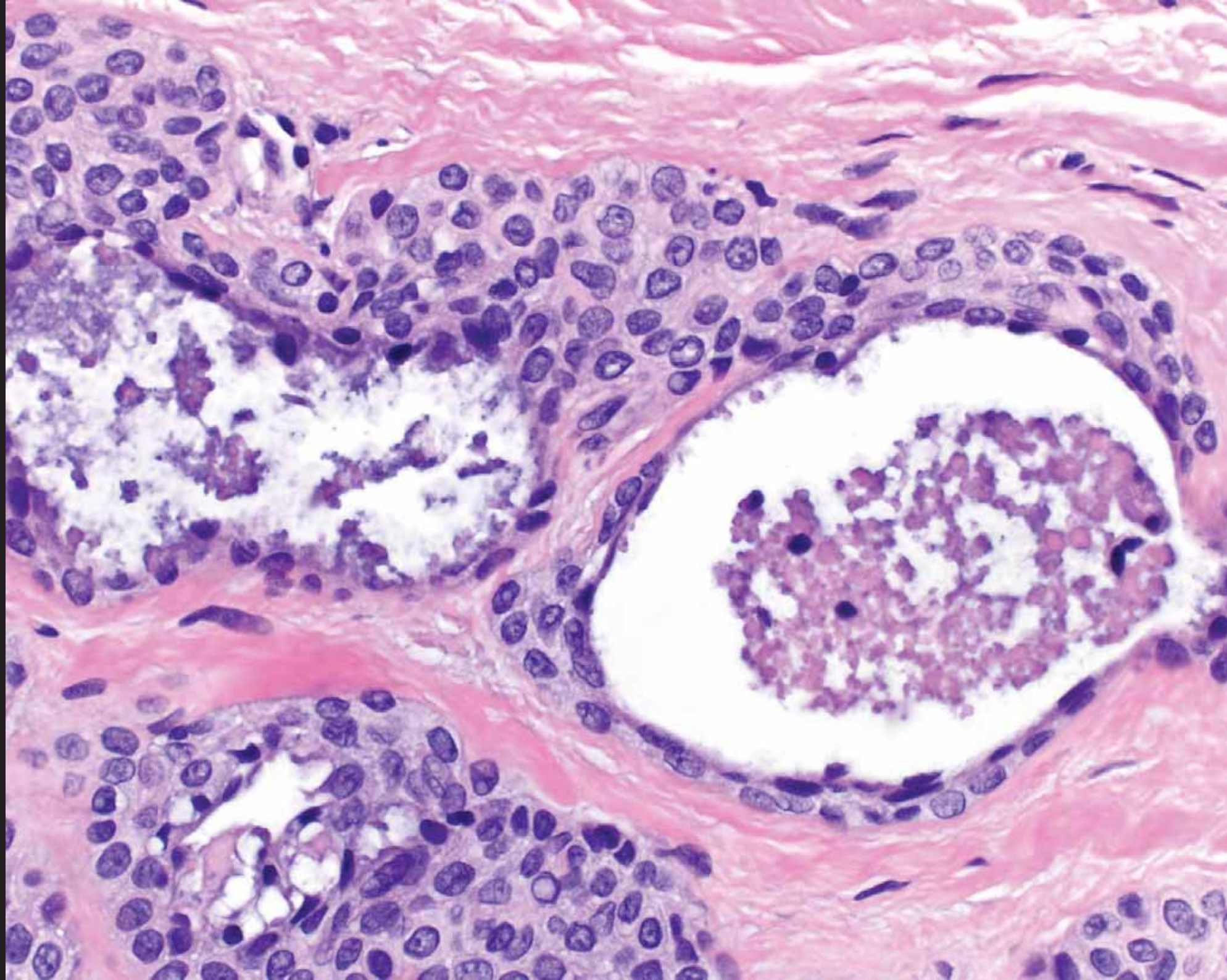


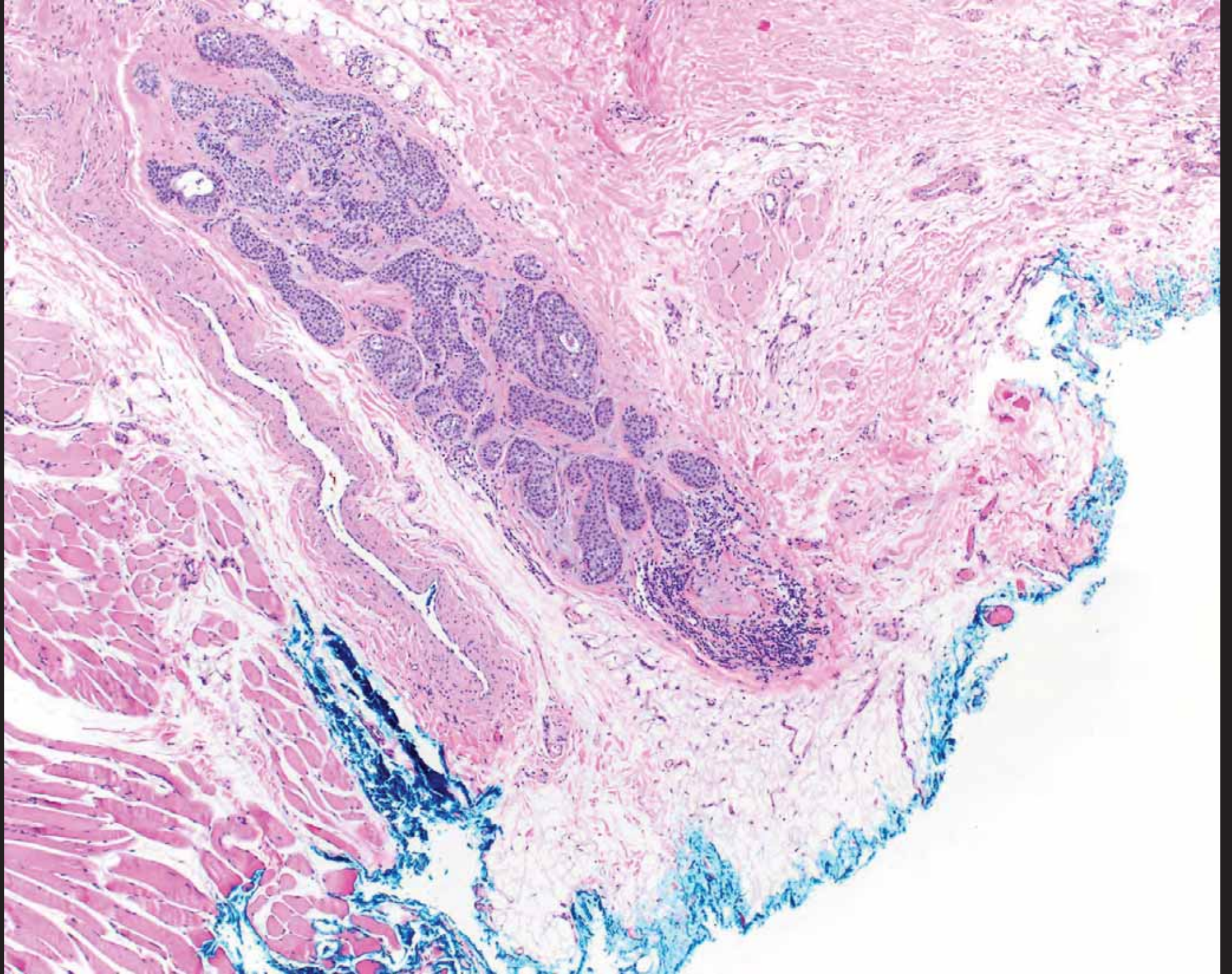


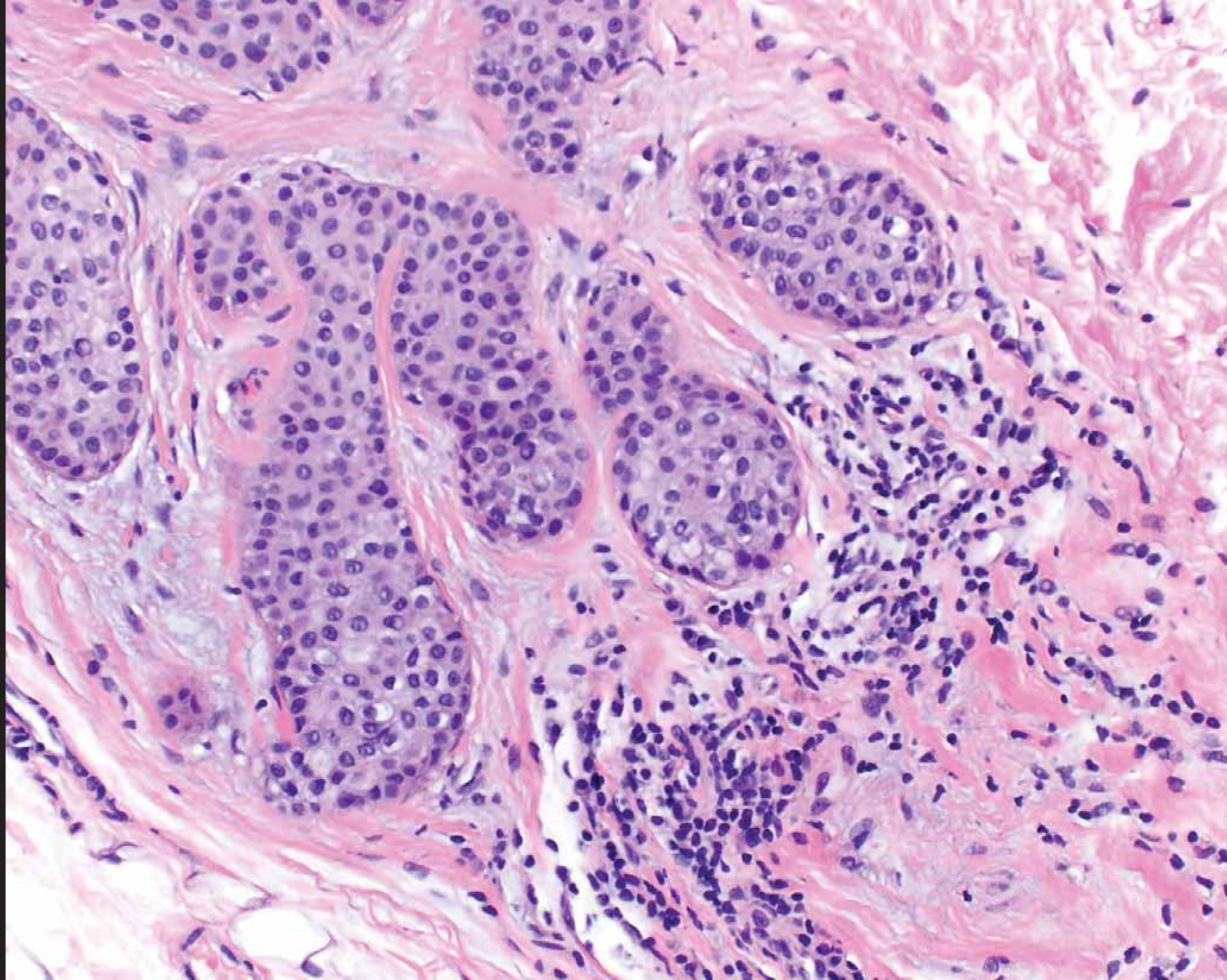










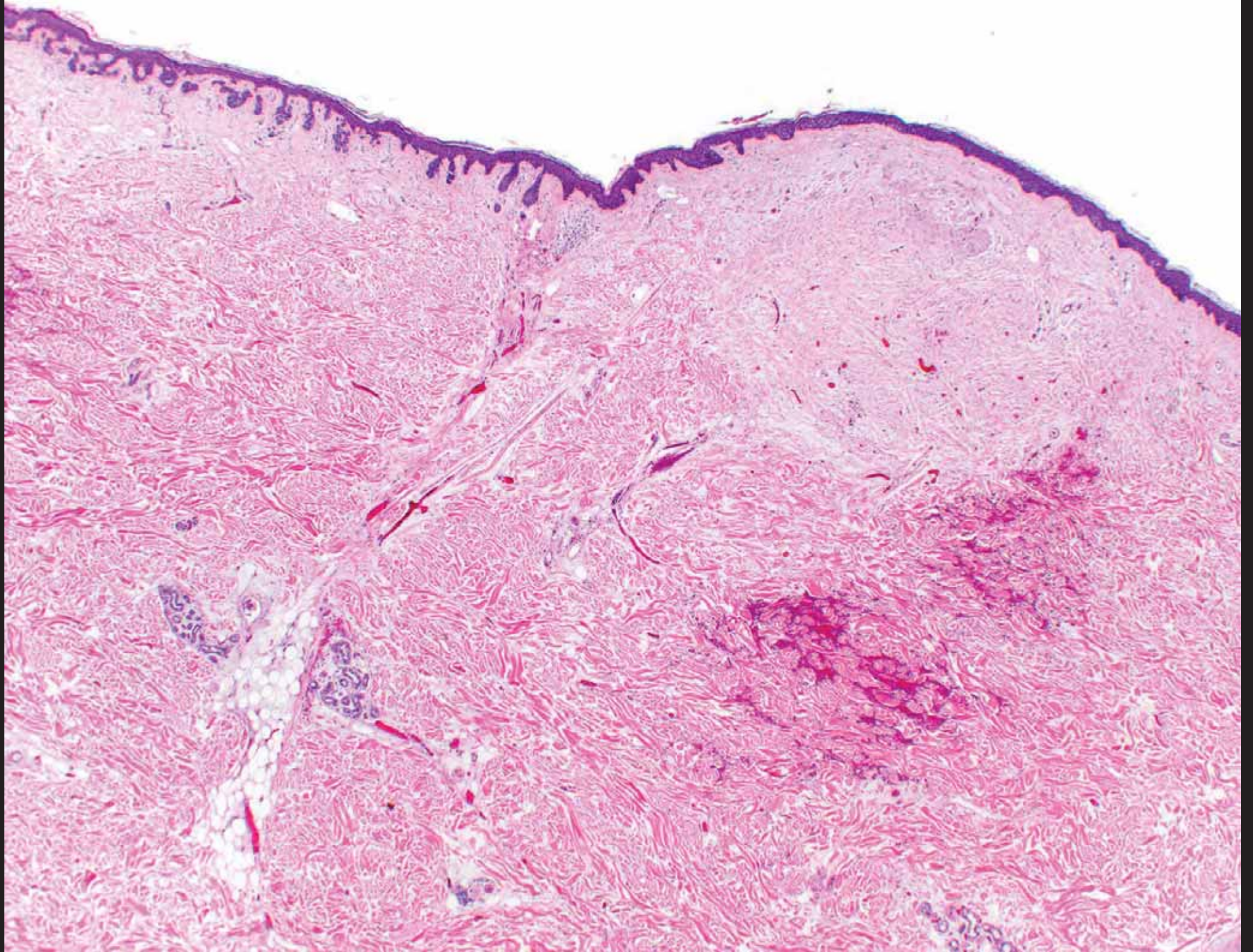


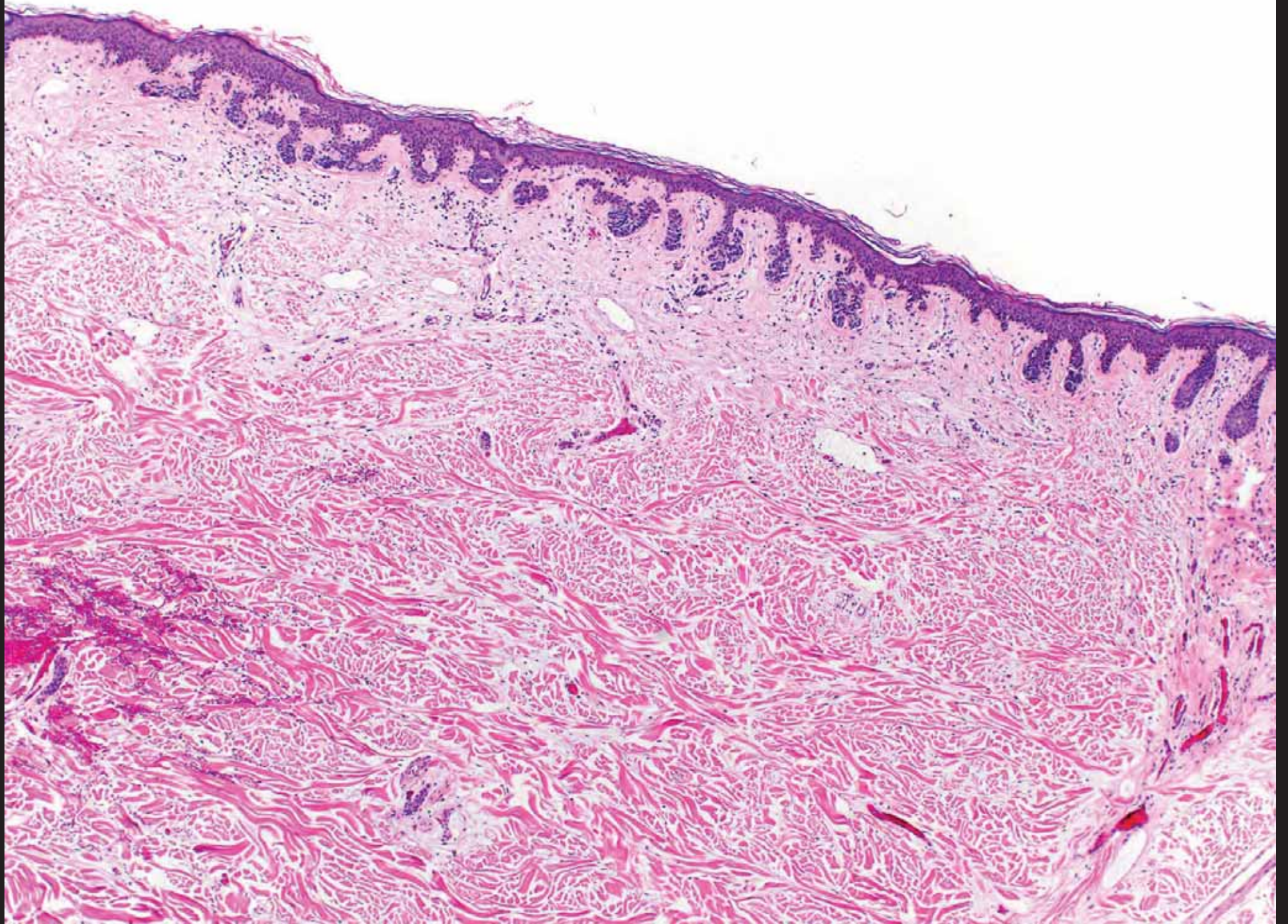
