

EYE PATHOLOGY:

Cornea, conjunctiva, and eyelid

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Disclosures

- I have no relevant financial relationships to disclose



Learning Objectives

Summary: We will cover three areas of eye pathology: **cornea, conjunctiva, and eyelid**. We will discuss the differential diagnosis of lesions at these sites, focusing on the most common entities.

1. Identify the key pathologic features and differential diagnosis of **corneal** lesions.
2. Identify the key pathologic features and differential diagnosis of **conjunctival** lesions.
3. Identify the key pathologic features and differential diagnosis of select **eyelid** lesions.



NO PHOTOGRAPHY OR SOCIAL MEDIA SHARING



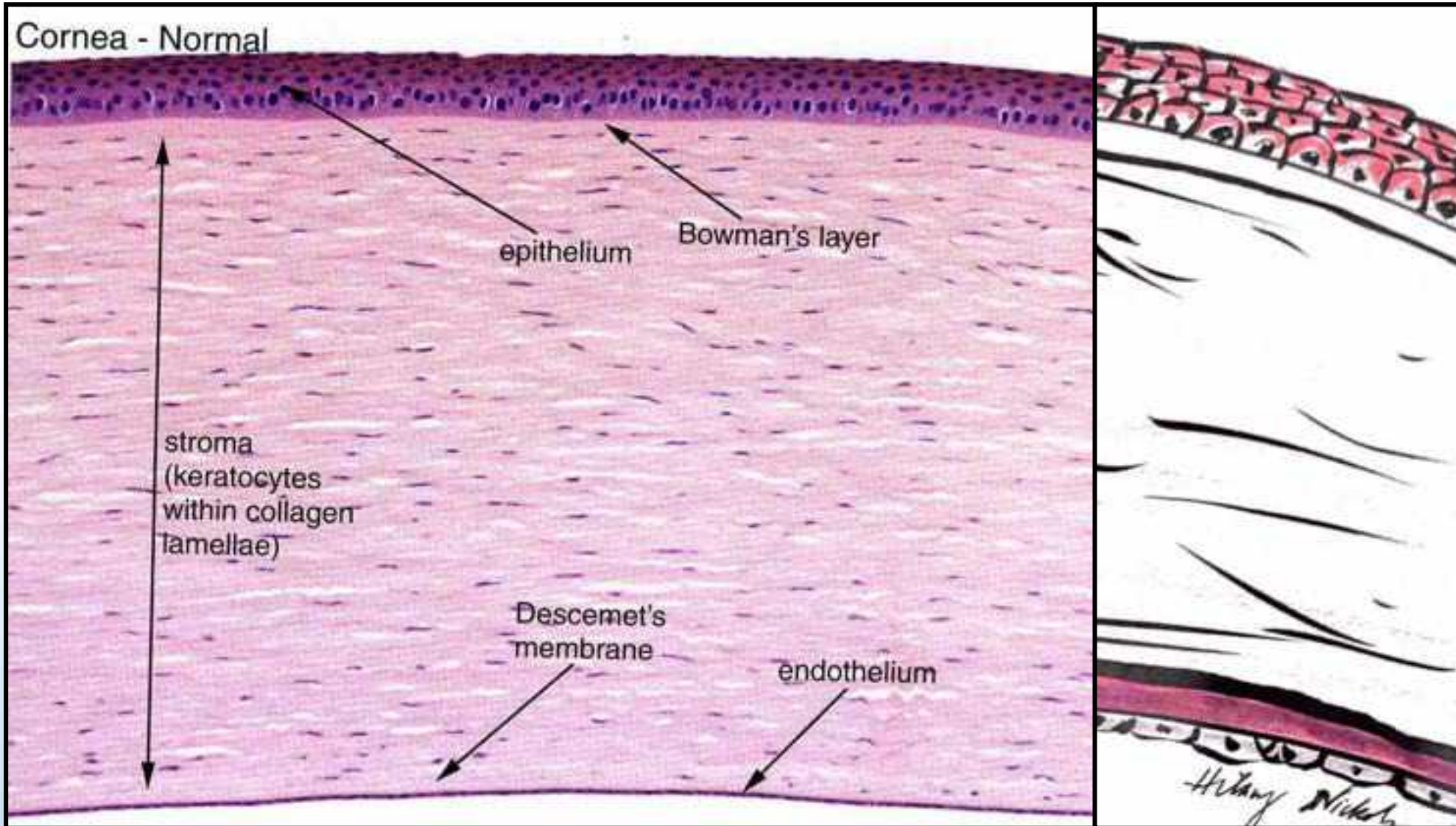
Thank you!



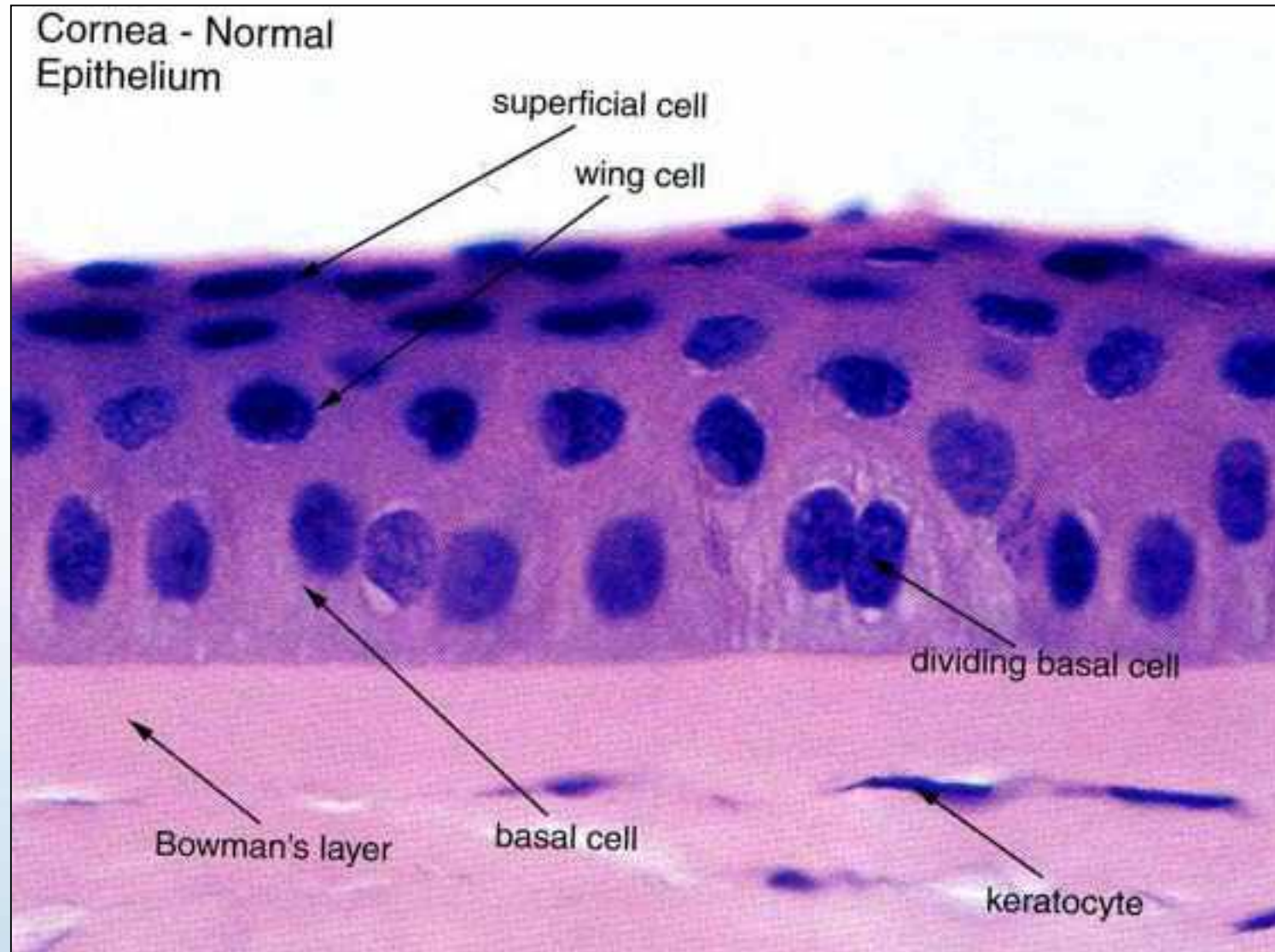
CORNEA



CORNEA



Epithelium



Eagle, Ralph C. (2017), *Eye Pathology: An Atlas and Text*, 3rd ed. Wolters Kluwer

Epithelium



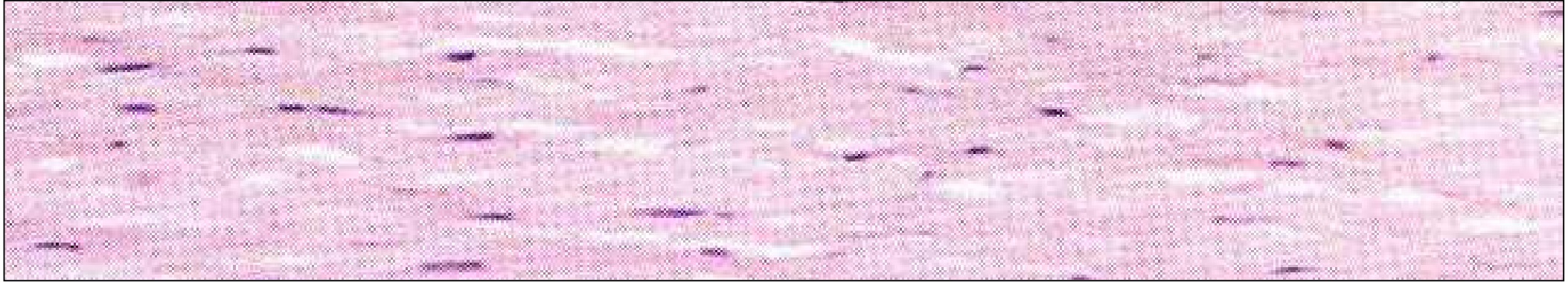
- **stratified squamous, non-keratinizing,**
- **5-7 layers thick; stains with PAS**
- **mitotically active basal layer attached to basement membrane**
- **wing cells second layer designed to fit over the rounded anterior surface of basal cells**
- **superficial cells, flatten as reach surface**
- **basal layers connected to Bowman's by an anchoring complex**
- **cells connected by desmosomes**

Bowman's layer



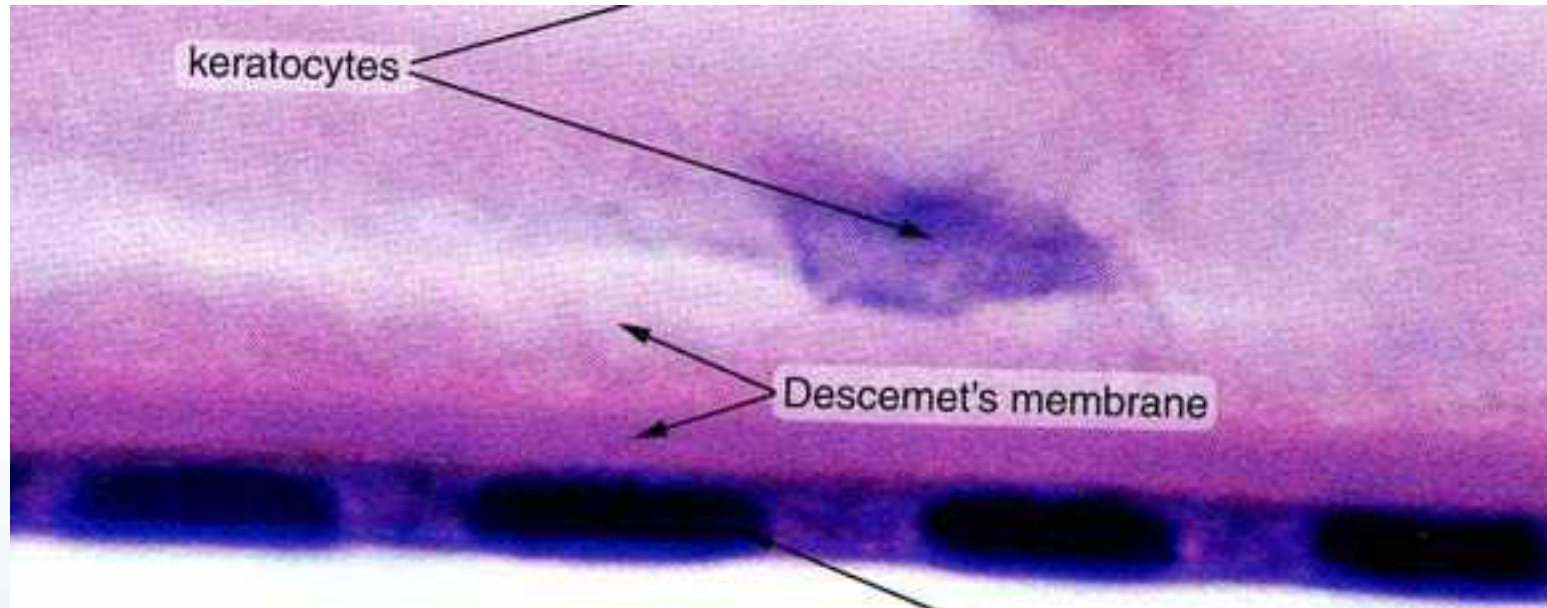
- beneath epithelial basement membrane (PAS+)
- acellular collagen layer
- 8-14nm thick
- once destroyed (trauma/ulcer), never replaced

Stromal layer



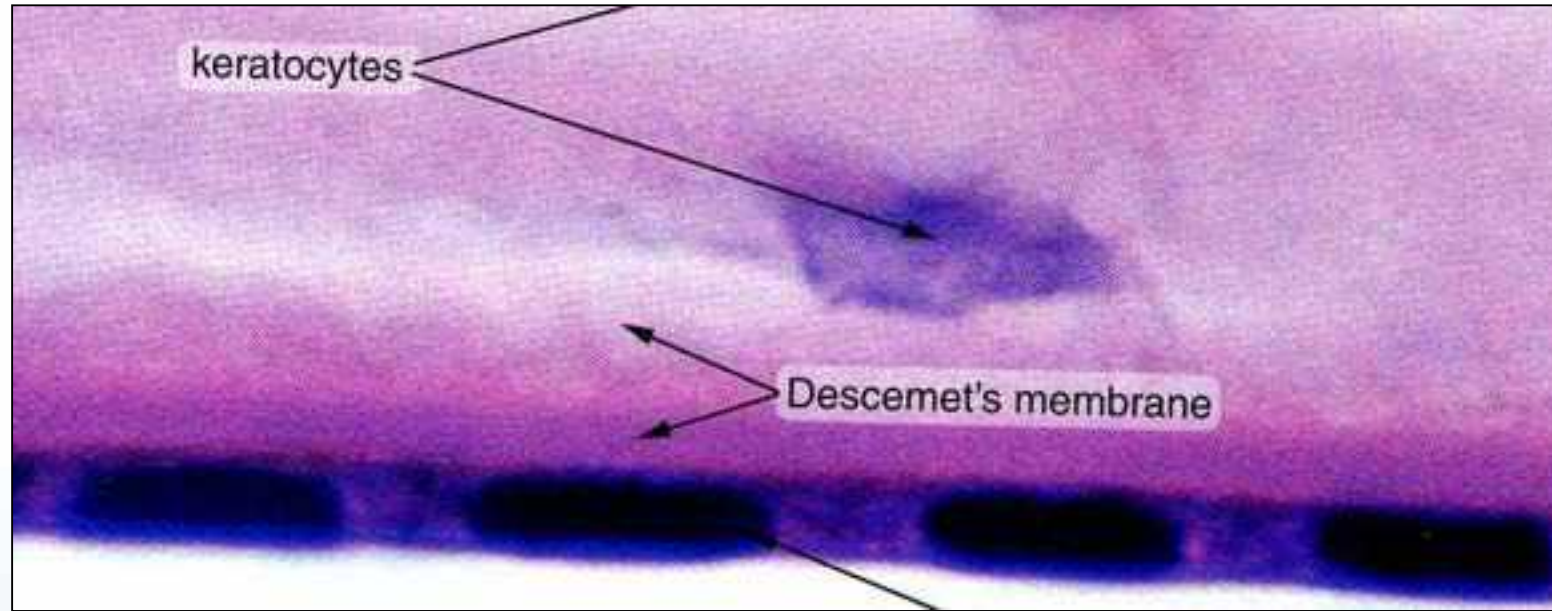
- 90% of corneal thickness (500mM)
- spindled keratocytes with long branching interconnected processes, lie between lamellae with uniformly spaced collagen fibrils
- elongate collagenous lamellae
- ground substance of mucoprotein and glycoprotein coats each fibril

Descemet's membrane



- Thin elastic membrane of high tensile strength
- Contains proteoglycans and glycoproteins in addition to collagen
- Basement membrane elaborated by endothelium
- Composed of type IV collagen
- Strongly PAS positive

Endothelium

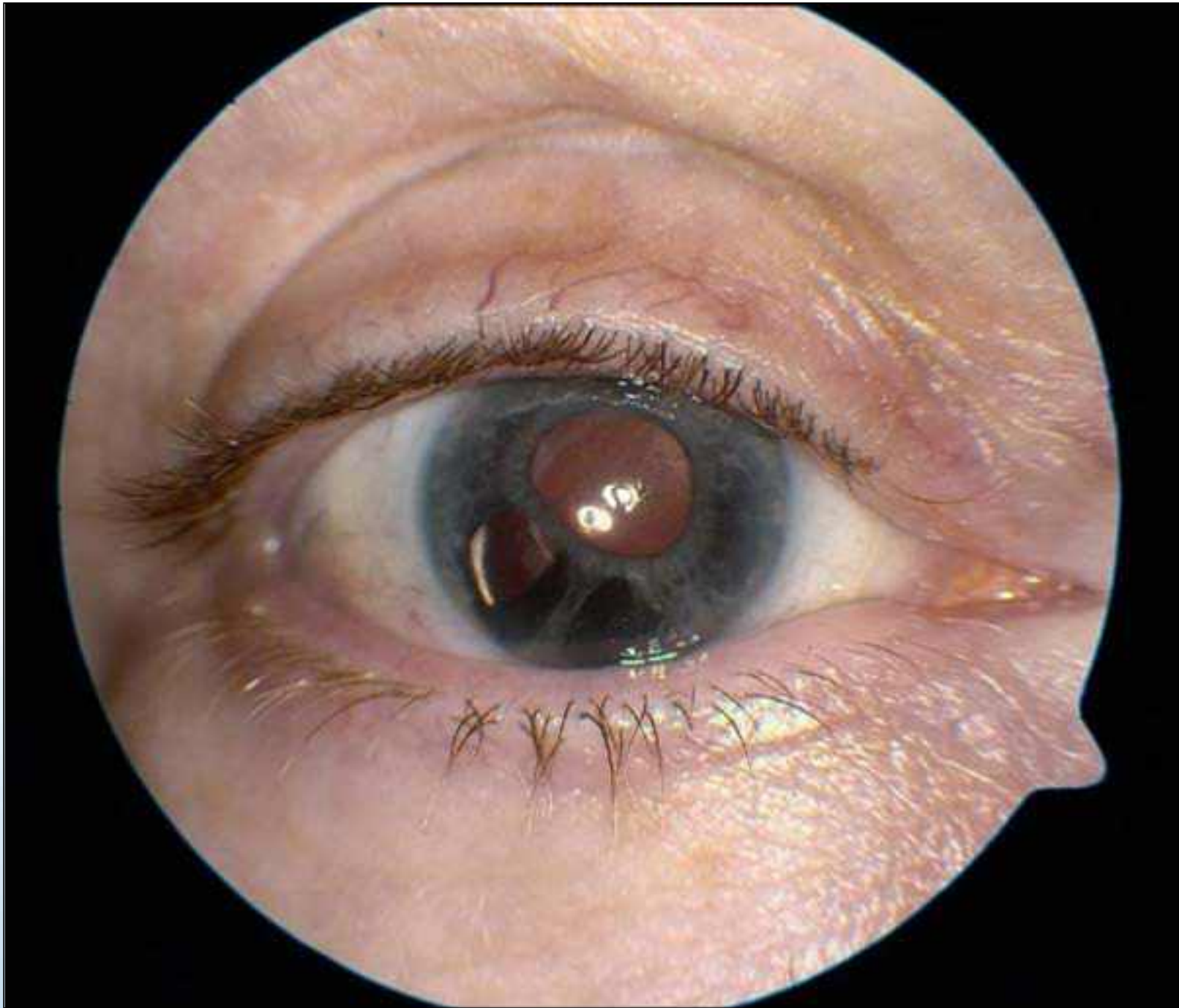


- Monolayer of flattened cuboidal cells
- Derived from neural crest
- Maintains corneal clarity by removing water from stromal layer
- Cell population declines with age

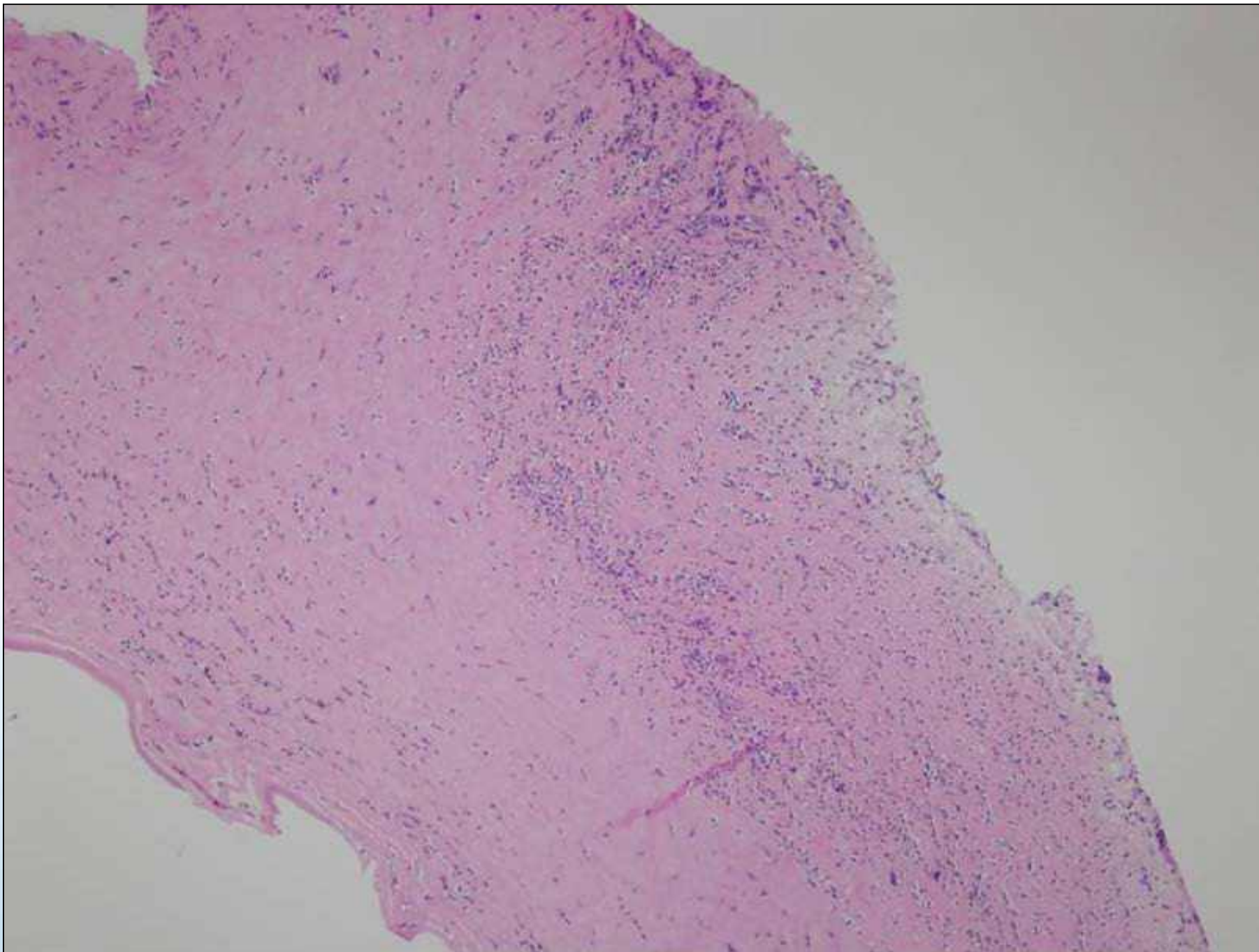
Corneal infection

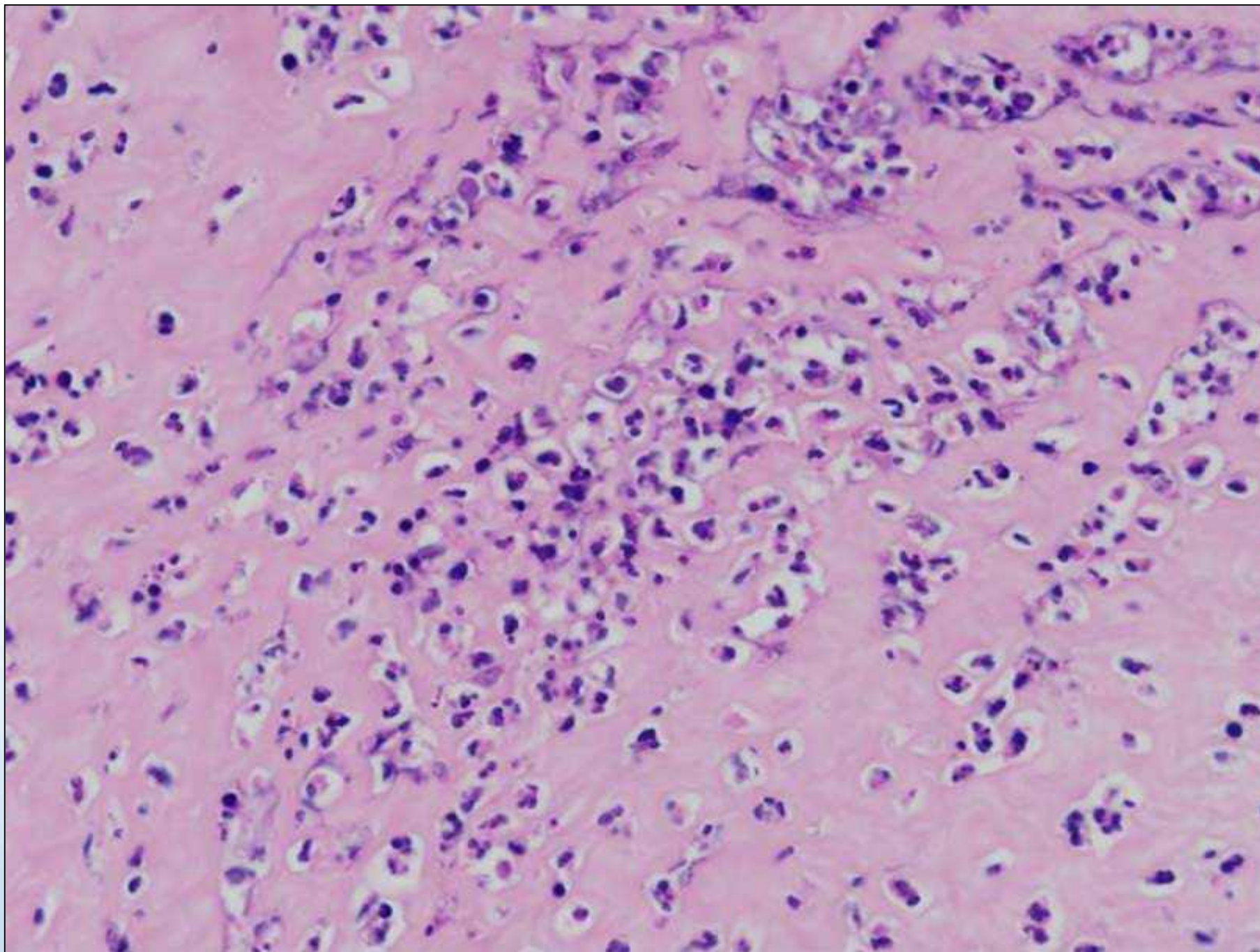
- Mechanisms protecting cornea from infection
 - Corneal epithelium
 - Mucous strands of conjunctival epithelium ensnare microorganisms
 - Tears: lactoferrin, lysozyme, antibodies (lymphs/PCs)
- Compromise of protective mechanisms
 - Inoculation by foreign body
 - Epithelial abrasion
 - Invasive organisms (*Neisseria gonorrhoeae*)

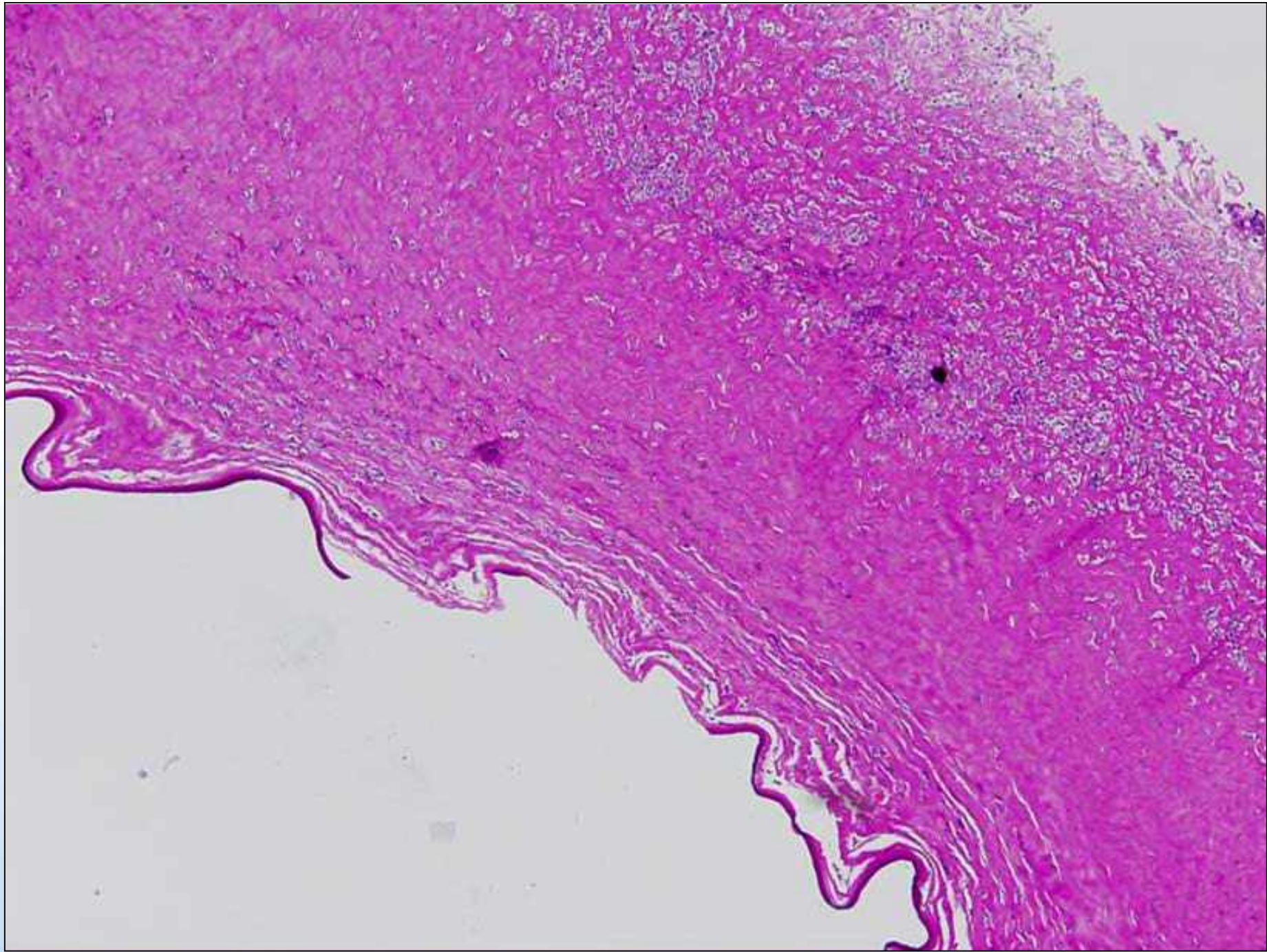








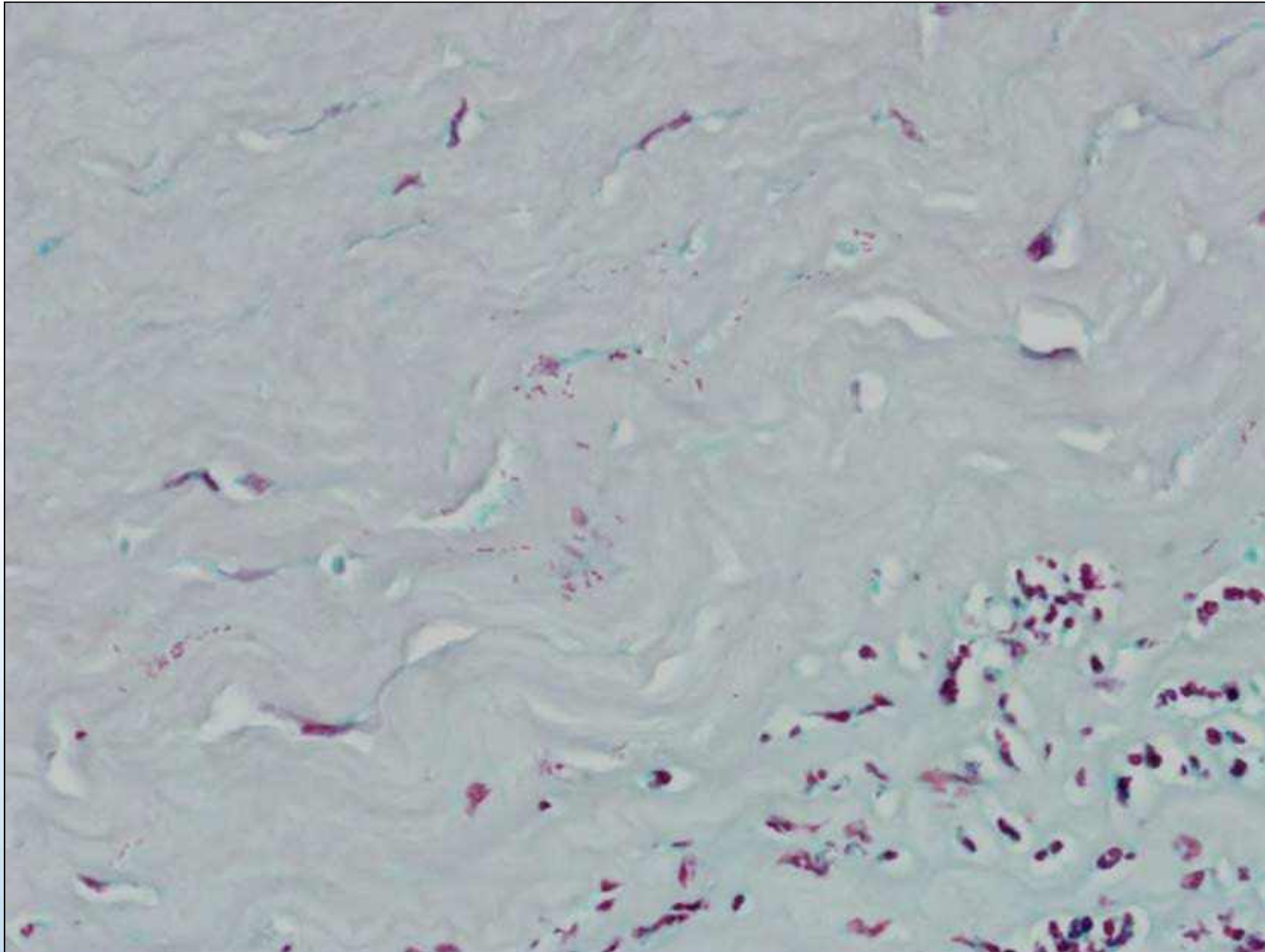




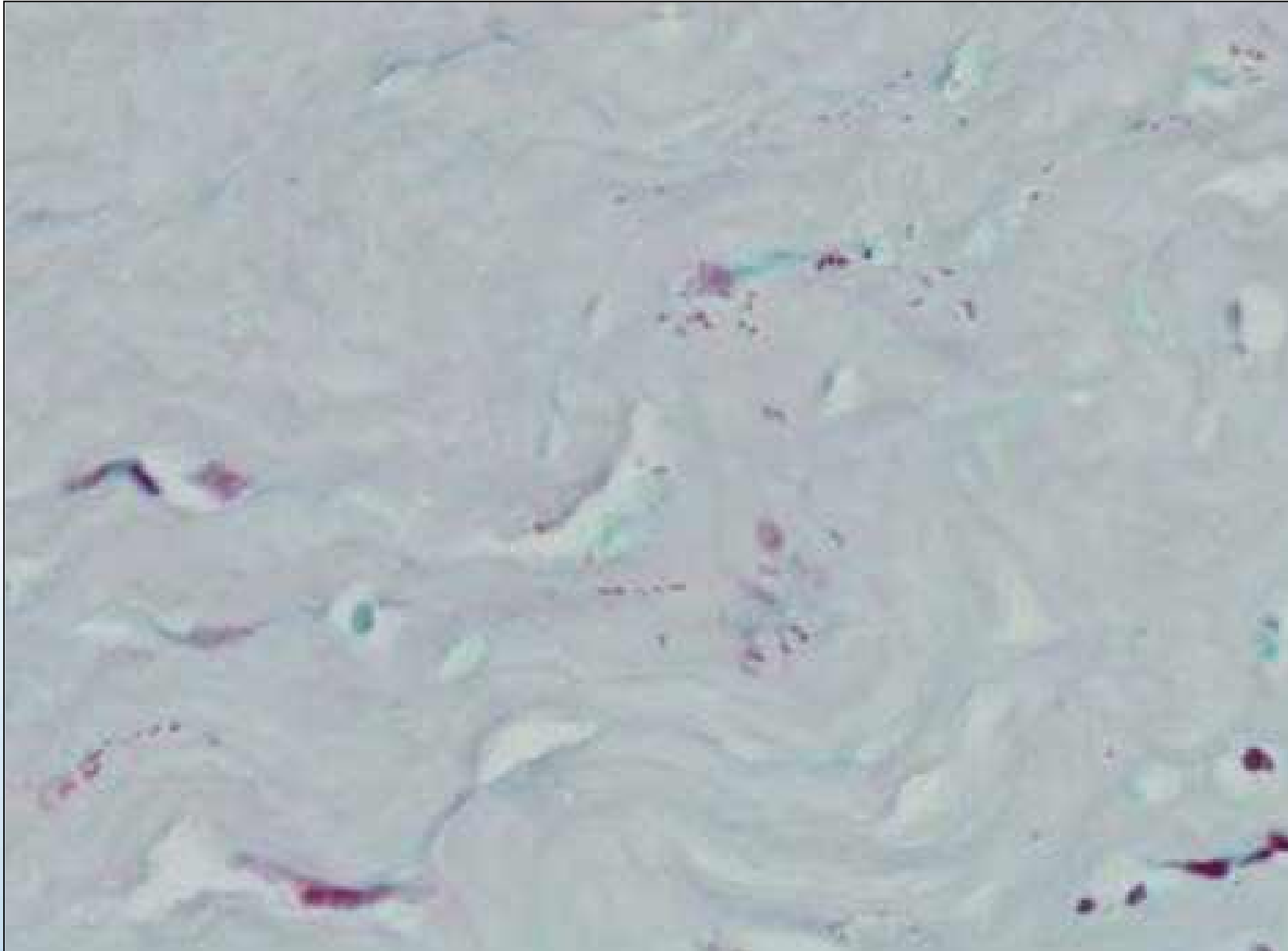
PAS



Gram Stain



Gram Stain



Acute bacterial keratitis

- Infiltration of stroma by PMNs occurs in acute bacterial keratitis
- PMNs collect in clefts between adjacent stromal lamellae
- Digestive enzymes released by dying inflammatory cause stromal necrosis

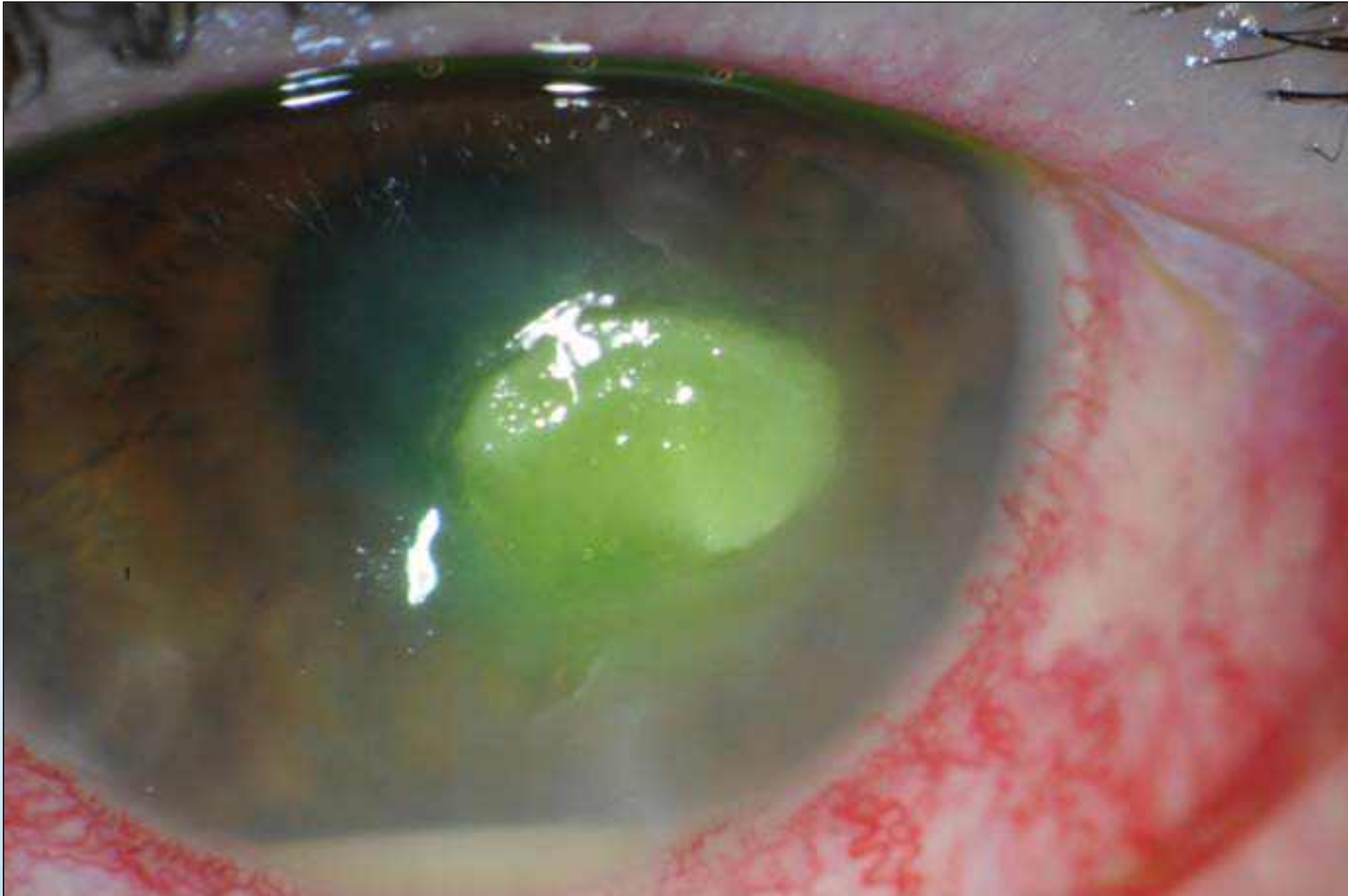


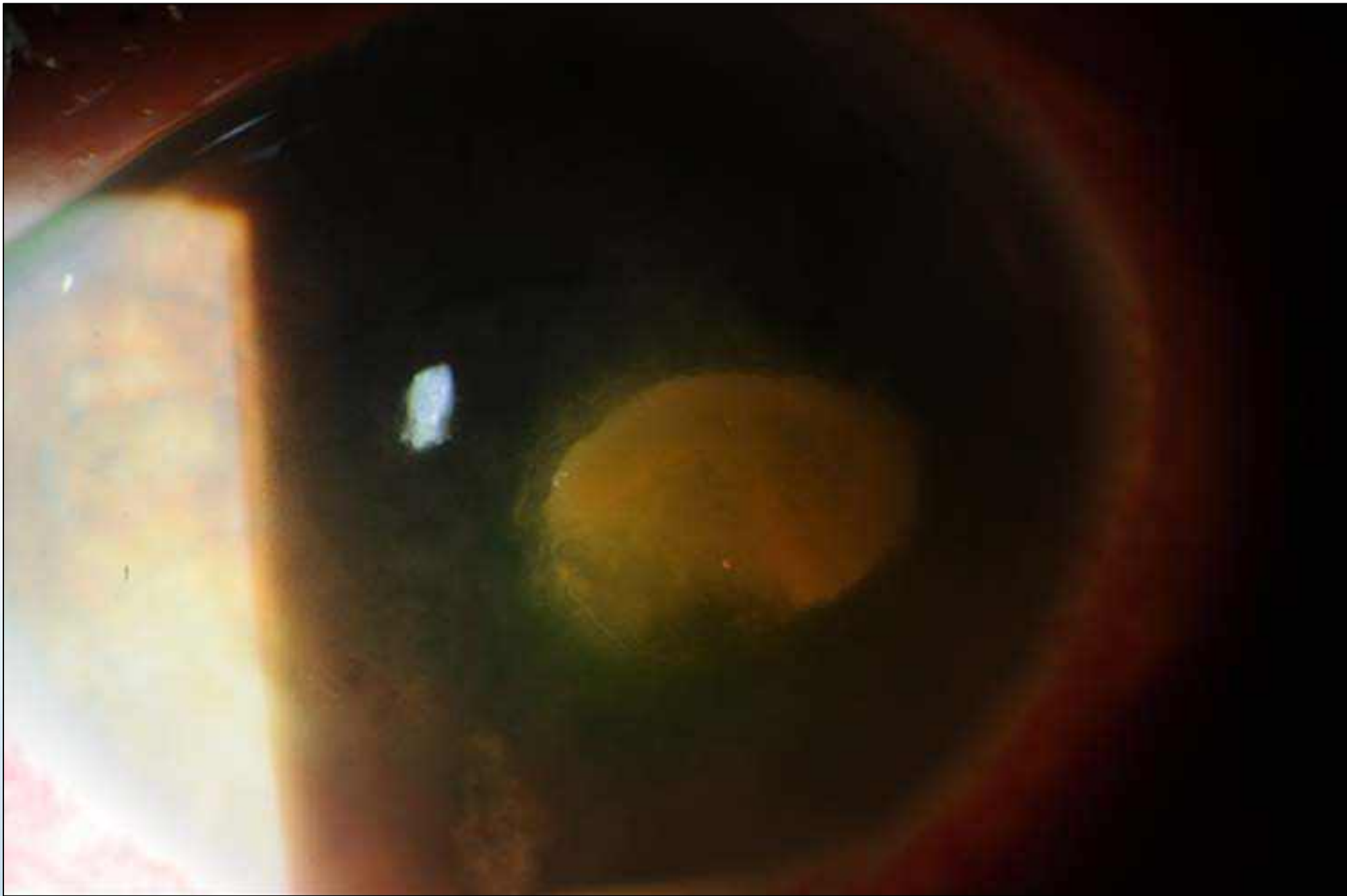
Bacterial Keratitis: Pseudomonas

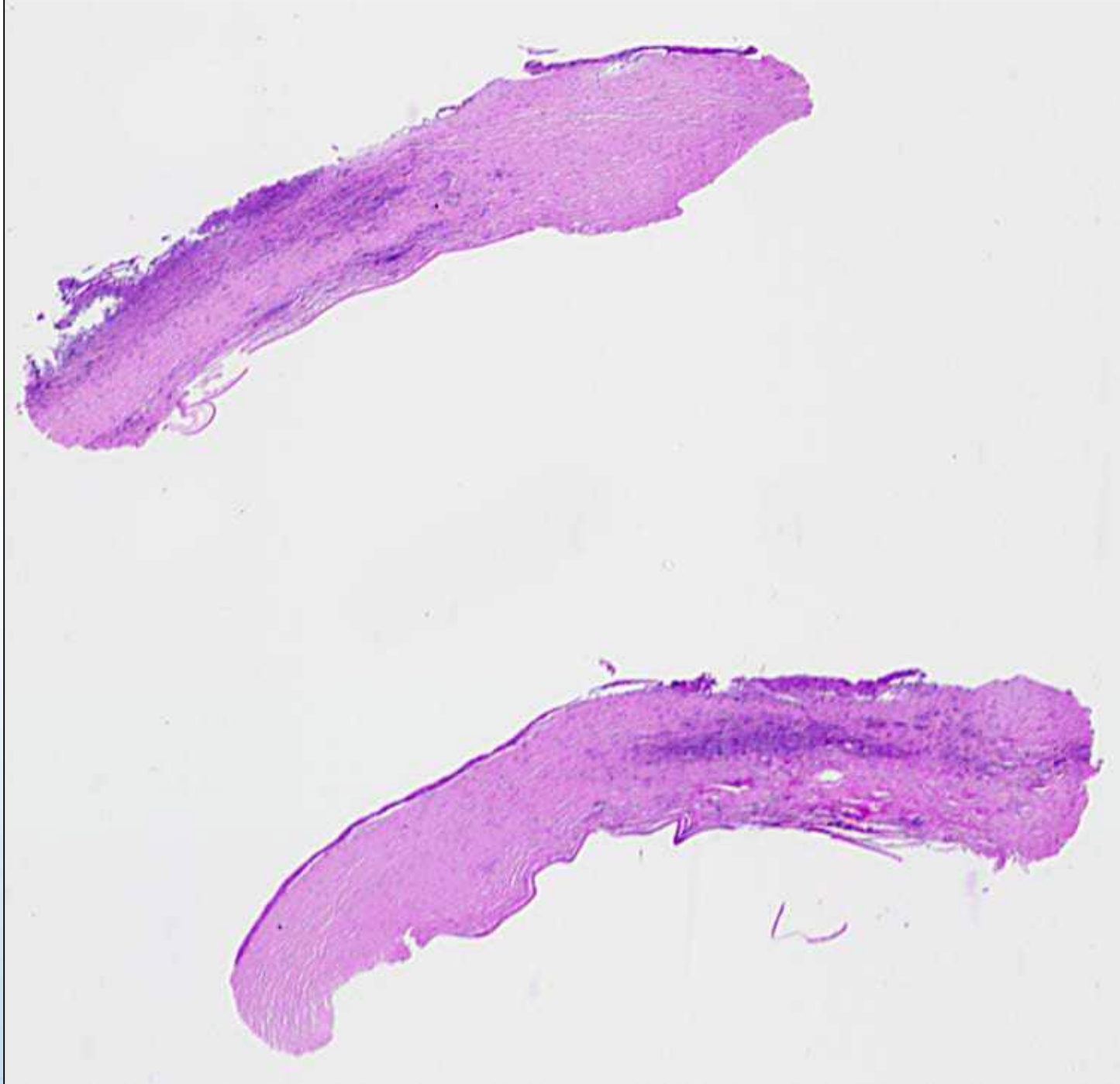
- Pseudomonas produces enzymes that destroy cornea
- Marked stromal edema and dissolution
- Stromal infiltration by neutrophils and necrosis
- Rapid corneal perforation
- Infection can spread posteriorly → sclerokeratitis
- Most frequent cause of contact lens-associated microbial keratitis
- Organisms invade and replicate within surface corneal epithelial cells in animal models of infection

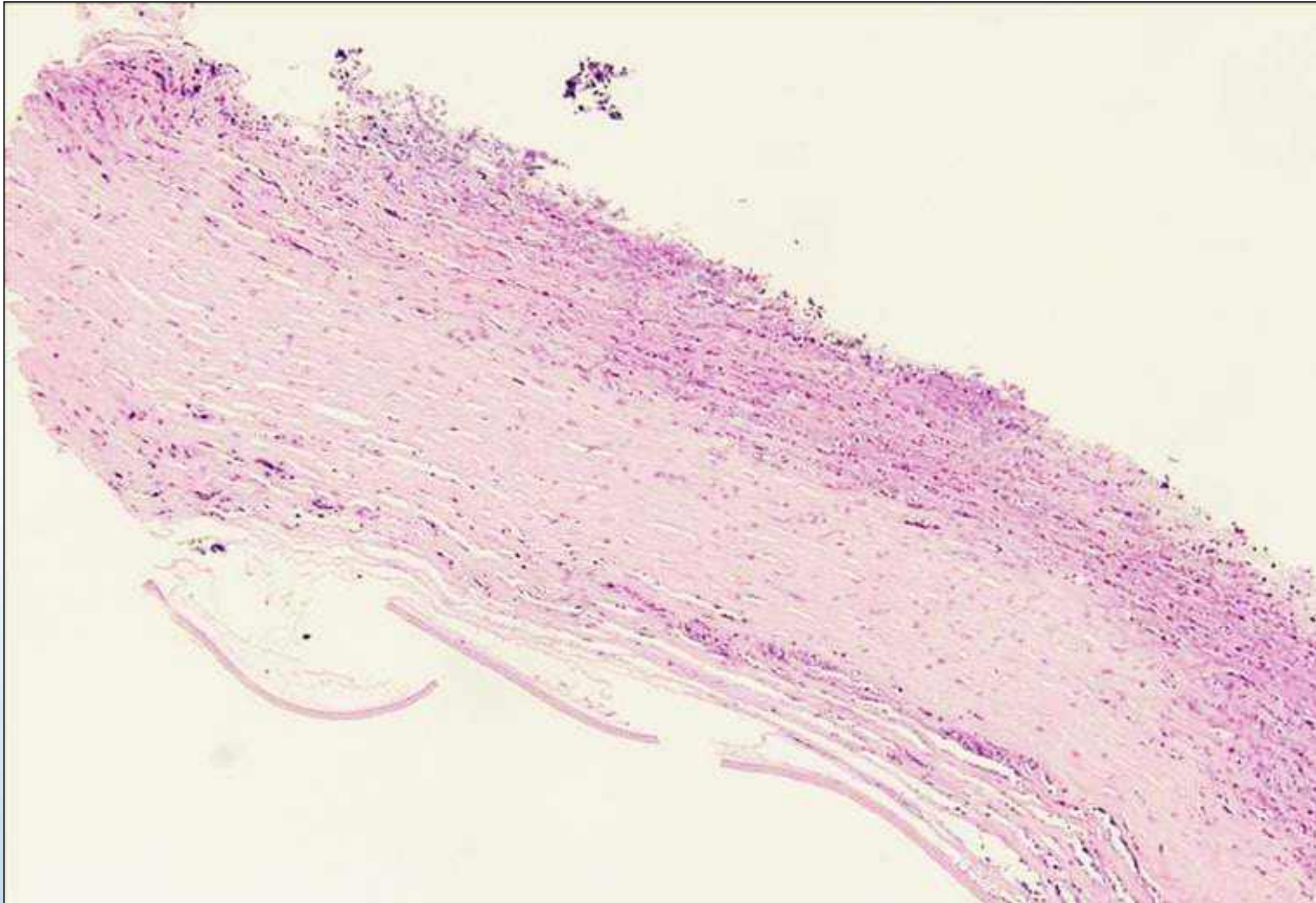


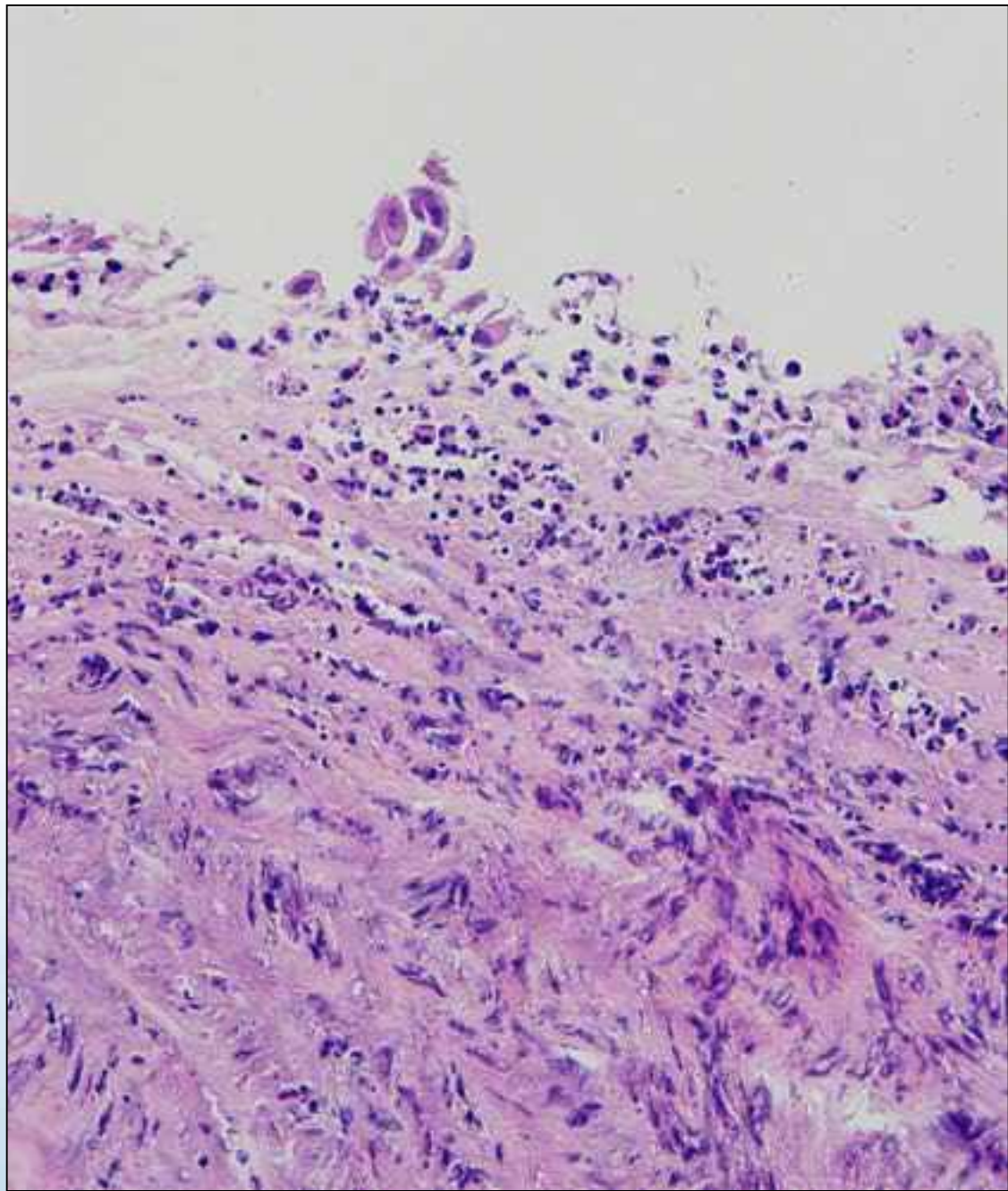
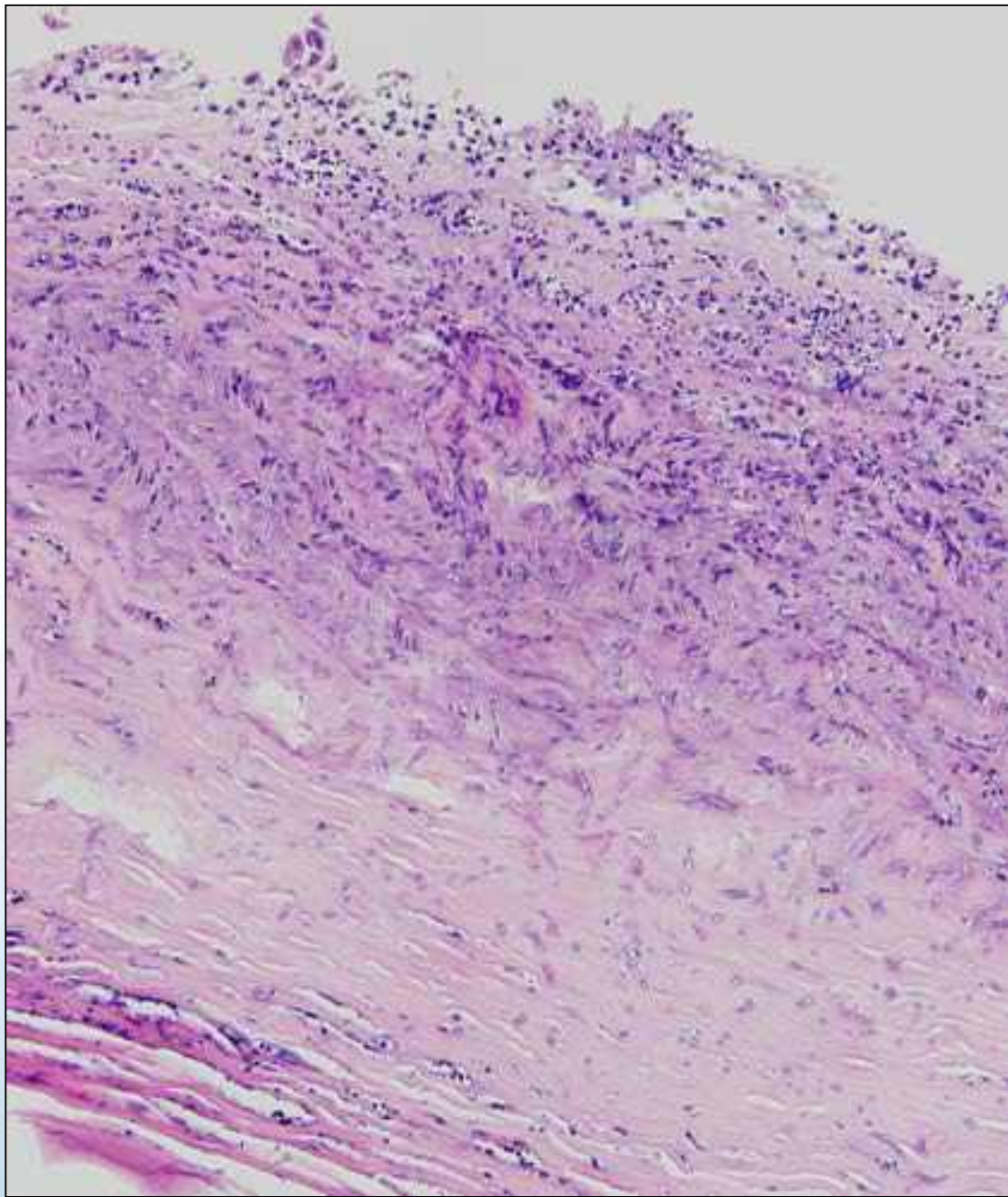
Slit Lamp Photo

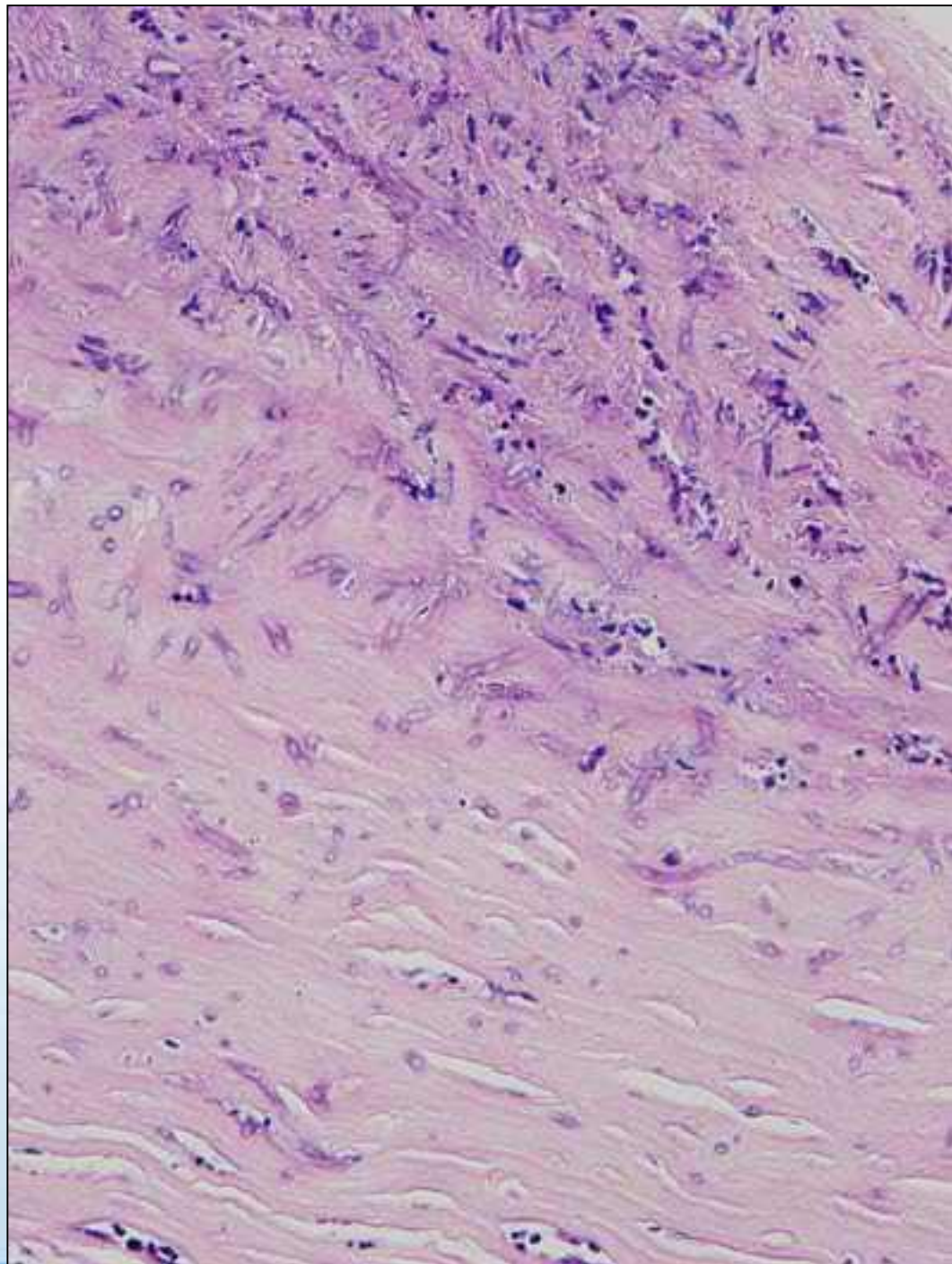


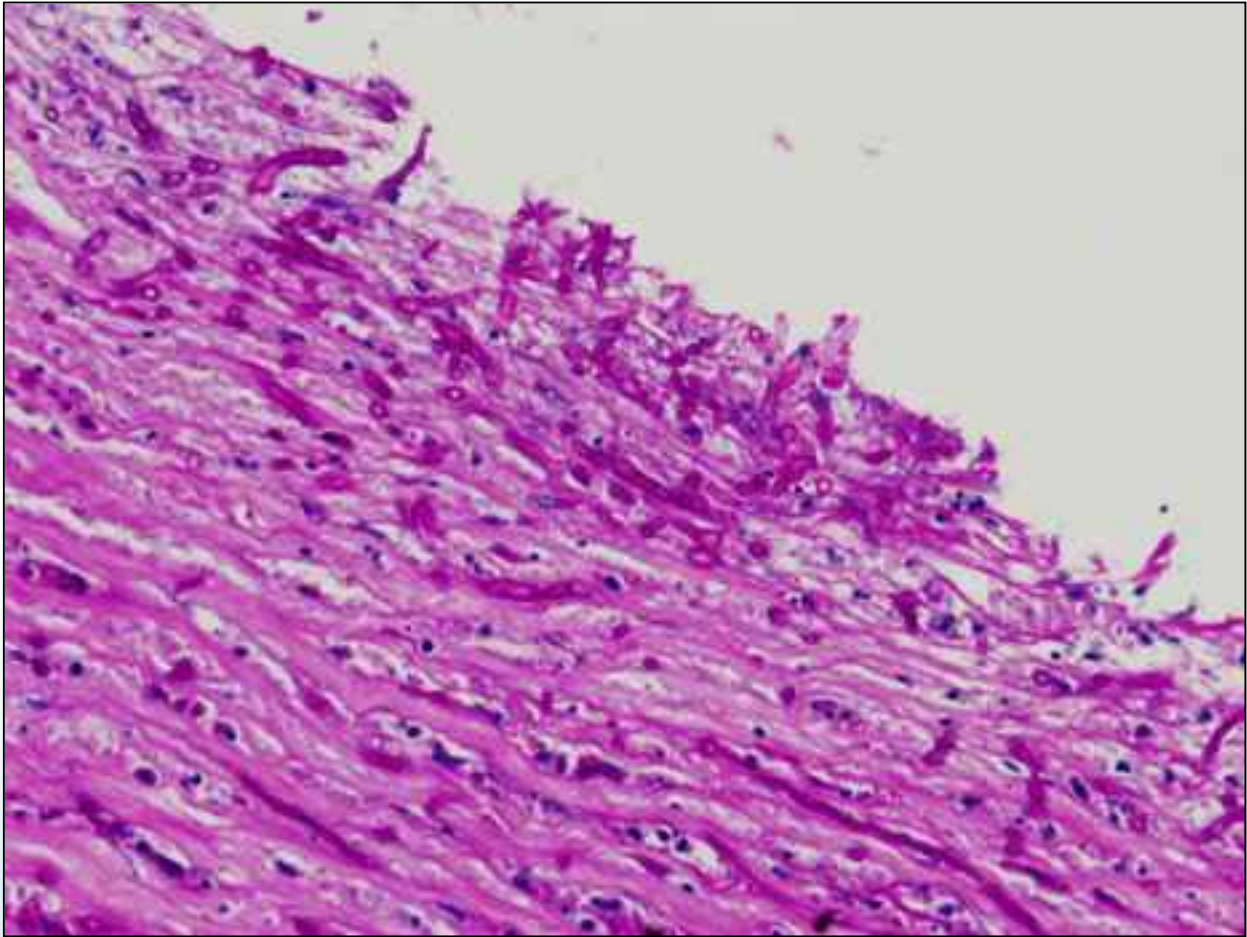
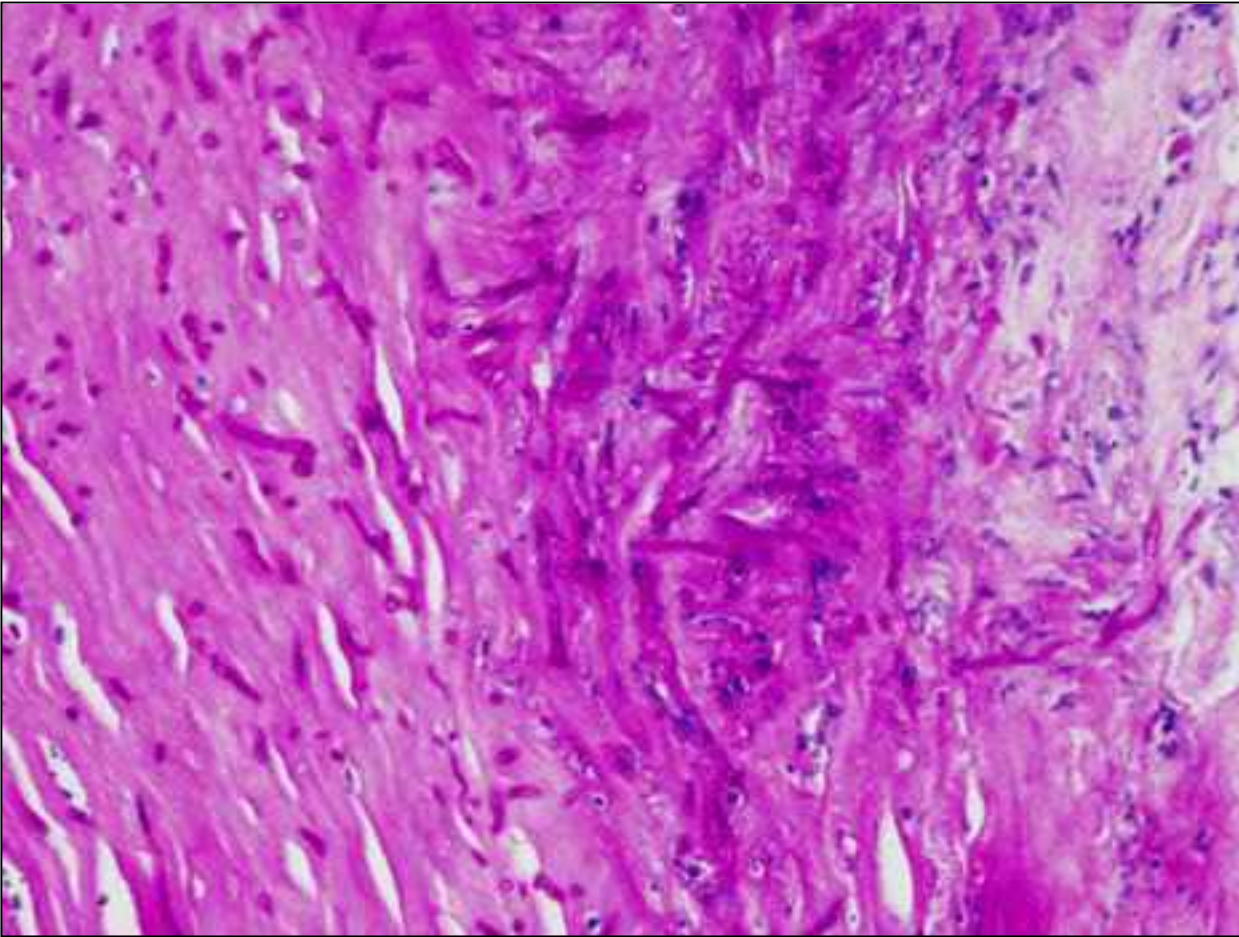










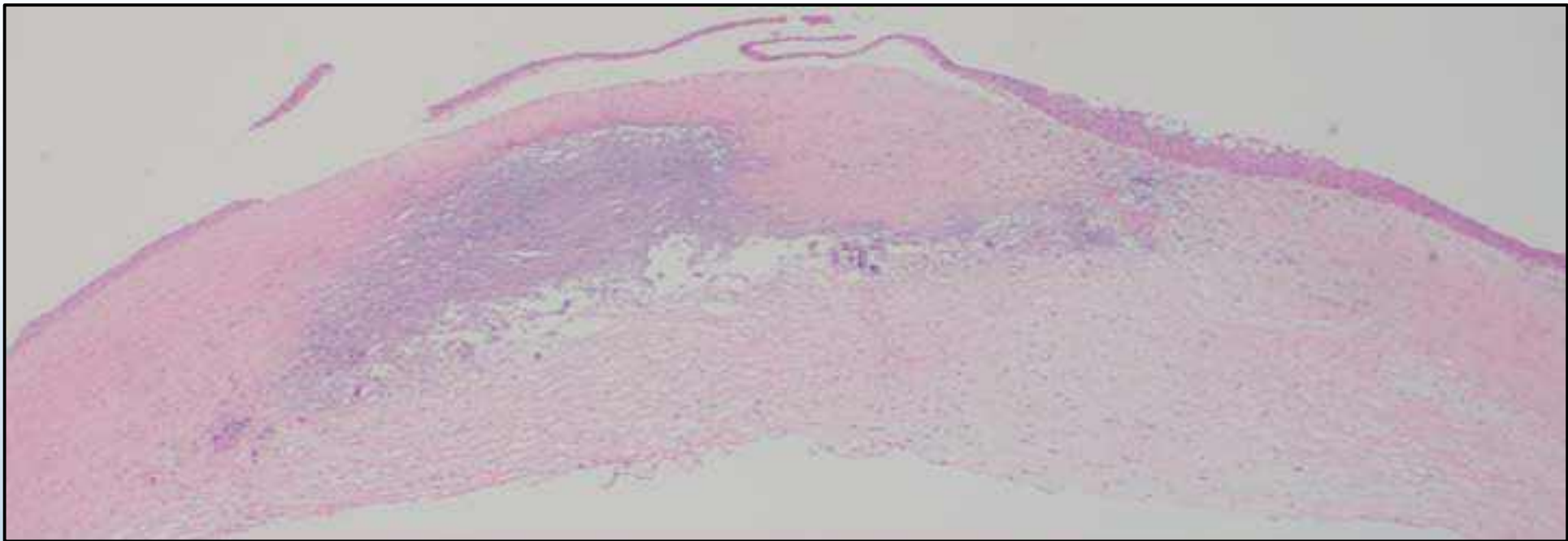
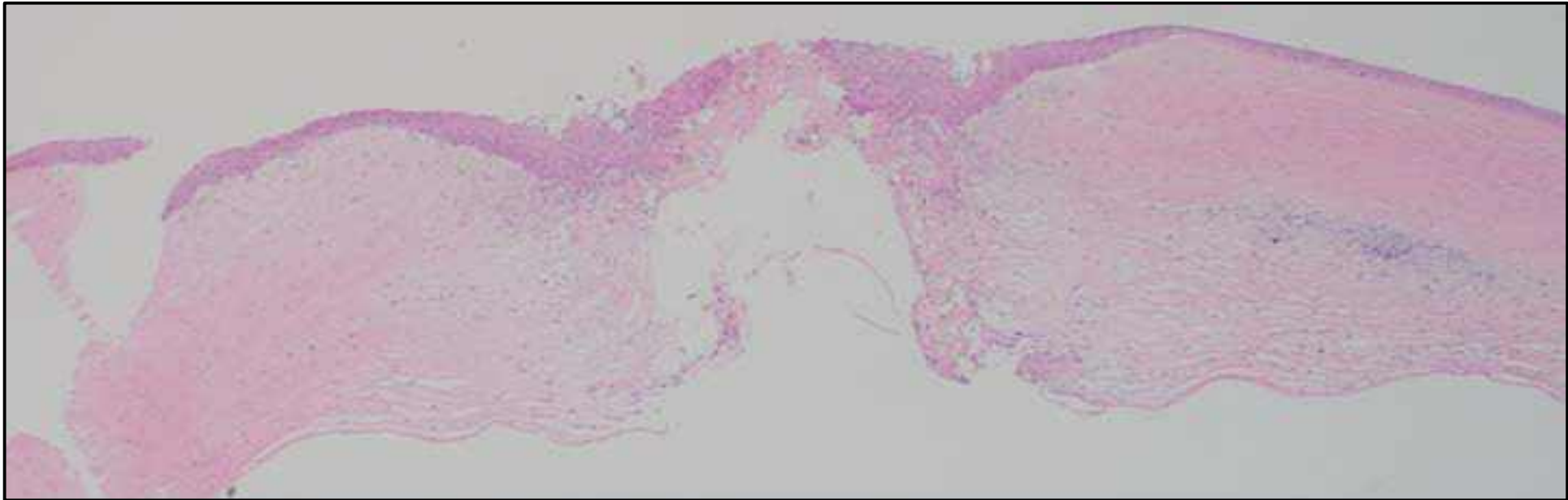


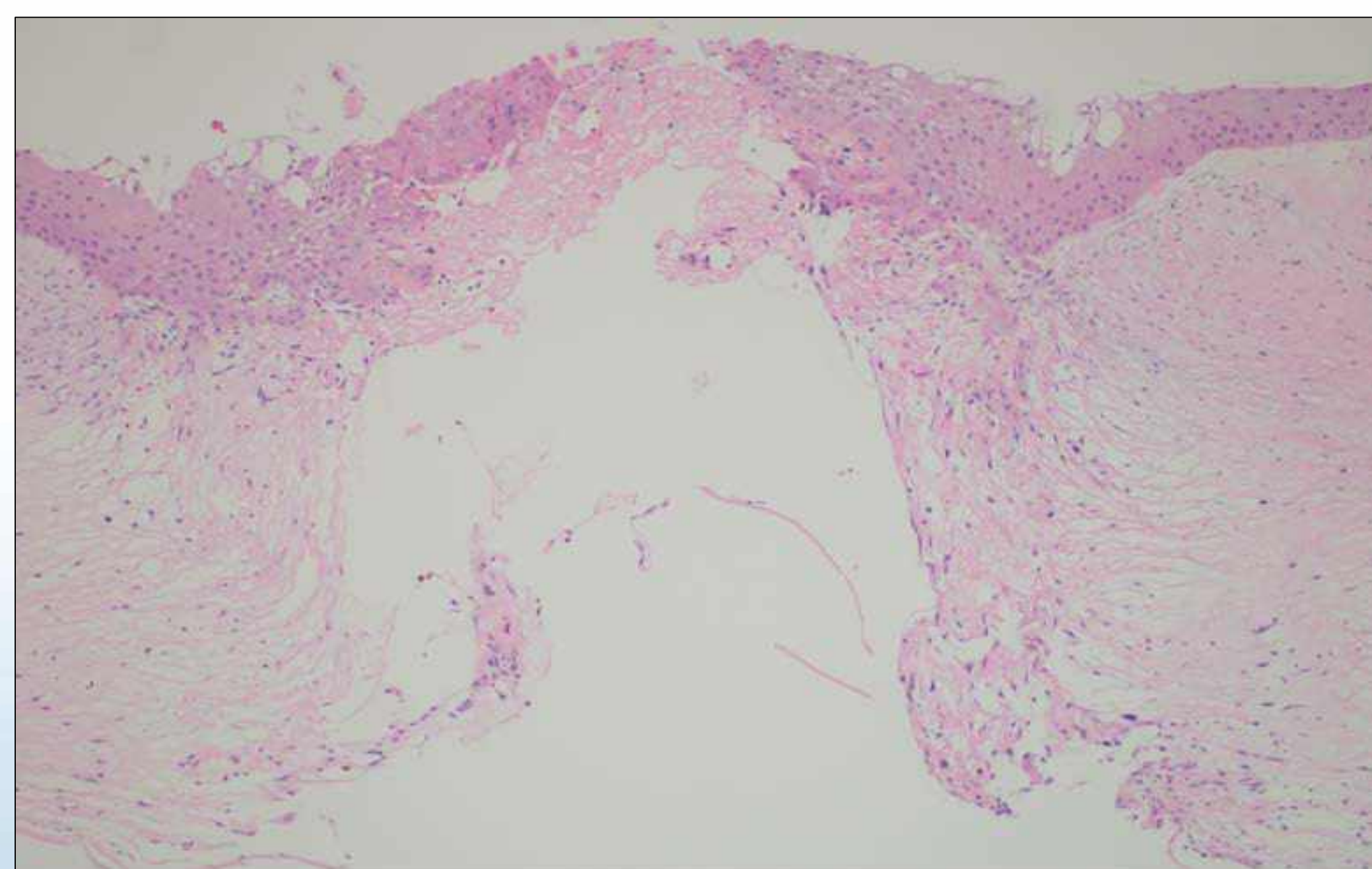
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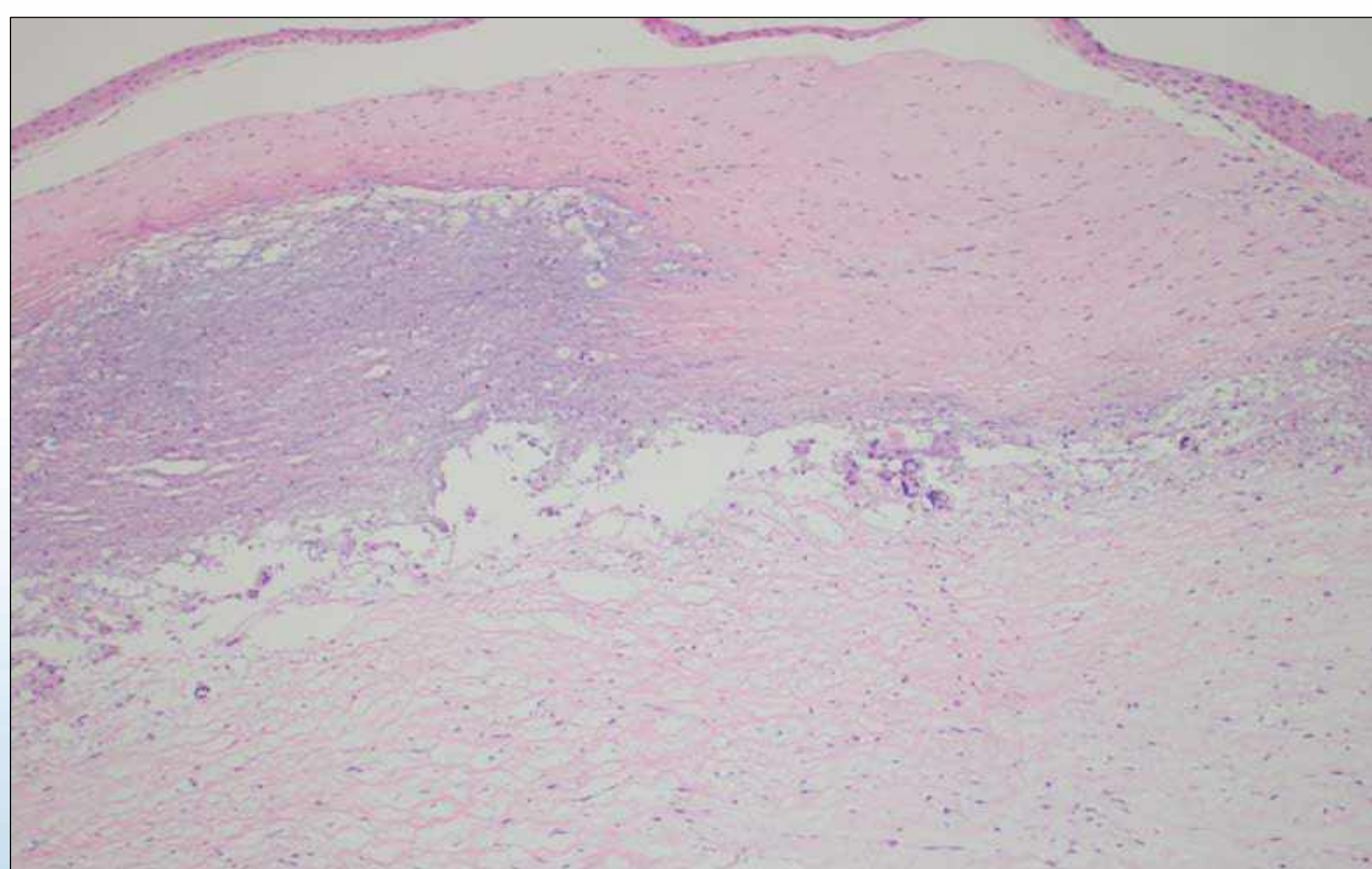


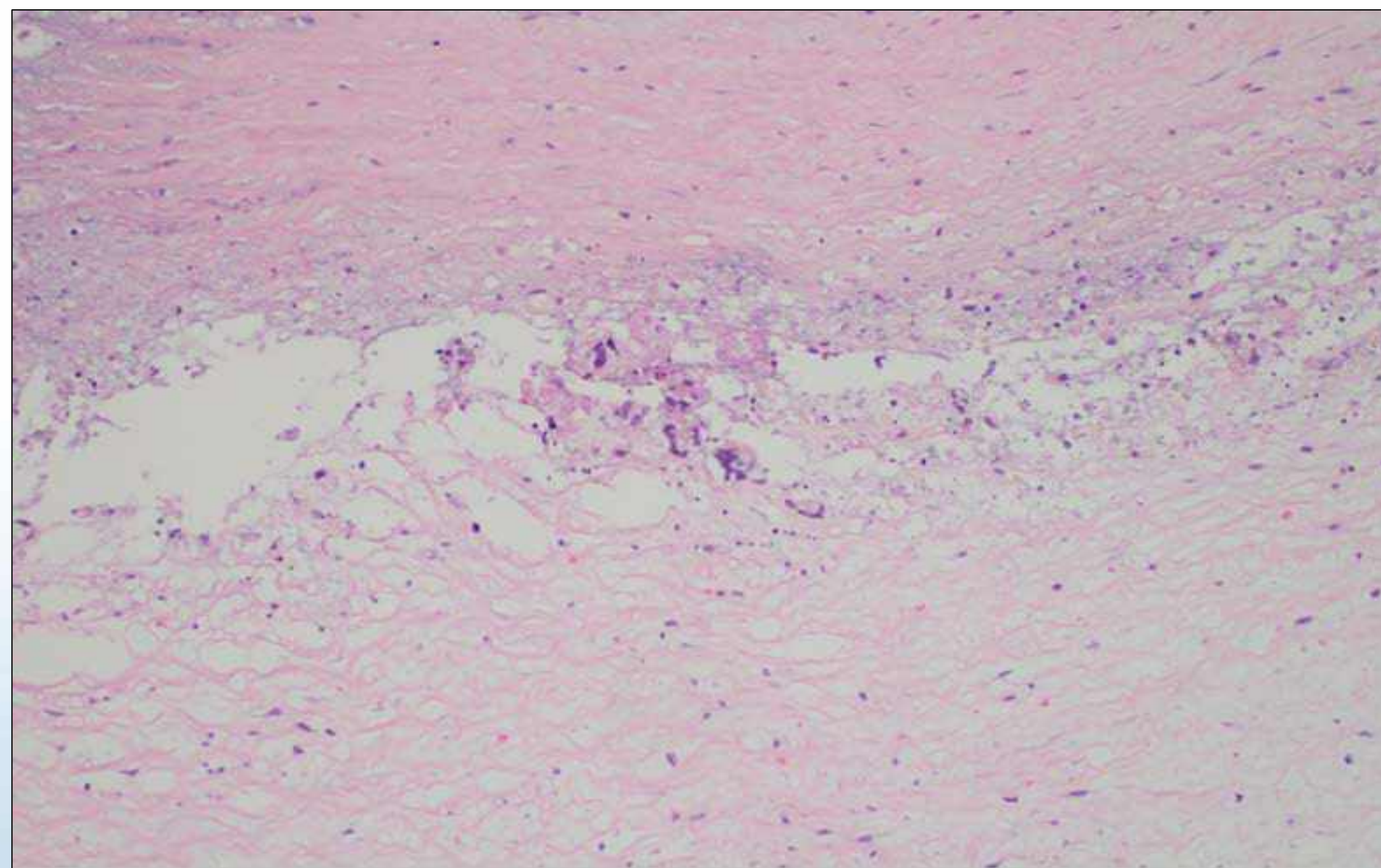
Corneal graft, 61 yo F

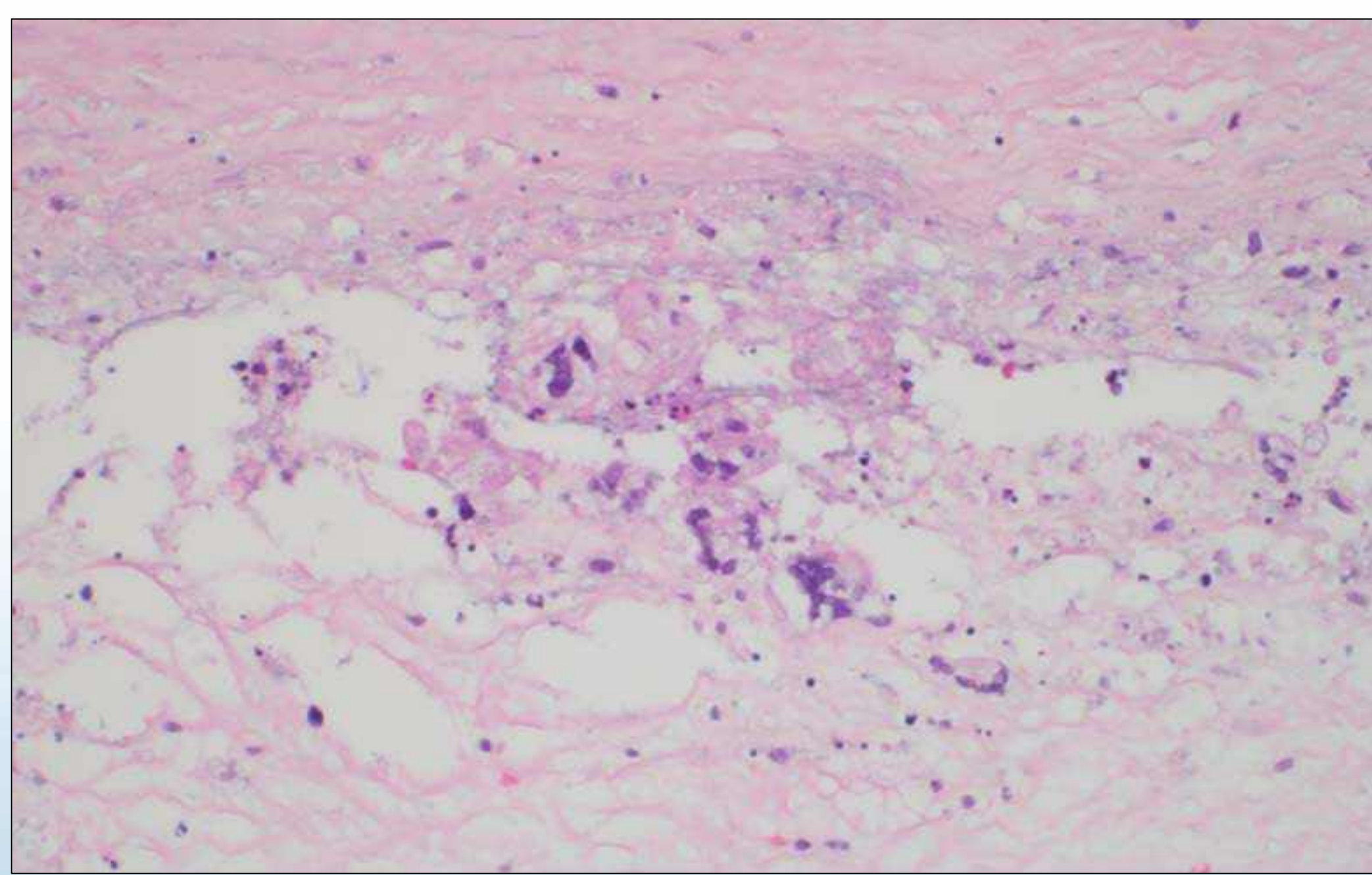


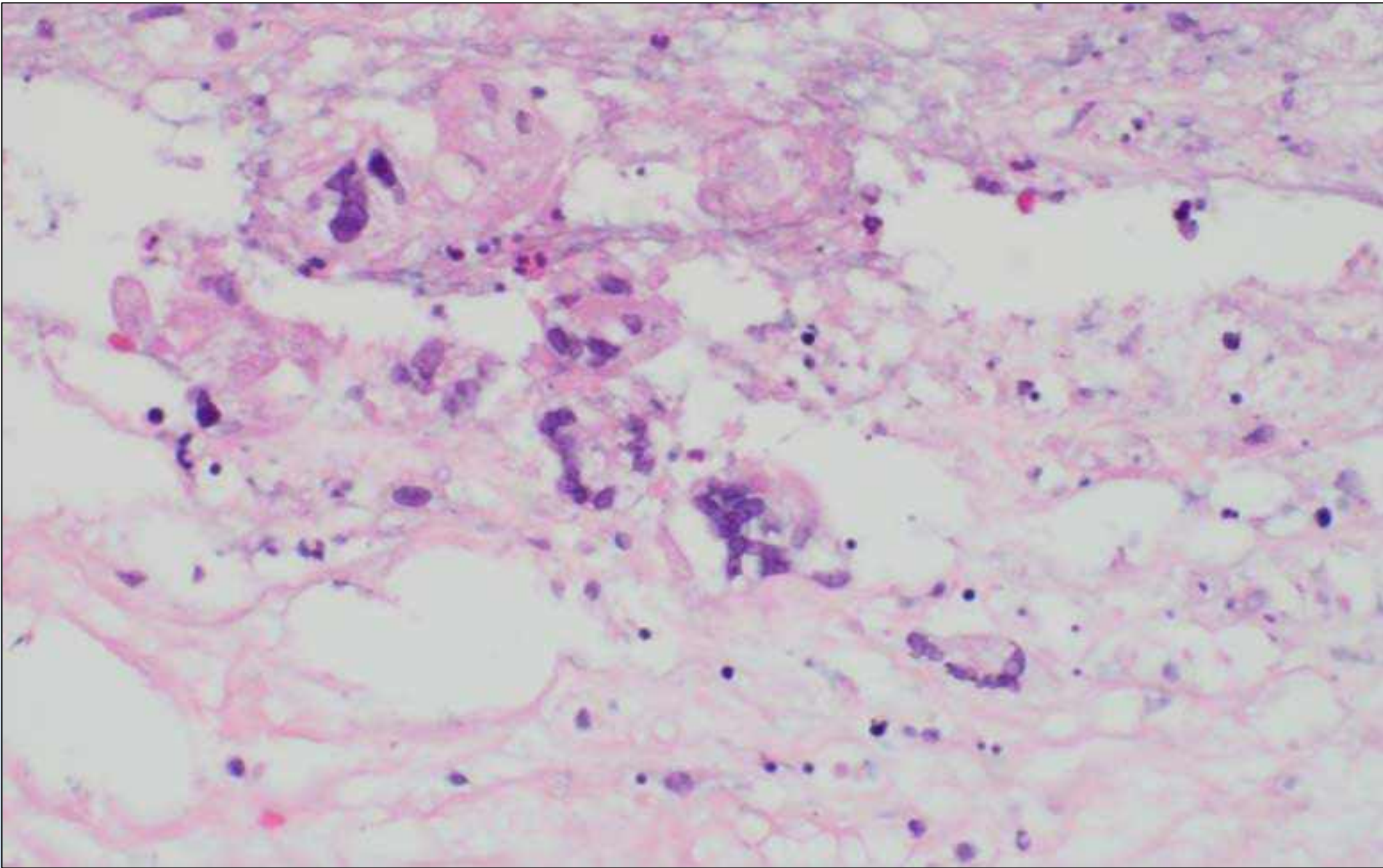


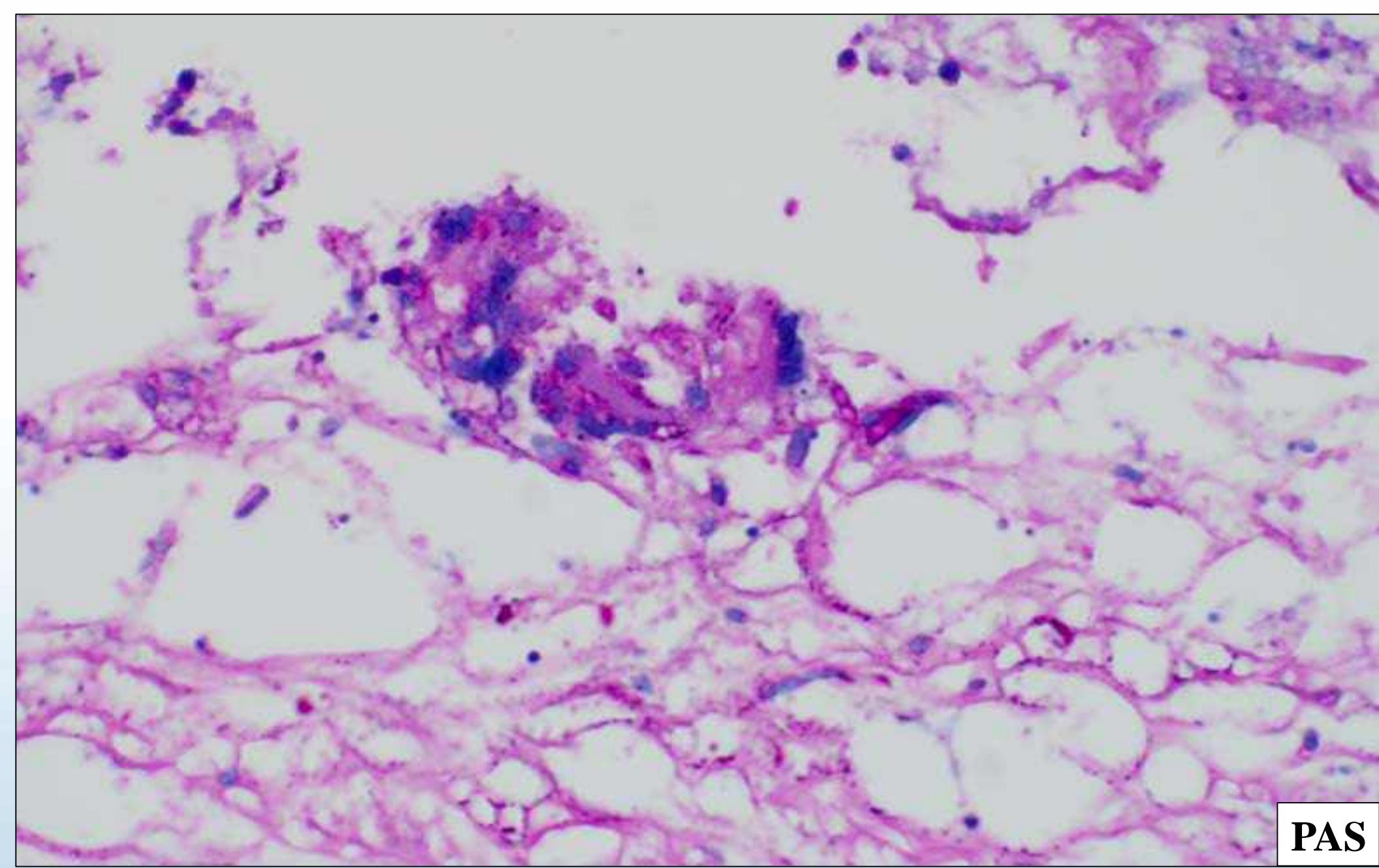






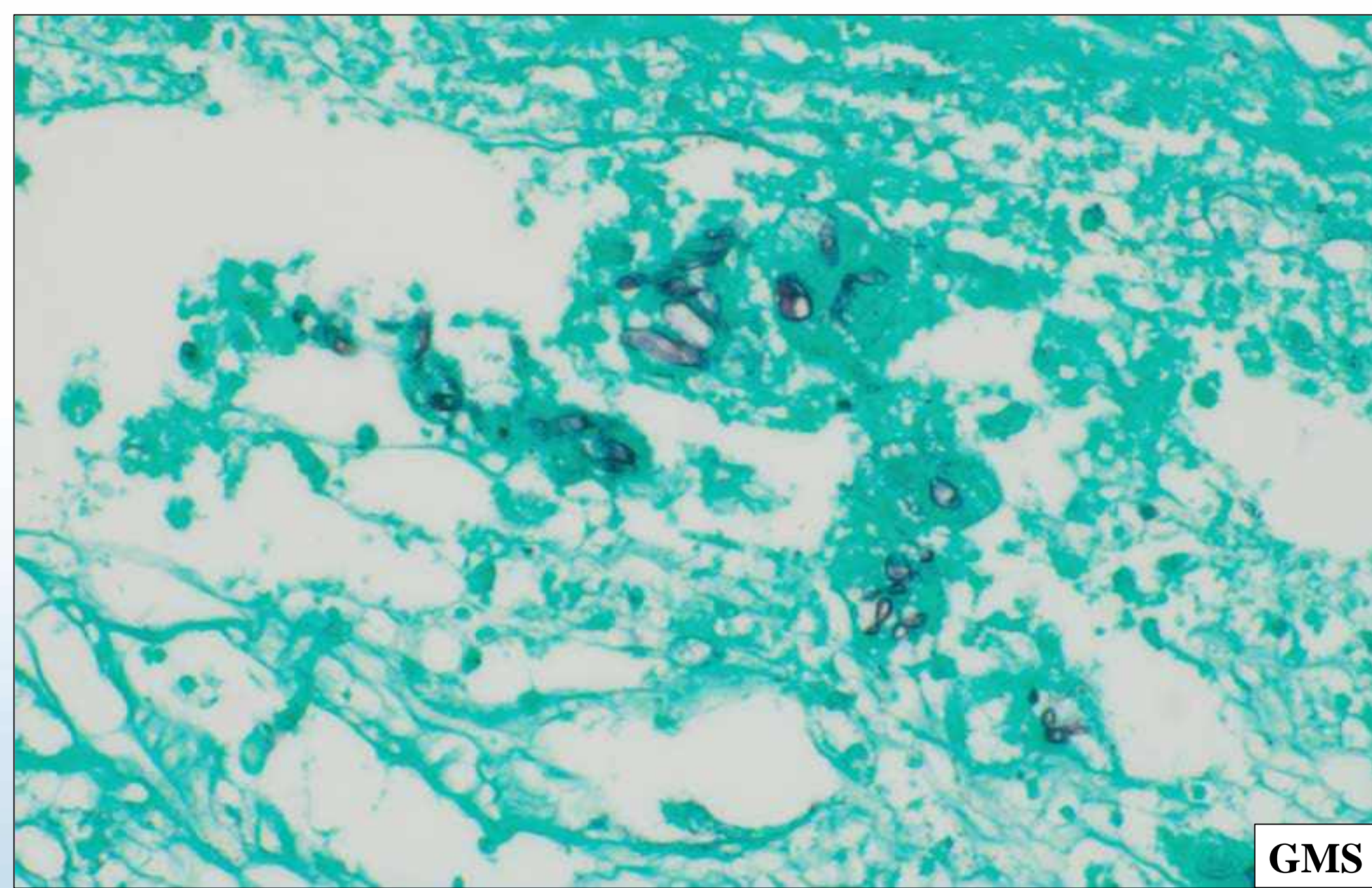






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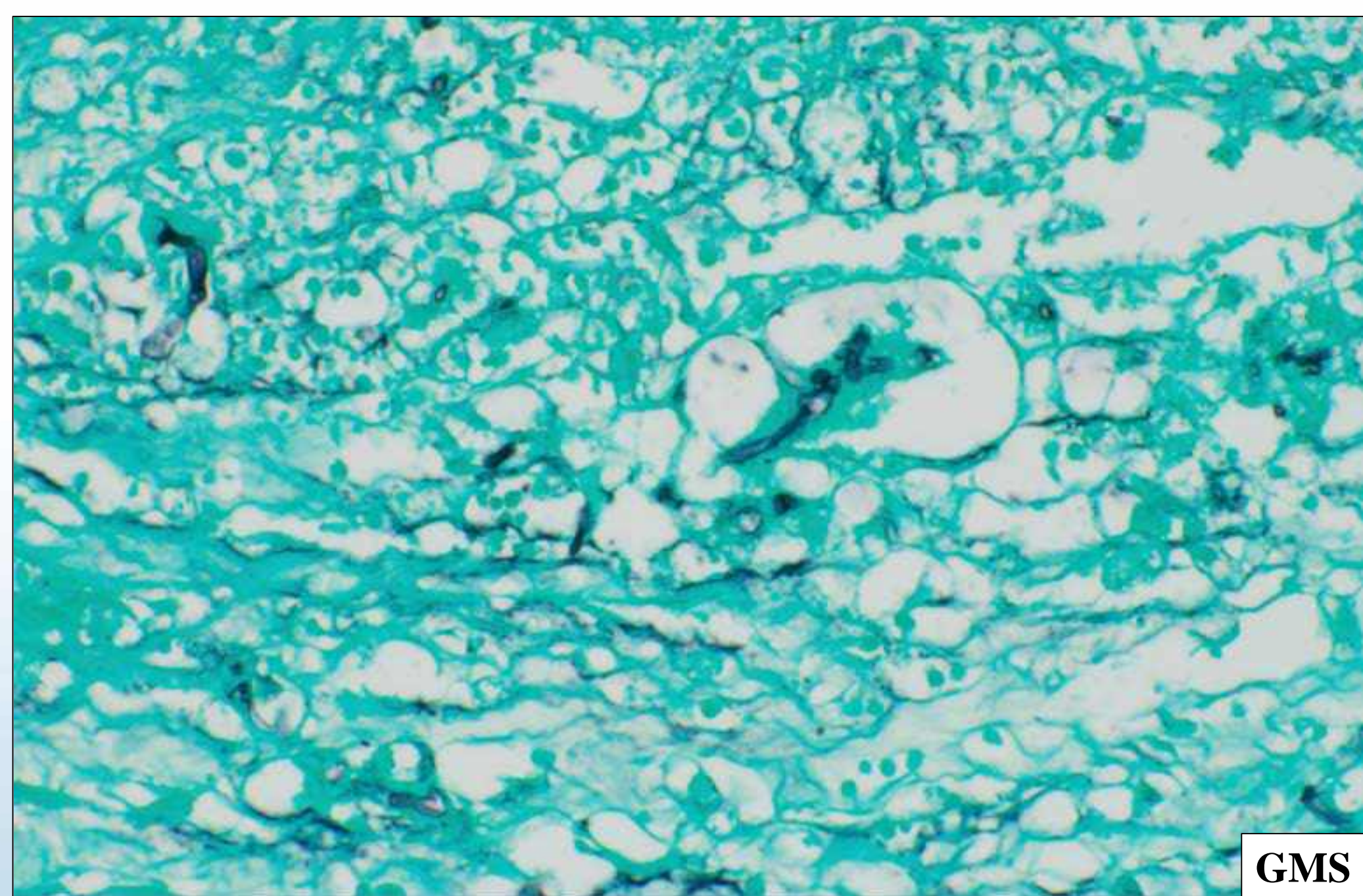