Does the ddx between trichilemmoma and microcystic adnexal CA with clear cells matter?

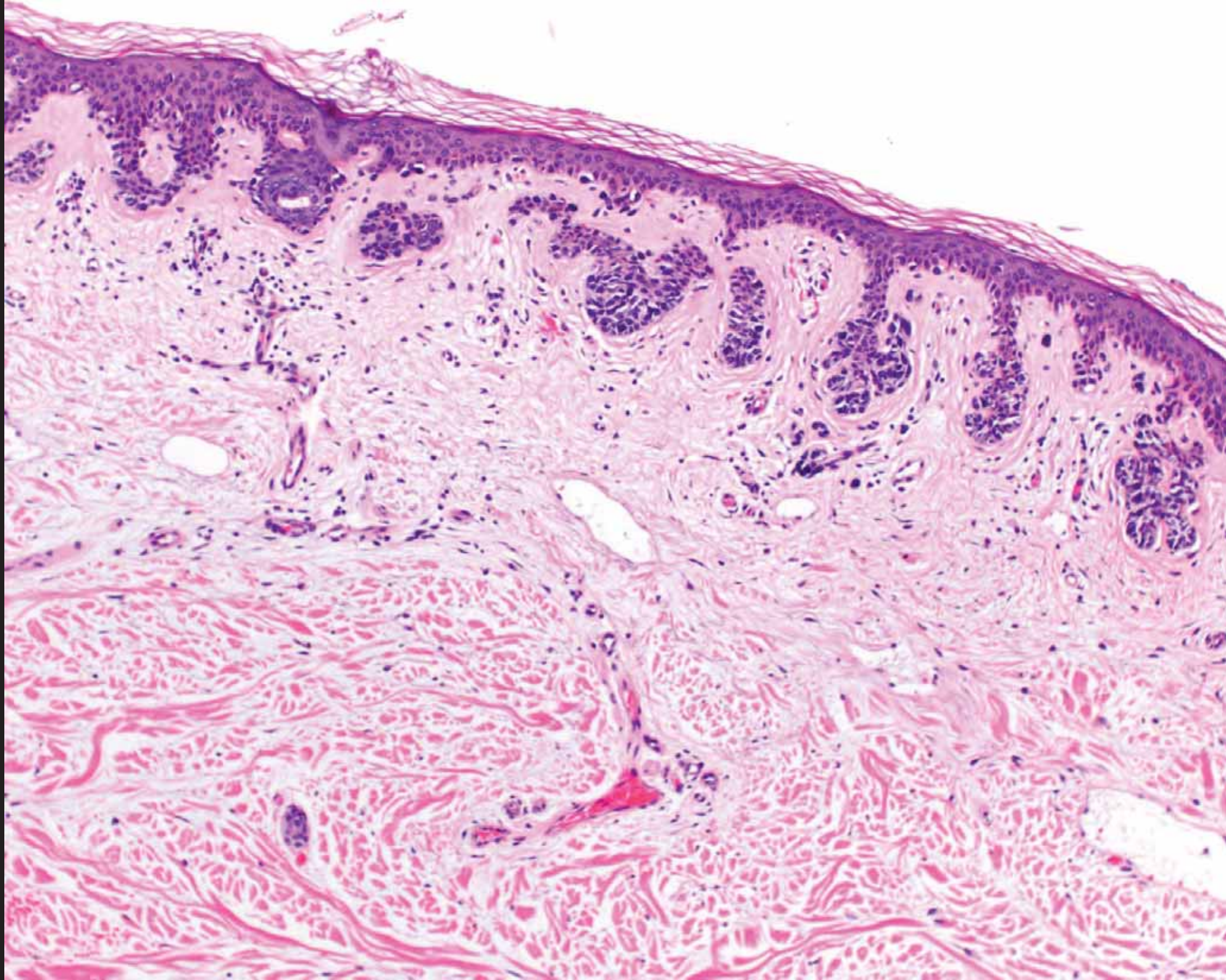
- Yes! For the first, no further treatment, for the second, clear margins
- Needs assessment- If my experienced colleague could call the initial biopsy trichilemmoma, we need better tools¹
- Small superficial bxs of adnexal tumors on the face are more common than good bxs of them