GMS



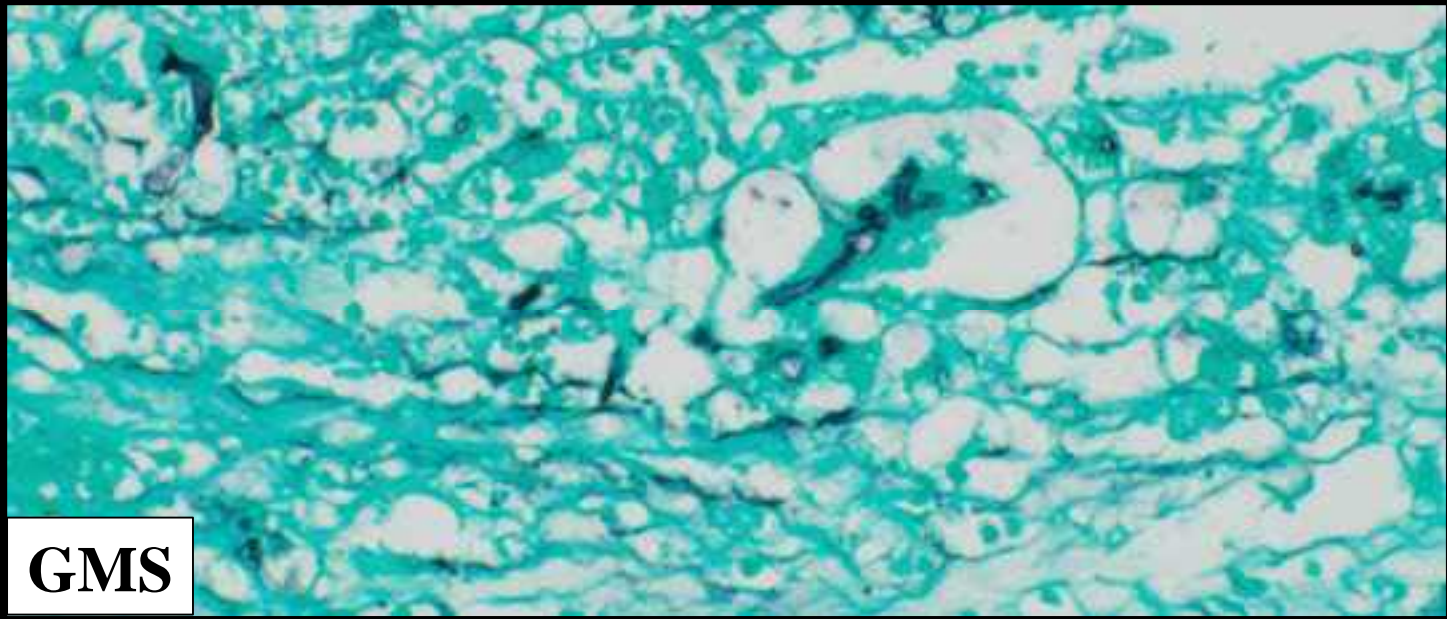
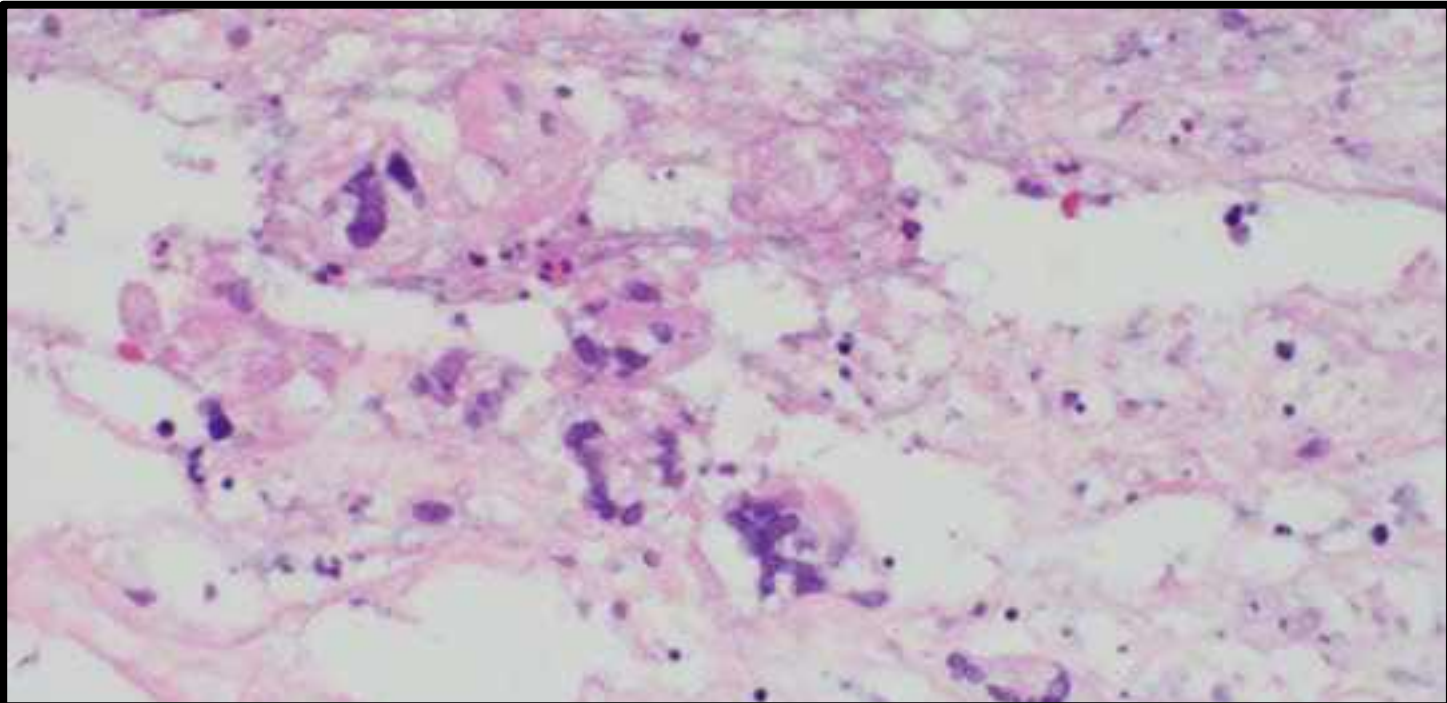
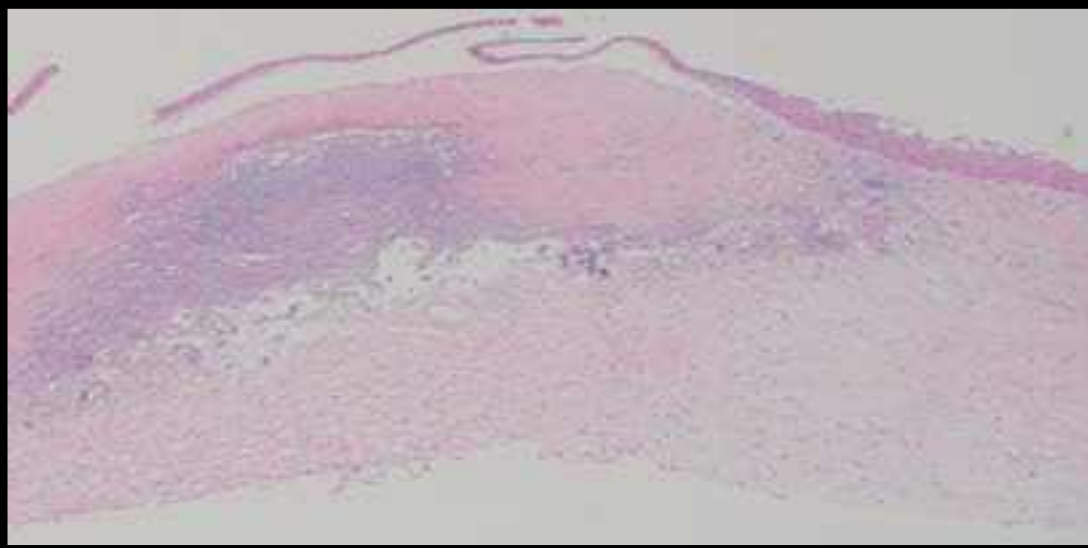
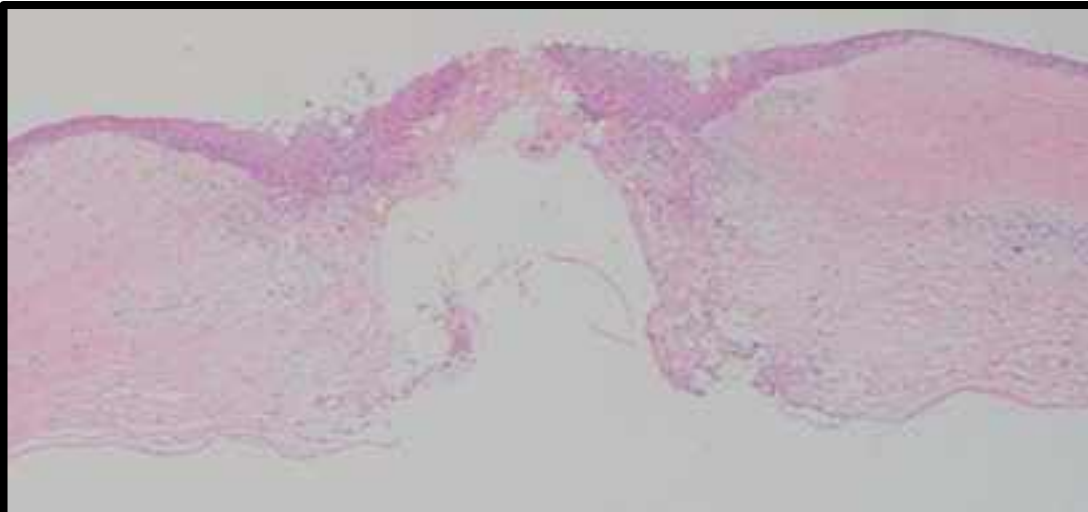
AANP



GMS



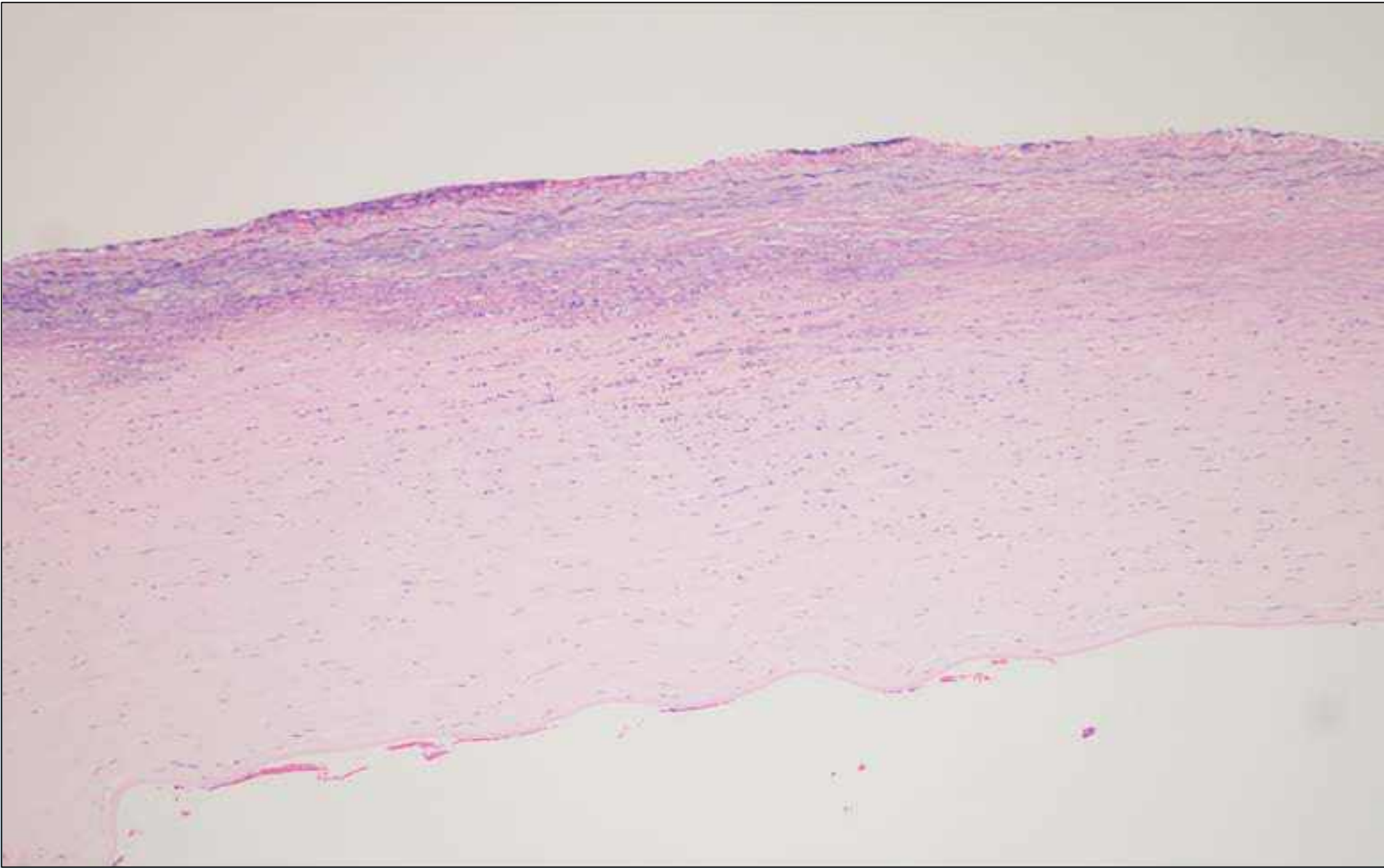
Fungal keratitis: Fusarium

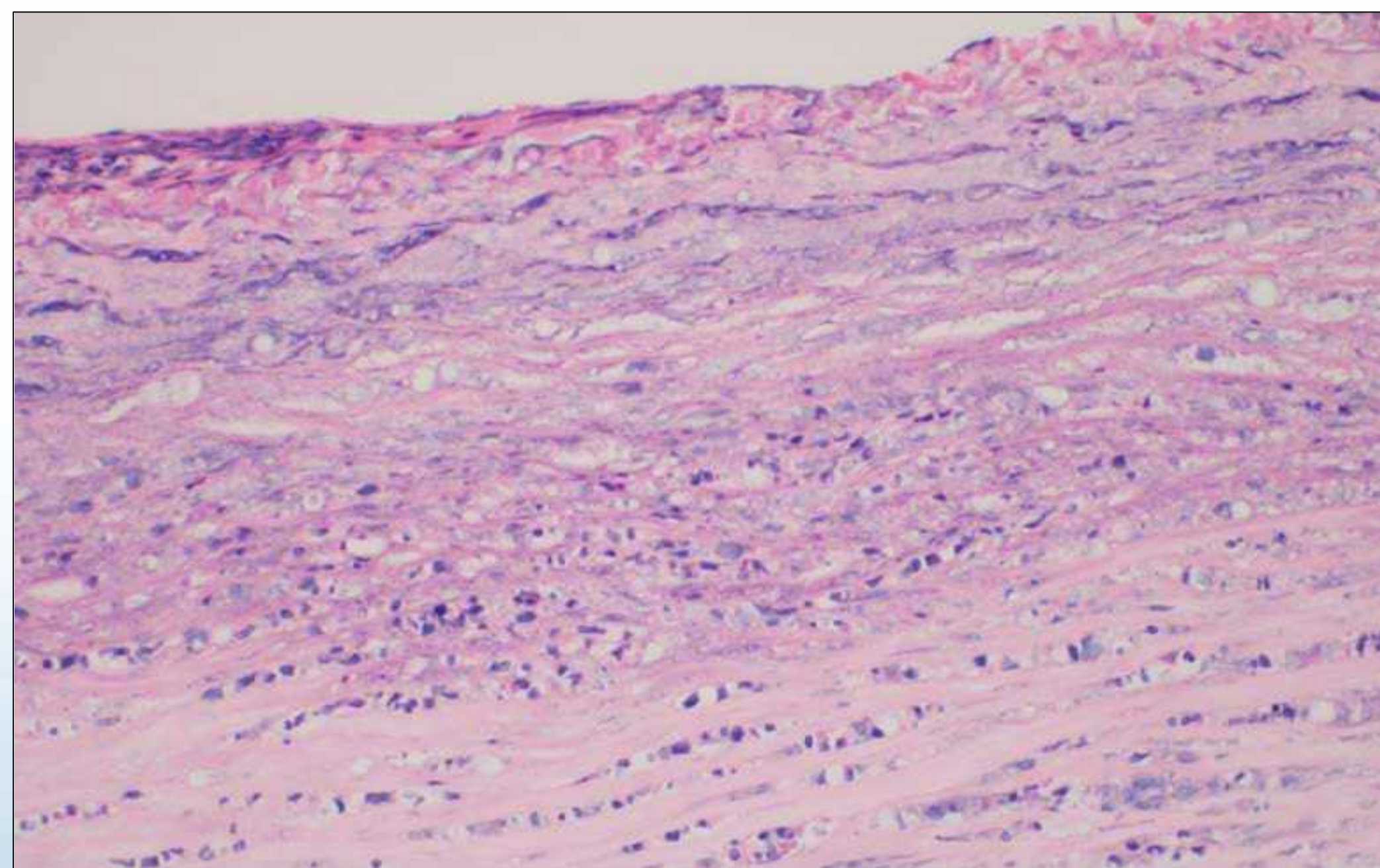


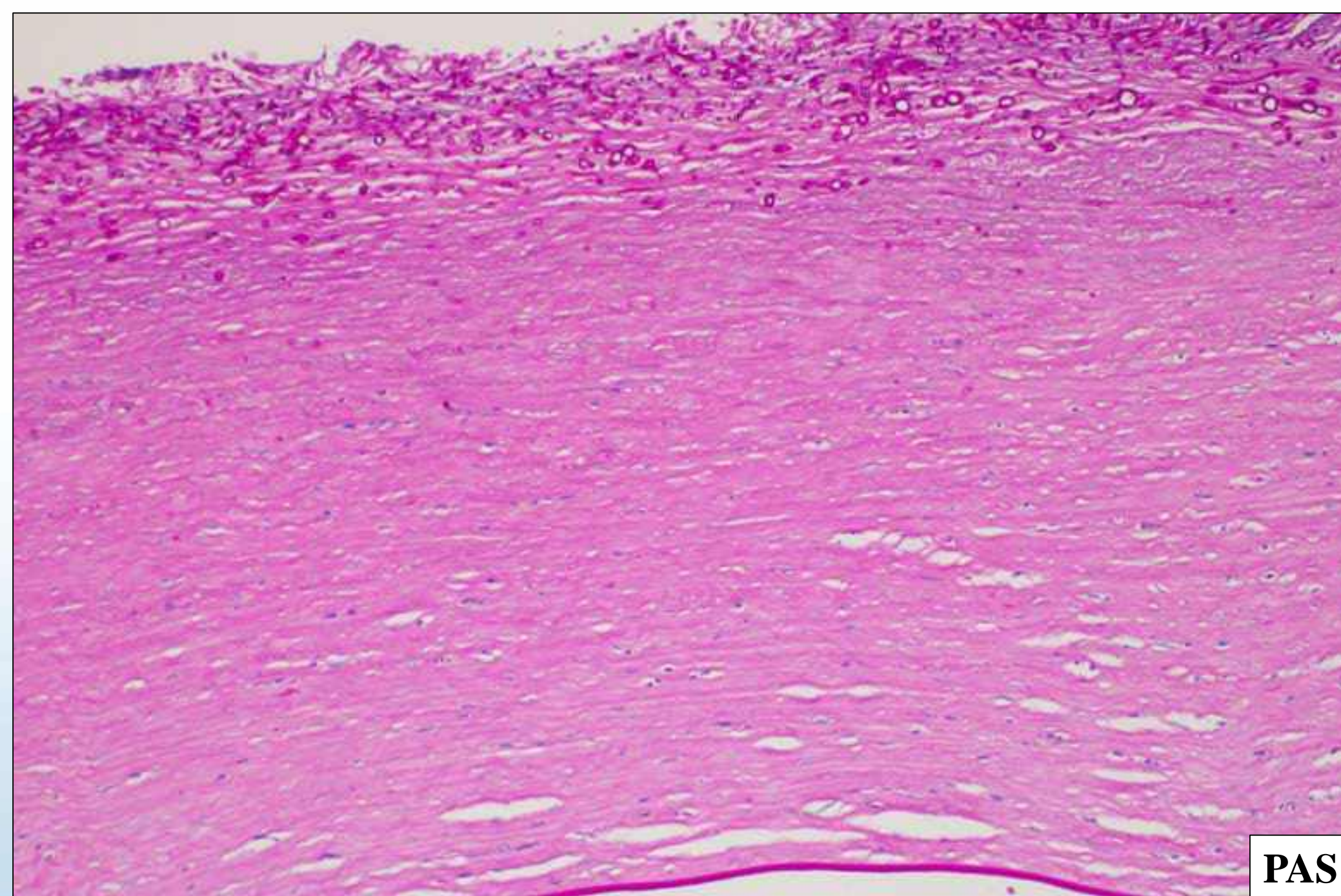
GMS

Corneal graft



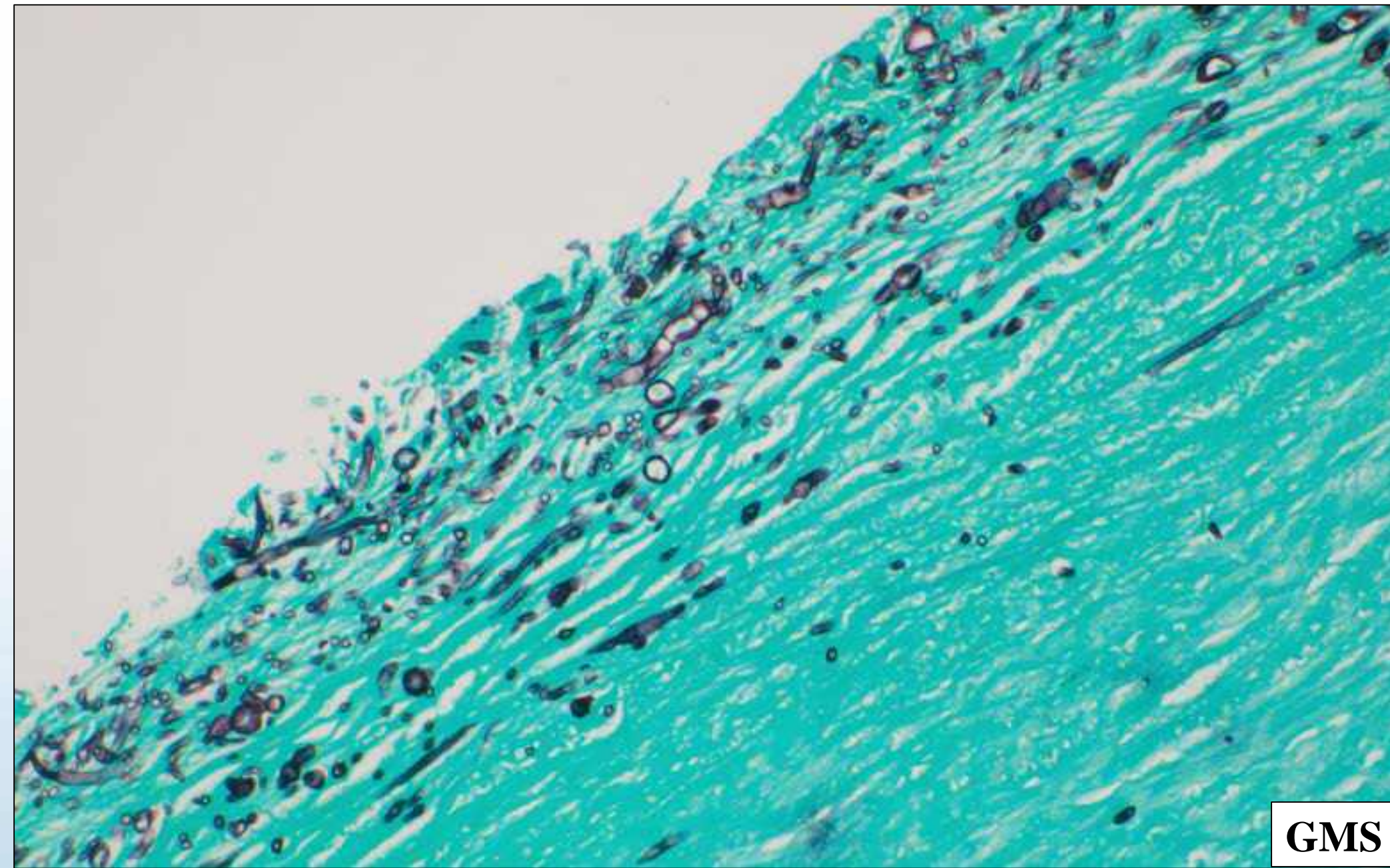






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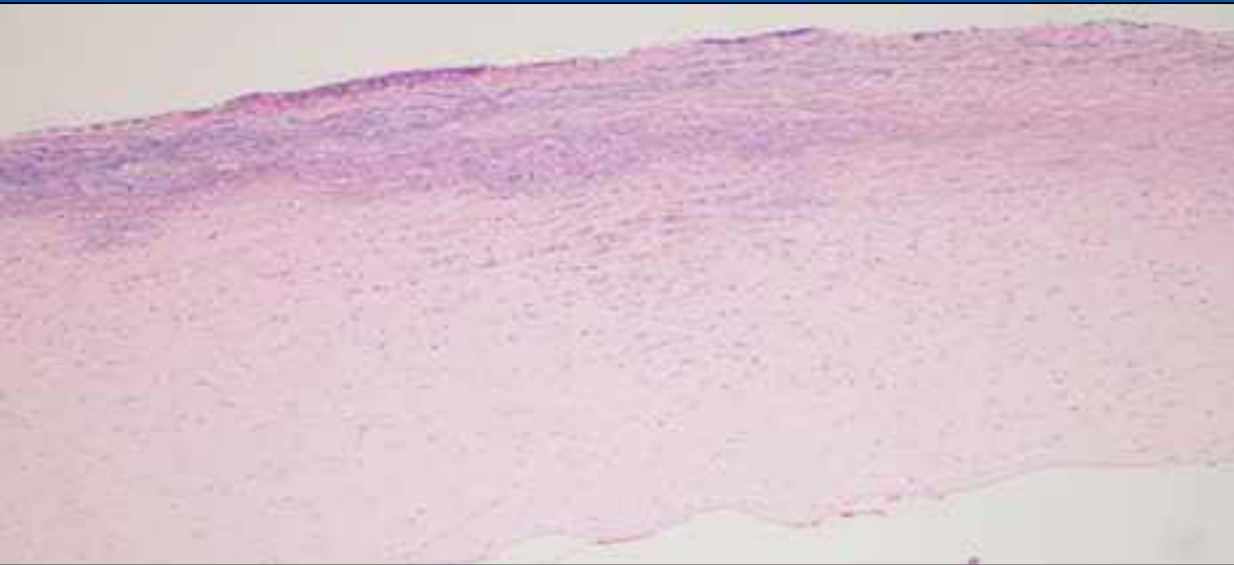


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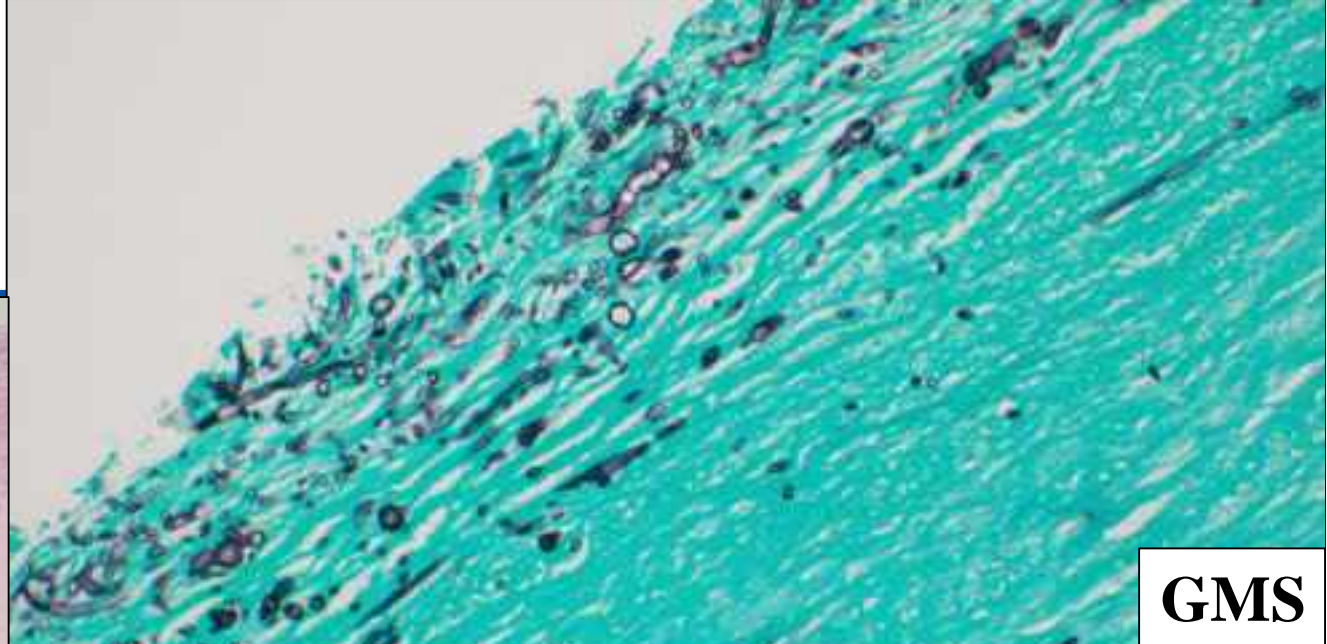




Fungal keratitis: Fusarium



PAS



GMS



Hilary Highfield
10/11/24

Fungal keratitis

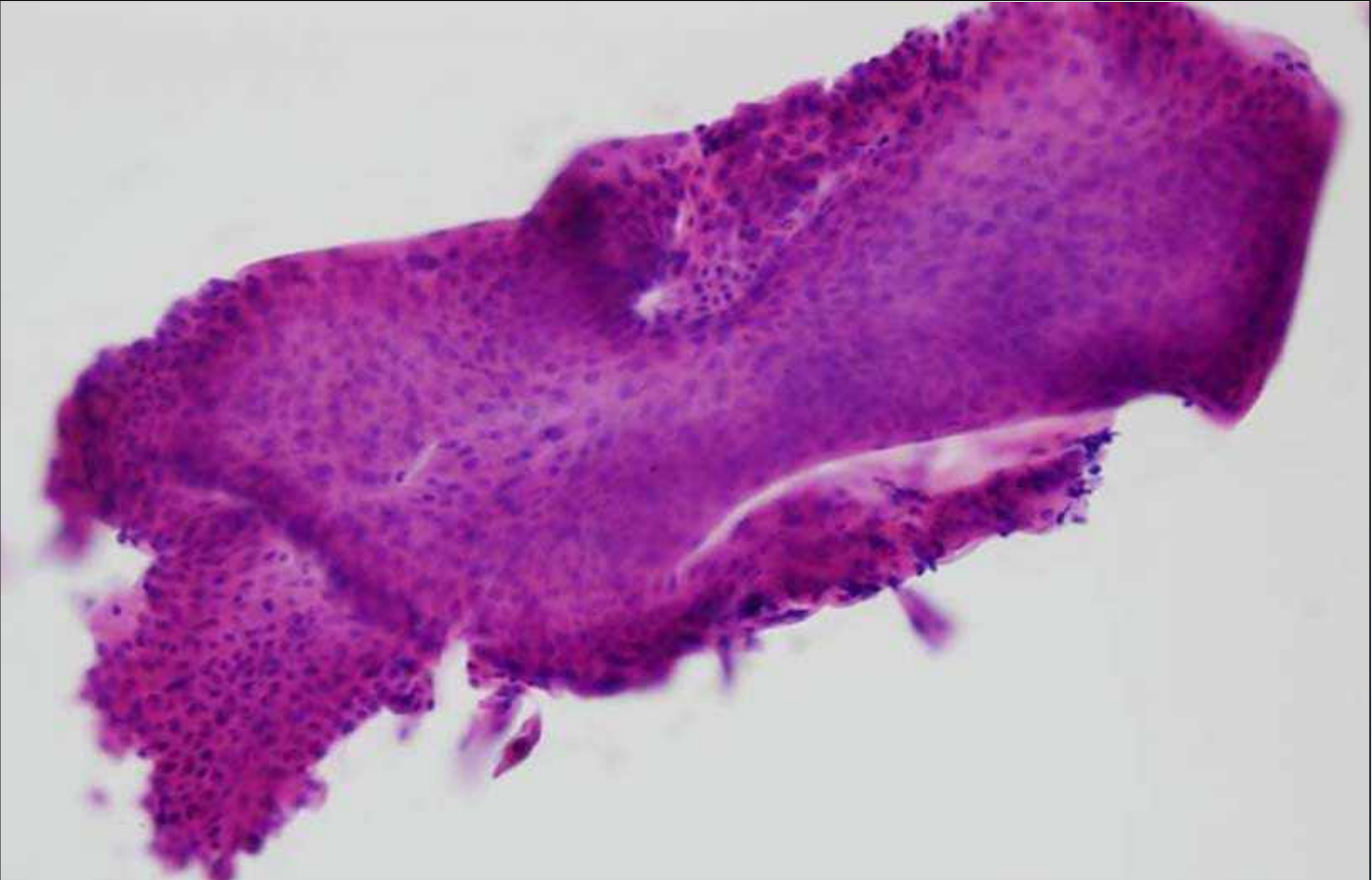
- More prevalent in South
- Rarer than bacterial
- Complicates corneal injury by vegetable matter
- Complicates steroid therapy in debilitated hosts
- 80% *Aspergillus*, *Candida*, *Fusarium*
- Deep crater with raised edges

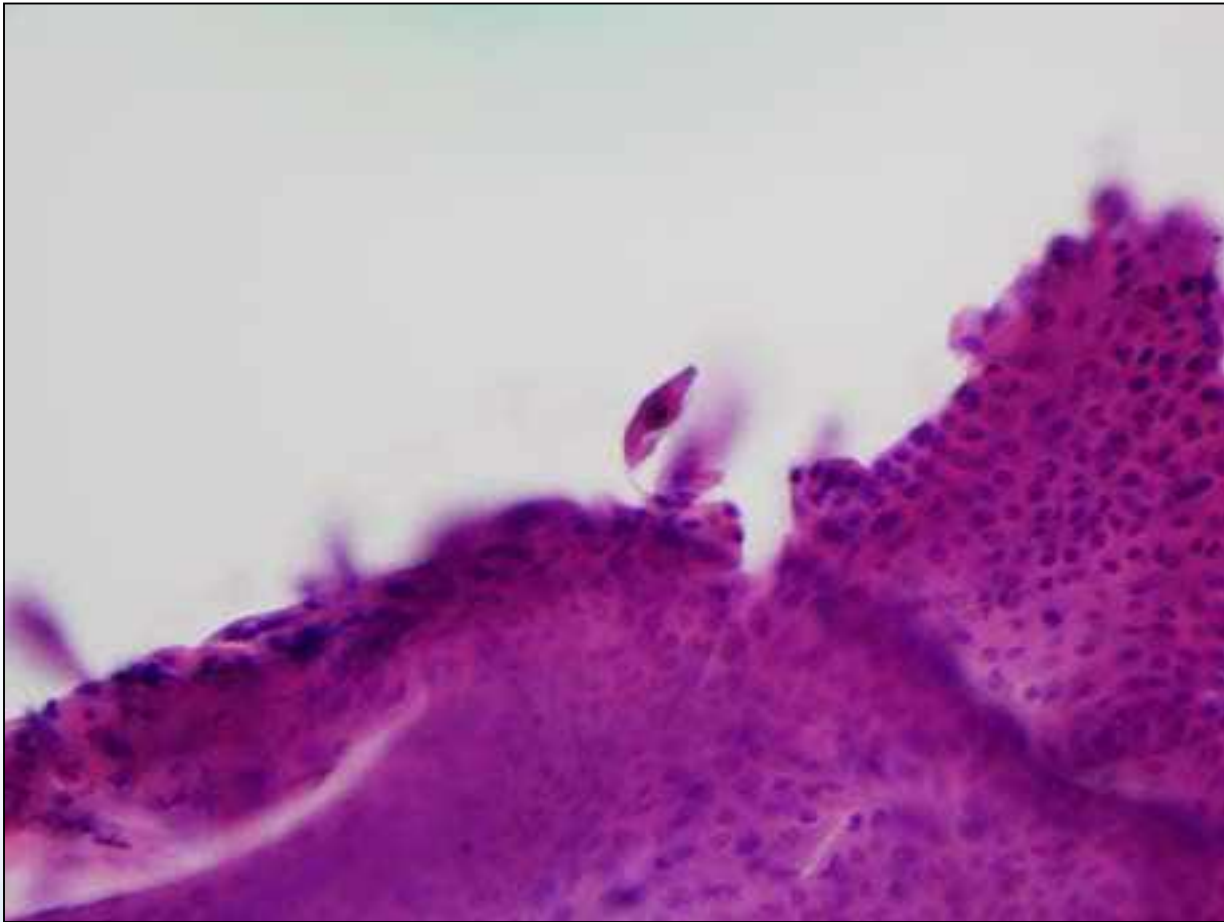
Clin Ophthal 2011; 5: 275-279.

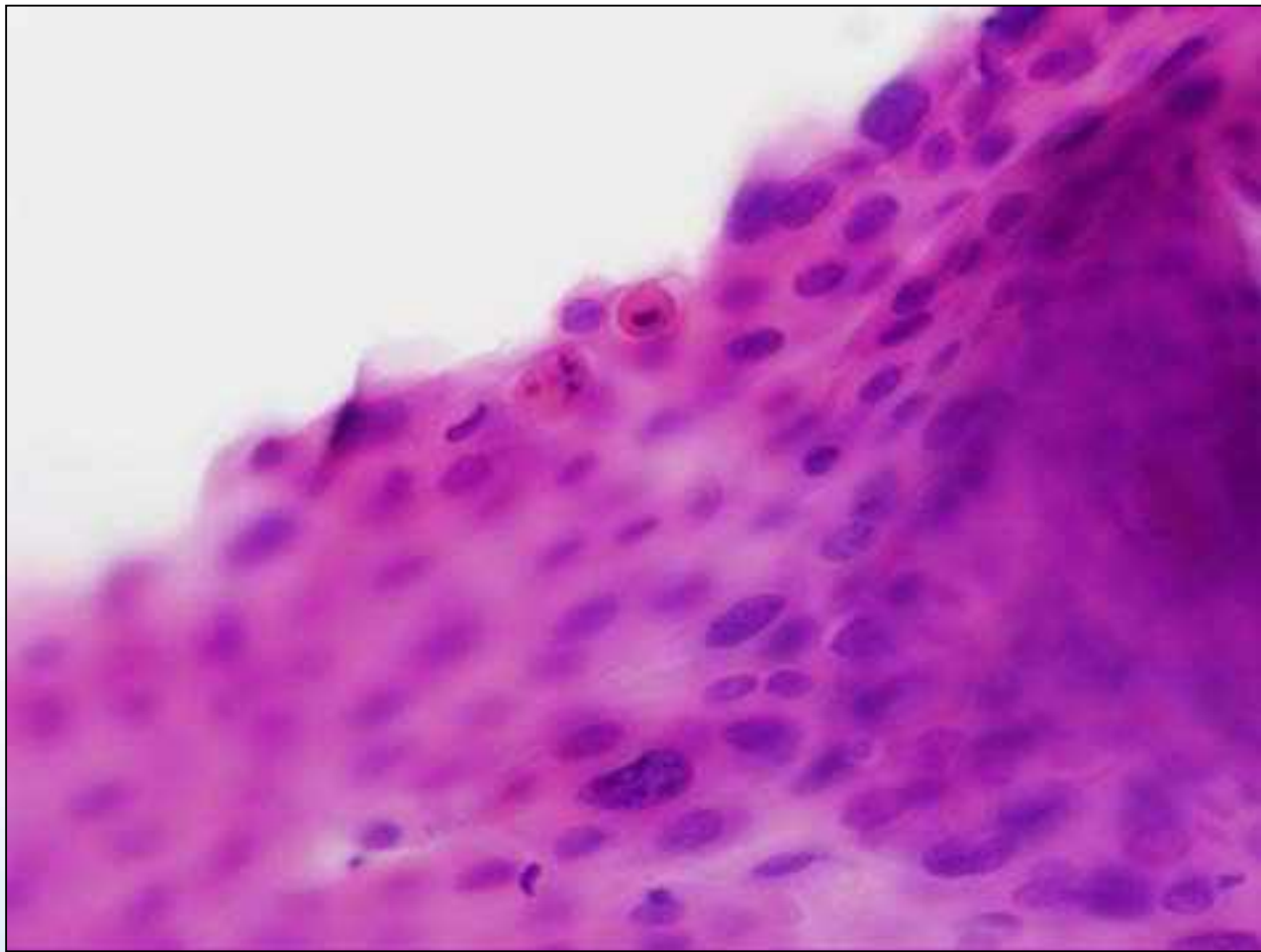


Contact lens wearing patient with corneal lesion

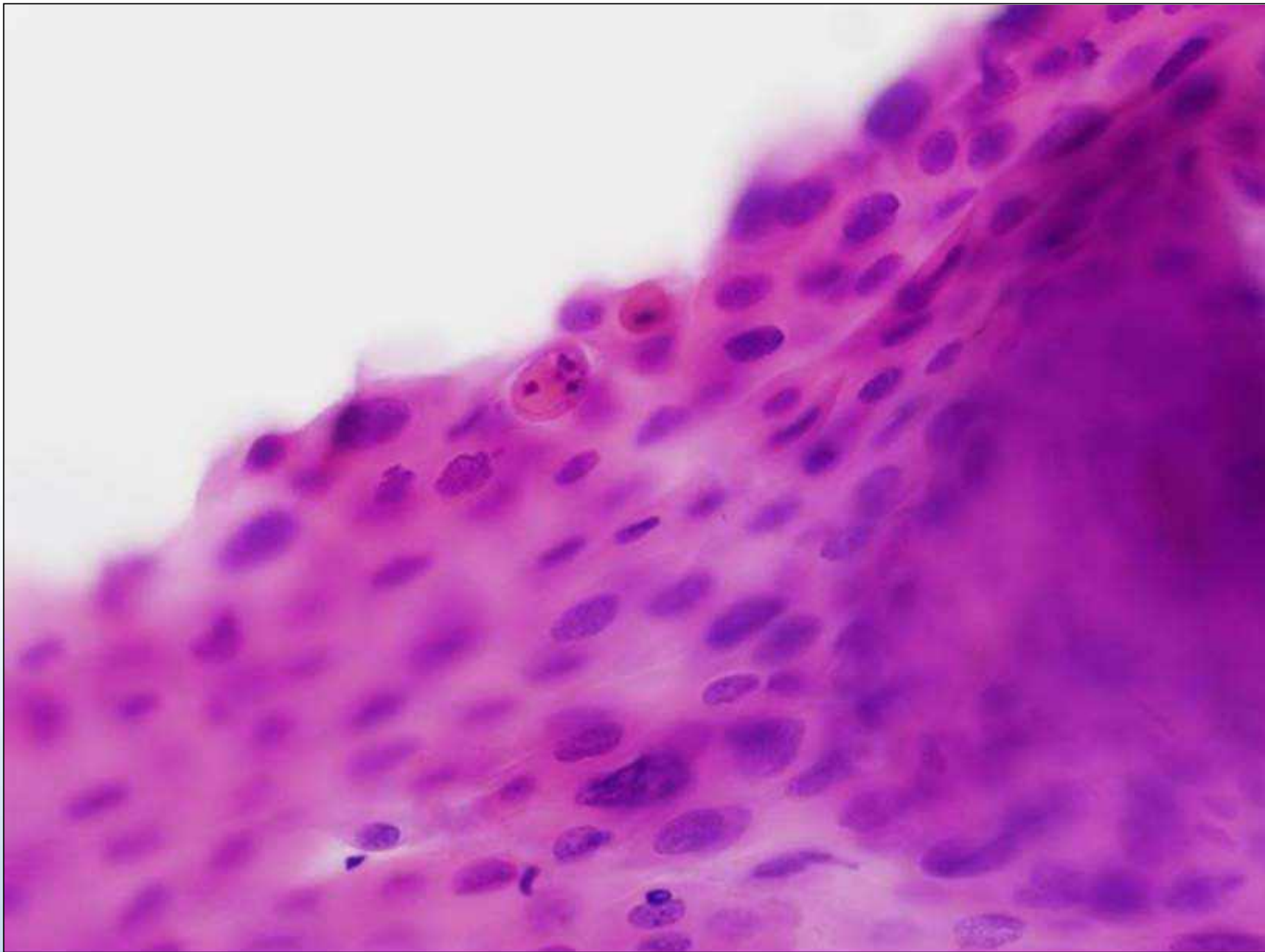








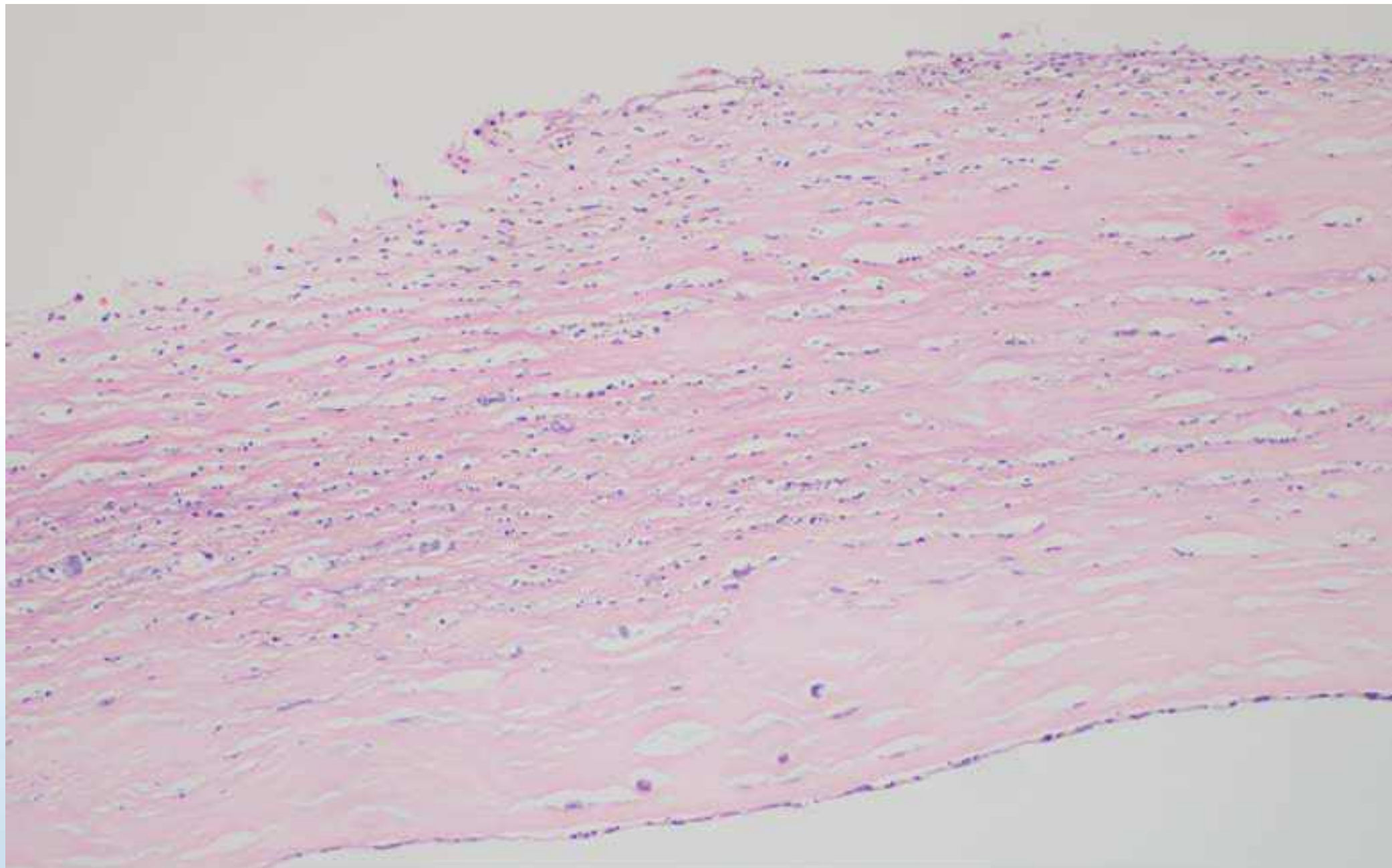
AANP

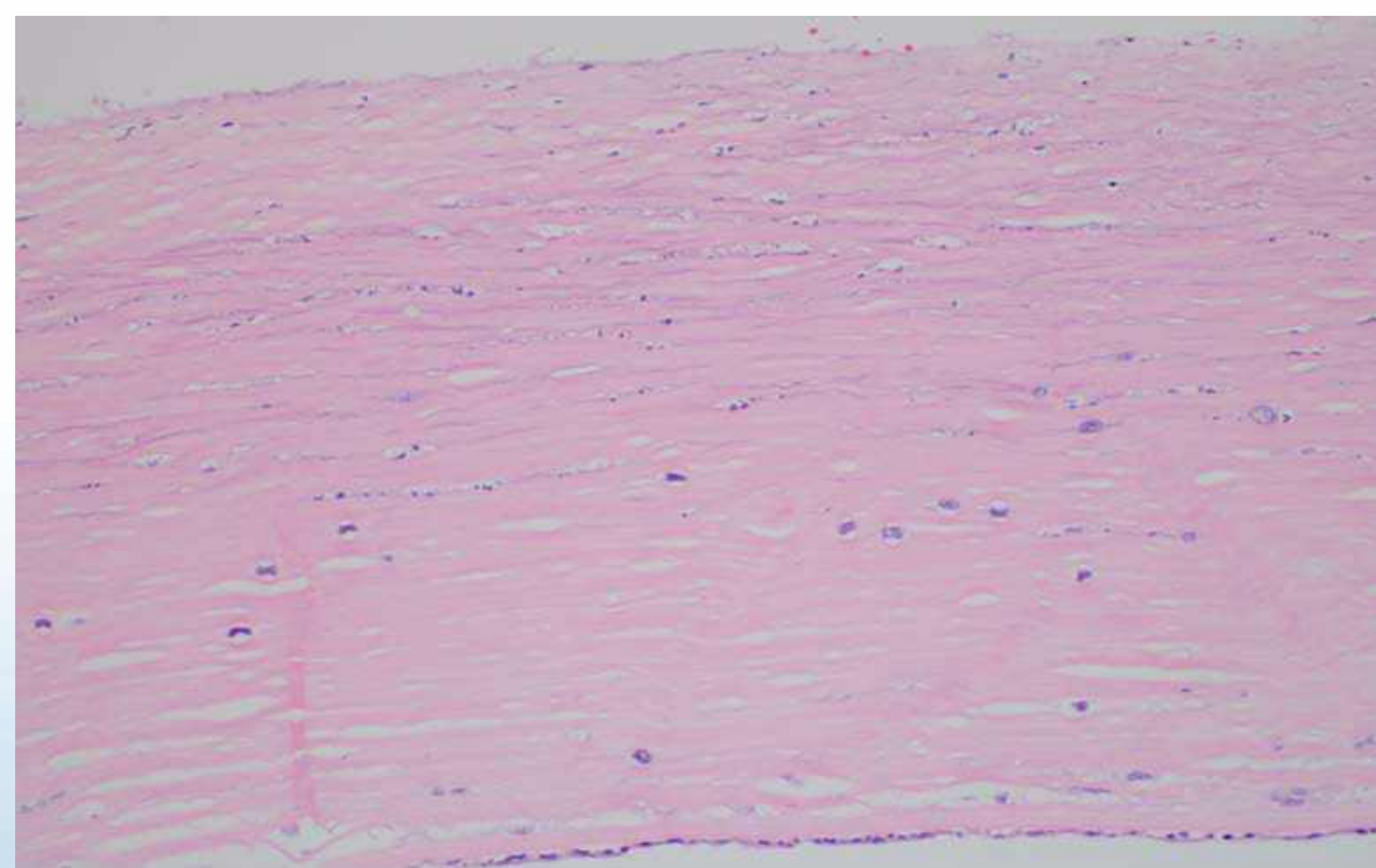


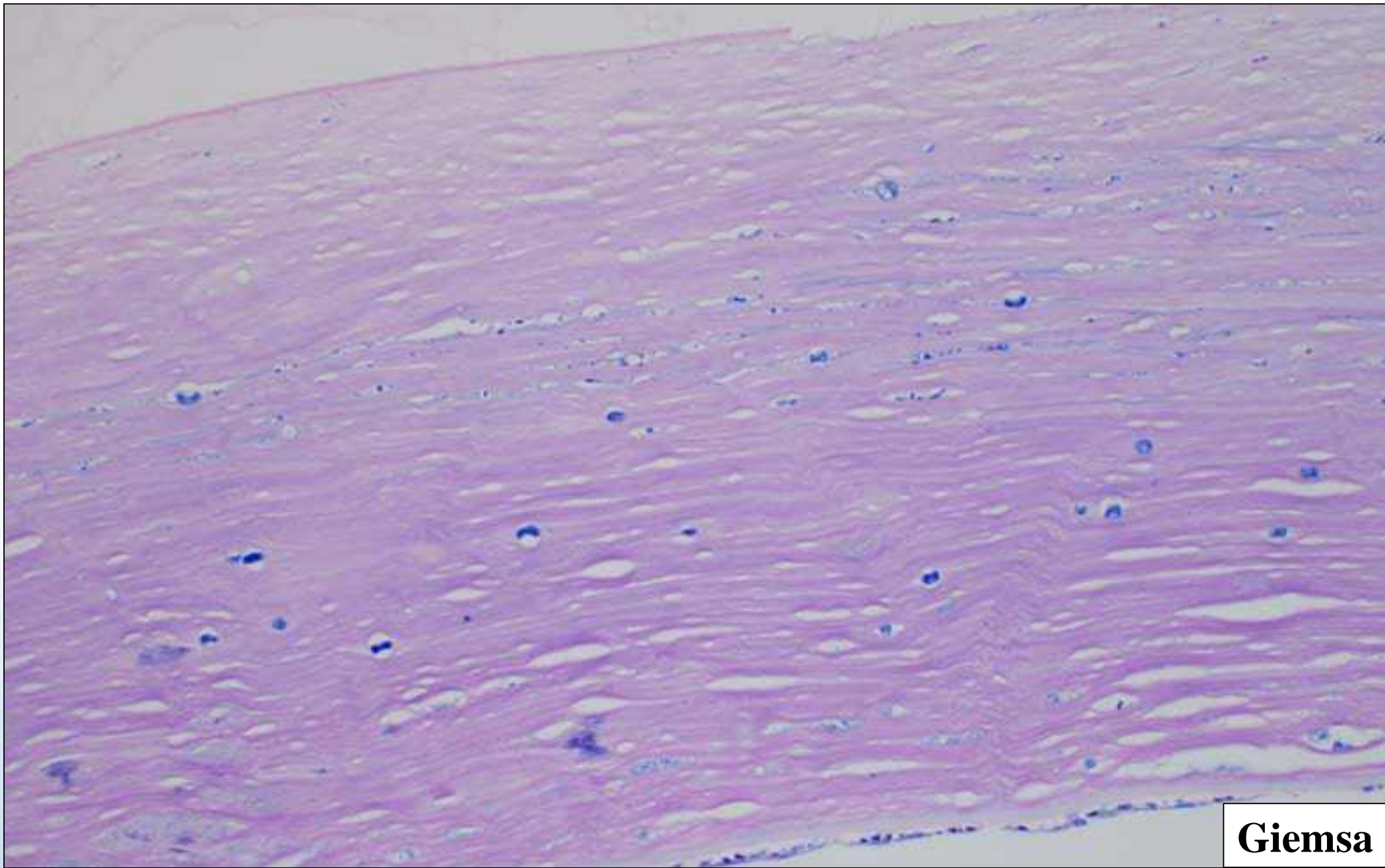


CDC: broad illumination with slit beam; high magnified slit beam, Typical advanced ring infiltrate.





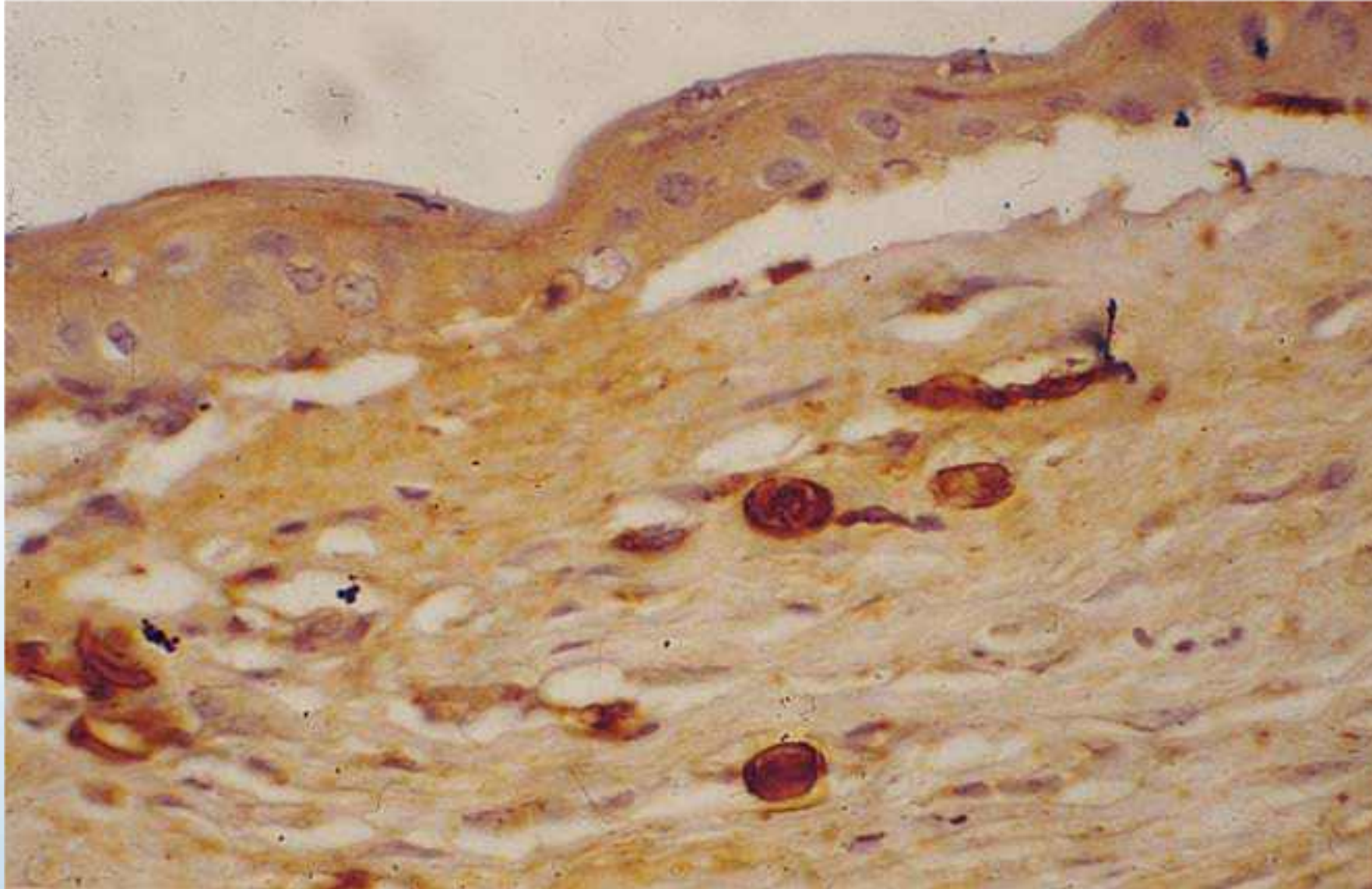




Giemsa



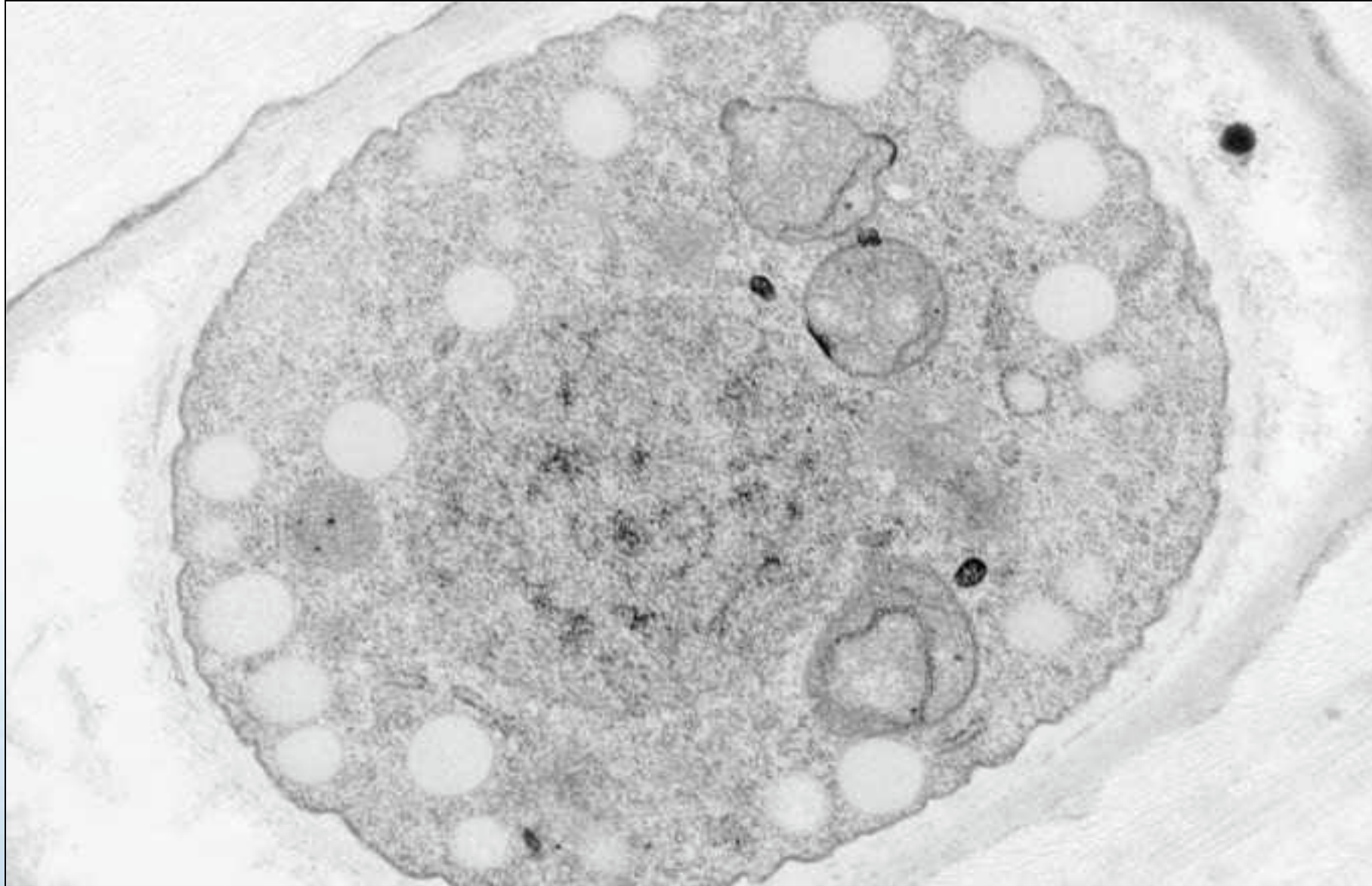
Immunohistochemistry



Sur ophthal (1998) 42:493



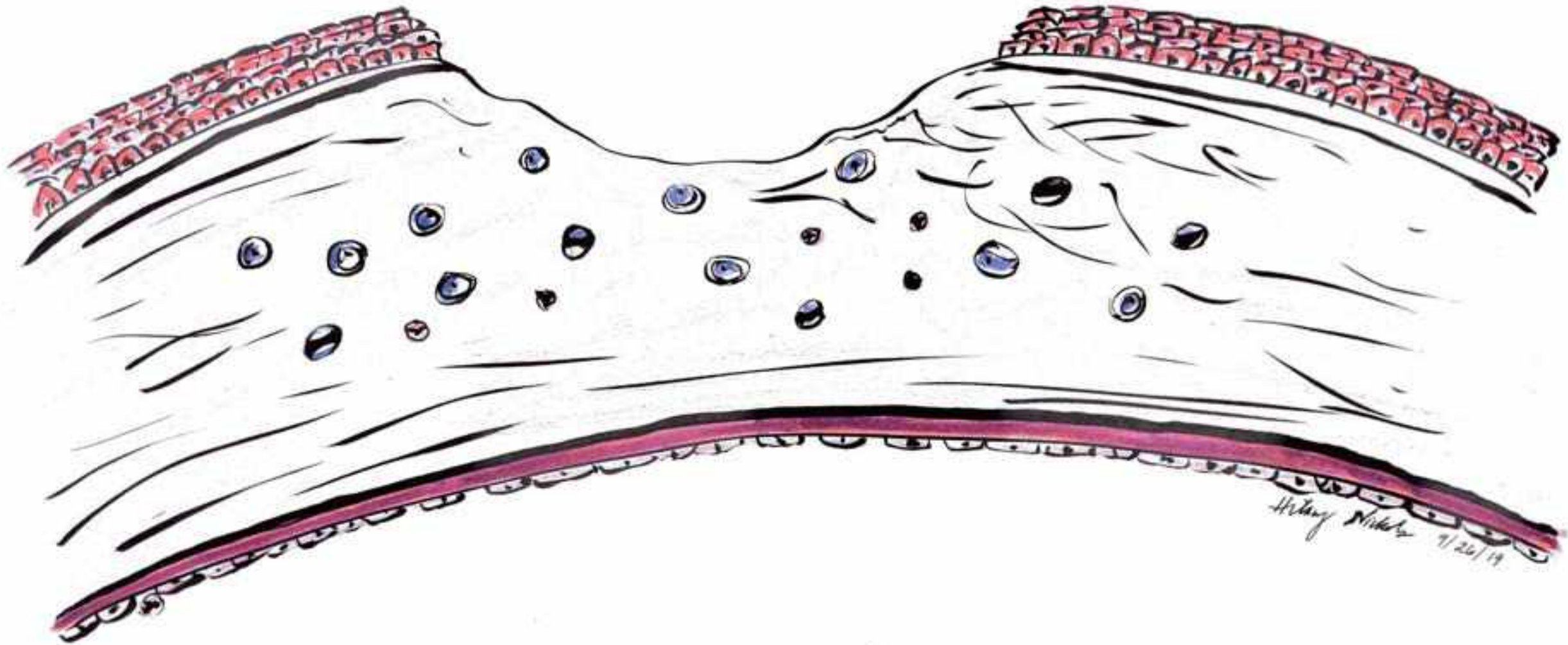
Electron microscopy



Sur ophthalm (1998) 42:493



Acanthamoeba keratitis



Acanthamoeba keratitis

- Protozoan parasite: cyst and trophozoite
- *A castellani* and *A polyphagia* most commonly involved species
- Closely linked to soft contact lens use
- Severely painful
- Interstitial keratitis: non-ulcerative stromal inflammation



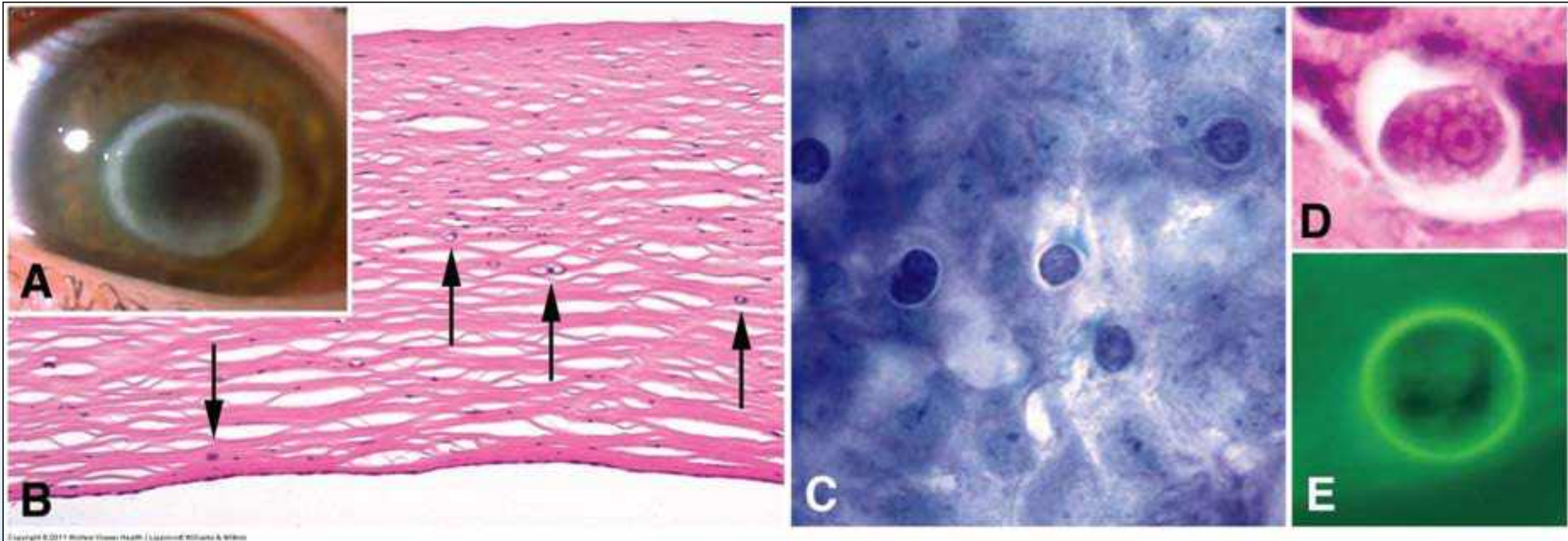
Acanthamoeba keratitis

- First described 1970s
- Dramatic increase 1980s with soft contact lens wear
- Risks:
 - Contact lens wear: 80%
 - Frequent-replacement soft contacts
 - Overnight orthokeratology patients
 - Swimming, tap water cleaning
- DDx includes *herpetic* and *fungal* keratitis

Curr Opin Ophthalmol 2006; 17:327-331.

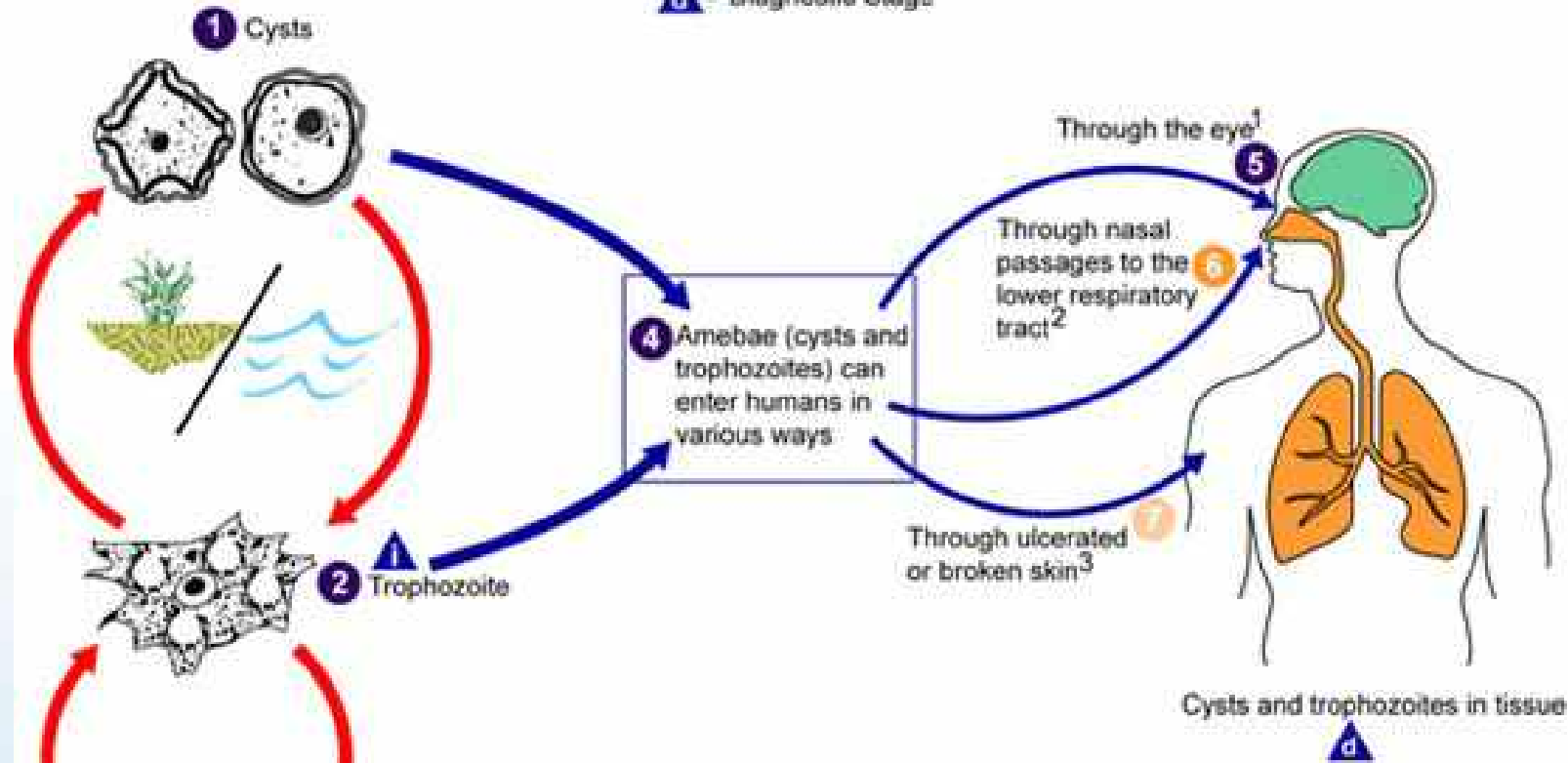


Acanthamoeba keratitis



Acanthamoeba lifecycle

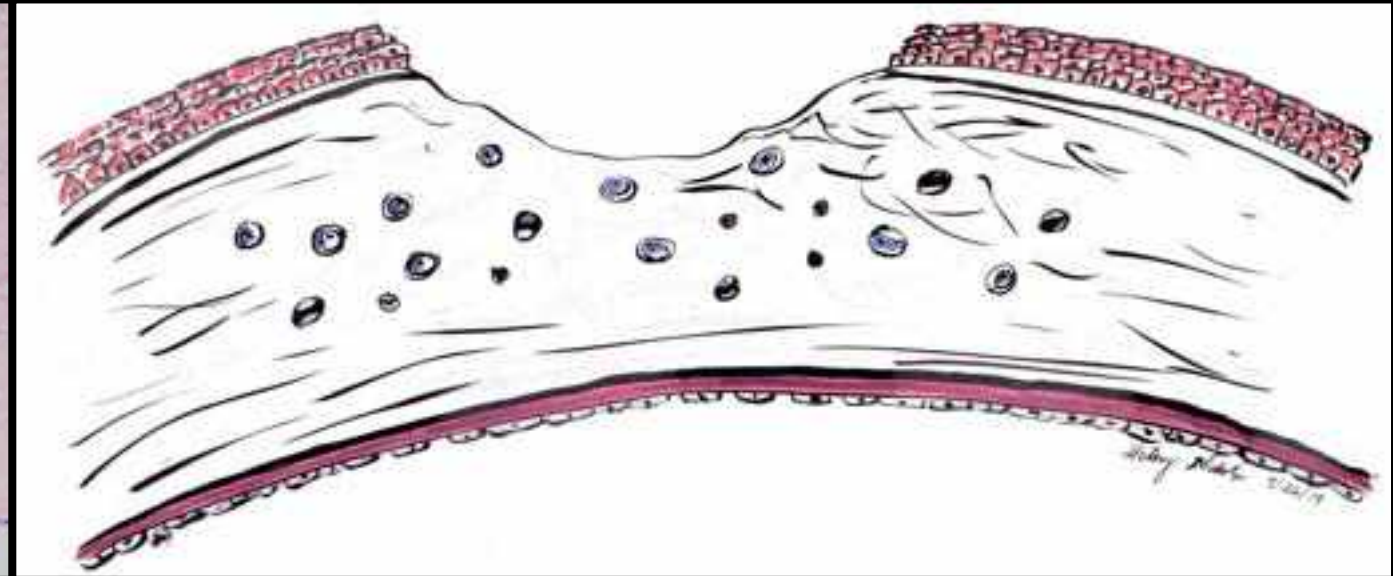
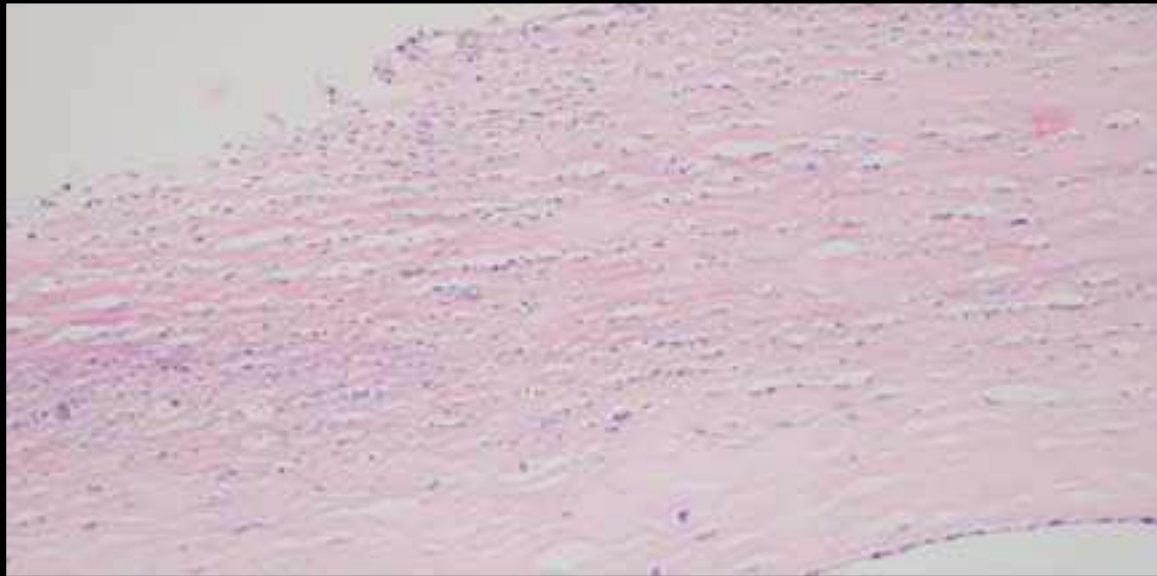
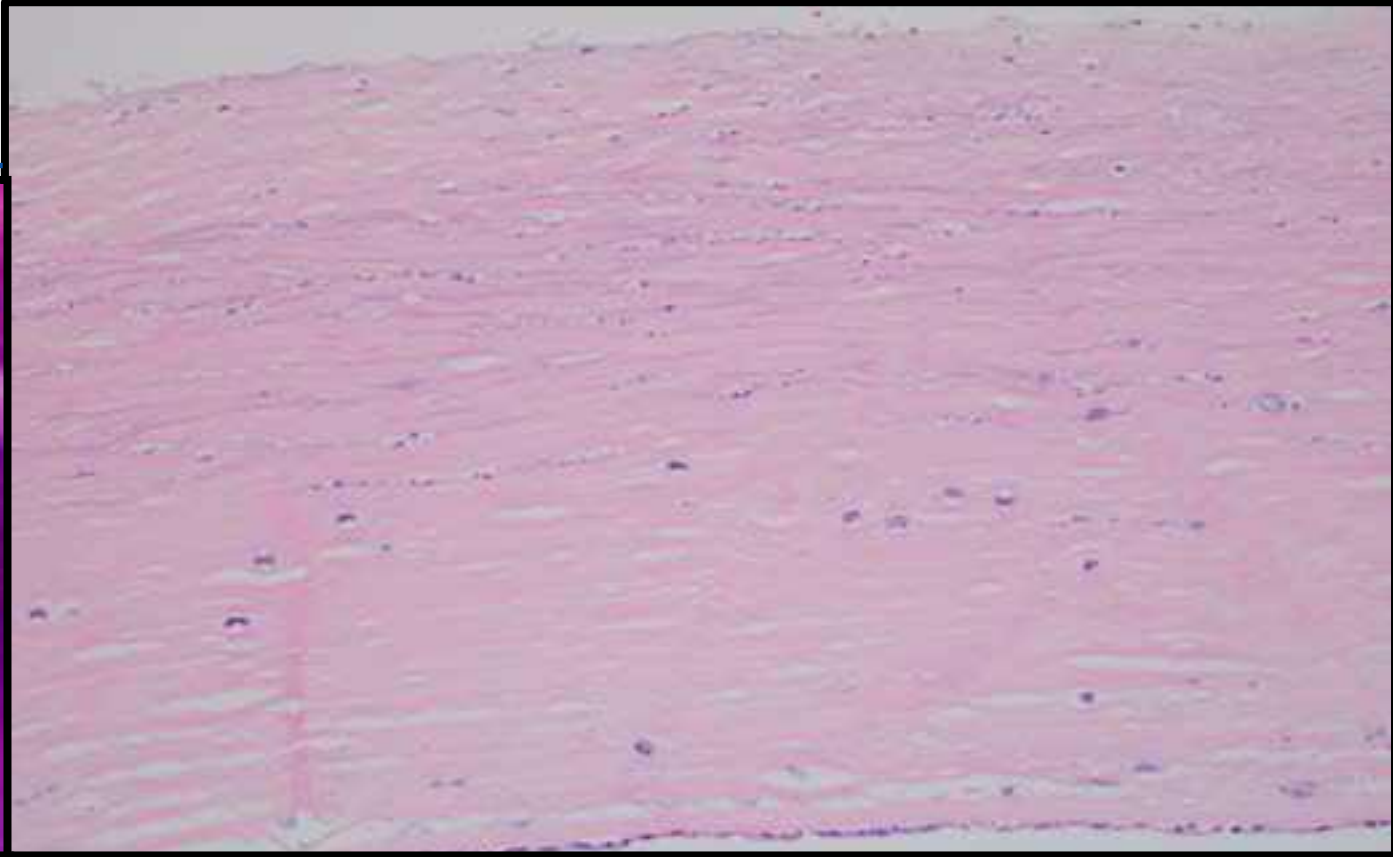
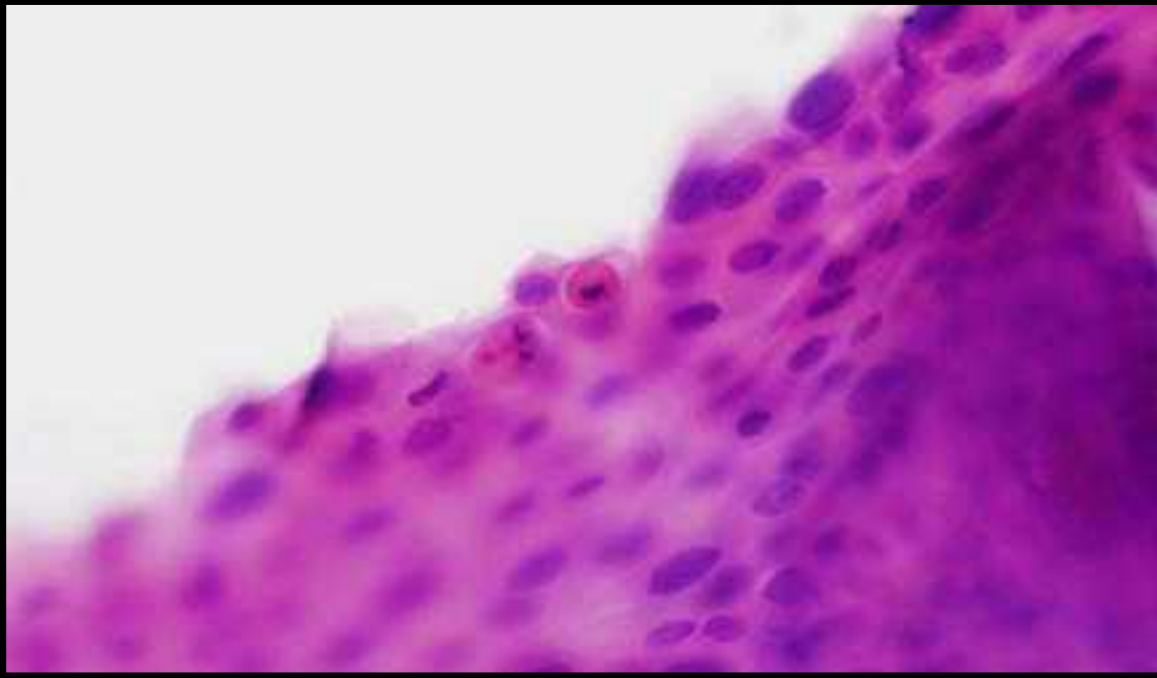
▲ = Infective Stage
 ▲ = Diagnostic Stage



- ¹ Results in severe keratitis of the eye. ⁸
- ² Results in granulomatous amebic encephalitis (GAE) and/or disseminated disease ¹⁰ in individuals with compromised immune systems. ⁹
- ³ Results granulomatous amebic encephalitis (GAE), disseminated disease ¹⁰, or skin lesions individuals with compromised immune systems. ¹¹



Acanthamoeba keratitis

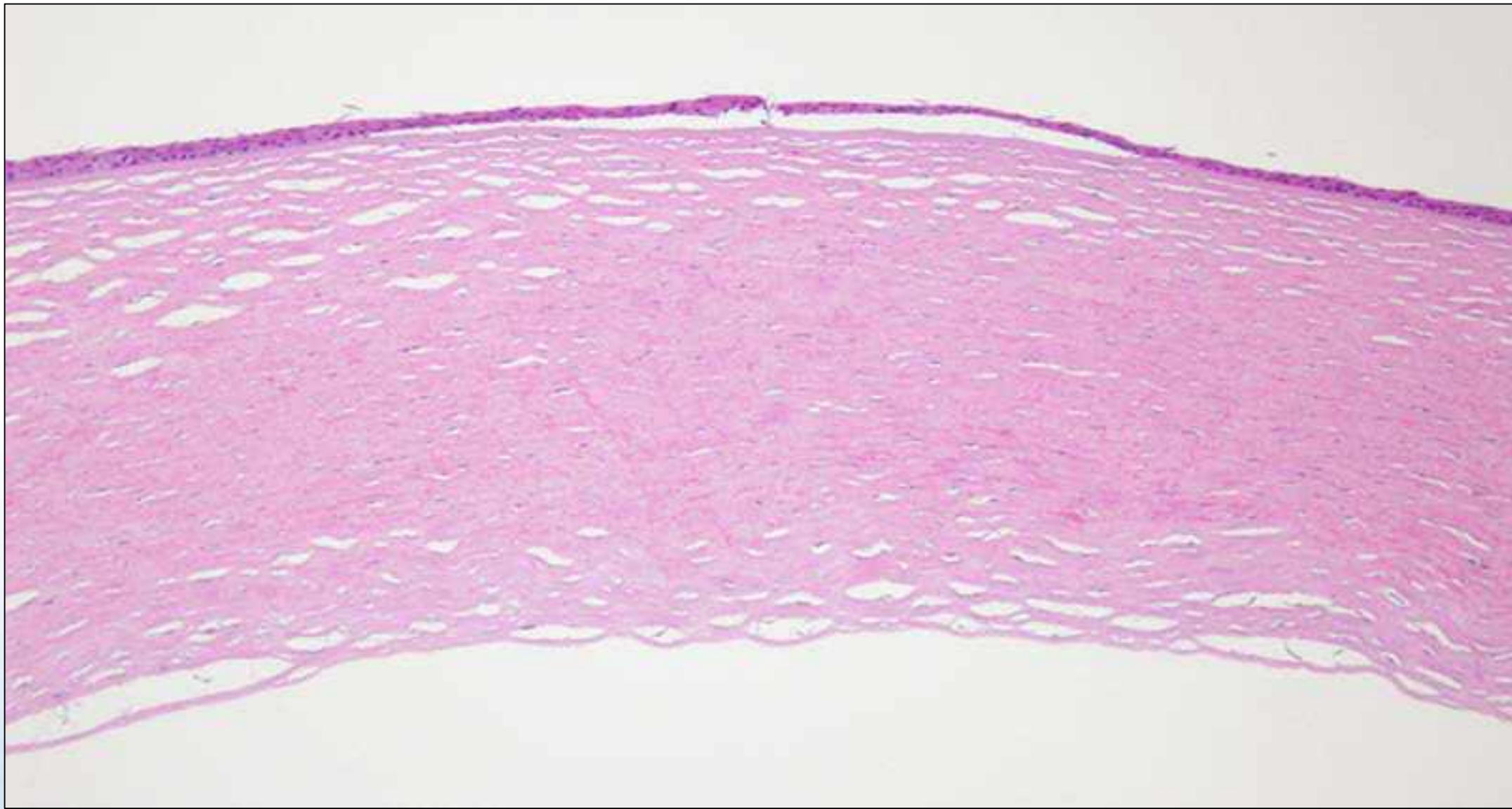


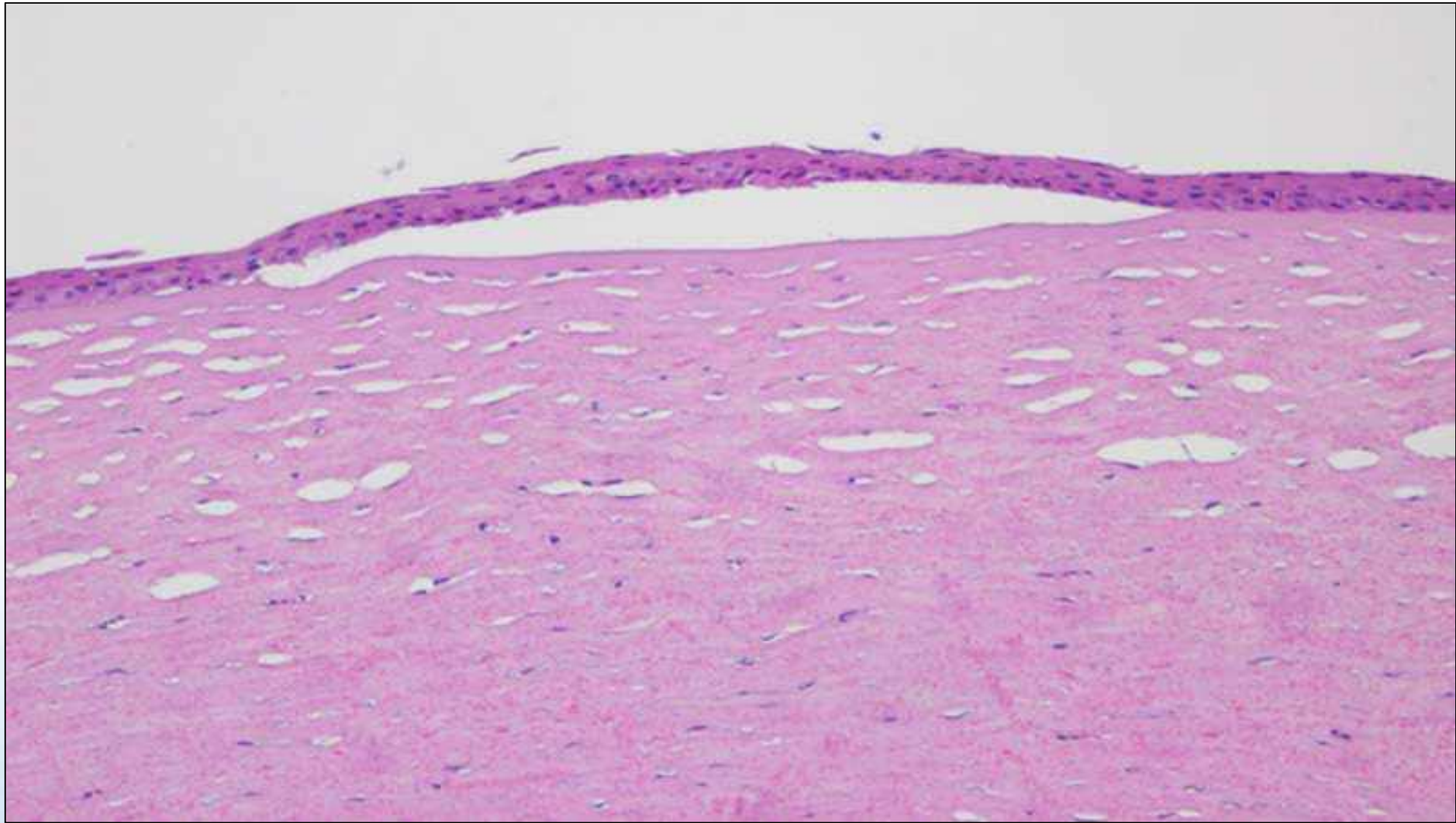
Hyphema





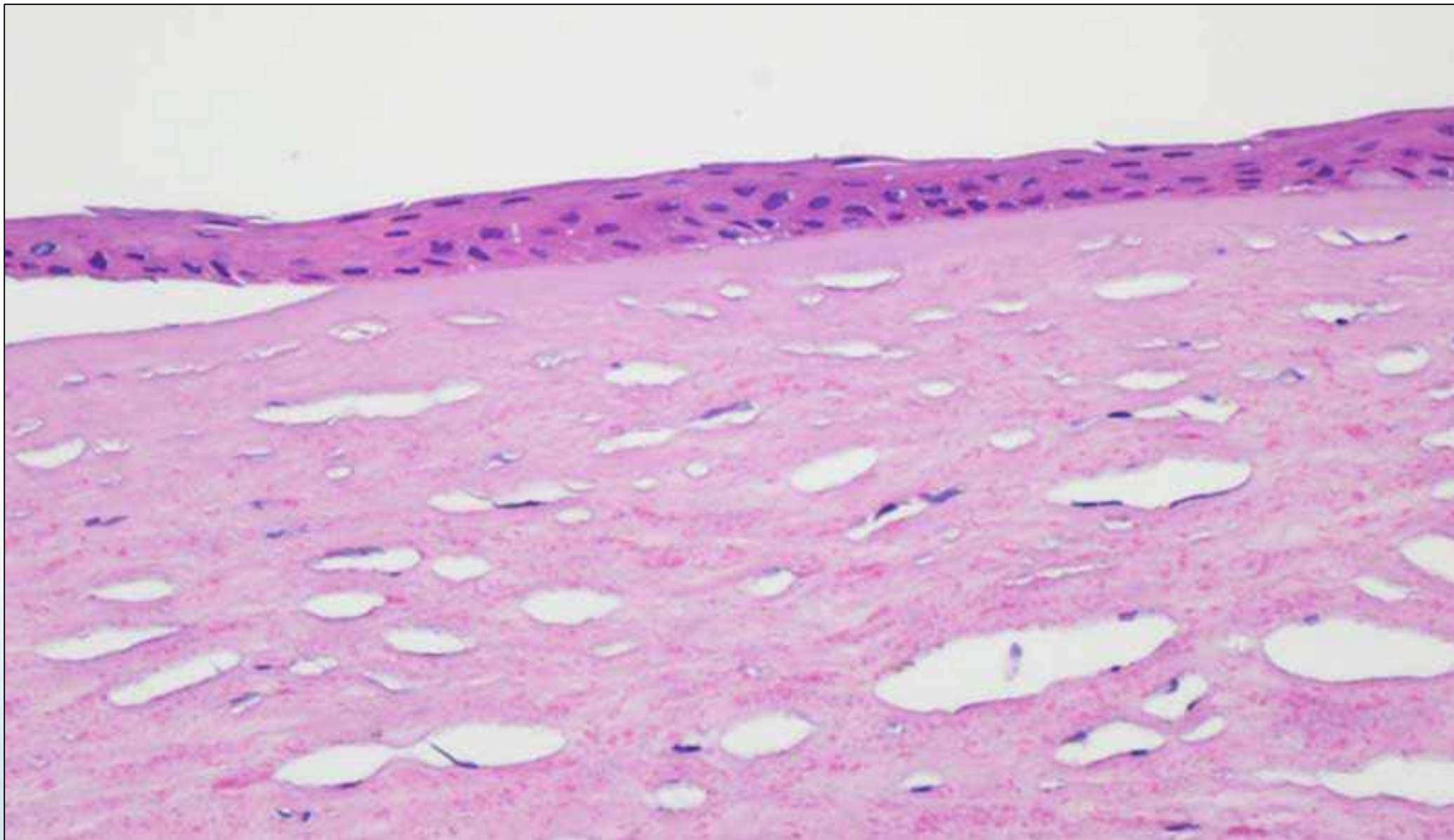




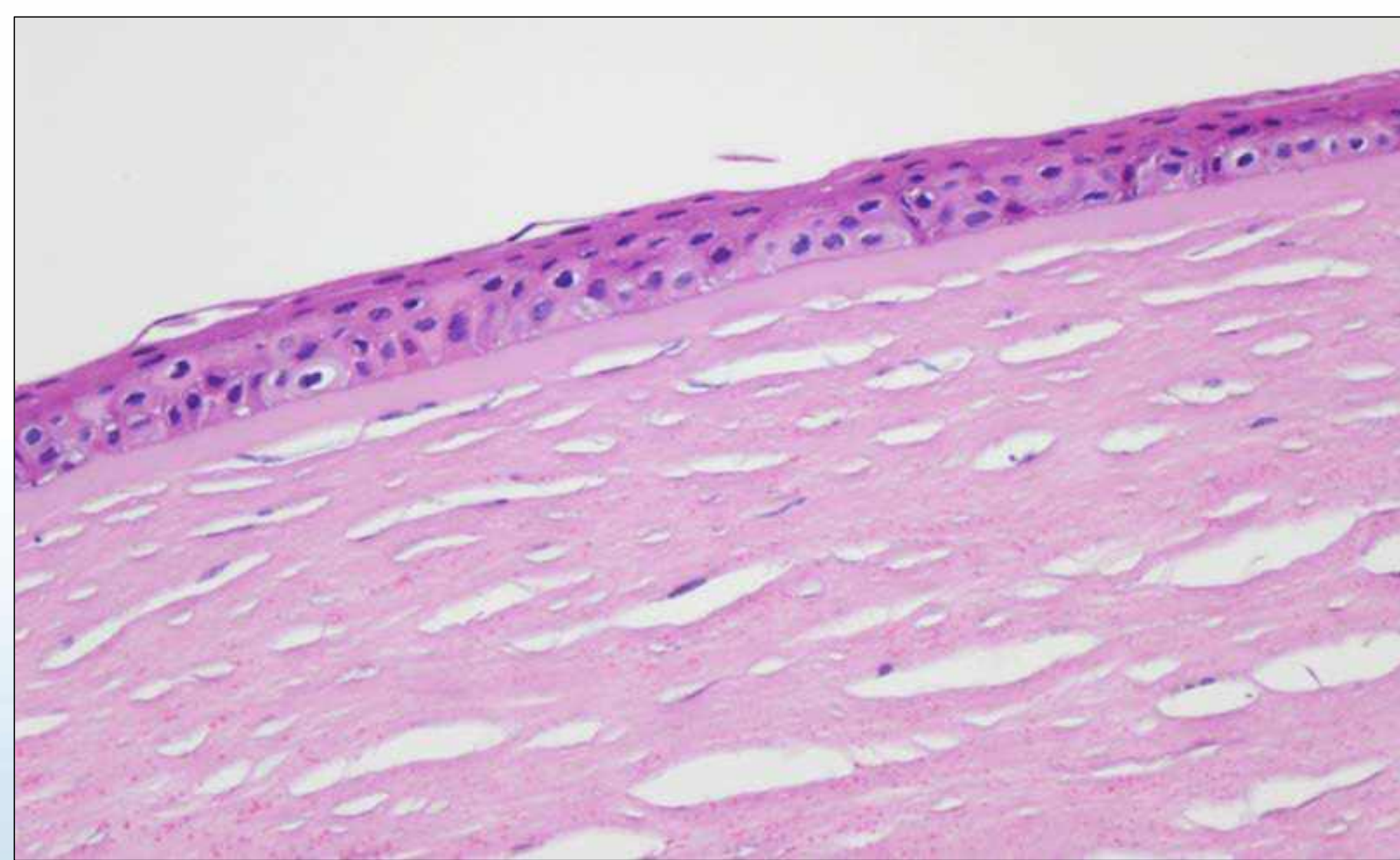


Bullae AANP

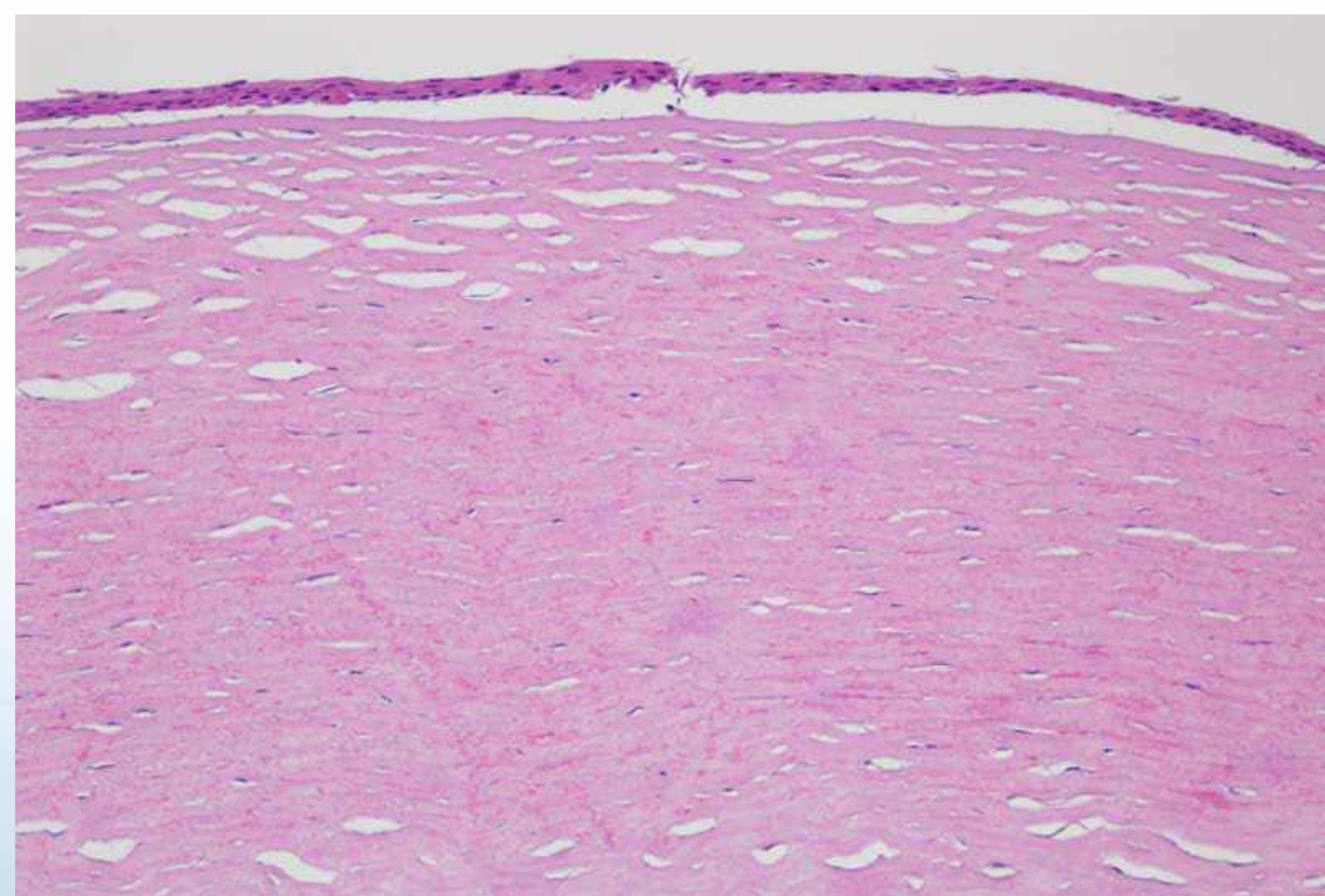




Subepithelial vacuoles

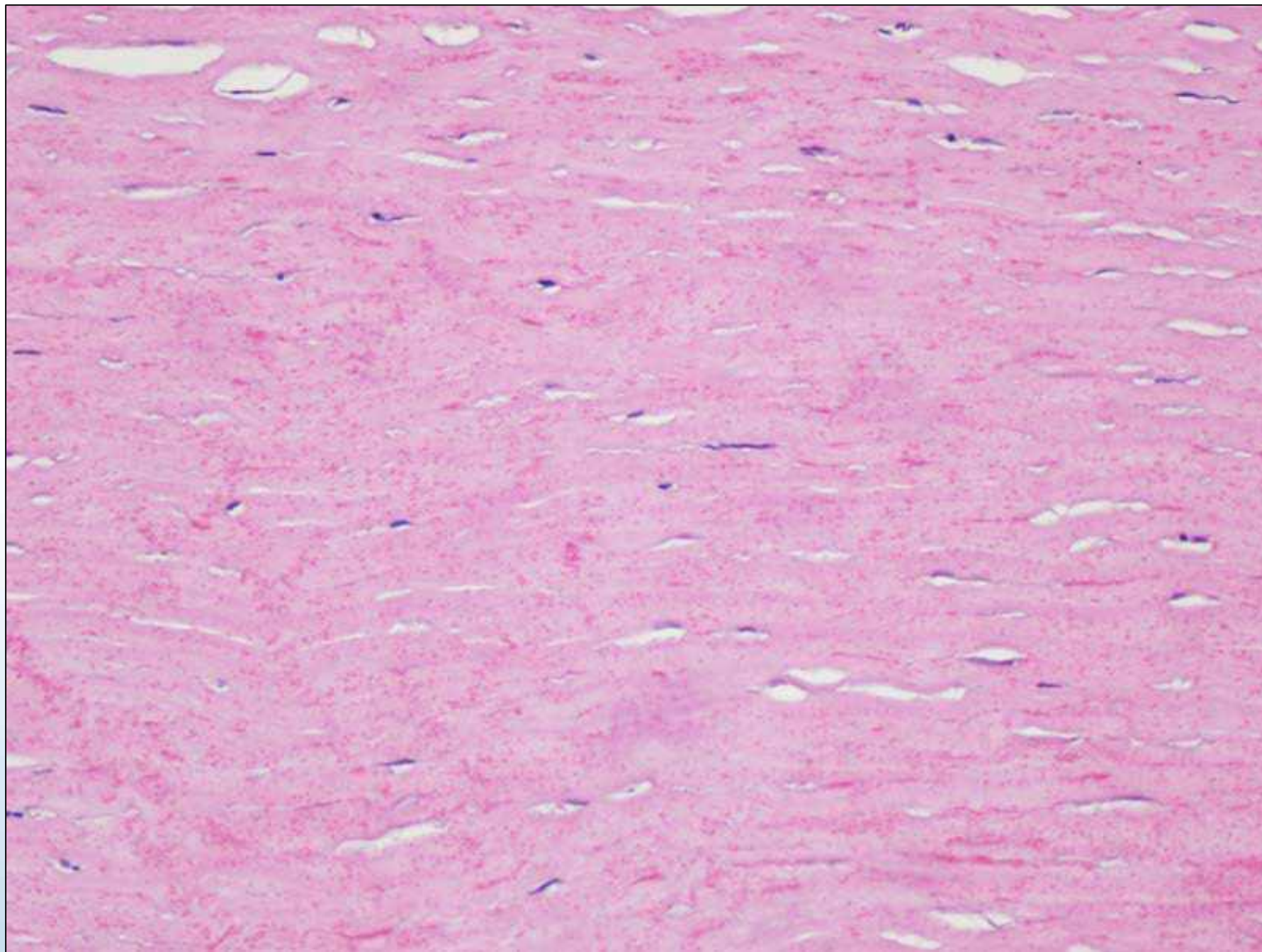


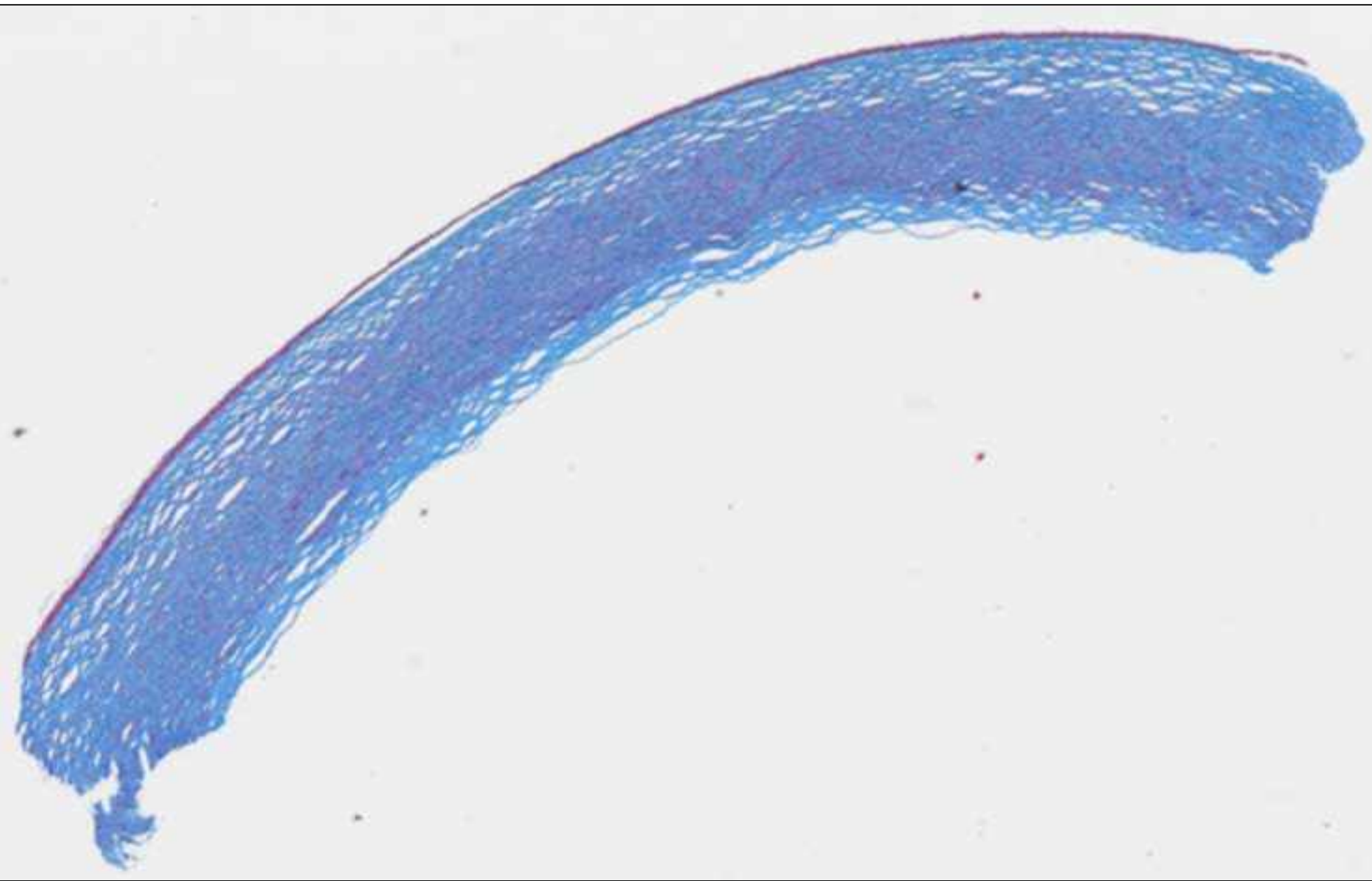
Hydropic change



Degenerating RBCs

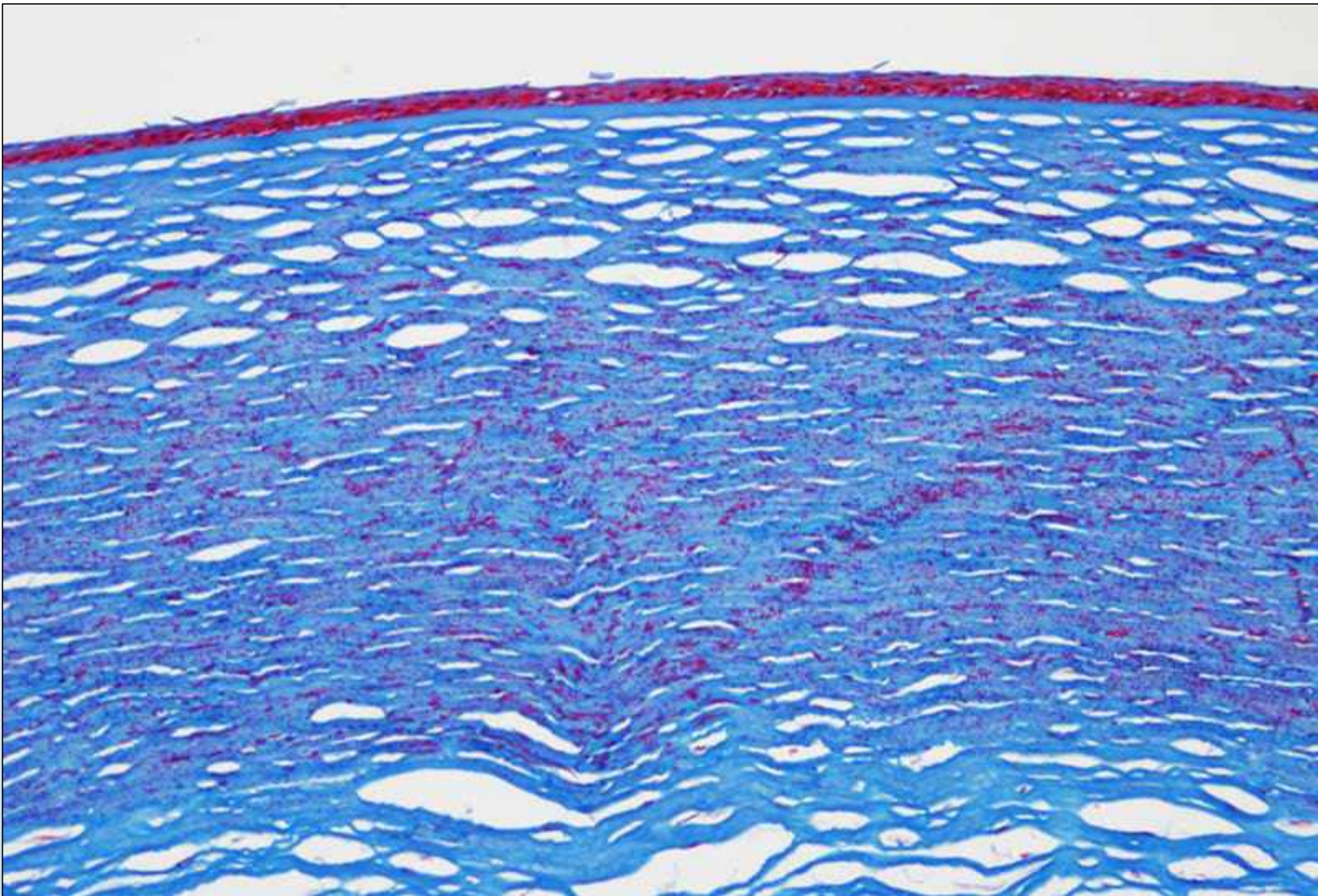


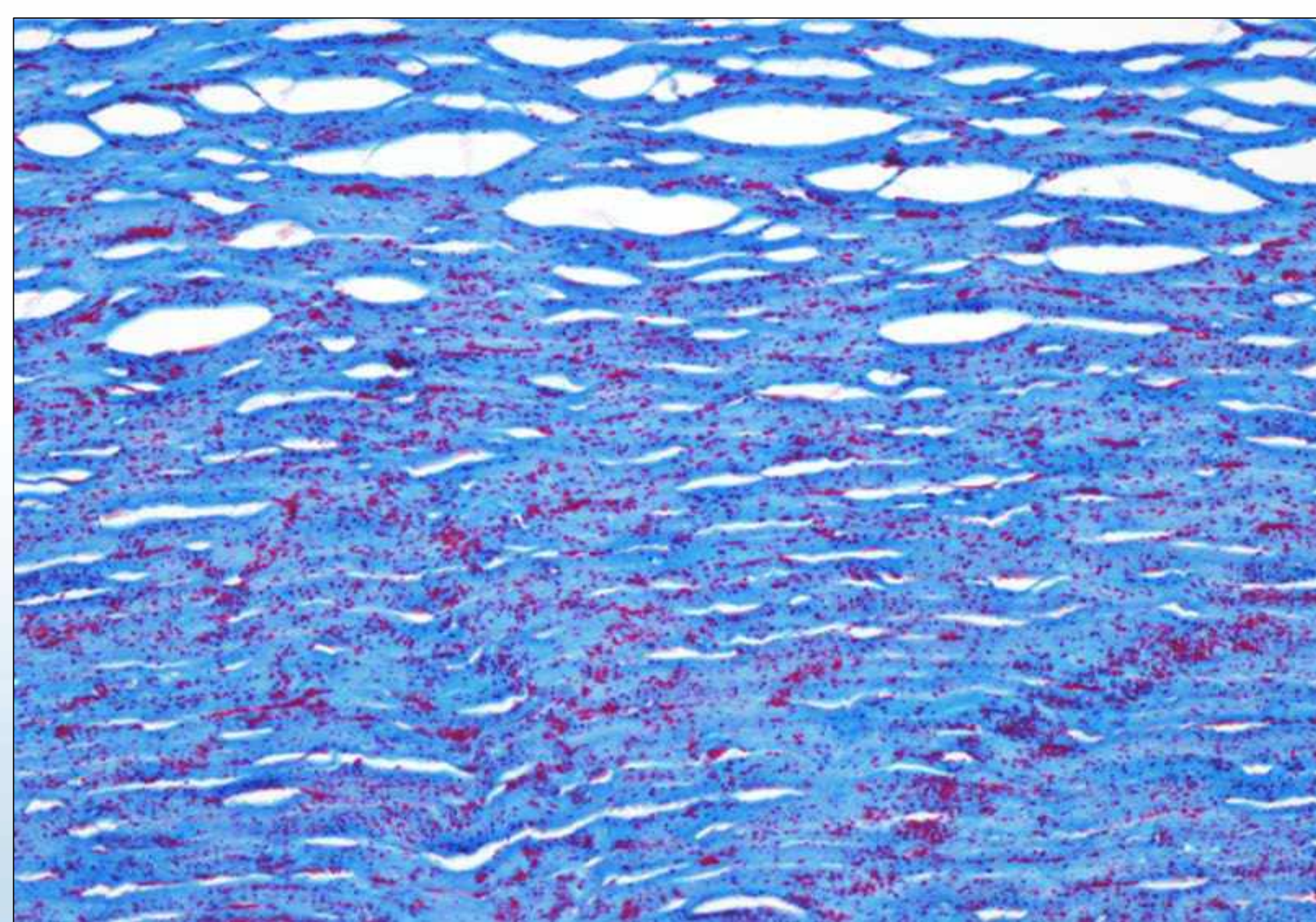


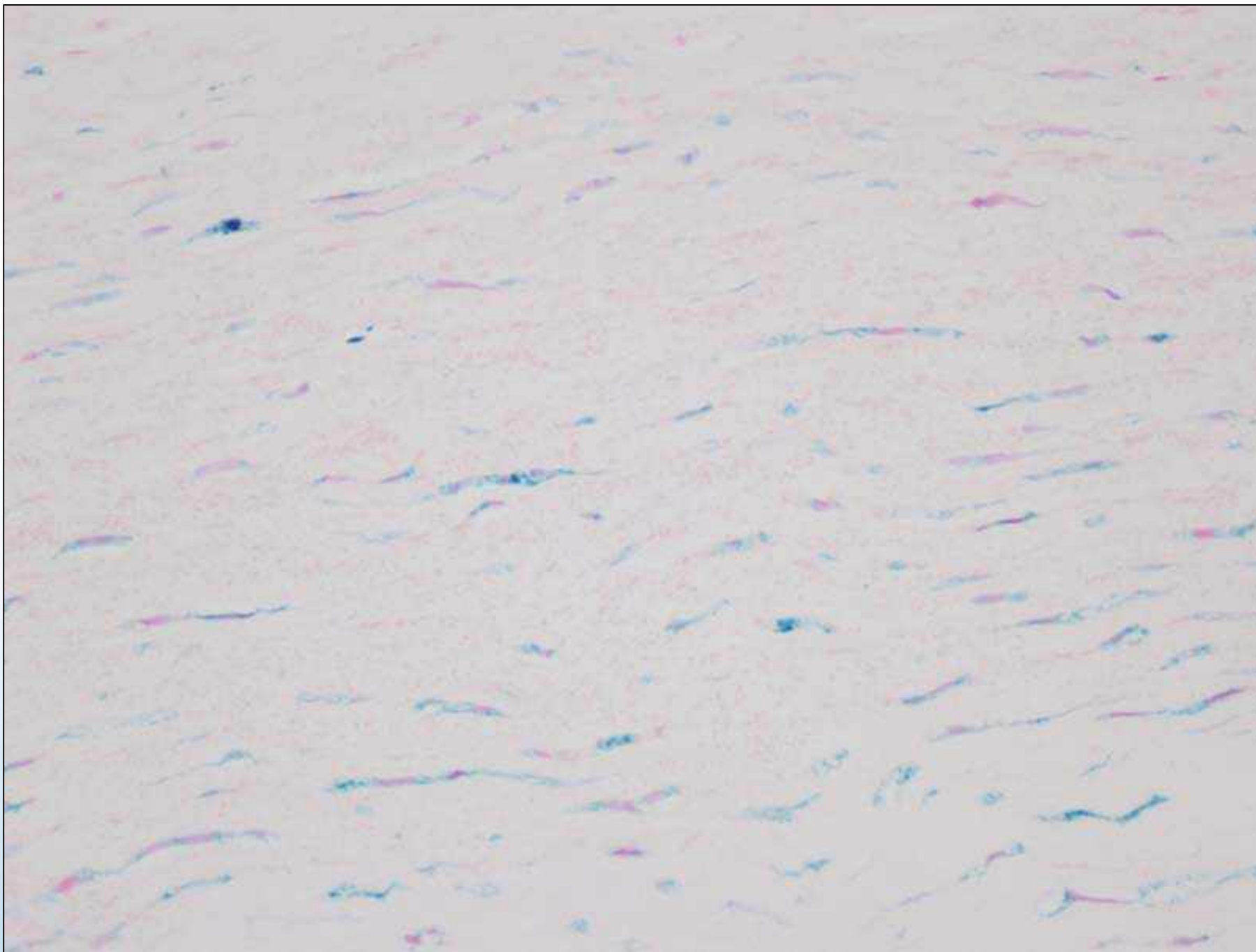


Trichome









Iron stain



Hyphema

- High IOP can cause blood staining of the cornea
- Can occur at normal IOP if the endothelium is damaged
- Blood cells break into hemoglobin particles

Gottsch JD et al. Arch Ophthalmol 1989;107:1497-500

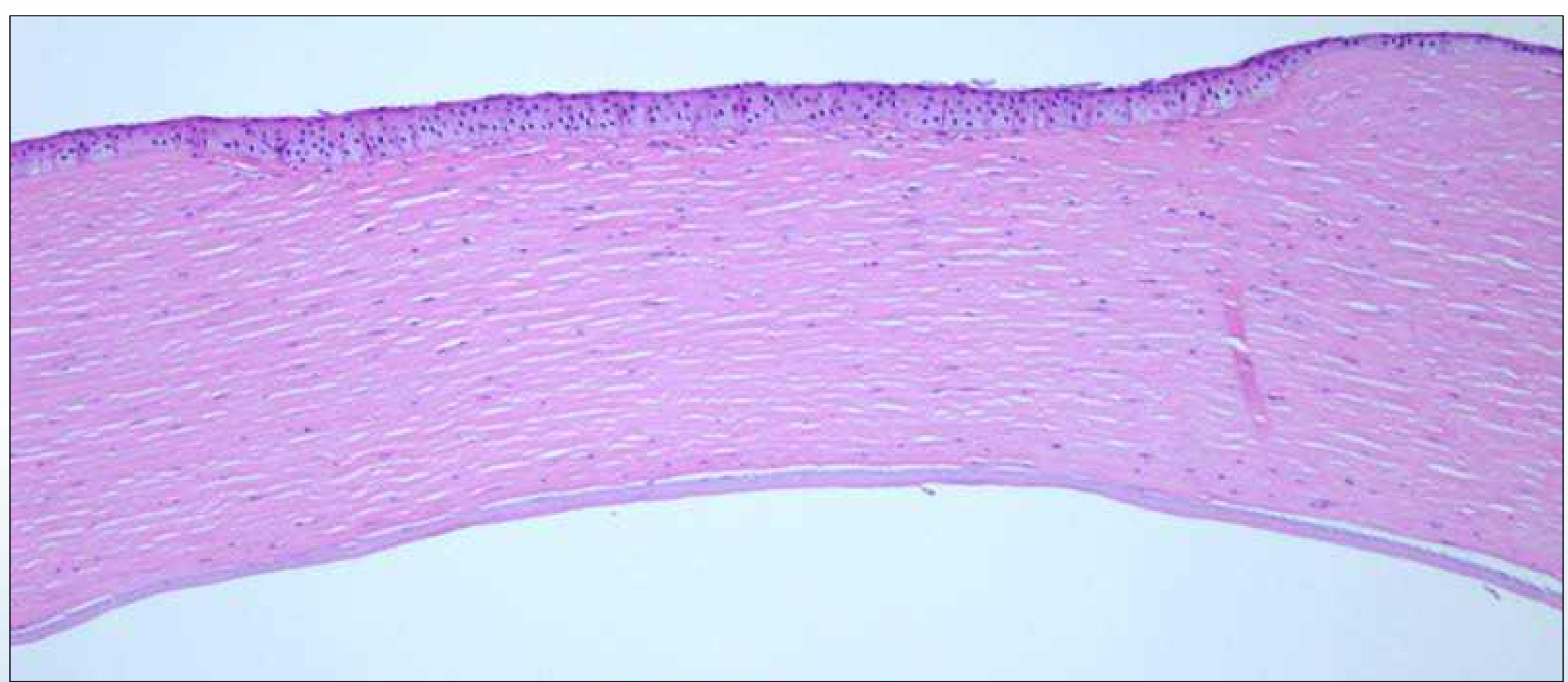
Fraser C. Et. Al. Spontaneous resolution of corneal blood staining. Clin Experiment Ophthalmol 2006; 34: 279-80.

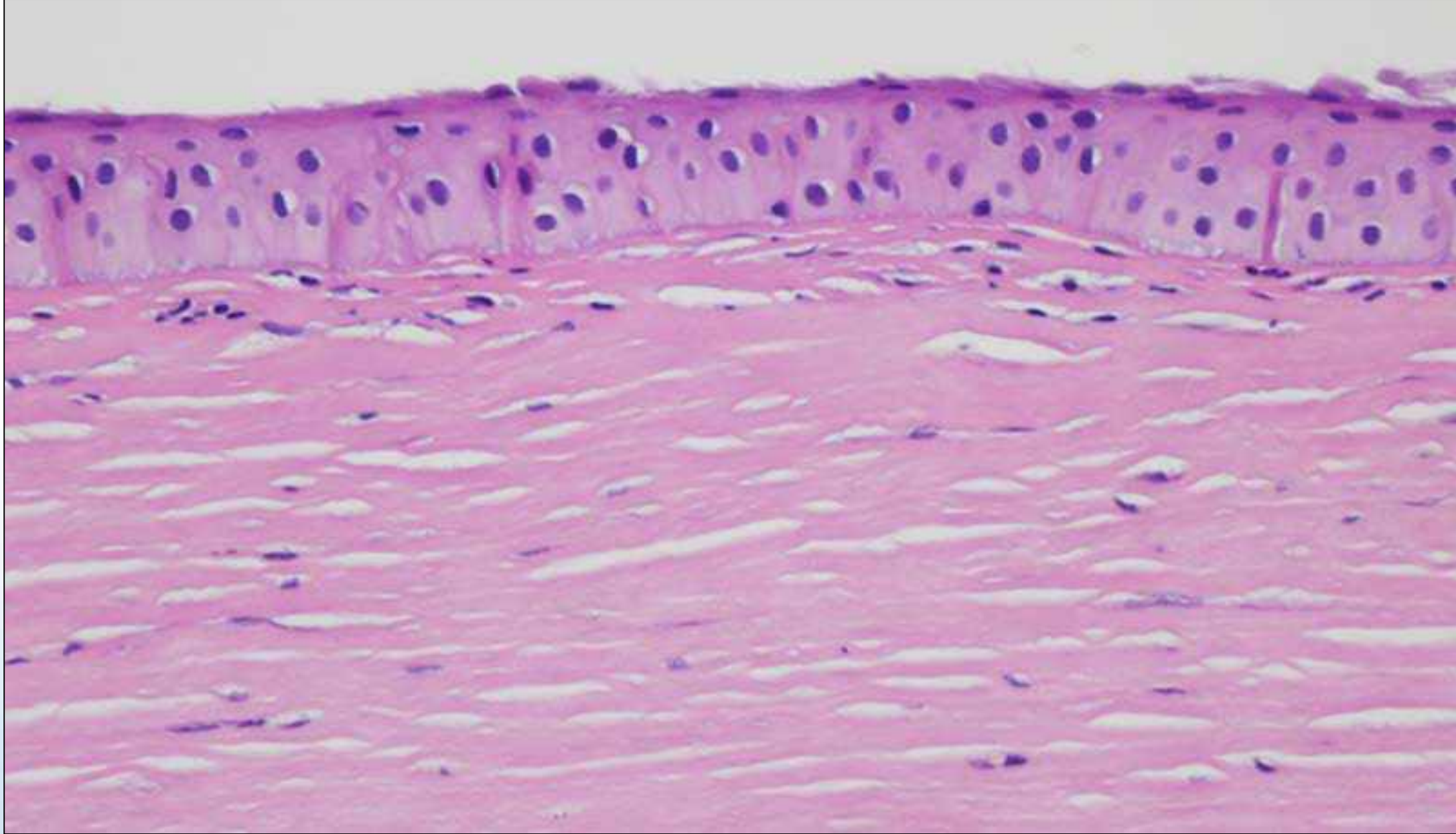


BULLOUS KERATOPATHY

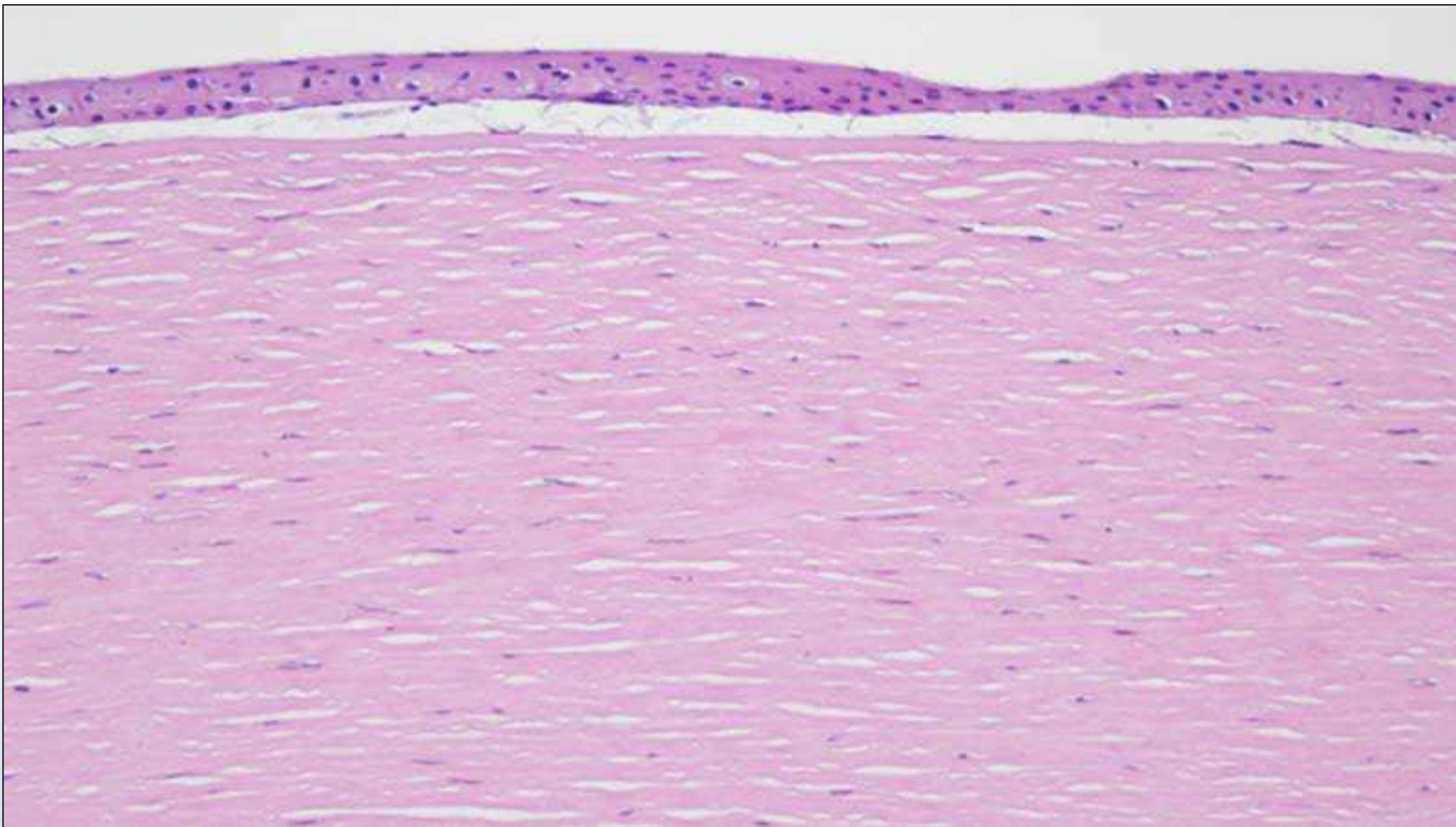




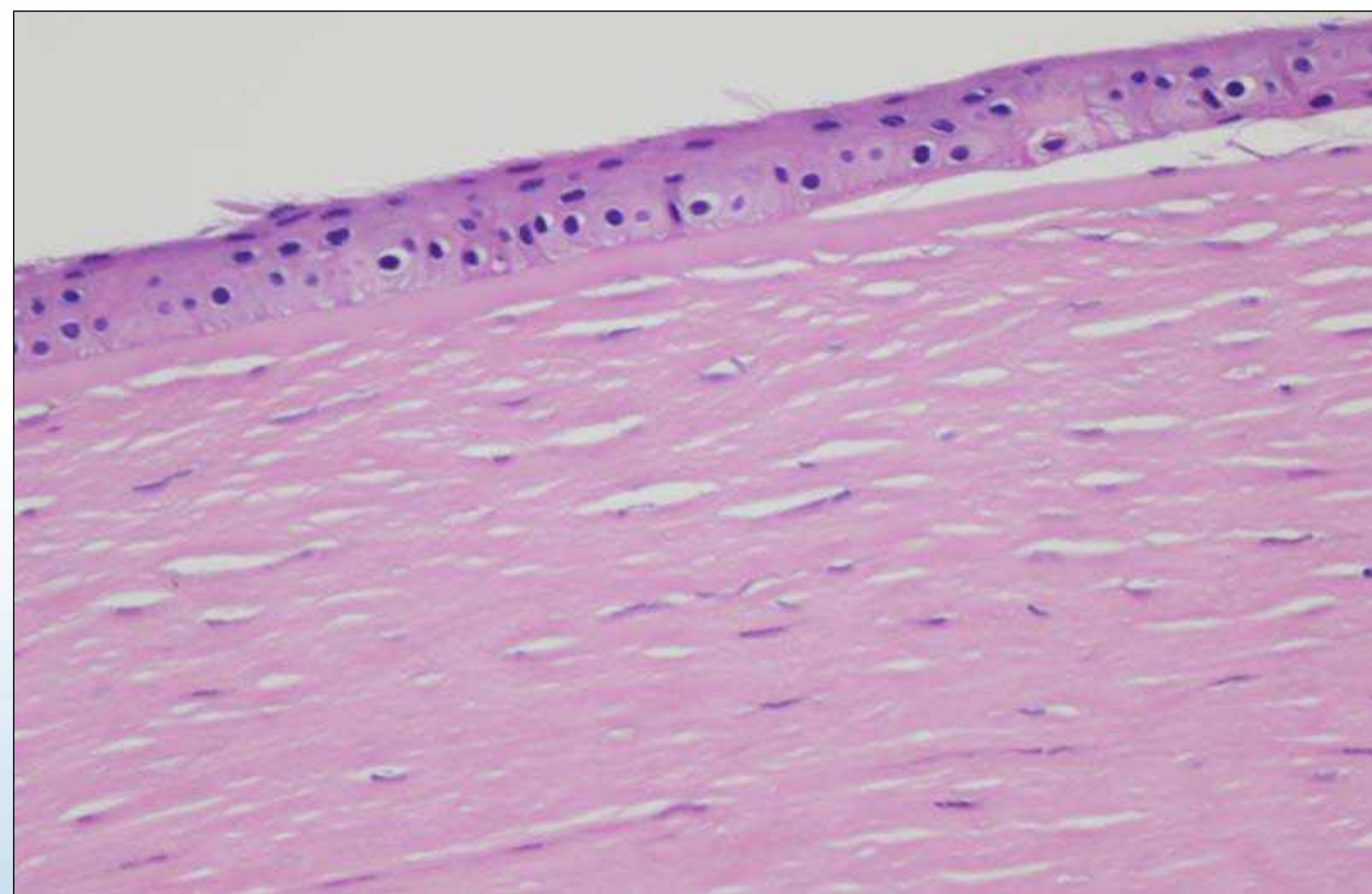




Loss of Bowman's layer

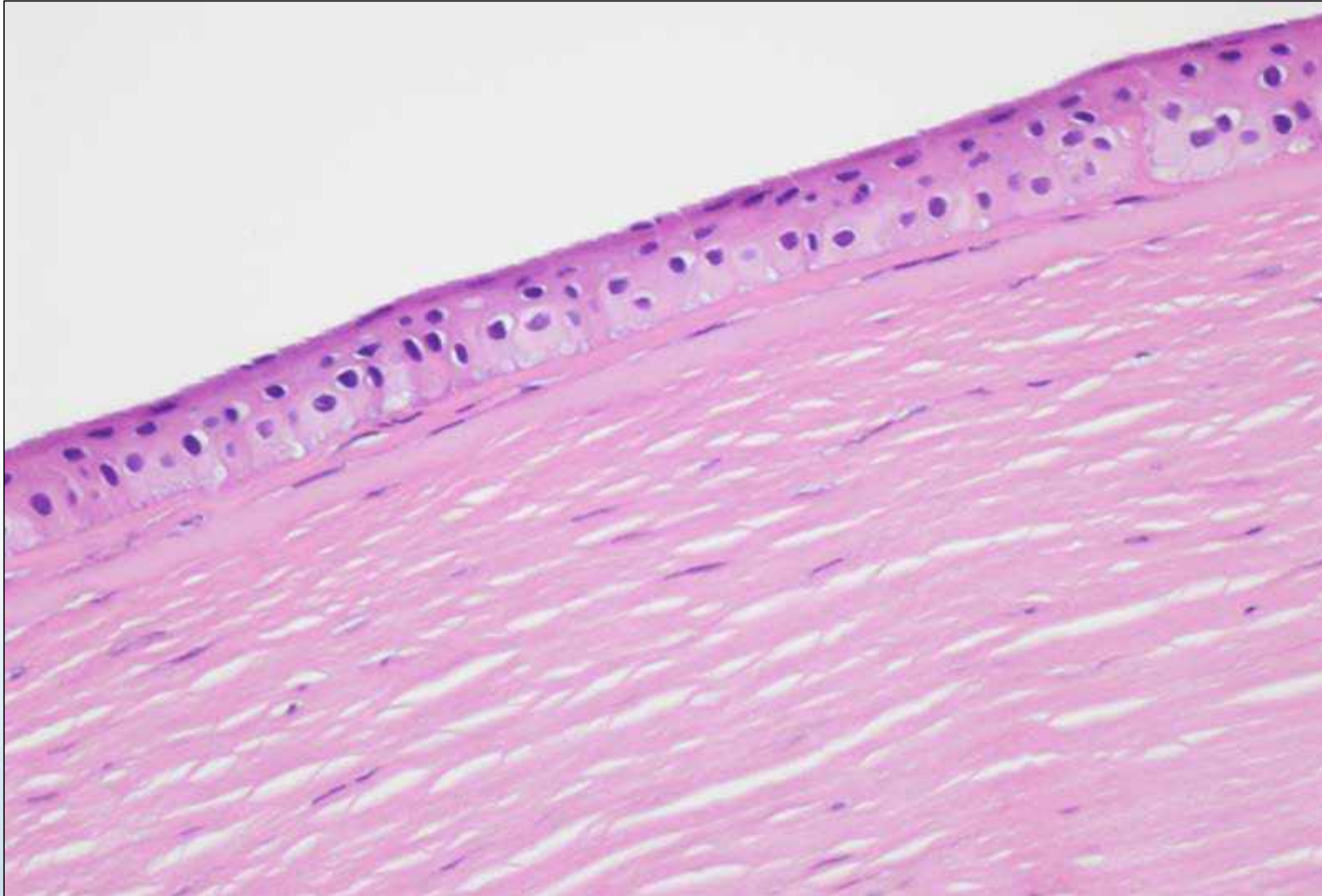


Subepithelial bullae

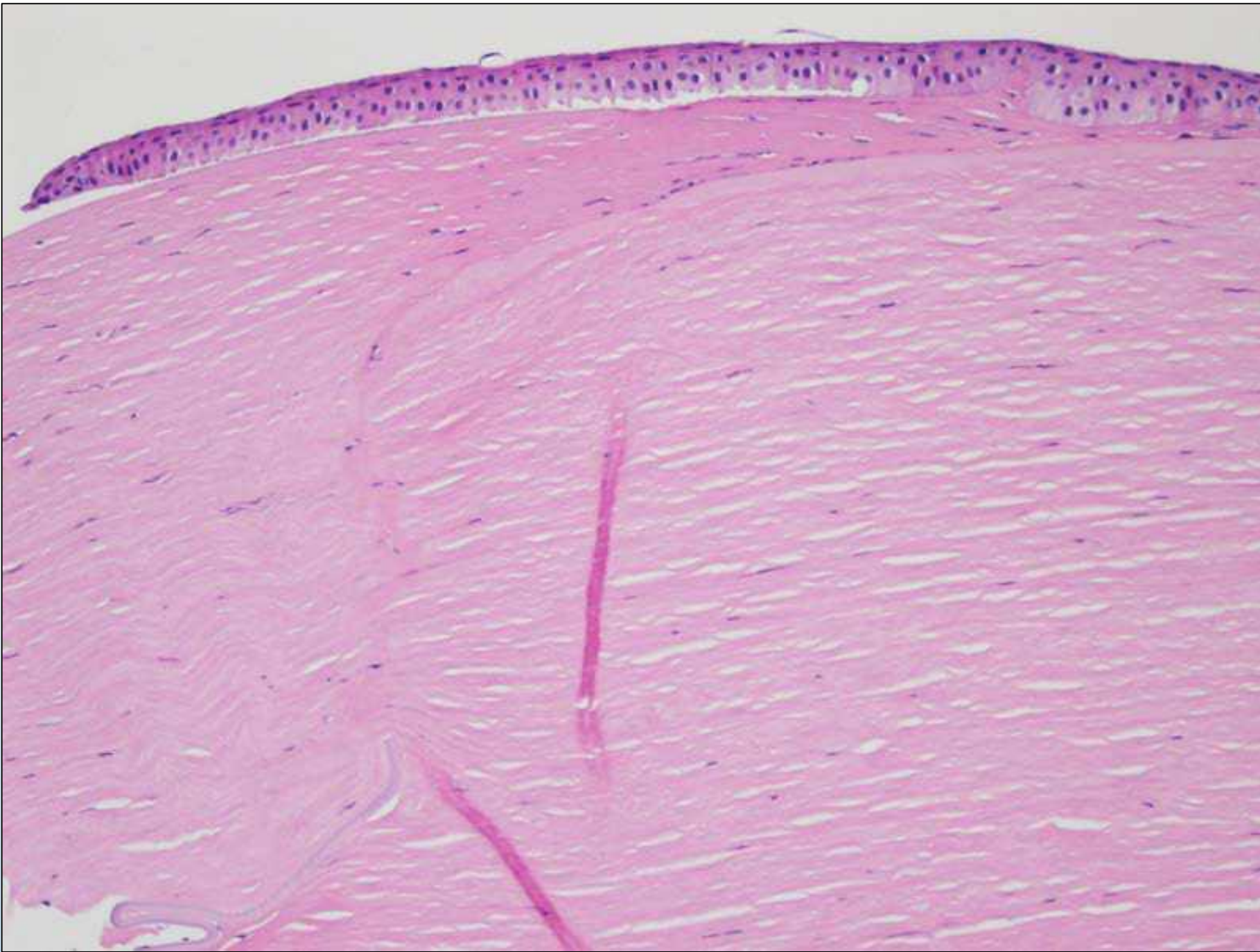


Epithelial basal cell hydropic change



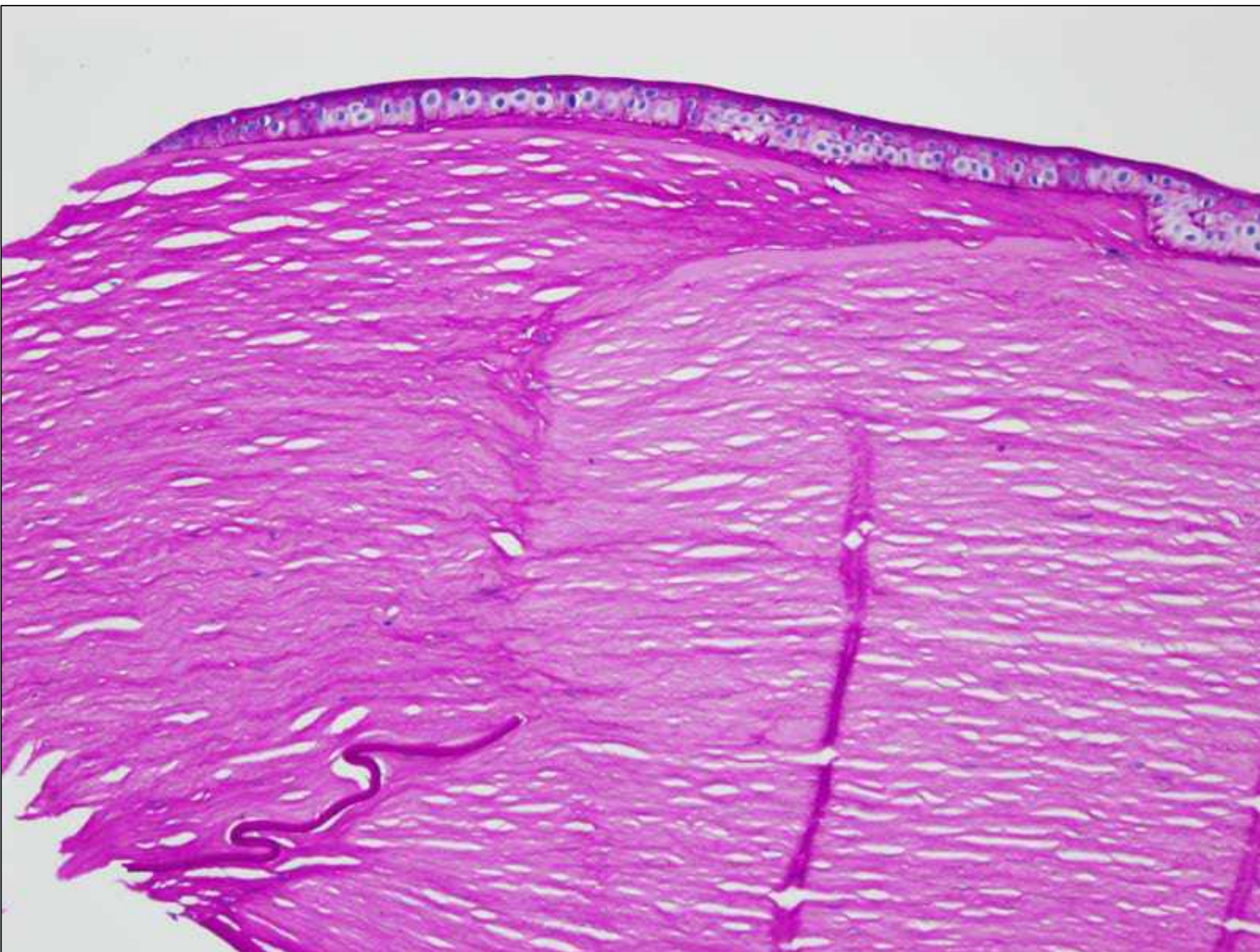


Subepithelial fibrous pannus



Surgical changes





PAS

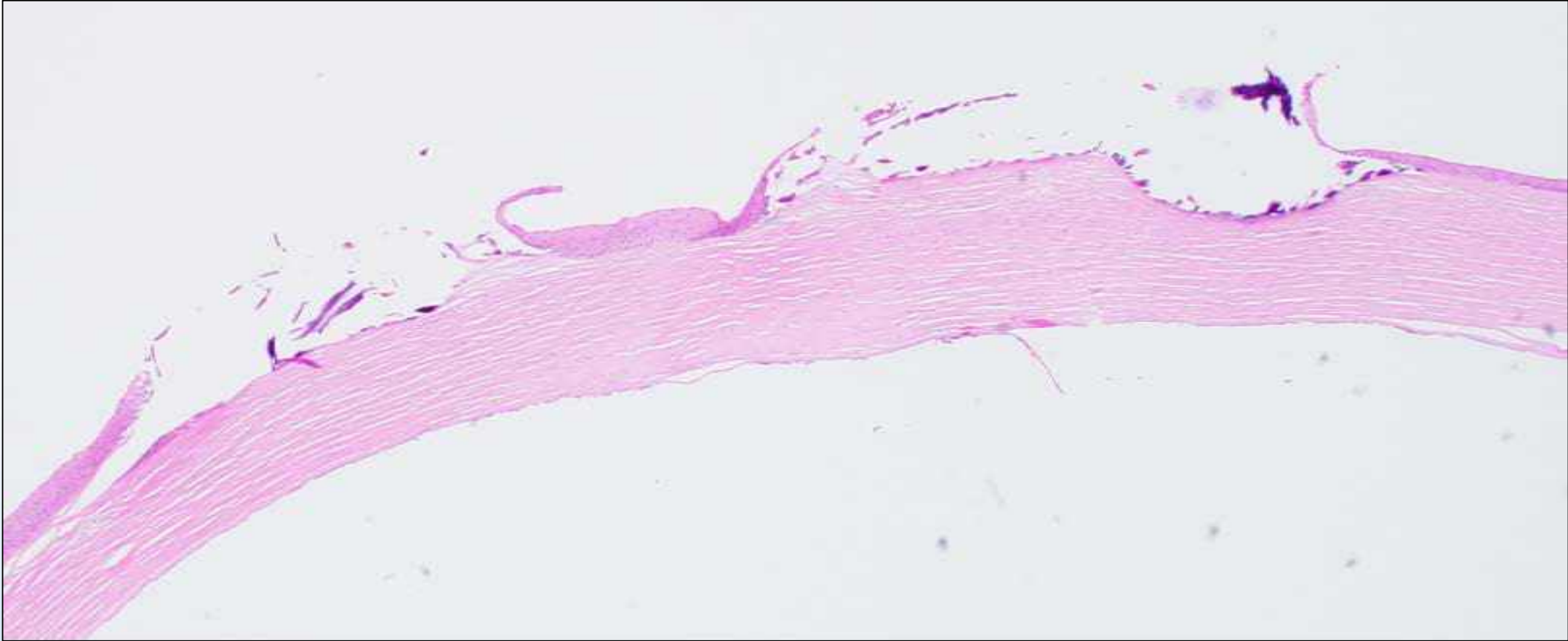


Bullous Keratopathy

- End result of persistent corneal edema
- Often caused by failure of corneal endothelium
 - Abnormal cell function
 - Decreased number of cells
- Seen following cataract surgery

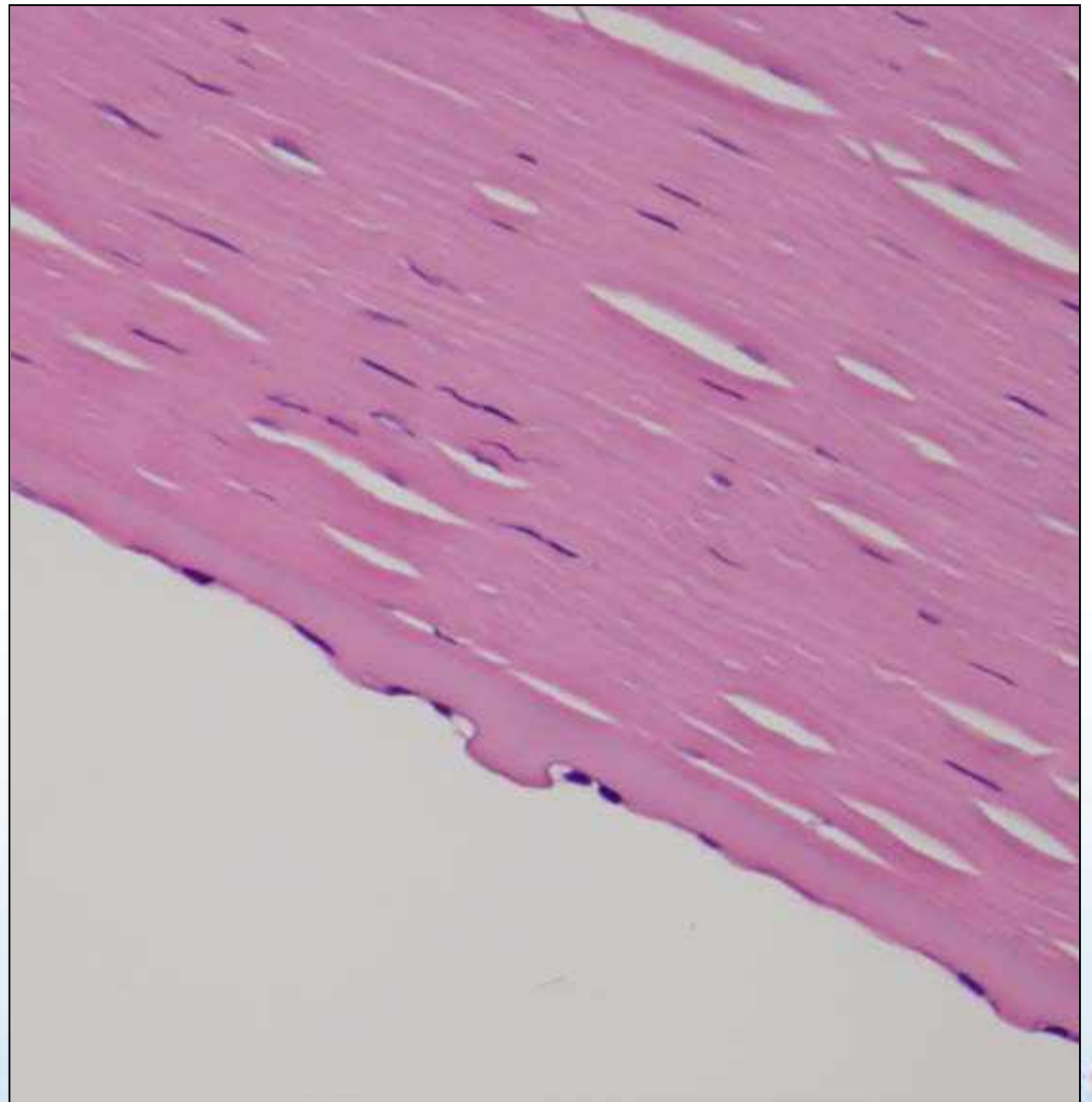
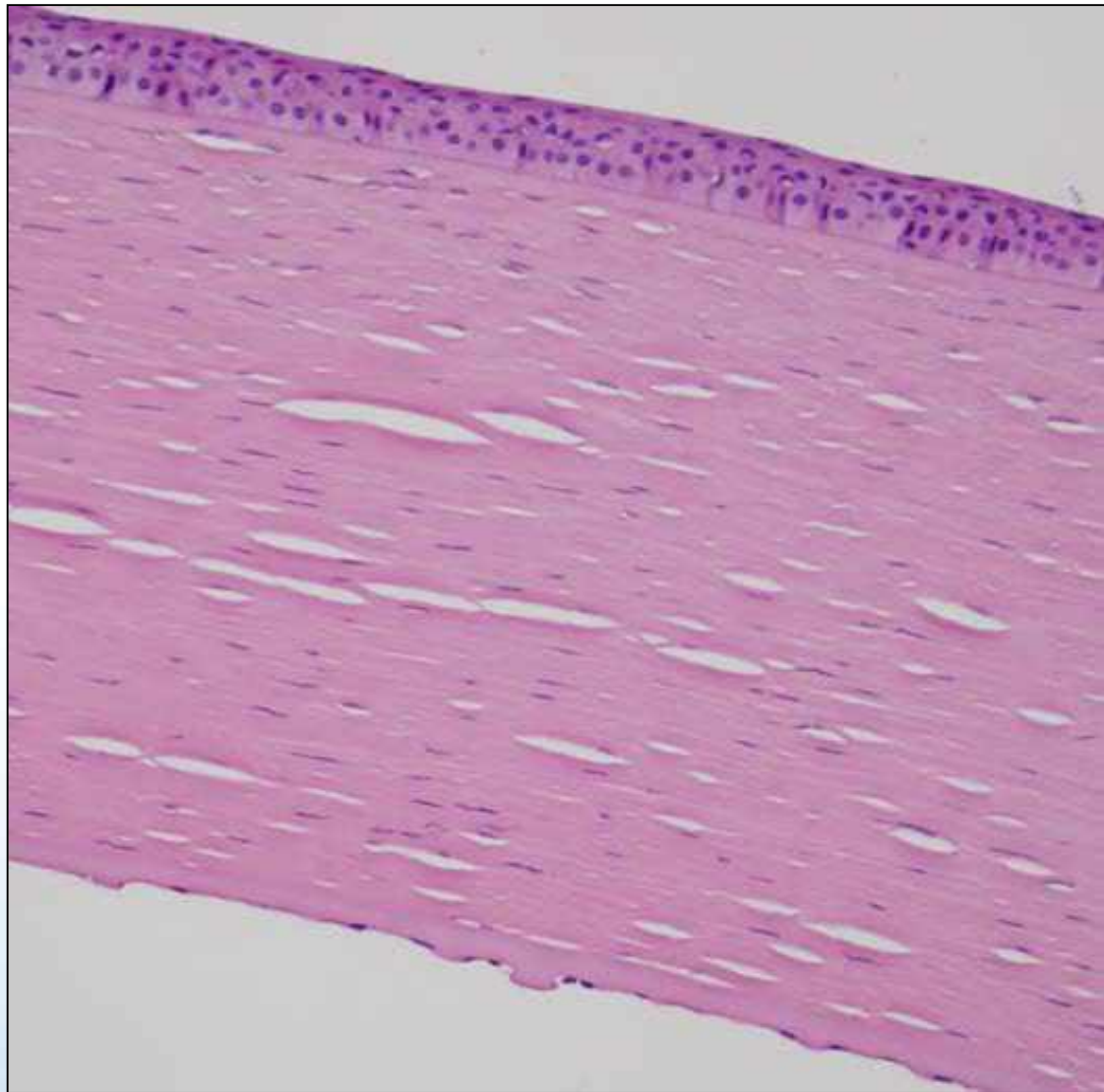


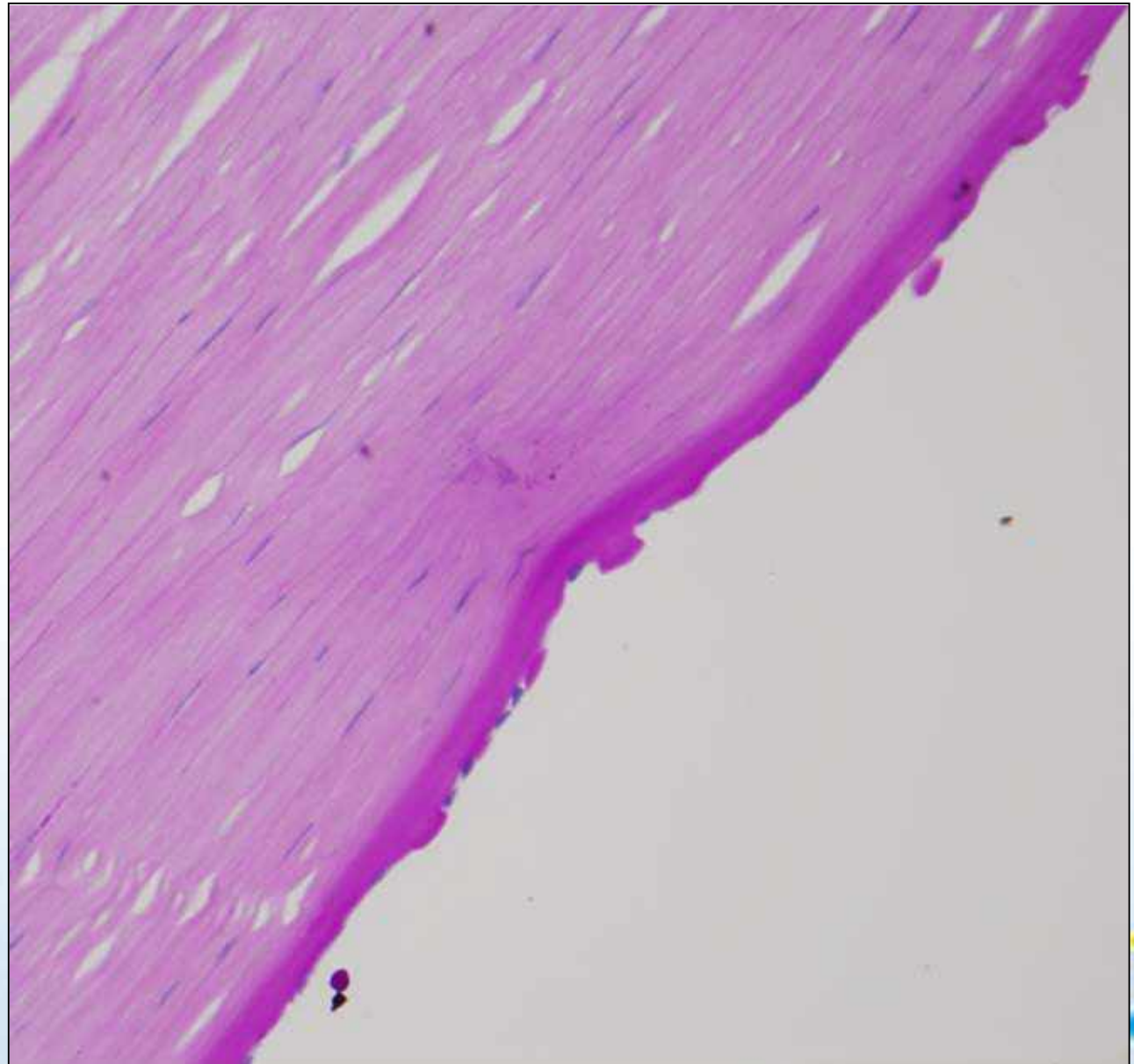
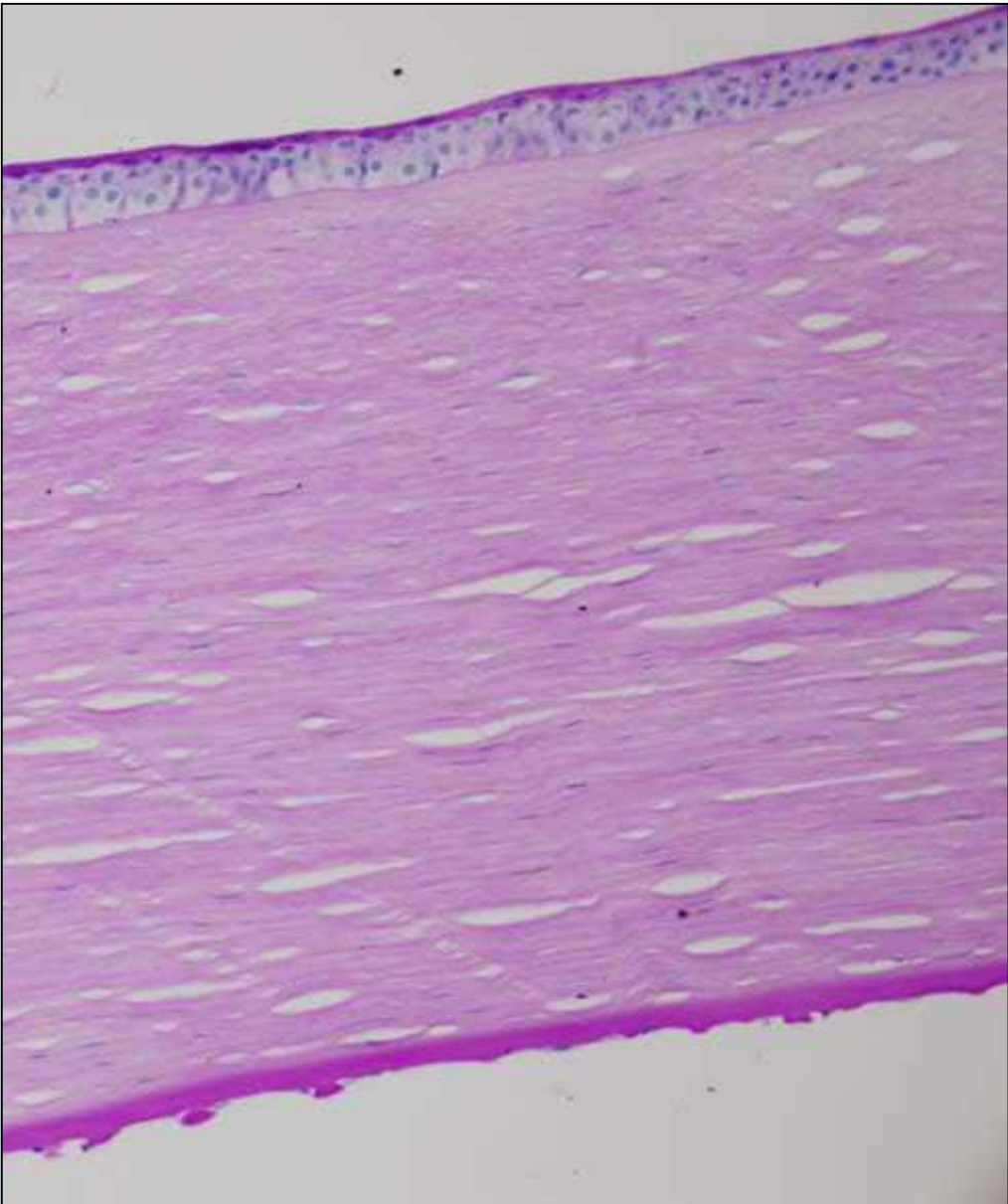
Calcific Band Keratopathy



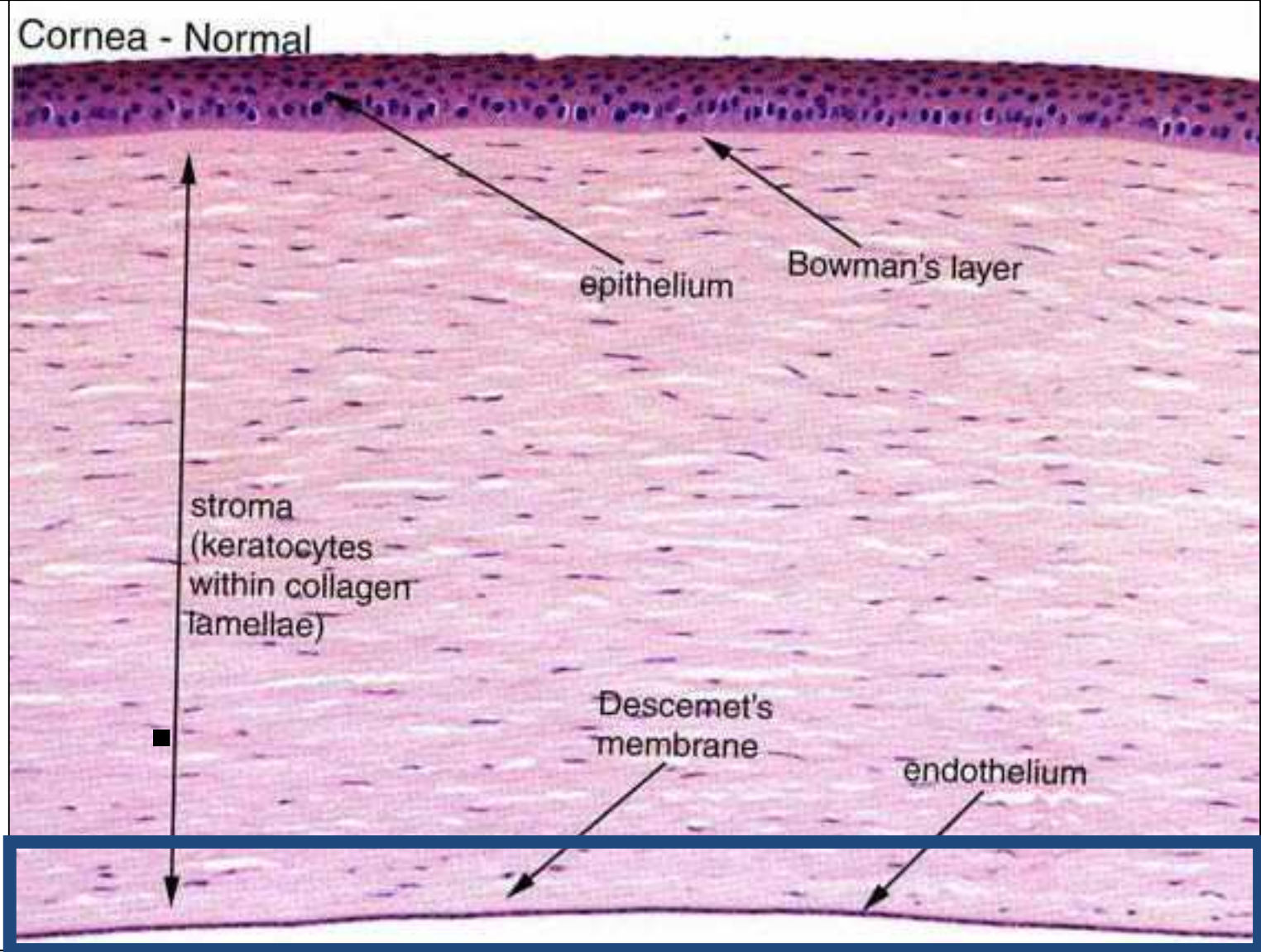
FUCHS DYSTROPHY







DSAEK -

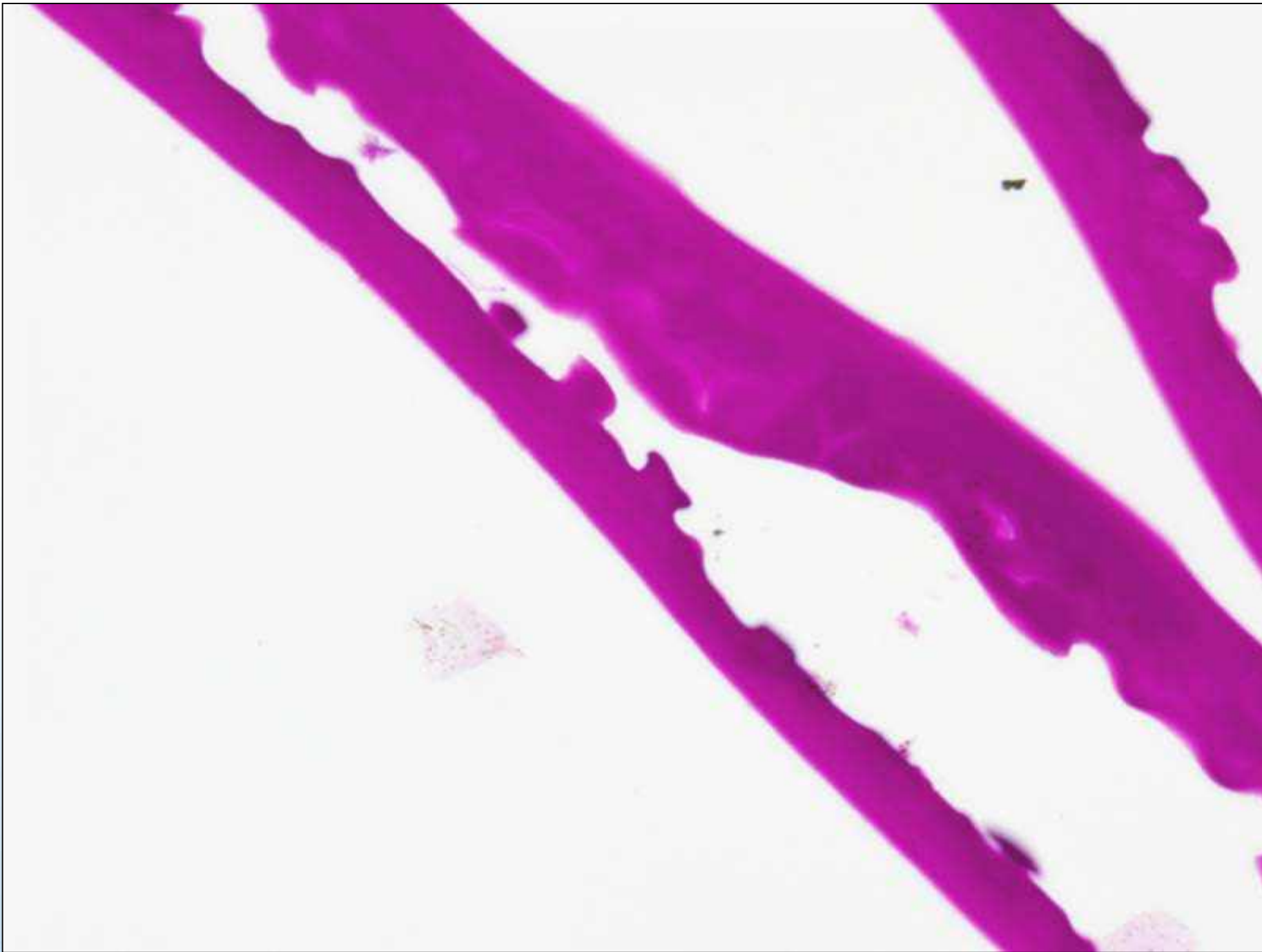


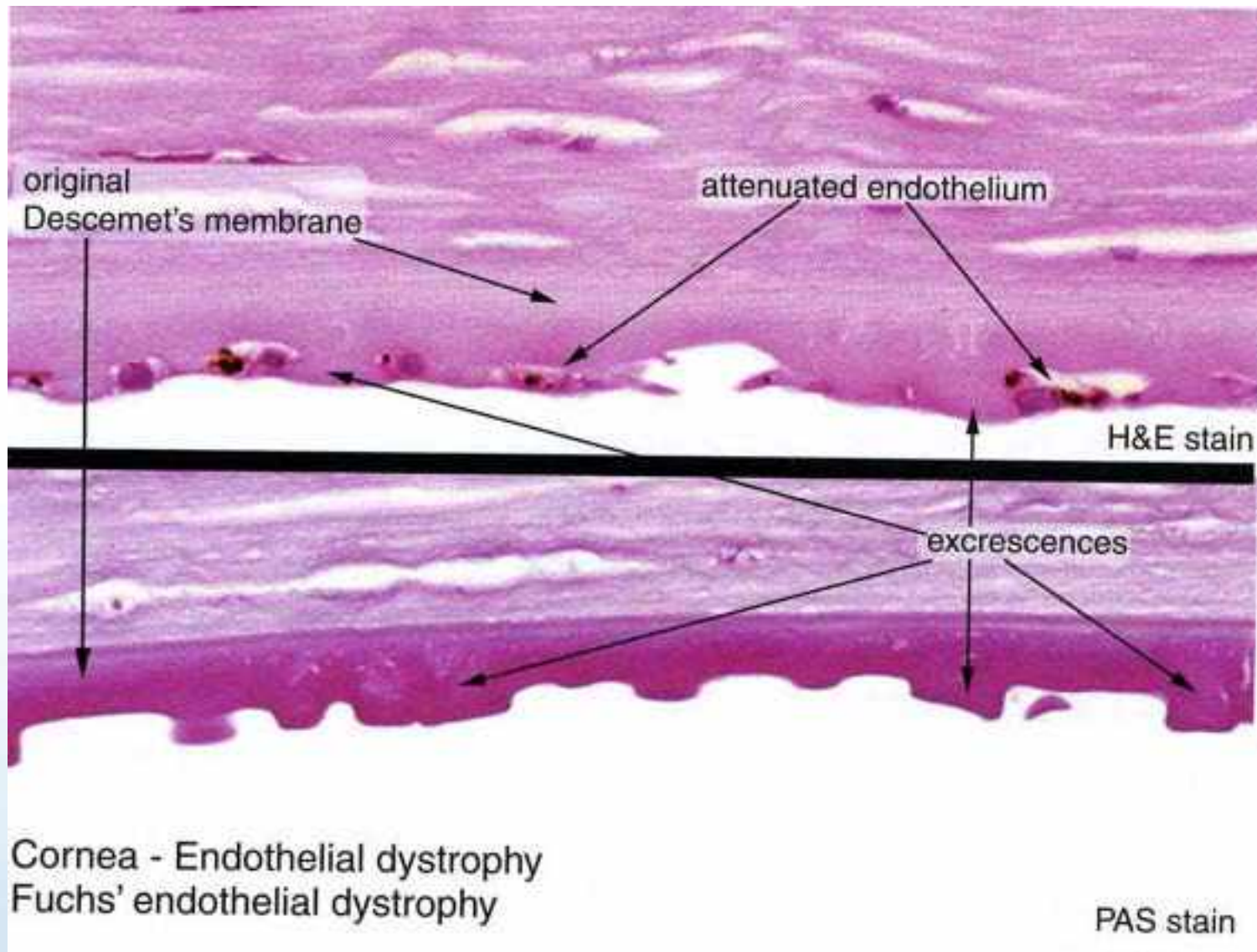


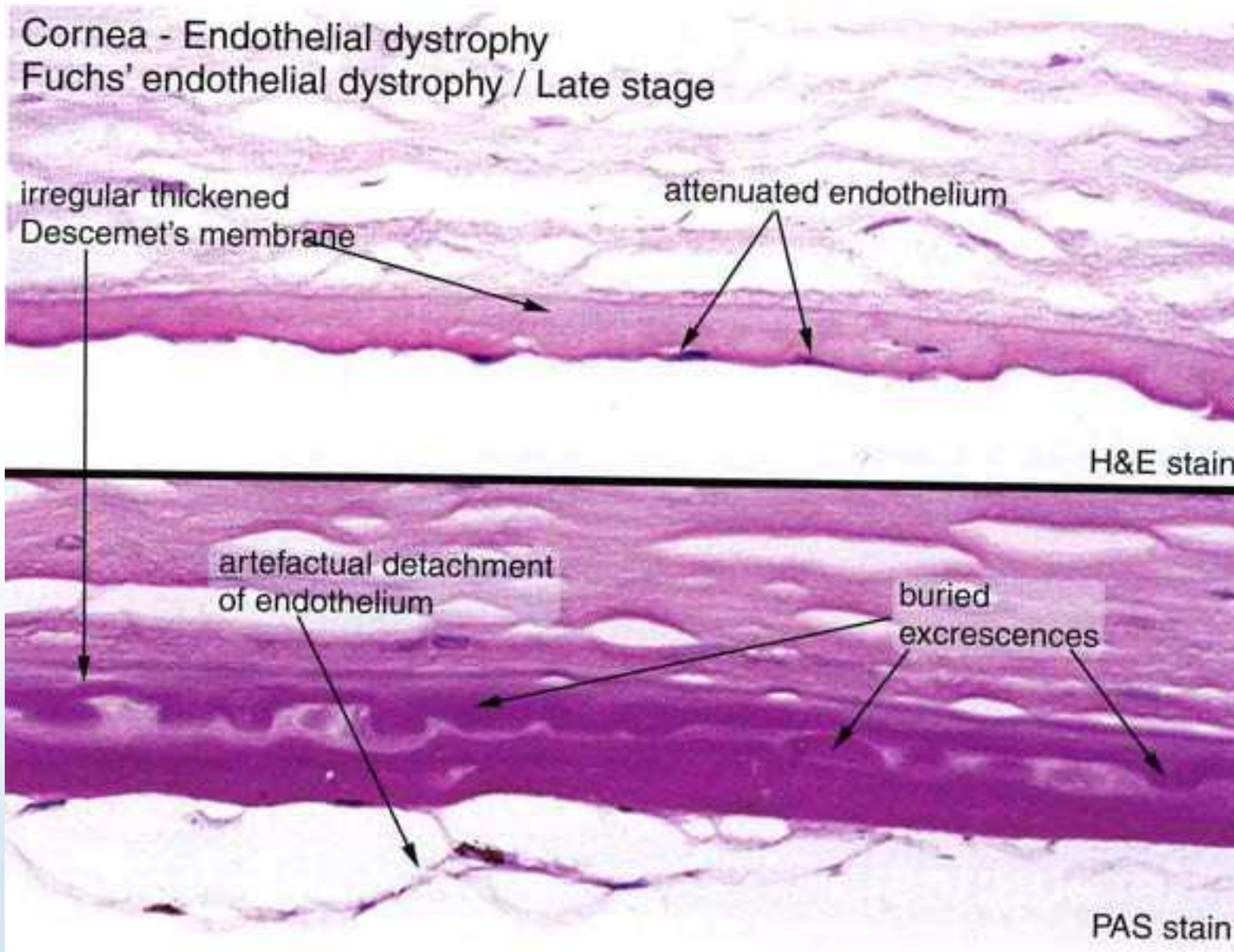
AANP





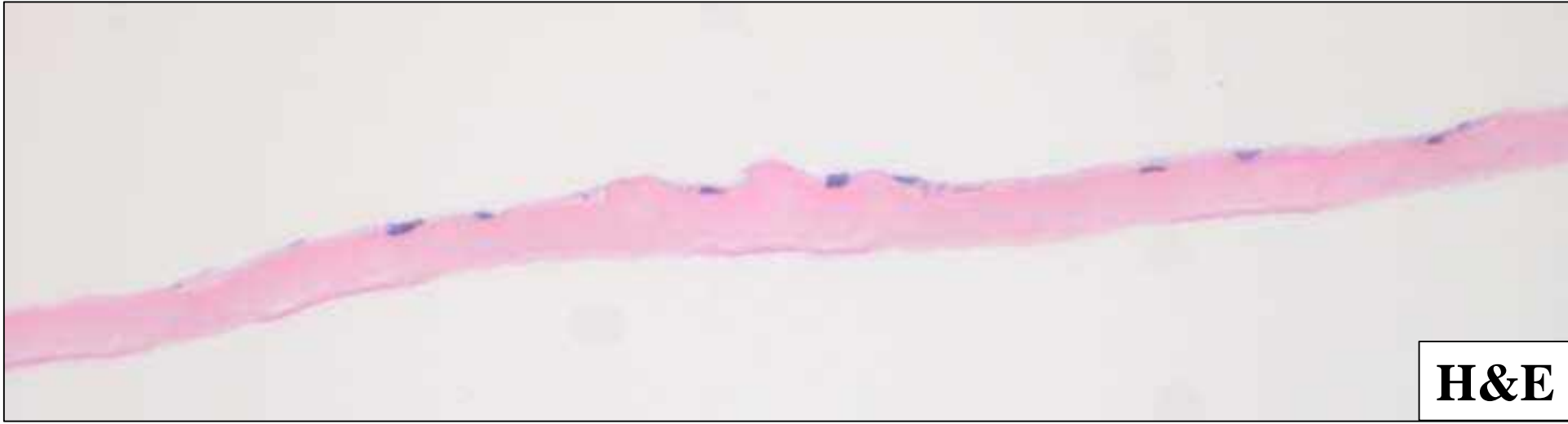


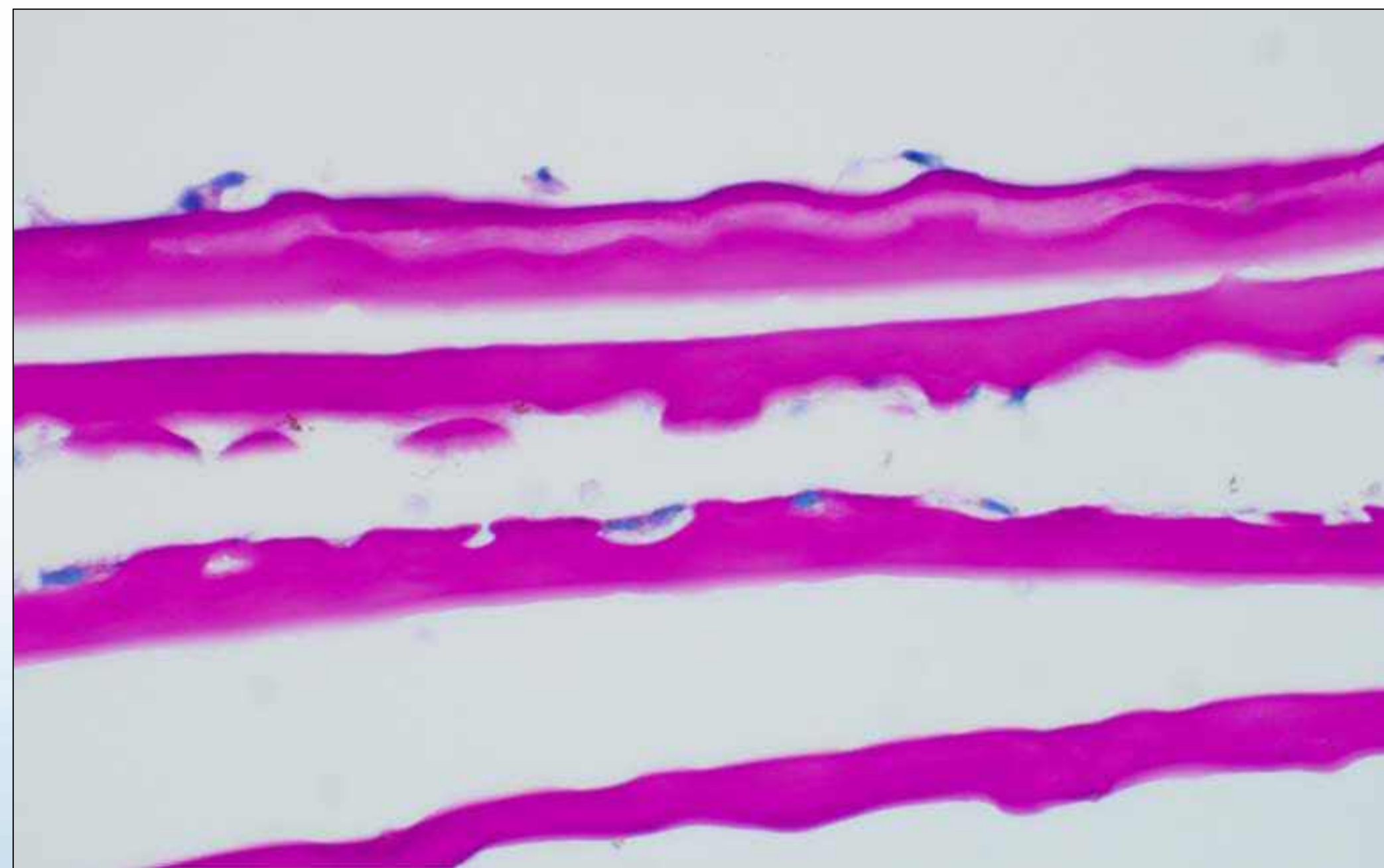




Eagle, Ralph C. (2017), *Eye Pathology: An Atlas and Text*, 3rd ed. Wolters Kluwer









Hilary Highfield
8/10/22



Fuchs dystrophy



Fuchs endothelial dystrophy

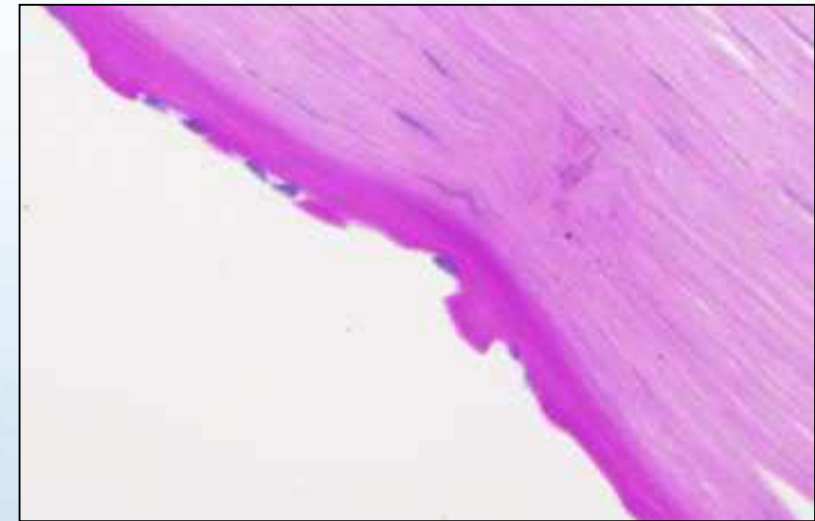
- Affects 5% of Americans over age 40
- Bilateral
- Middle-aged to elderly; (40-50's)
- Occurs more commonly in women
- Variable inheritance pattern
- Most common indication for penetrating keratoplasty for corneal dystrophy
- One of the leading causes of bullous keratopathy

NEJM 2010; 363: 1016



Fuchs endothelial dystrophy

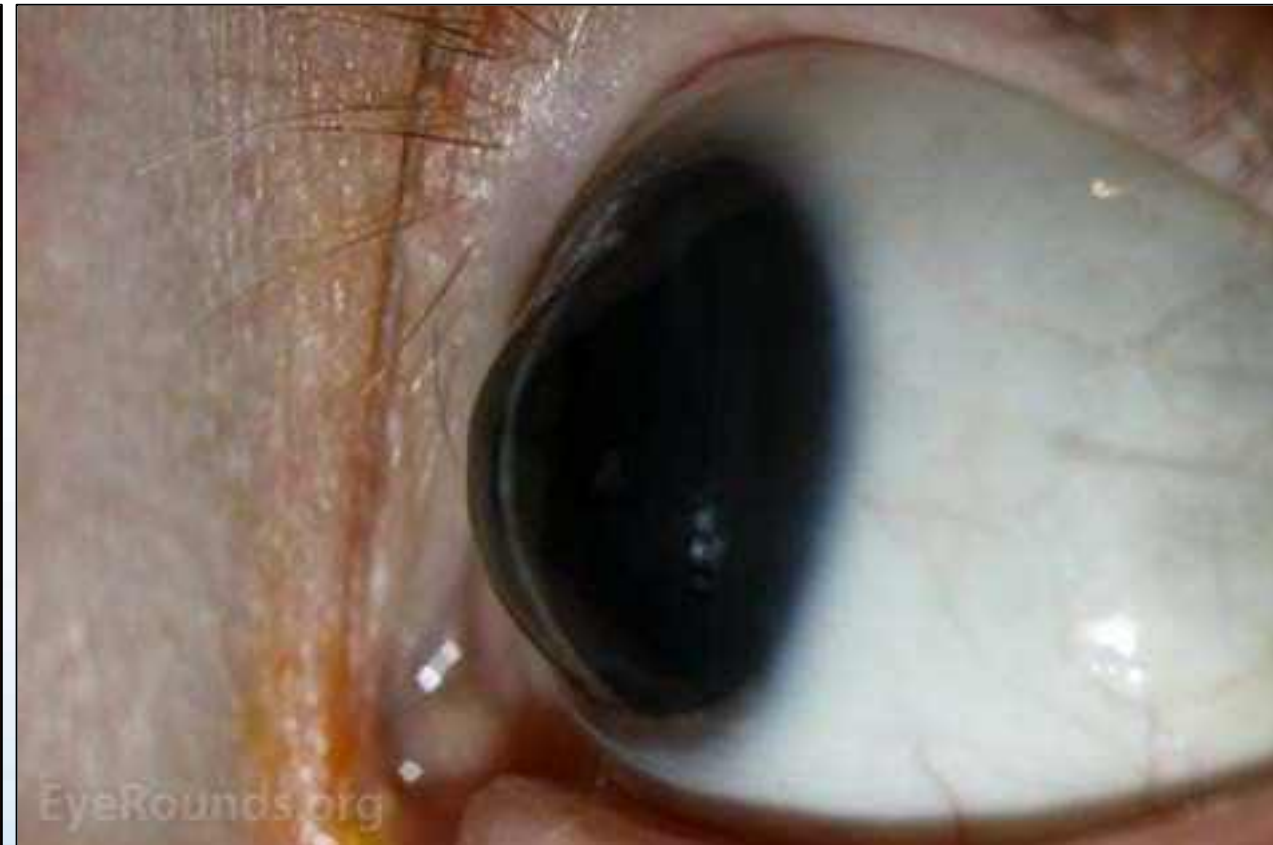
- Anvil-shaped excrescences of Descemet's membrane: Guttae “spotted/speckled”
- Endothelial cells sparse to absent (attenuated endothelium) with basement membrane thickening
- Diffuse edema: corneal epithelium and stroma



KERATOCONUS



Munson's sign



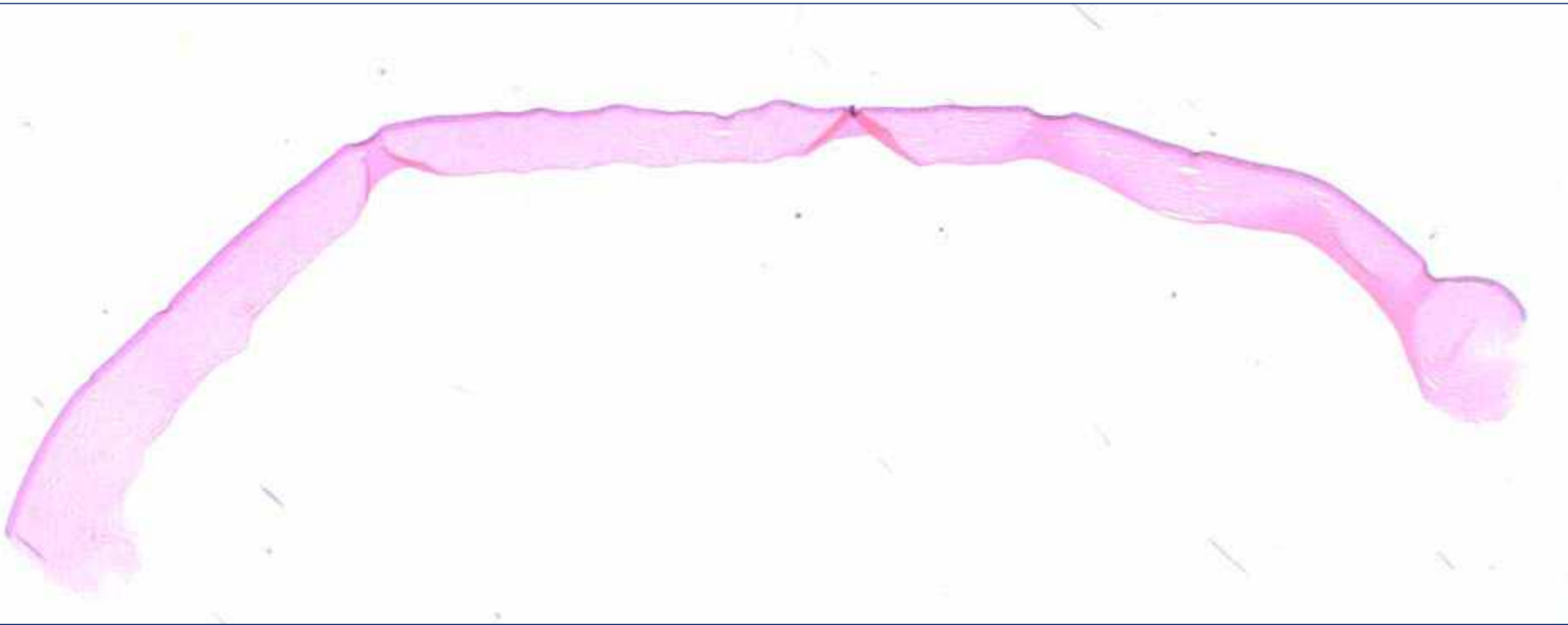
University of Iowa
EyeRounds.org



Keratoconus



Keratoconus



Keratoconus

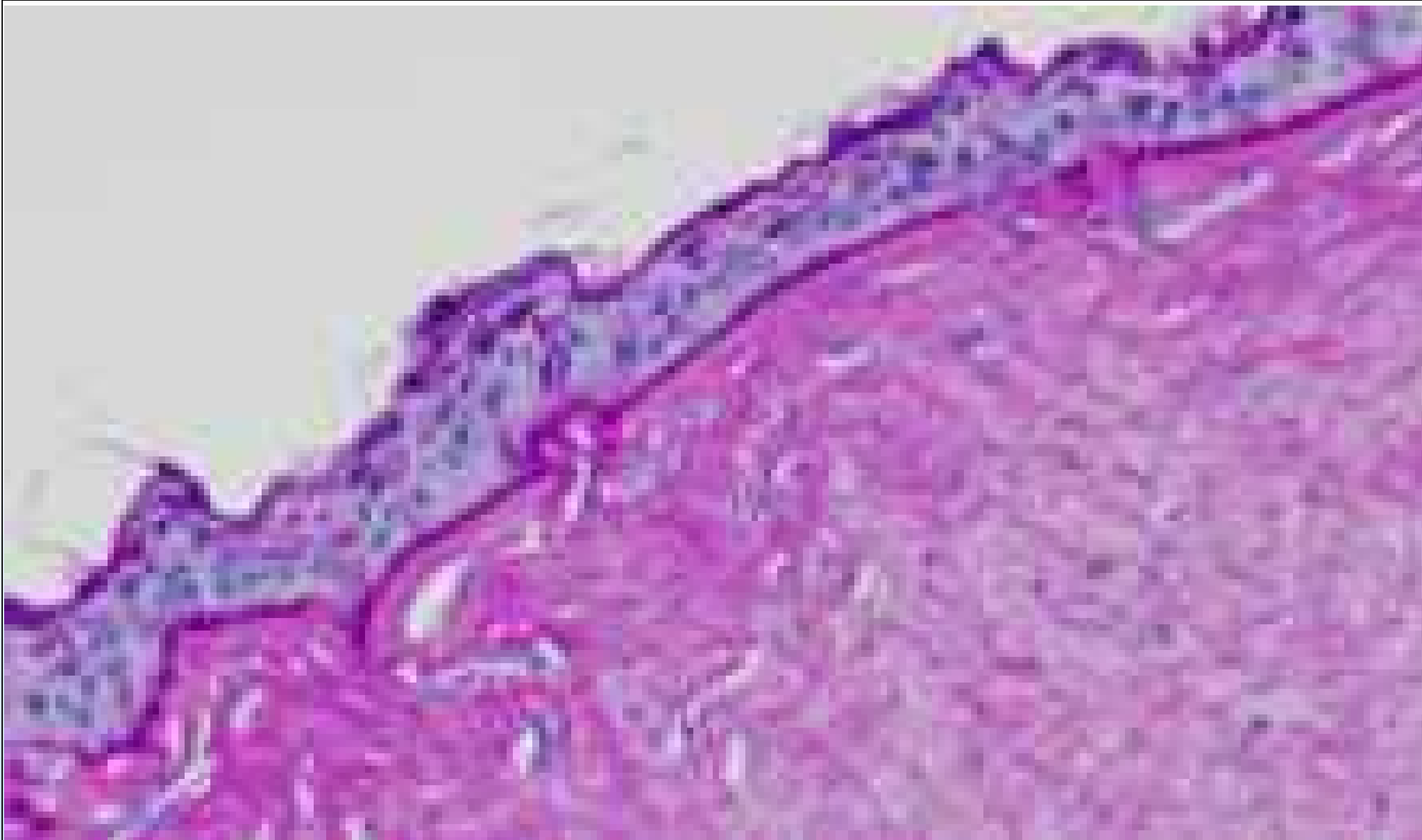


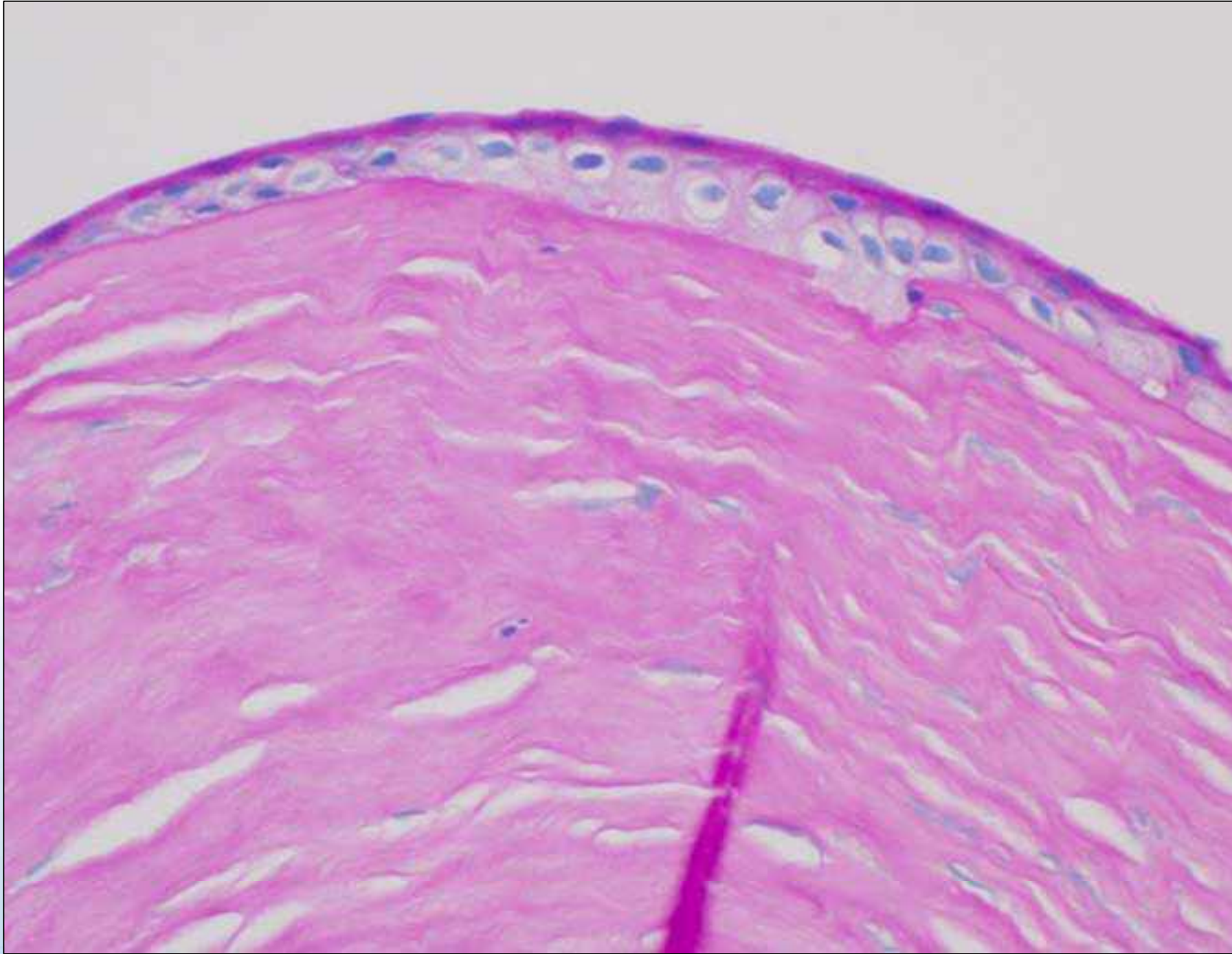
Keratoconus

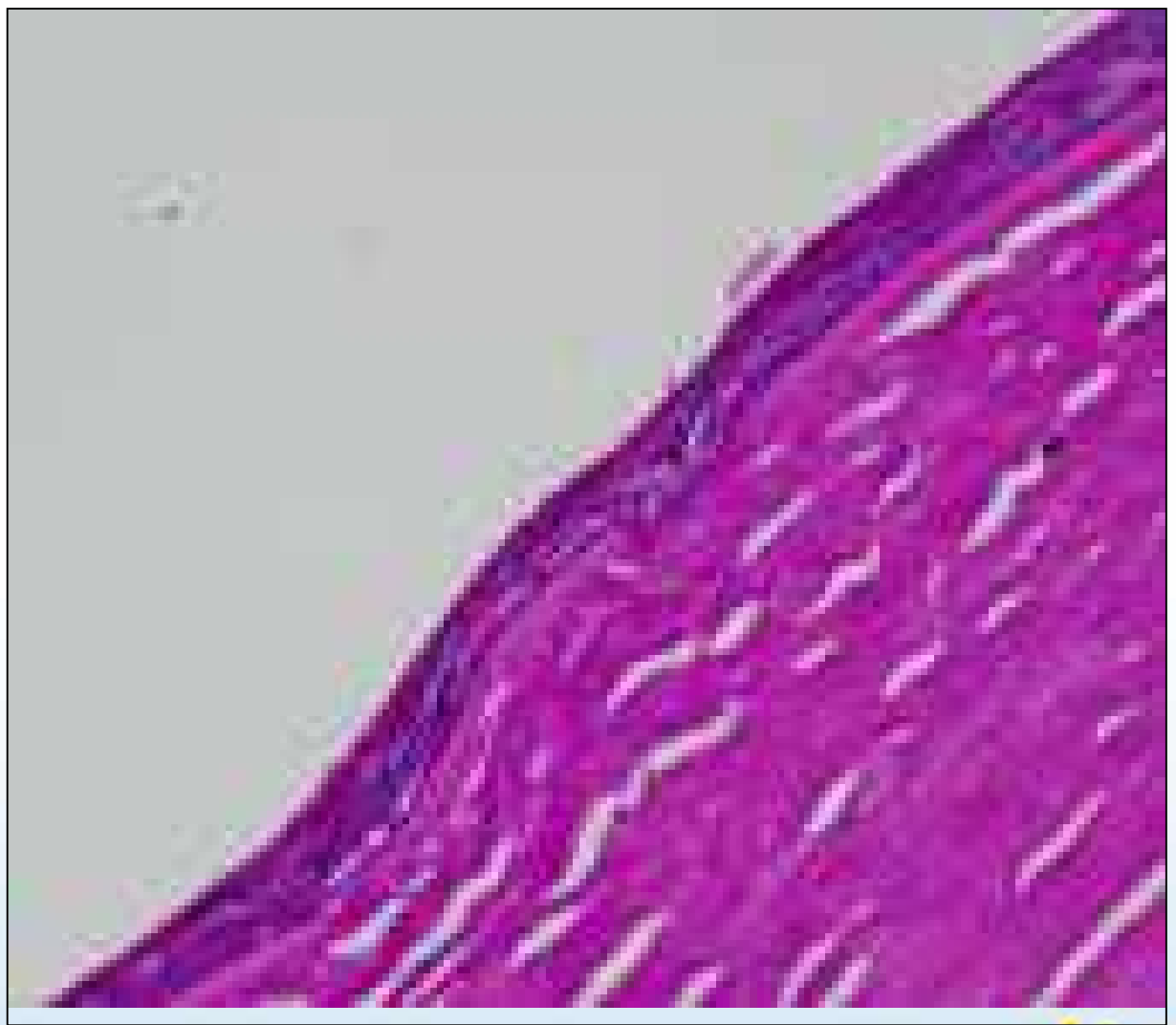




Bowman's Layer Breaks

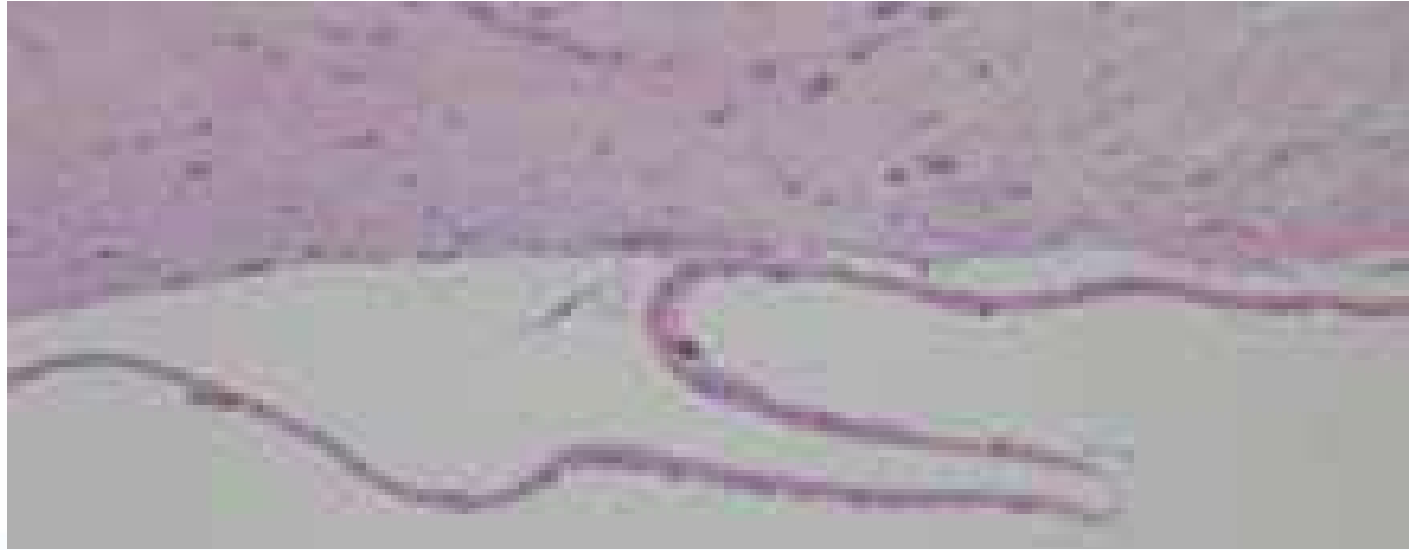




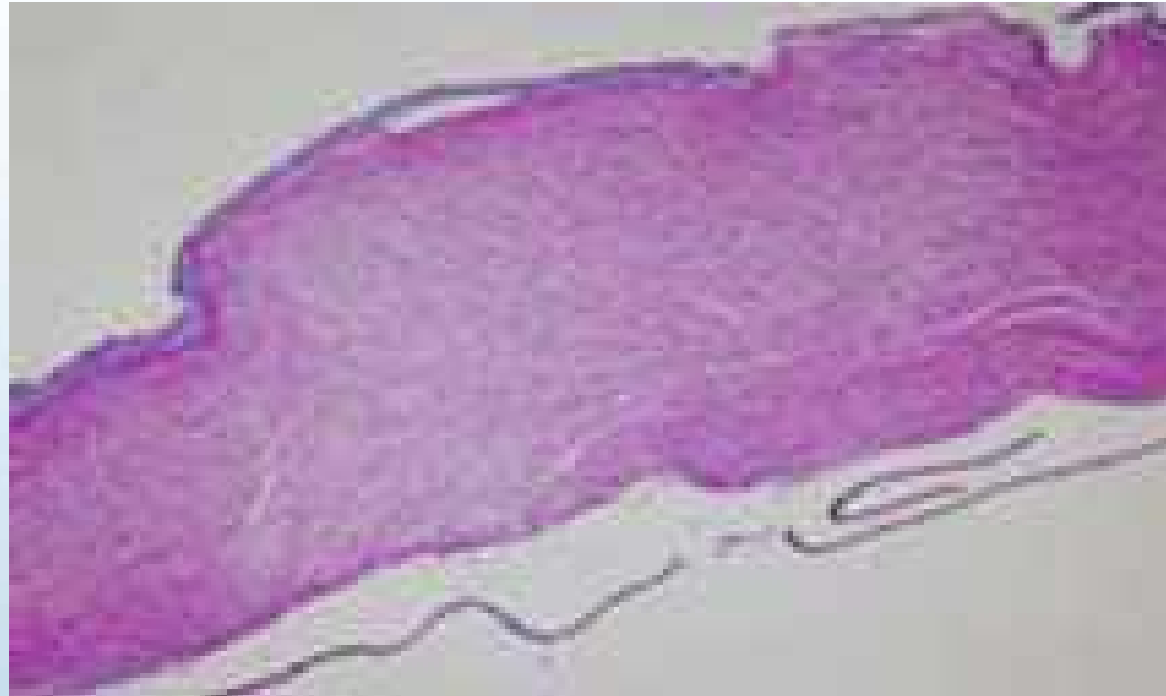


Hydrops





**Decemet's
membrane
breaks**



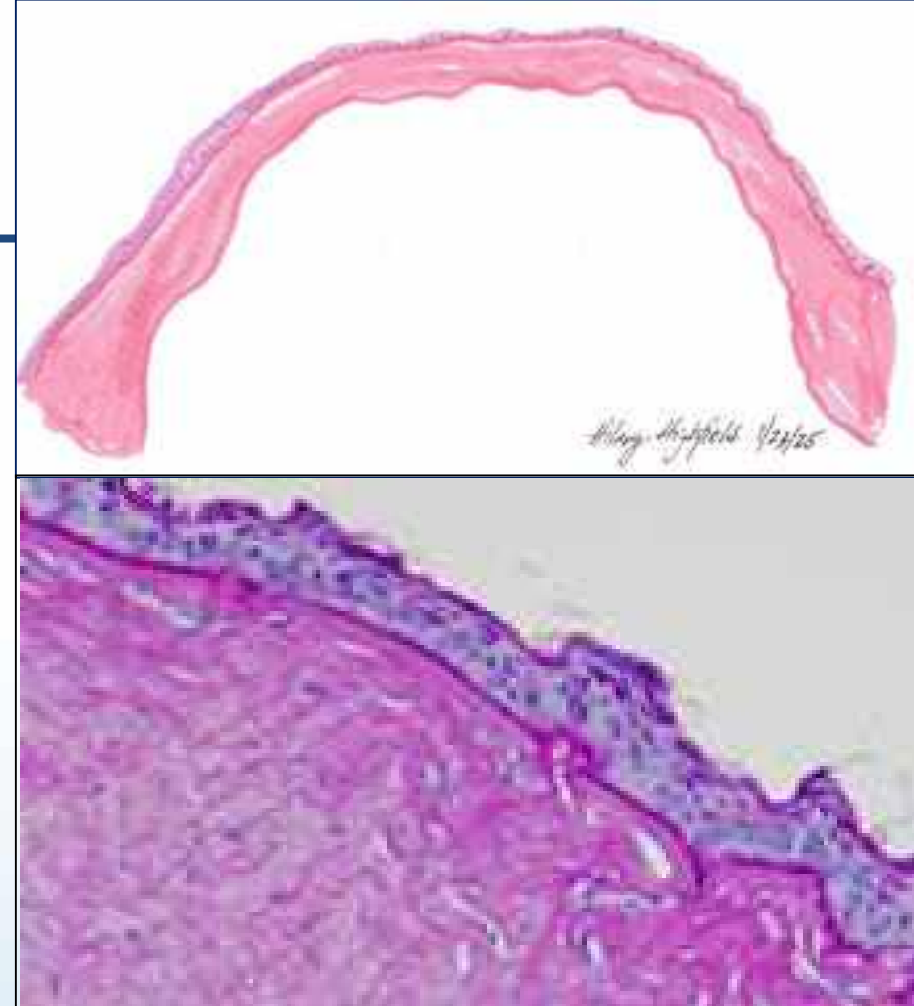
Keratoconus

- Non-inflammatory degenerative disorder
- Bilateral central corneal ectasia, with anterior protrusion of cornea (Mason's sign)
- Progressive thinning of central stroma
- Usually presents around puberty
- May be associated with systemic disorders
 - Atopy
 - Down syndrome
 - Marfan syndrome

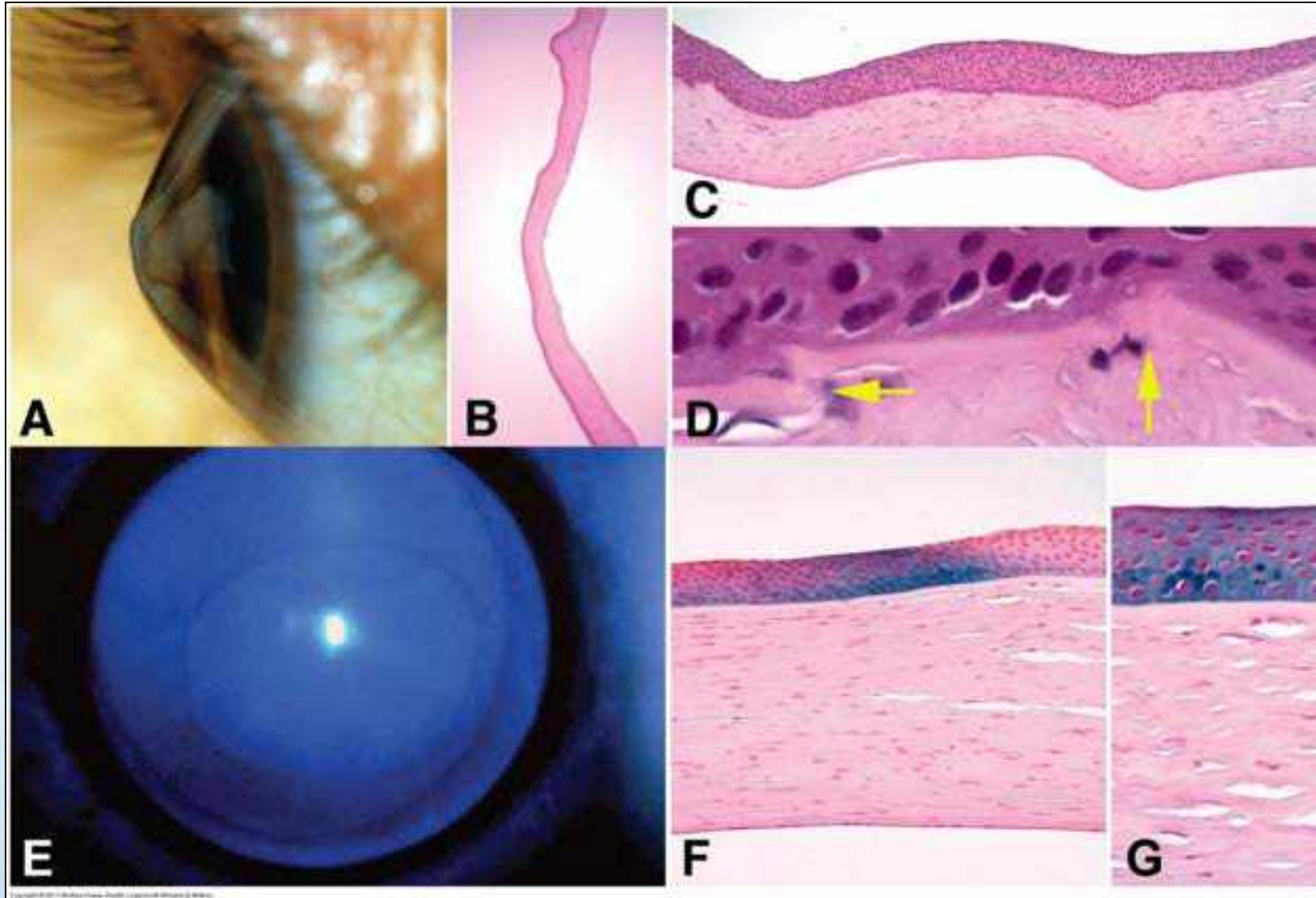


Keratoconus: Histologic findings

- Focal discontinuity of epithelial basement membrane and Bowman's layer
- Central stromal thinning
- Anterior stromal scarring
- Breaks in Descemet's membrane lead to acute stromal edema: hydrops
- Iron staining shows focal iron deposition in basal epithelium (Fleischer ring)



Keratoconus



Eagle, Ralph C. (2017), *Eye Pathology: An Atlas and Text*, 3rd ed. Wolters Kluwer

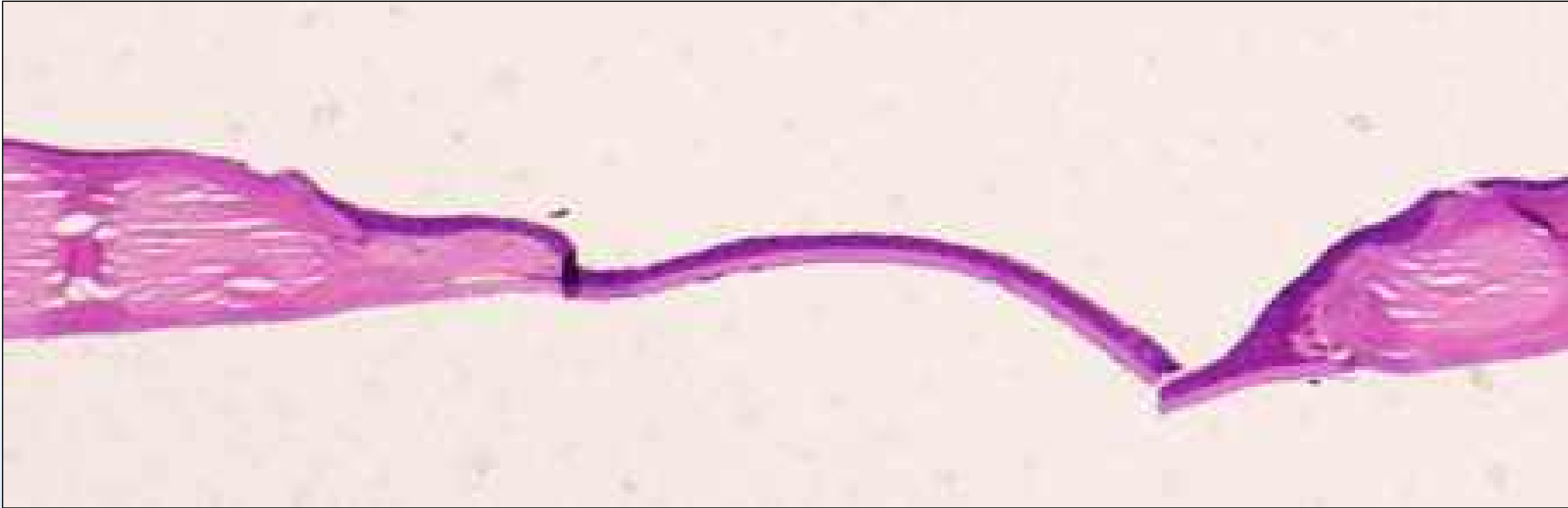
DESCEMETOCELE



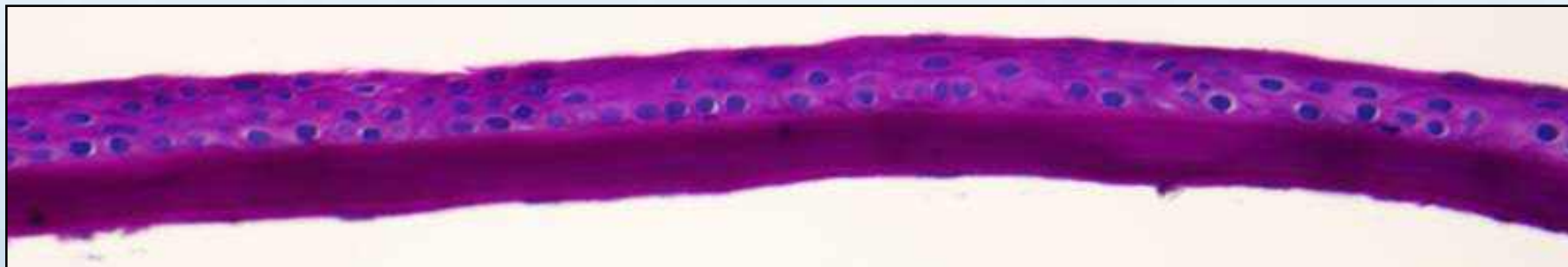
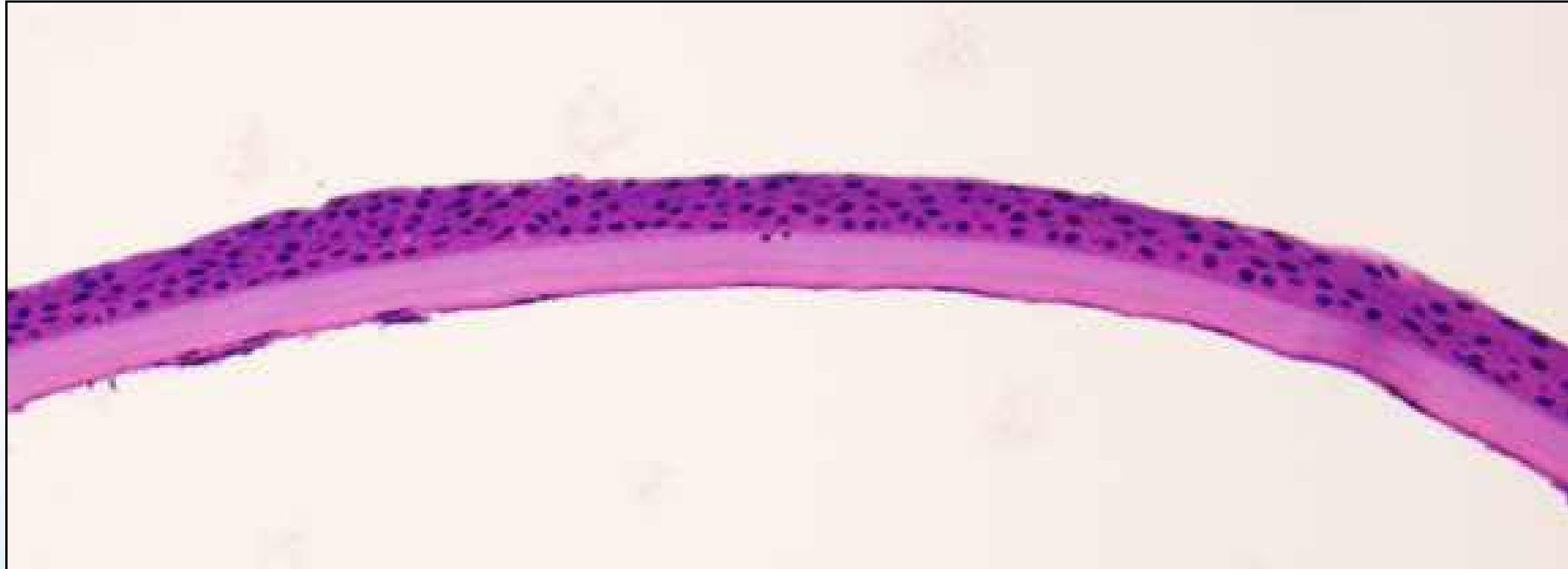
Descemetocoele



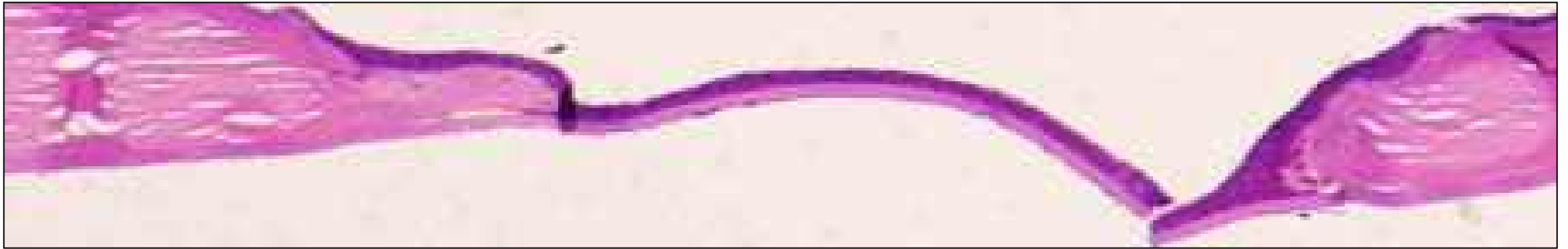
Descemetocèle



Descemetocèle



Descemetocoele



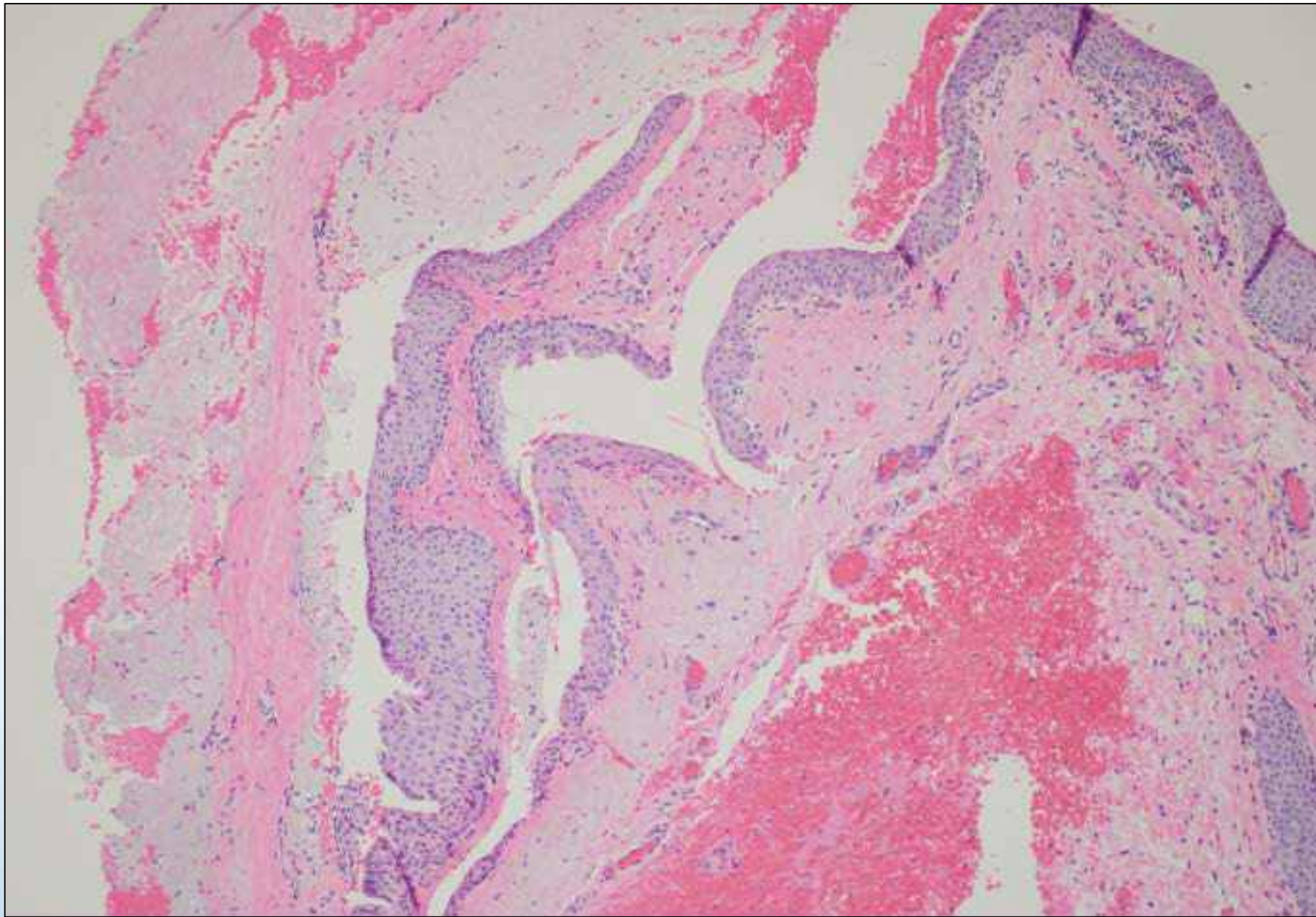
- An intact Descemet's membrane with an overlying absence of stroma
- Results in anterior corneal herniation.
- Descemet's membrane is the sole corneal layer maintaining the integrity of the eye.
- Often the result of corneal ulceration
- **There is imminent risk of corneal perforation.**
- Clinical scenarios resulting in corneal ulceration include microbial keratitis (bacterial and herpetic), neurotrophic ulceration (cranial nerve injury), dry eye disorders, trauma, and immune-related disorders.

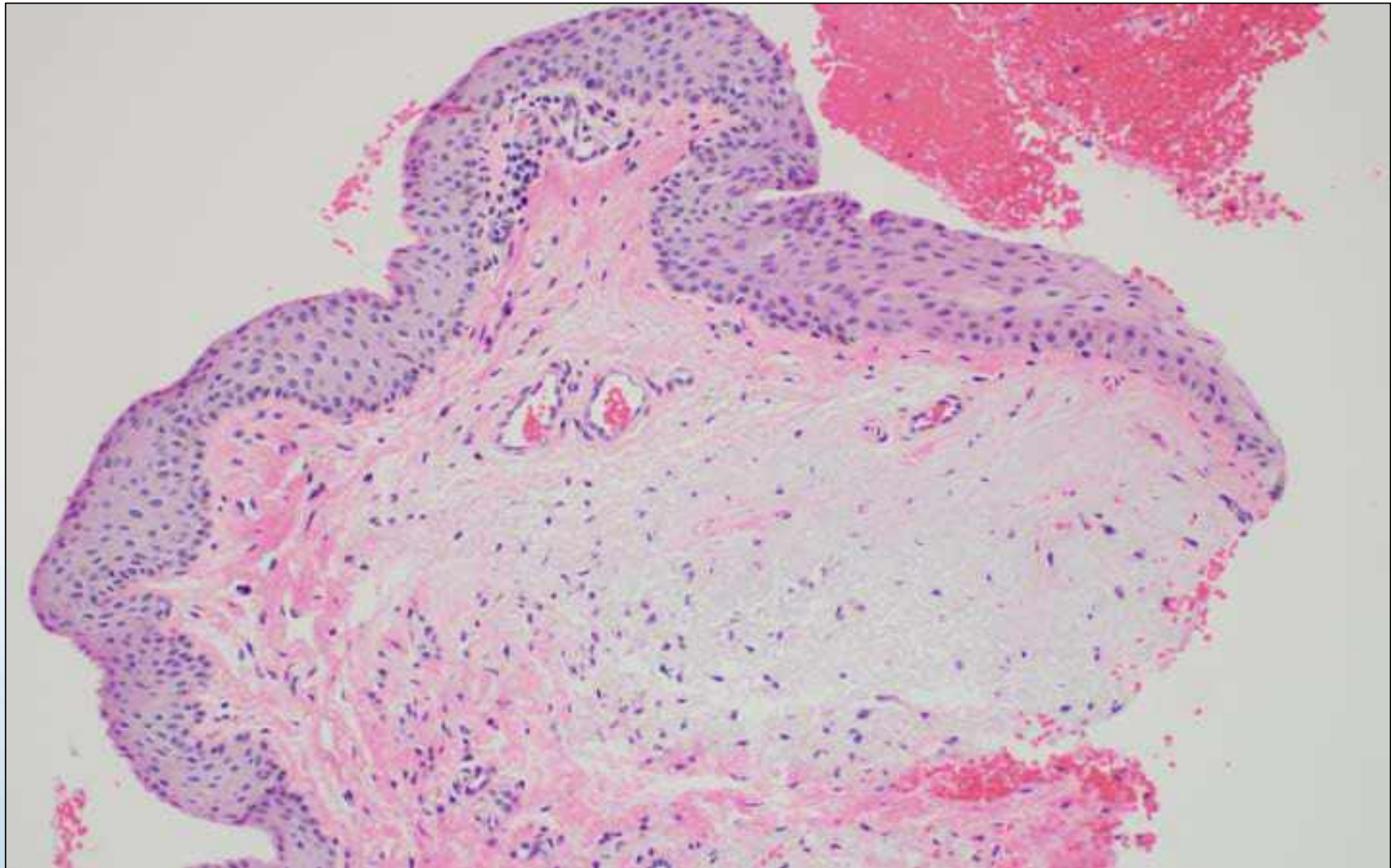
CONJUNCTIVA

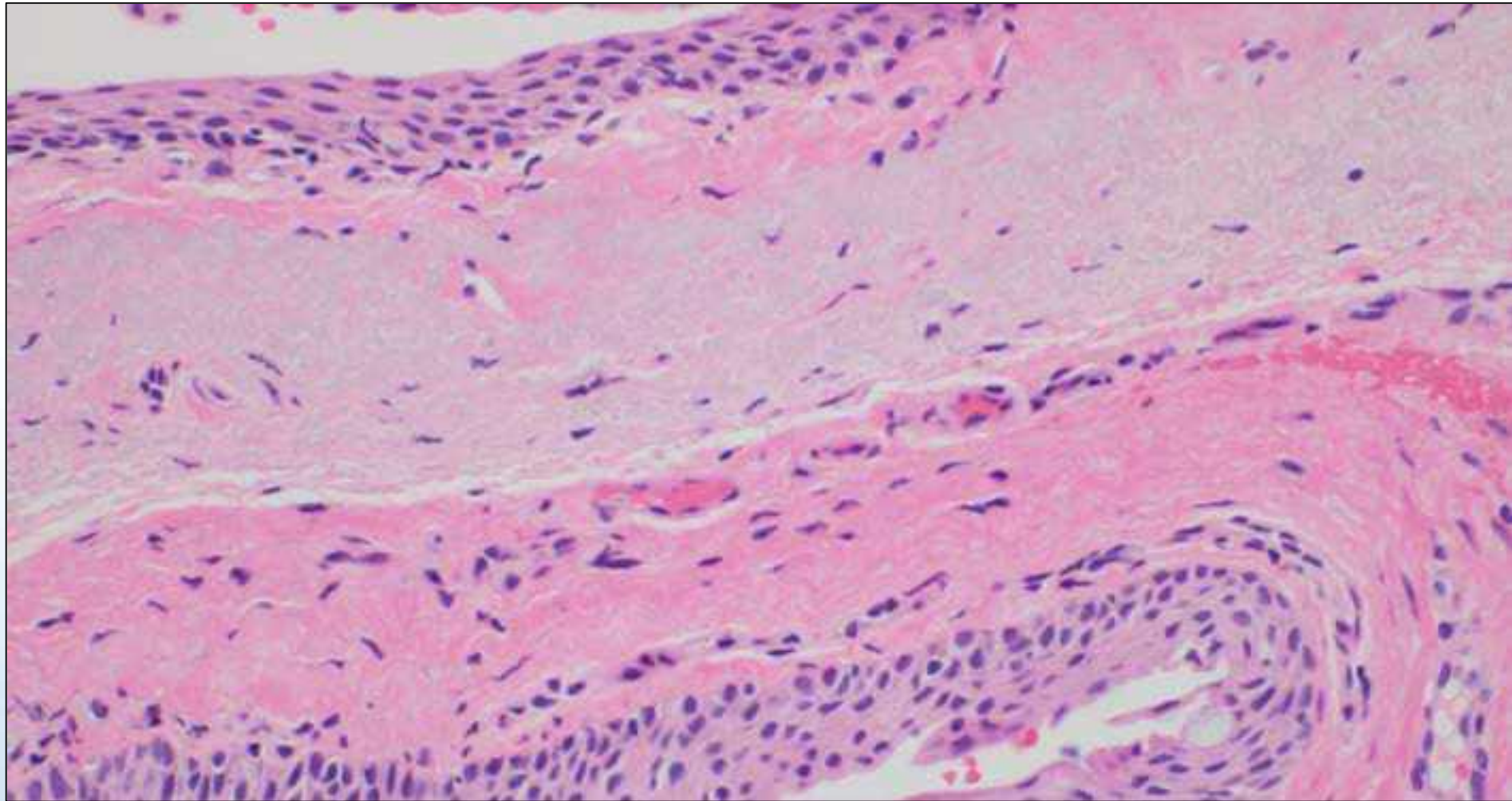


PTERYGIUM

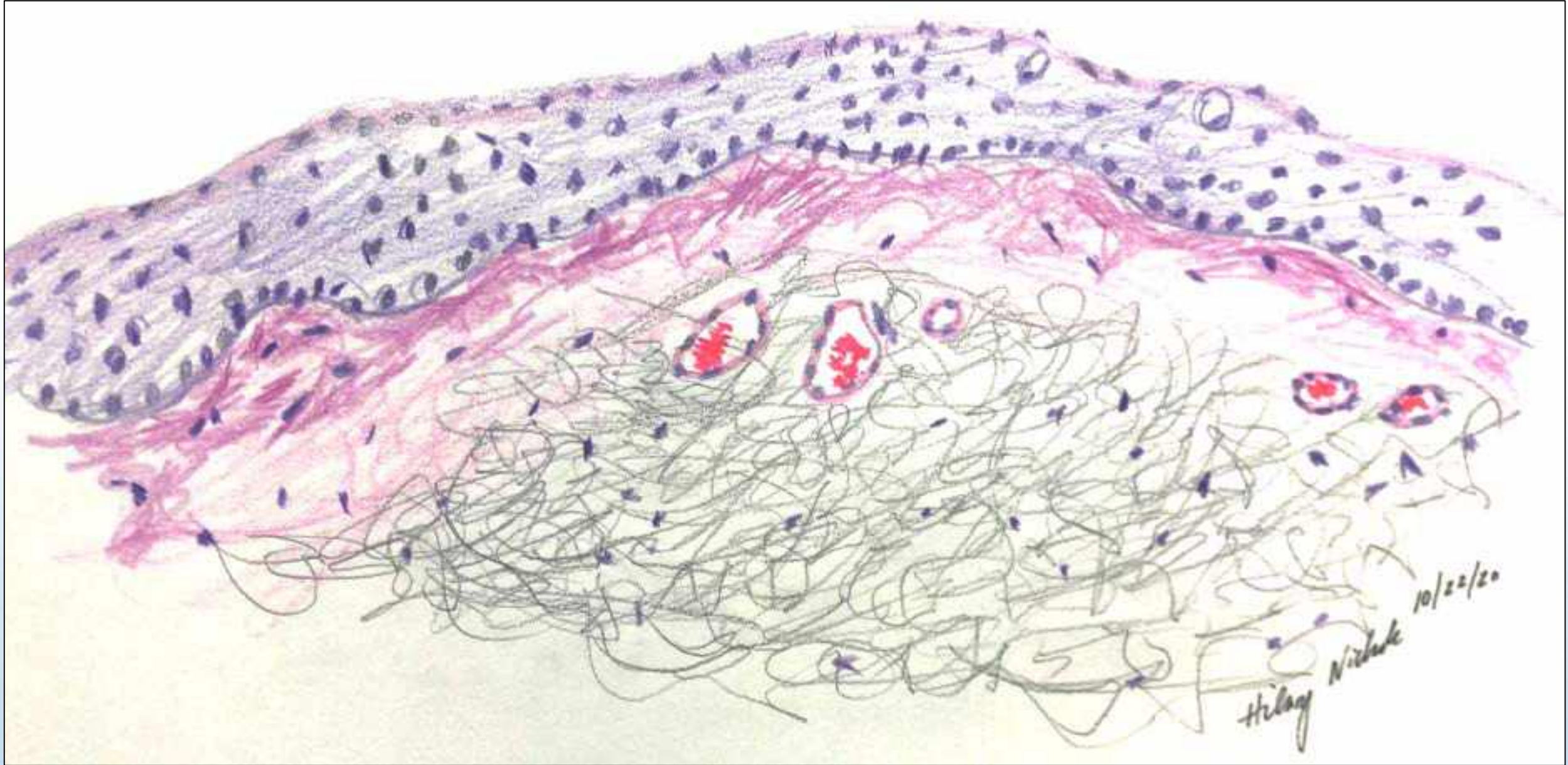


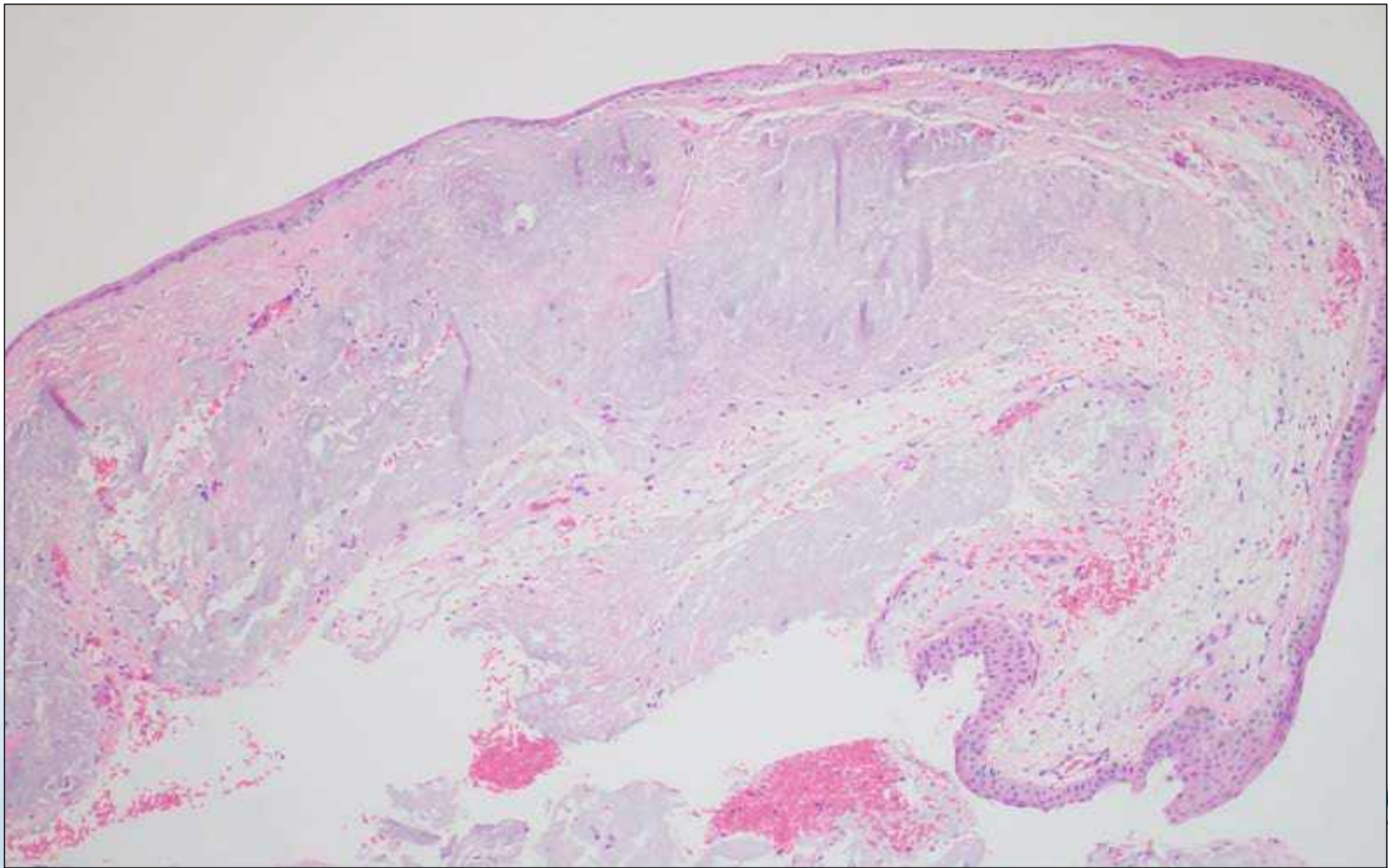


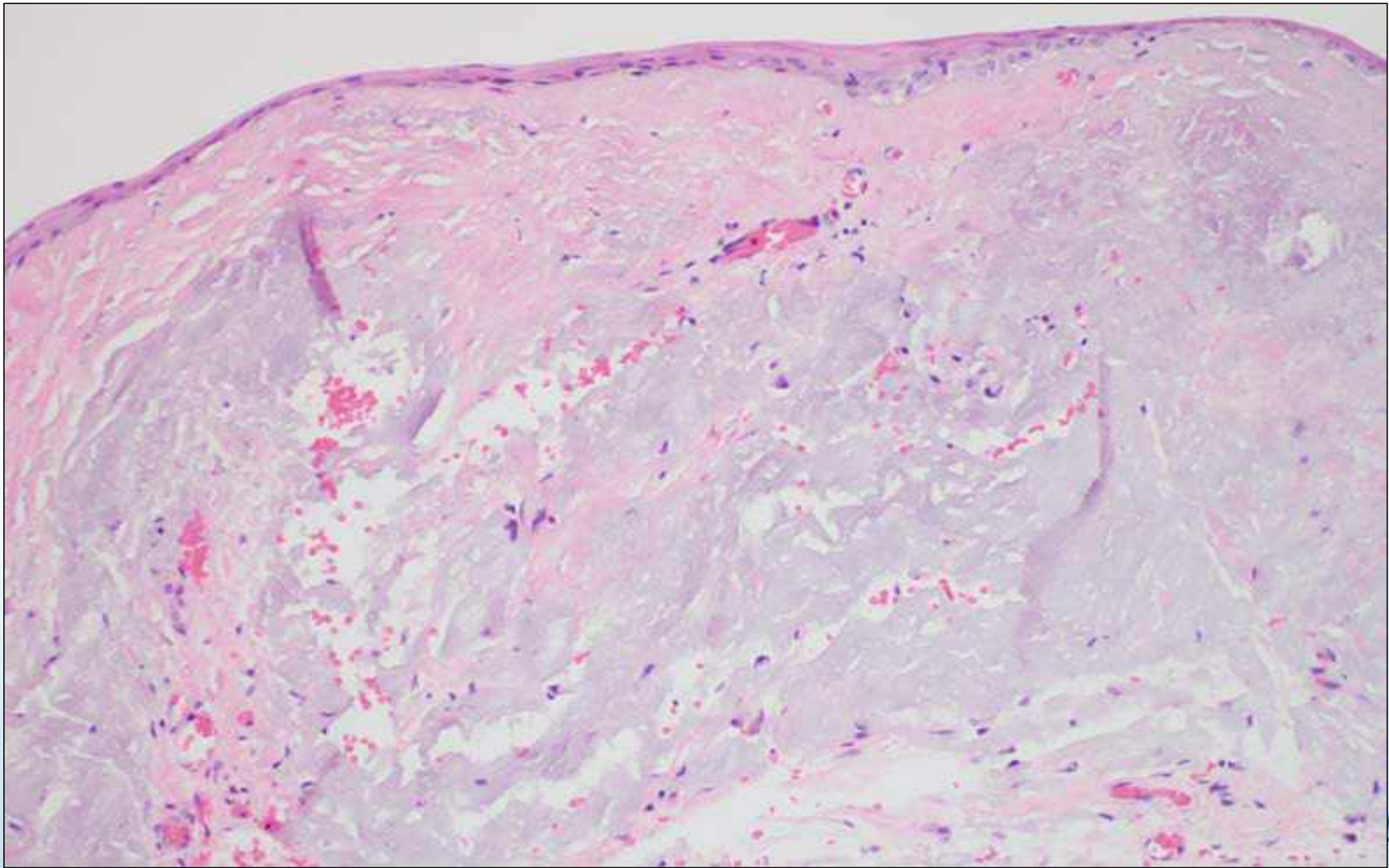




Pterygium



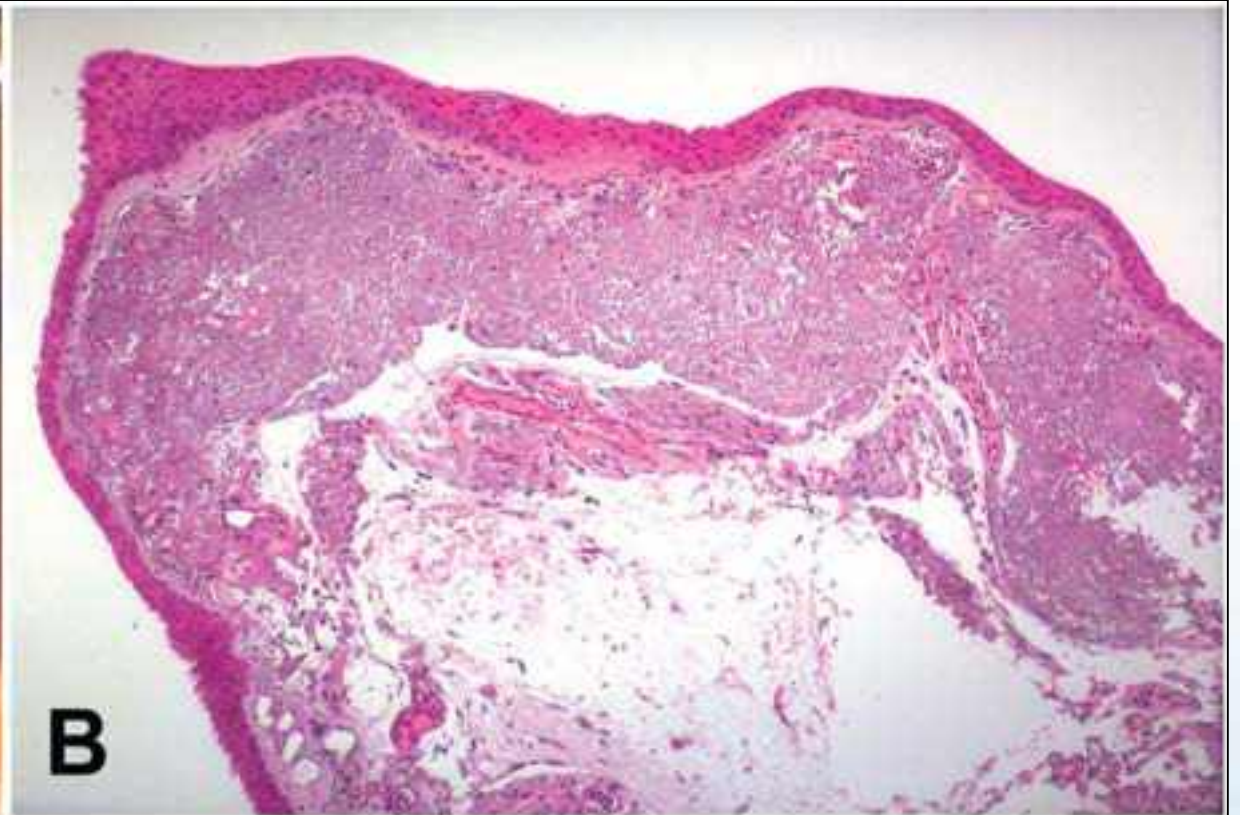




Pterygium



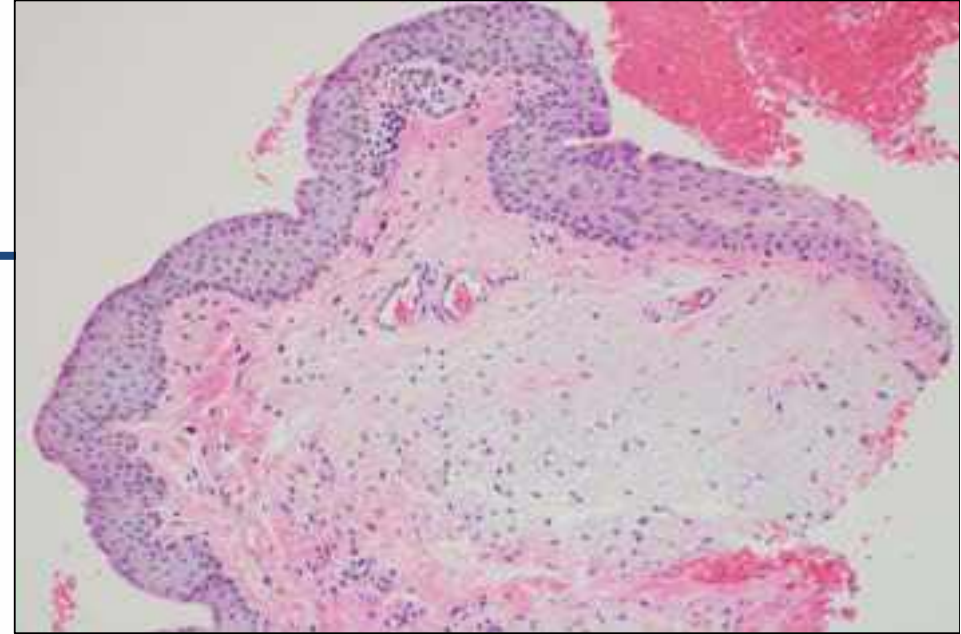
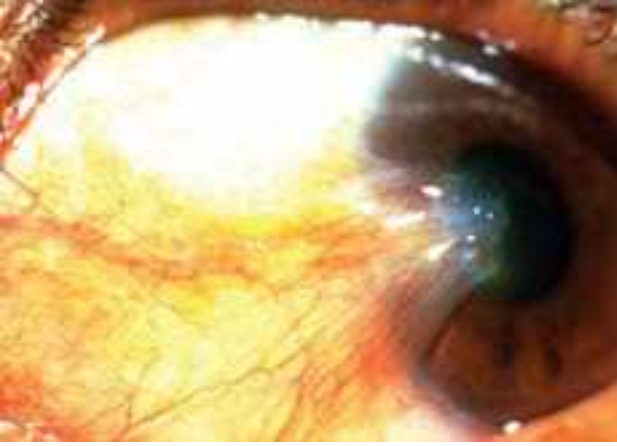
Pterygium



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Pterygium



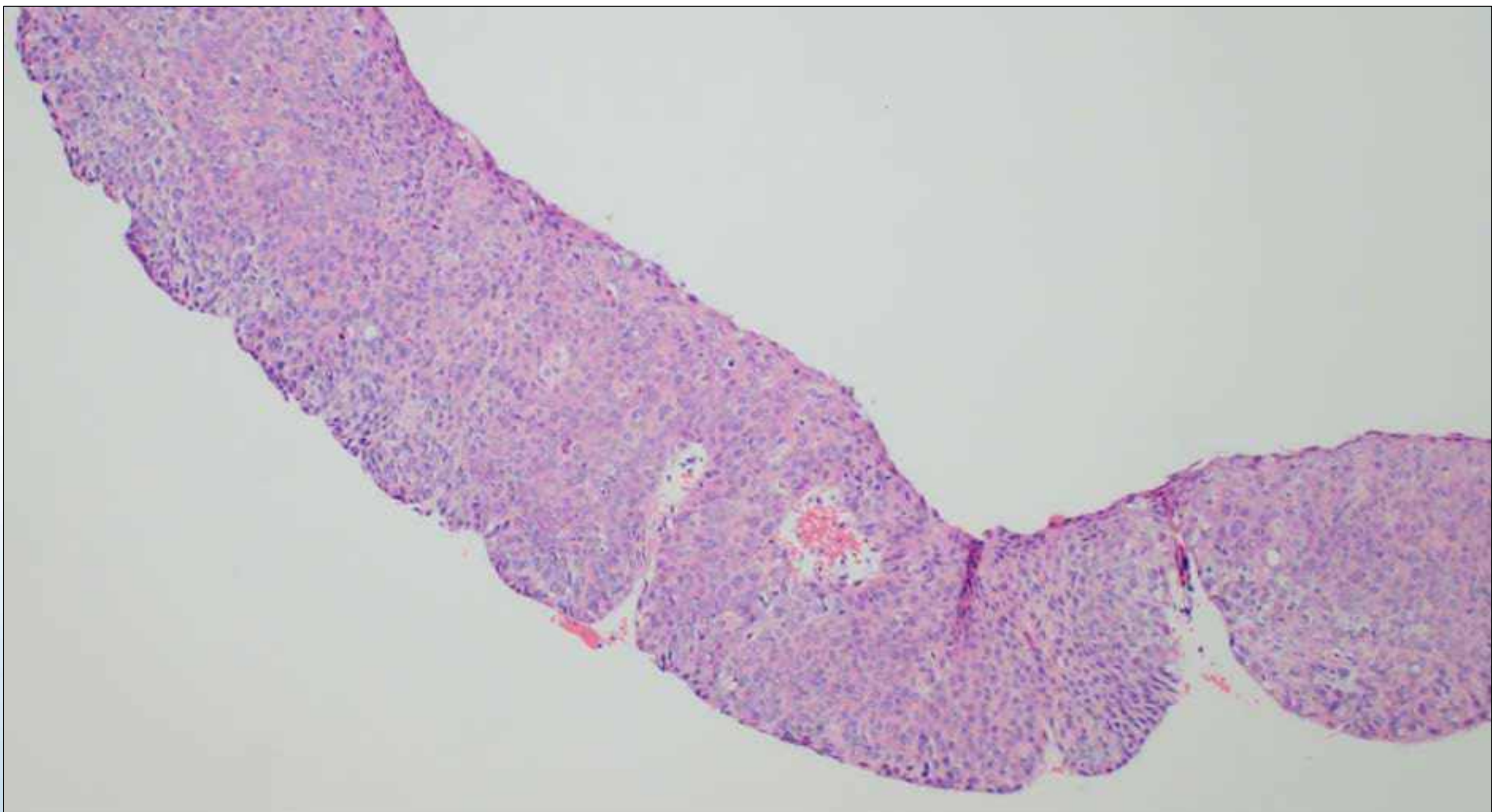
- *pter* is Greek word for wing
- Degenerative
- Wedge-shaped ingrowth conjunctival tissue
- Invades peripheral cornea
- UV/environmental exposure
- Solar elastosis/actinic damage



CIN/OSSN

Conjunctival Intraepithelial neoplasia
Ocular surface squamous neoplasia

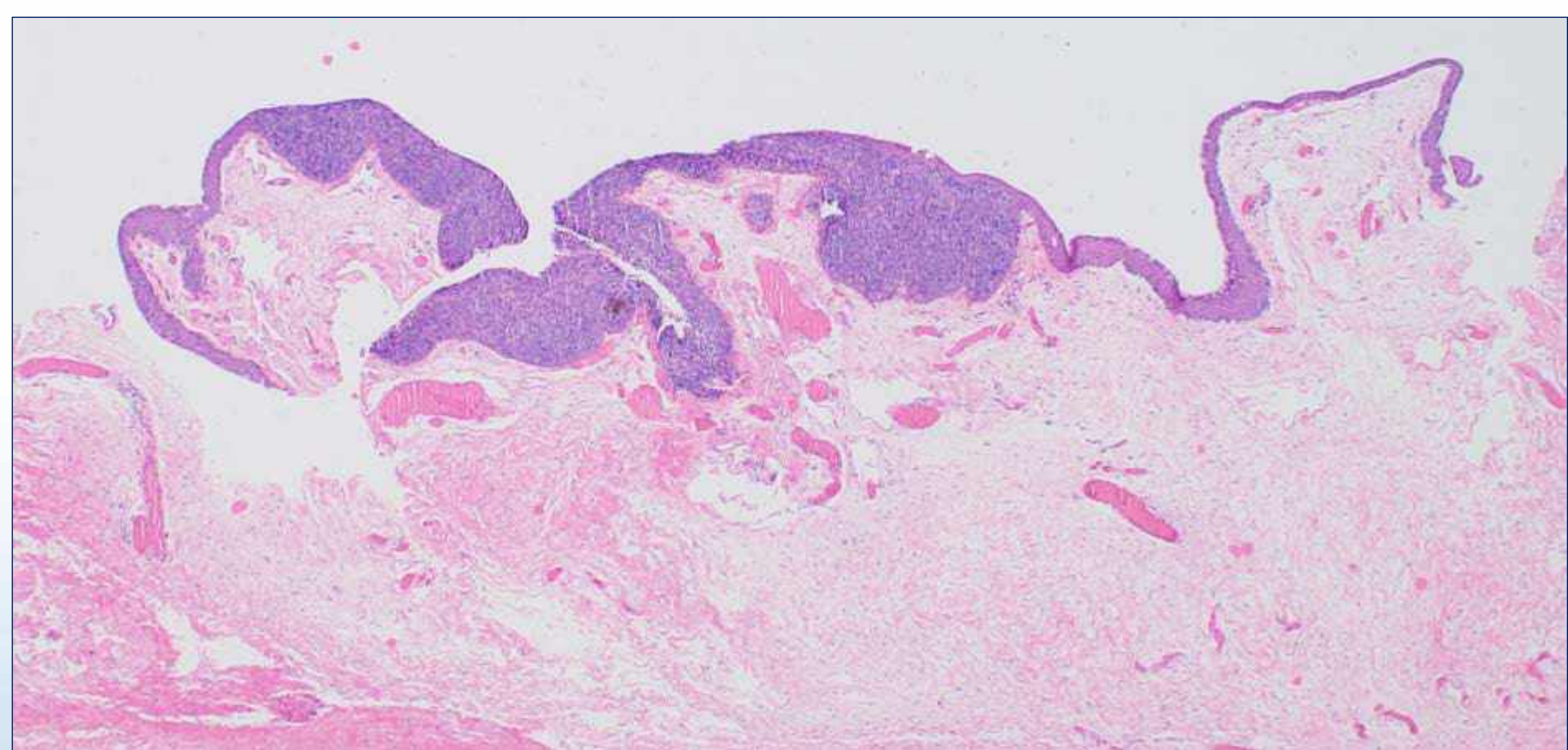


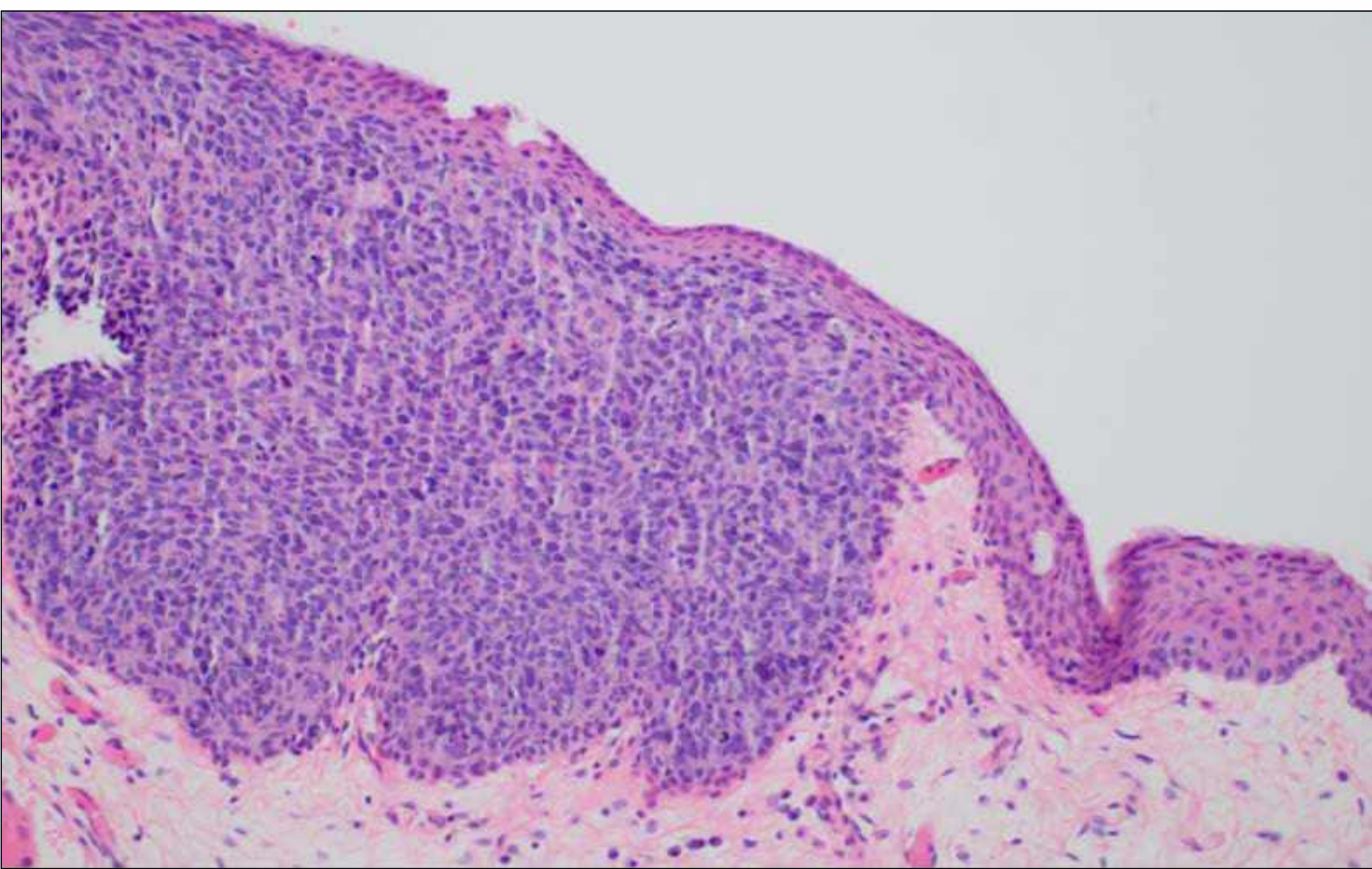


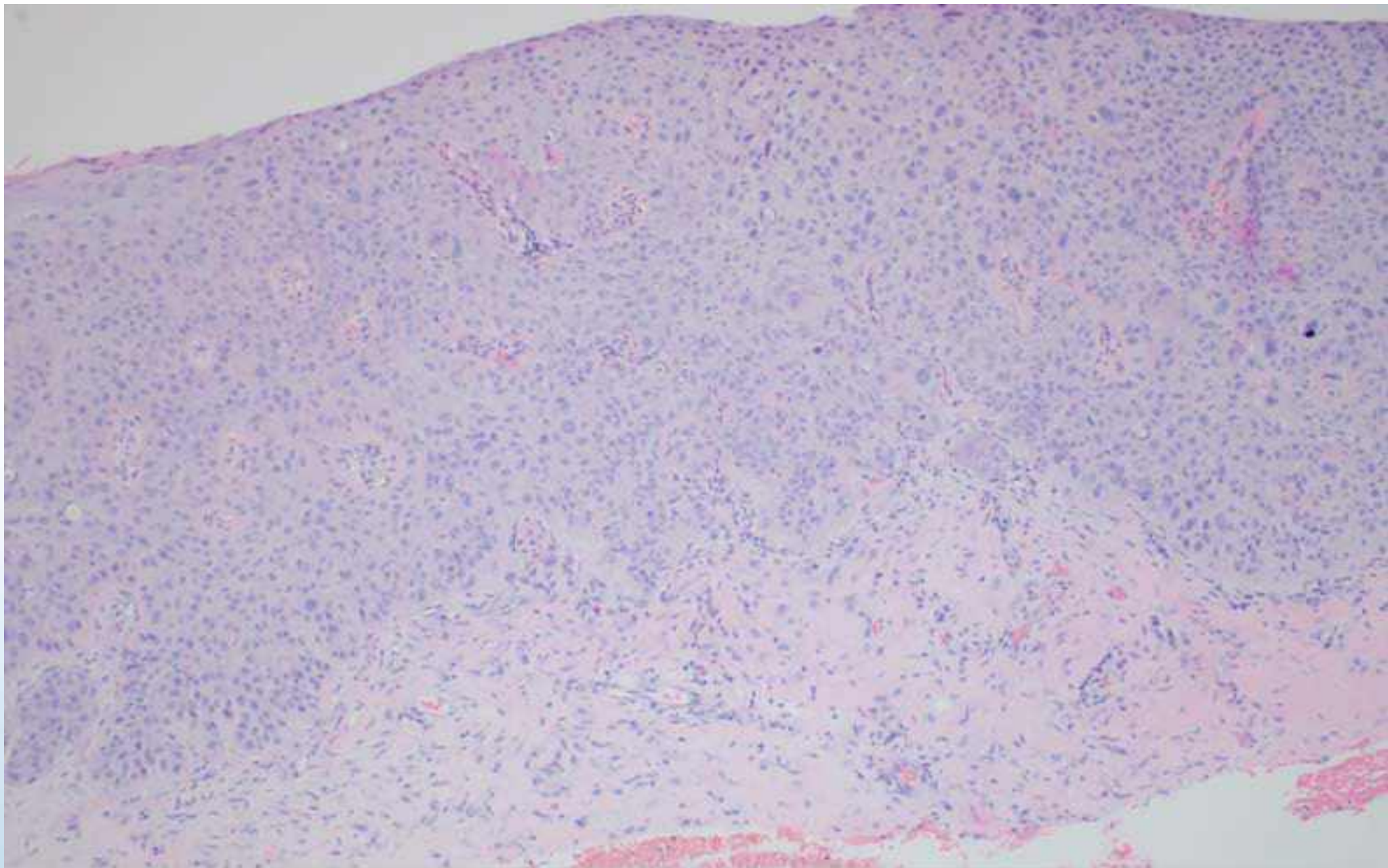


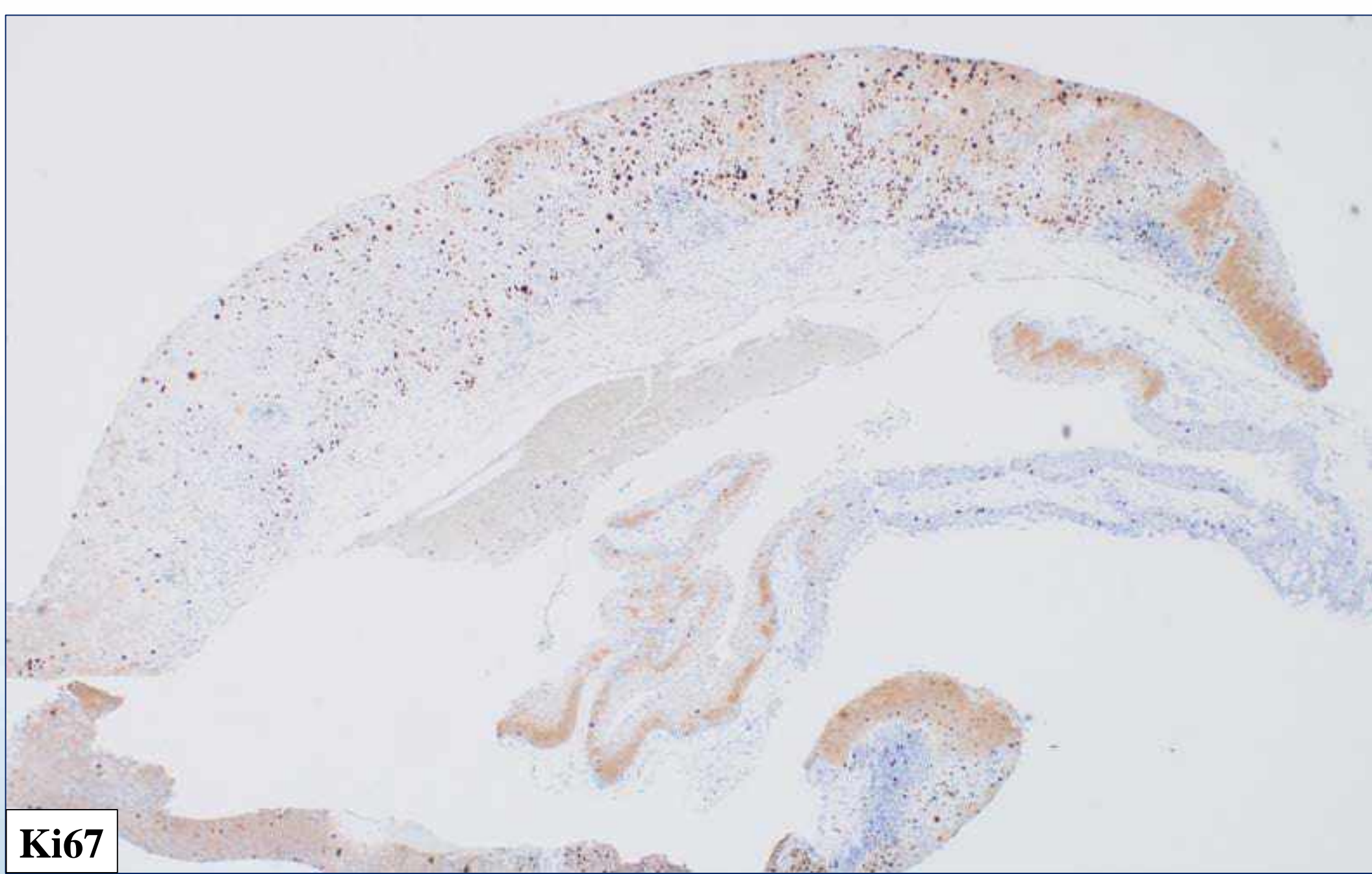
Ki67











Ki67



CIN/OSSN

- Conjunctival squamous cell carcinoma (CIN)/ Ocular surface squamous neoplasia (OSSN)
- Typically affects middle aged males, and is slow growing
- Complete surgical excision an effective treatment
- Younger individuals, particularly in equatorial Africa, there is a risk of HIV infection, and these lesions behave in a much more aggressive manner
- Risk factors for the development of CIN include
 - ultraviolet A and B exposure
 - ionizing radiation
 - HPV infection (6,11,16,18)
 - immune suppression (HIV, medication, organ transplant)
 - ocular injury
 - vitamin A deficiency

Sun, J. et al. Hum Path, 2017: 64-68.

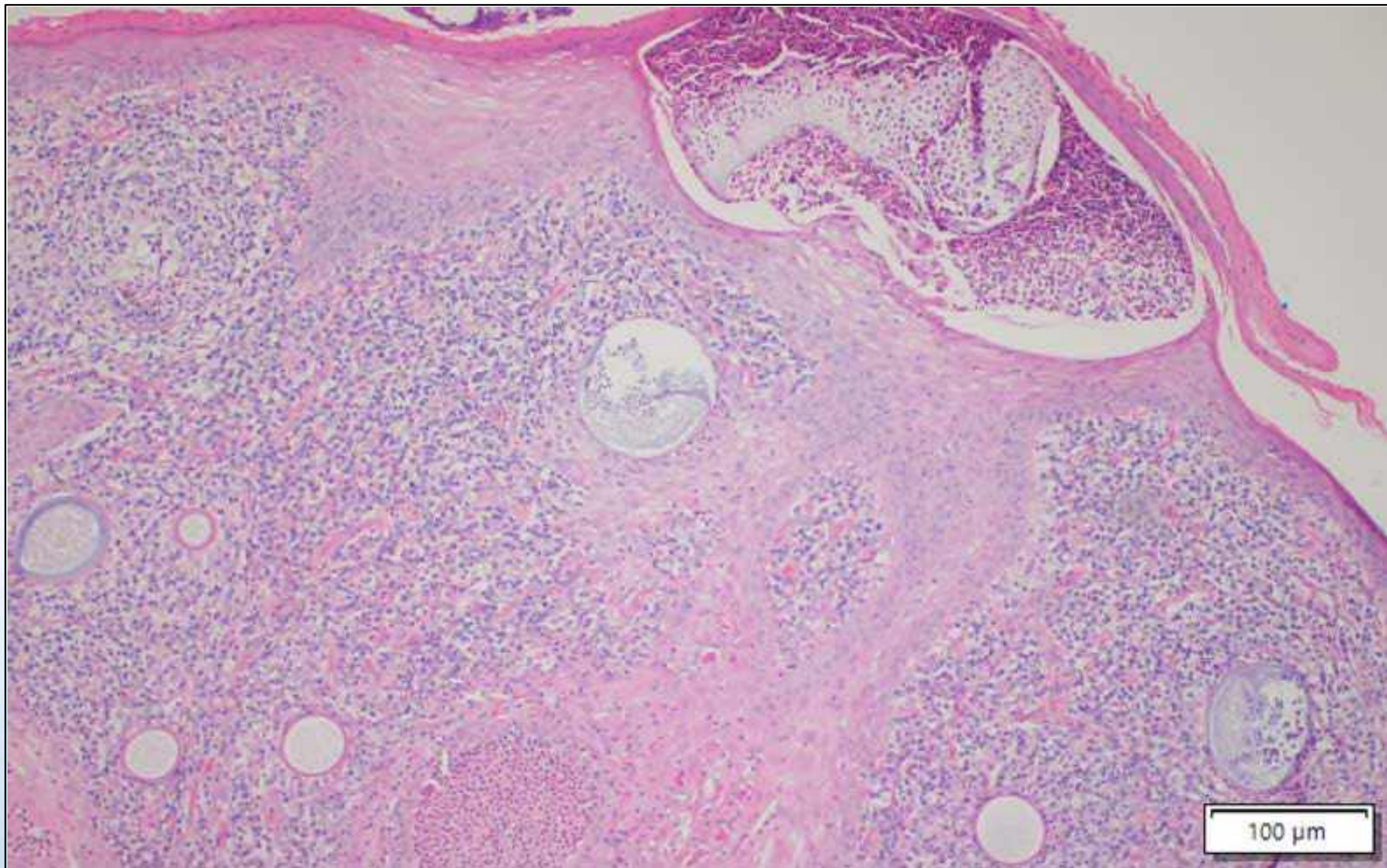
Shields, C. et al. 2011: 118 (11) pp 2133-2137.

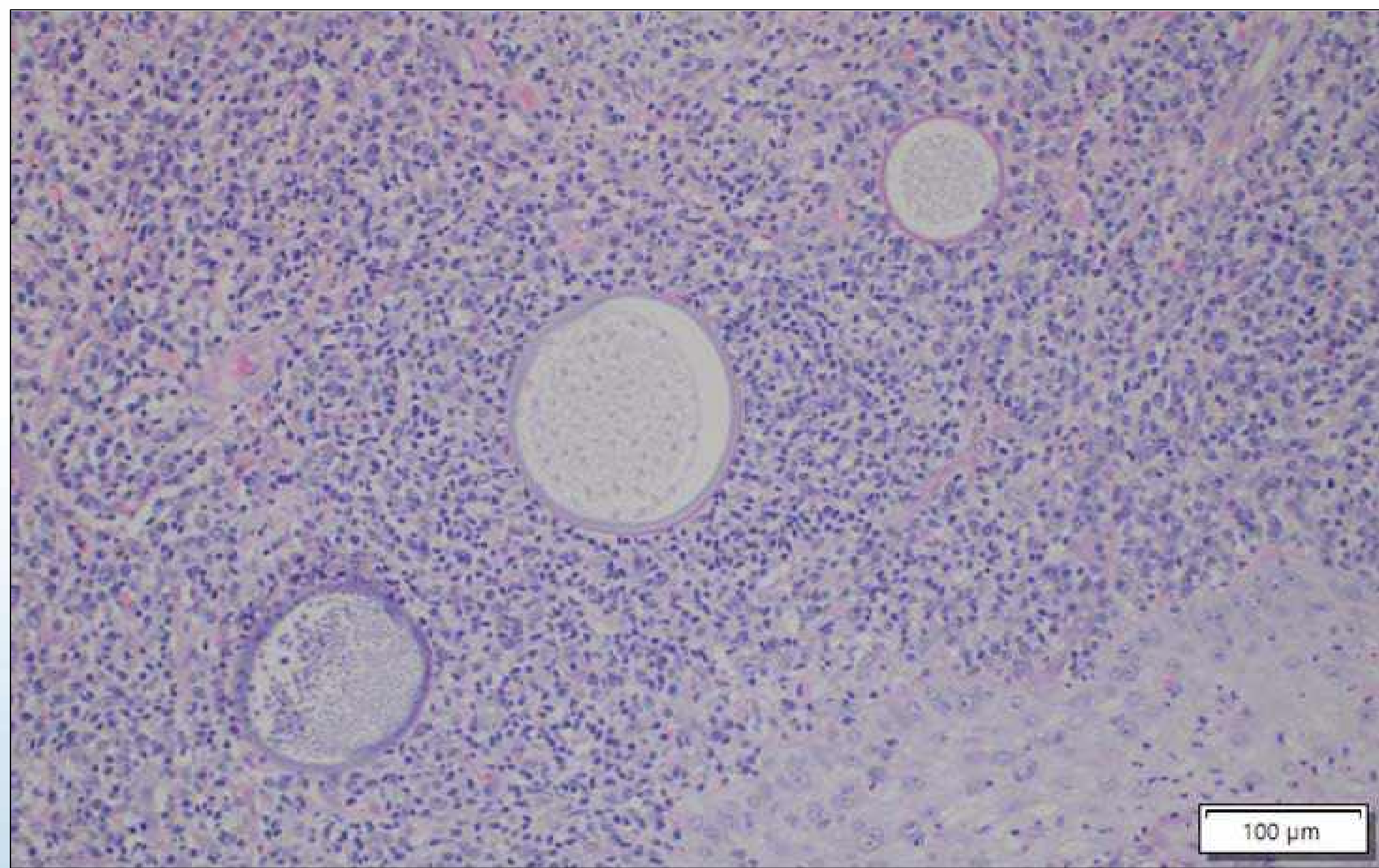


Case presentation



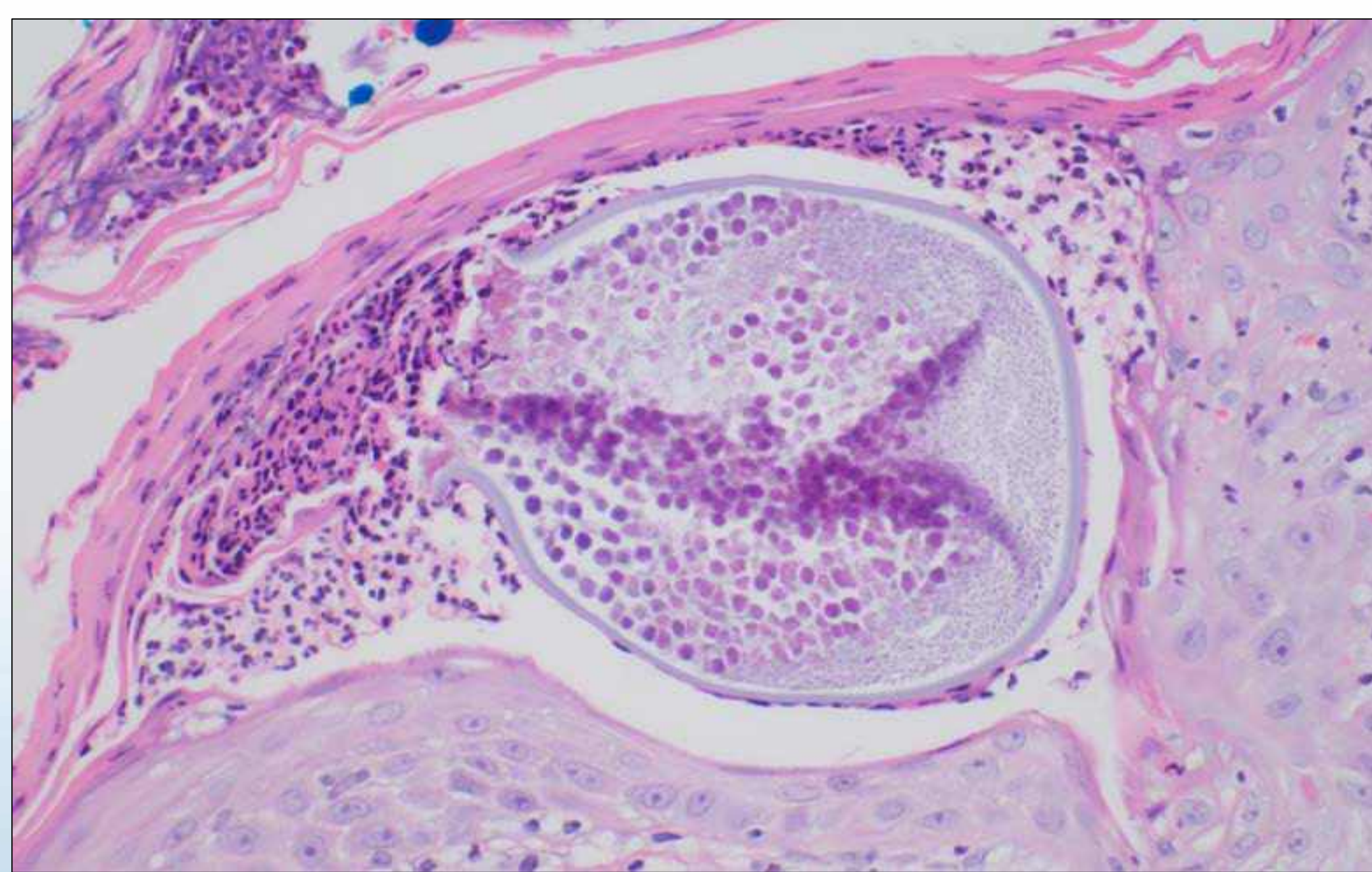
Bulbar conjunctival mass in a 62 year old man, clinically consistent with pyogenic granuloma.

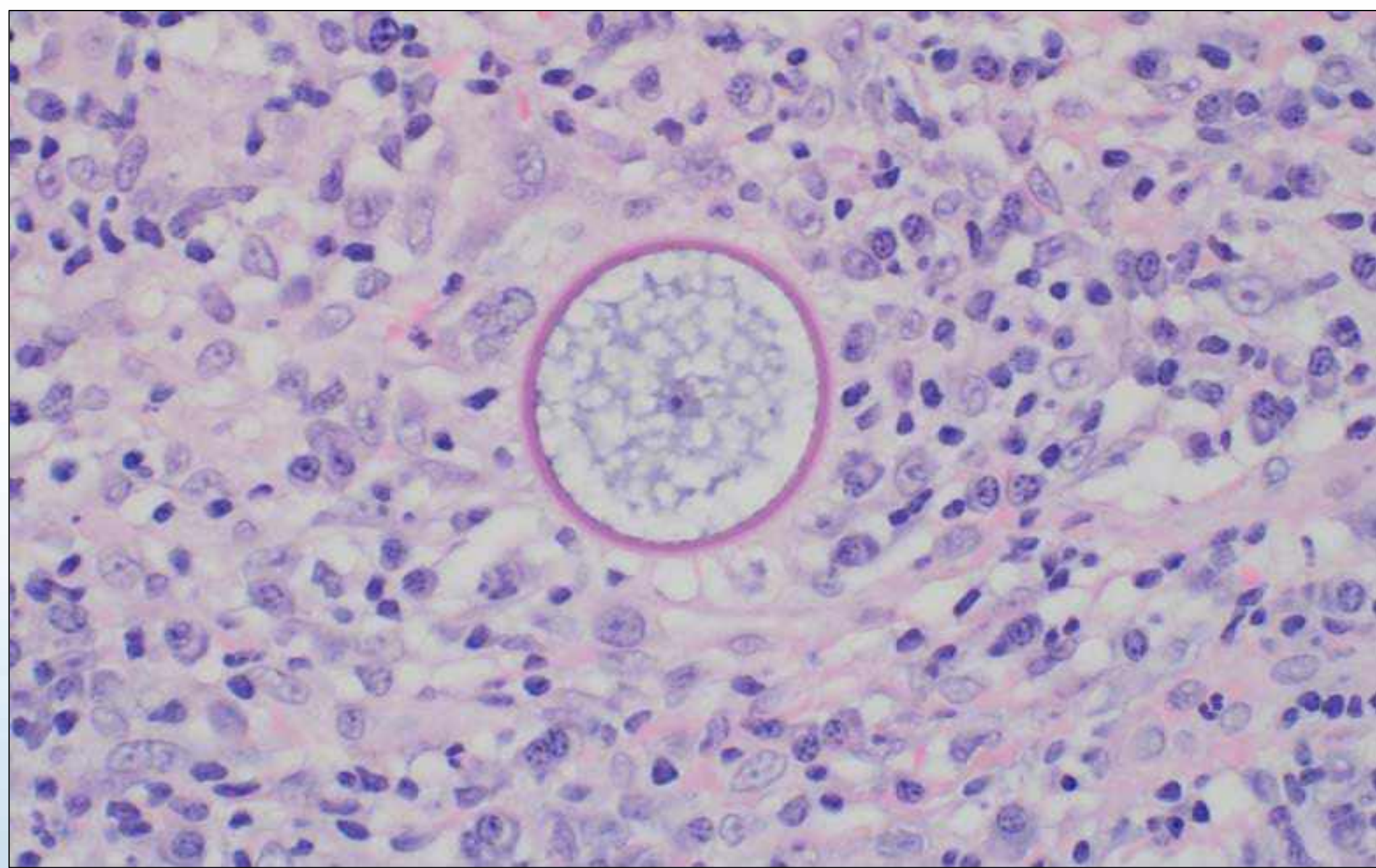


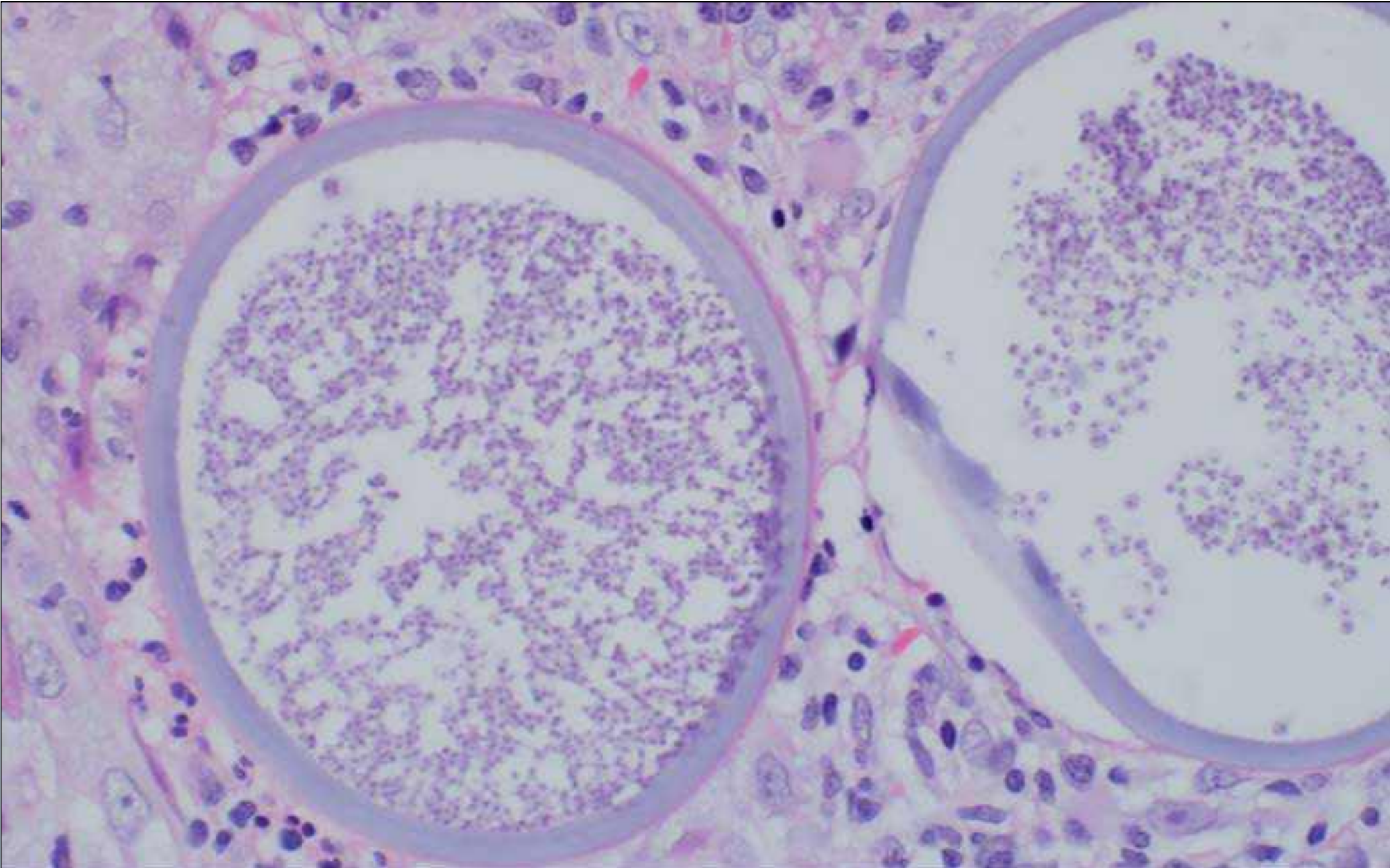


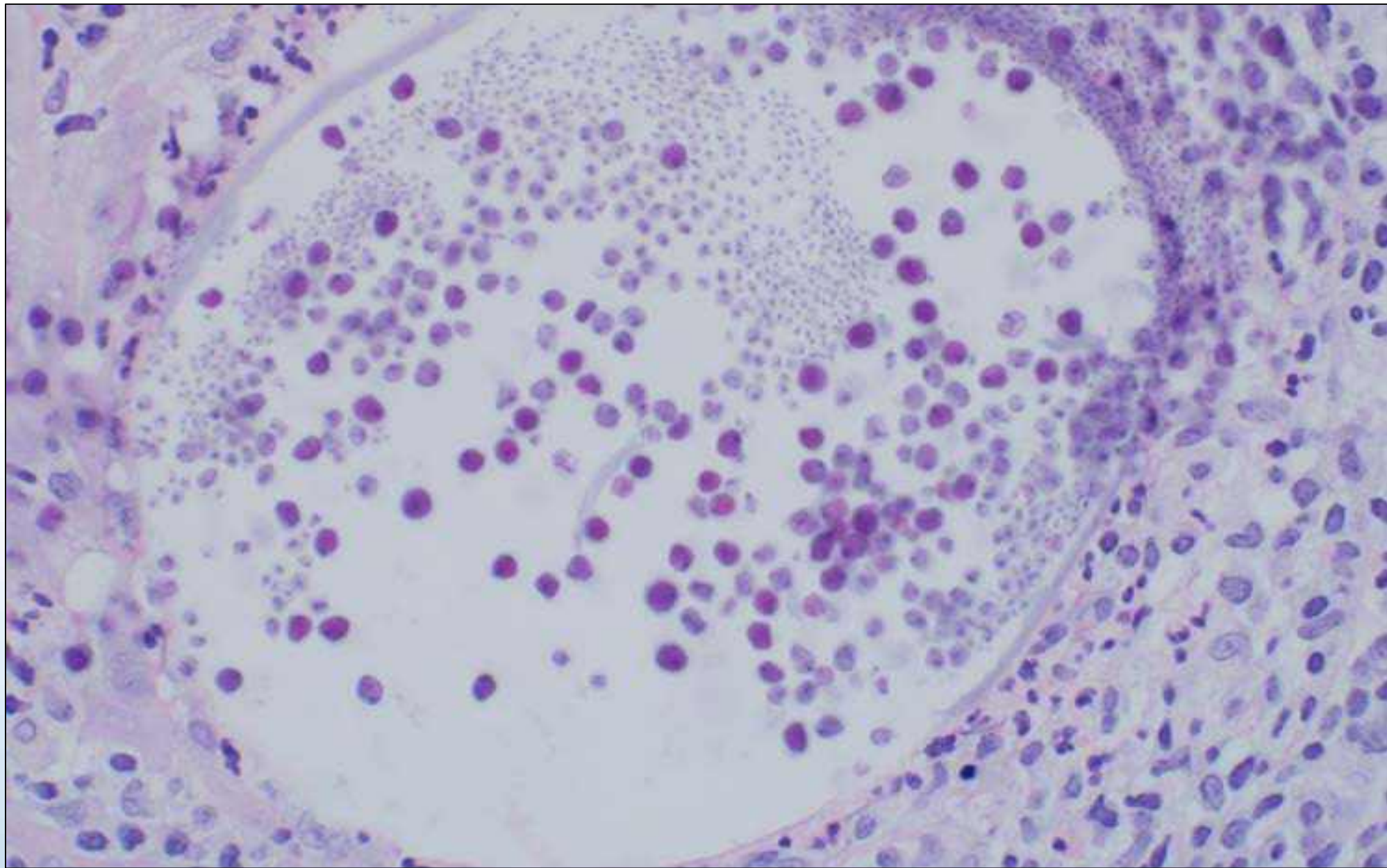
100 μ m

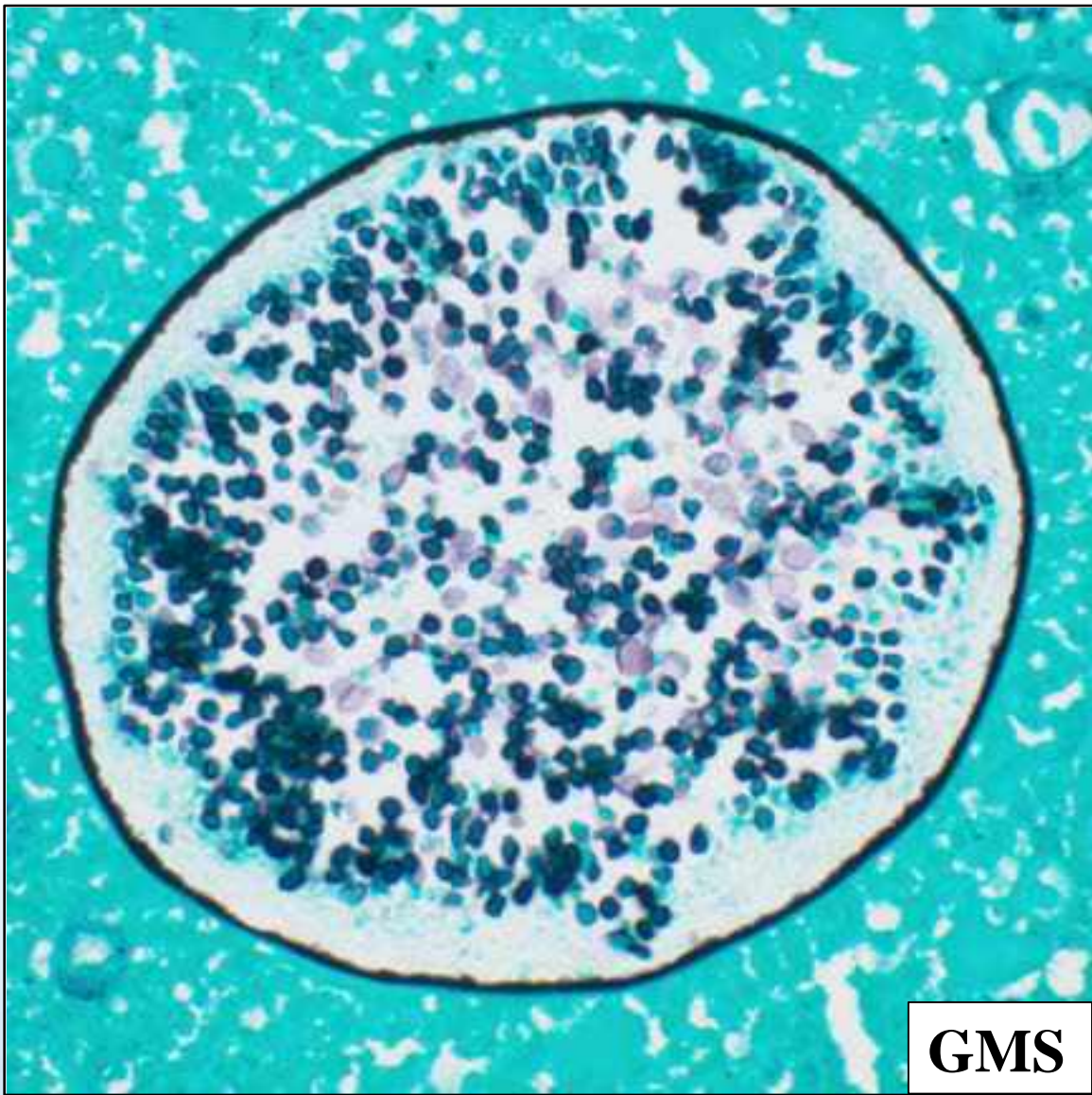








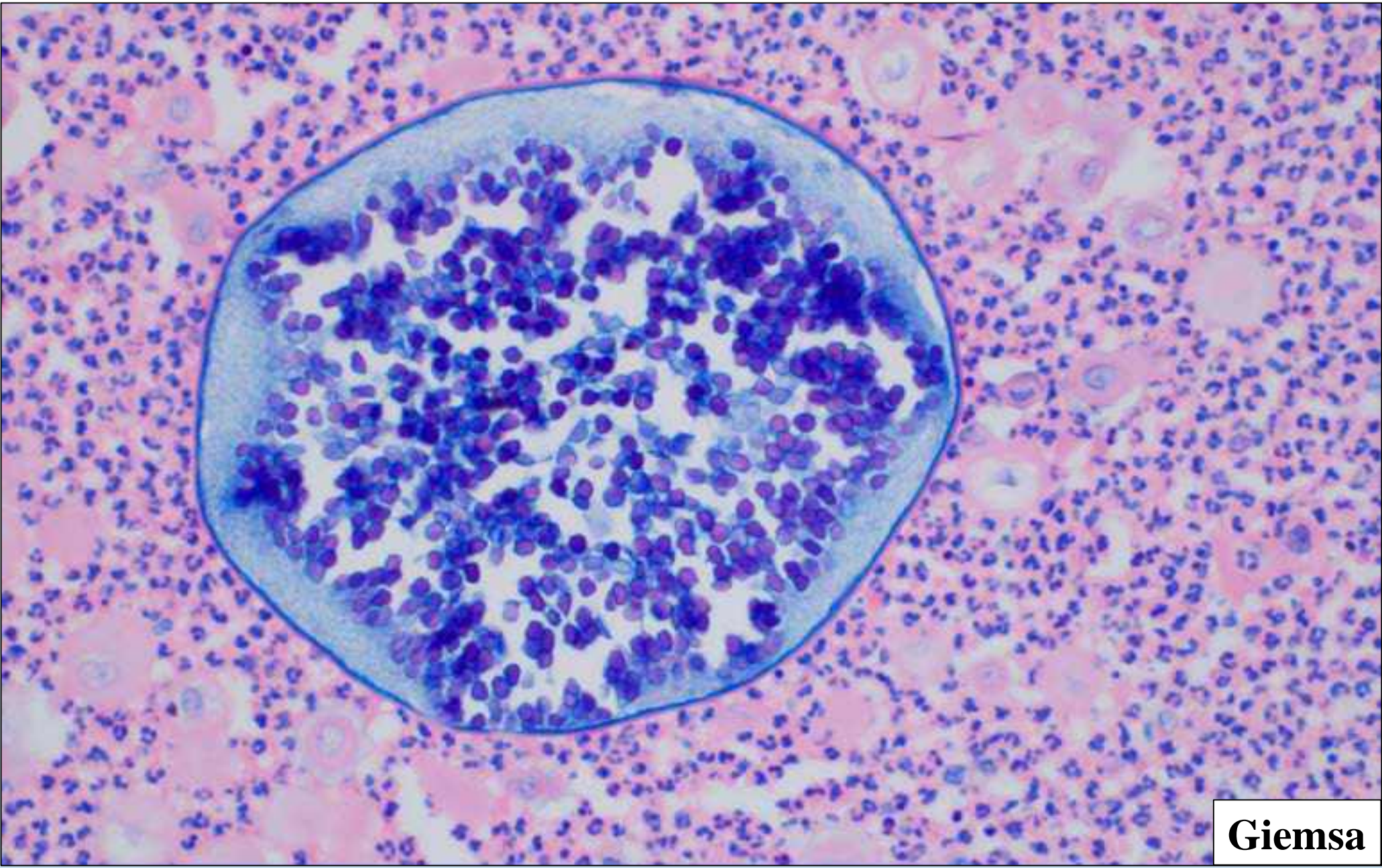




GMS



Mucicarmine

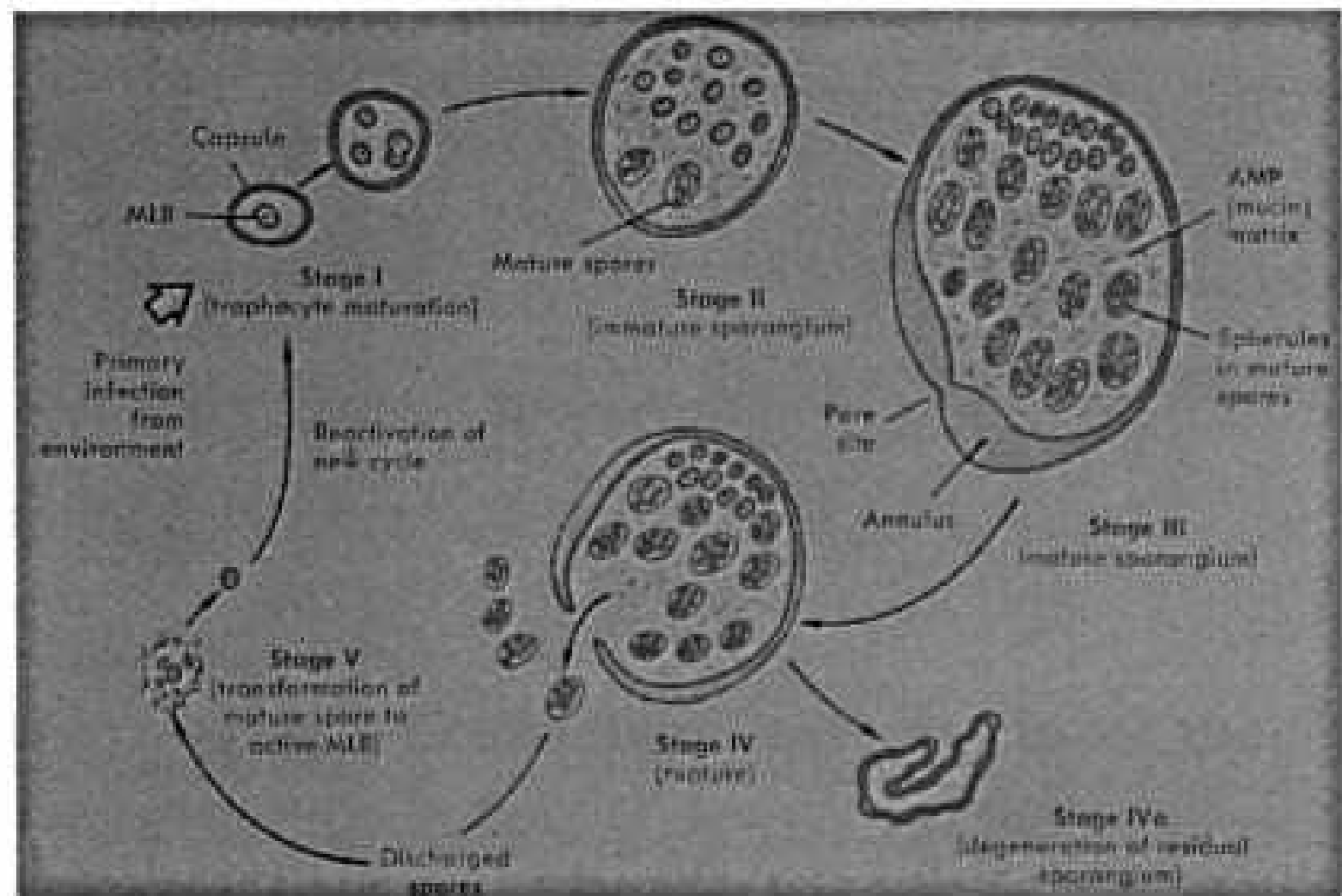


Giemsa

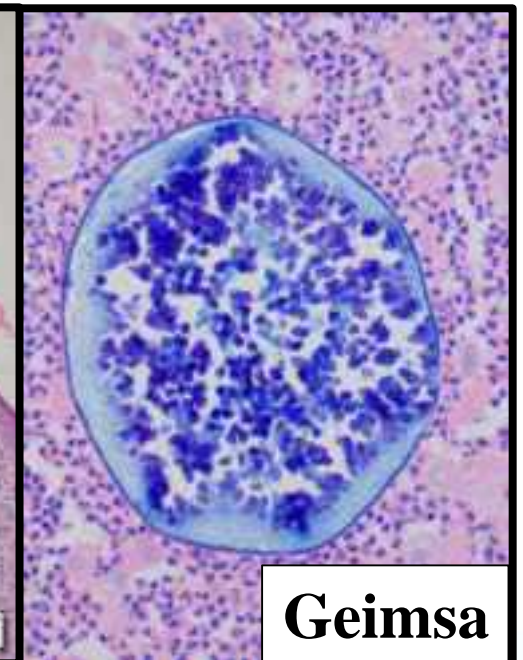
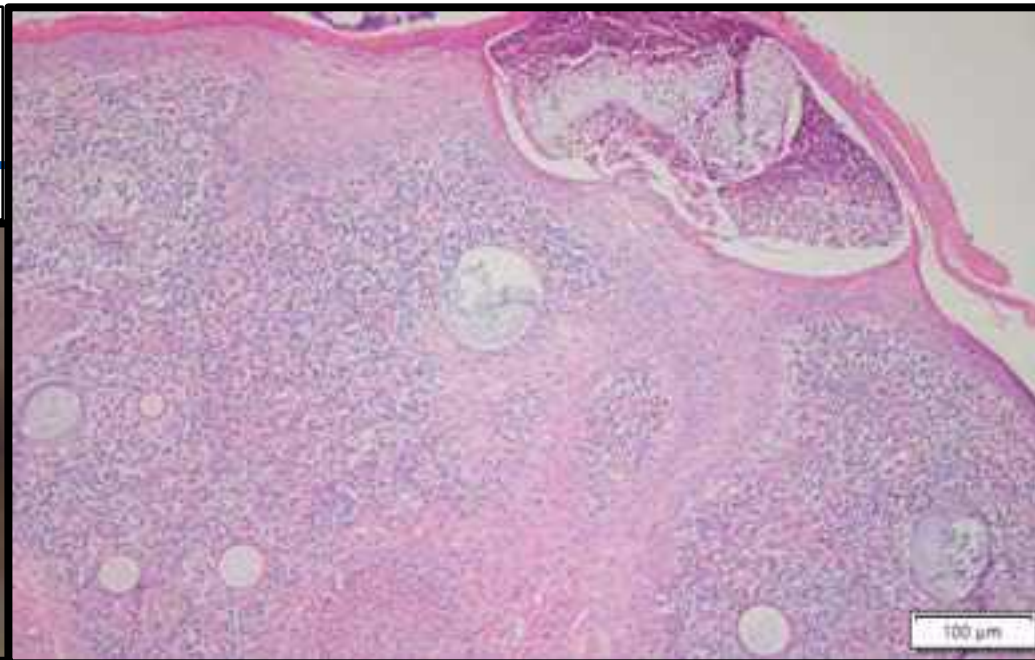
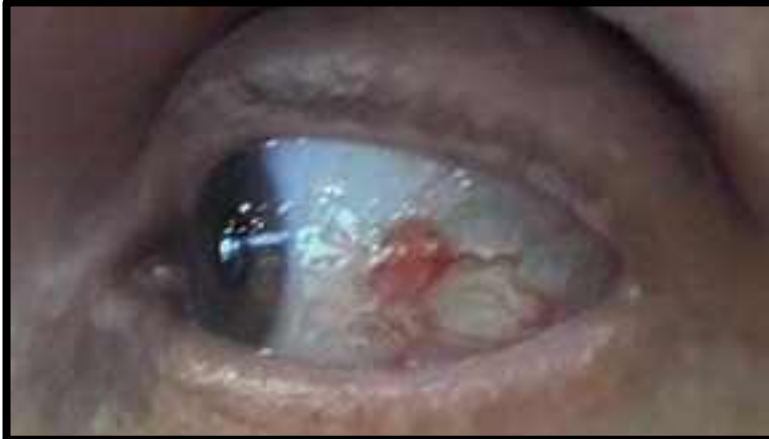




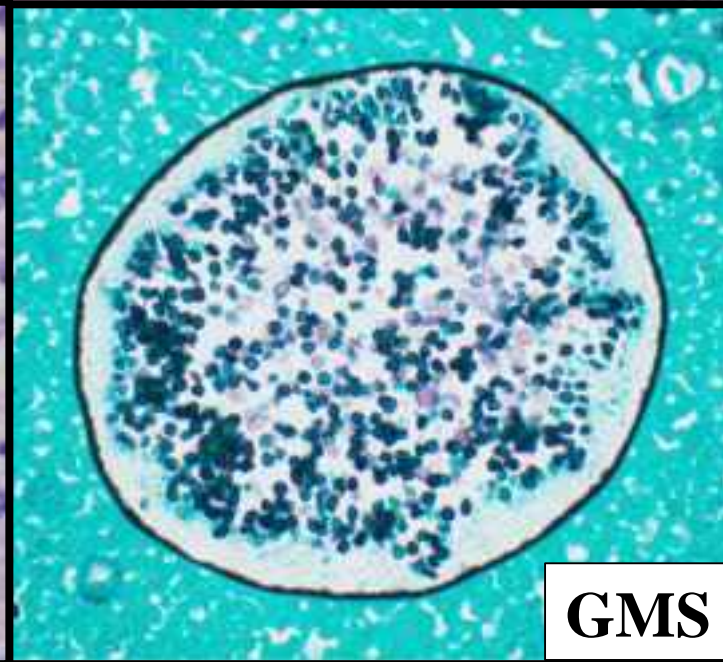
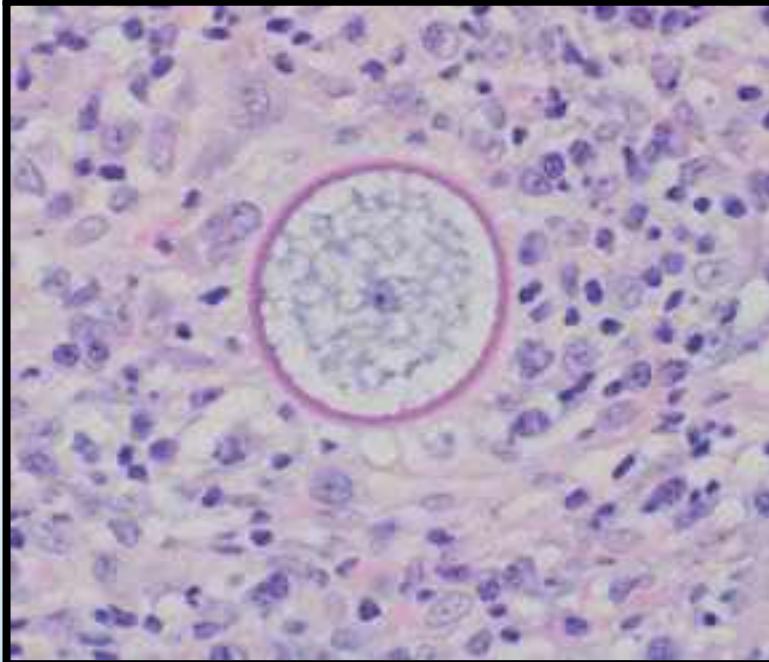
Fig 1. Life cycle of rhinosporidiosis. MLB- multilamellar body, the nucleic acid core of the organism. (Reprinted from Apple DJ, Rabb, MF: Ocular Pathology: Clinical Applications and Self-Assessment. St Louis, CV Mosby, 1991, ed 4, with permission of the author and publisher.)



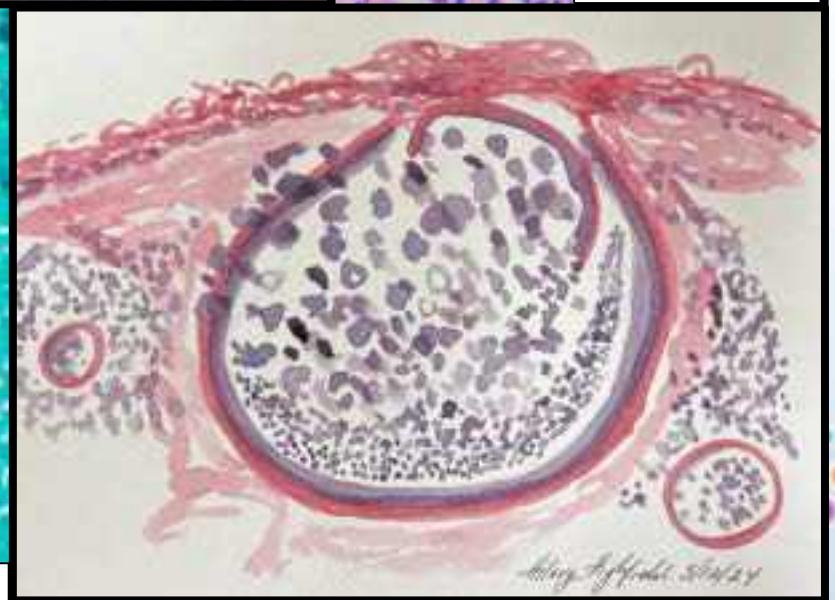
Rhinosporidiosis



Geimsa



GMS



Rhinosporidiosis

- Rhinosporidiosis is a rare infection caused by the organism *Rhinosporidium seeberi*.
- This endosporulating microorganism affects mucous membranes, causing a slow growing chronic granulomatous disease.
- The organism is in the class **Mesomycetozoea**, which includes microorganisms with features of both animals and fungi.
- Contact of damaged epithelium with contaminated water or inhalation of spore contaminated field dust are the proposed mechanisms of infection.
- The majority of reported cases are from India, followed by endemic parts of South America, with additional cases in tropical and subtropical areas of North America and Europe.
- Rhinosporidiosis is considered an emerging infectious disease.



Rhinosporidiosis: Clinical Spectrum

- Localized vascularized masses with chronic granulomatous reaction in the sinonasal cavity or eye.
- Can involve the genitourinary tract, anal canal, lung, liver, spleen, bone, and brain.
- Disseminated skin disease is a rare presentation.
- Endemic in tropical and subtropical areas, specifically South Asia (3).
- Diagnosis is via tissue biopsy demonstrating mature sporangia containing endospores.
- Treatment is by surgical resection with careful examination of mucous membranes.



Rhinosporidiosis: Summary

- Rhinosporidiosis typically presents as a chronic mucosal infection caused by ***Rhinosporidium seeberi***.
- Rhinosporidiosis should be considered in patients with painless conjunctival or sinonasal lesions clinically resembling pyogenic granuloma.
- Exposure risk factors include contact of damaged epithelium with contaminated stagnant water or inhalation of spore contaminated field dust.
- Complete surgical excision and examination of sinonasal mucous membranes are recommended to assess for additional lesions.



Rhinosporidiosis: References

1. Fredricks, DN et al. Rhinosporidium seeberi: A human pathogen from a novel group of aquatic protistan parasites”. *Emerg Infect Dis.* 2000; 6(3): 273-282.
2. Gopinathan, A et al. “Rhinosporidiosis of the tarsal conjunctiva”. *Indian J of Ophthal.* 2015; 63(5): pp. 462-3.
3. Pengos S et al. “Rhinosporidiosis in the Americas: A Systemic Review of Native Cases”. *Am J Trop Med Hyg.* 2021; 105(1): 171-175.
4. Arias, A et al. “Case report: Rhinosporidiosis literature review”. *Am. J. of Trop. Med Hyg.* 2021; 104(2): pp. 708-711.
5. Sood, N et al. “Ocular rhiosporidiosis: a case report from Delhi”. *J Infect Dev Ctries.* 2012; 6(11): 825-827.



Pigmented Conjunctival Lesions

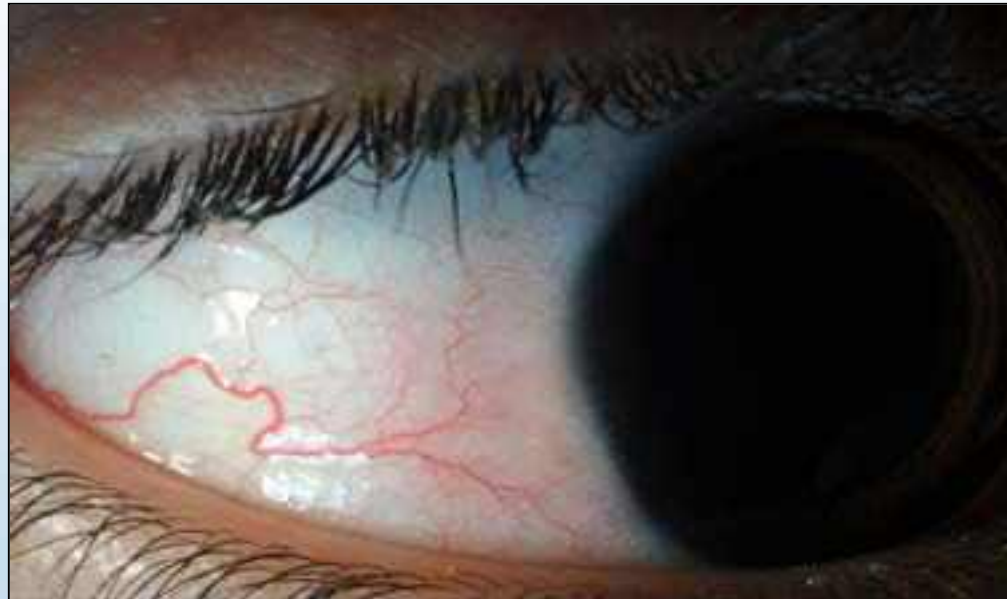
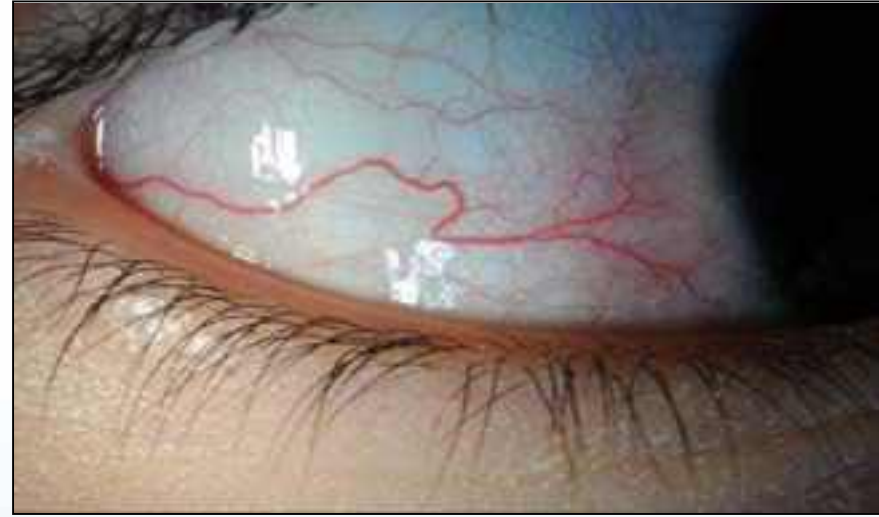
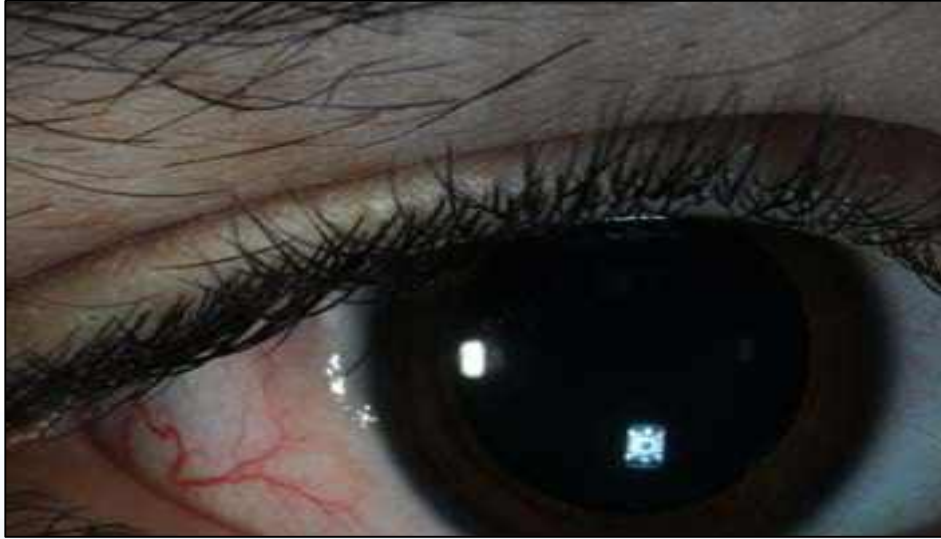


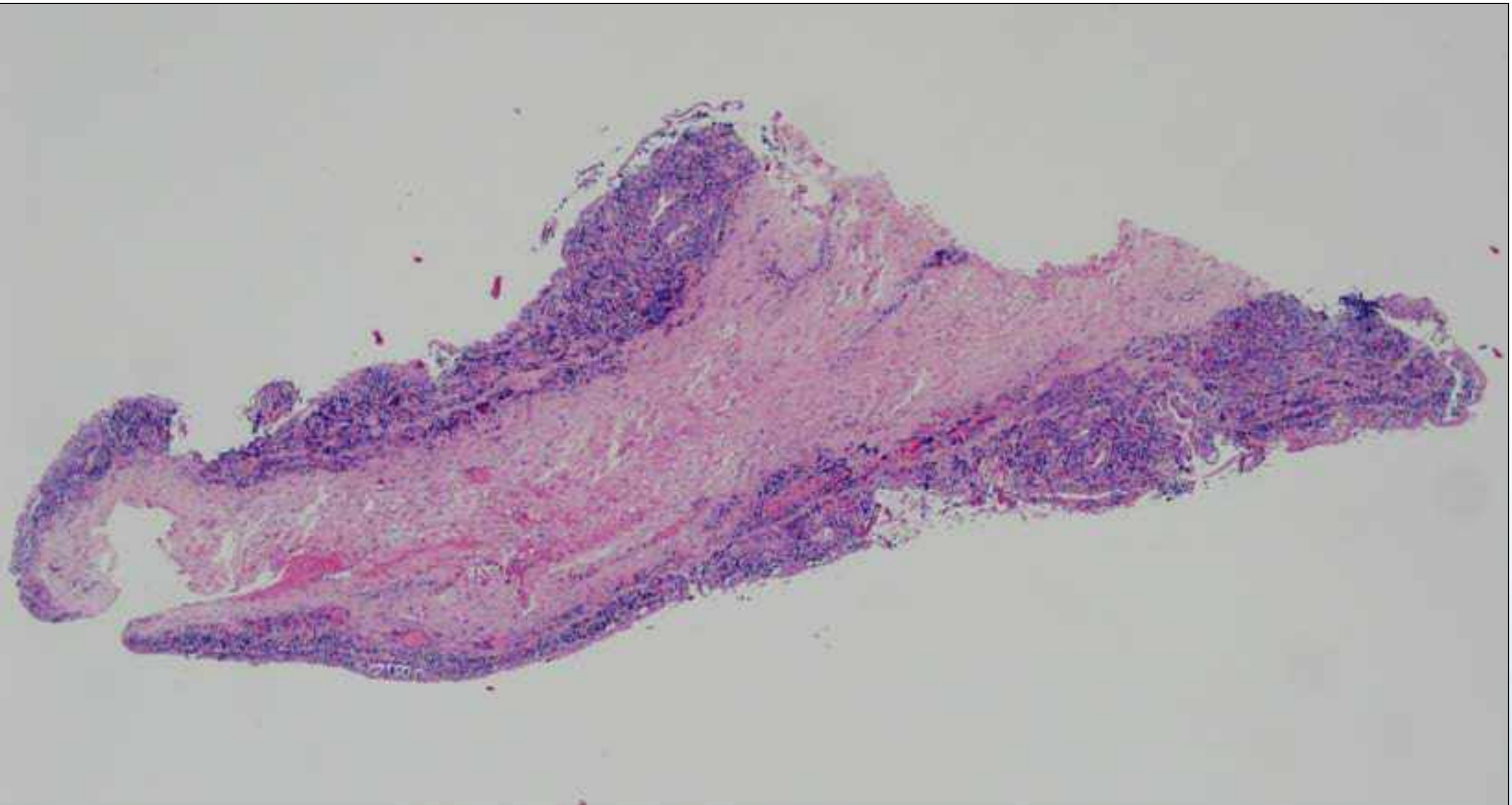
Temporal conjunctival lesion

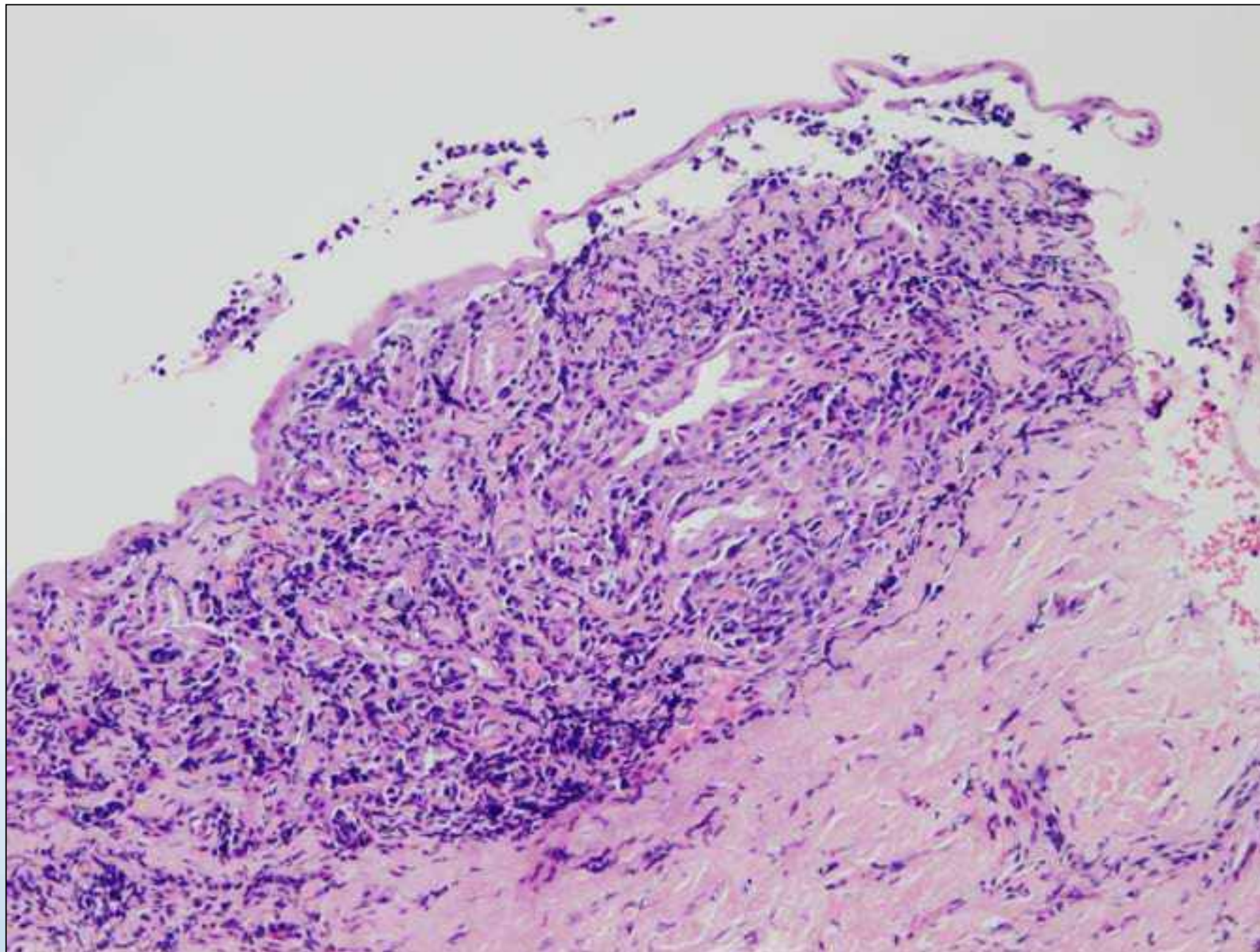
- **10 yo Caucasian male presents with 2 year history of temporal conjunctival lesion OD.**
- **Associated with occasional redness, blurry vision and eye irritation OD.**
- **Pt. evaluated by multiple eye doctors who diagnosed pt. with pinguecula, allergic conjunctivitis and a “dry spot.”**
- **Pt. prescribed Ciprofloxacin, Pataday, Alrex, PredForte and Lotemax in alternating concentrations and schedules.**
 - **Drops make the redness and irritation go away, but symptoms resume when drops are stopped.**

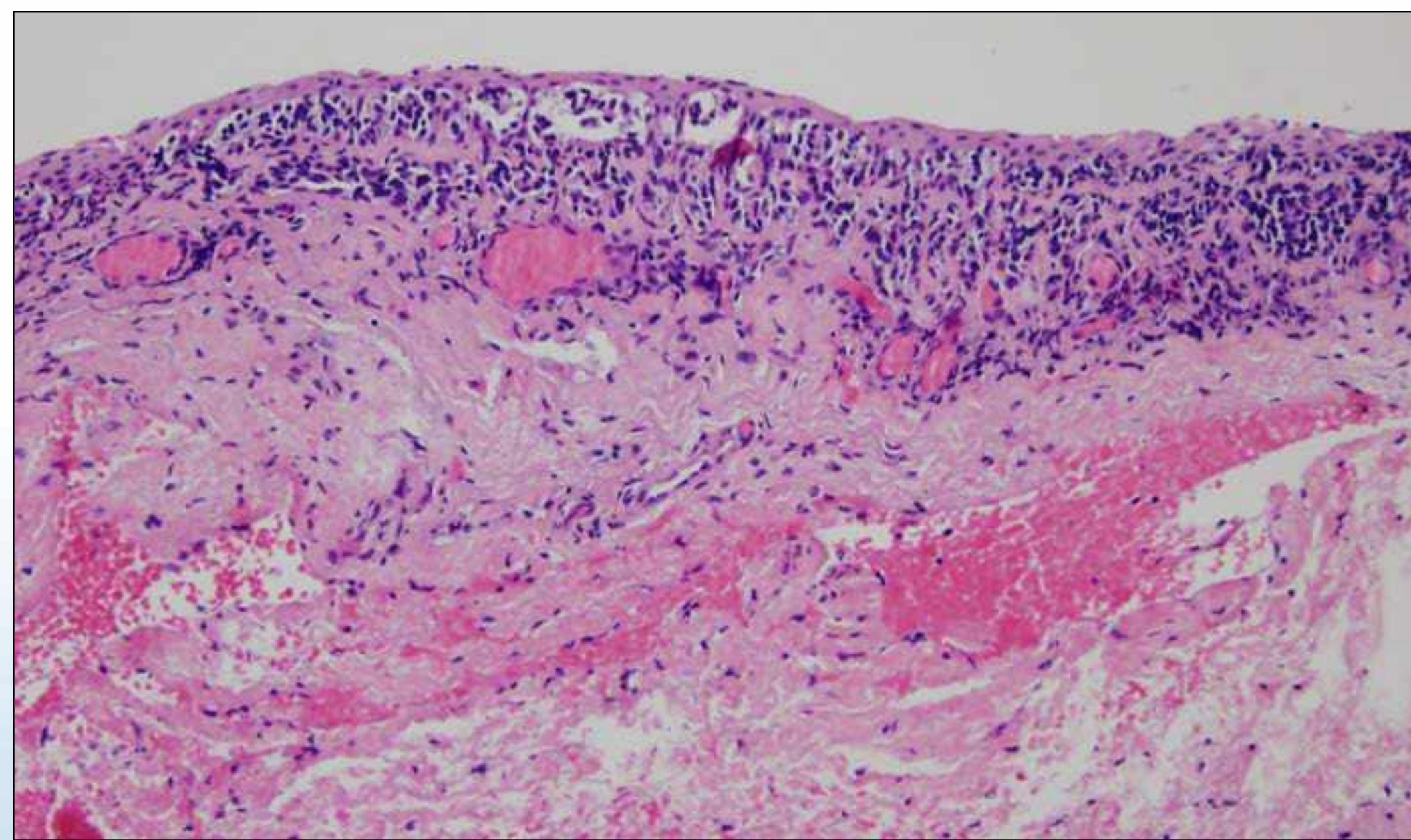


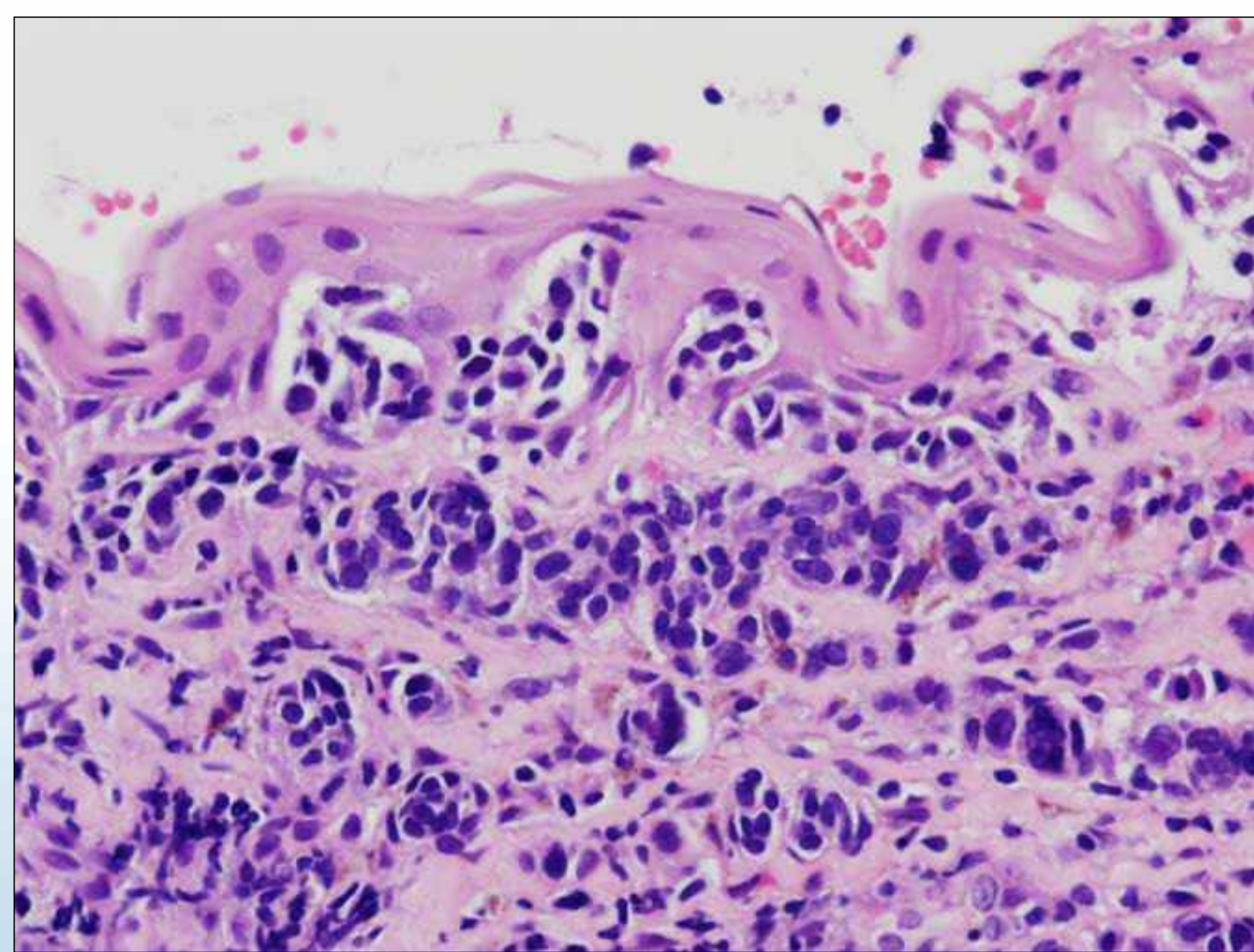
Slit Lamp Photos











Conjunctival Nevus

- Typically present at birth, cystic and located near limbus.
- Mostly are brown, but rarely red.
- Can become enlarged and affected by hormonal changes such as puberty.
- Benign



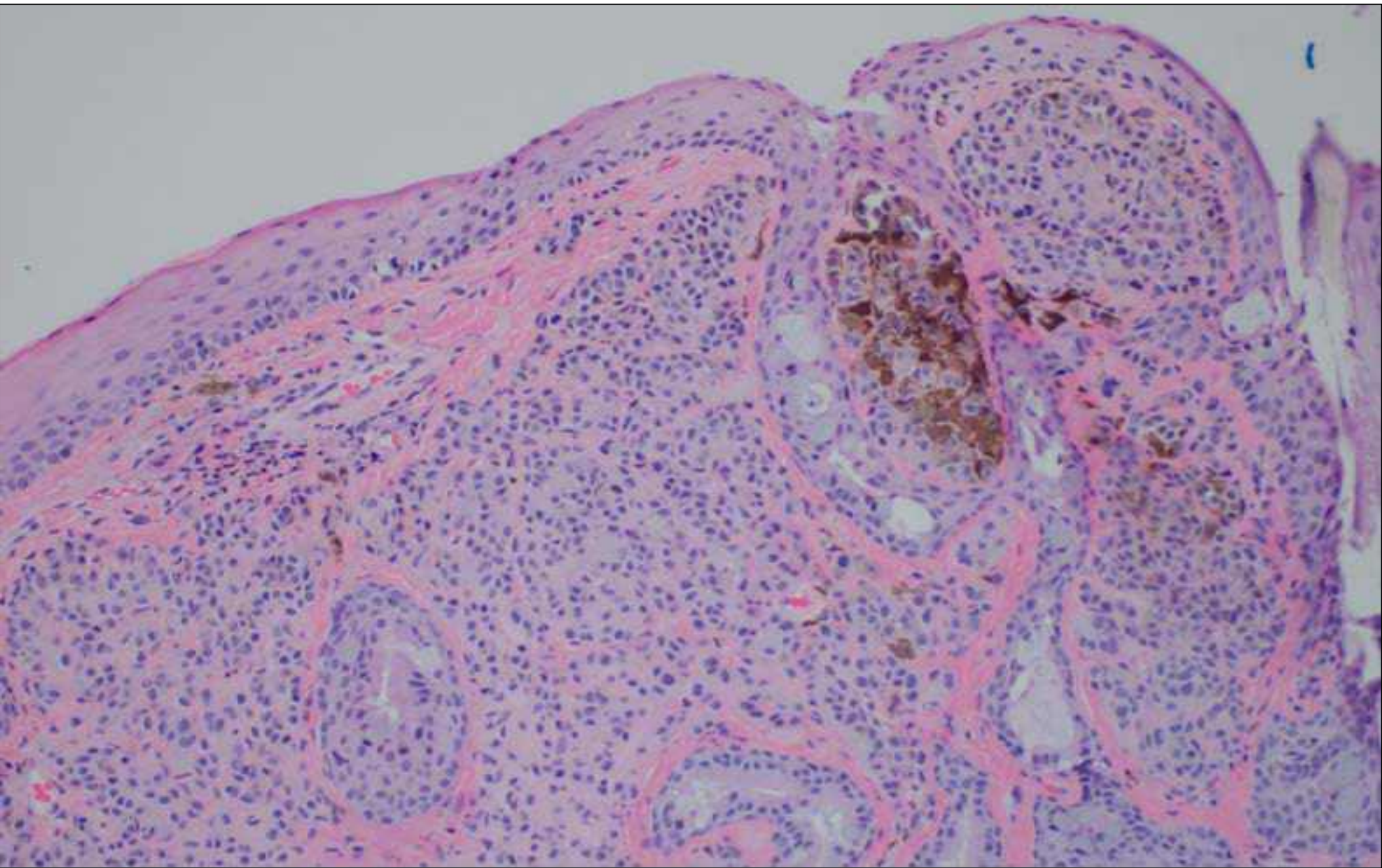
Conjunctival Nevi

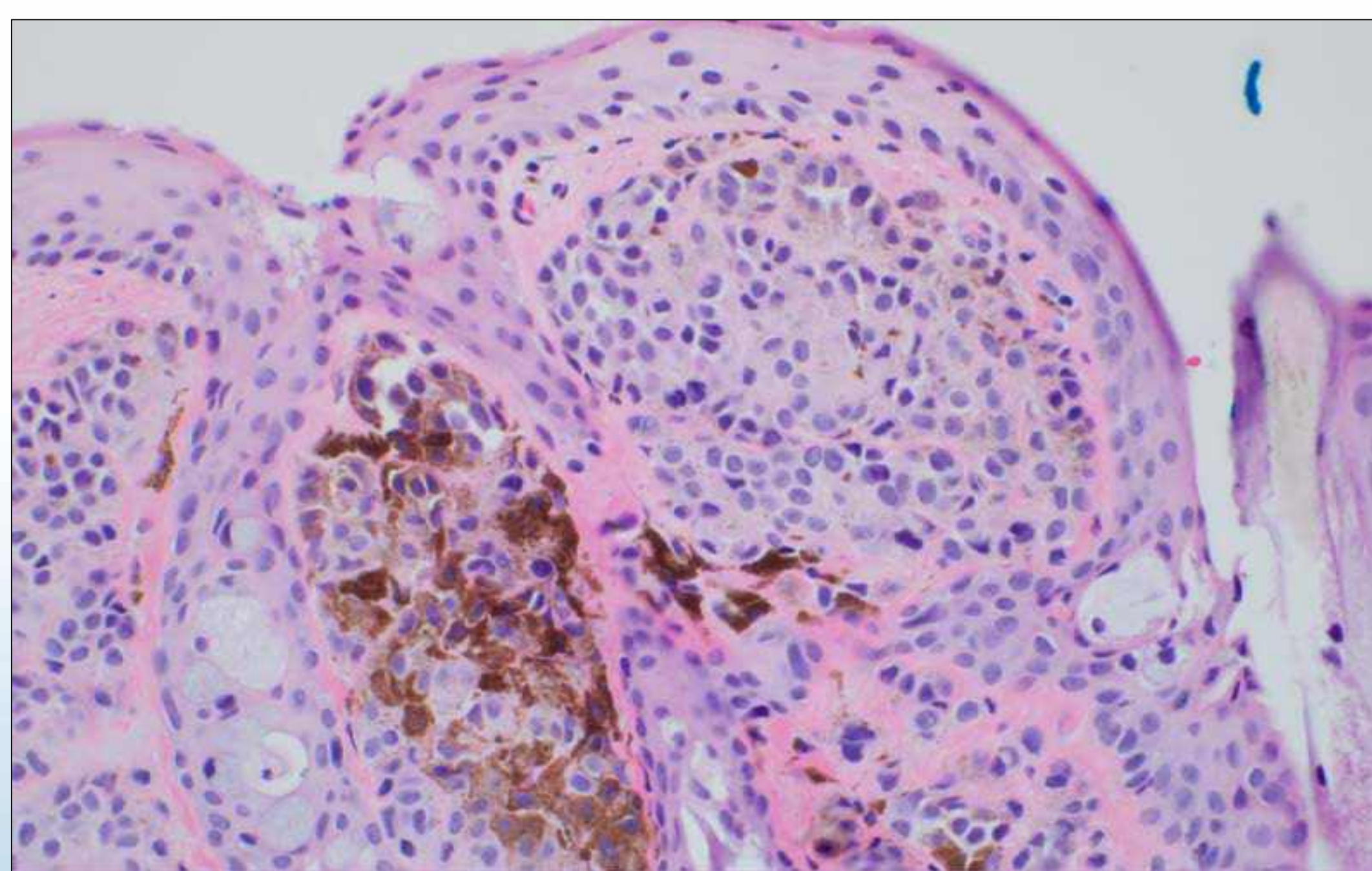
- Benign hamartomatous tumors
- Modified melanocytes = nevus cells (neural crest) Often detected towards end of 2nd decade
- Growth and increased pigmentation caused by elevated hormones in puberty and pregnancy
- Most common conjunctival tumor in children
- Fewer than 1% result in melanoma
- Classification based on nevus cell localization (junctional, compound, subepithelial)

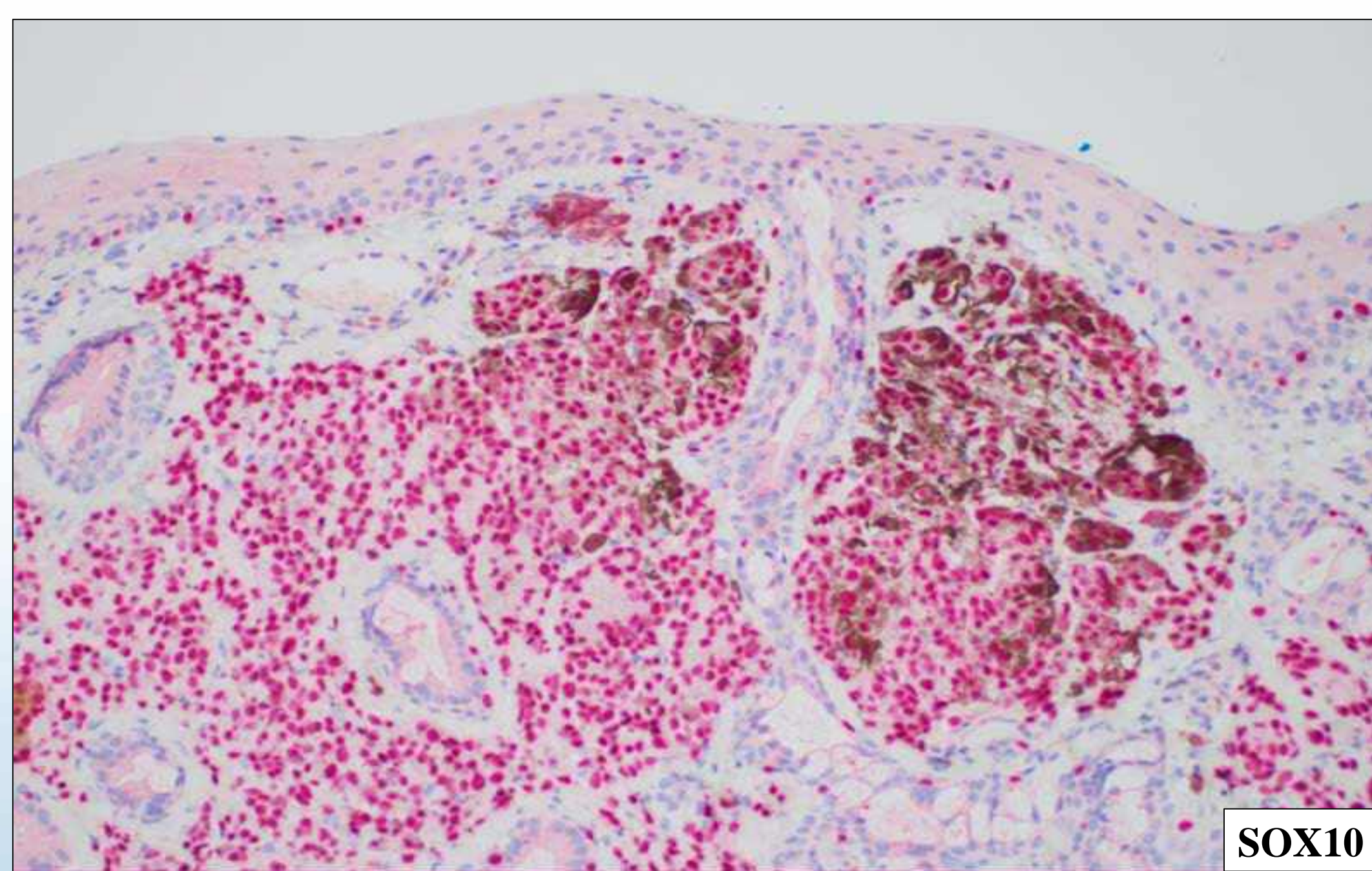


Compound cystic melanocytic nevus (Conjunctiva)



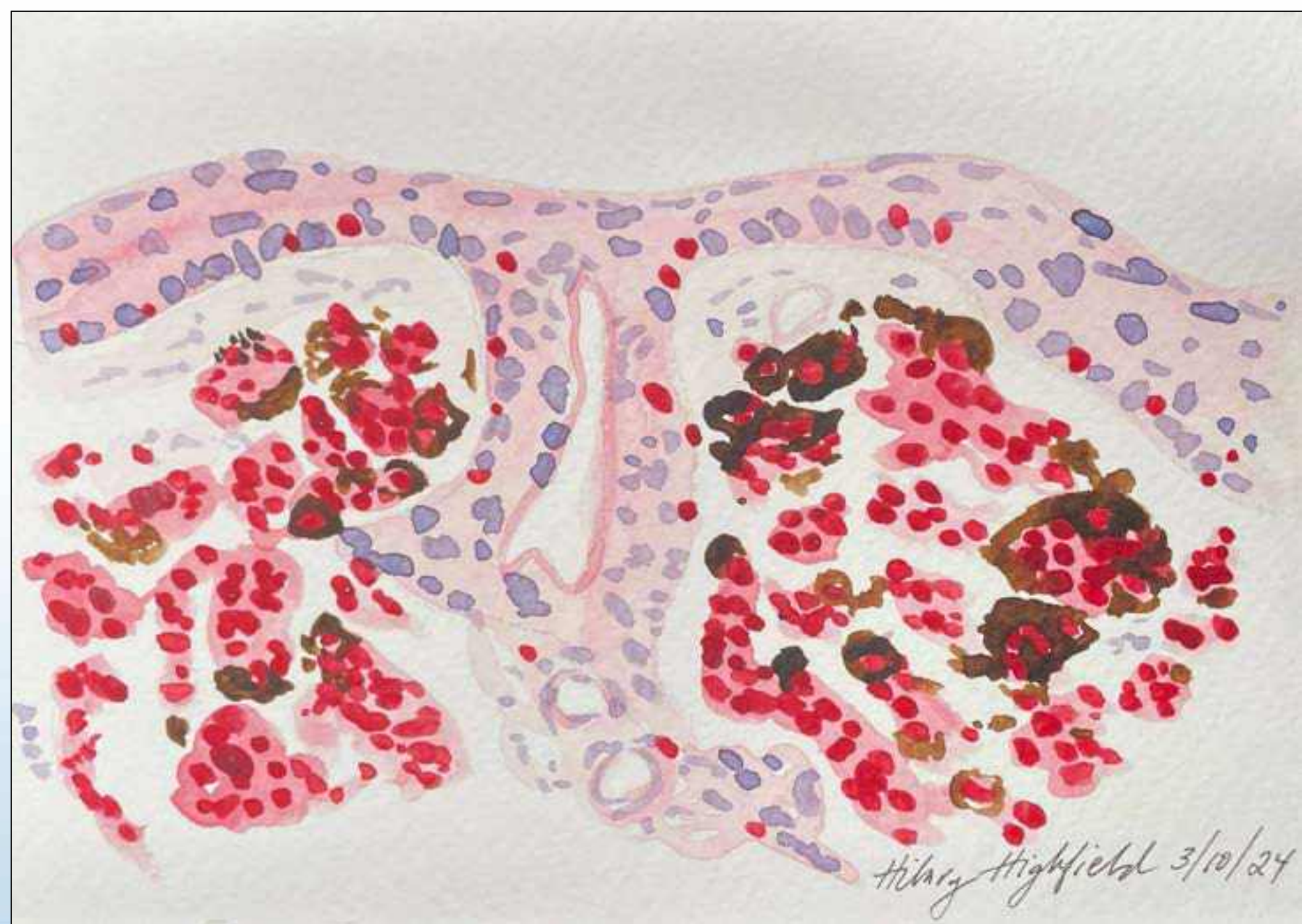




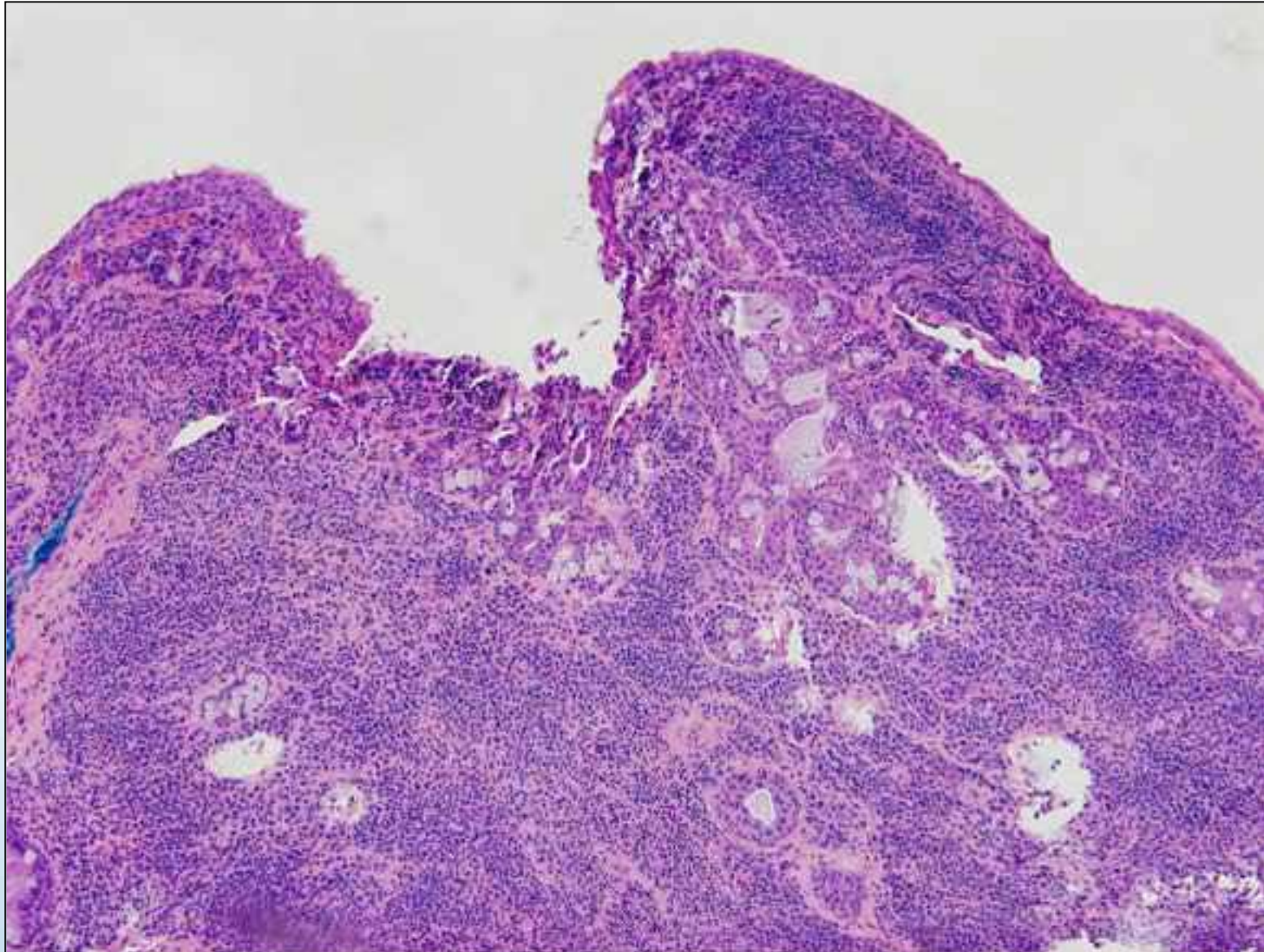


SOX10 AANP

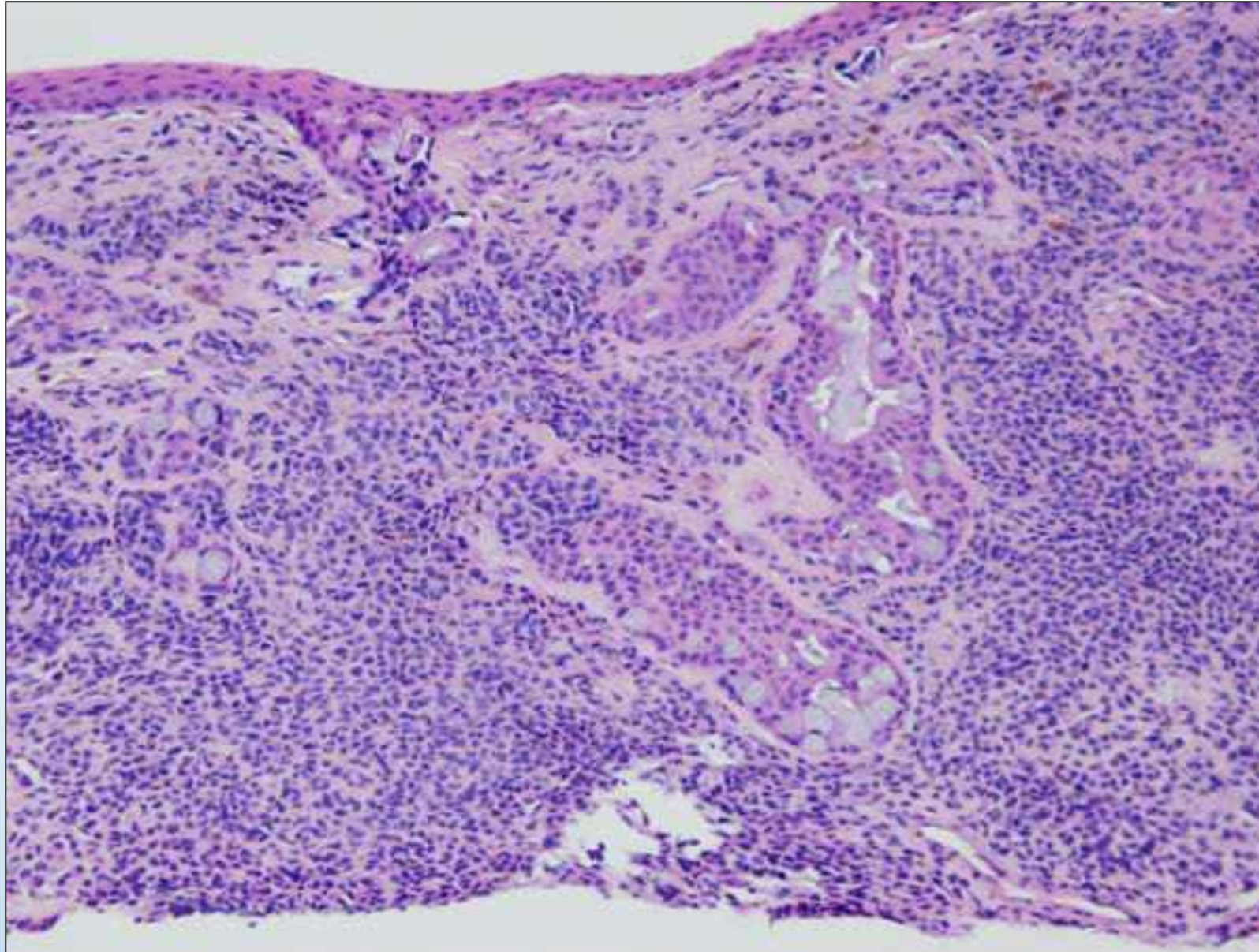




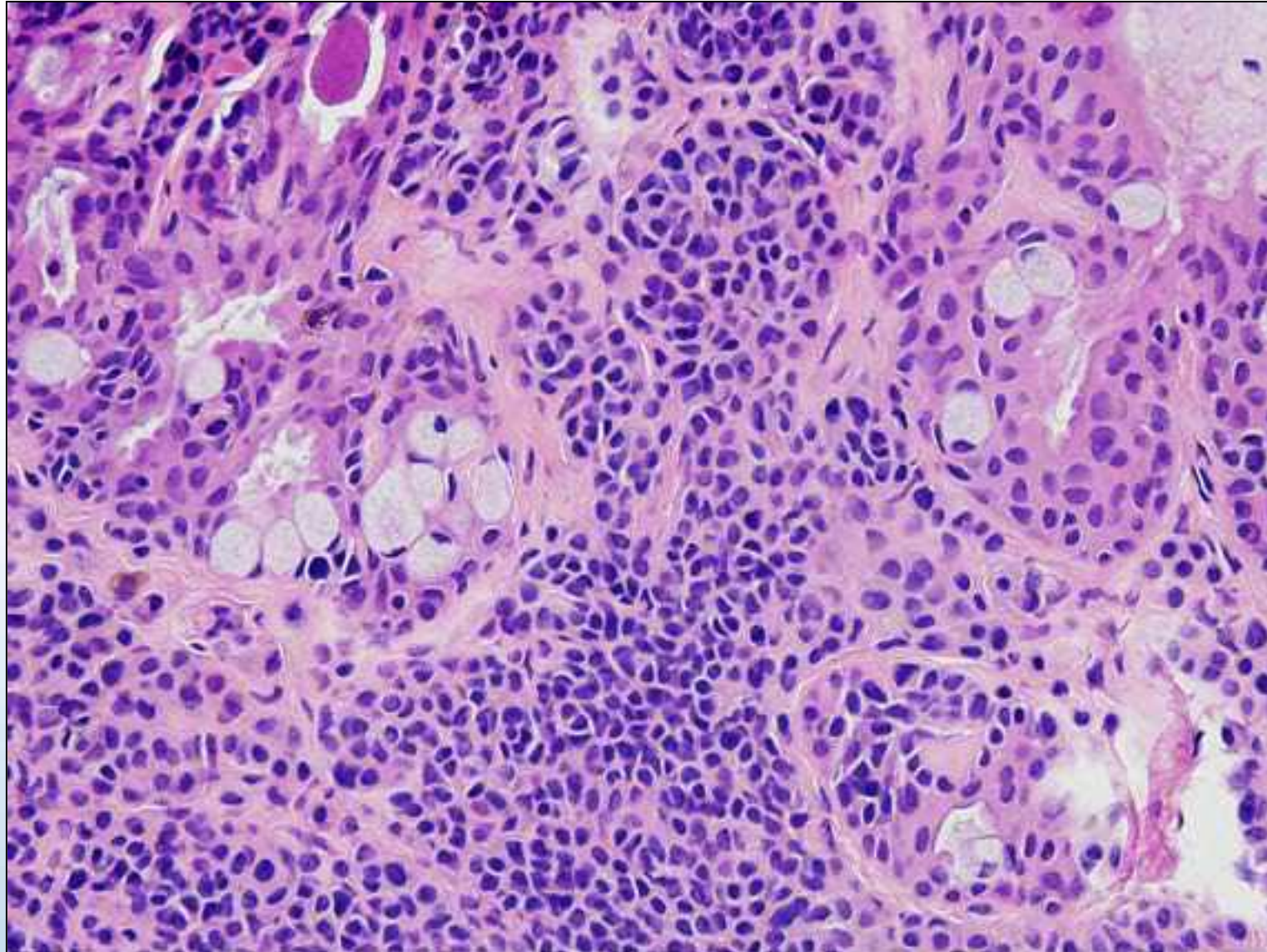
Compound nevus

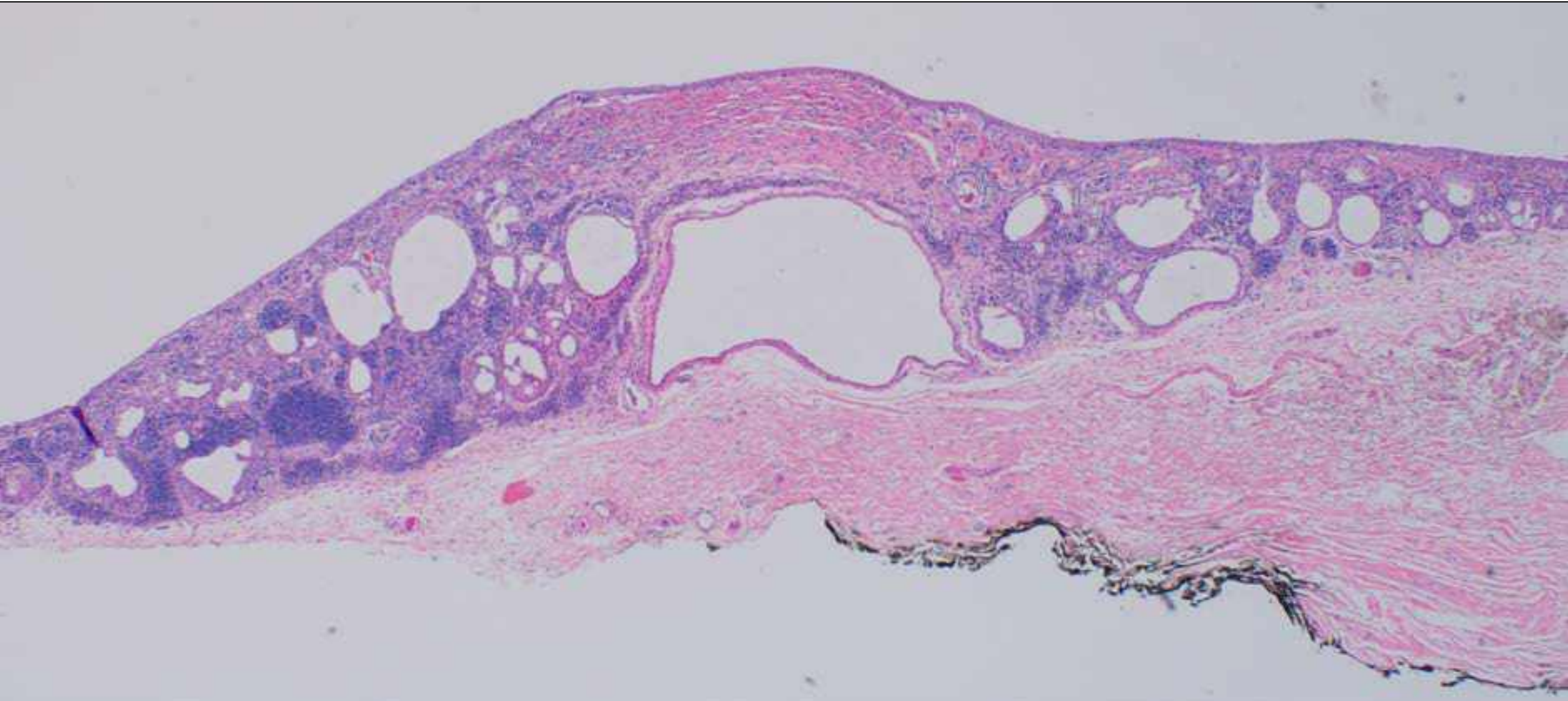


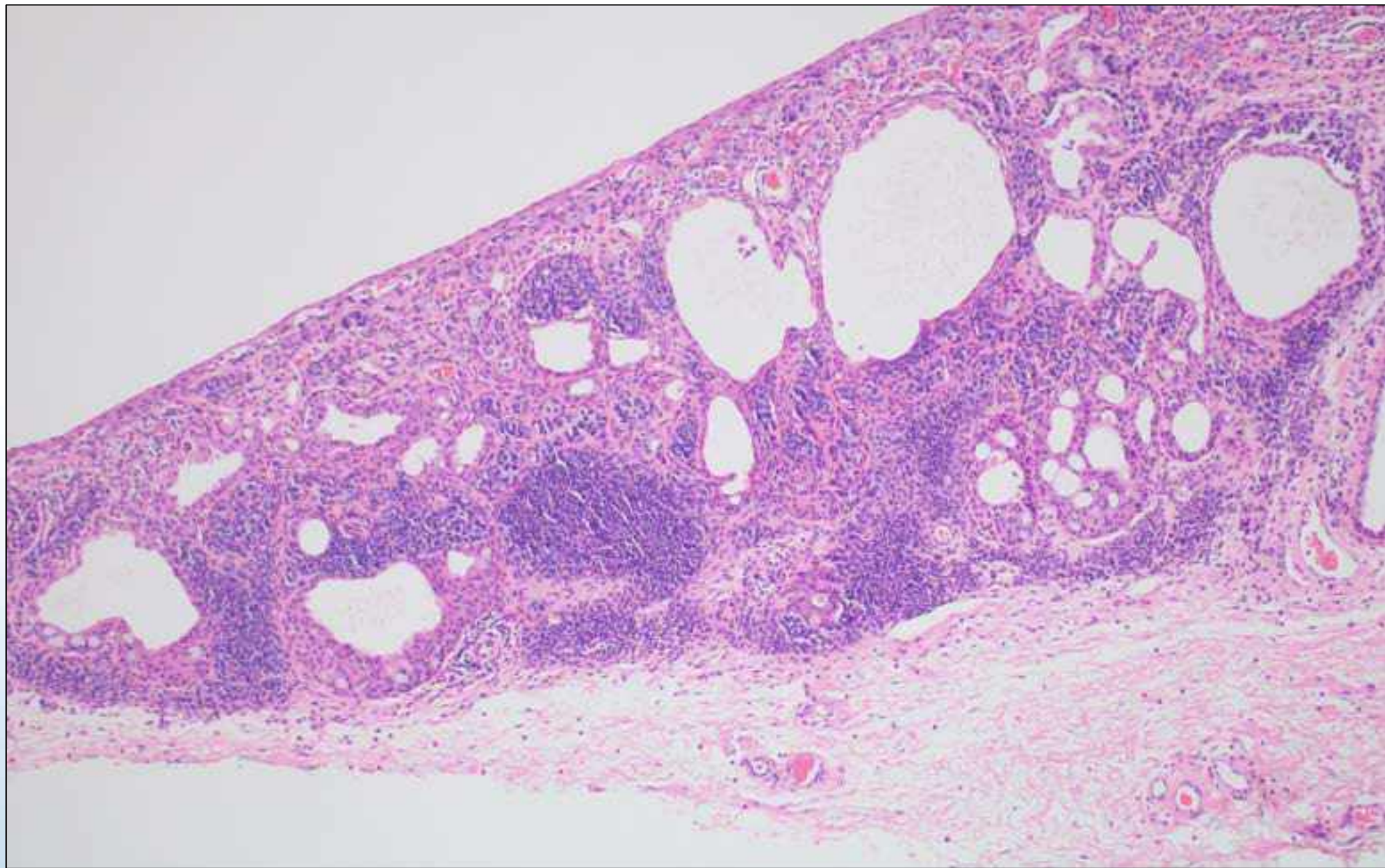
Compound nevus

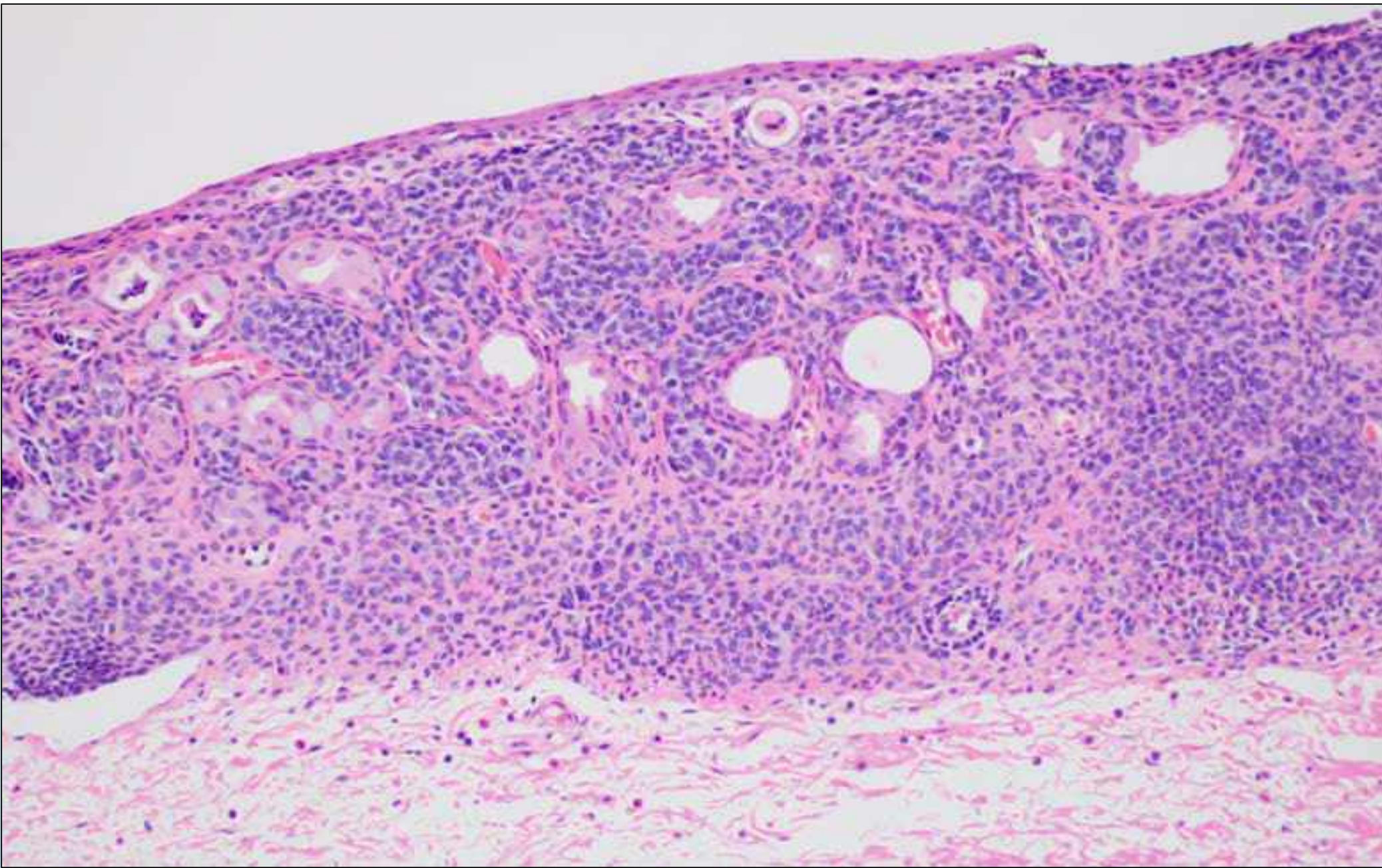


Compound nevus











SOX10



Compound nevus (conjunctiva)

- Benign melanocytic nevi
- Composed of modified melanocytes: nevus cells (derived from neural crest)
- Most common nevus type
- Median age 21-23 years
- Nevus cells within the epithelium and substantia propria
- Maturation towards the deeper layers
- Solid and cystic epithelial rests common



Conjunctival pigmented lesion

- HPI: 44 YOF
- Referred for evaluation of conjunctival pigmented lesion OS.
- Present since birth and stable until growth over past 5-6 weeks.
- No pain, bleeding, or injection.

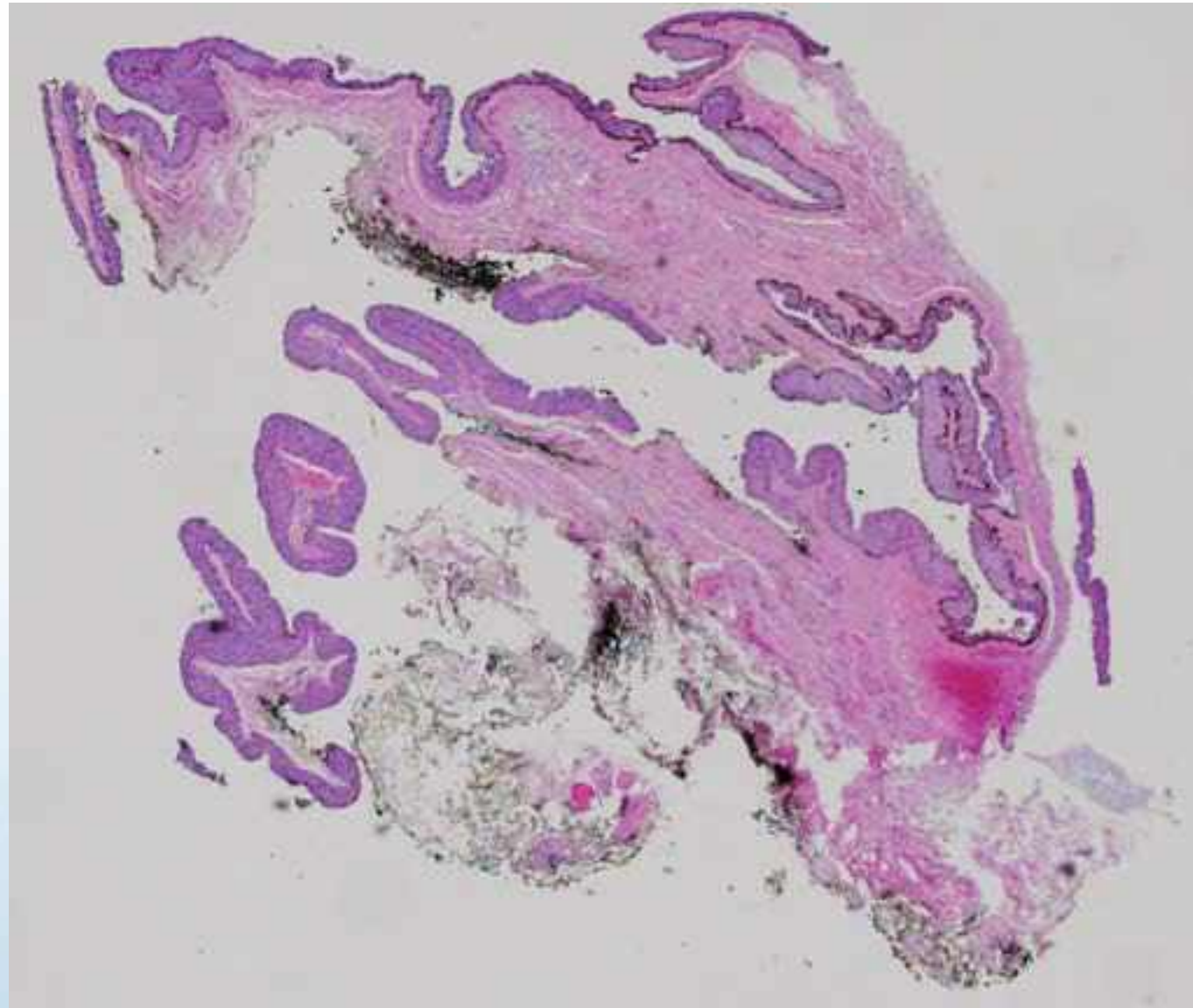




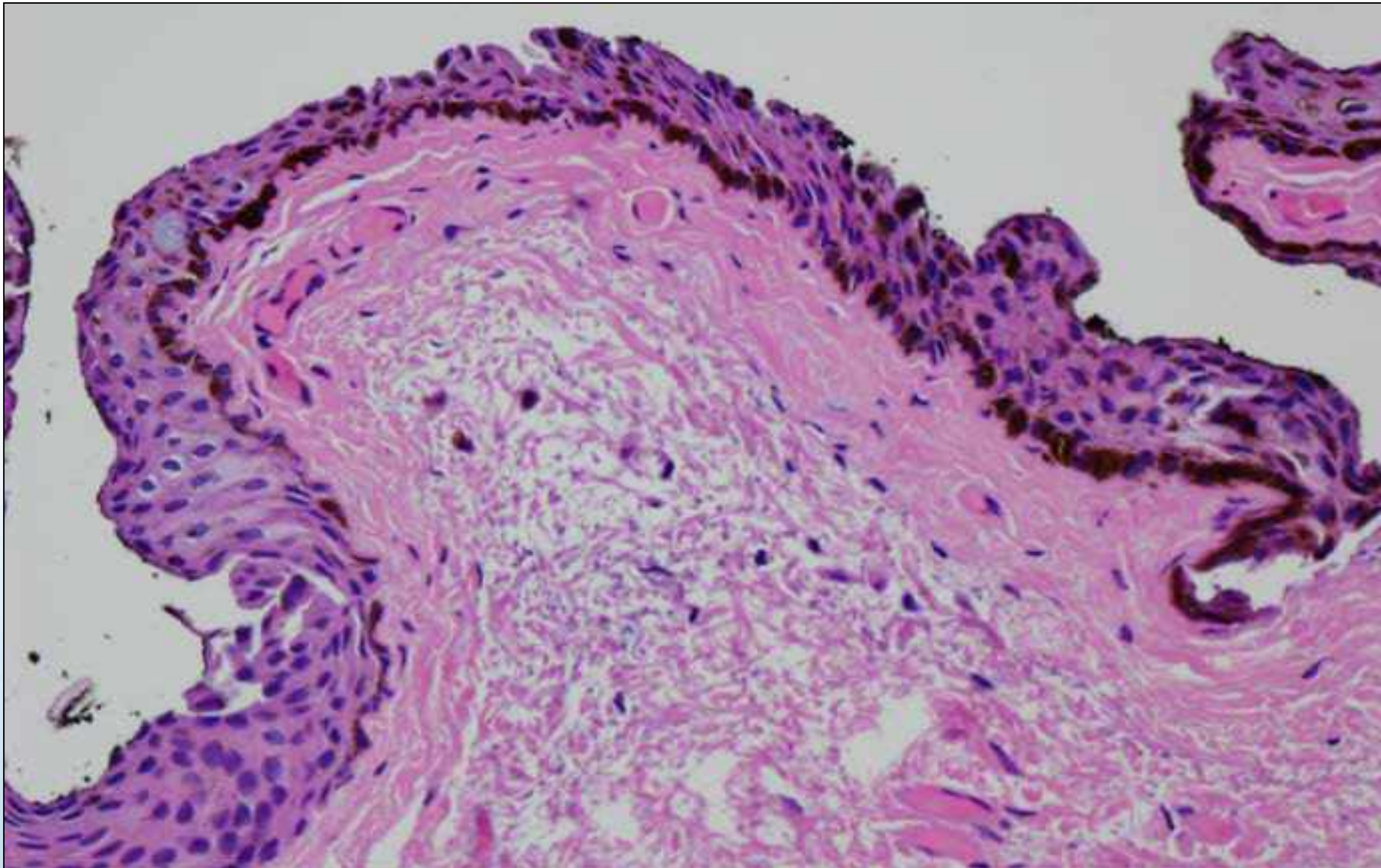




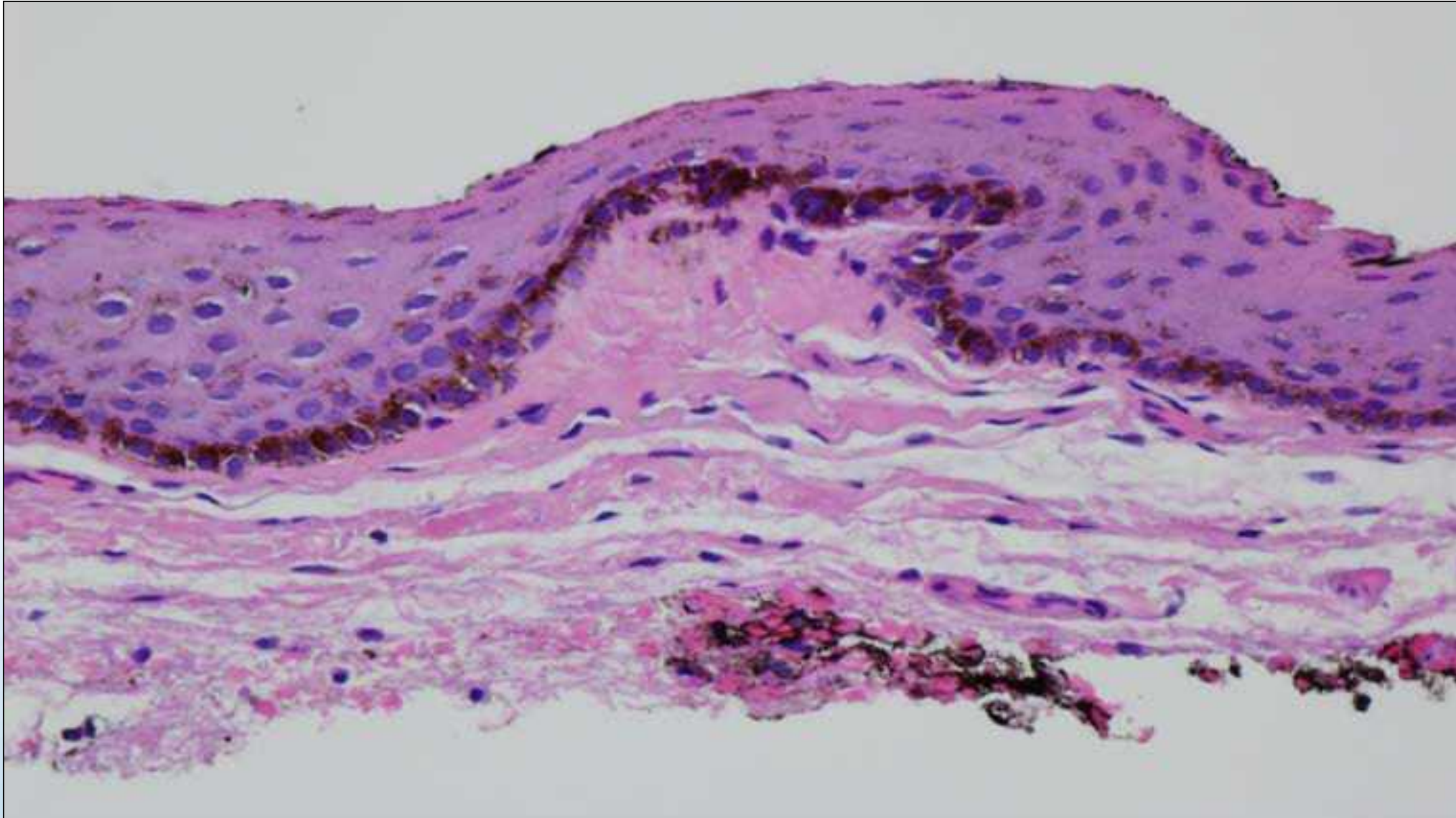
Conjunctival pigmented lesion



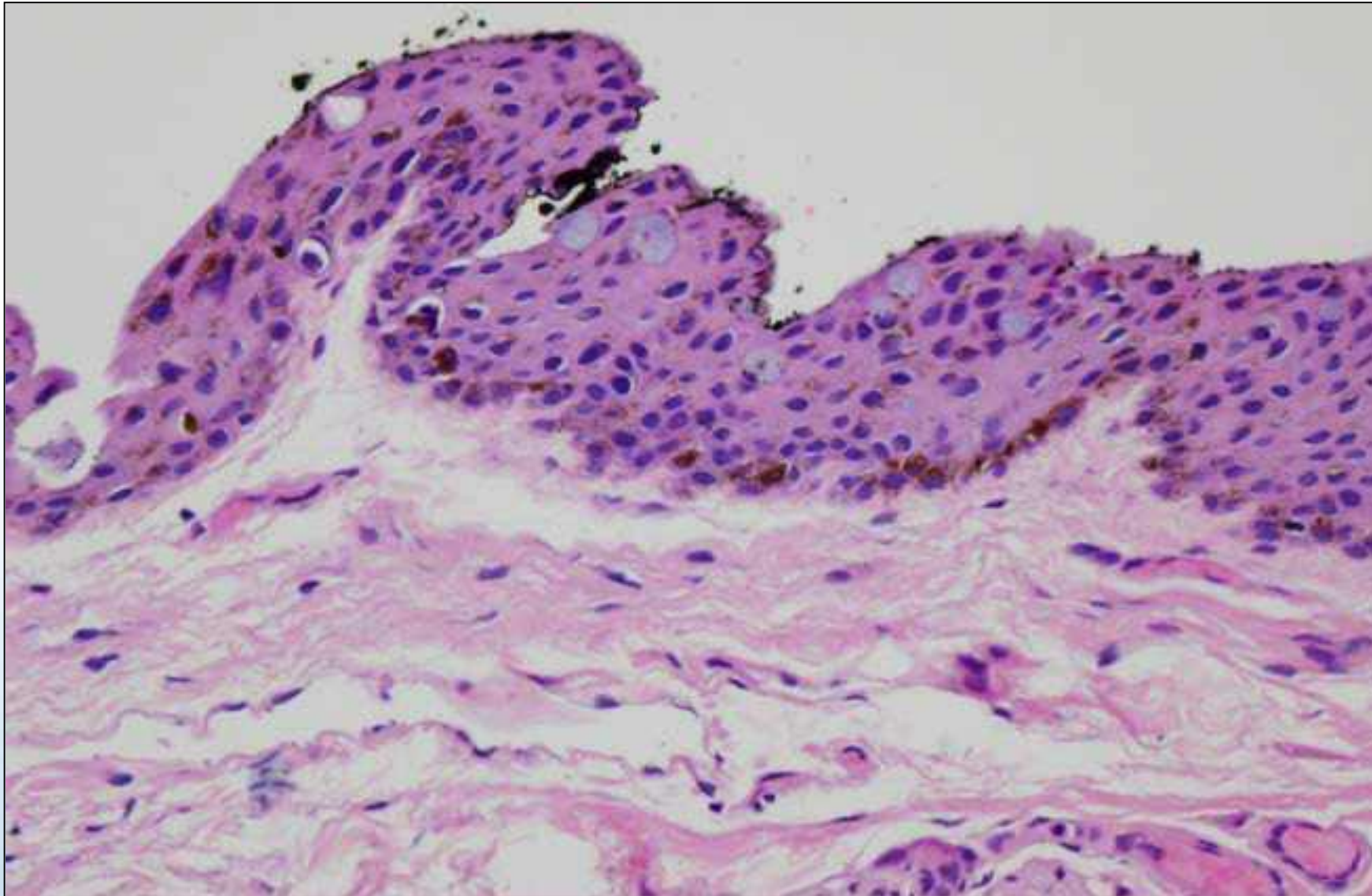
Conjunctival pigmented lesion



Conjunctival pigmented lesion



Conjunctival pigmented lesion



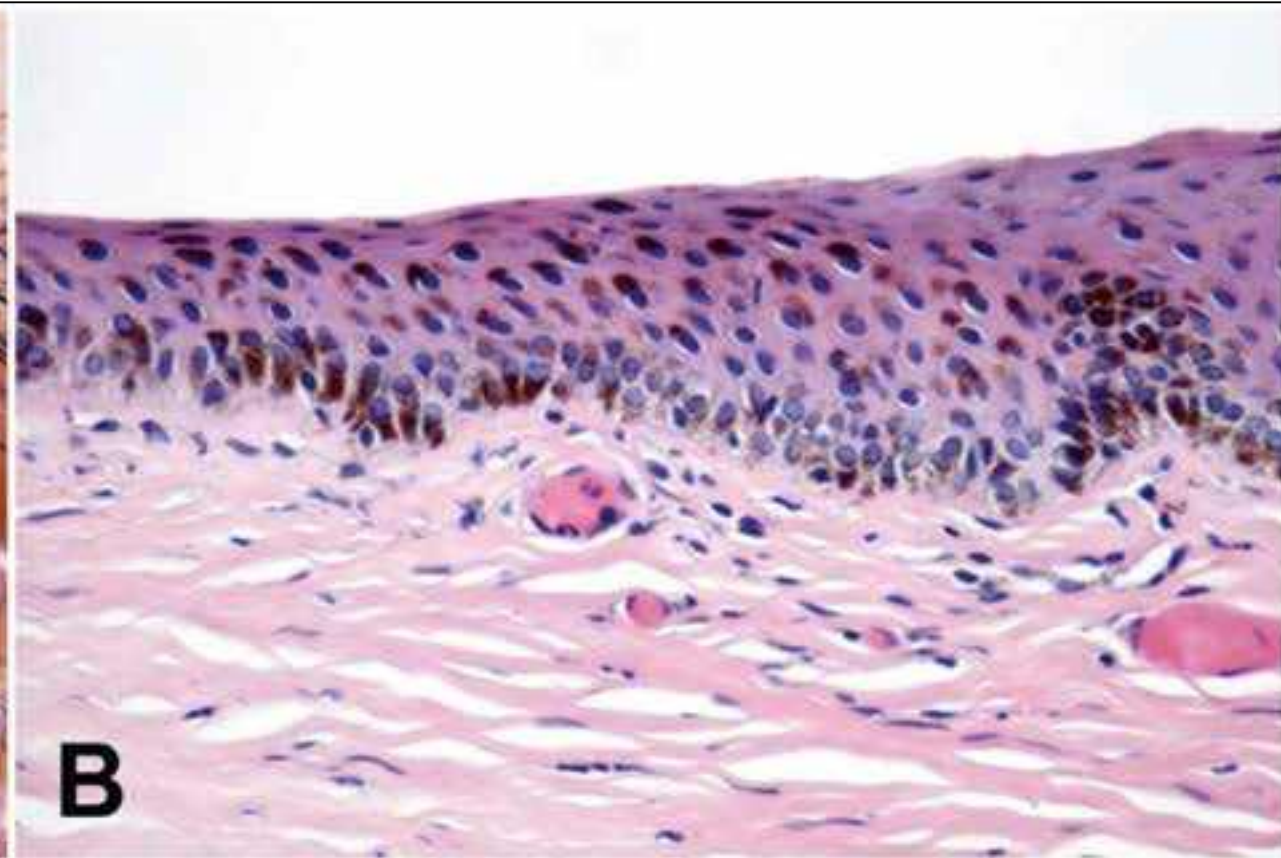
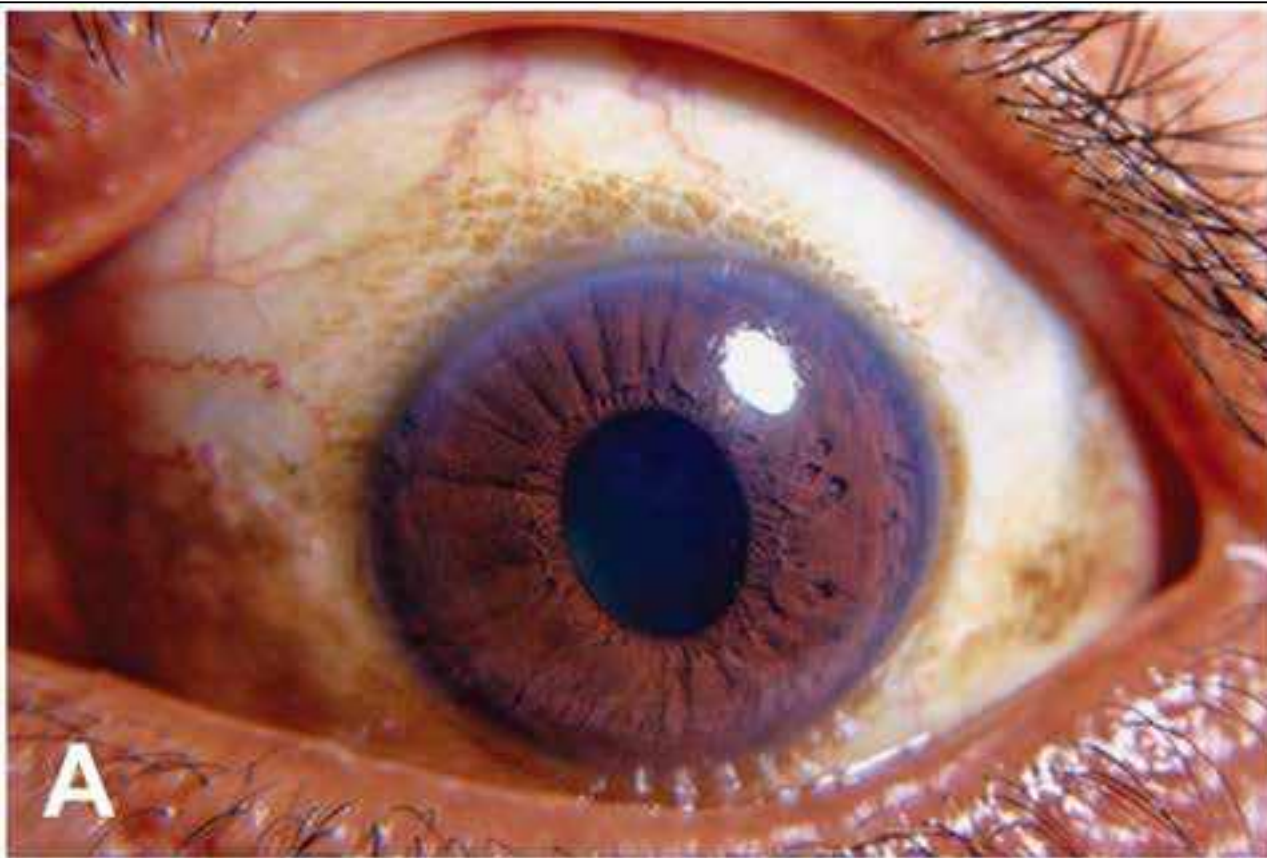
Secondary (Pigment-associated) Melanosis

Conjunctival Skin-Tone Associated Melanosis (C-STAM)

- **Constitutional melanosis, complexion-associated conjunctival pigmentation, benign epithelial melanosis**
- **92.5% Blacks, 36% Asians, 28% Hispanics, 4.9% Caucasians**
- **Bilateral pigmentation, denser at junction between peripheral cornea and limbus**



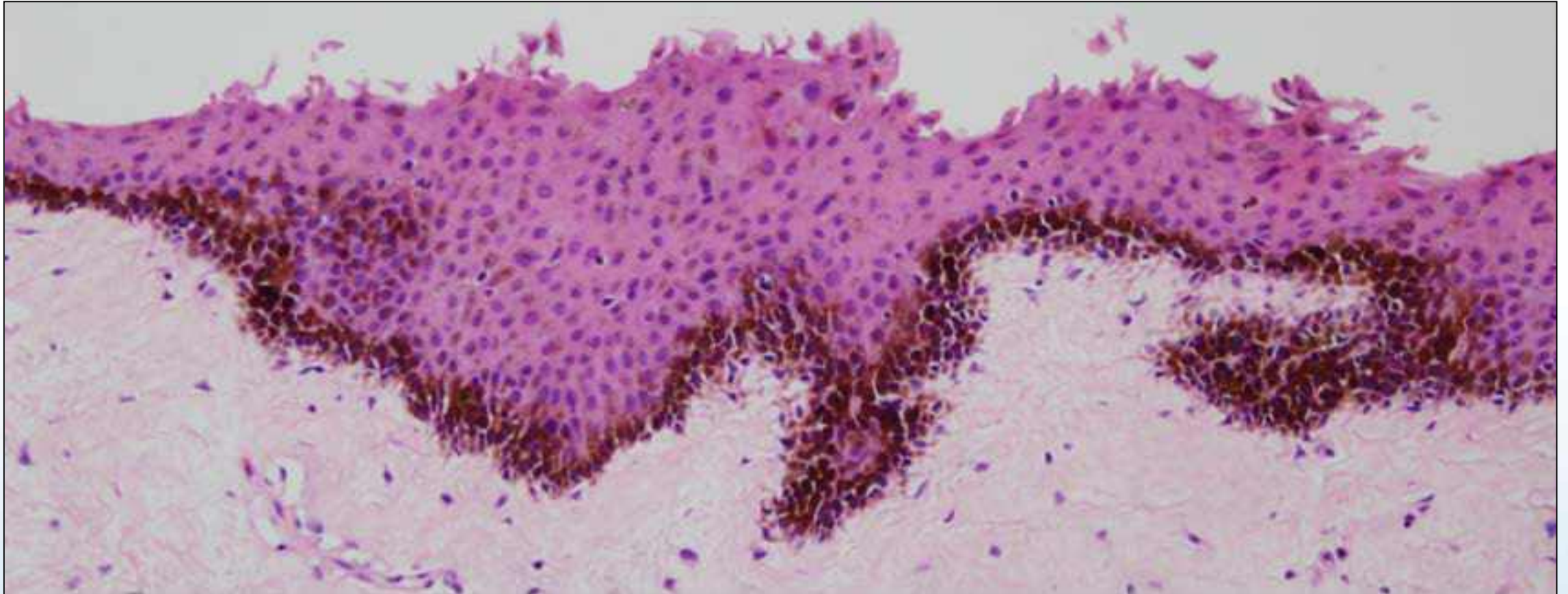
Constitutional melanosis



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Epithelial Basal Layer Pigmentation

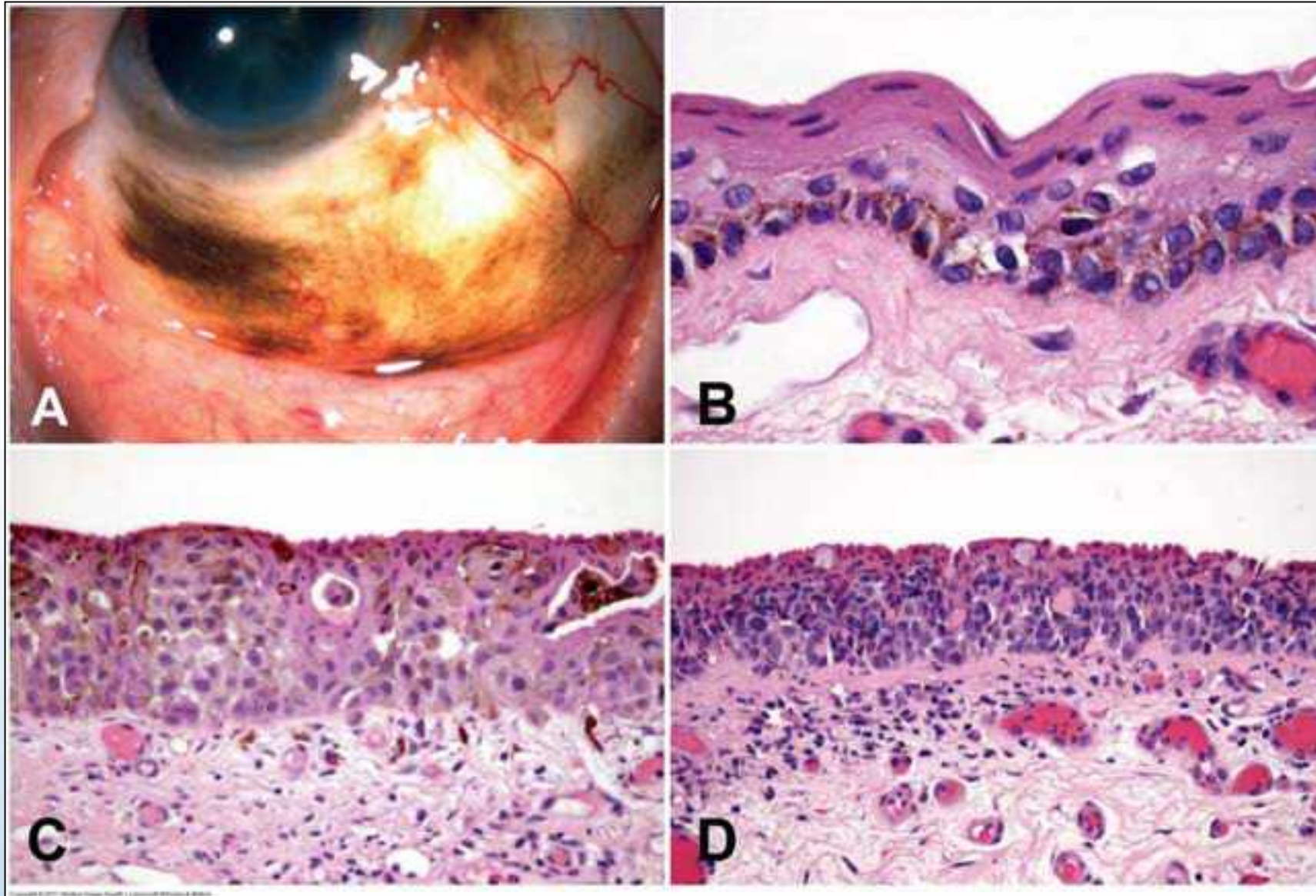


- **40s Asian female**
- **Conjunctival pigmentation; growing**
- **Flat brown mobile lesion**

Primary Acquired Melanosis



Primary Acquired Melanosis



Primary Acquired Melanosis (PAM)

- **Precursor of conjunctival melanoma**
- **75% of cases of malignant melanoma arise in cases of PAM with atypia**
- **Patchy acquired unilateral pigmentation older**
- **Most important risk factor for recurrence and progression – extent of pigmentation in clock hours**



Primary Acquired Melanosis (PAM)

- **PAM without atypia**
 - Pigment confined to conjunctival epithelial cells
 - No evidence of melanocytic hyperplasia
 - Indistinguishable from complexion melanosis/freckle
- **PAM with mild atypia**
 - Atypical melanocytes confined to epithelial basal layer
 - Single cell lentiginous pattern
 - Low risk for progression



Primary Acquired Melanosis (PAM)

- **PAM with severe atypia**
 - Melanocytes involve more superficial layers
 - Form pagetoid nests
 - Pagetoid involvement associated with 95% risk of progression

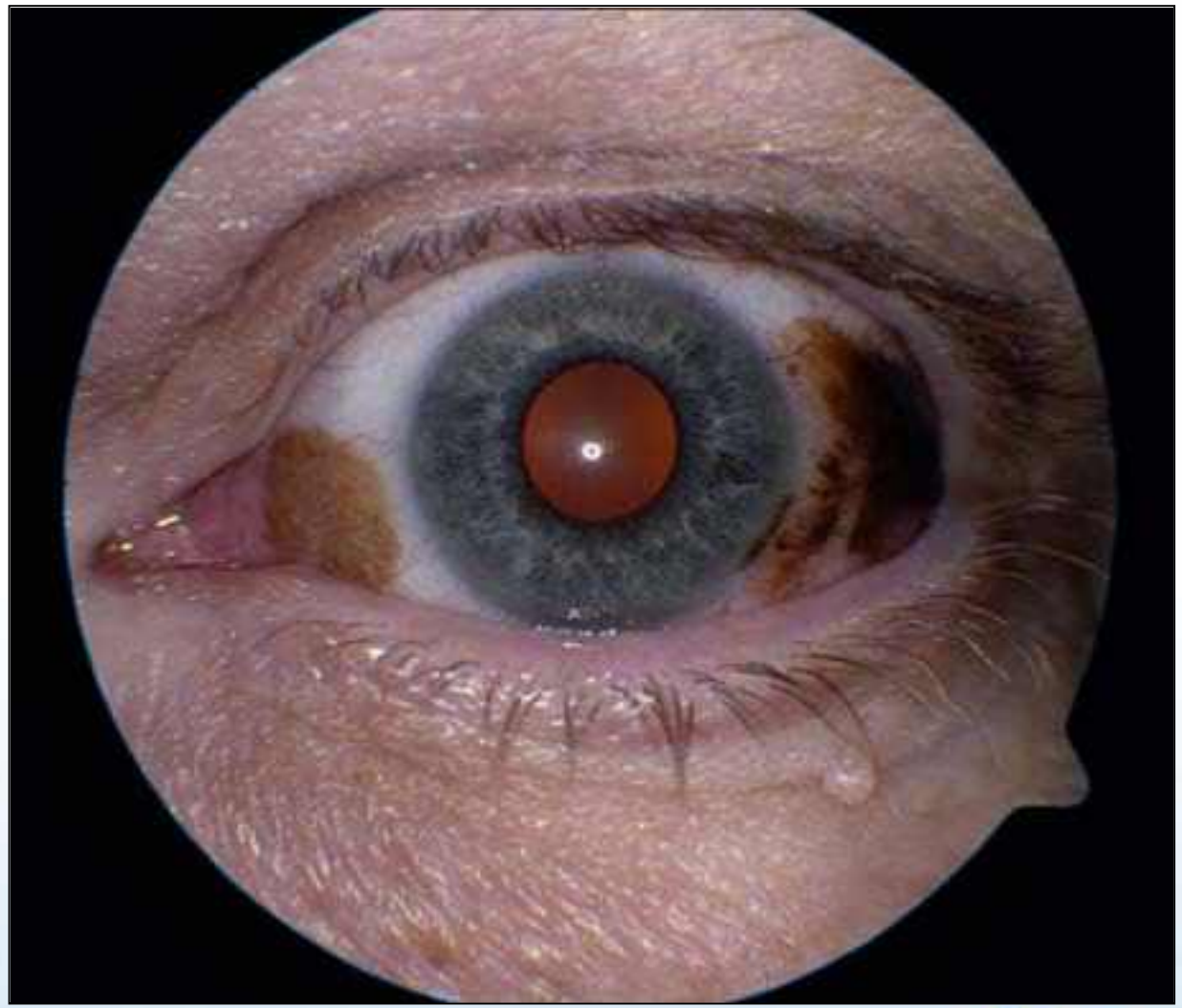
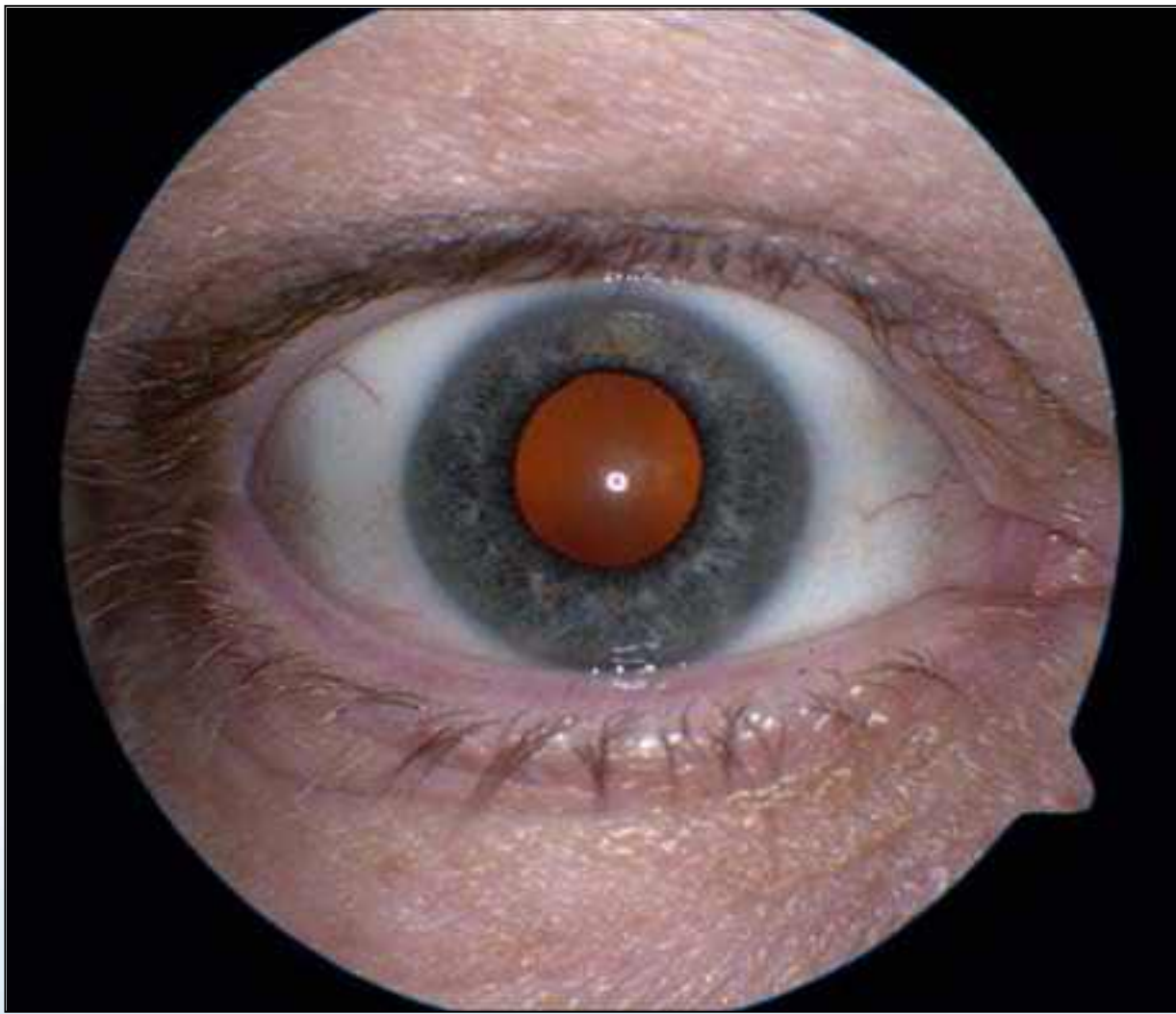
- **Invasive melanoma**
 - Extension through epithelial basement membrane
 - Vertical growth into substantia propria

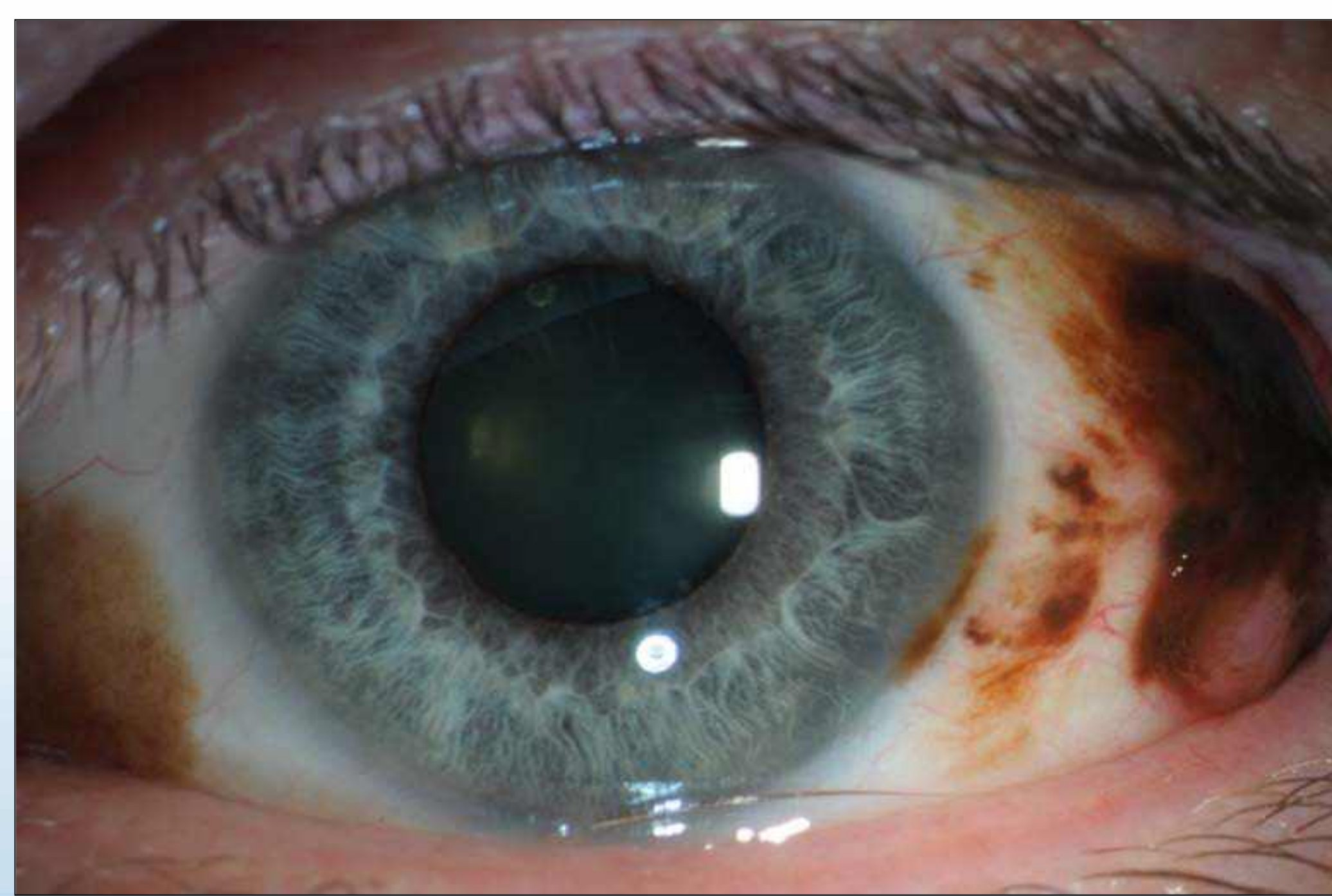


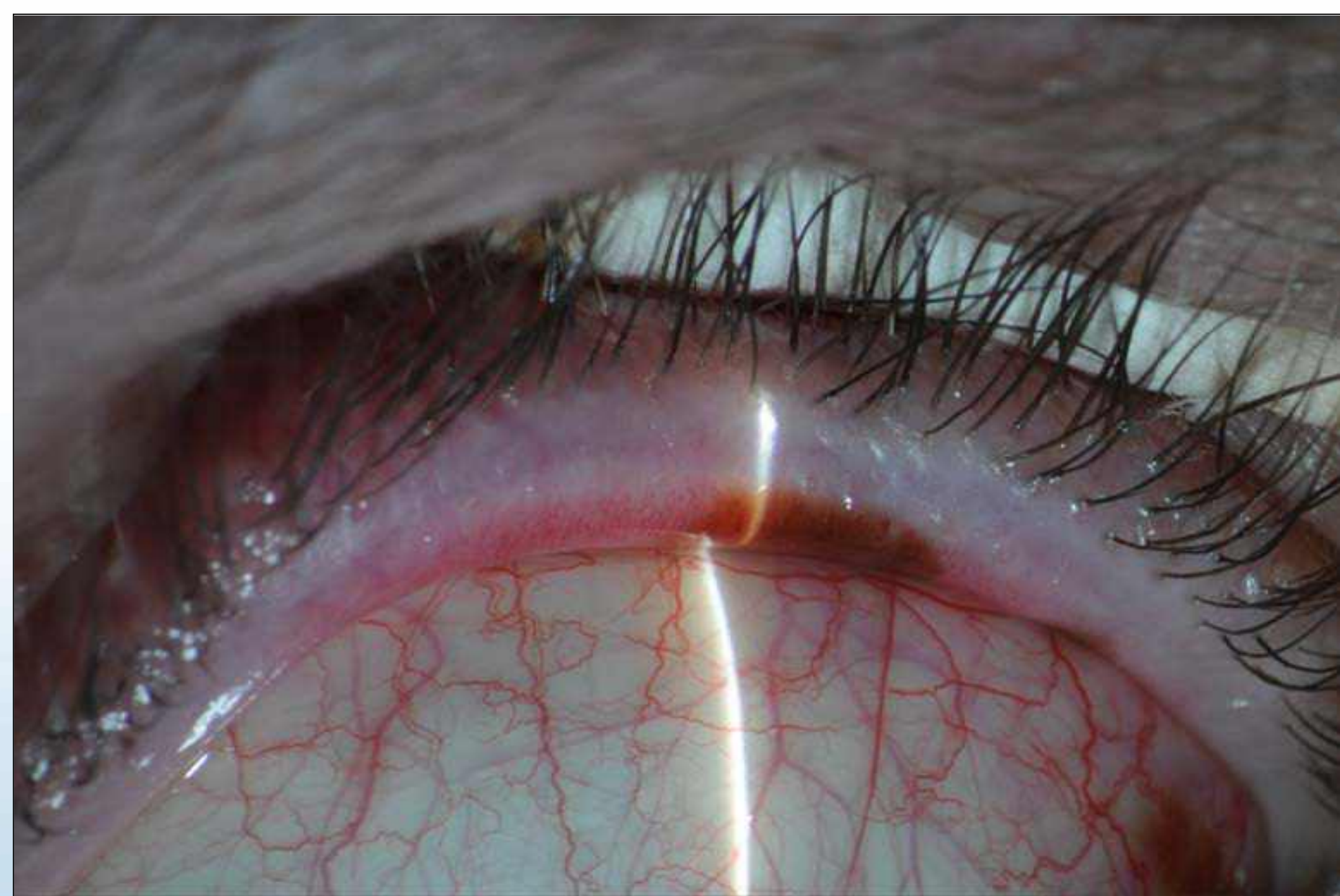
Pigmented spot on OS

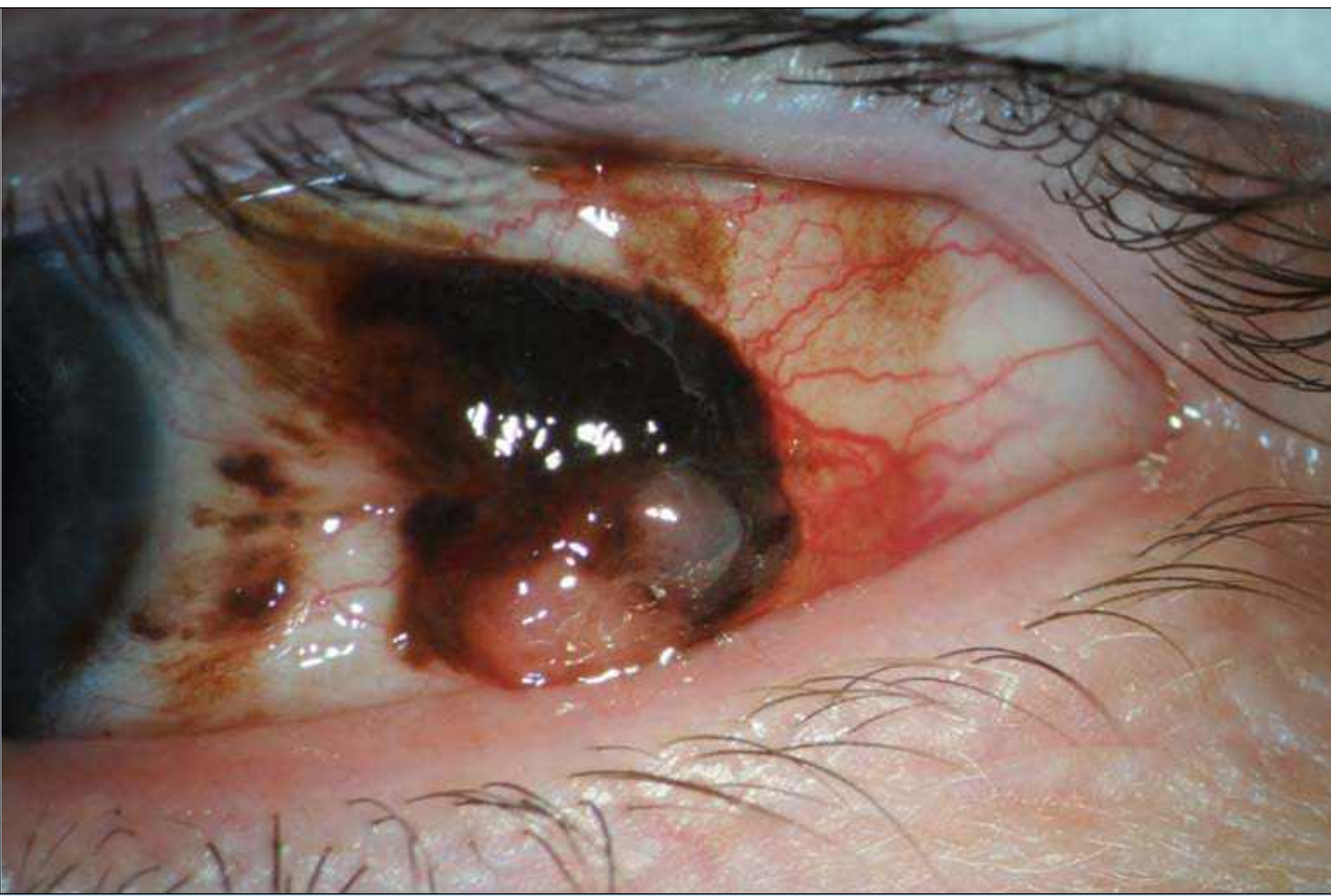
- 54yo M referred from local ophthalmologist
- CC: Pigmented spot on OS x 1 ½ years
- Growing larger in past several weeks
- + Sore, blurred VA, photophobia, tearing OS

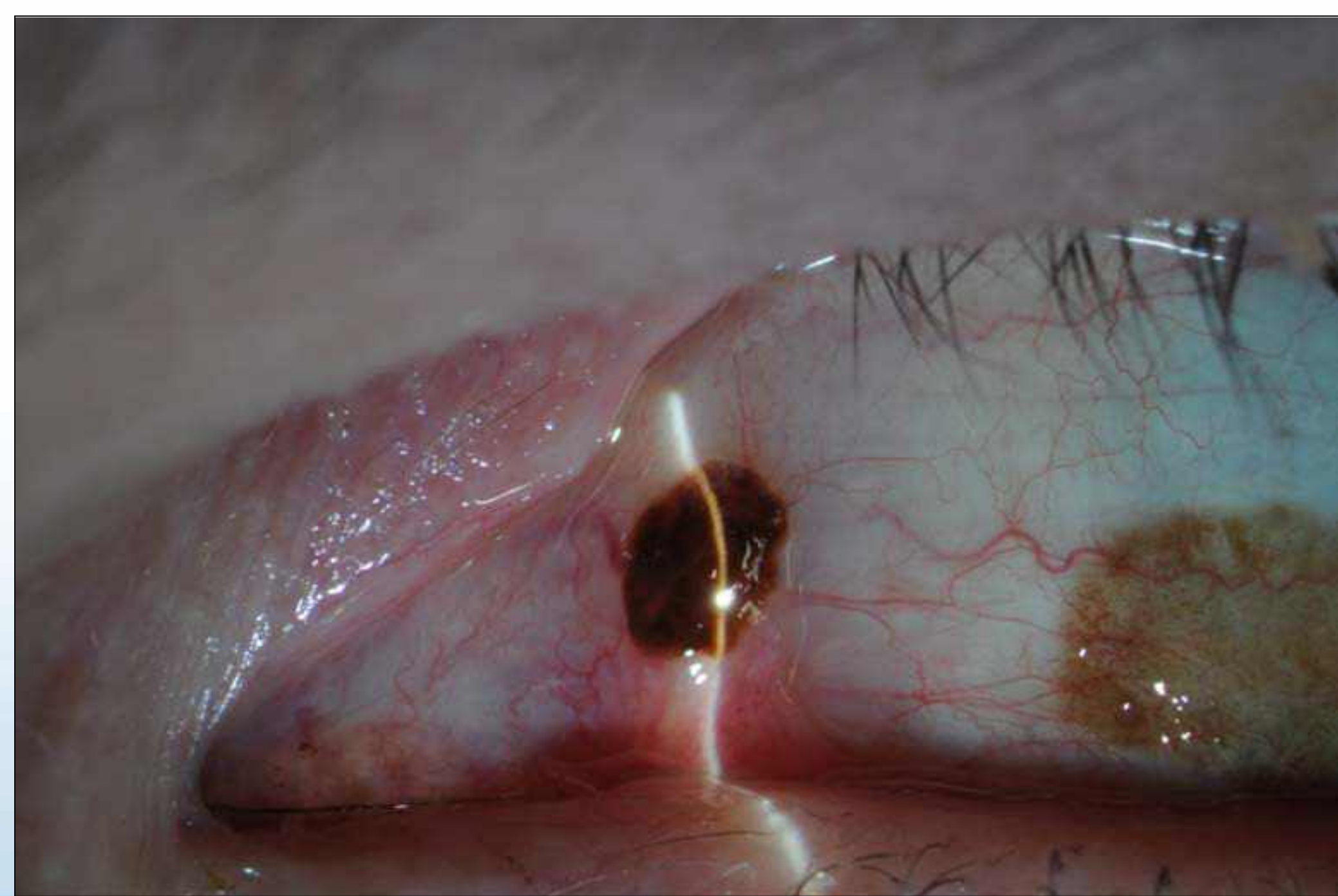


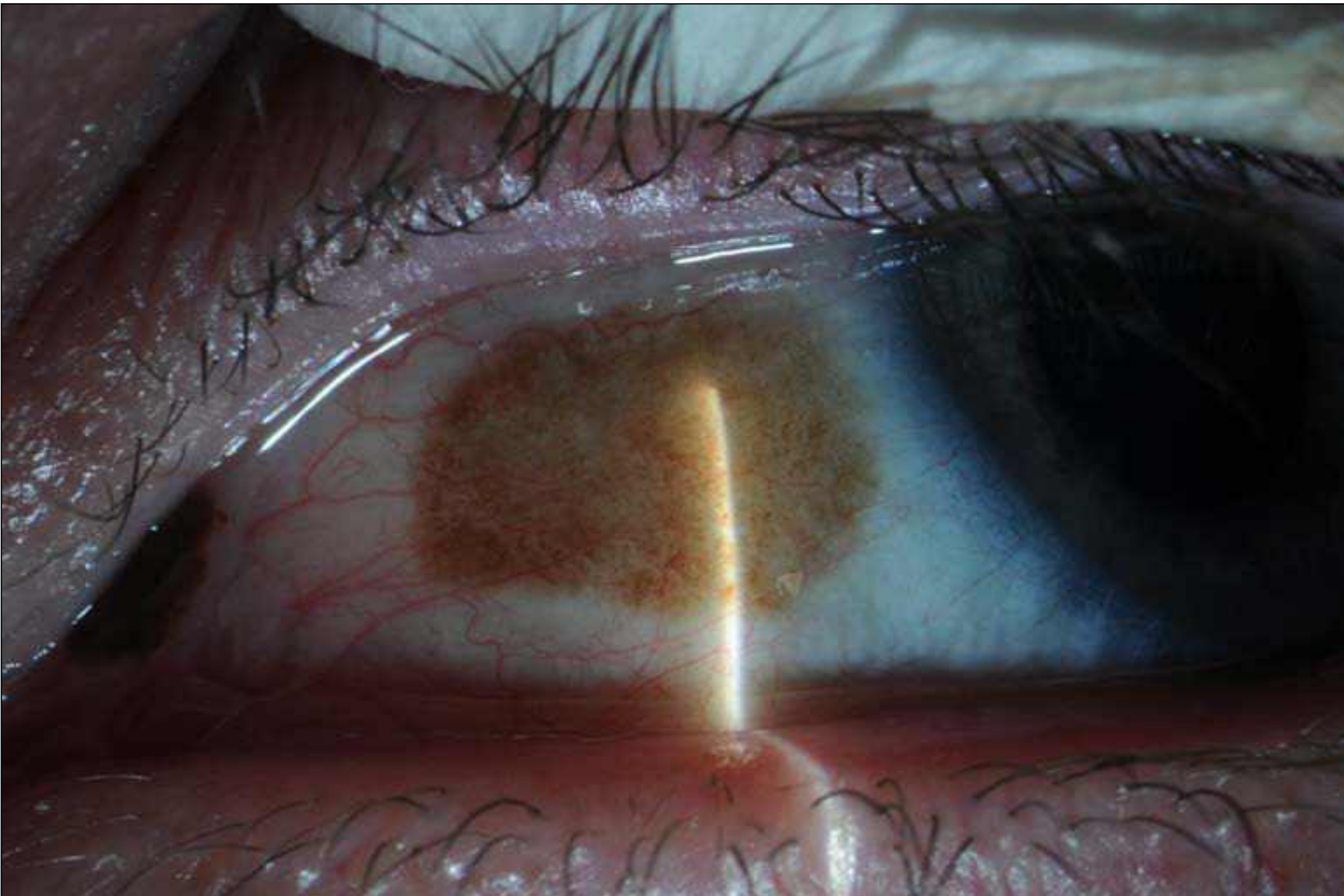


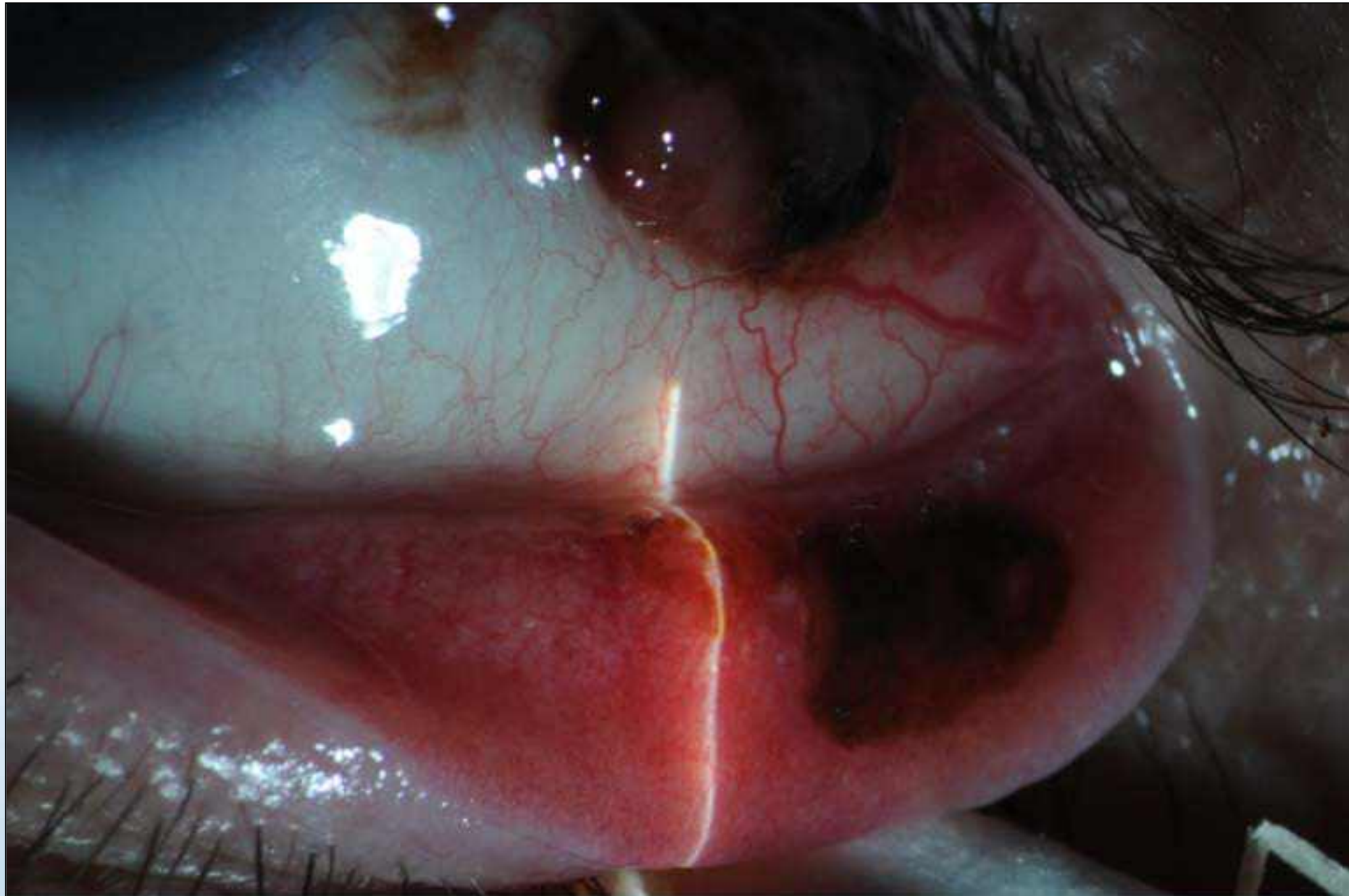




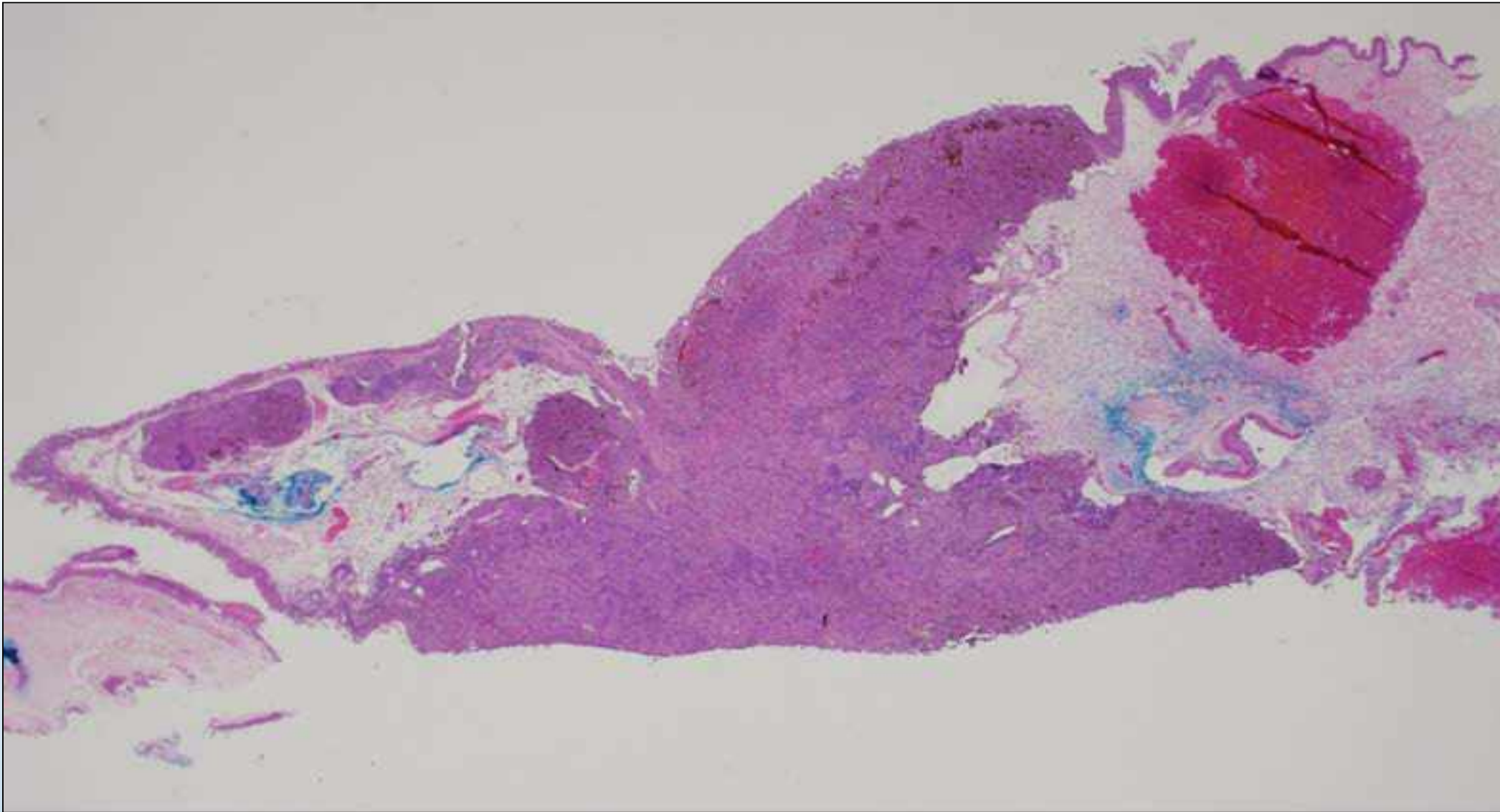




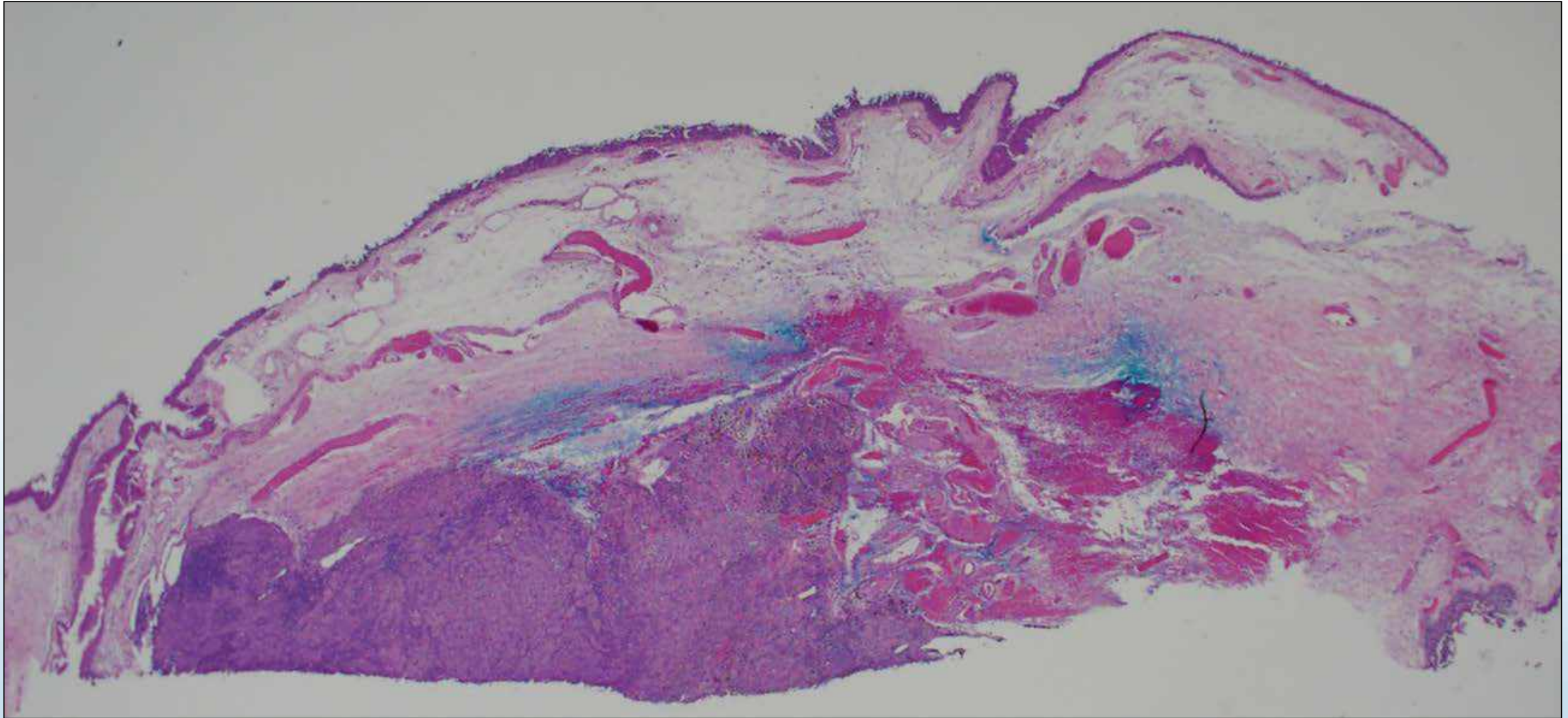




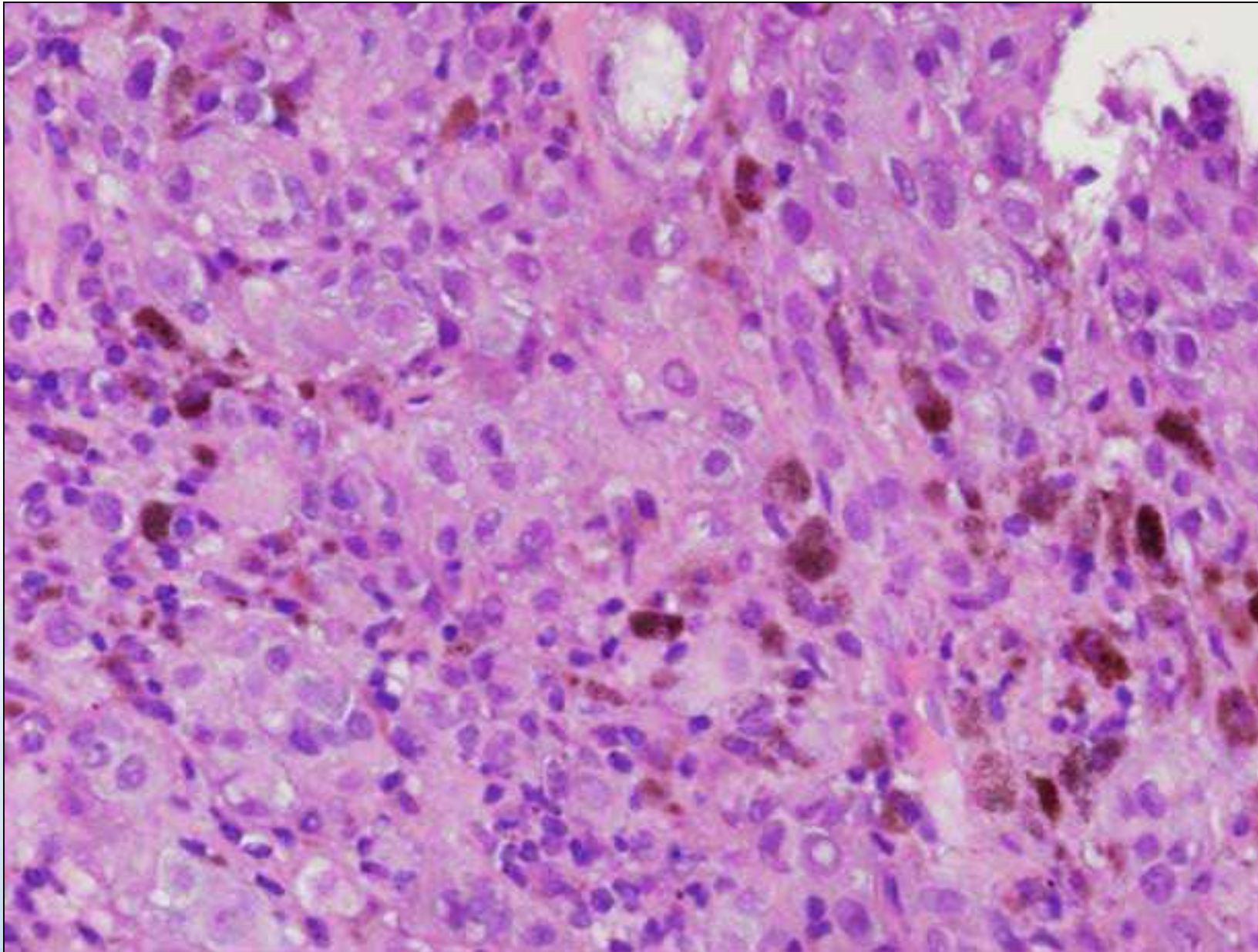
Left Temporal



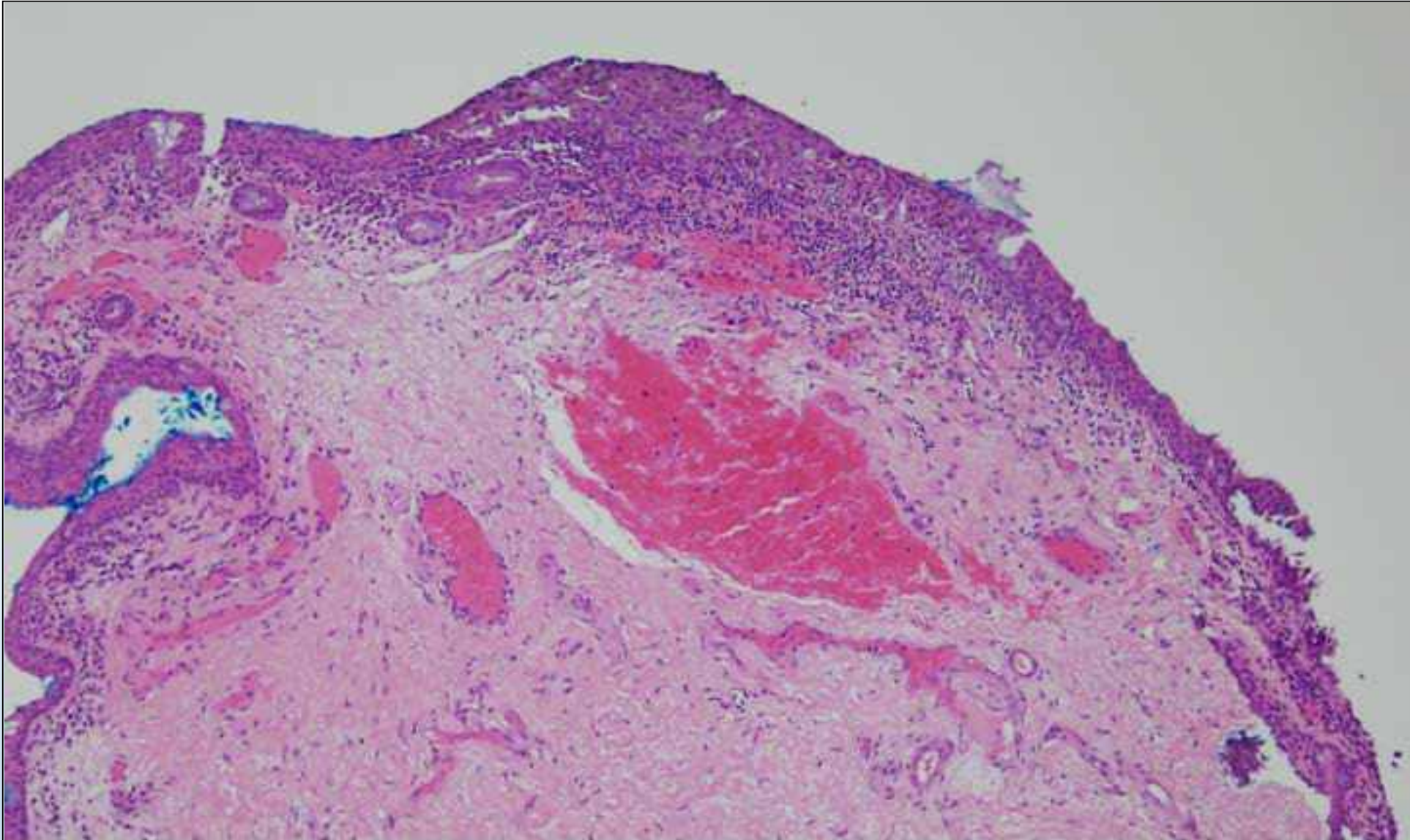
Left Temporal



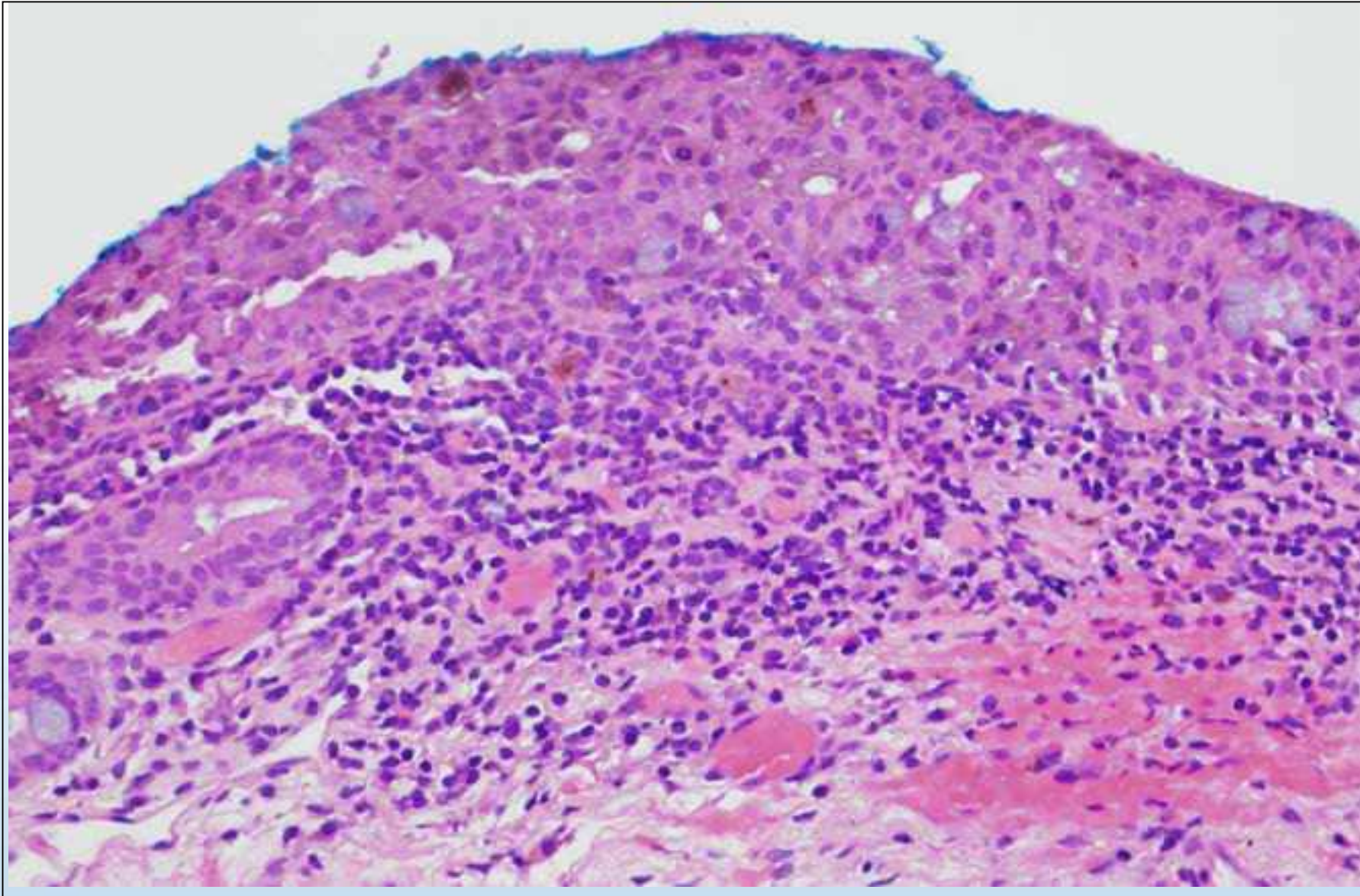
Left Temporal



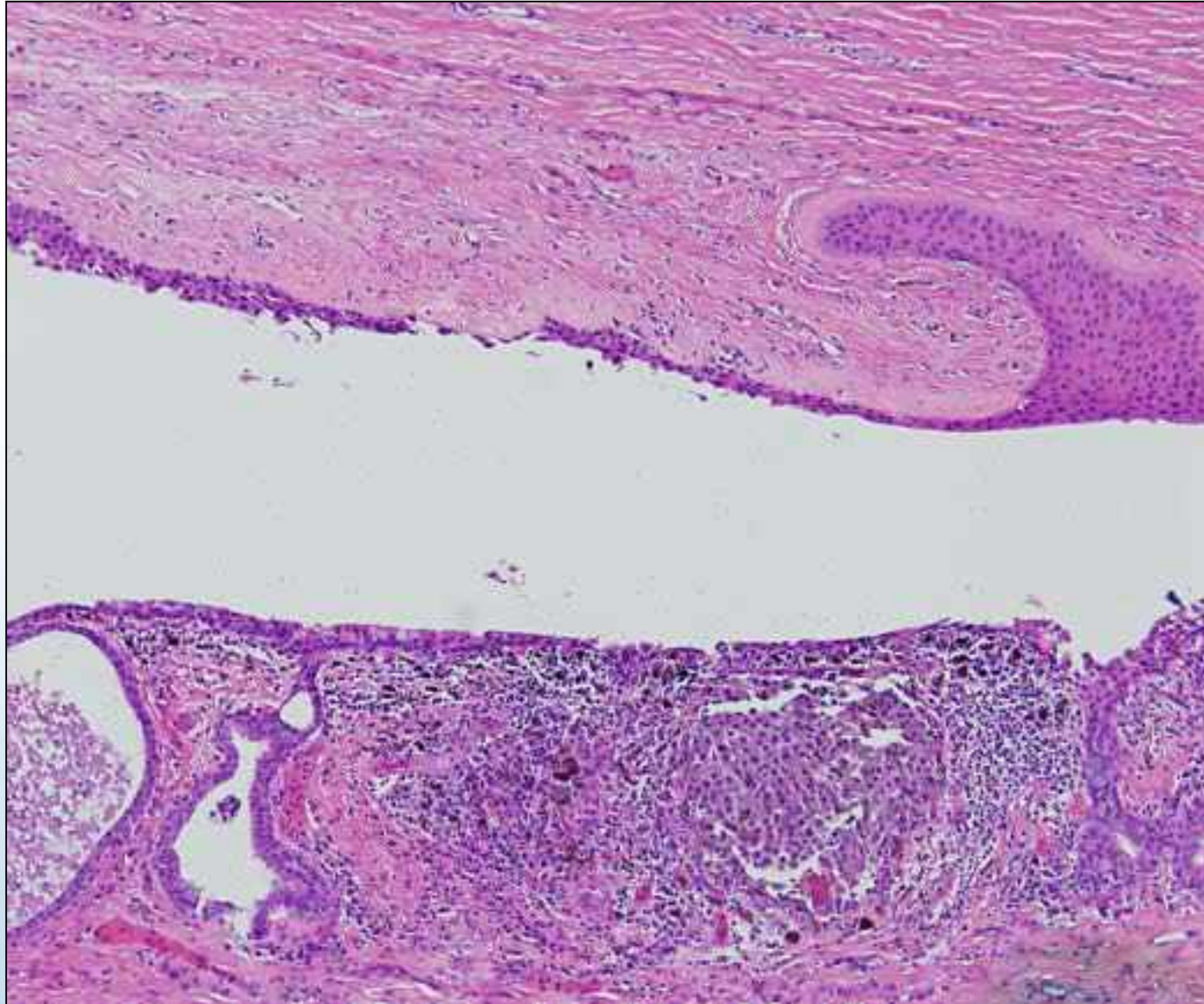
Left Nasal



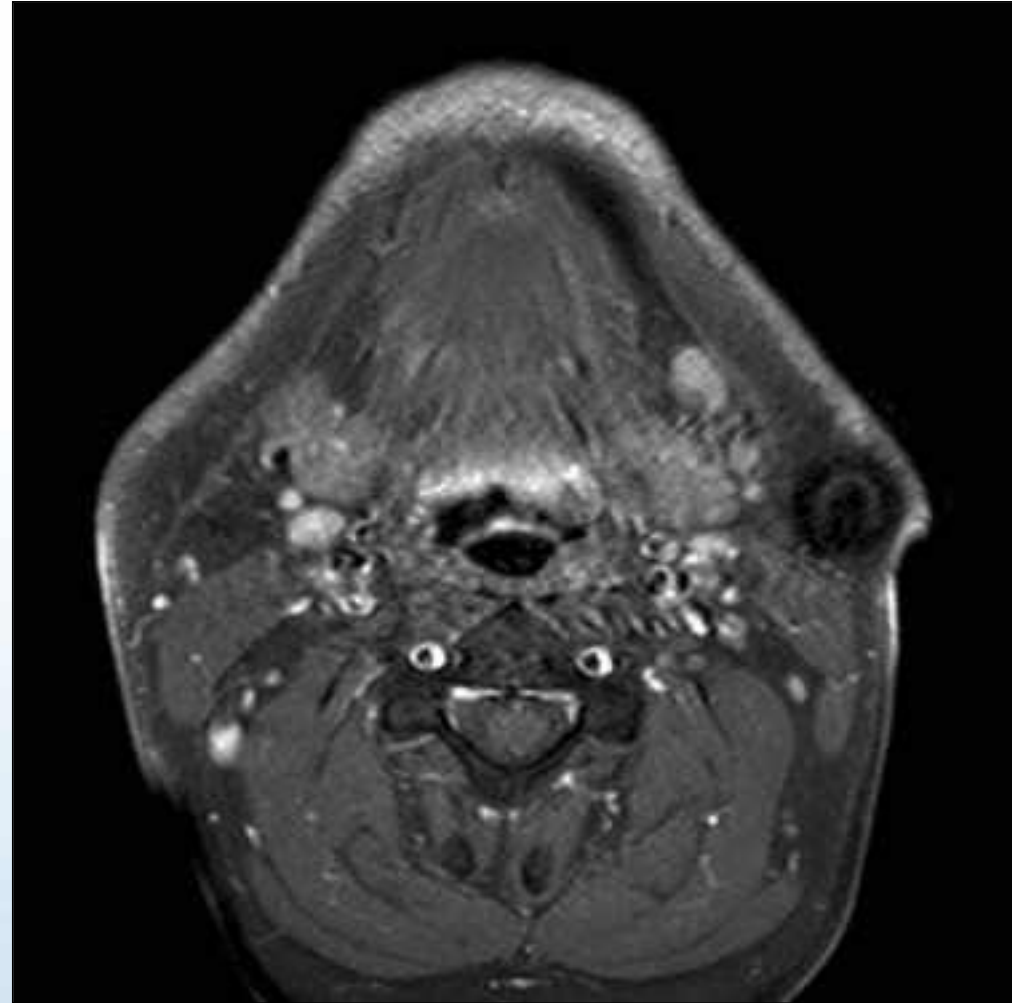
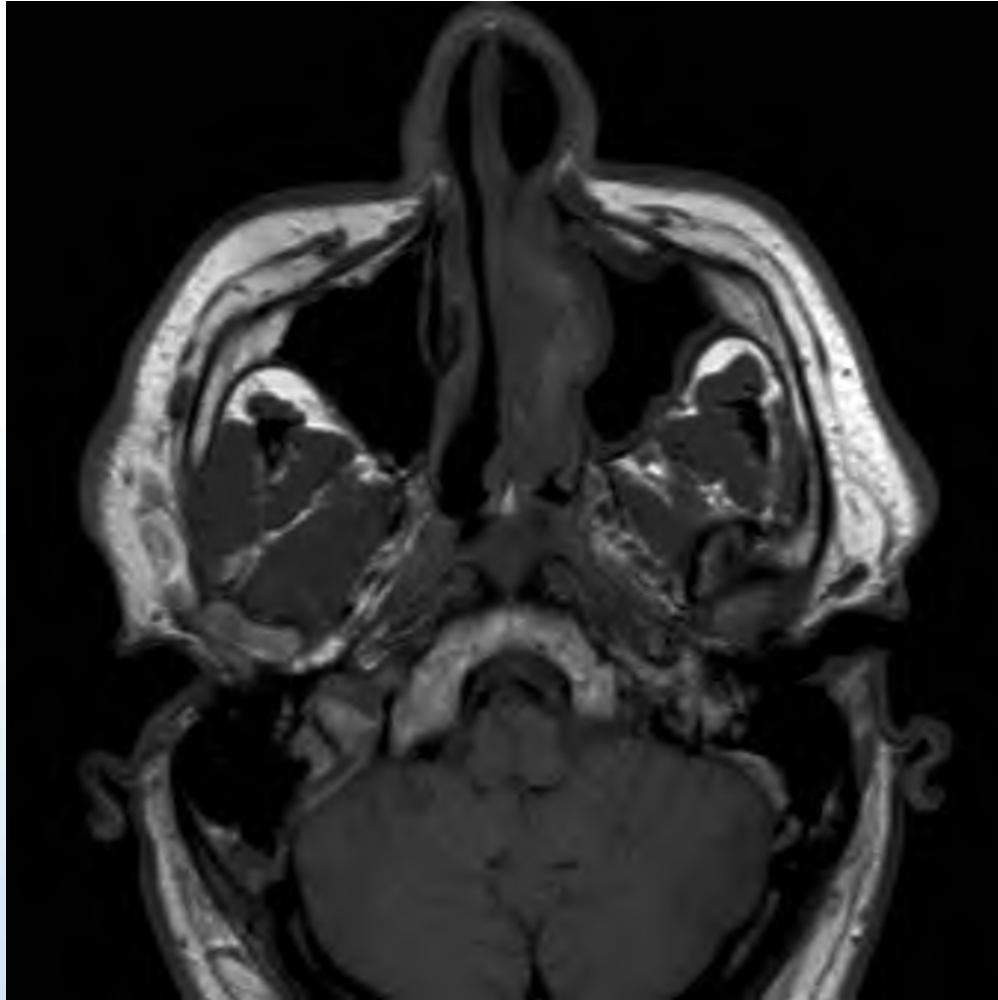
Left Temporal



Palpebral conjunctiva



MRI/PET

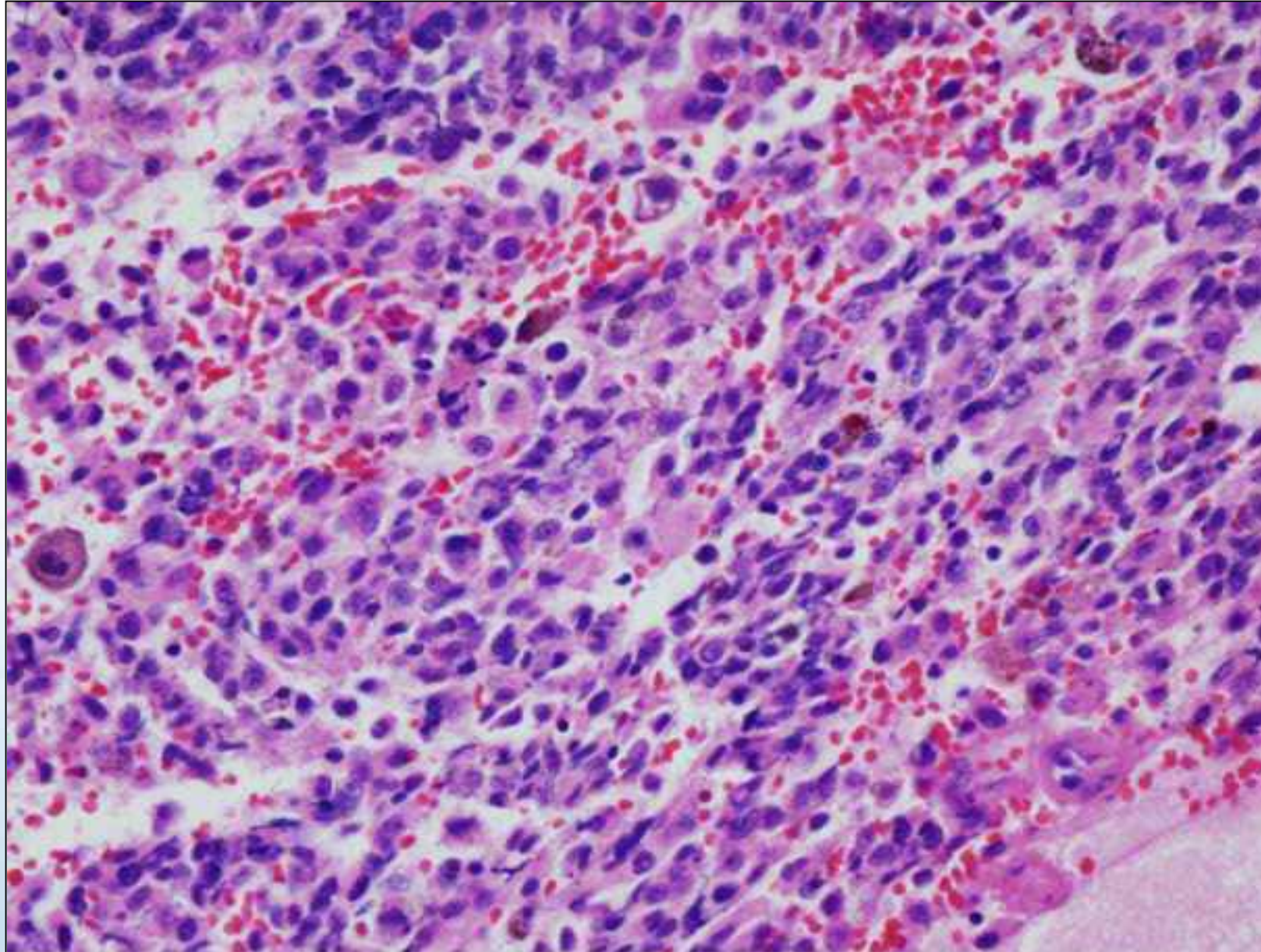


Clinical progression

- **6mo after extenteration:**
 - **CC: epistaxis**
 - **Referred to ENT – soft tissue mass in left nasal cavity**
 - **Biopsied in office**
- **Underwent endoscopy with tumor removal**
- **Continued radiation**
- **Died 6 mo later**



Nasal sinus

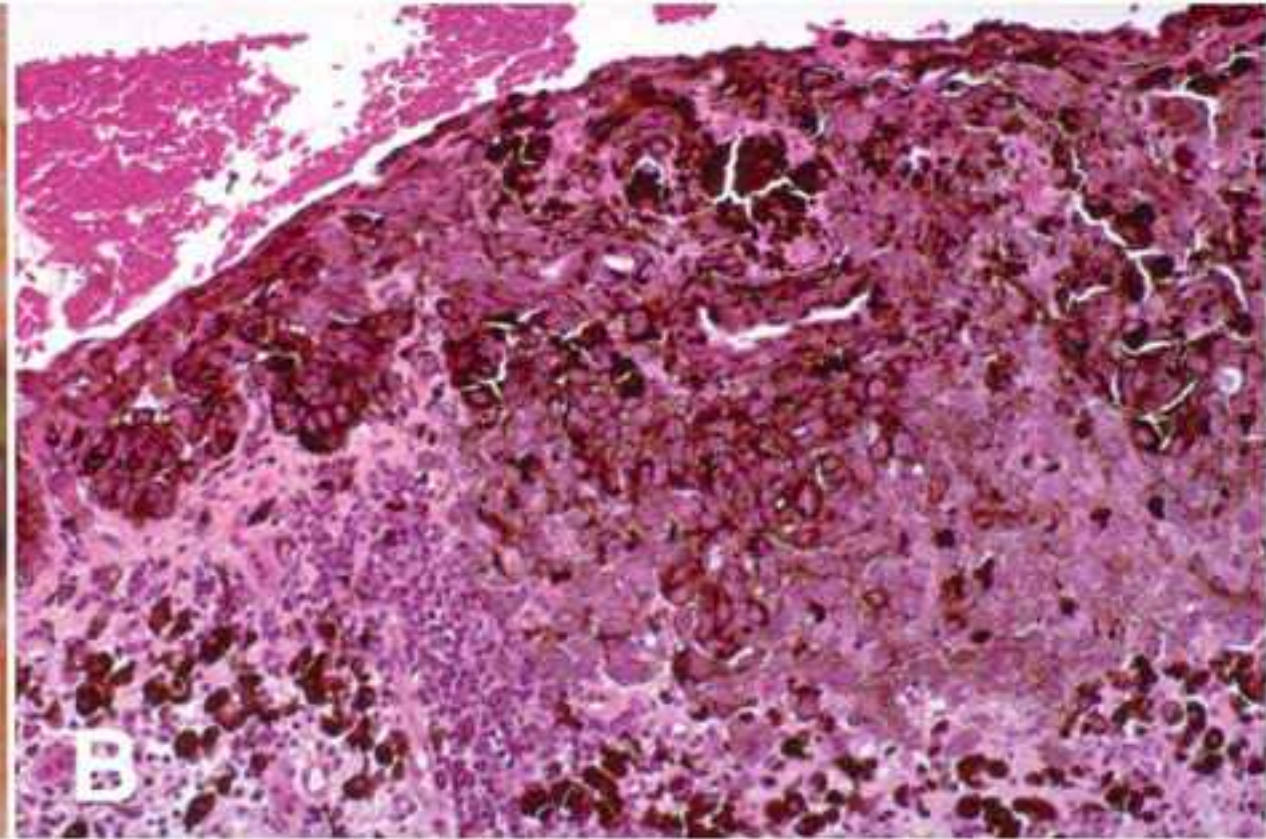


Conjunctival Melanoma

- Rare lesions: 5% of ocular melanomas
- Better prognosis than uveal melanoma
- Mortality 26%
- Most (75%) arise from PAM with atypia
- May arise from nevi or de novo
- Spread to regional lymph nodes (preauricular, intraparotid)



Malignant Melanoma Arising in PAM with Atypia



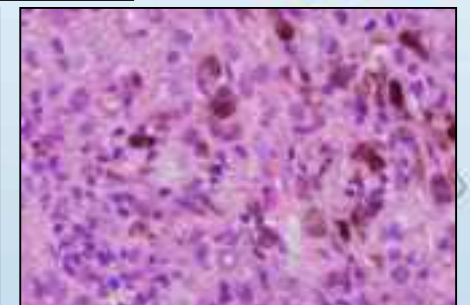
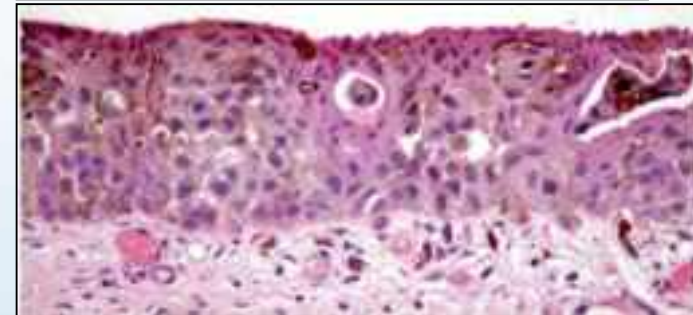
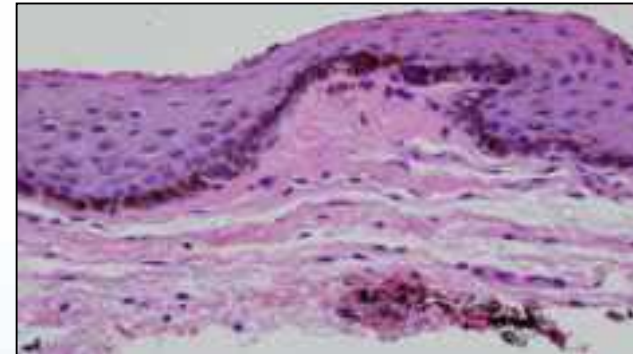
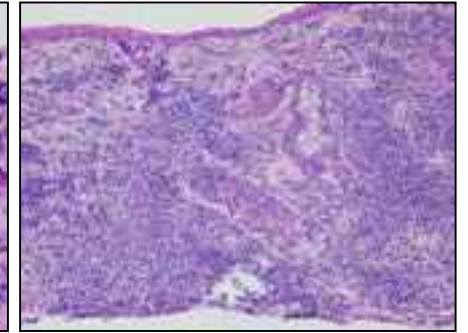
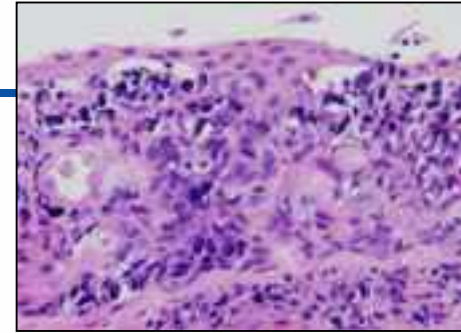
Conjunctival melanocytic intraepithelial lesions

- **Conjunctival pigmented lesions**
- **Primary acquired melanosis (PAM)**
- **Conjunctival melanocytic intraepithelial neoplasia (CMIL) – WHO classification**
 - **Benign melanosis, PAM w/ mild atypia = low grade CMIL**
 - **PAM w/ mod/severe atypia = high grade CMIL**



Pigmented Conjunctival Lesions

- **Conjunctival Nevi**
- **Flat pigmented patches**
 - Complexion melanosis
 - Freckles and lentiginos
 - PAM (early stages)
- **PAM with Atypia**
- **Malignant Melanoma**



EYELID



EYELID LESIONS: Differential Diagnosis

- **Epidermis**

- Benign: SK, Papilloma, EIC
- AK, KA
- Malignant: BCC, SCC

- **Adnexal**

- Sebaceous
- Sweat glands
- Hair follicles

- **Melanocytic**

- Nevi
- Melanoma

- **Vascular**

- Hemangioma

- **Neural**

- Neurofibroma
- Traumatic neuroma

- **Miscellaneous**

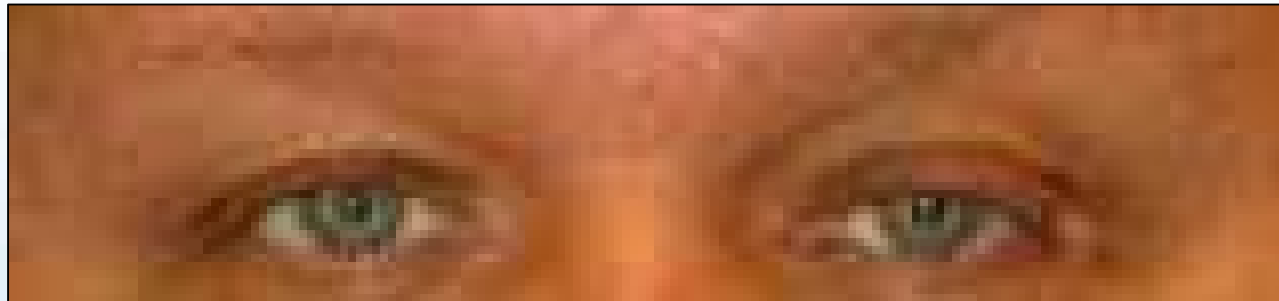
- Lymphoid
- Chalazion
- Xanthelasma

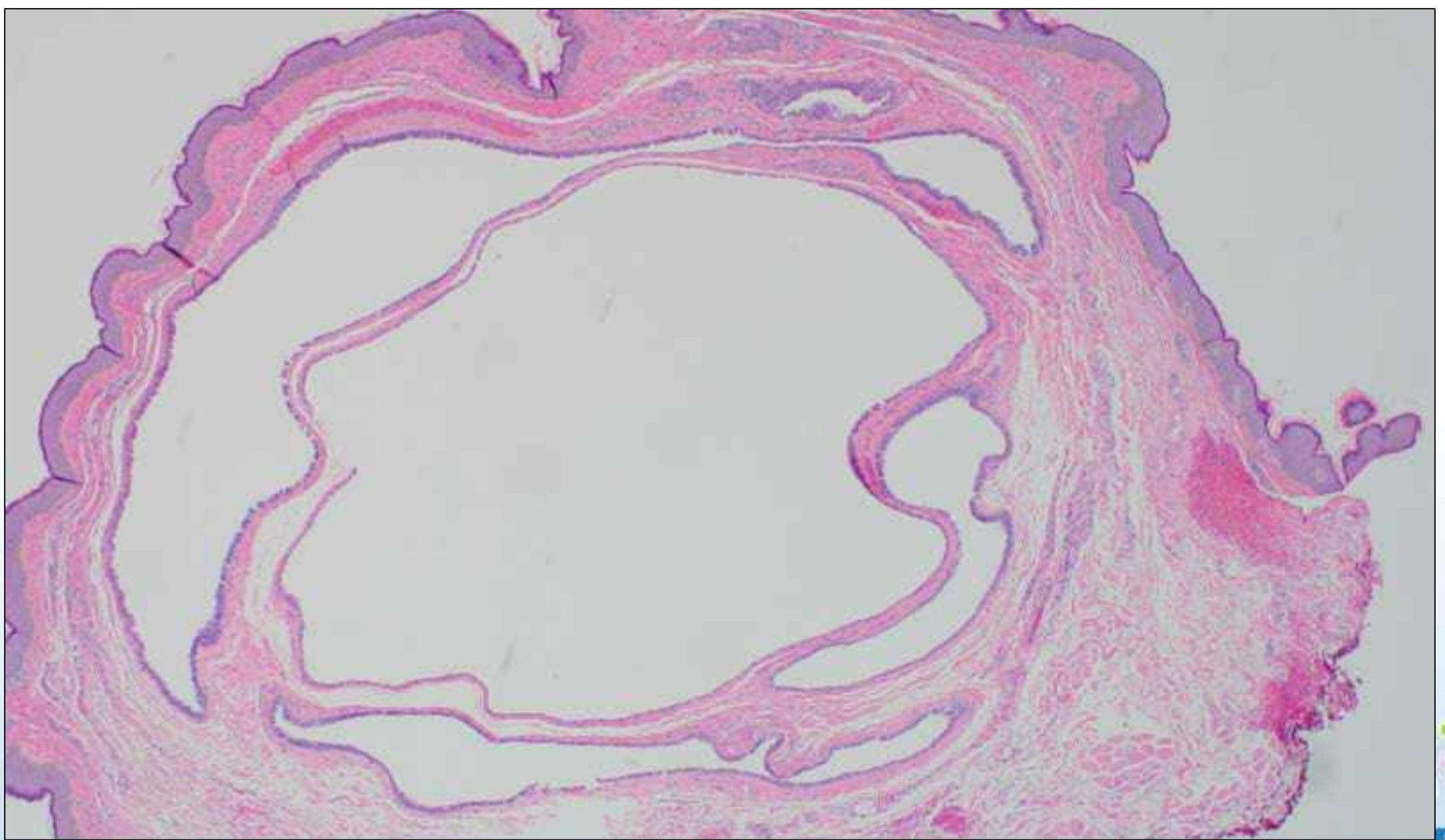
- **Metastatic**

- Breast (50%), lung, prostate, melanoma, RCC

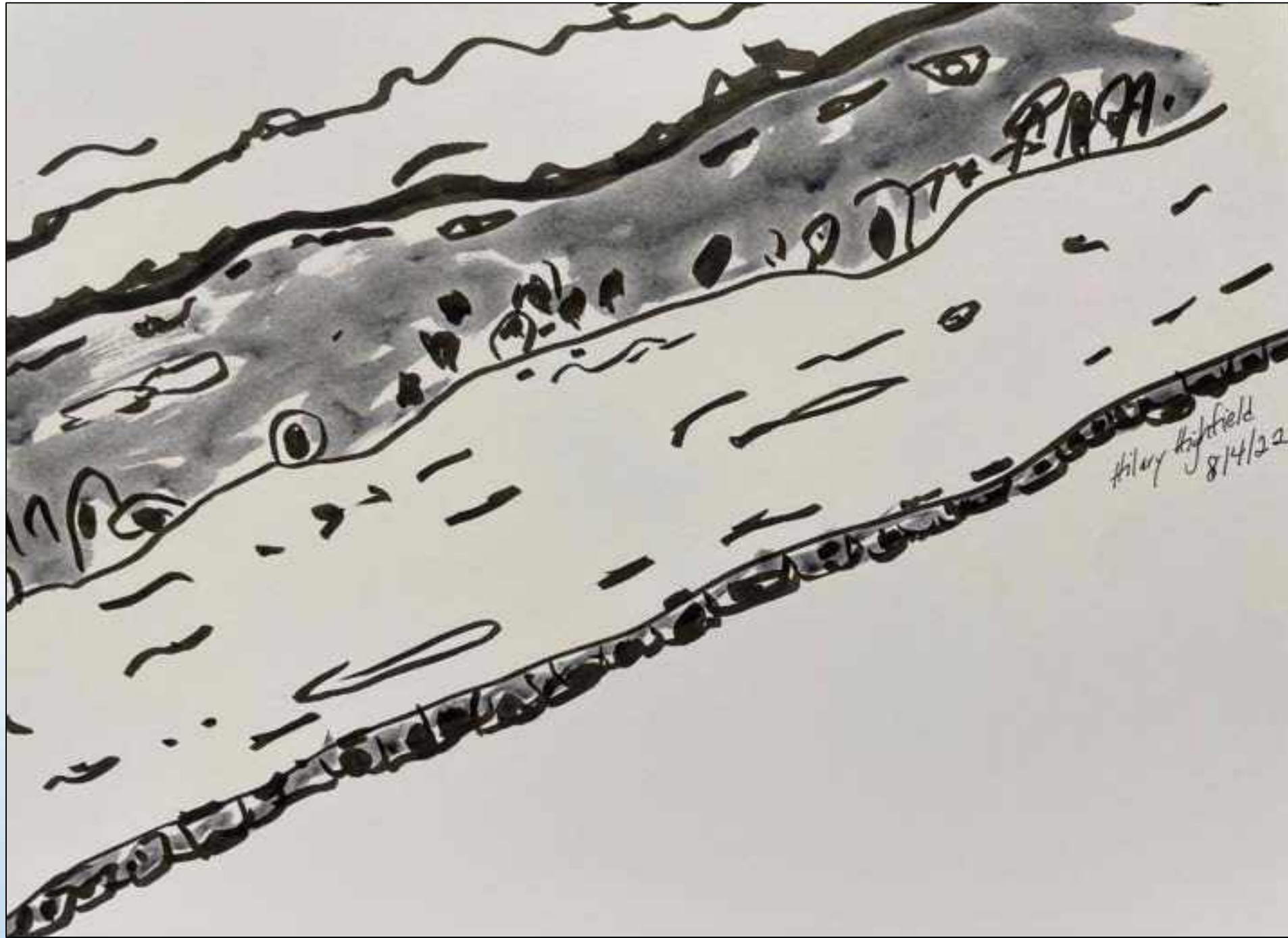


HIDROCYSTOMA

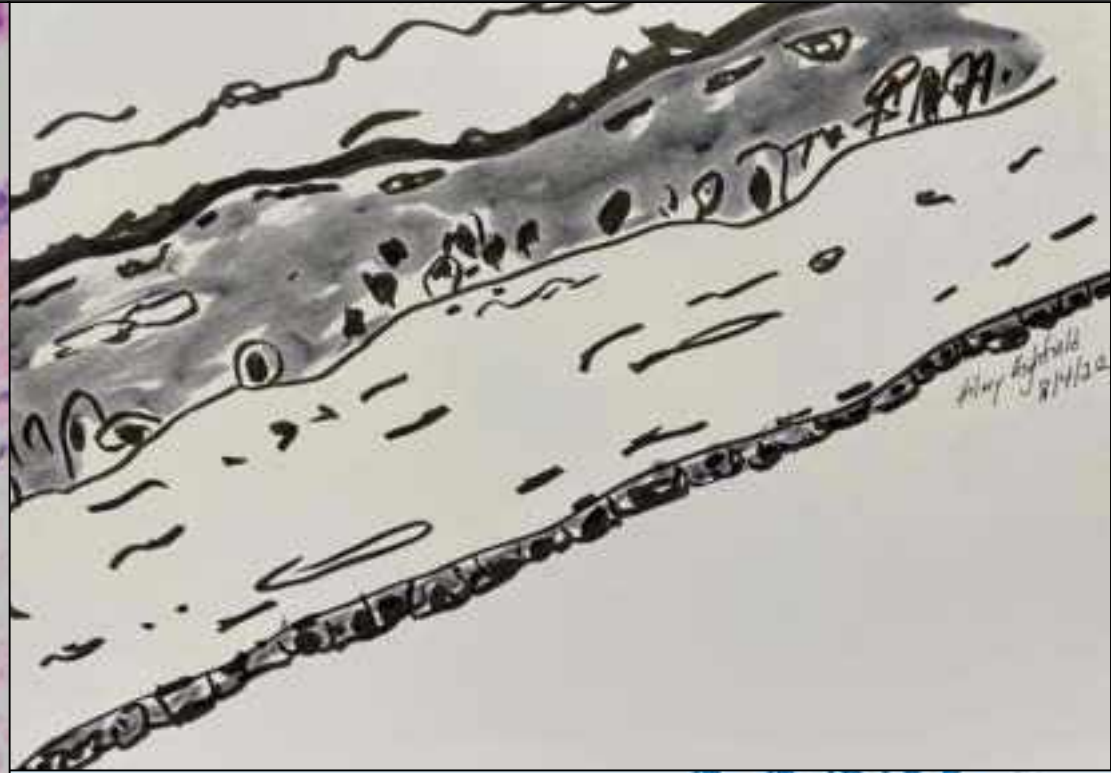
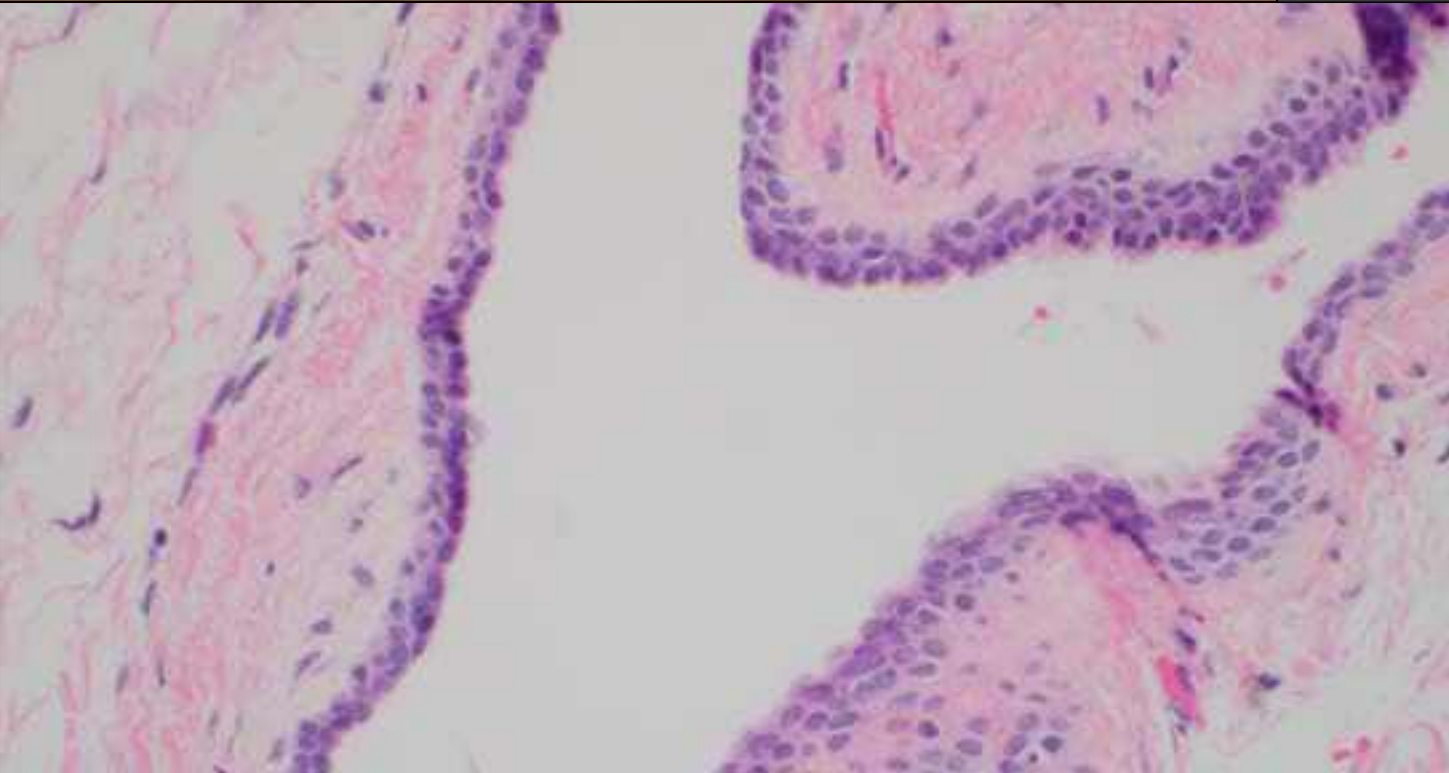
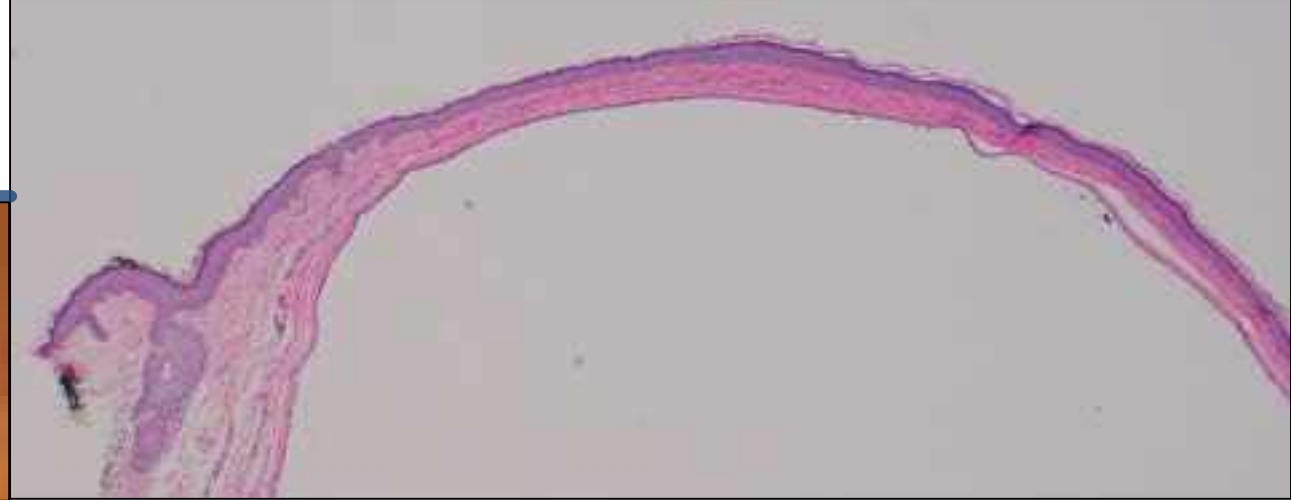
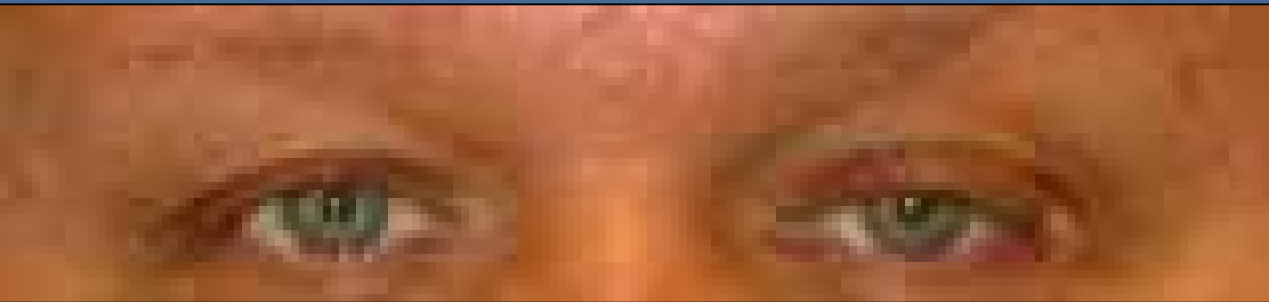


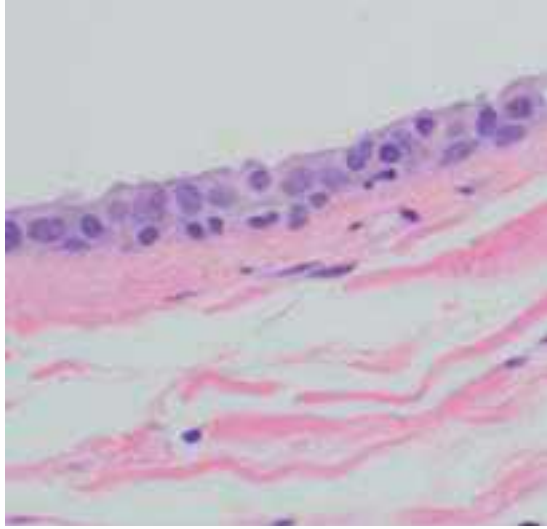




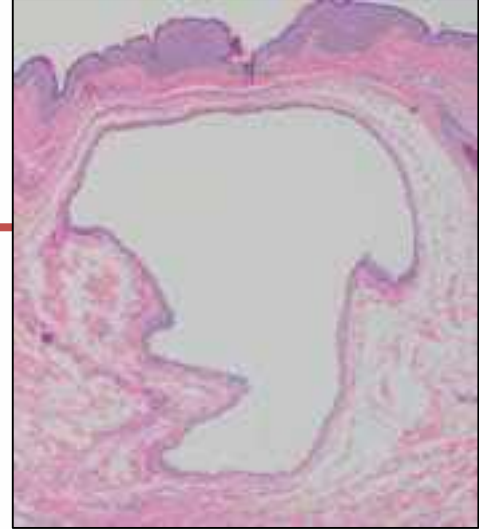


HIDROCYSTOMA



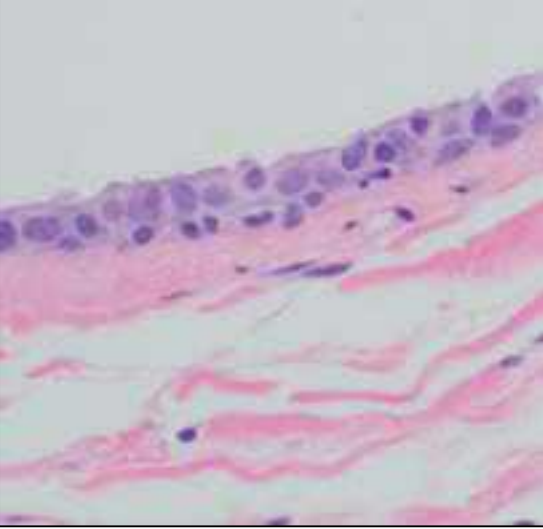


Hidrocystoma



- Cystadenoma or Moll's gland cyst
- Cyst of sweat duct
- Eccrine or apocrine
- Low cuboidal epithelium
- Apocrine with decapitation secretion

HIDROCYSTOMA



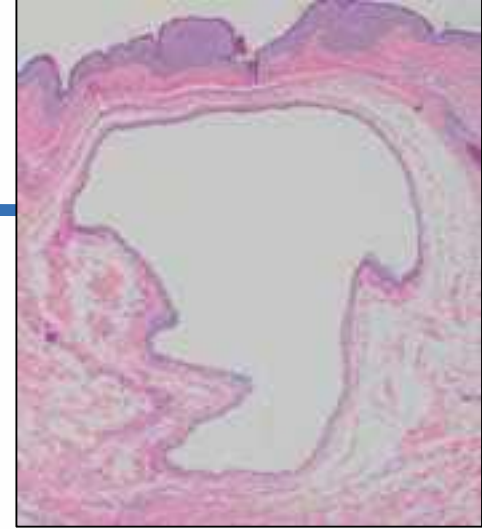
Eccrine: retention cyst of eccrine sweat gland

Common in eyelid region; often multiple

Clear translucent lesion near eyelid margin

Smooth shiny overlying skin

Clear cystic lesion lined by bilayer cuboidal cells



Apocrine: retention cyst of apocrine gland

Usually solitary

Apocrine glands of eyelid (glands of Moll)

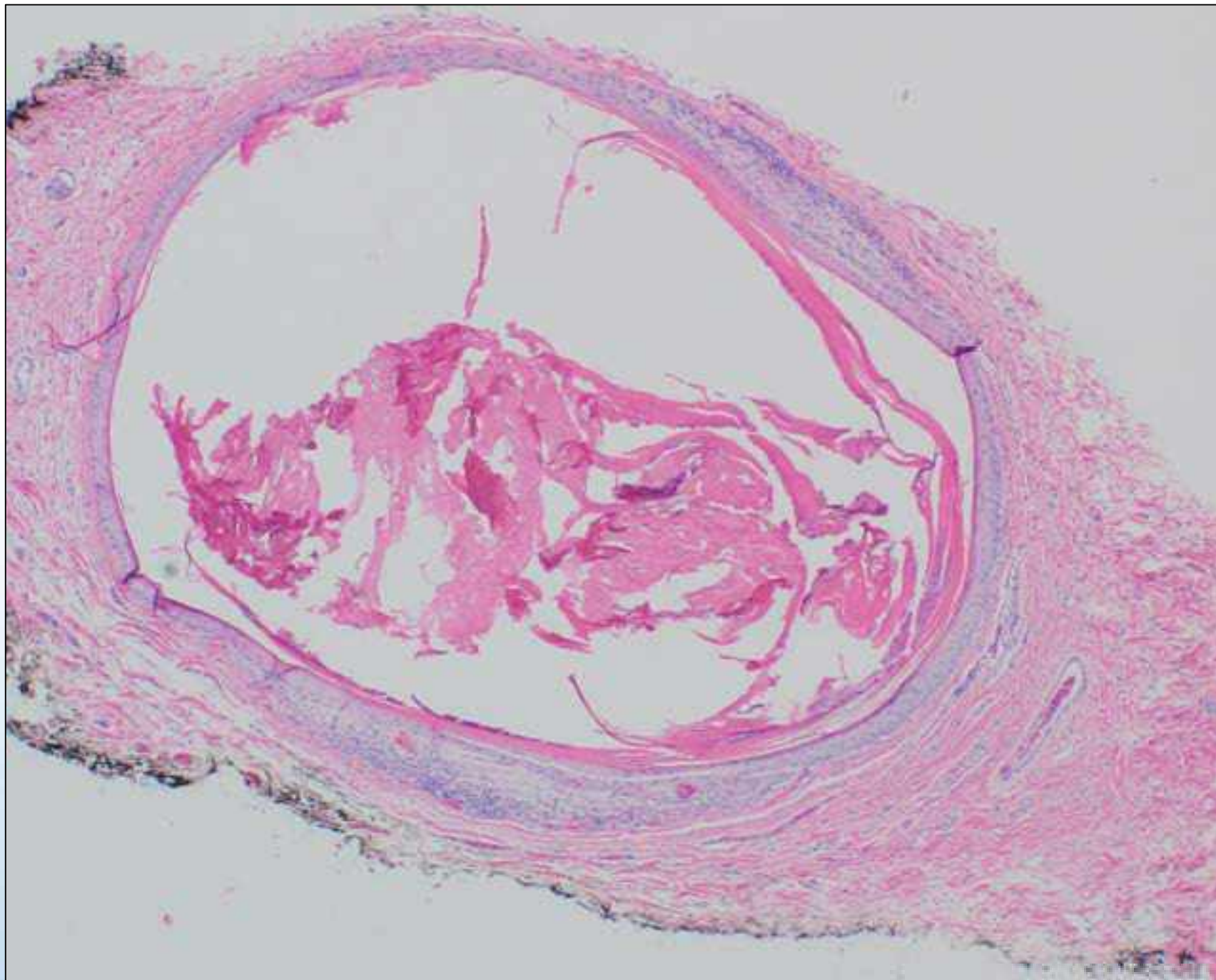
Frequently bluish

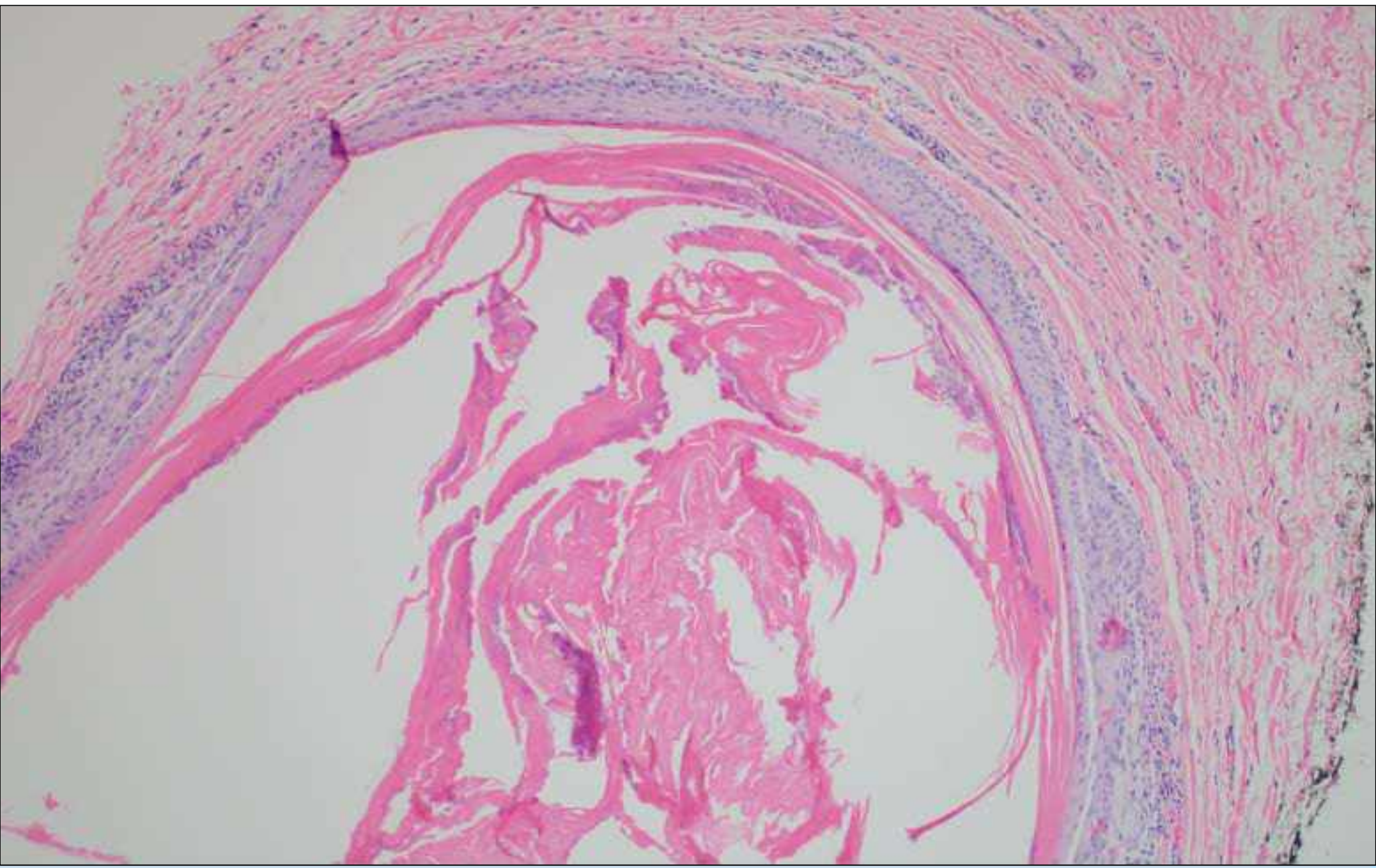
Decapitation secretion

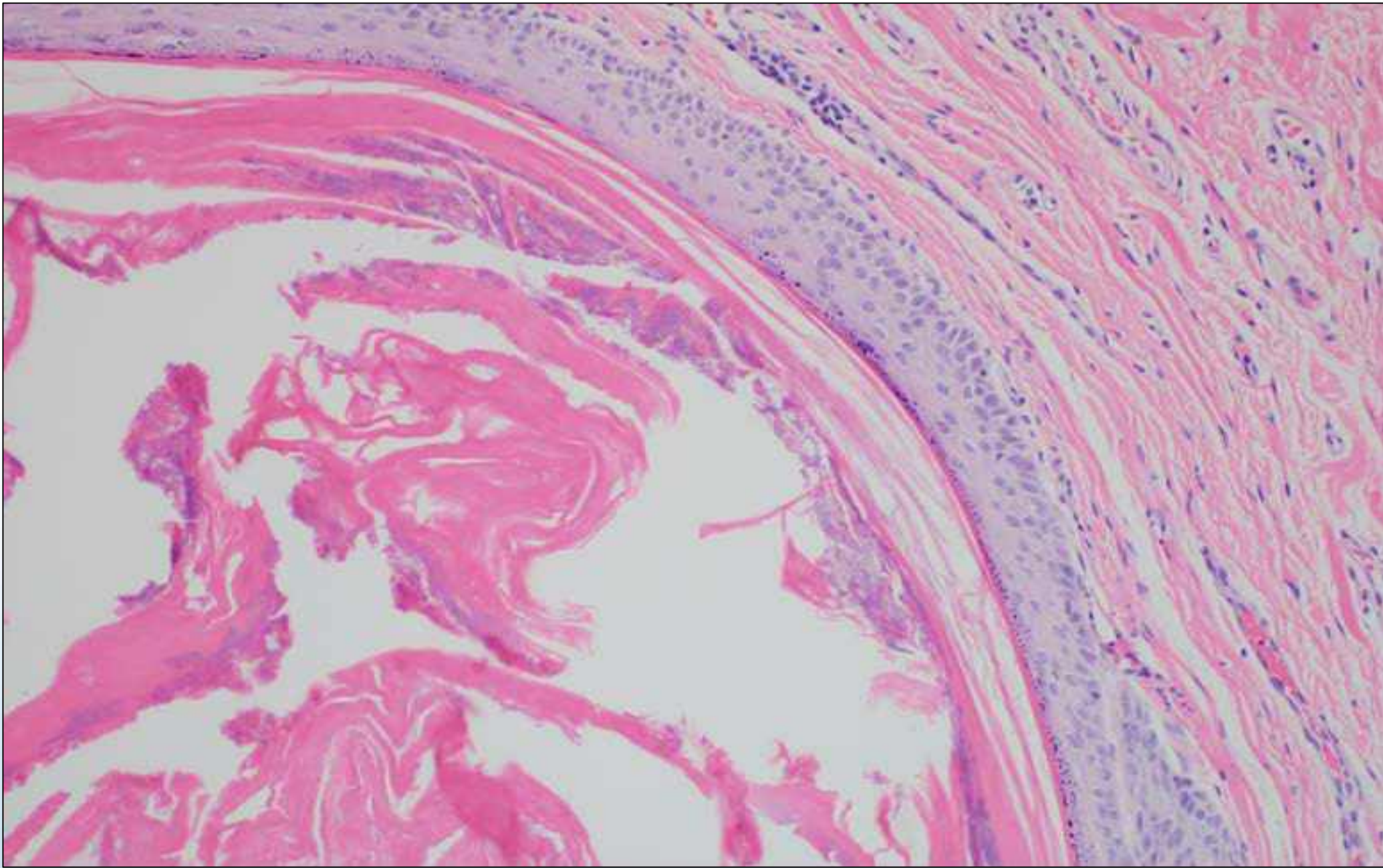


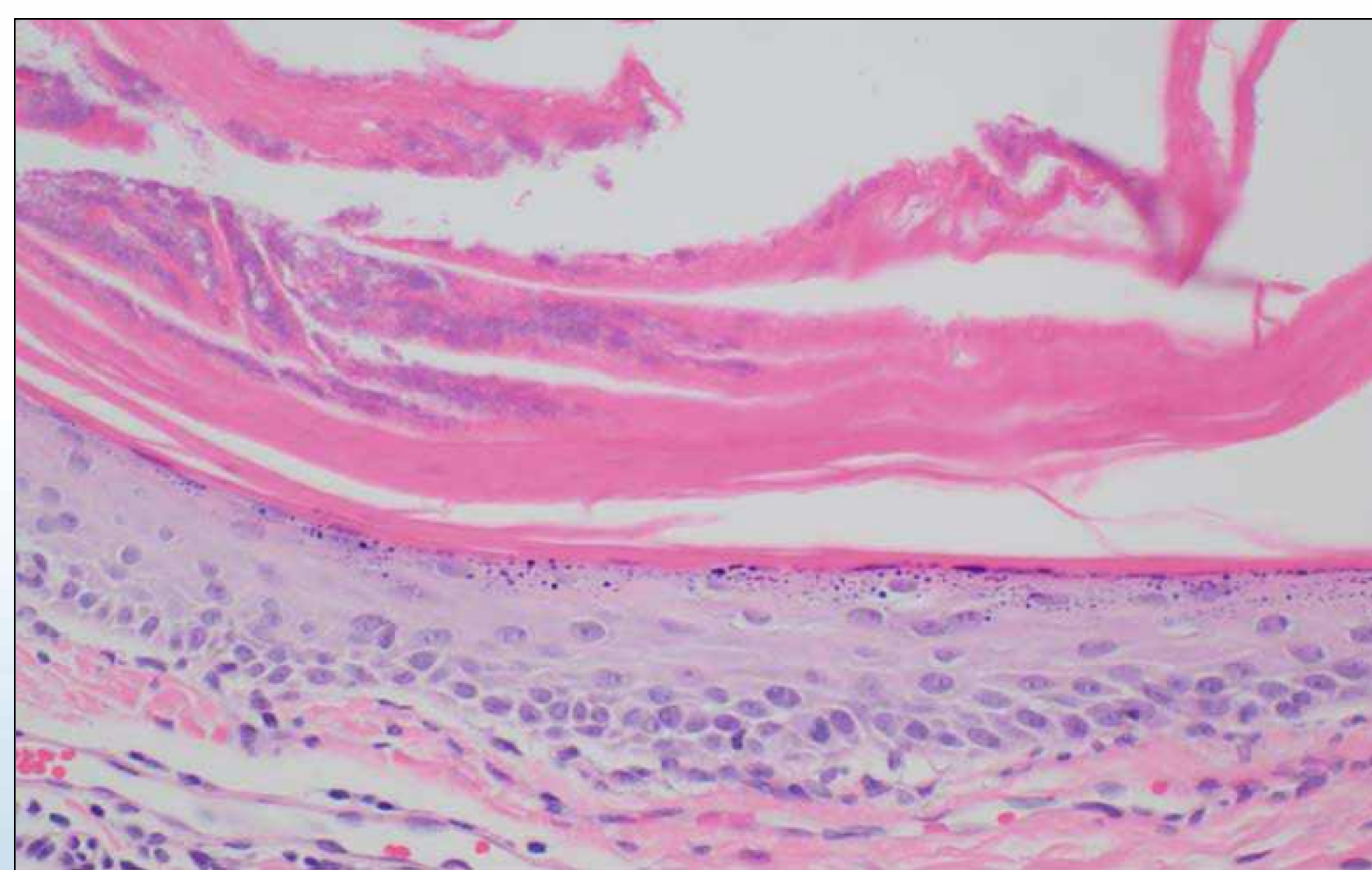
Epidermal Inclusion Cyst

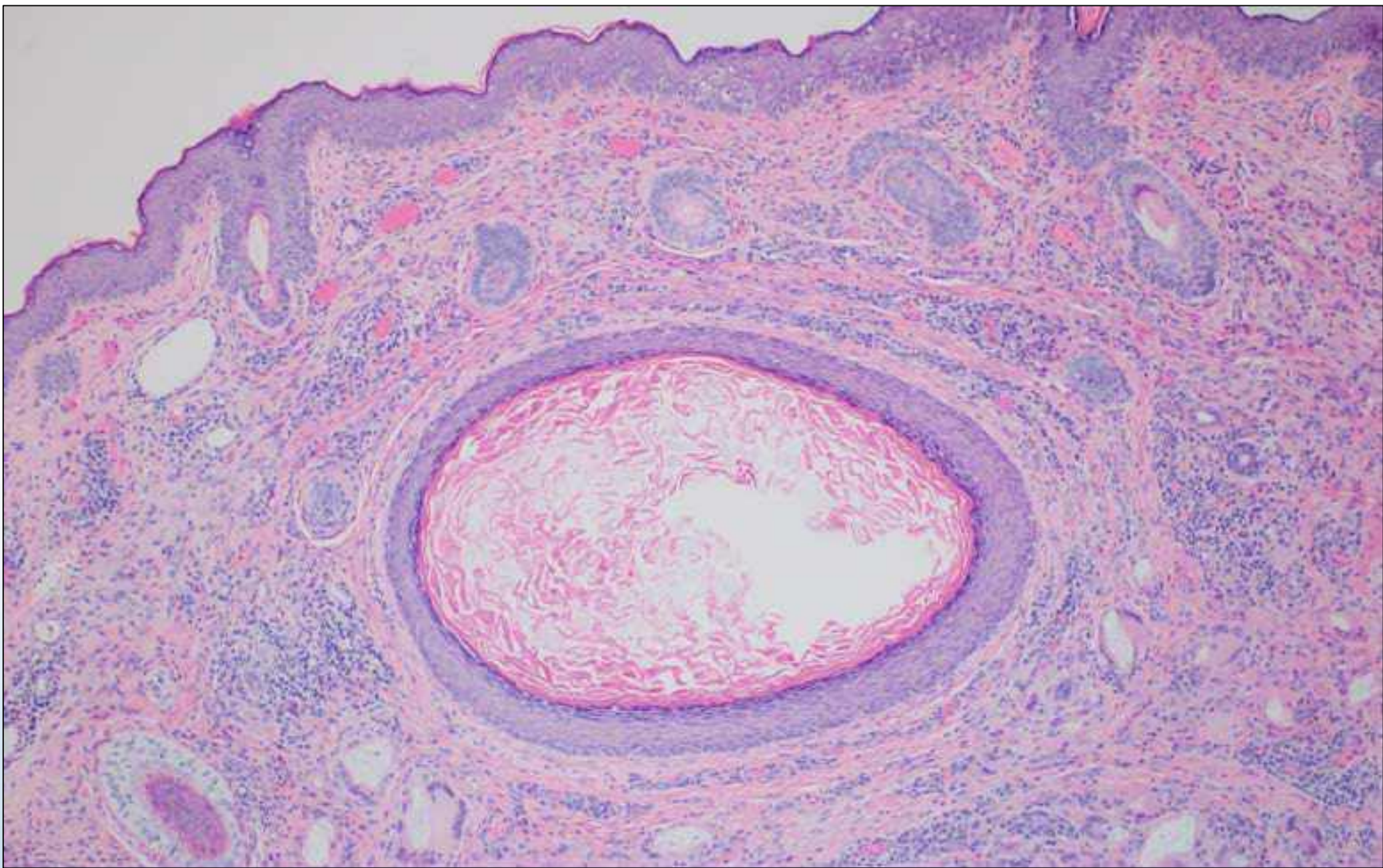


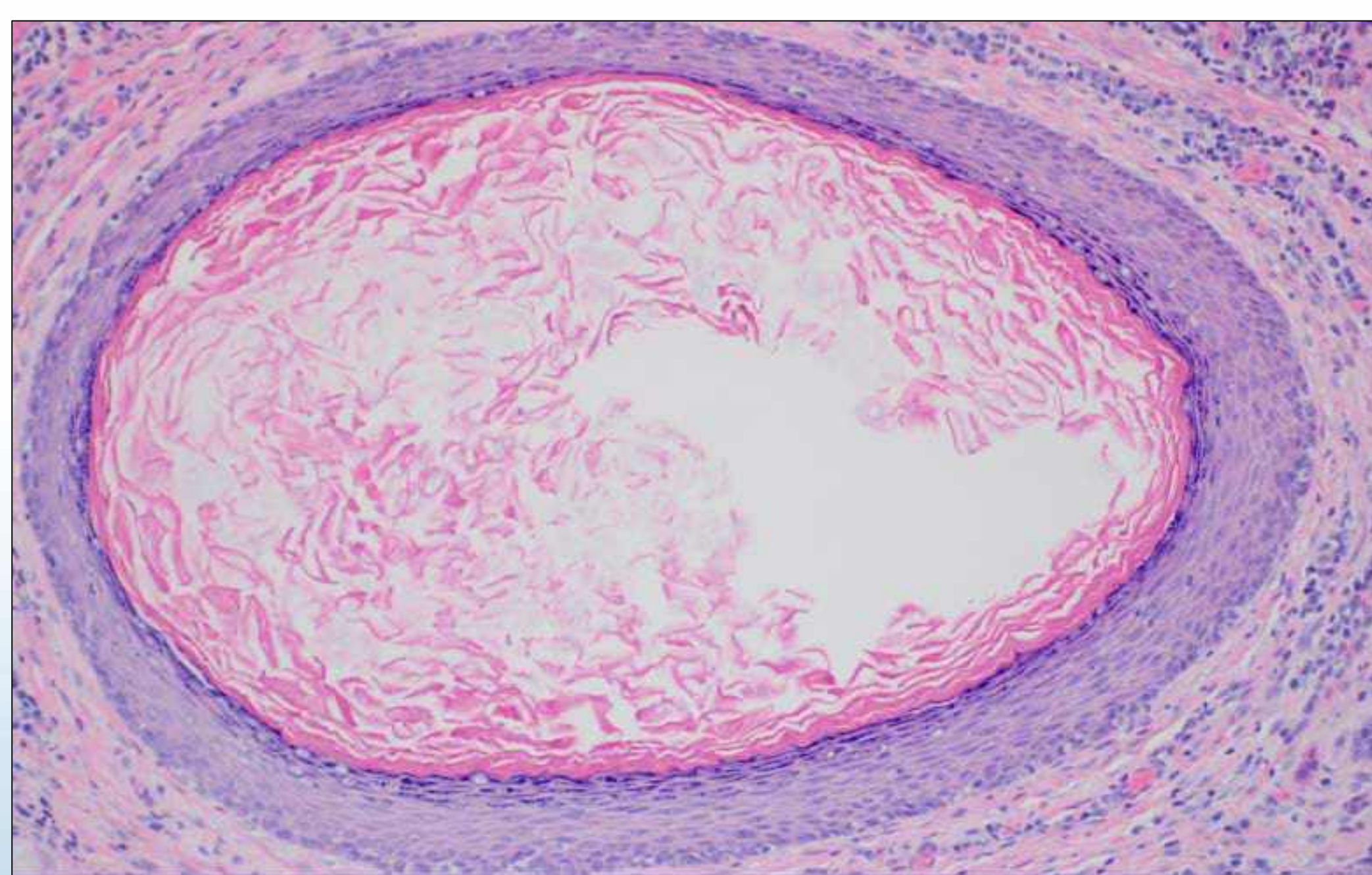


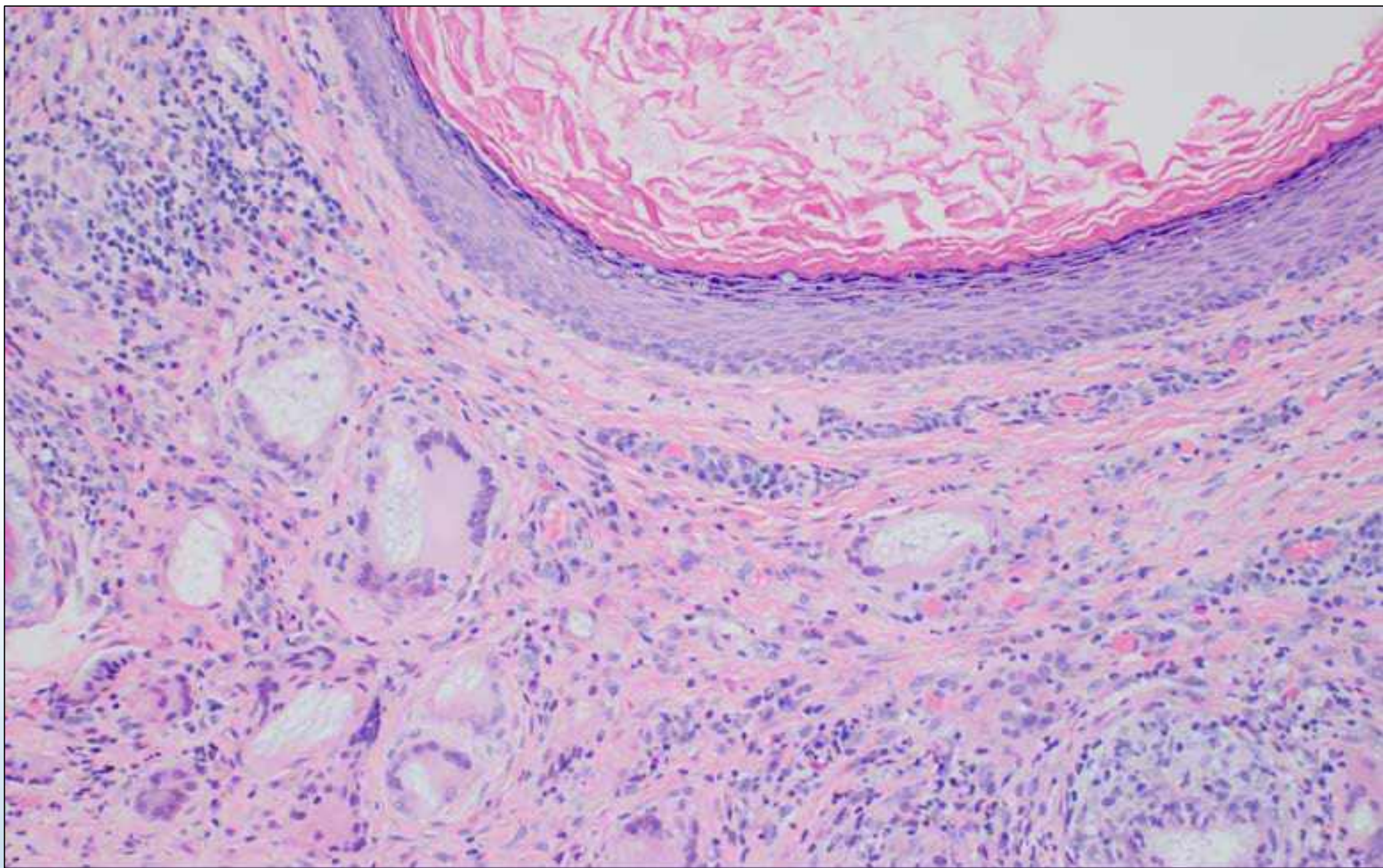


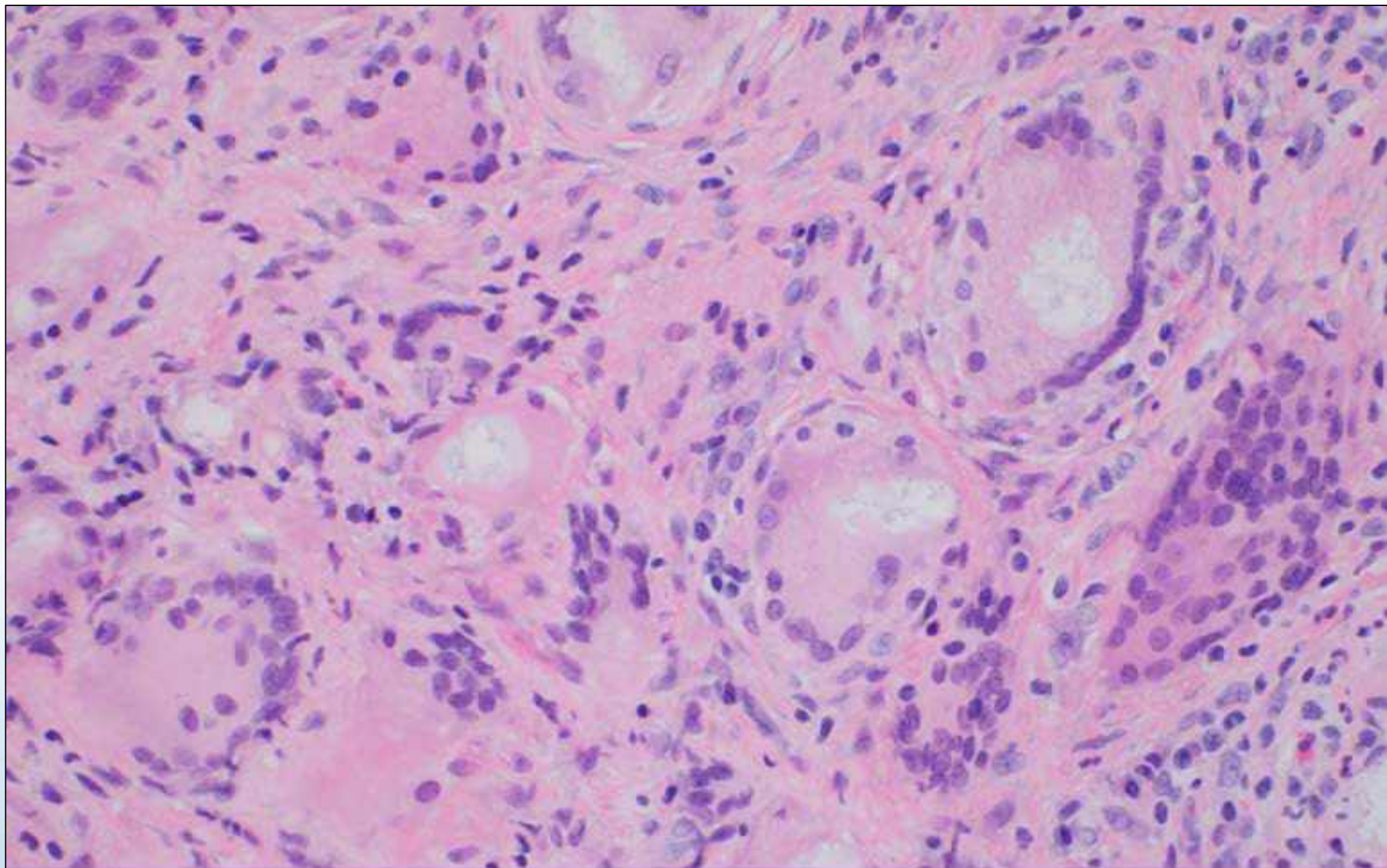


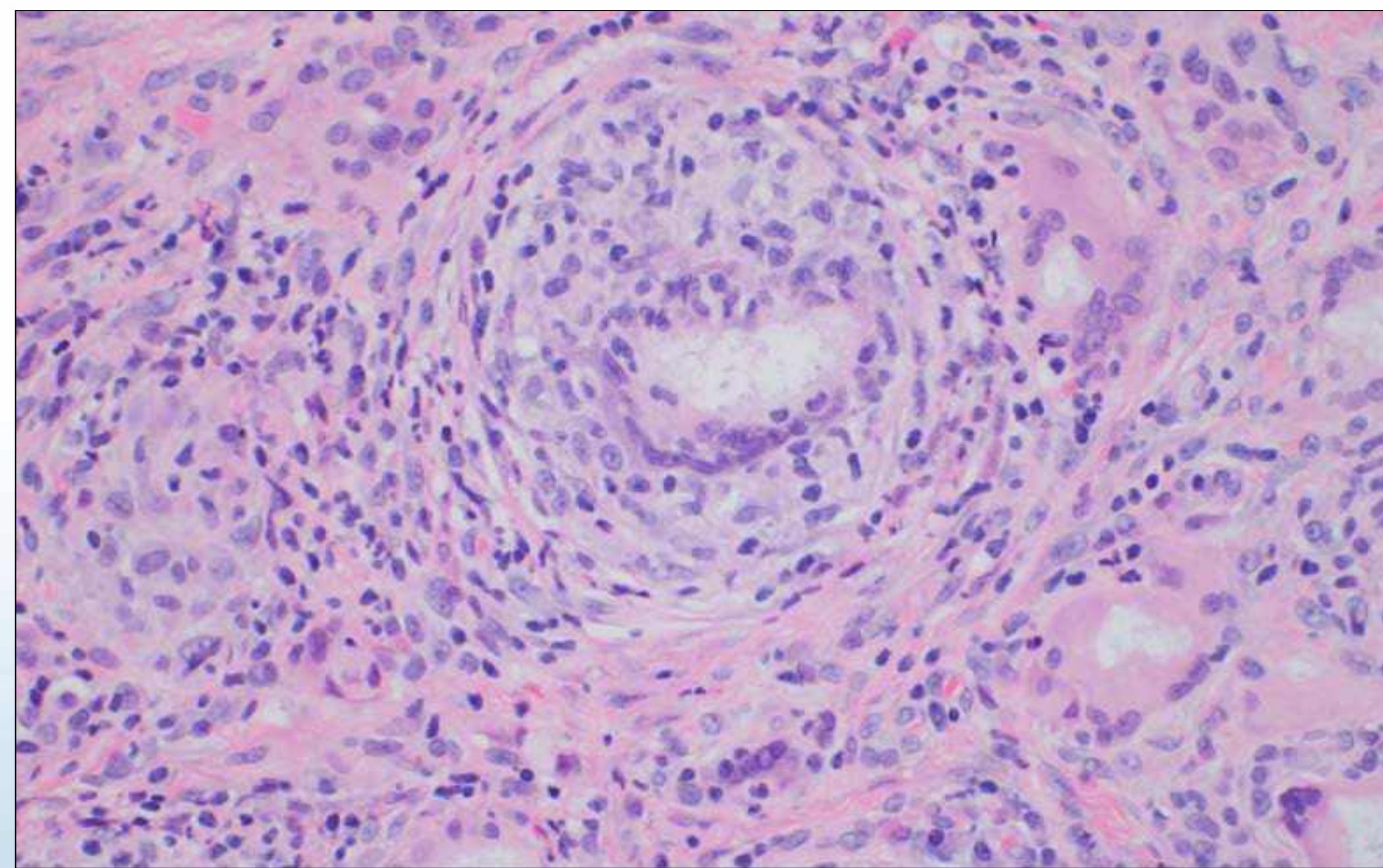














Epidermal inclusion cyst

- Cyst lined by squamous epithelium with granular cell layer containing lamellated keratin
- Rupture leads to granulomatous/foreign body giant cell reaction
- Multiple: syndromic
(Bowel cancer and internal/cutaneous lesions)
 - Muir-Torre: Lynch syndrome
 - Gardner syndrome: form of FAP (AD)

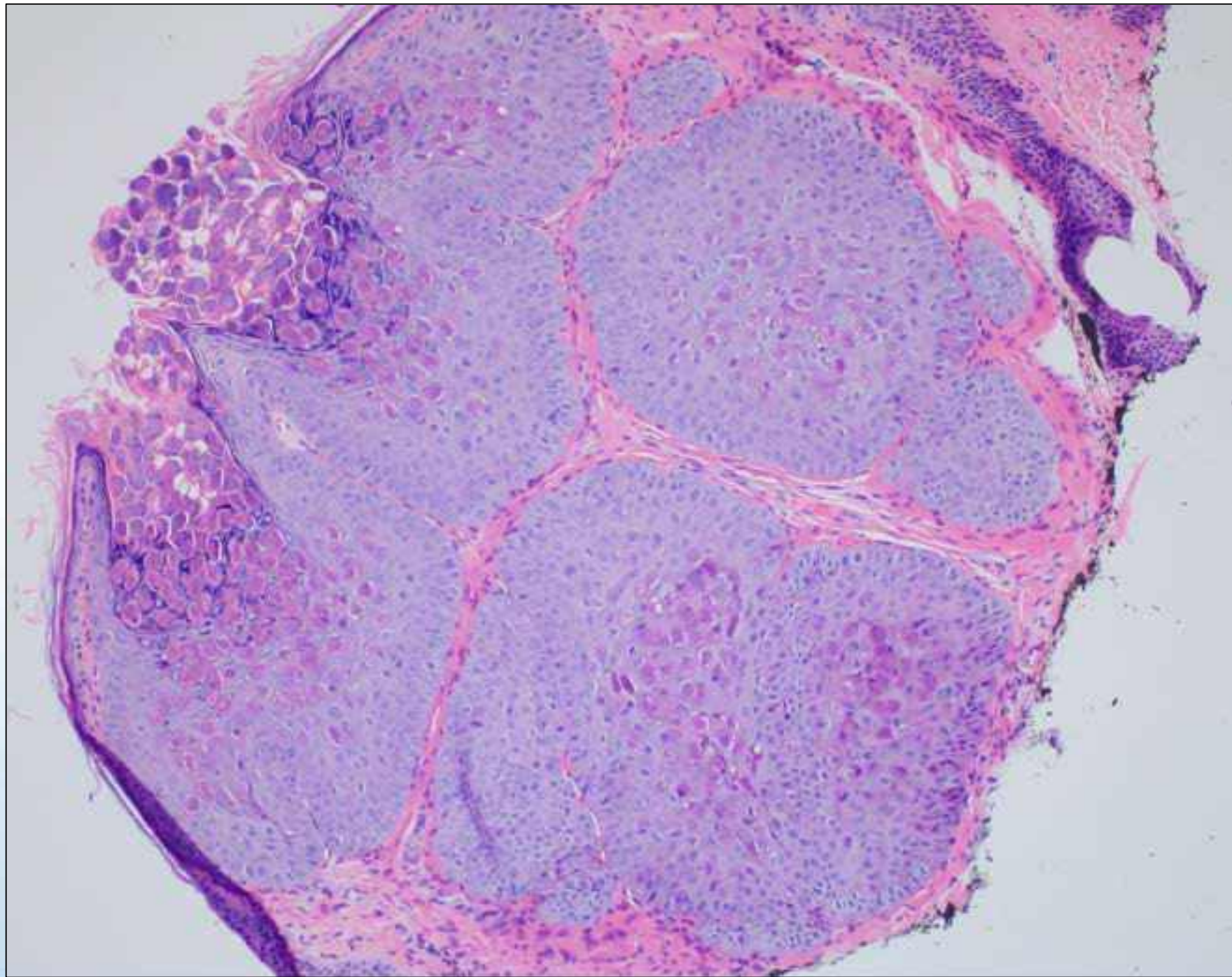


Epidermal inclusion cyst: syndromes

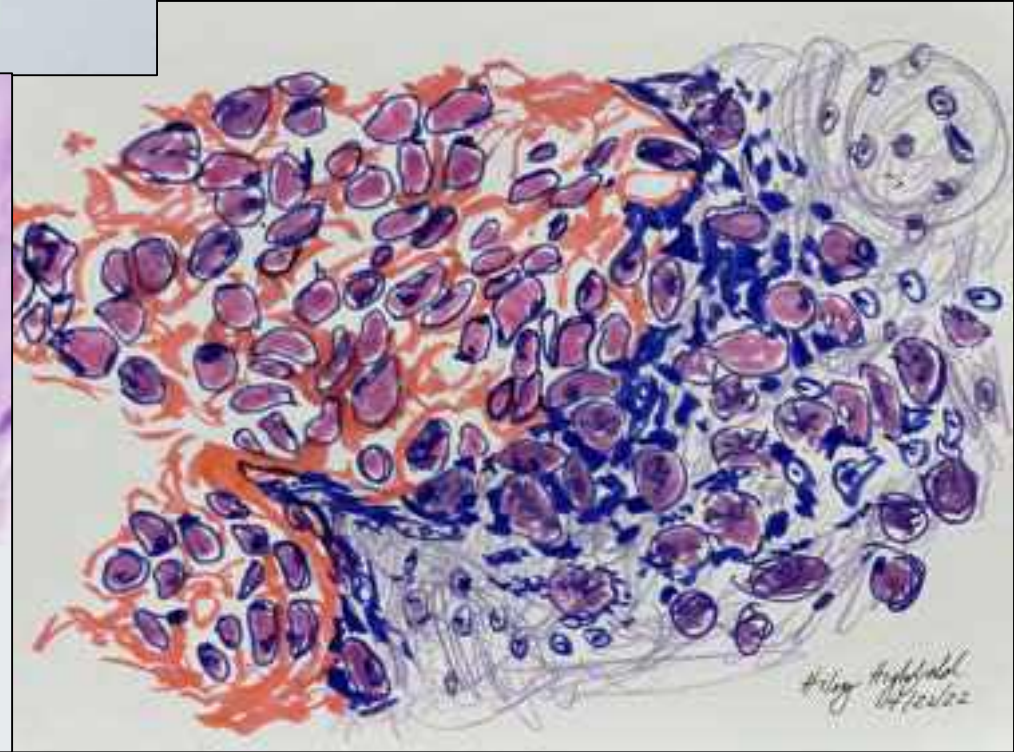
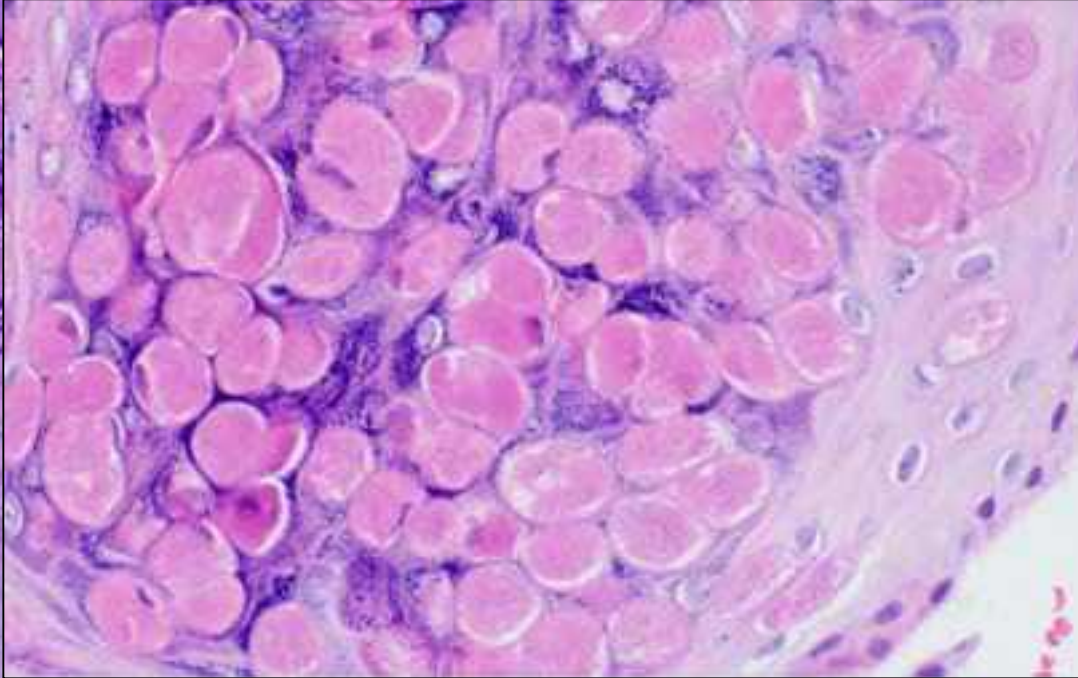
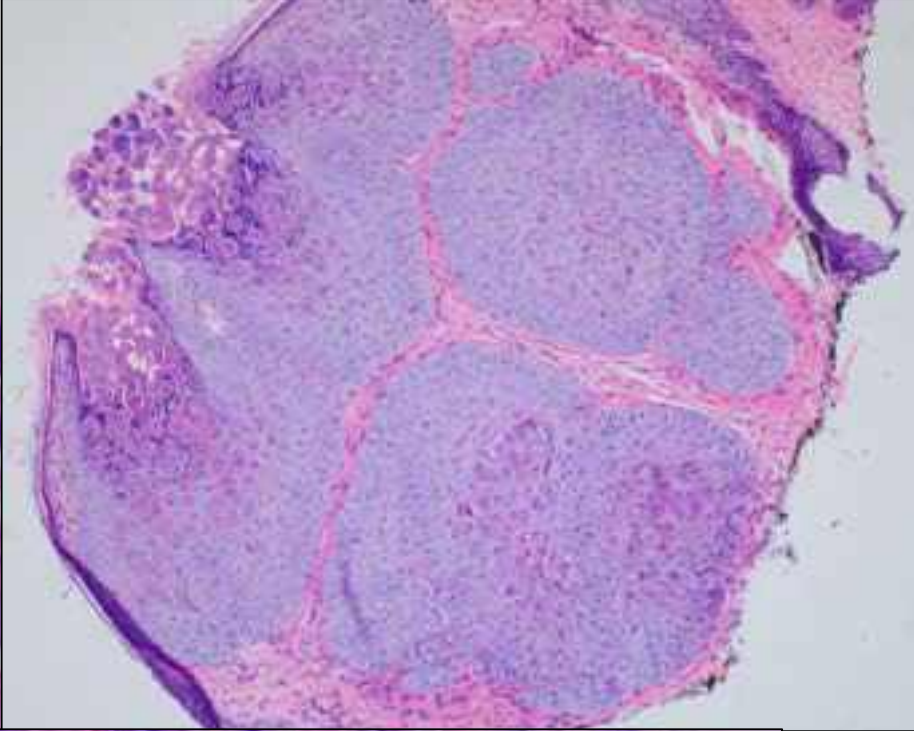
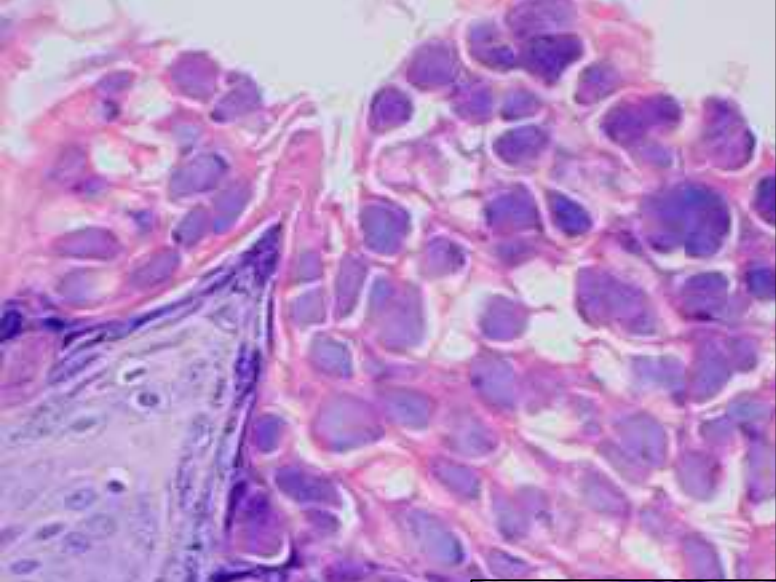
- Muir-Torre syndrome (AD):
 - Lynch syndrome: hereditary non-polyposis colorectal cancer (HNPCC)
 - Increased risk of Colorectal/GI cancer and others
 - DNA mismatch repair (*MLH1*, *MSH2*, *MSH6*, *PMS2* or *EPCAM* gene)

- Gardner syndrome (AD):
 - form of Familial Adenomatosis Polyposis (AD)
 - Colorectal polyps, tumors, dental, osteomas, fibromas, lipomas
 - Tumors: colorectal, thyroid, liver, bile ducts, adrenal
 - APC gene: tumor suppressor gene cell growth/division



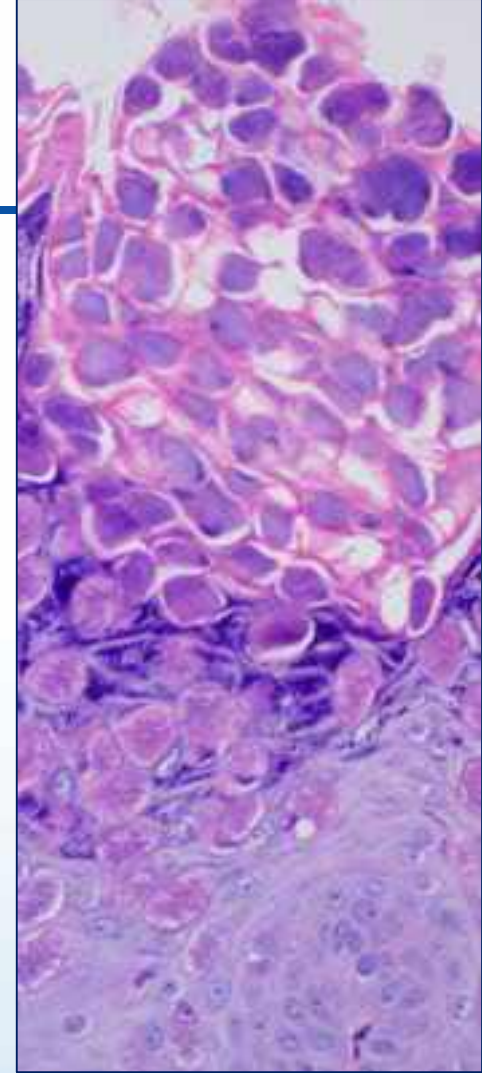


Molluscum



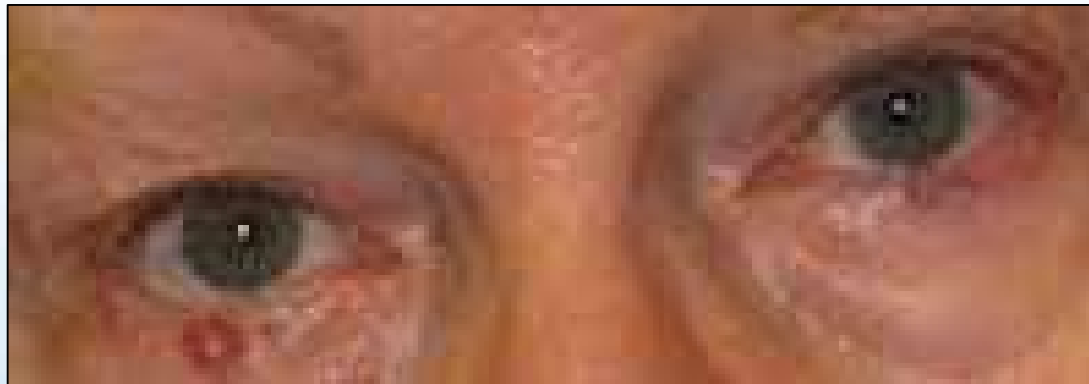
MOLLUSCUM CONTAGIOSUM

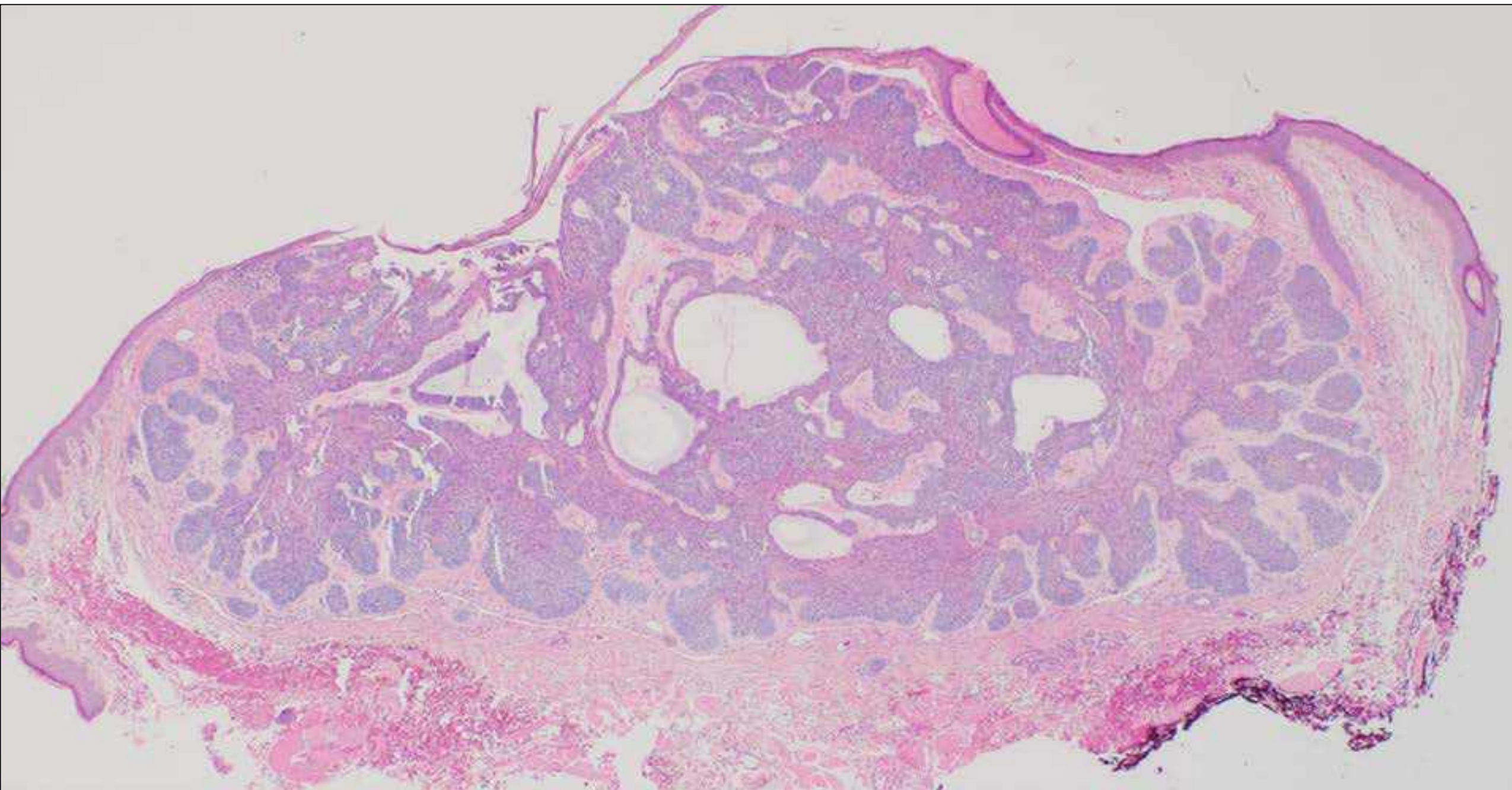
- Pox virus
- Elevated smooth nodule with central umbilication
- Crateriform shape of acanthotic epithelium
- Large intracytoplasmic inclusion bodies:
Henderson-Patterson corpscules
- Inclusions become more basophilic as they mature

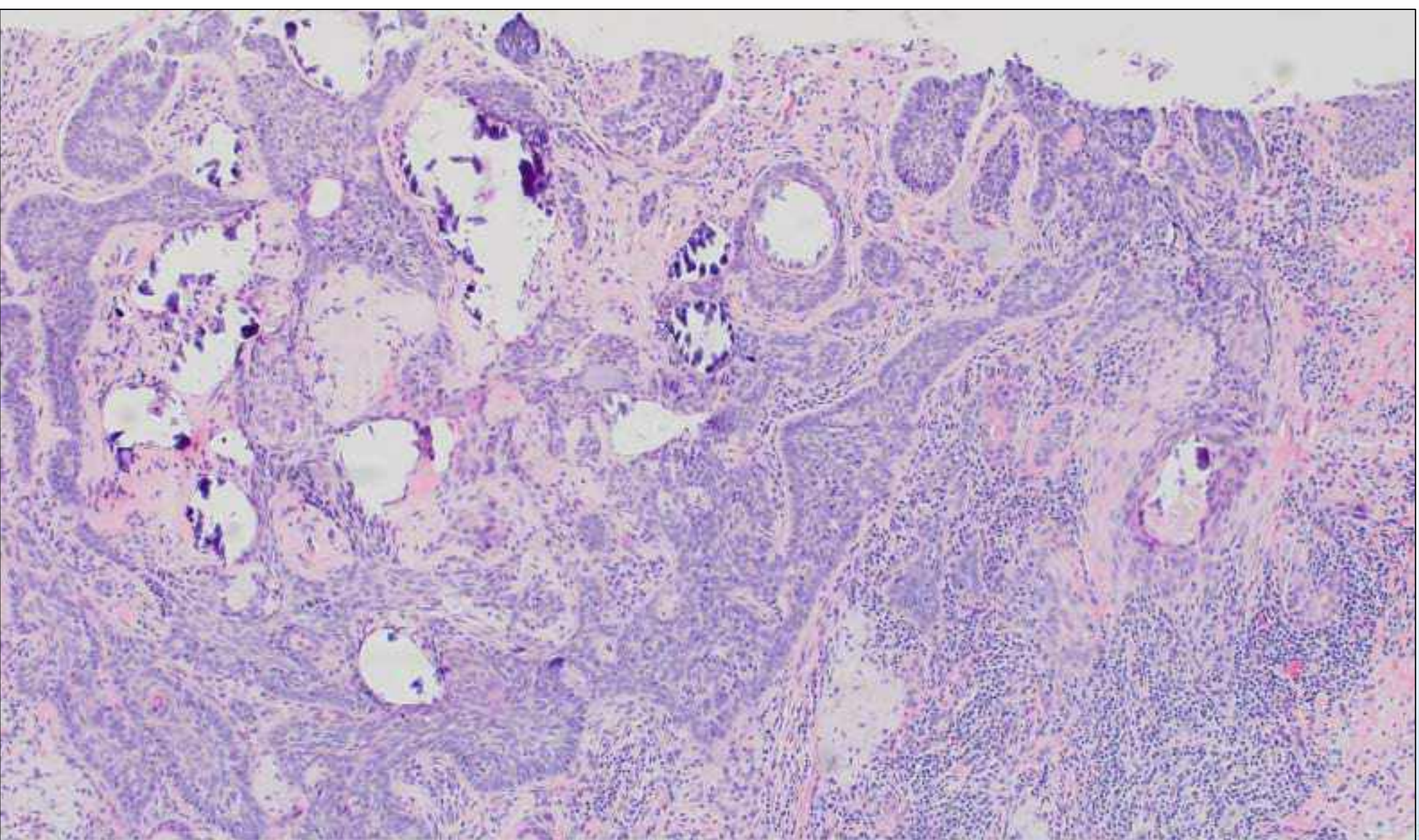


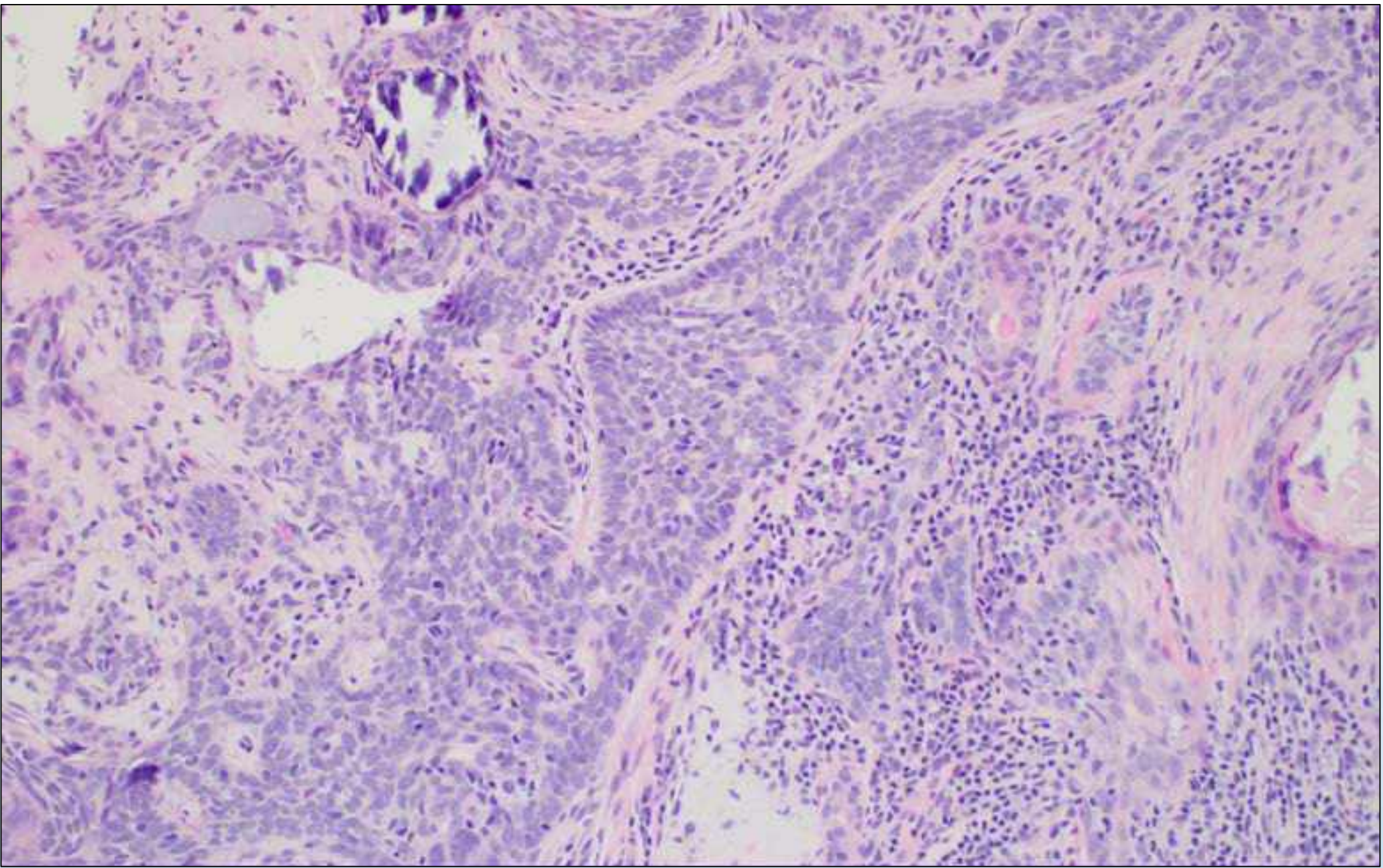


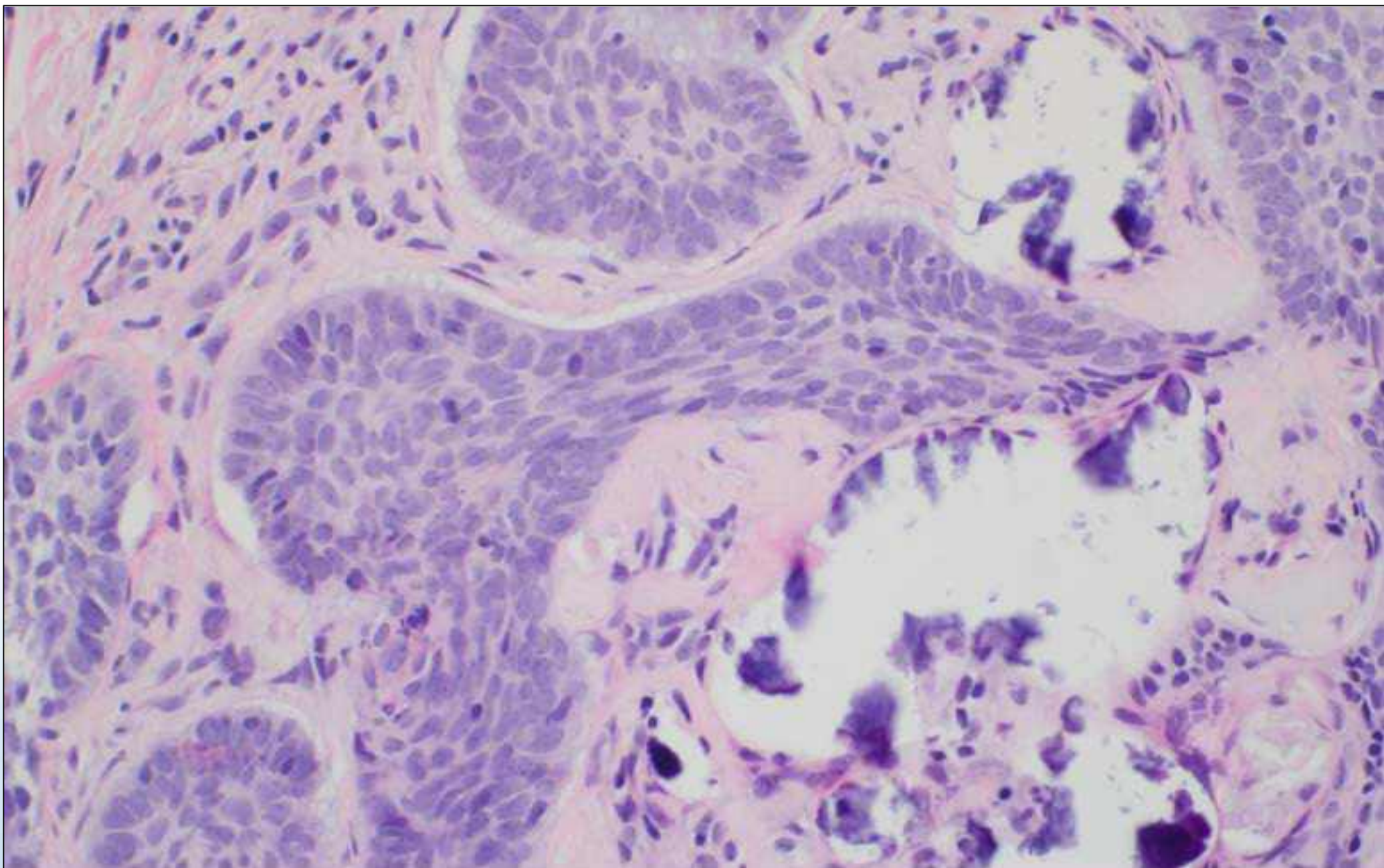
BASAL CELL CARCINOMA

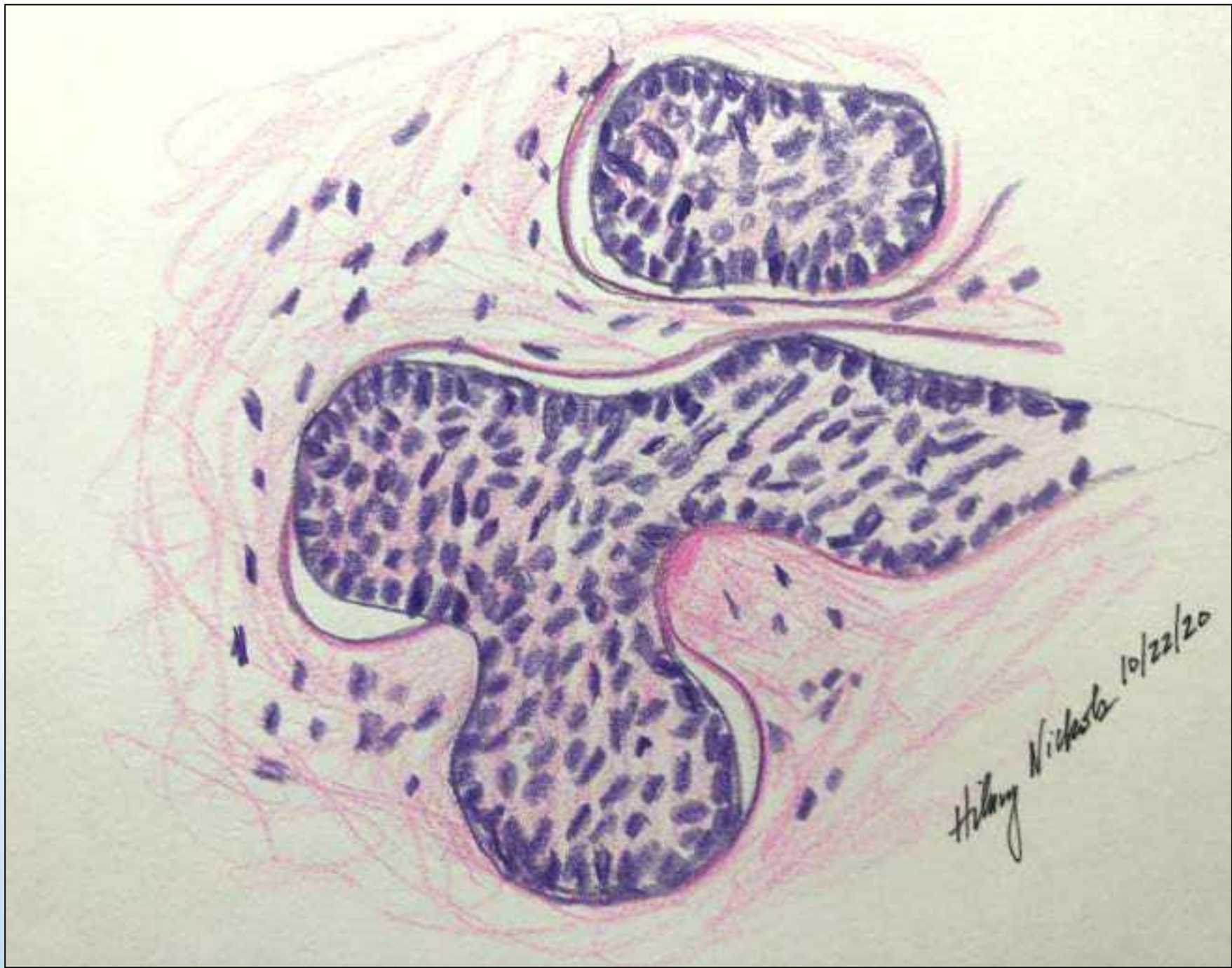












Hilary Nicholas 10/22/20

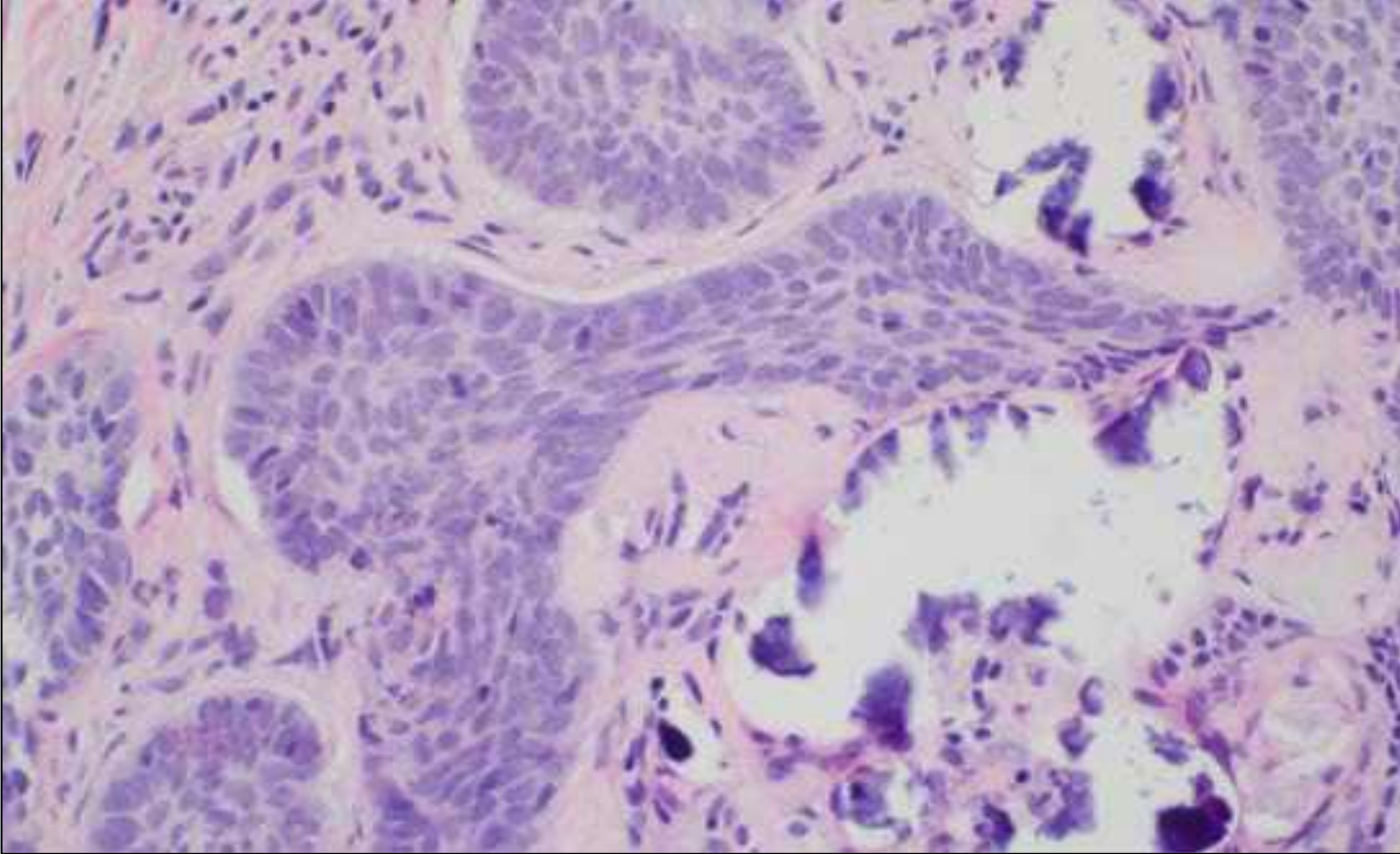
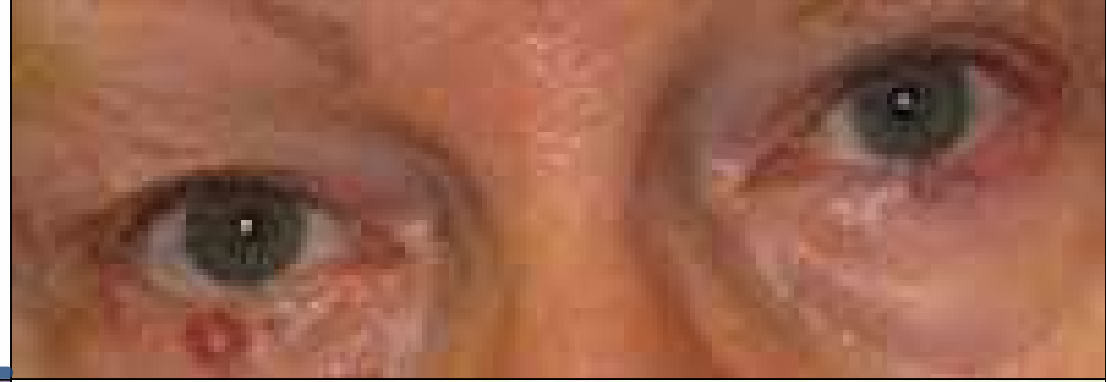


BASAL CELL CARCINOMA

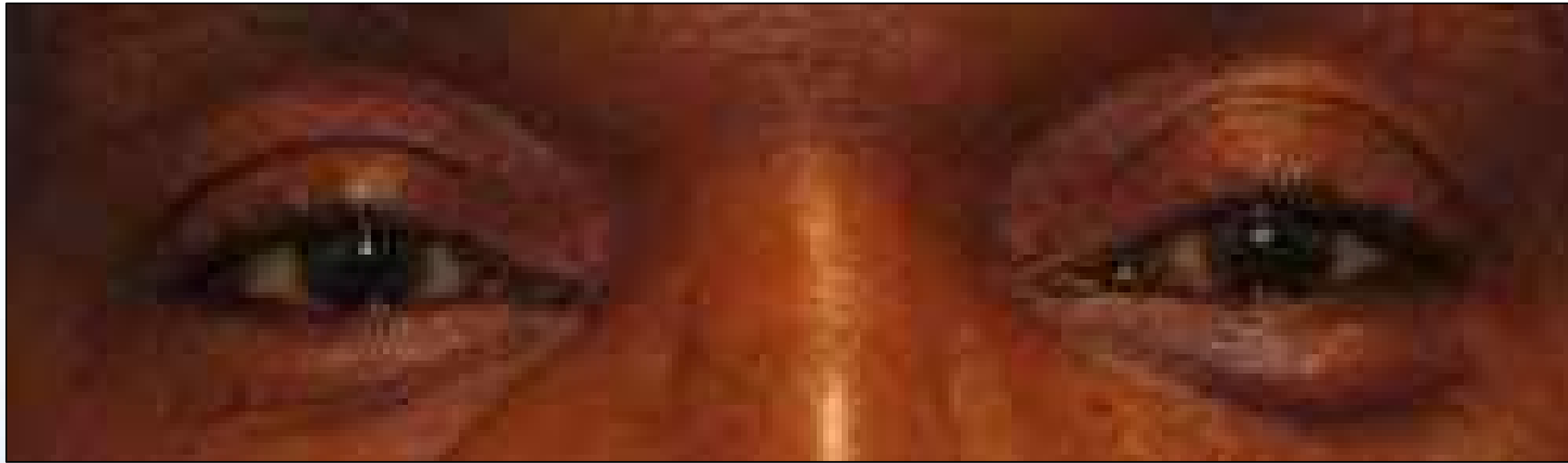
- Most common malignant tumor periocular skin
- Incidence highest in fair-skinned adults (sun exposed)
- Lower > inner canthus > upper > outer canthus
- Basaloid nests with peripheral palisading

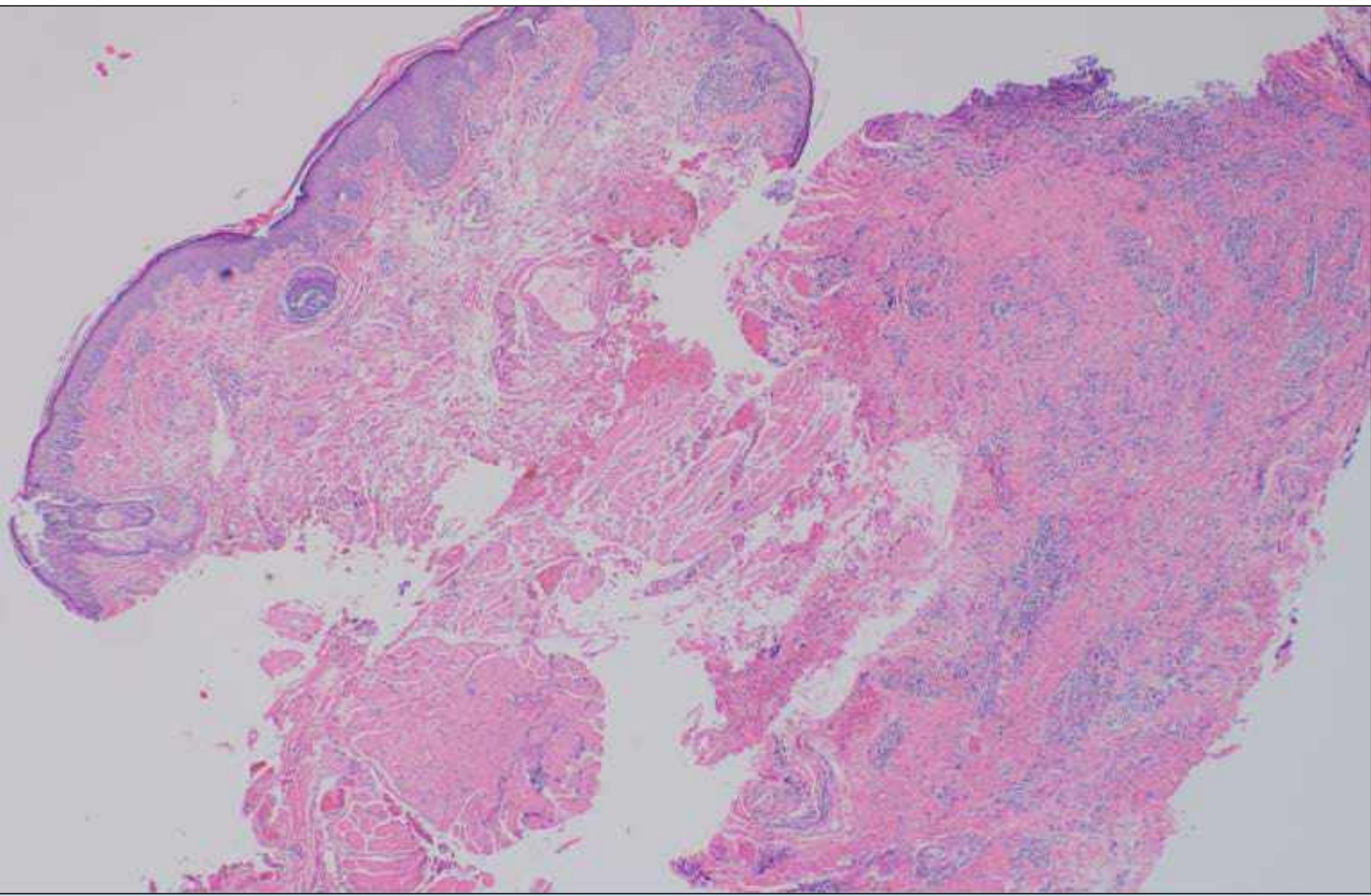


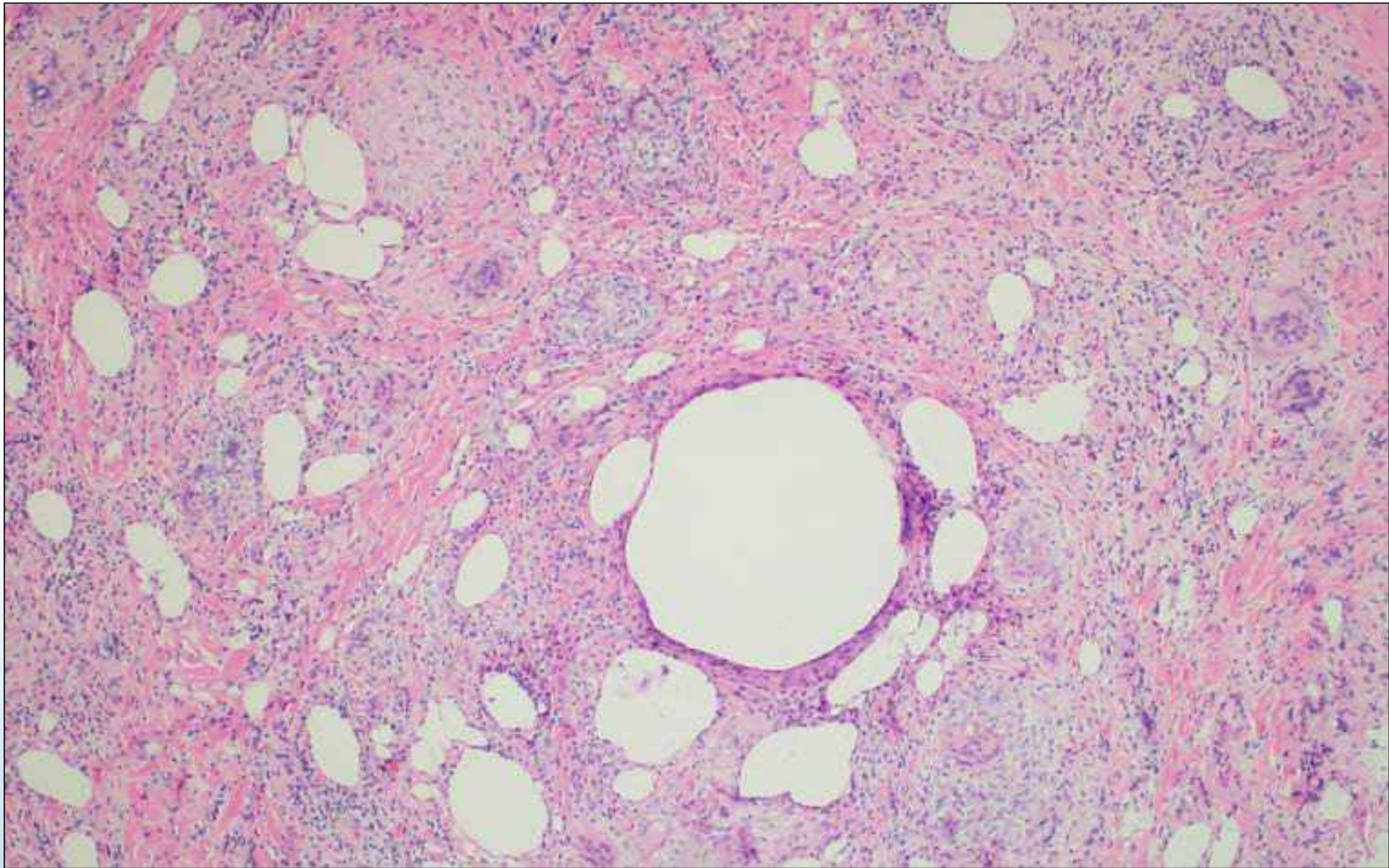
BASAL CELL CARCINOMA

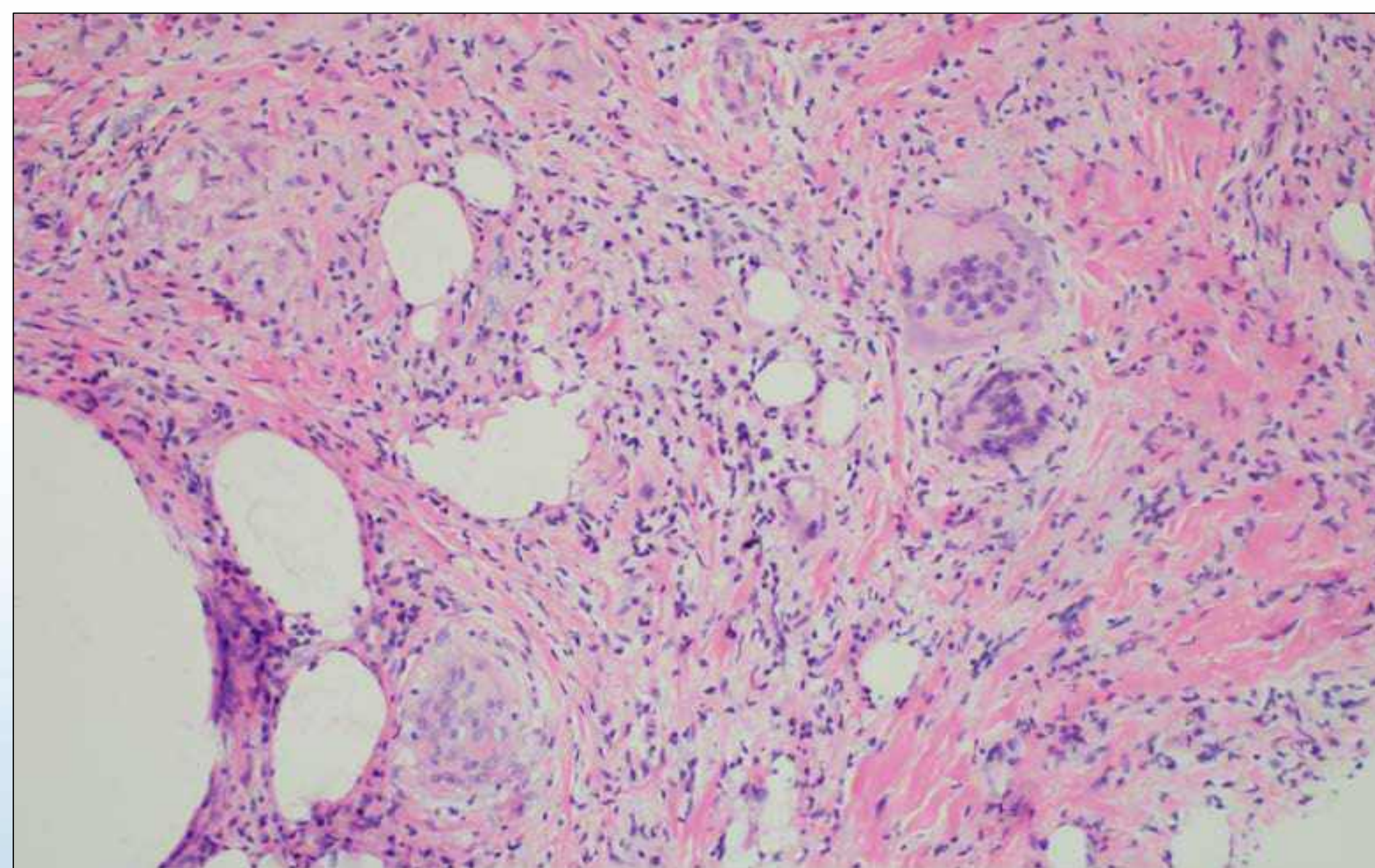


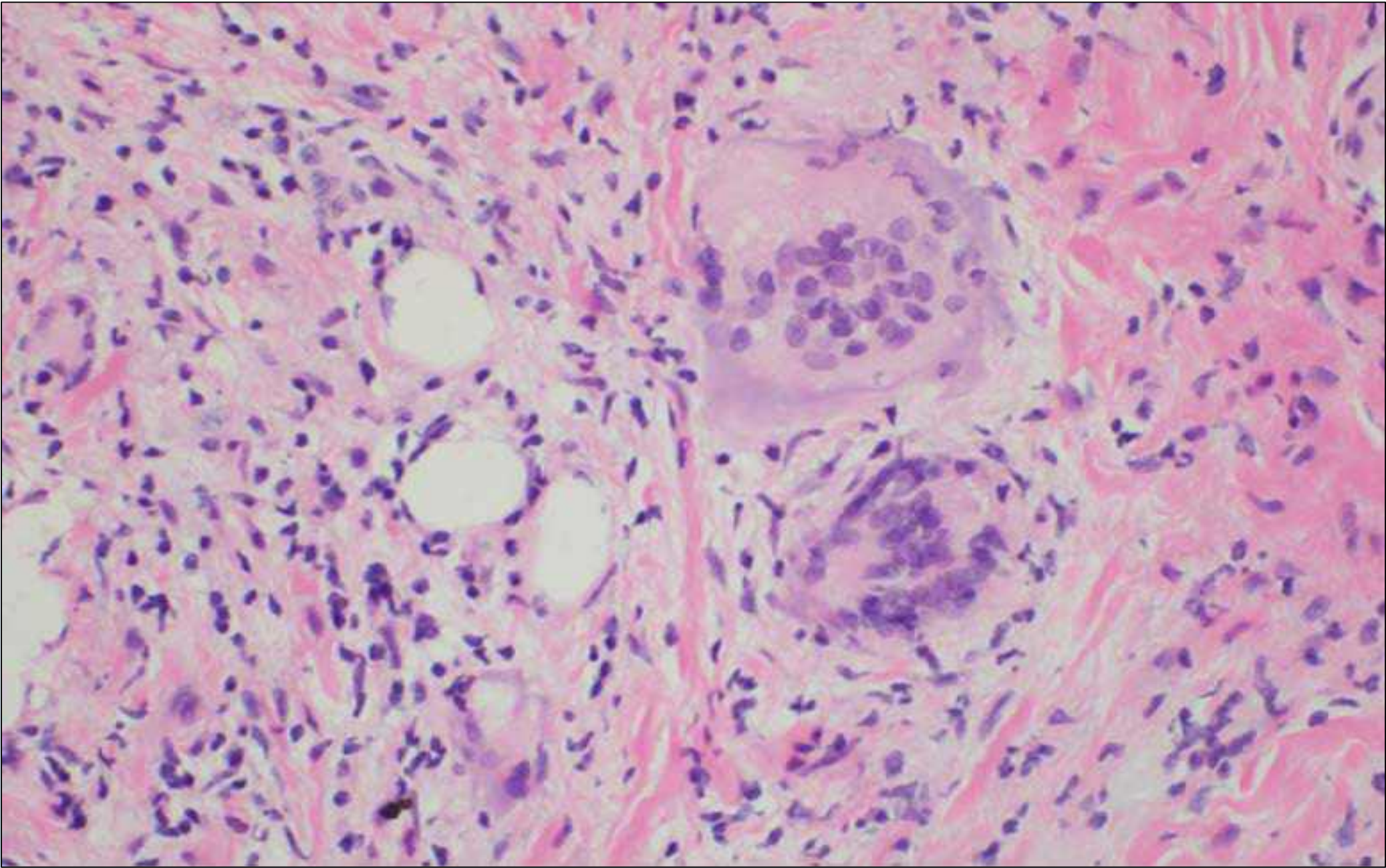
CHALAZION

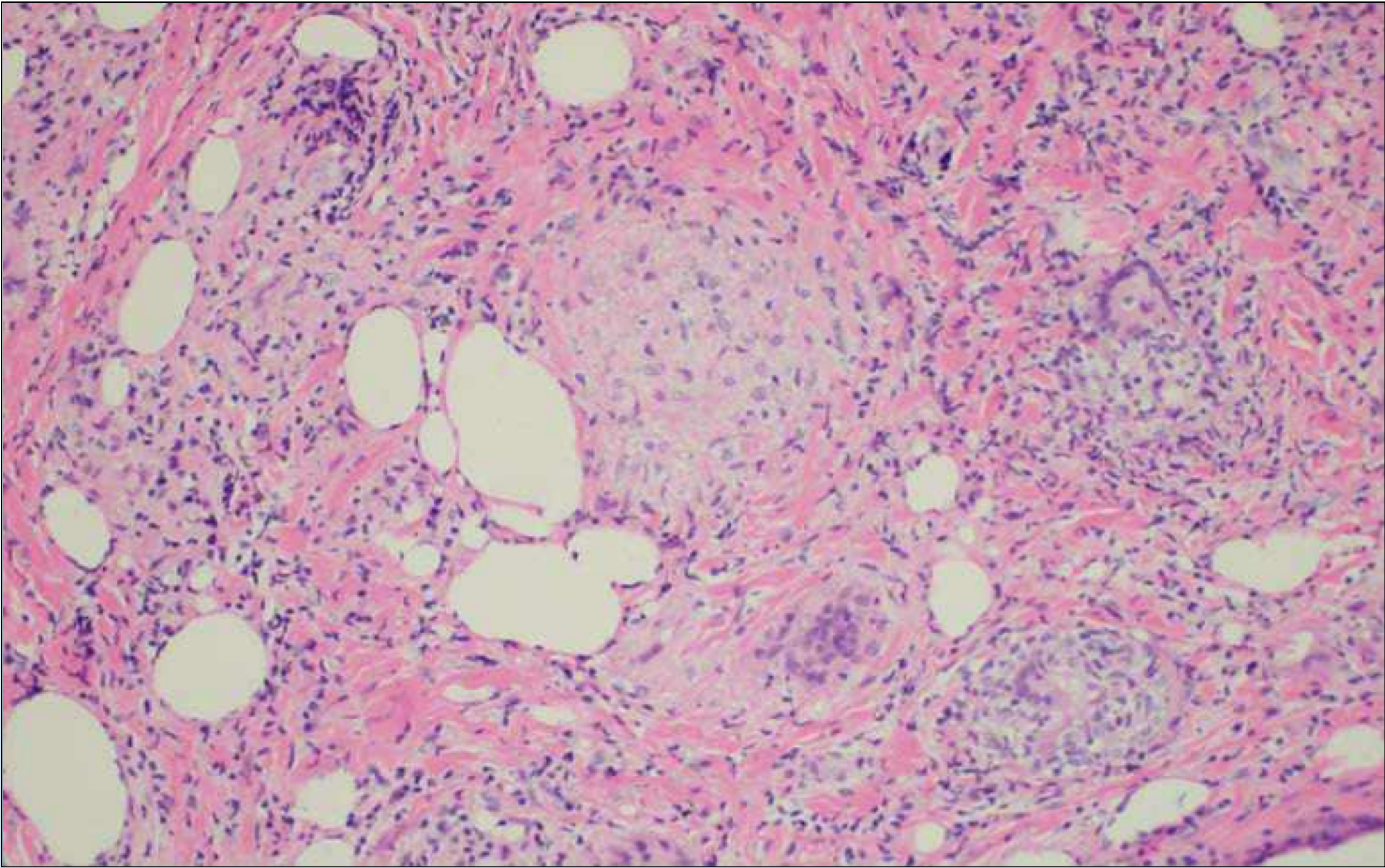


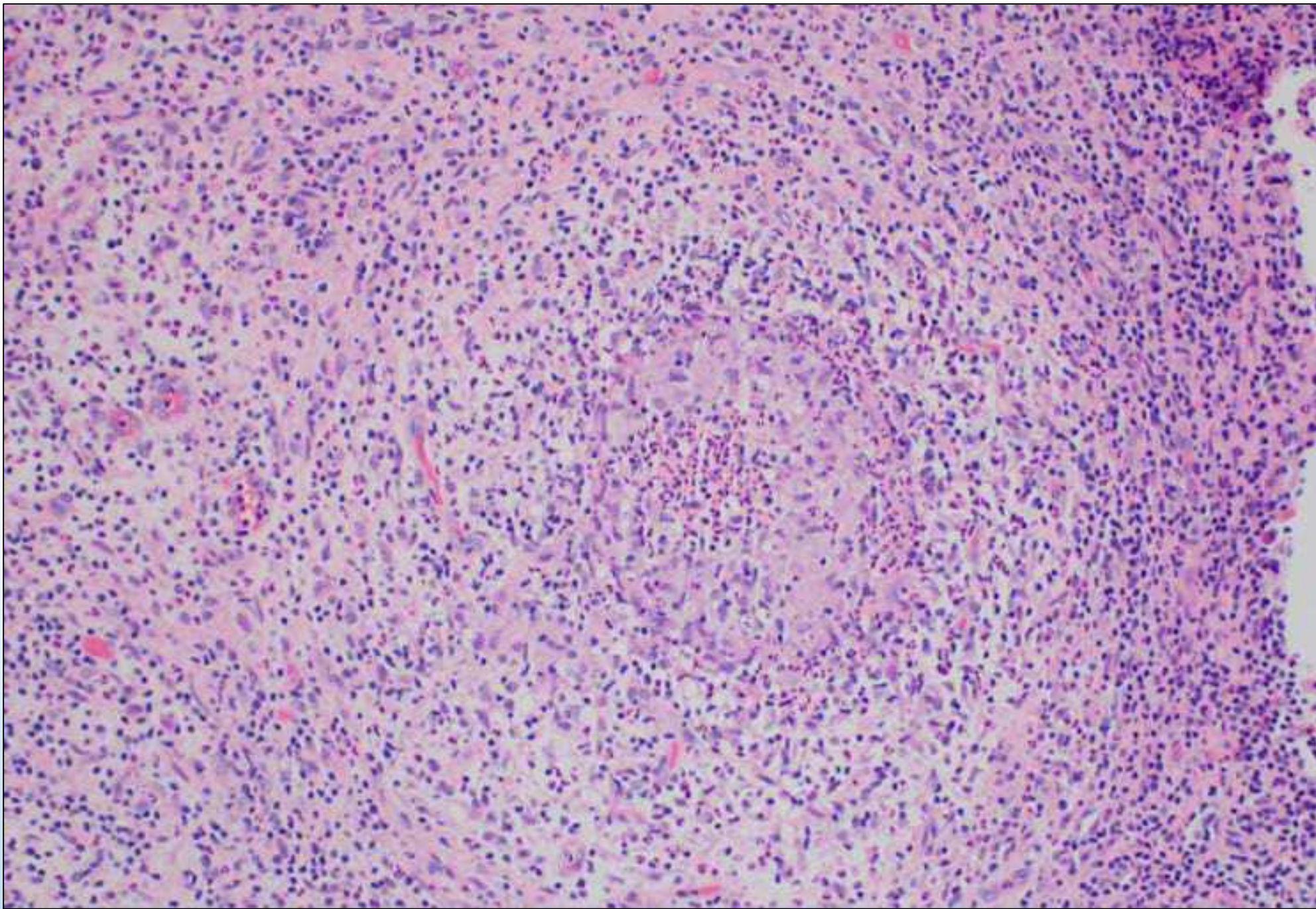


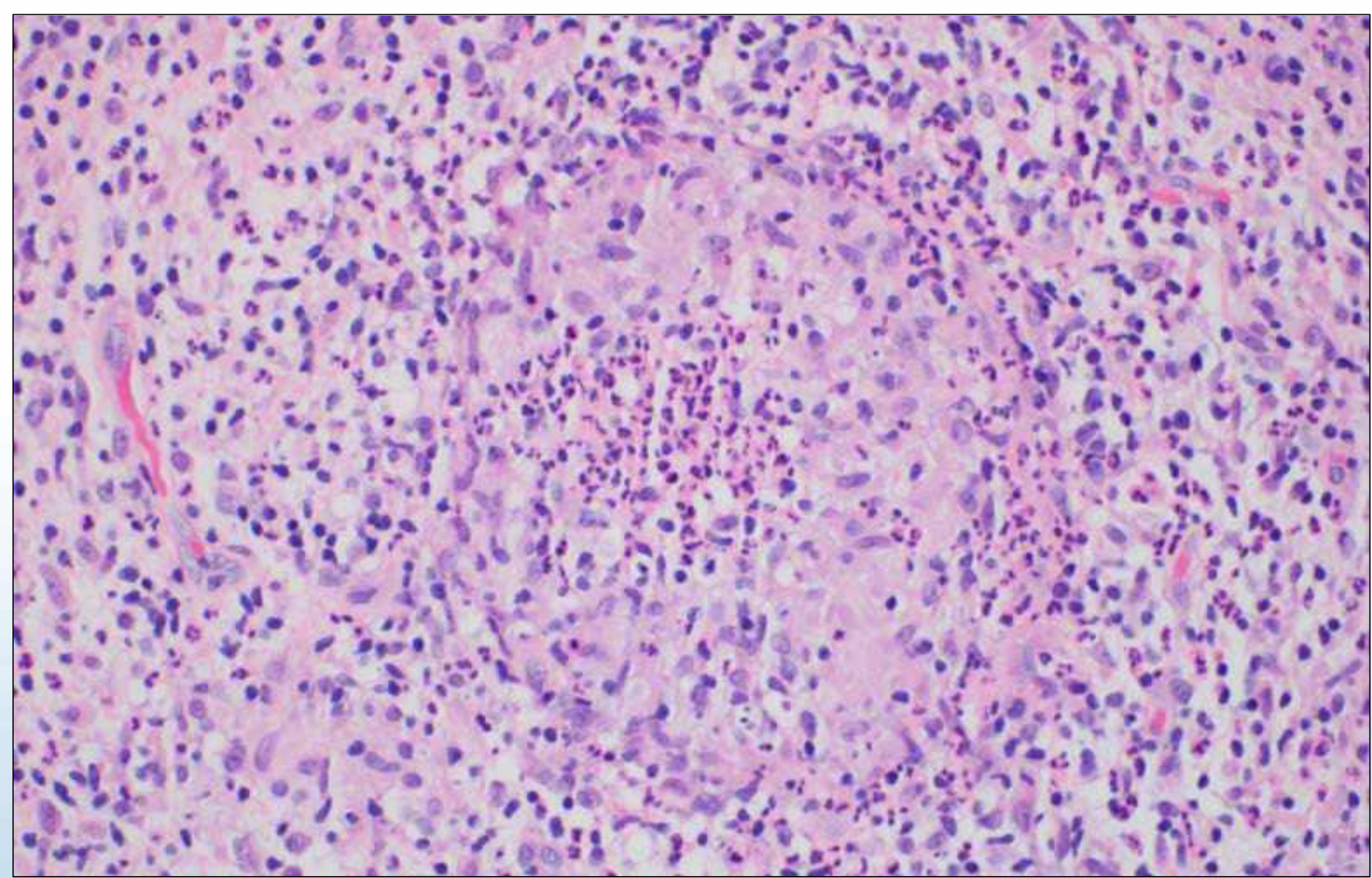


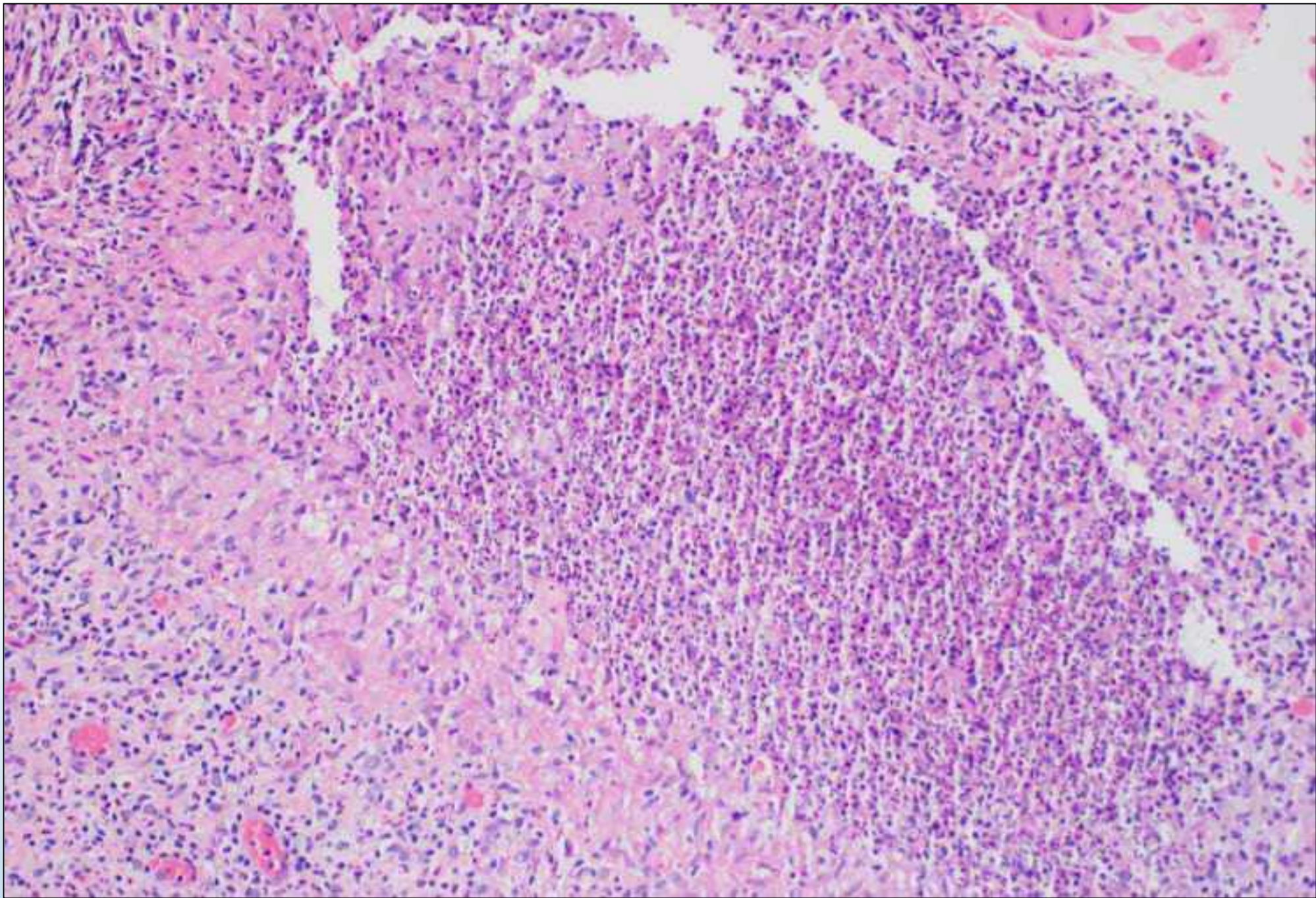


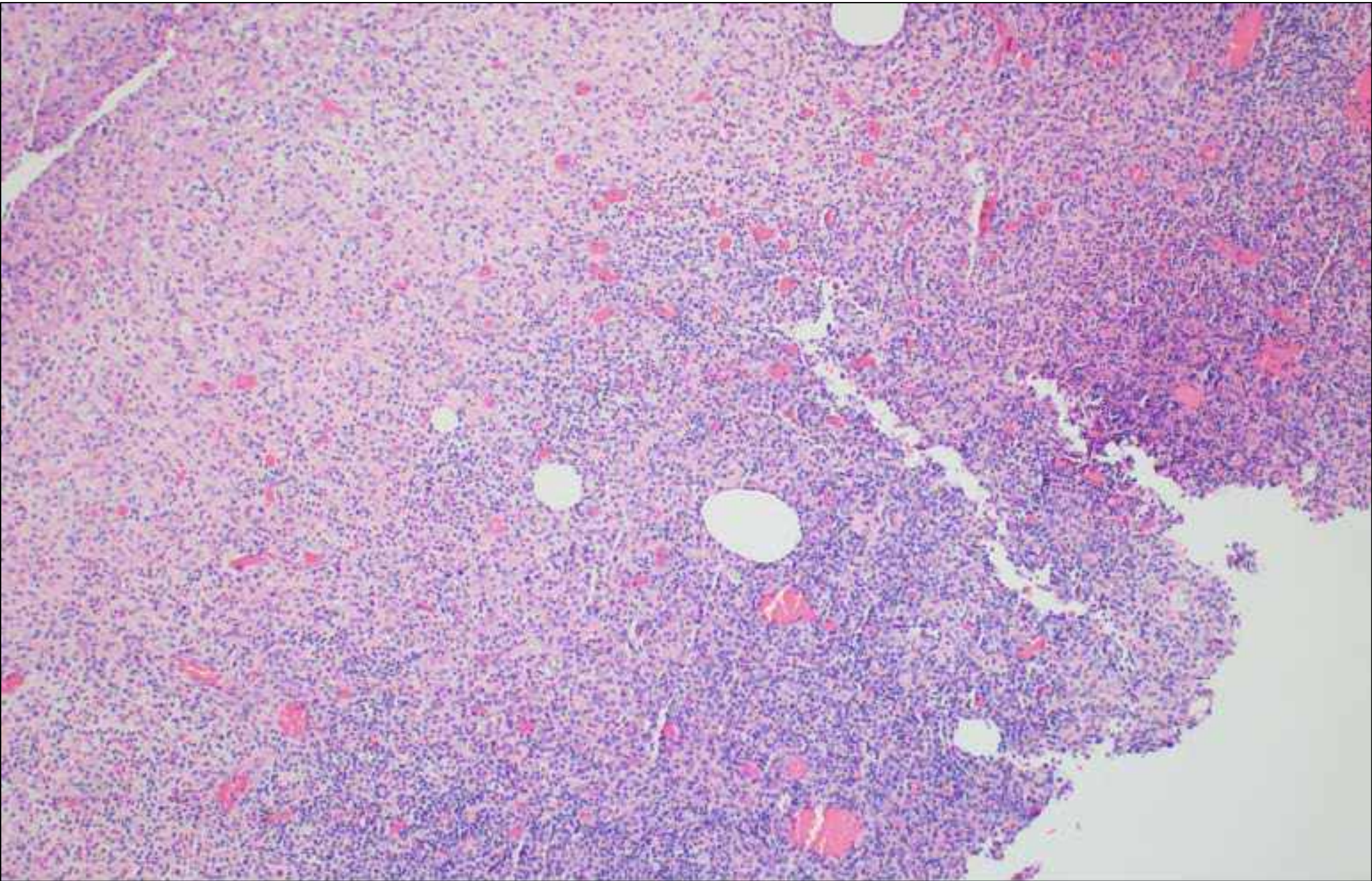


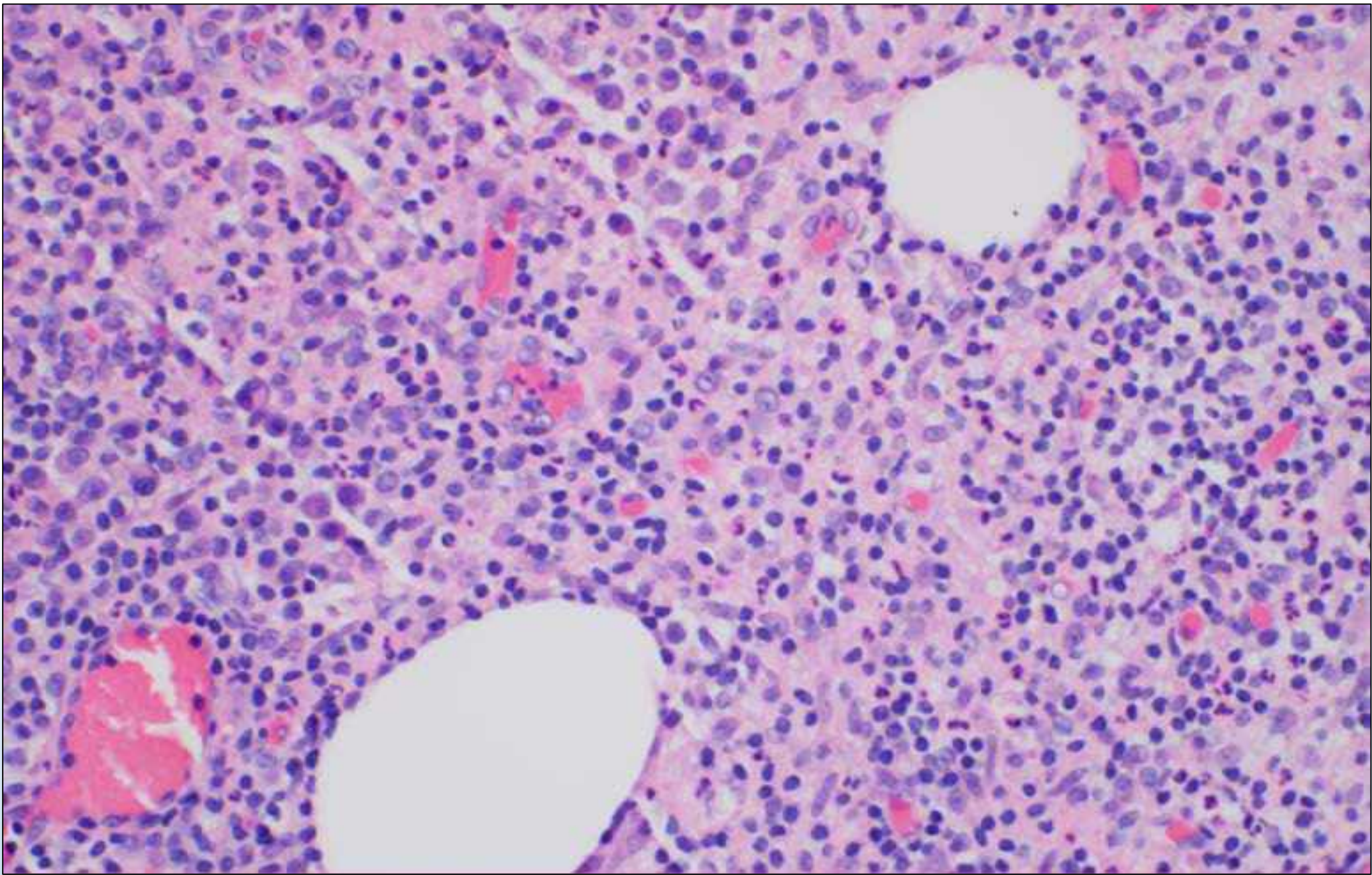


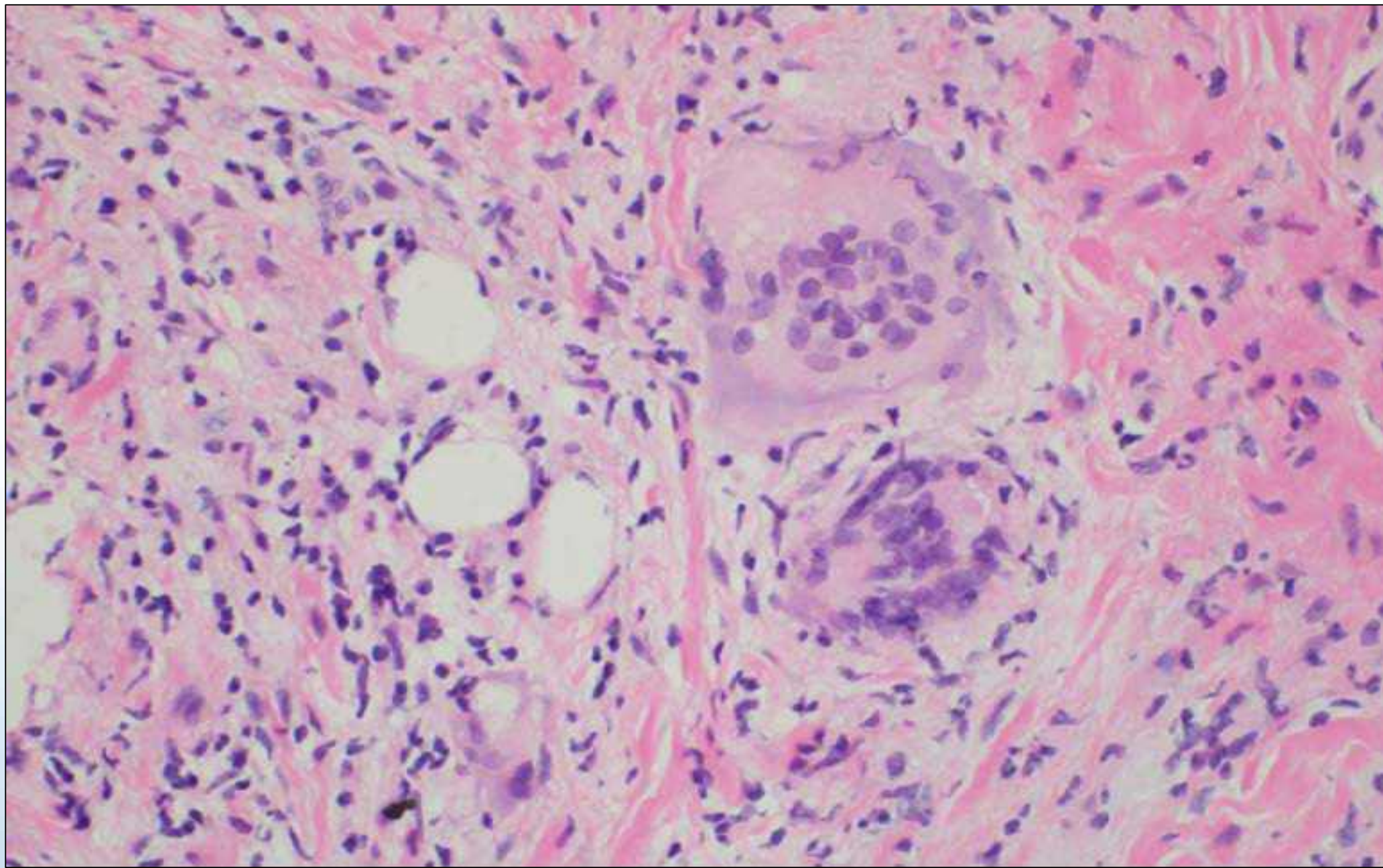






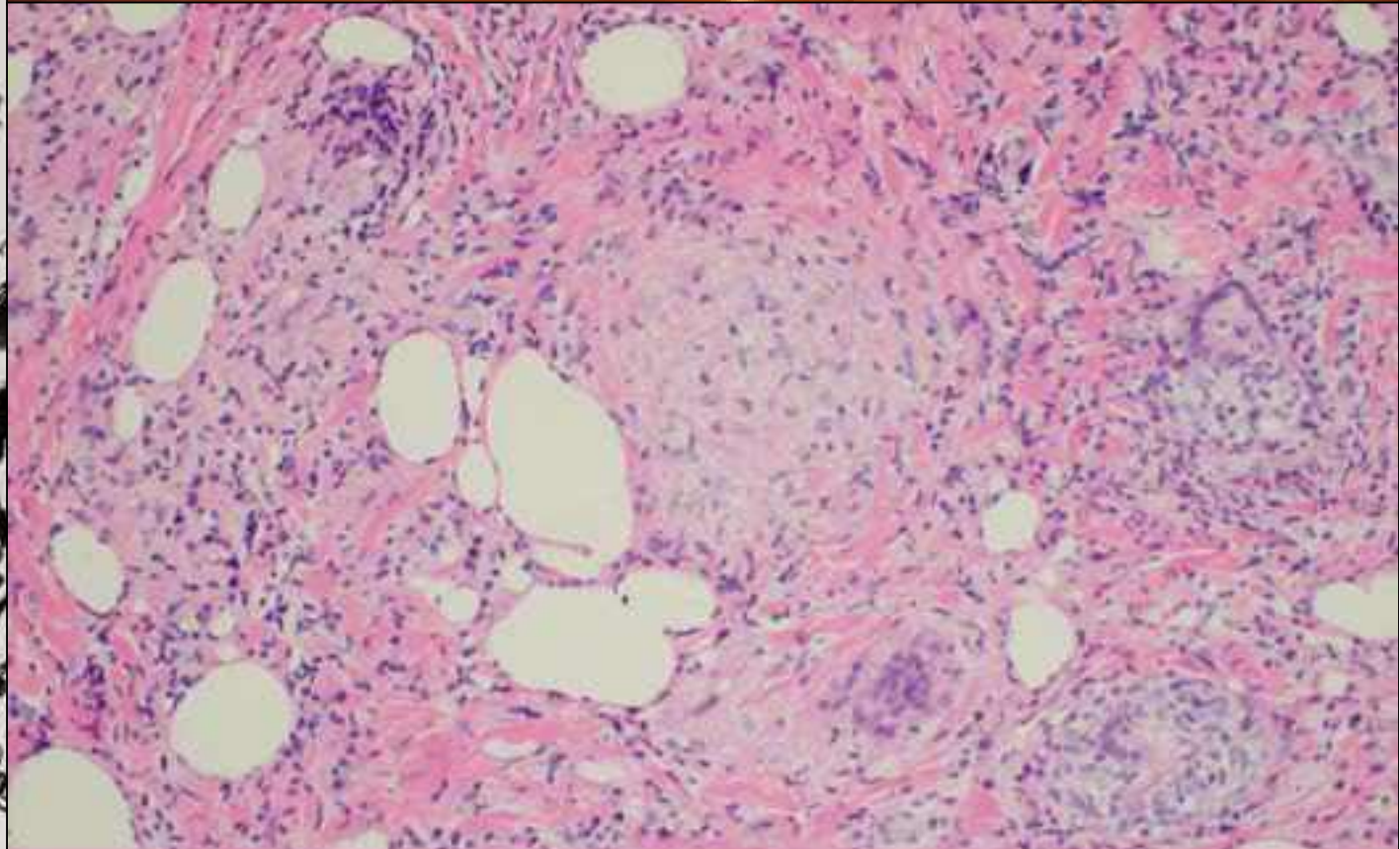








CHALAZION



CHALAZION

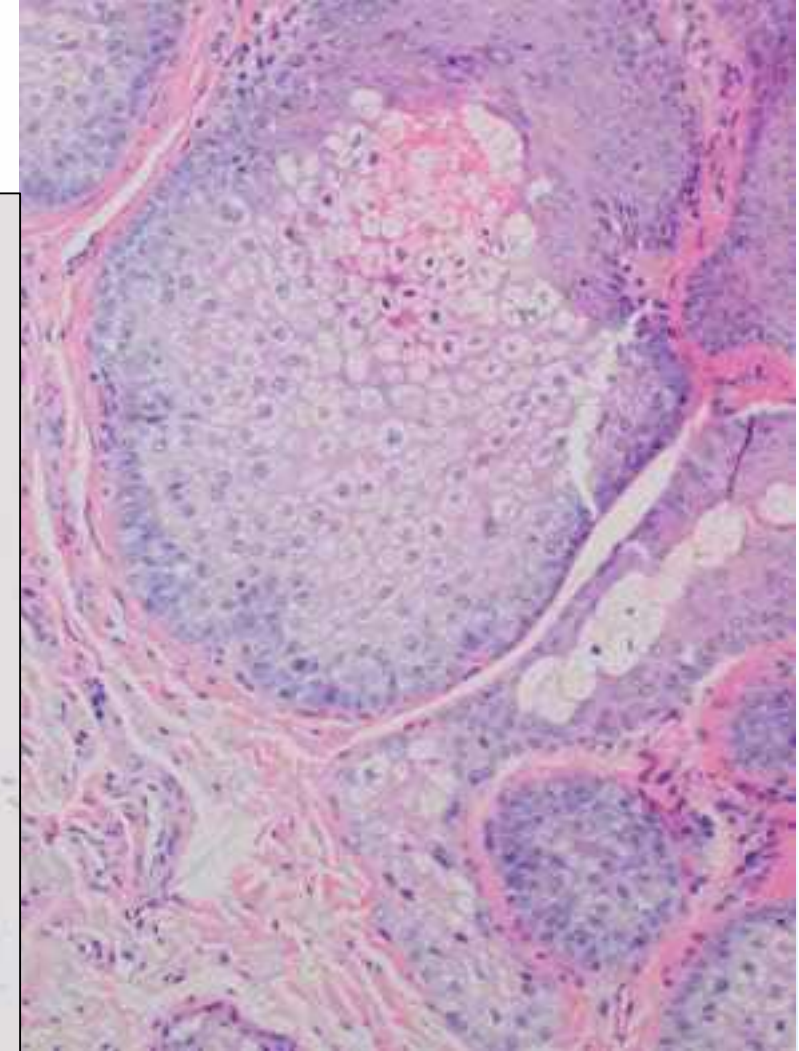
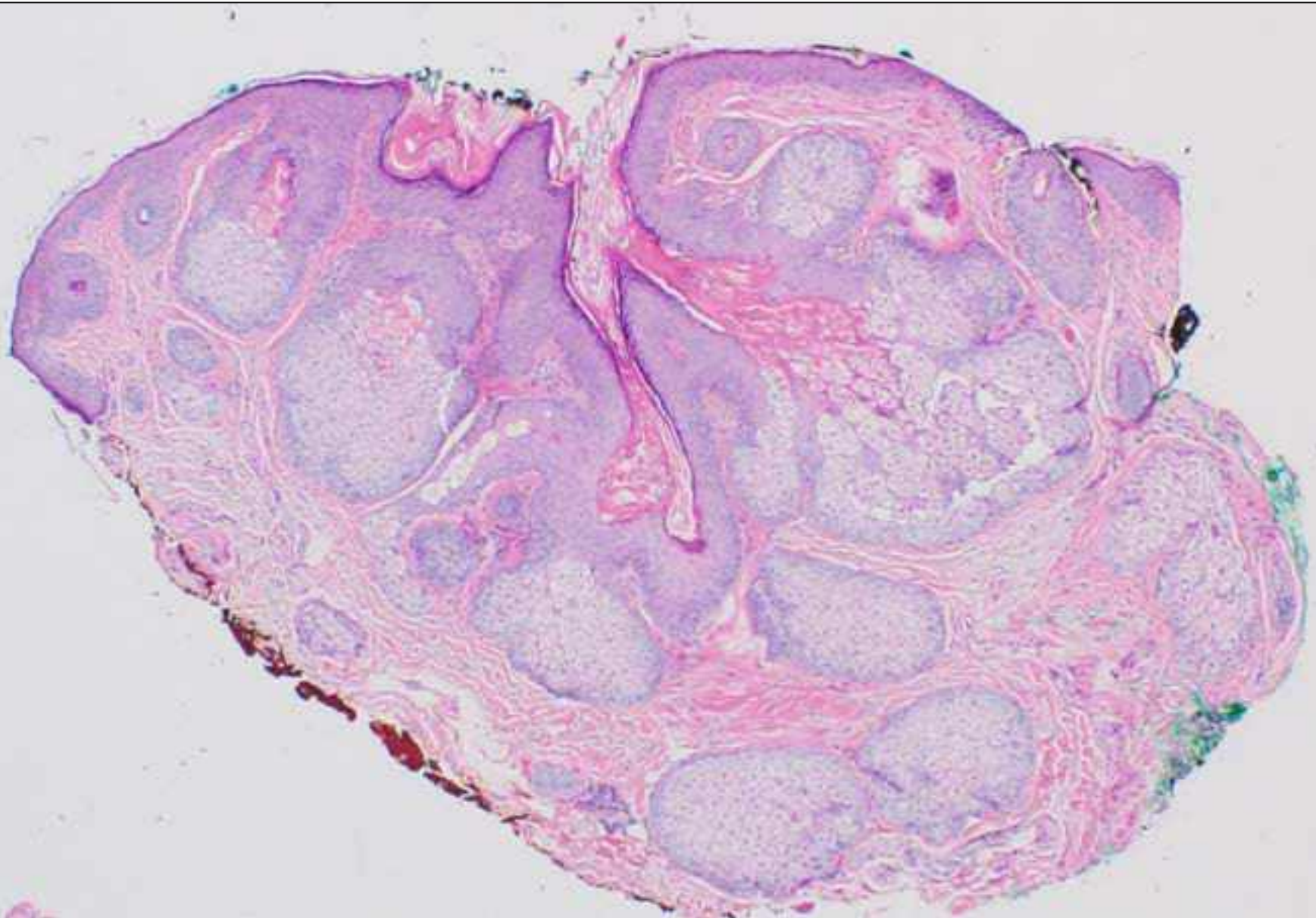
- Nodule(s) on eyelid
- Chronic lipogranuloma
 - Endogenous foreign body reaction to lipid-rich secretions of Meibomian/Zeiss glands
- Epithelioid histiocytes/giant cells surrounding empty spaces (lipid vacuoles), lymphocytes and plasma cells
- DDX: Sebaceous ca, Merkel cell ca, metastatic ca



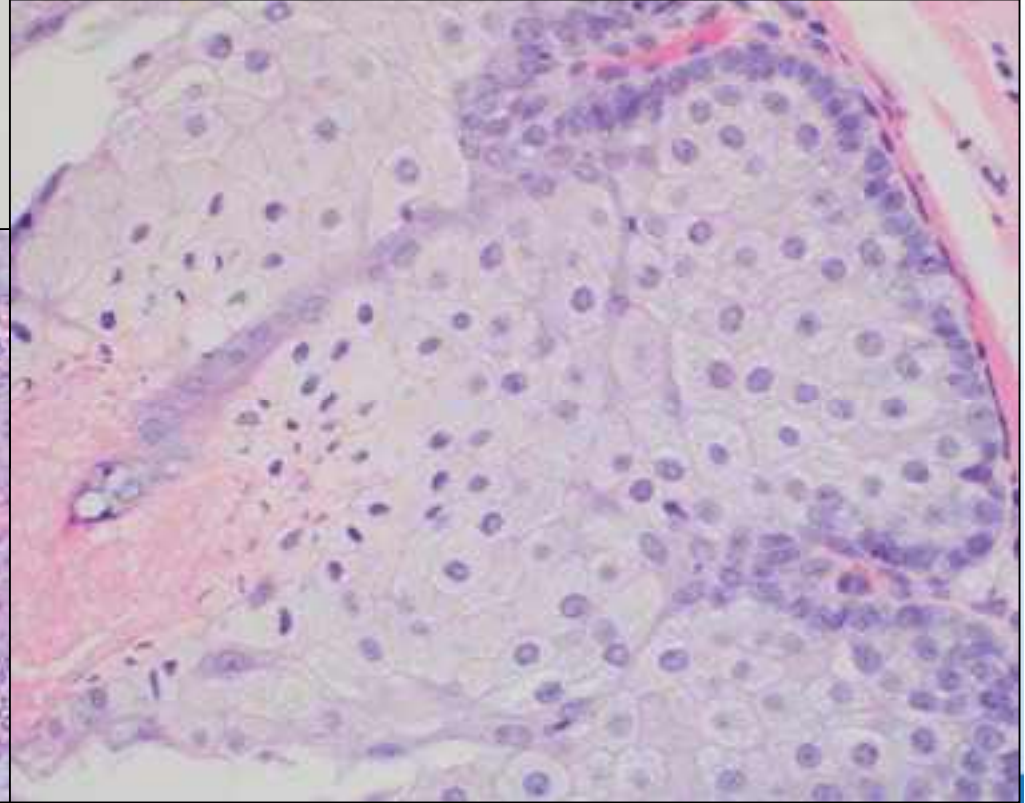
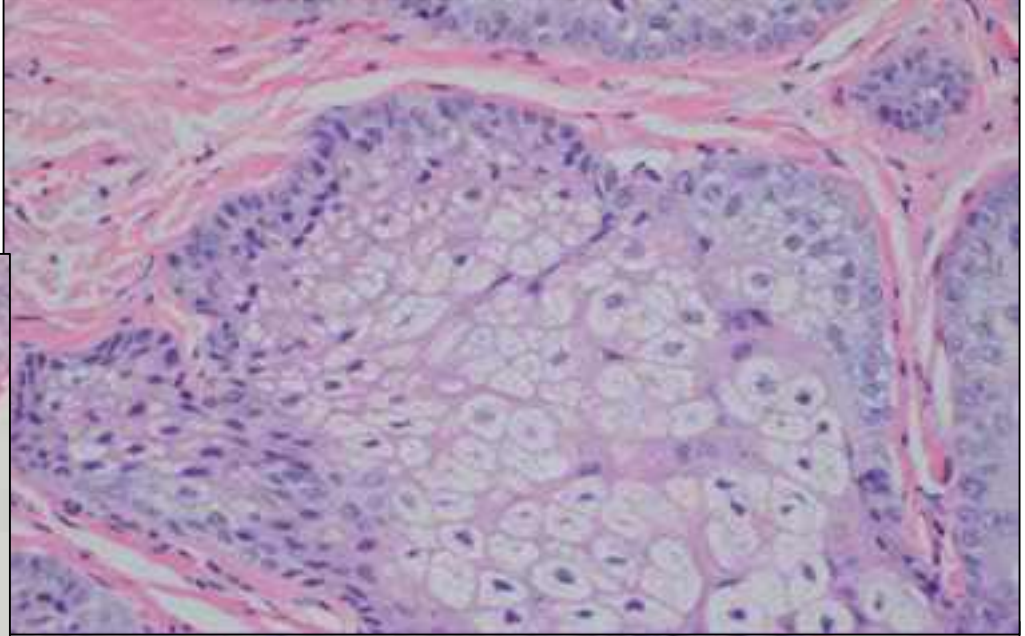
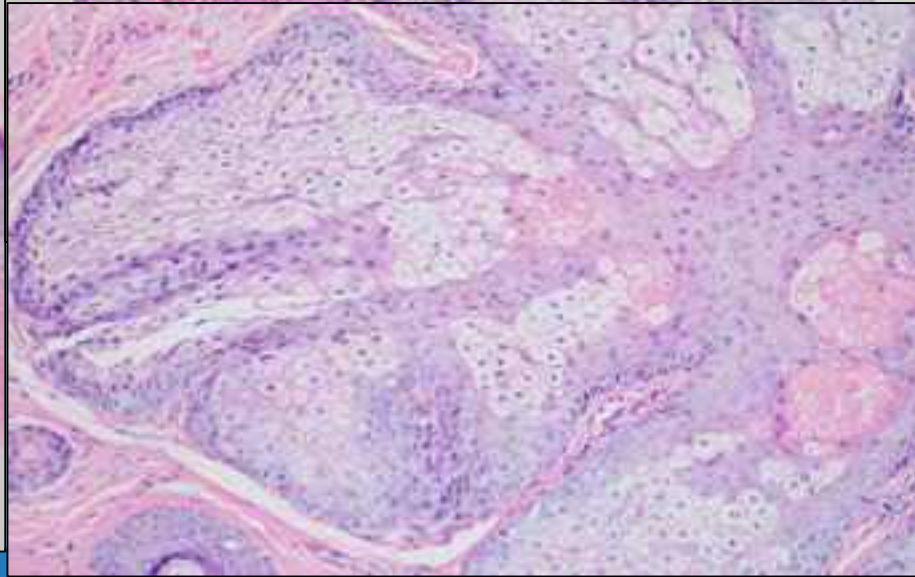
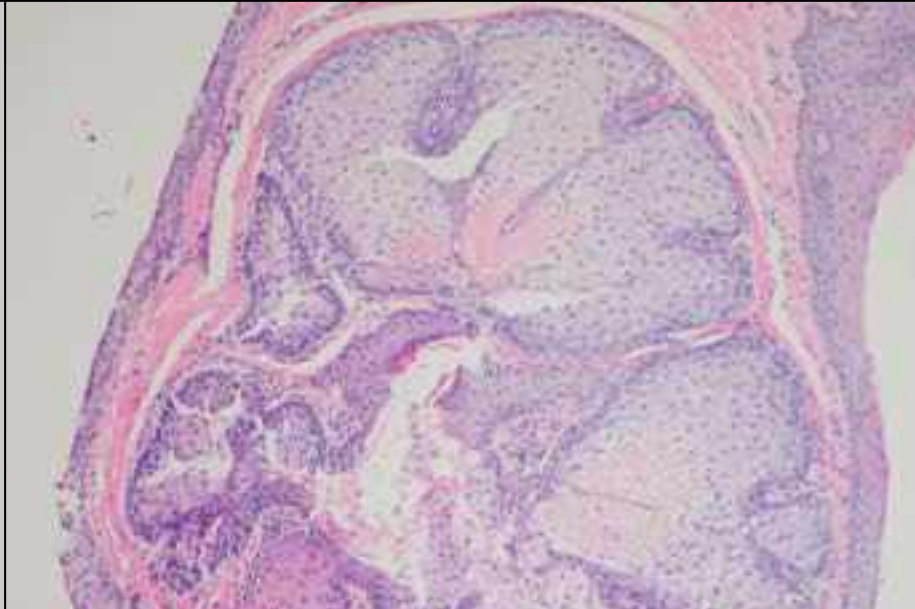
SEBACEOUS LESIONS



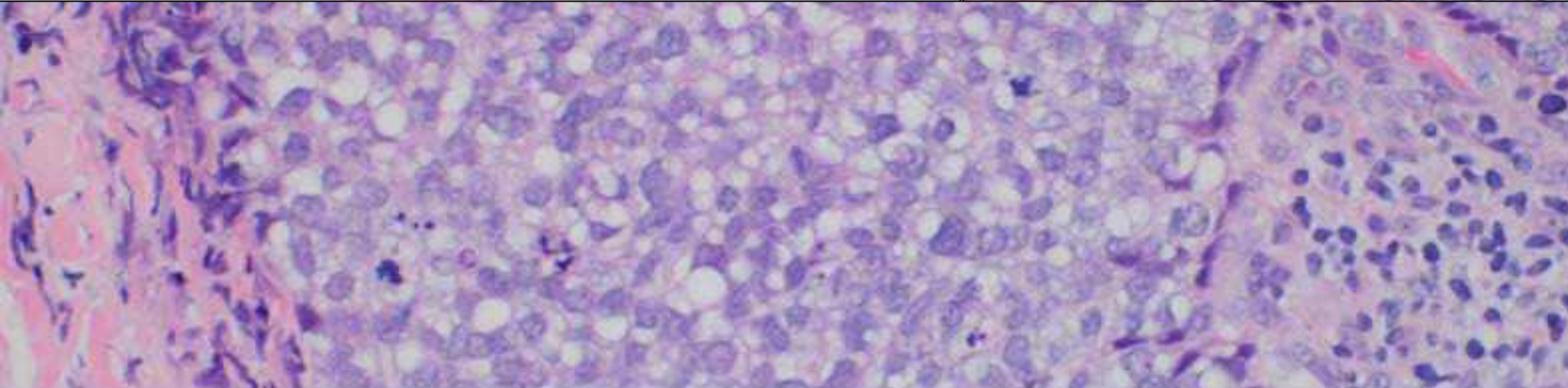
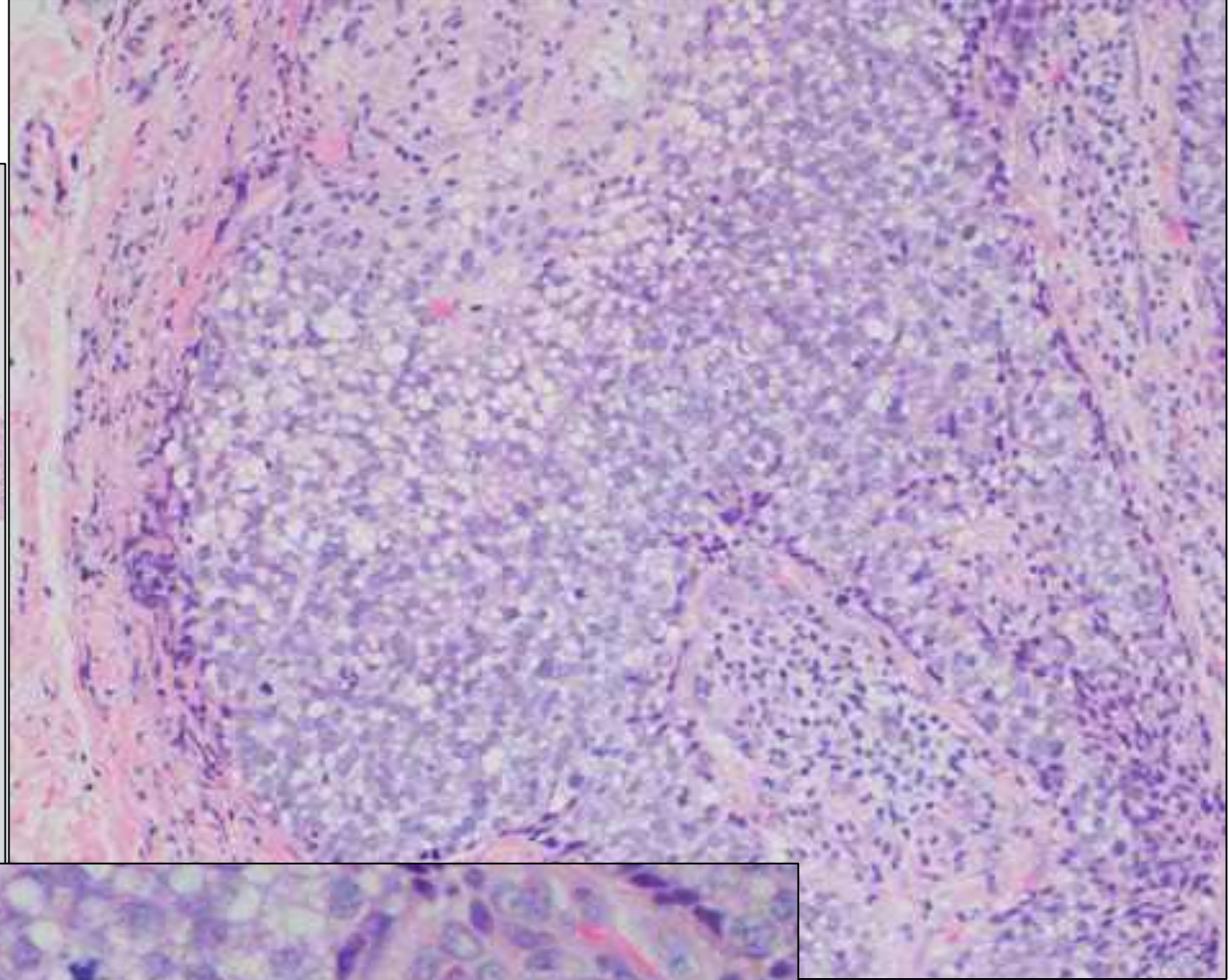
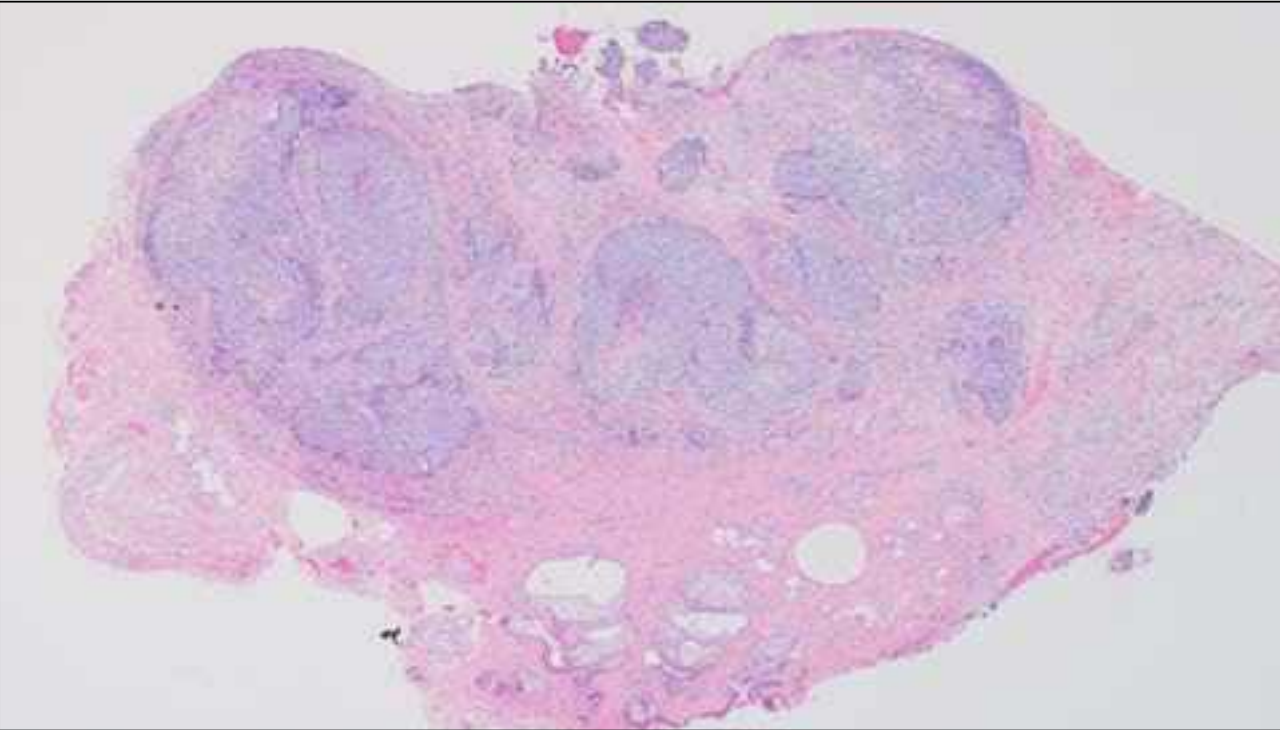
Sebaceous hyperplasia



Sebaceous adenoma



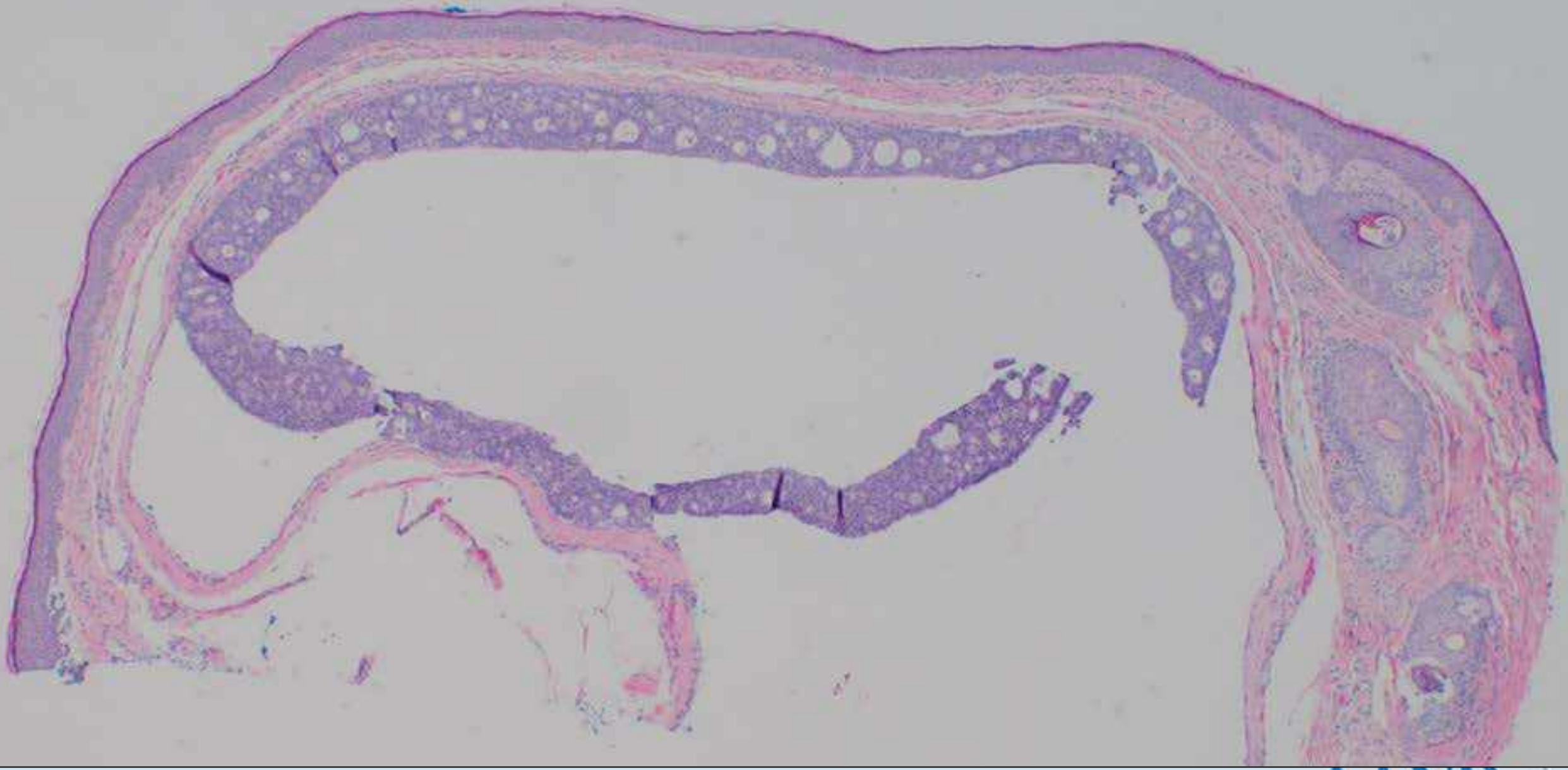
Sebaceous carcinoma

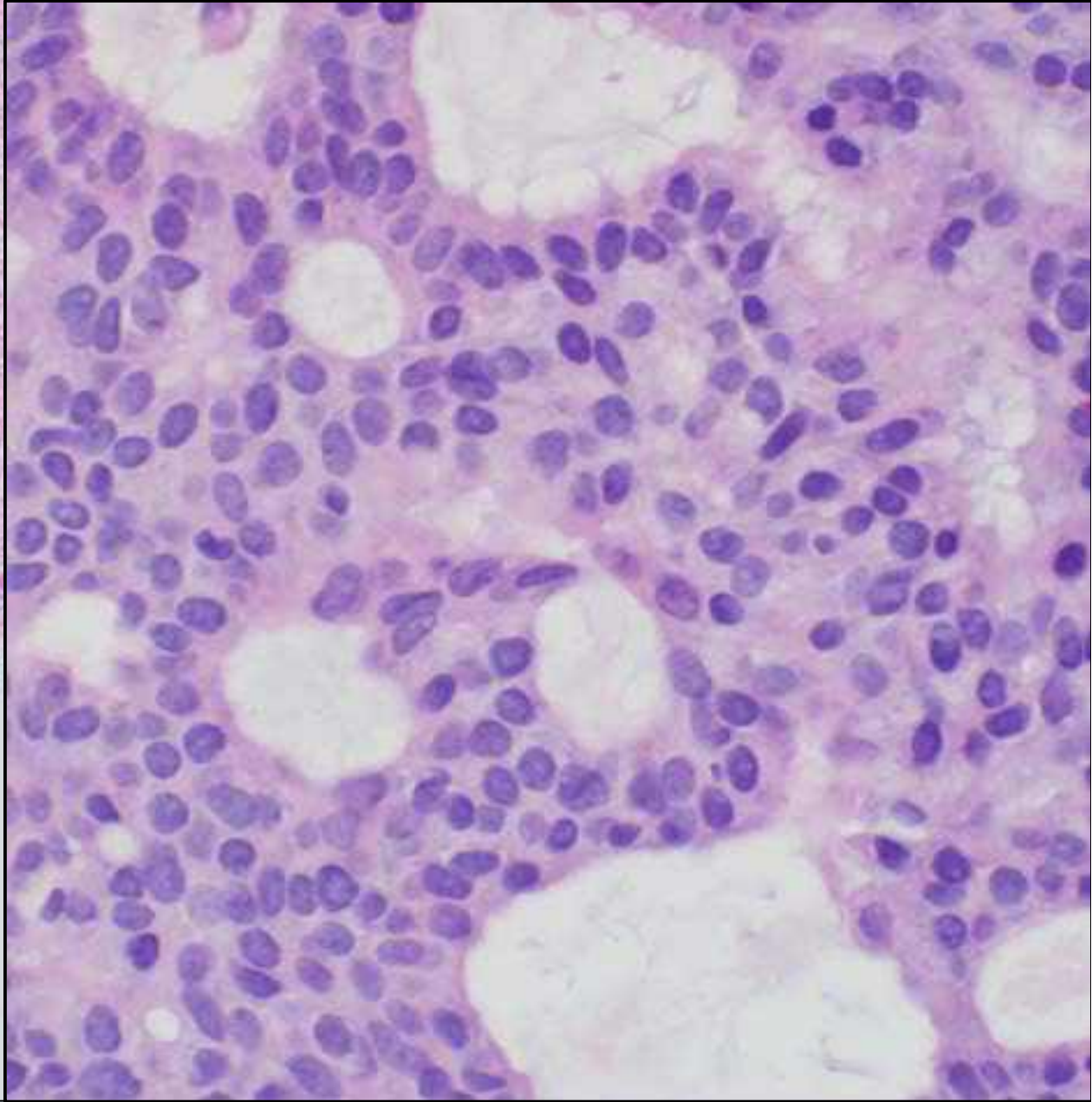
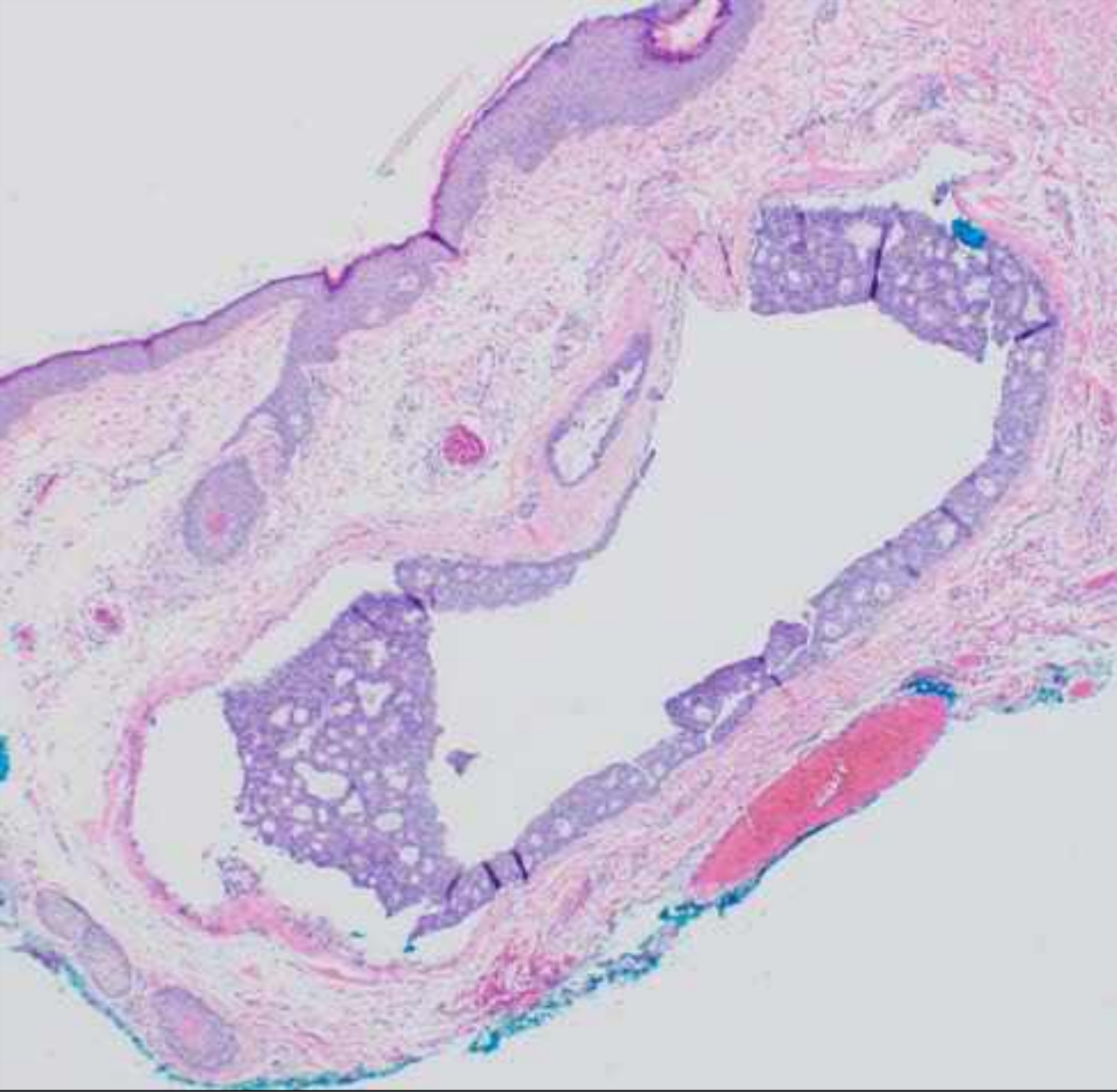


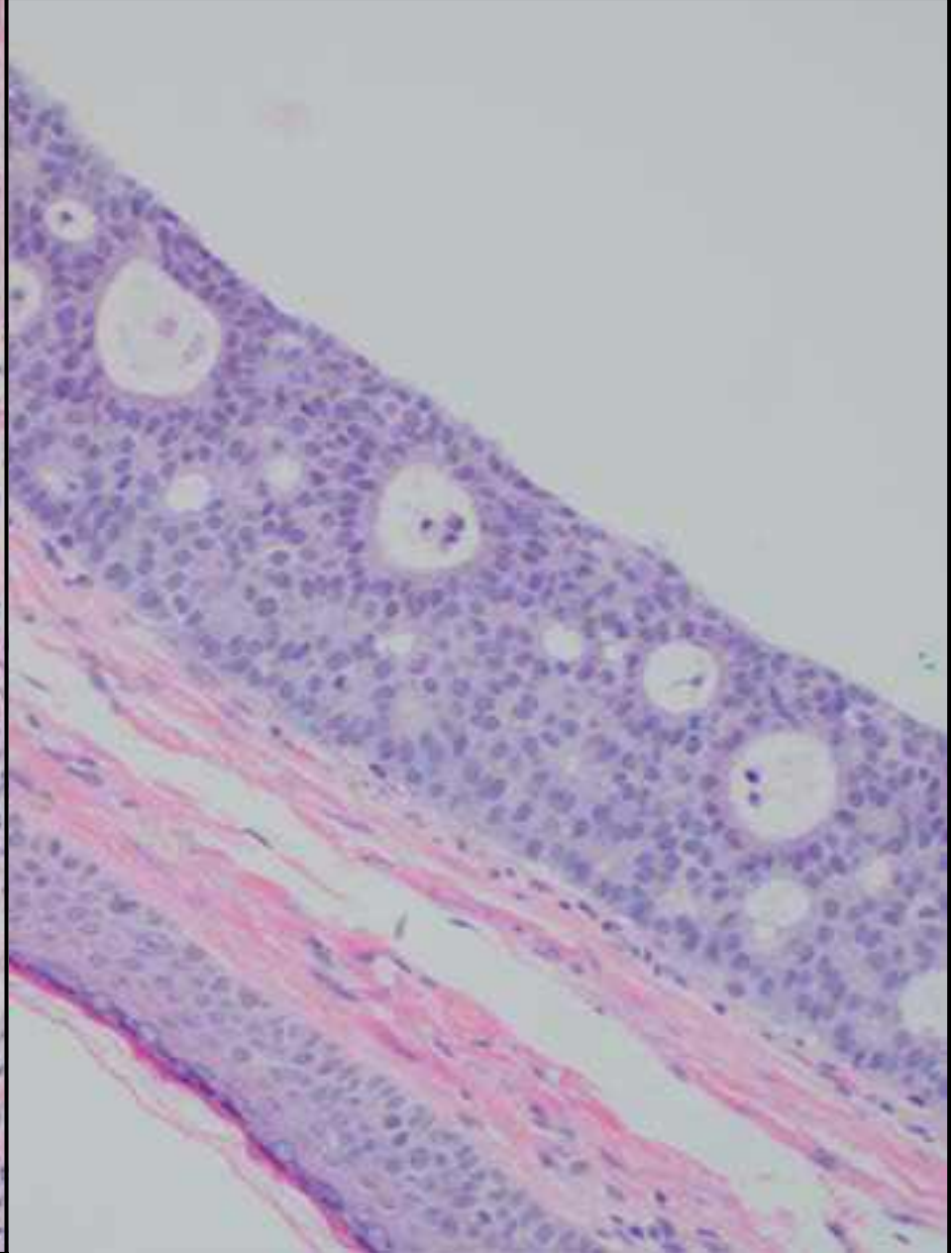
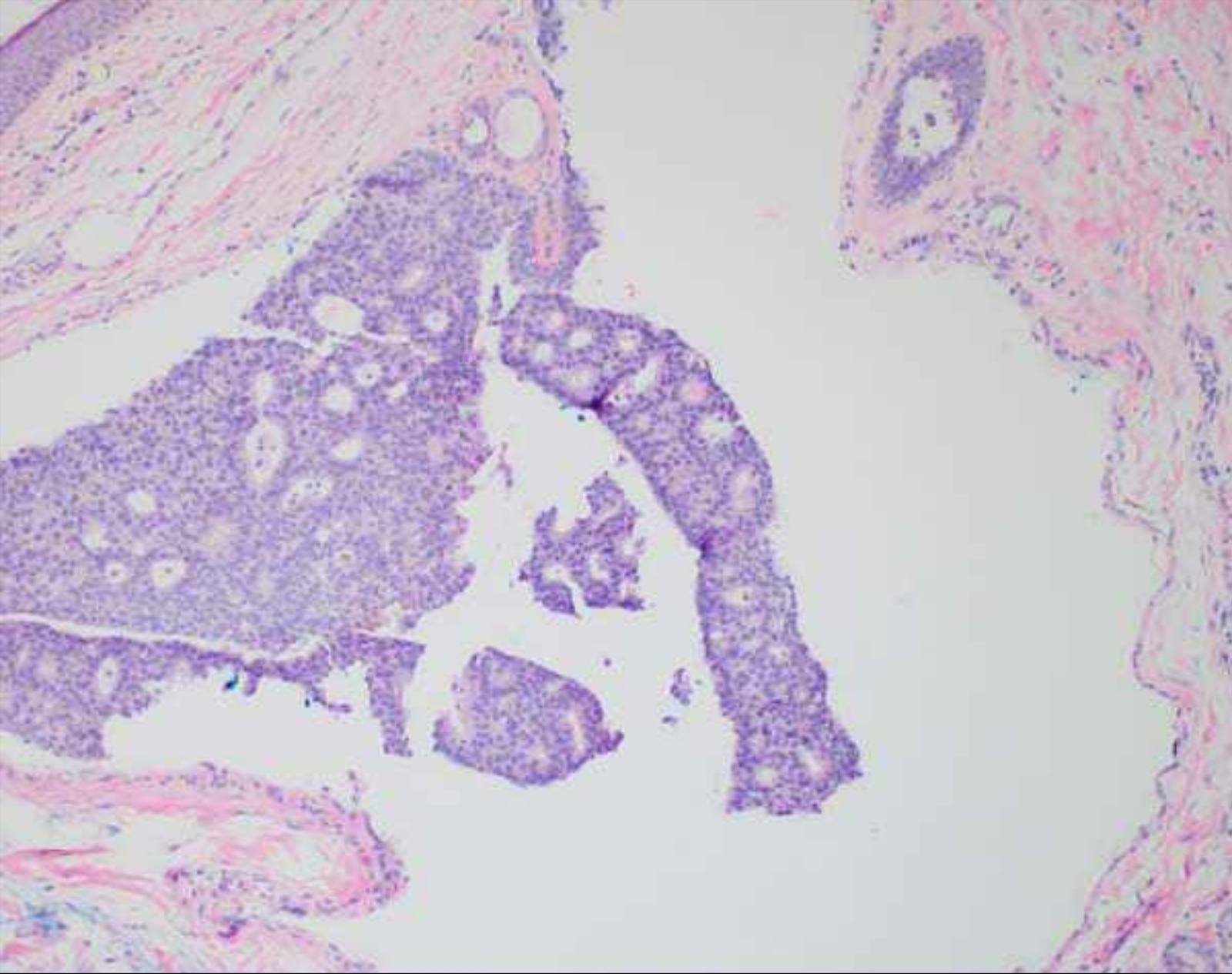
Patient History

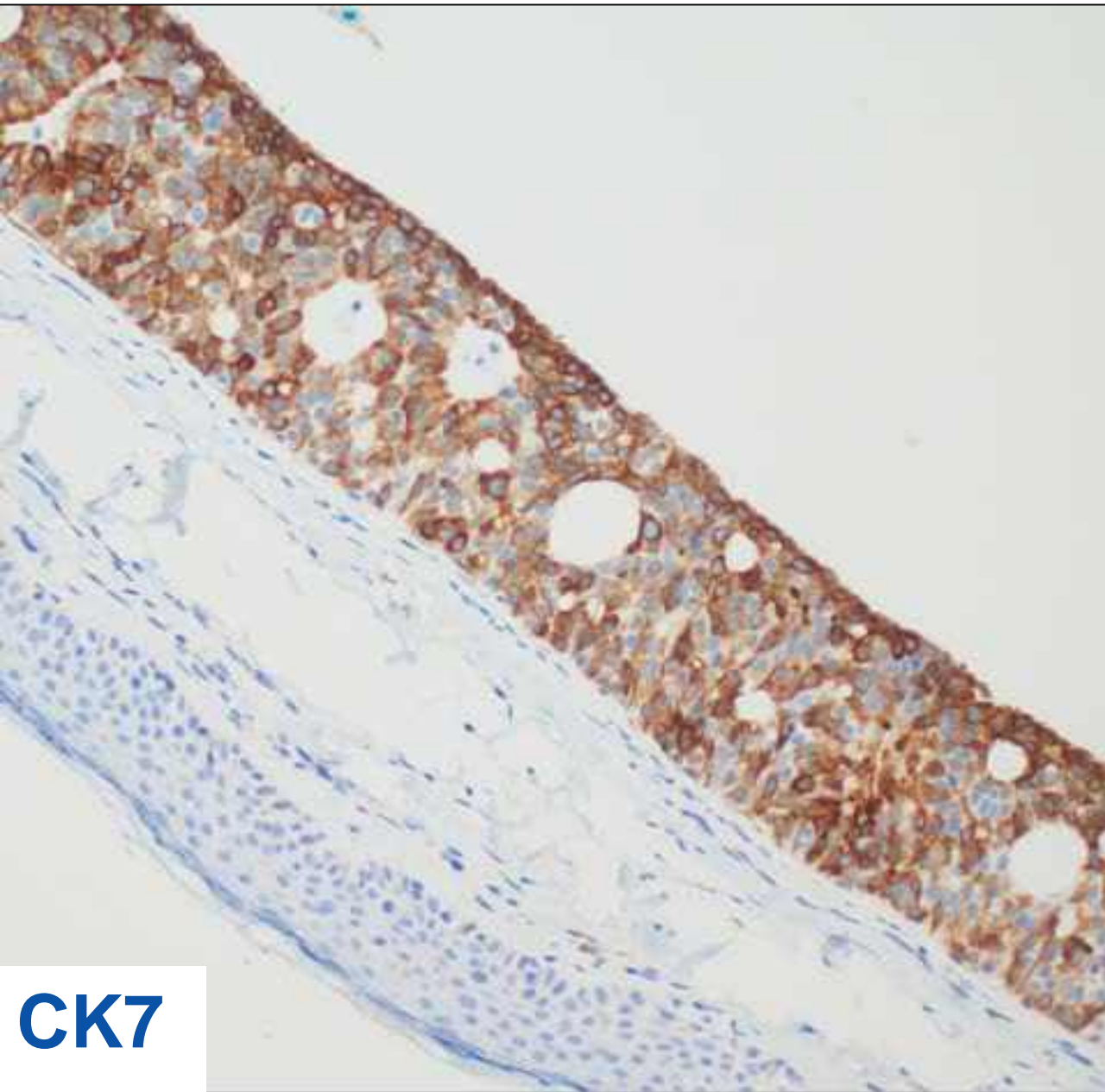


- **65 year old female with a RLL central violaceous lesion**
- **Cystic, 5 x 6 mm, with overlying telangiectasia**
- **Associated with 6 lashes**

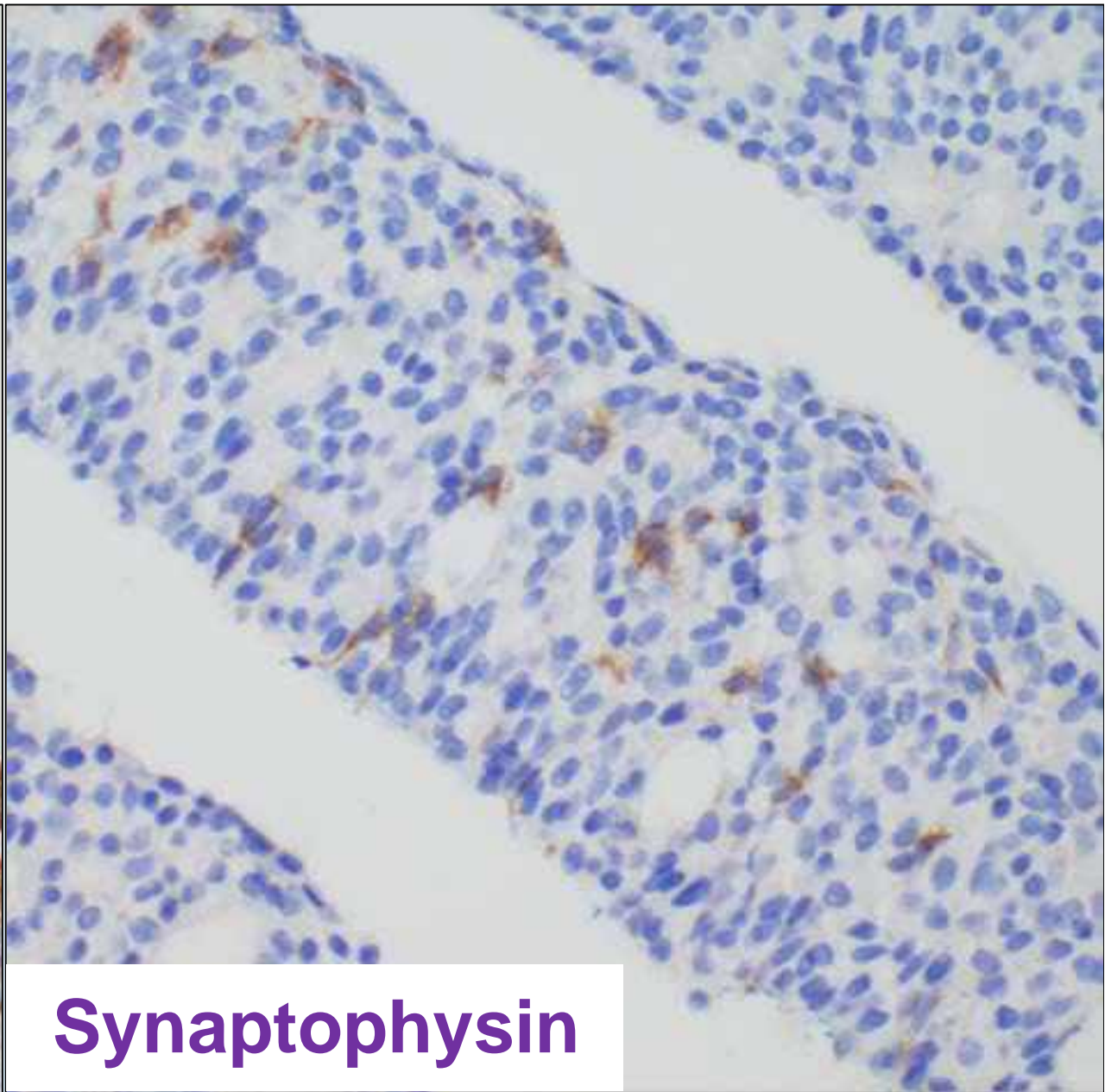




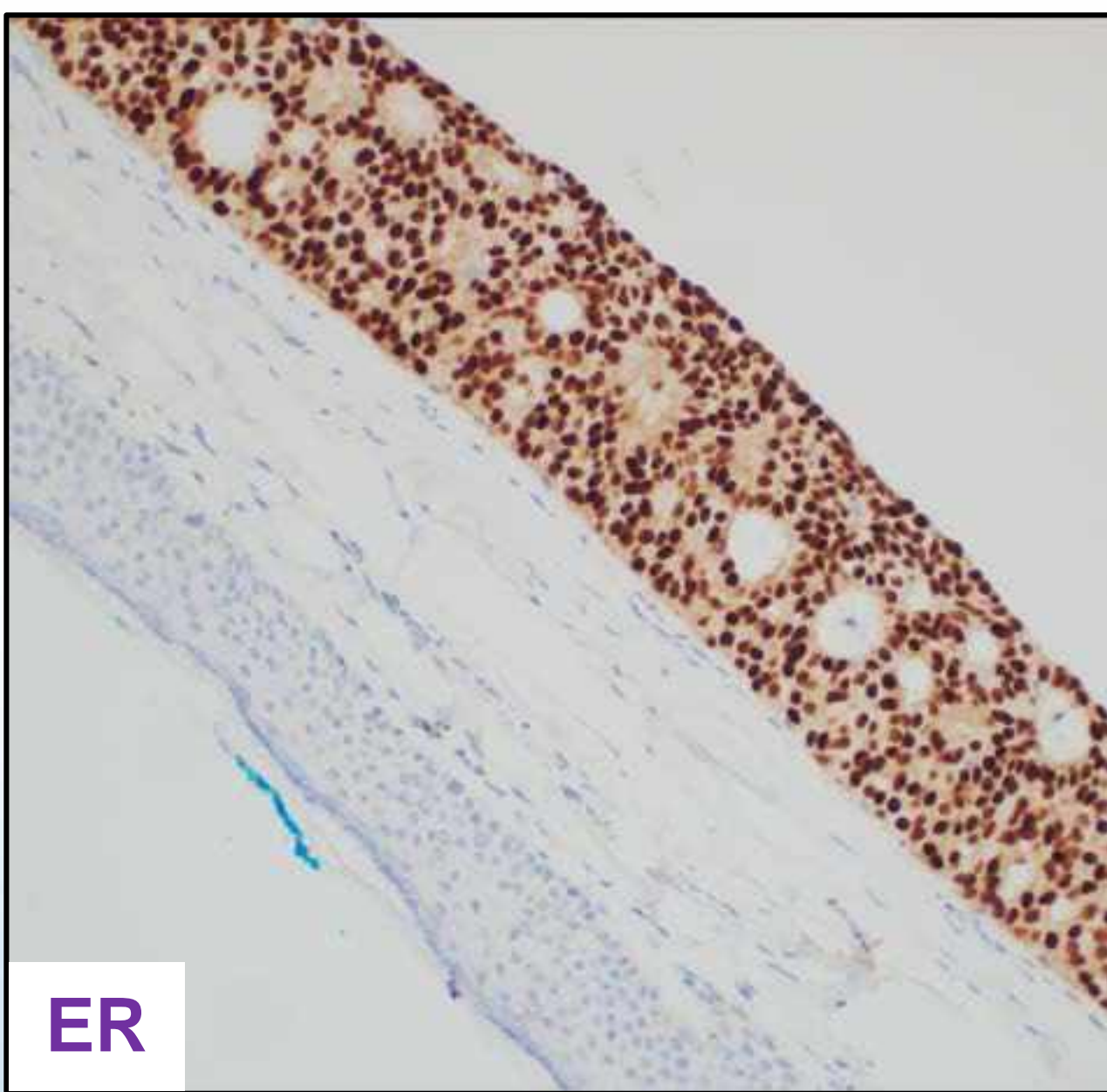




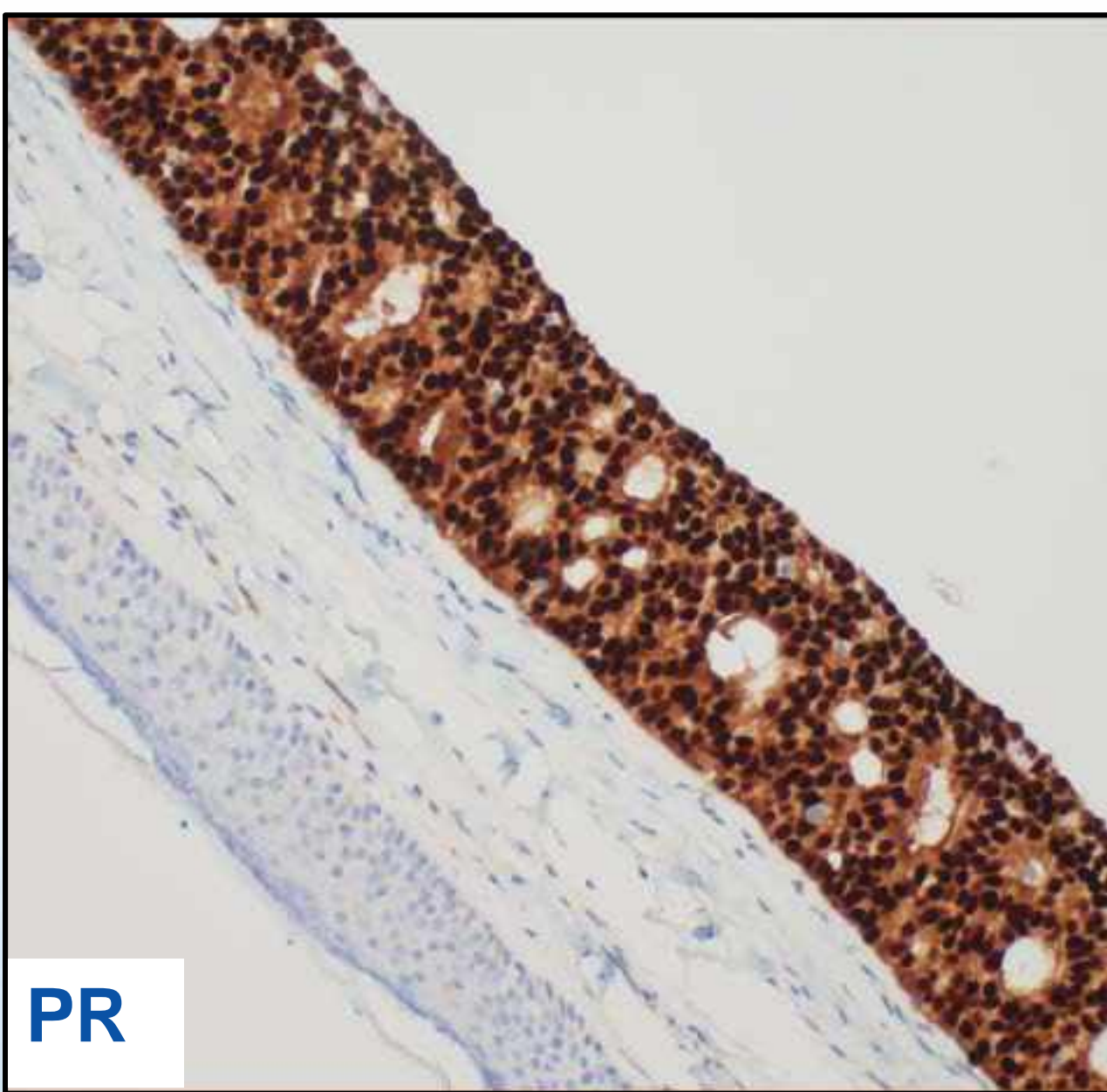
CK7



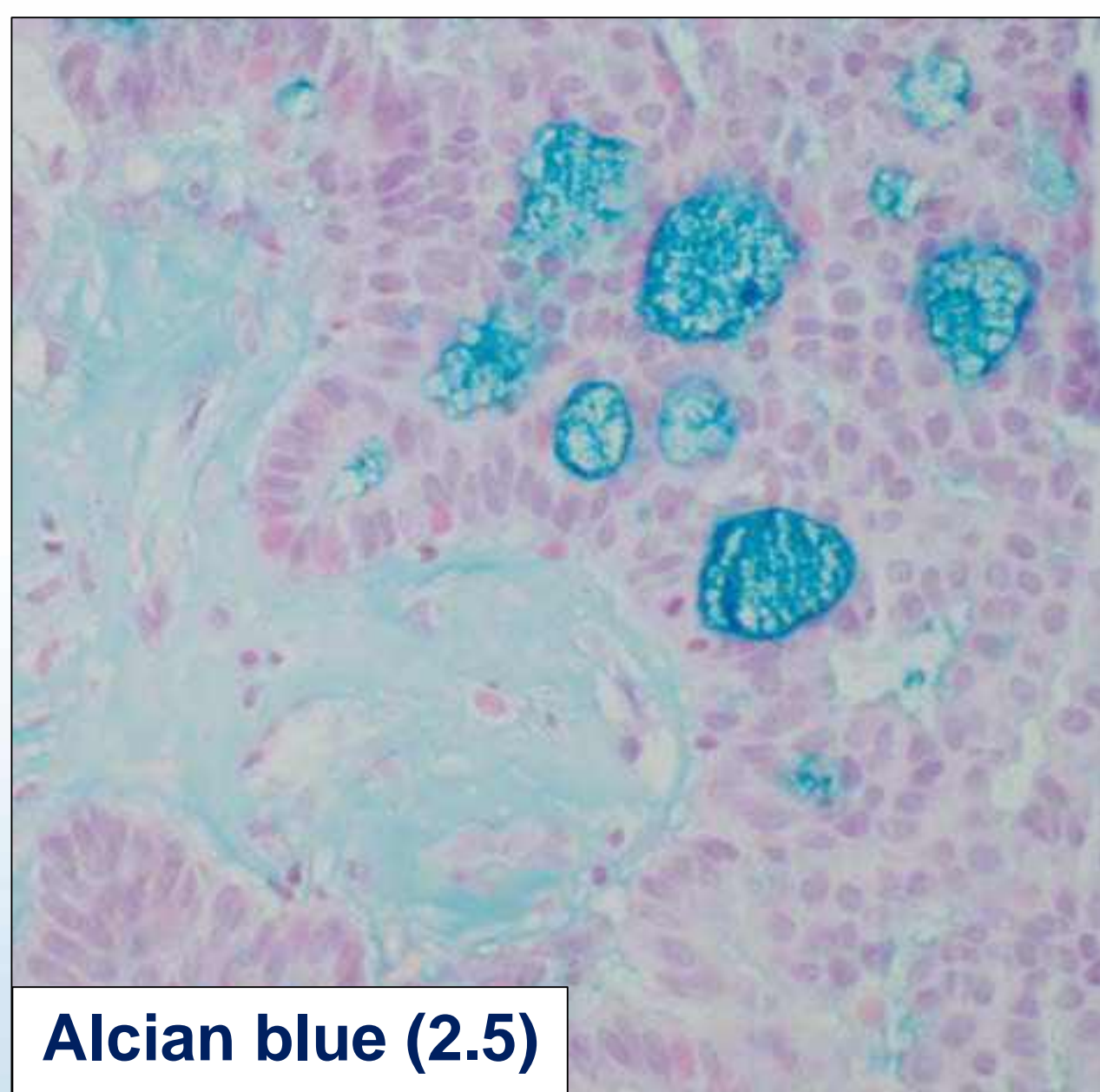
Synaptophysin



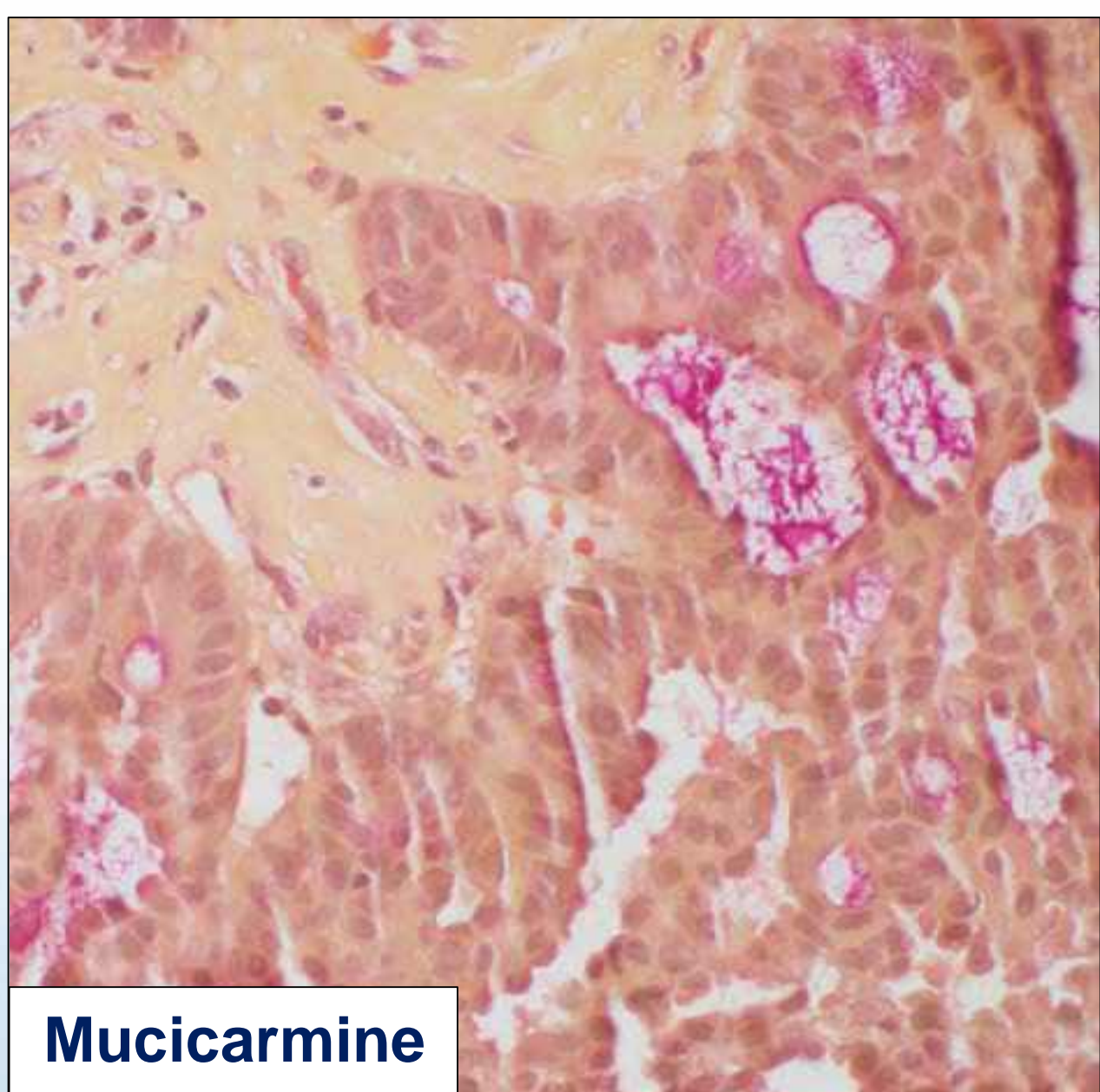
ER



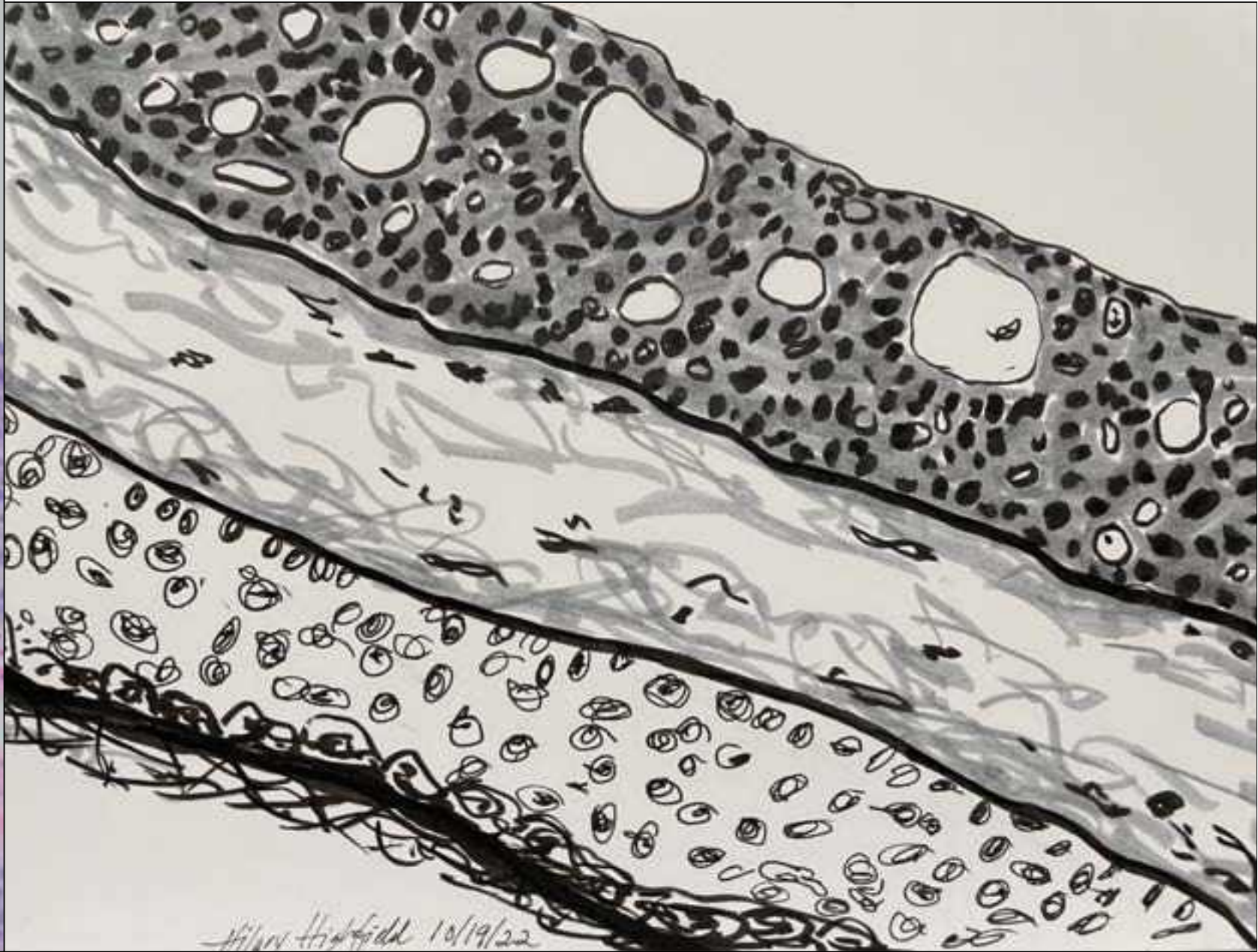
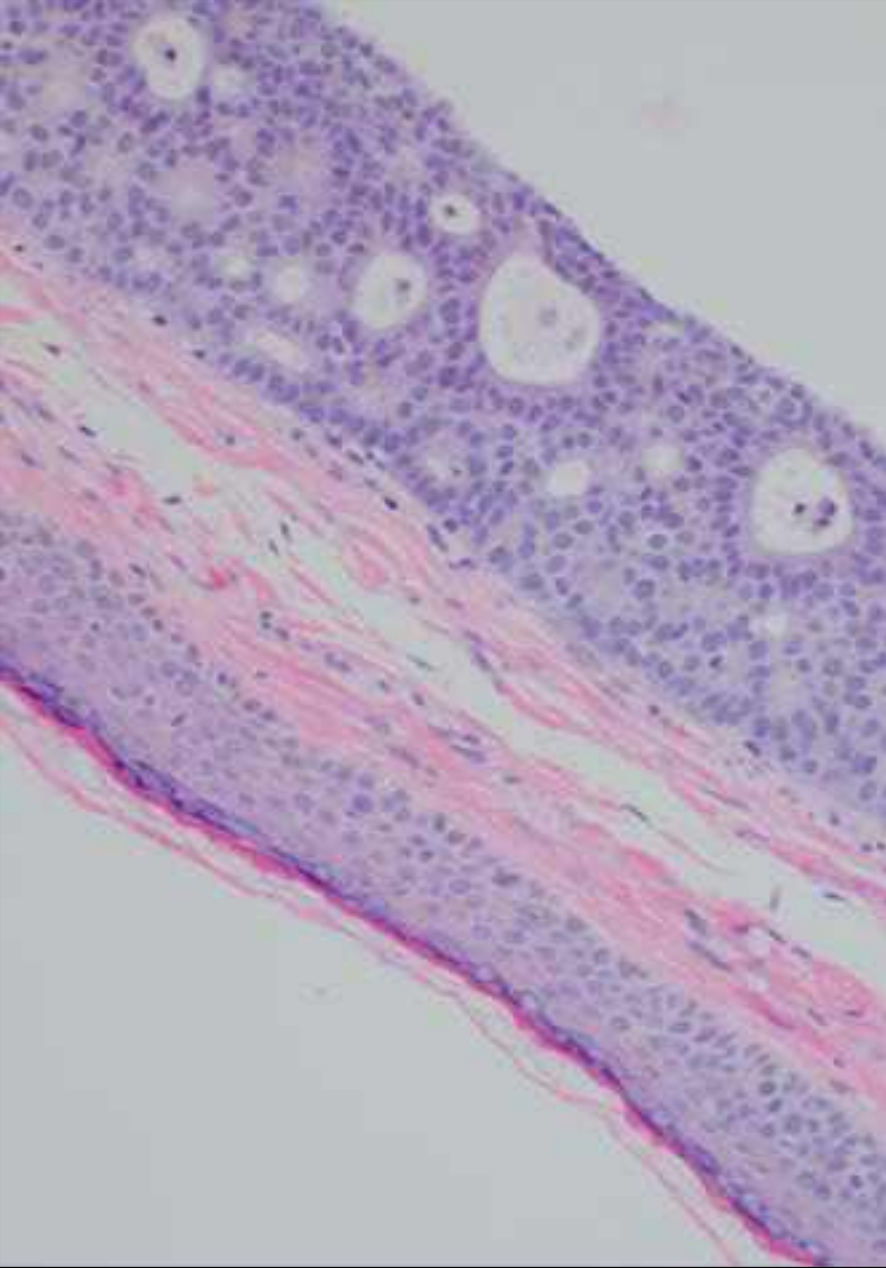
PR



Alcian blue (2.5)



Mucicarmine

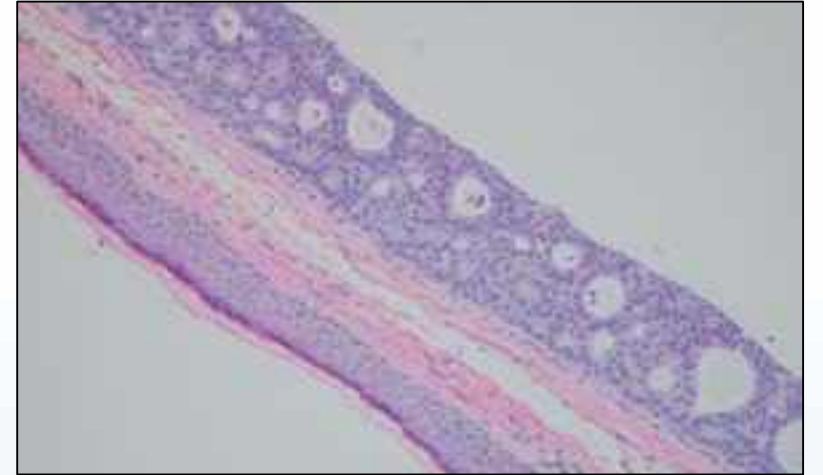


Diagnosis?



Endocrine Mucin-Producing Sweat Gland Carcinoma (EMPSGC)

- Low-grade carcinoma
- Eyelid lesion of older females
- Present as slow-growing lesion or cyst
- Can be associated with invasive mucinous component

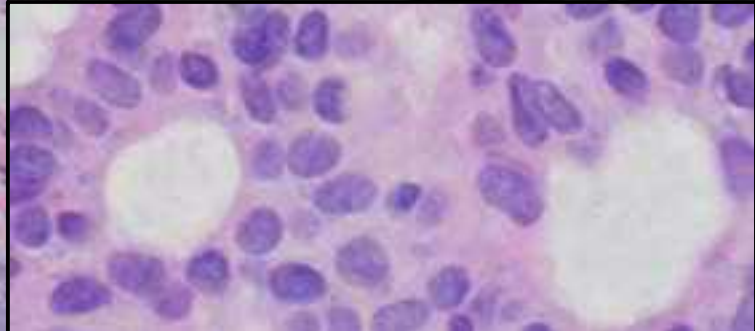
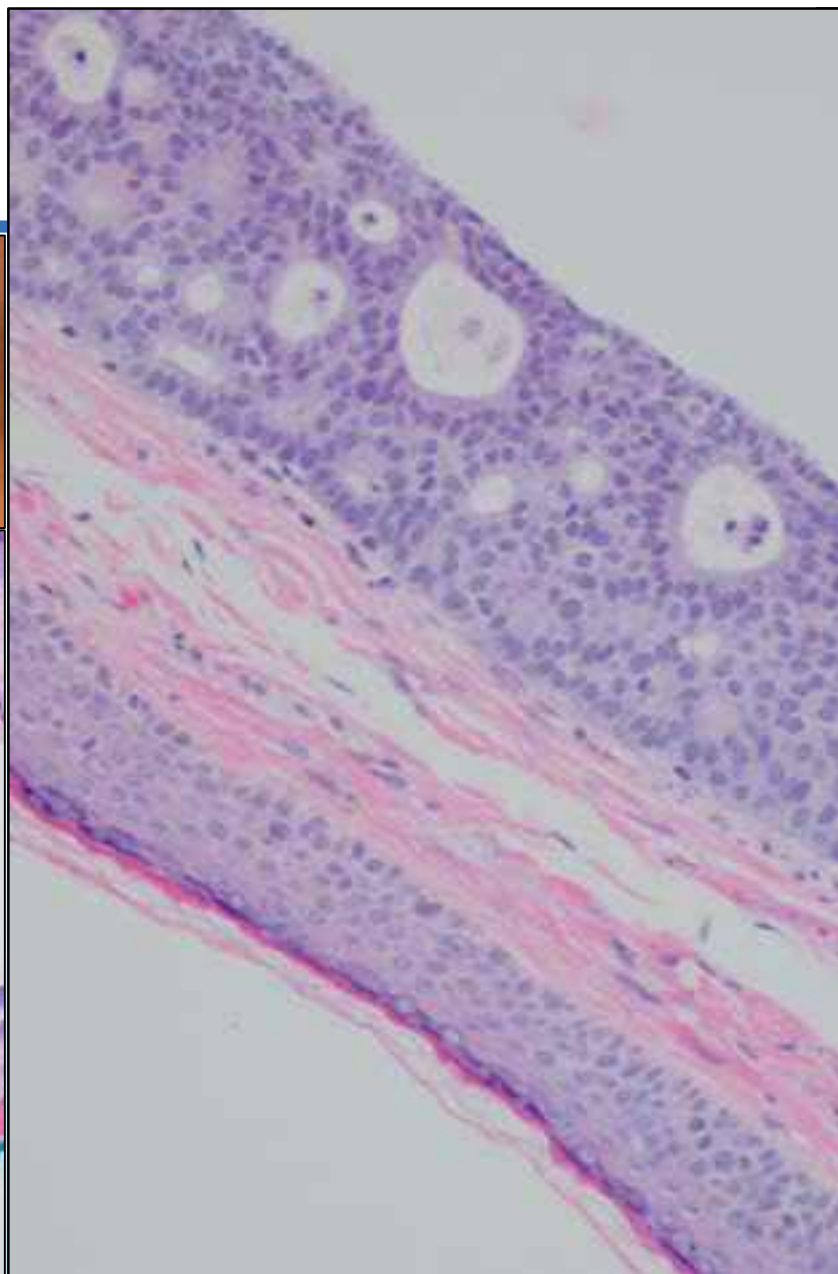
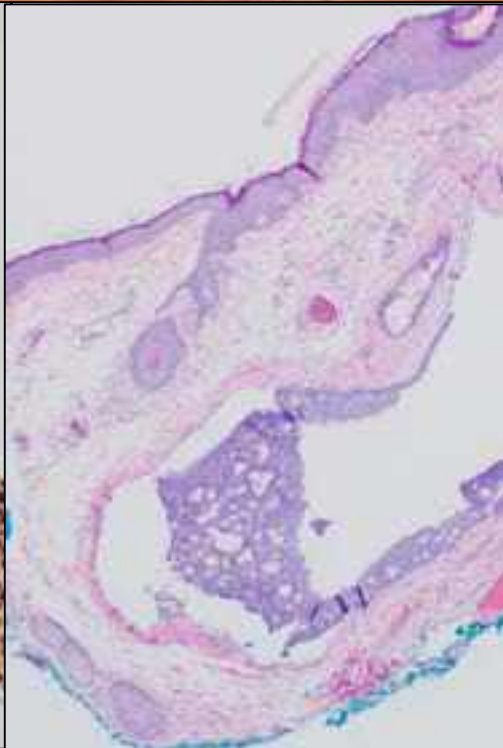
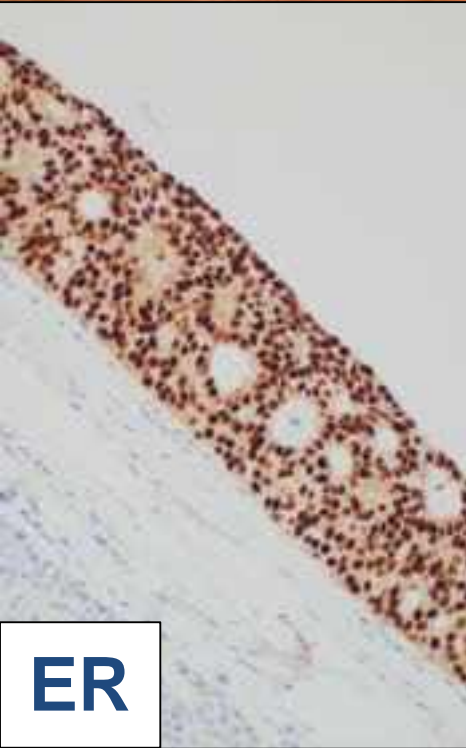


Differential Diagnosis:

- Benign Cyst (Hidrocystoma)
- Basal Cell Carcinoma
- Chalazion
- Metastatic Breast Carcinoma

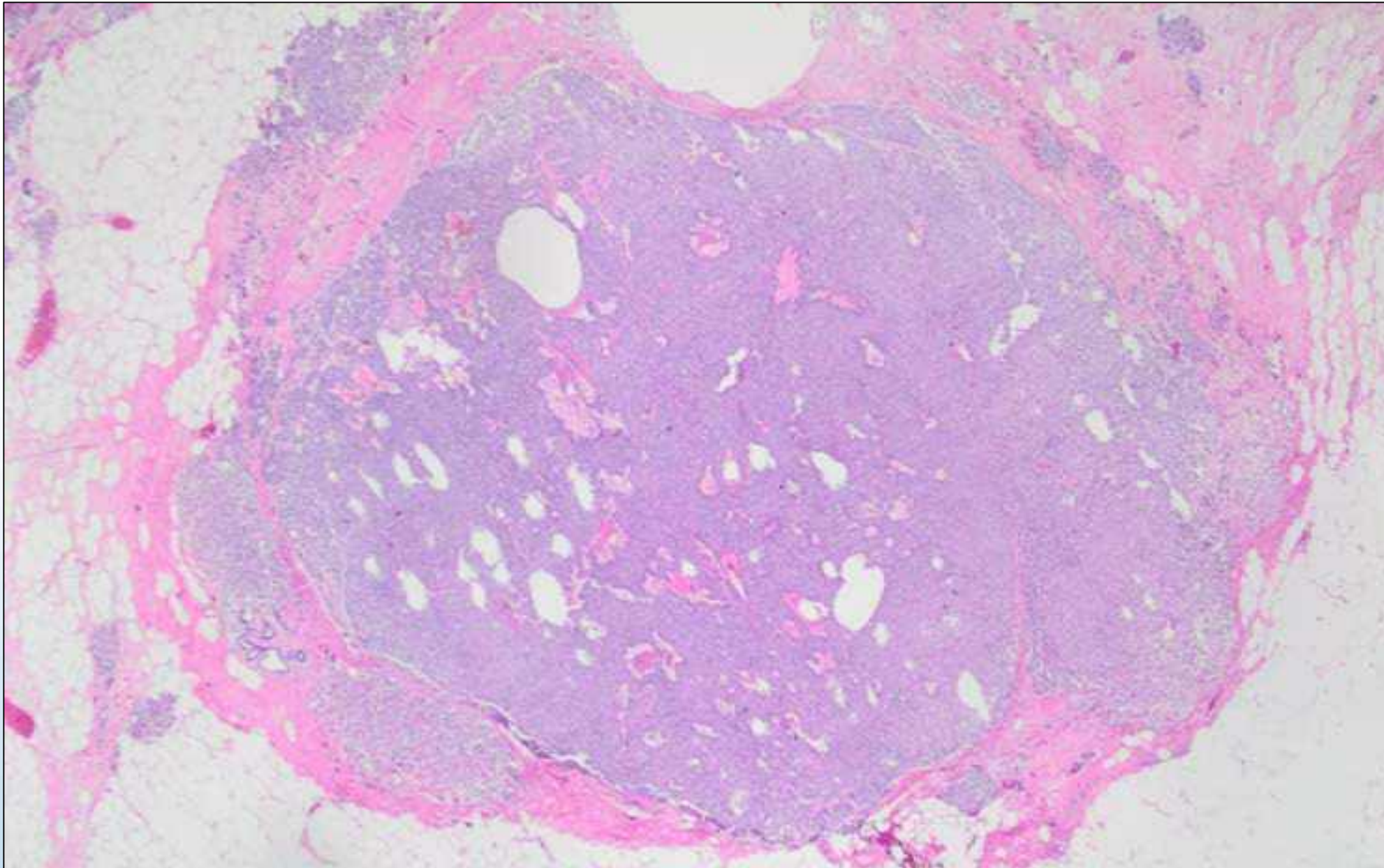


EMPSGC



ER

BREAST: SOLID PSEUDOPAPILLARY CARCINOMA



Endocrine Mucin-Producing Sweat Gland Carcinoma (EMPSGC)

- Rare low-grade carcinoma eyelid lesion of older(64yrs) females (2/3)
- Can be associated with an invasive mucinous component (1/3)
- Differential includes cystic lesions (hidrocystoma), BCC, chalazion, metastasis (breast)



Endocrine Mucin-Producing Sweat Gland Carcinoma (EMPSGC)

- Neuroendocrine appearance with mucin
 - + Synaptophysin/chromogranin/NSE/INSM-1
 - + Alcian blue (2.5), Mucicarmine, PAS (mucin)
 - + CAM5.2, CK7 (CK20 negative)
 - + ER/PR
- EMPSGC with invasive mucinous component → Primary cutaneous mucinous carcinoma (PCMC)
- Differential includes metastasis (breast) – consider systemic workup and plan for follow-up



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1. Agni, M et al. An Update on EMPSSGC. Am J Surg Pathol. 2020 44(8):1005-1016.
2. Zembowicz et al. EMPSSGC: Twelve cases that suggest that it is a precursor of Some Invasive Mucinous Carcinomas. Am J Surg Path. 2005. 29(10): pp1330-1339.
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5. Ravi, PY et al. EMPSSGC: Emerging evidence of multicentric cutaneous origin and occasional concurrence with analogous breast tumors. Am J Dermatopathol. 2002; 44(5):321-326.
6. Wung D et al. Bilateral Concurrent EMPSSGC and Mucinous Carcinoma of the Eyelids. Ophthalmic Plast Reconstr Surg. 2022; 38(4): e96-99.



Resources



AAO Pathology Atlas

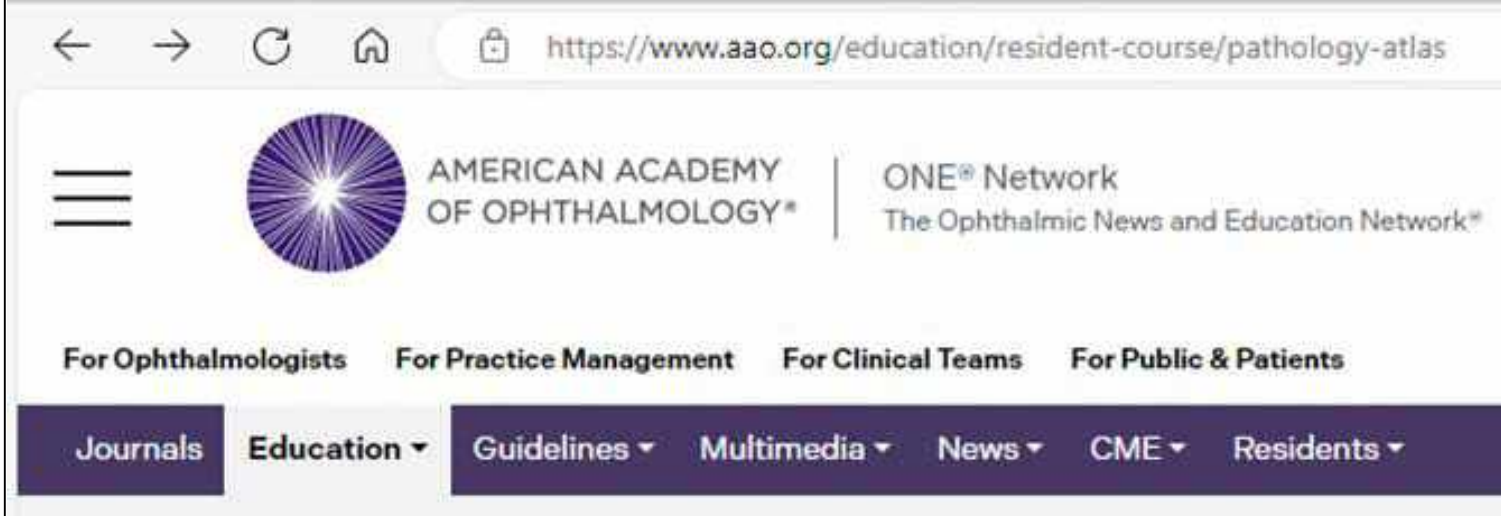
SEP 16, 2016

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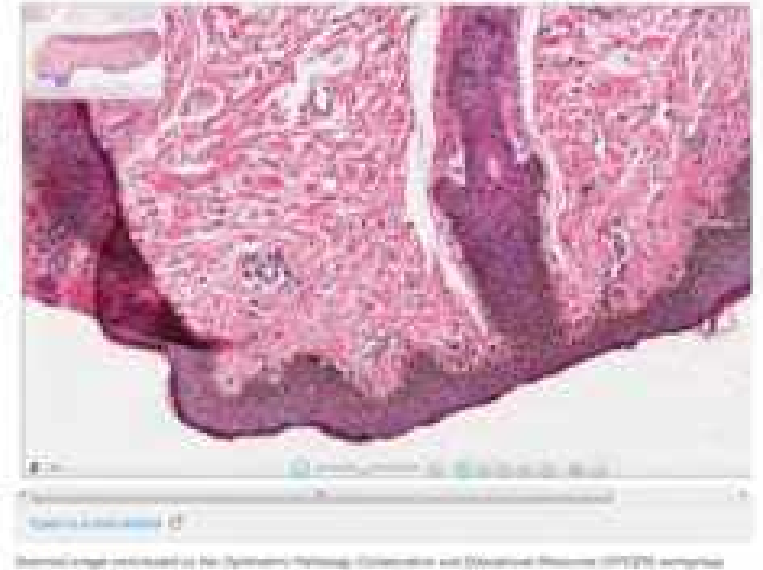
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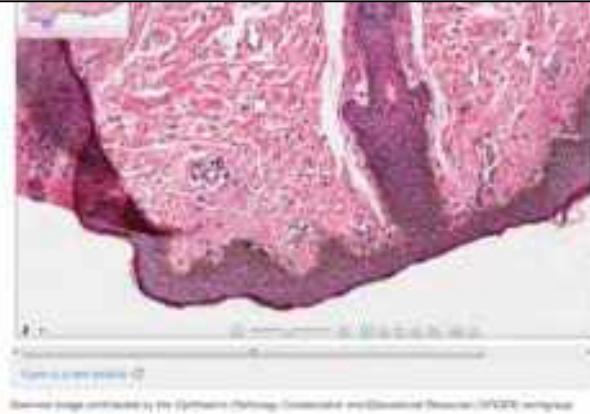
The screenshot shows the top portion of a web browser displaying the AAO Pathology Atlas website. The address bar shows the URL: <https://www.aao.org/education/resident-course/pathology-atlas>. The page features the AAO logo (a stylized sunburst) and the text "AMERICAN ACADEMY OF OPHTHALMOLOGY®". To the right, it says "ONE® Network | The Ophthalmic News and Education Network®". Below this, there are four navigation links: "For