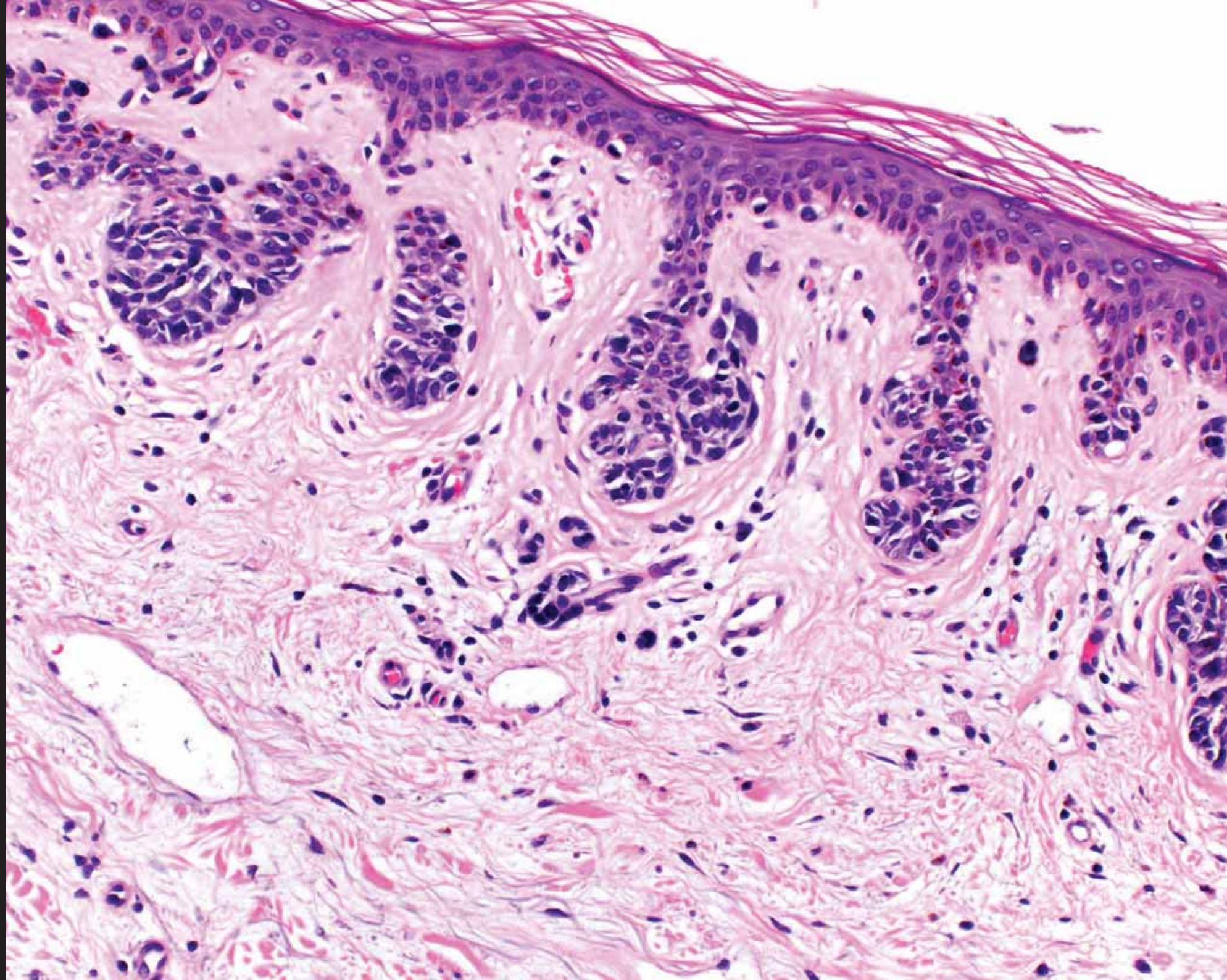
Case 4

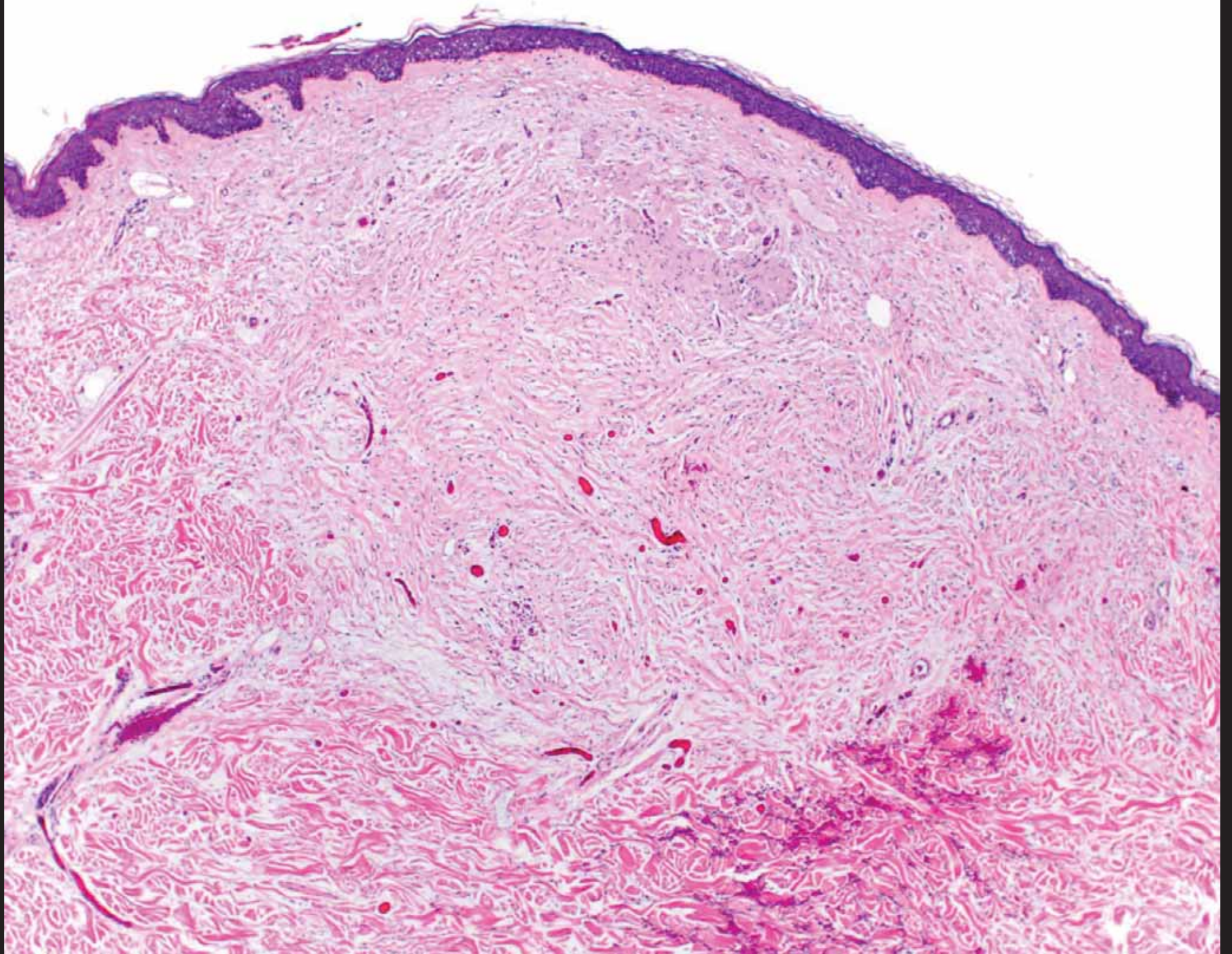
67 year old woman, rule out melanoma

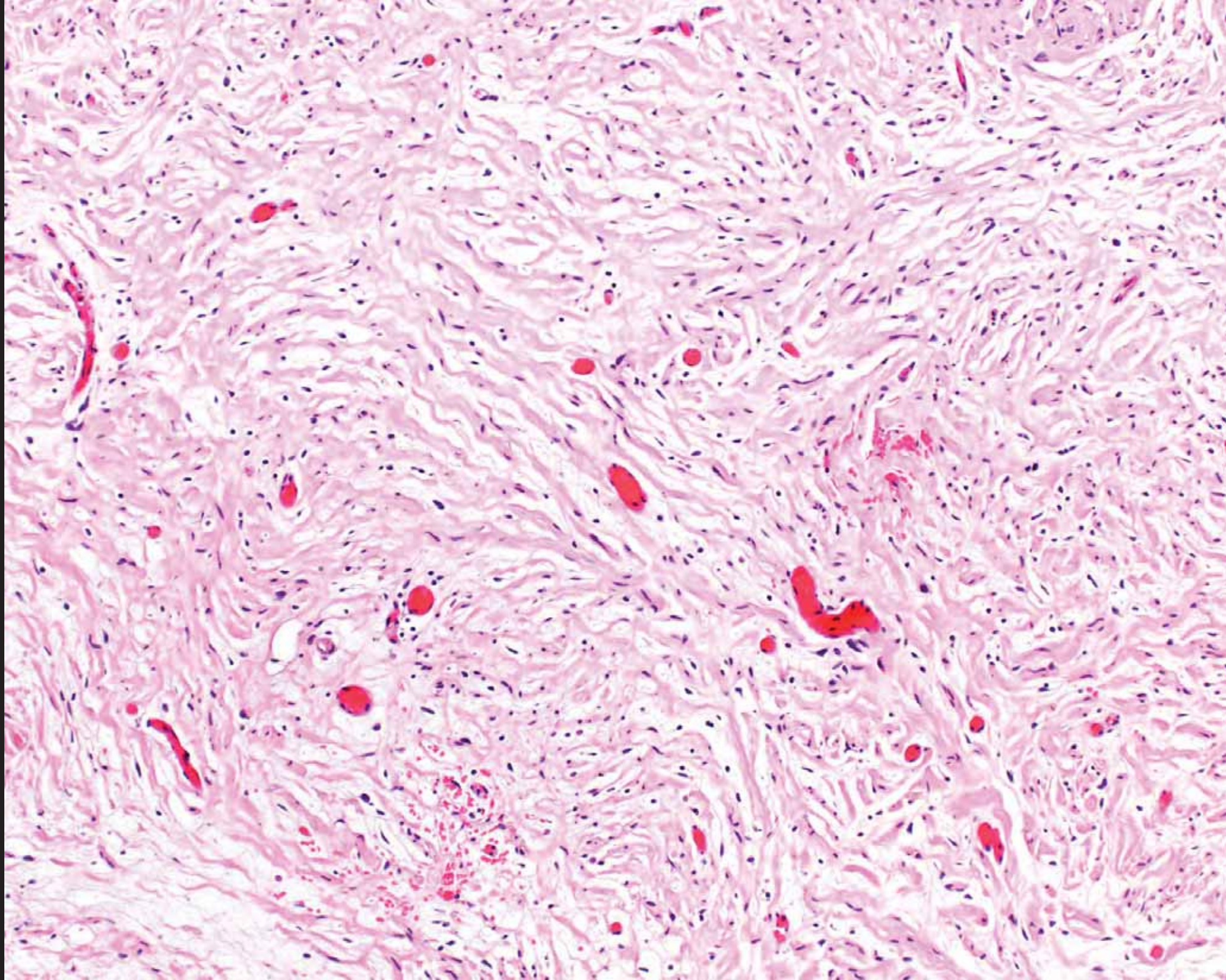


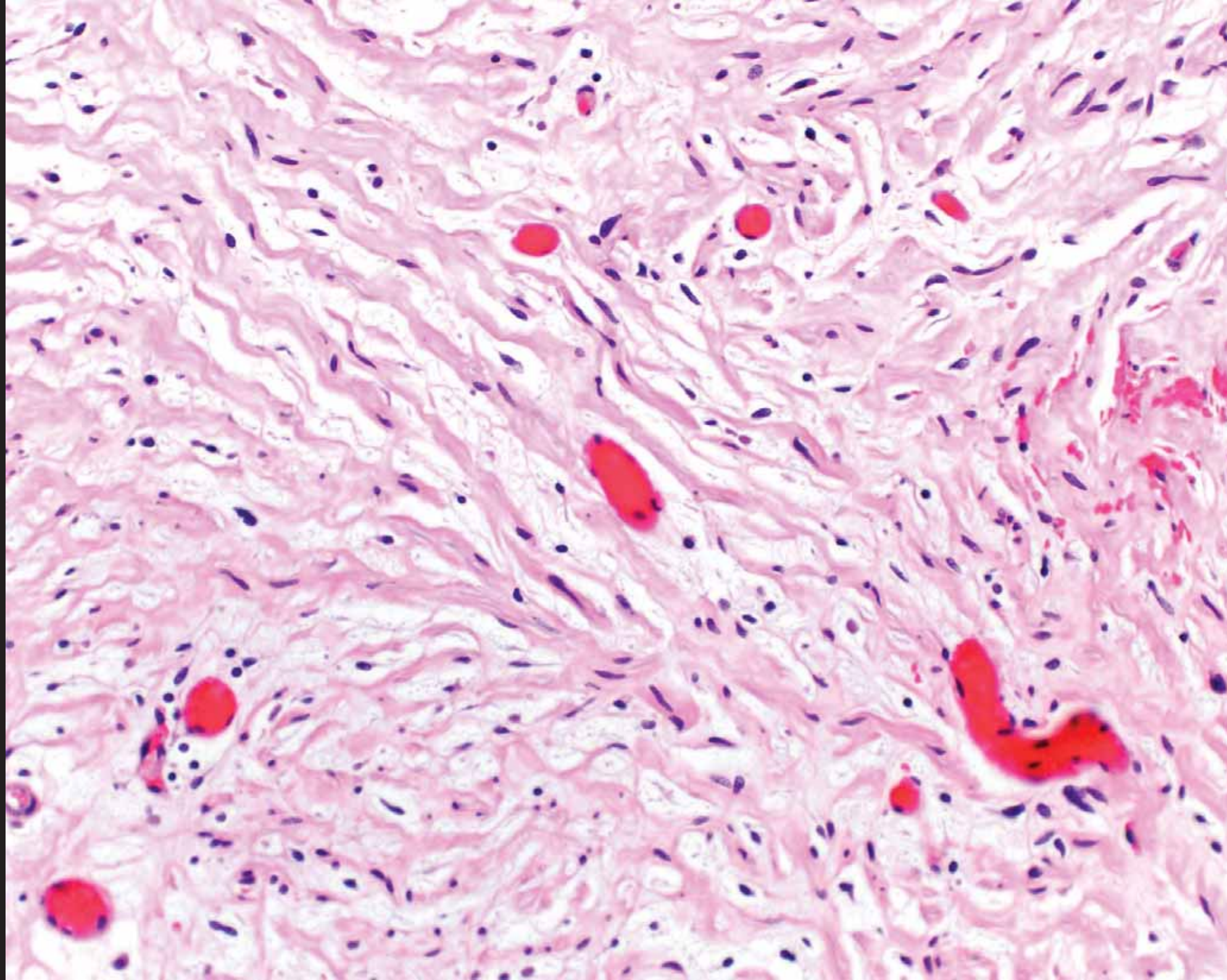


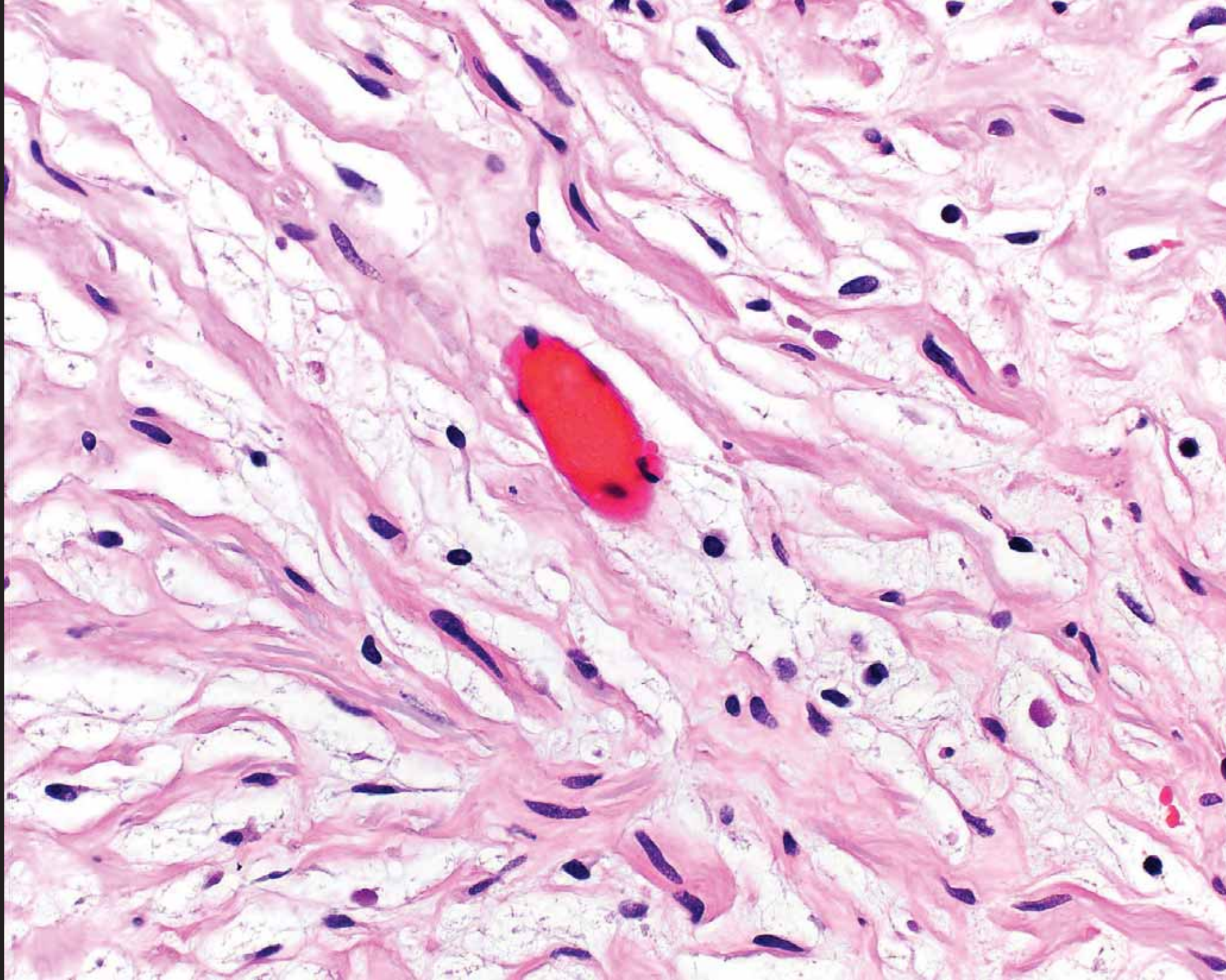


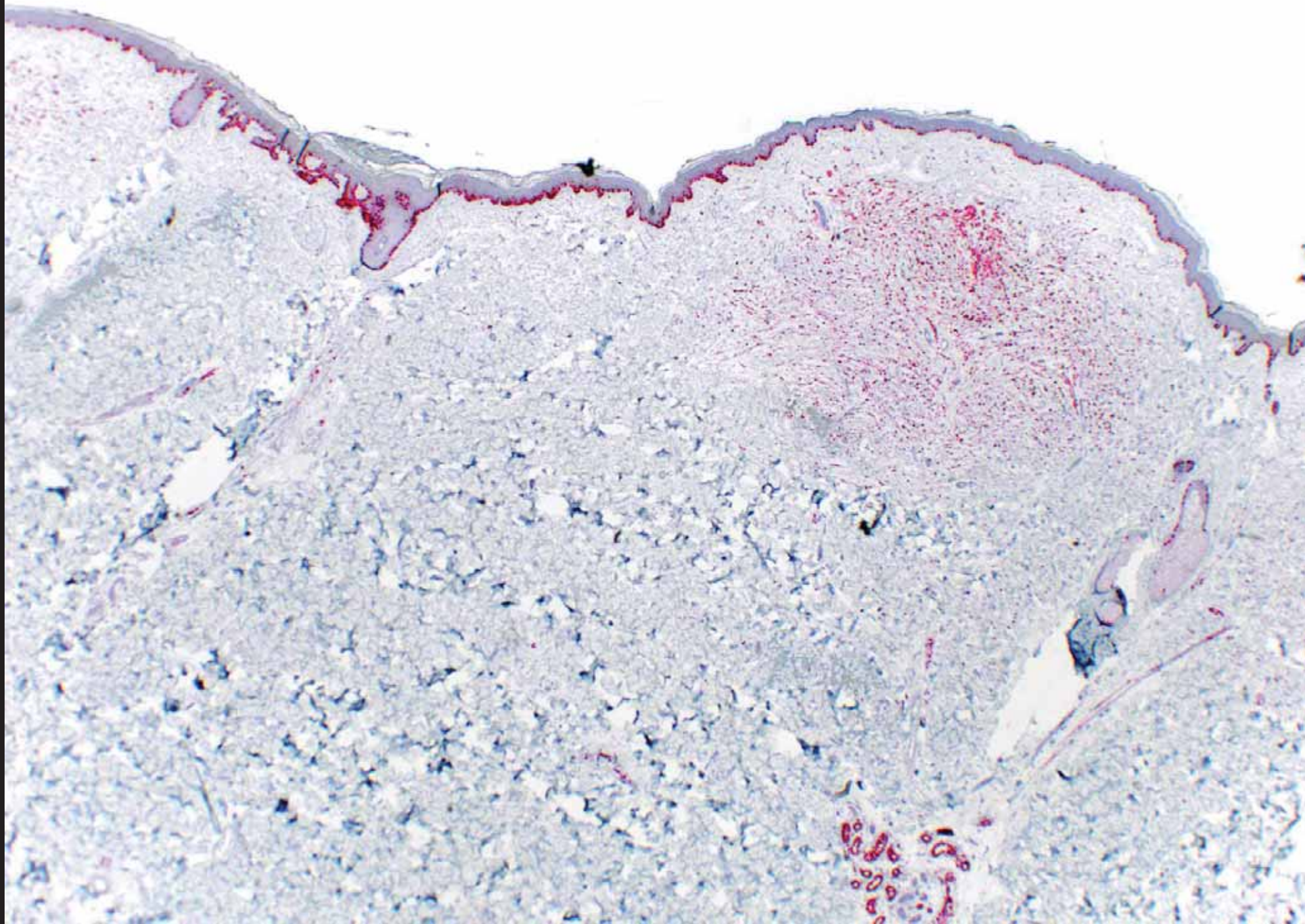




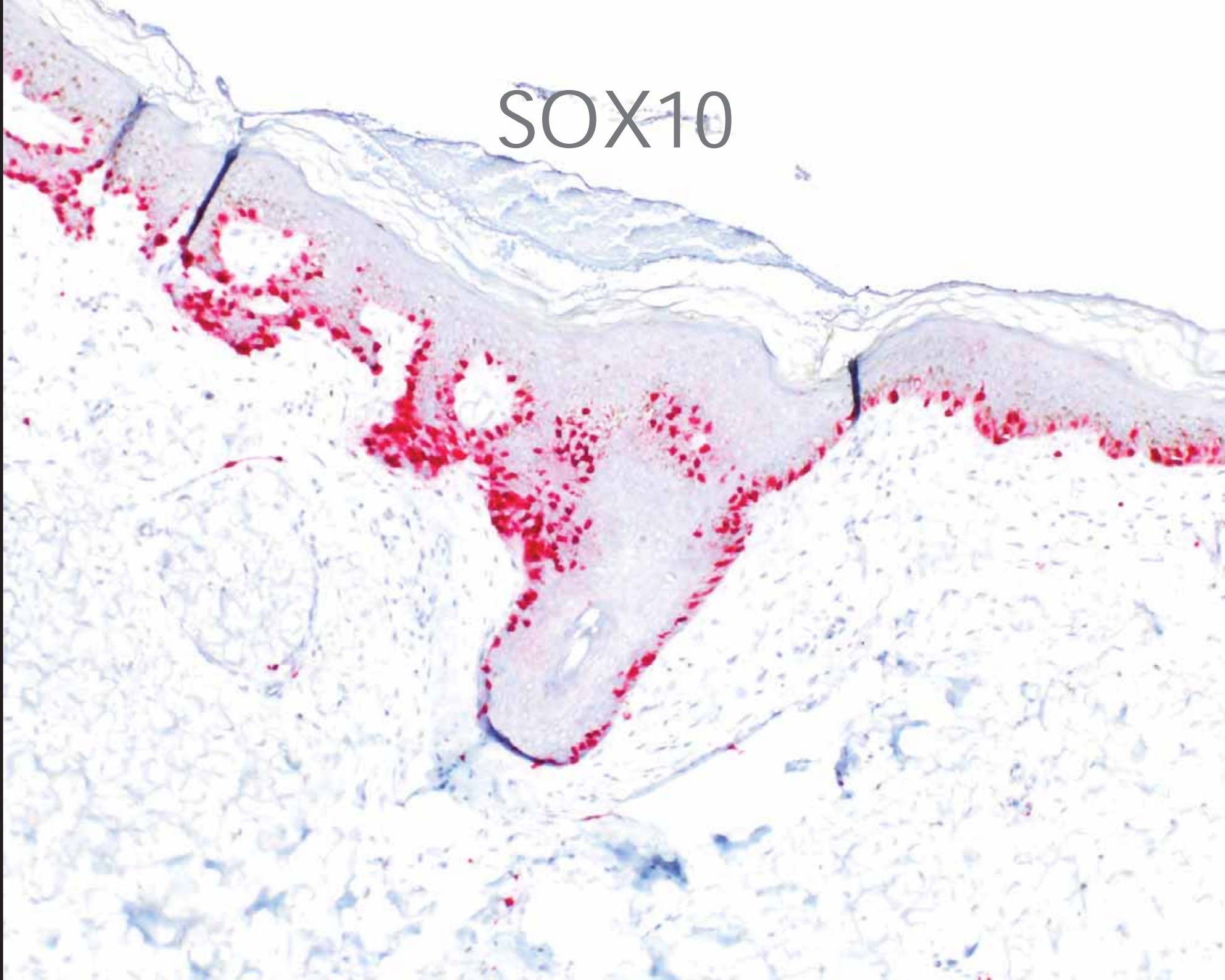




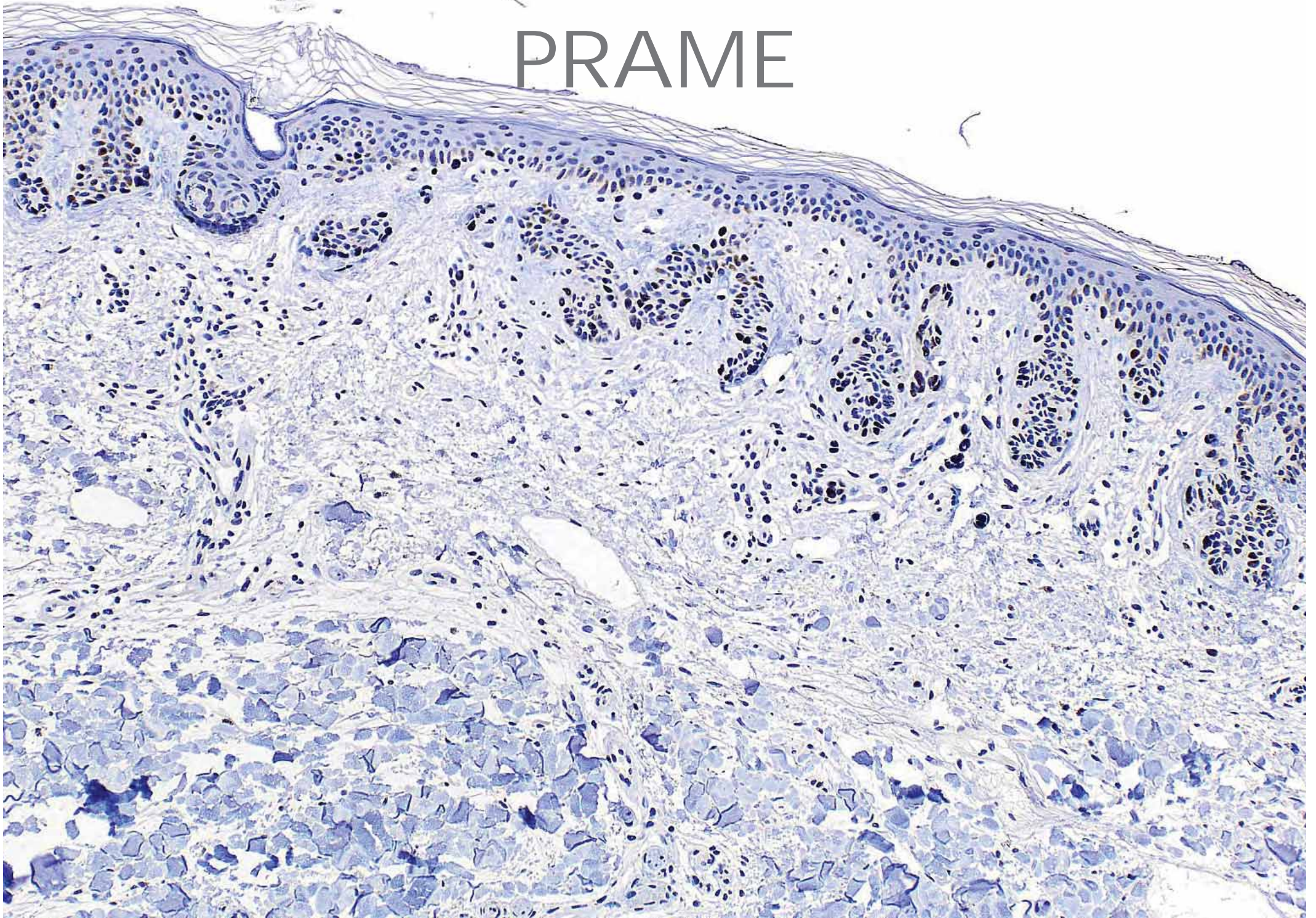


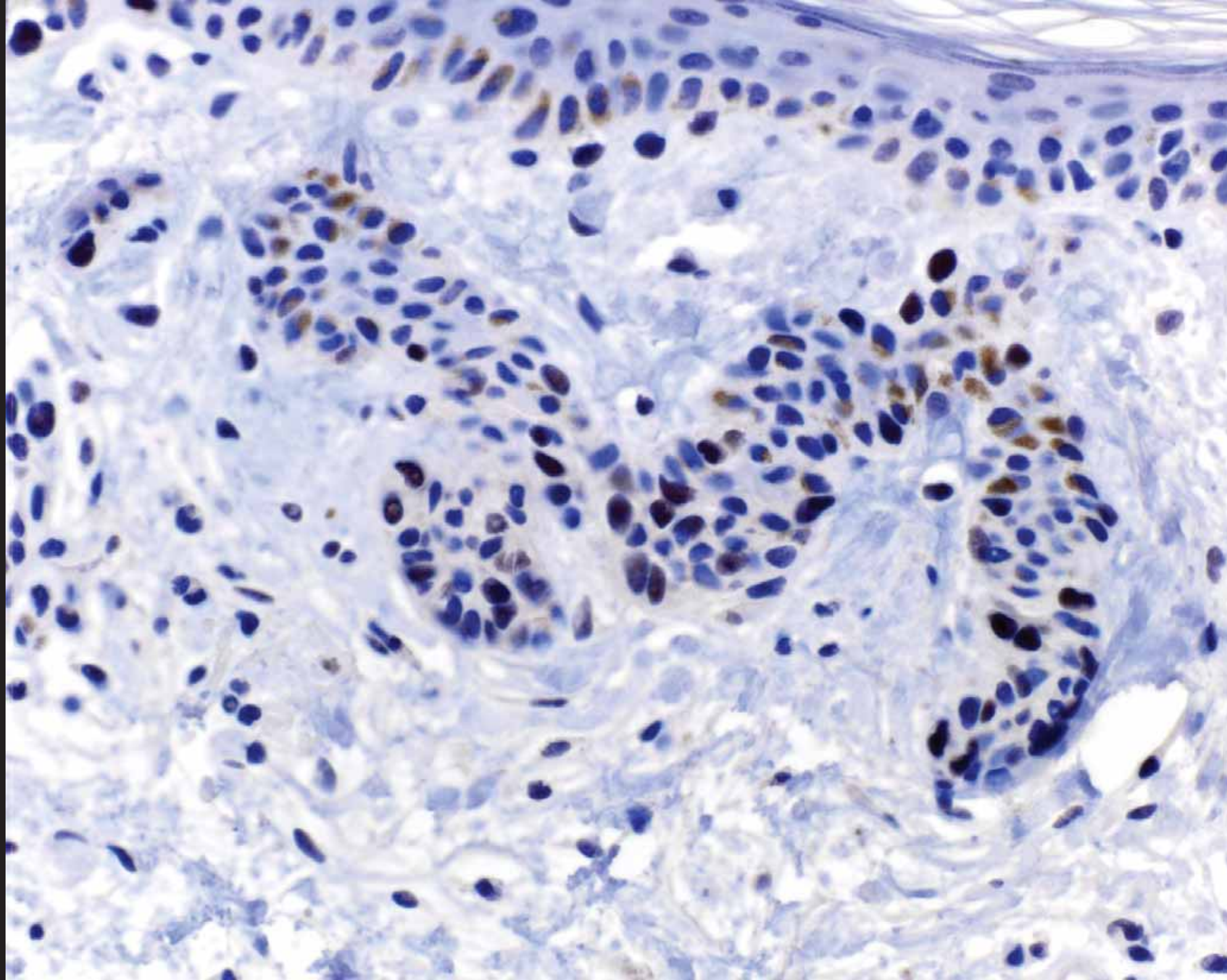


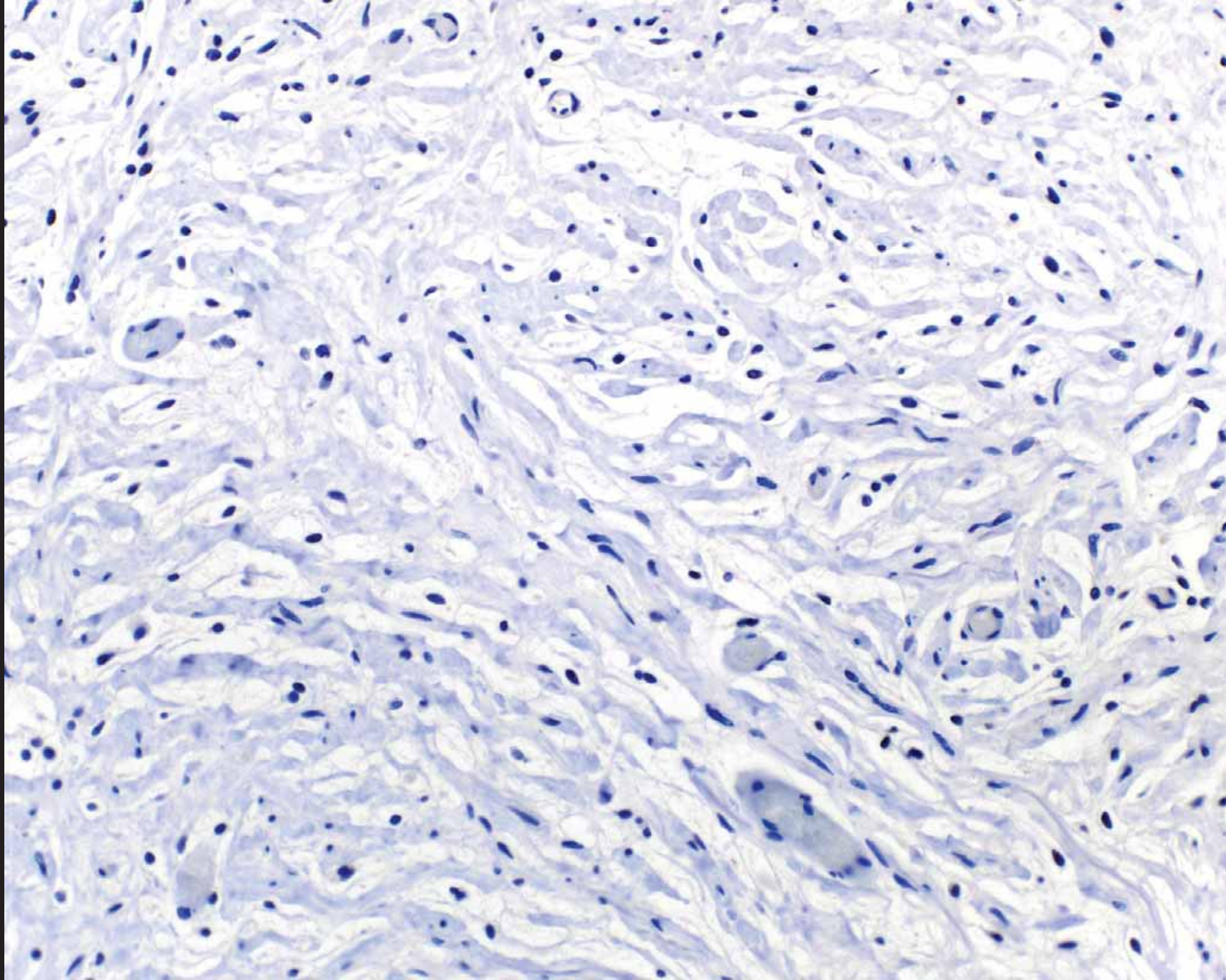
SOX10



PRAME







Neurofibroma-like desmoplastic melanoma

- Can have overlying melanoma in situ
- Dermal part resembles neurofibroma
- Clues said to include fibroplasia, poor circumscription, lymphoid aggregates, diffuse subcutaneous involvement
- No IHC markers reliably separate NF from NF-like DMM; PRAME usually (-) in DMM

Desmoplastic Melanomas Mimicking Neurofibromas

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Ayesha U. Khan, MBA,* Pedram Yazdan, MD,* Joan Guitart, MD,* and Klaus Busam, MD‡*

Abstract: Desmoplastic melanoma can be difficult to diagnose and on average have a significantly higher T stage at the time of diagnosis compared with conventional melanomas. Histologically, these tumors typically consist of spindle cells in a fibrous matrix. The spindle cells may display fibroblast and/or Schwann cell–like features. In this study, we describe the features of 12 cases of desmoplastic melanoma closely simulating neurofibroma. Although the spindle cells in these tumors may be indistinguishable from those of neurofibroma, features such as prominent fibroplasia (12/12), poor lateral circumscription (8/9), diffuse infiltration of subcutaneous tissue (7/9), and lymphoid aggregates (10/12) may be helpful clues to the diagnosis. No immunohistochemical markers were reliable in distinguishing neurofibroma–like desmoplastic melanomas from neurofibroma. Clinical follow-up was available in 8 cases, of which 4 were initially misdiagnosed as benign neoplasms and given no further re-excision. All 4 of these cases recurred; 2 of which showed transformation to a more aggressive phenotype.

Key Words: desmoplastic melanoma, neurofibroma, melanoma

(Am J Dermatopathol 2020;42:916–922)

fibrosing dermal spindle cell proliferation may be mistaken with a scar, dermatofibroma or neurofibroma (NF), especially on a partial superficial shave biopsy.

Although immunohistochemical (IHC) studies can readily distinguish a fibrosing melanocytic neoplasm from a scar or dermatofibroma, the distinction from a NF is more challenging, especially, because some variants of DM can display features of “neural transformation” or Schwannian differentiation that closely simulate or are in foci indistinguishable from a NF. Few studies in the literature have addressed this diagnostic challenge.^{1–3} Furthermore, there is little information regarding the natural course of neurofibroma–like desmoplastic melanomas (NFLDMs). Although some IHC stains have been assessed for their potential to distinguish a NF from a classic DM, which do not closely resemble a NF, there are little data regarding how these IHC markers would perform for the distinction of NFs from NFLDMs. In this study, we present 12 cases of NFLDMs, 8 of which had clinical follow-up. Four of 8 cases had a clinical recurrence; of which 2 had transformation to a more aggressive phenotype. We also examined PRAME, P53, and neurofilament for their potential to distinguish NFs

But there is a technique that reliably distinguished NF and NF-like DMM in this case

But there is a technique that reliably distinguished NF and NF-like DMM in this case







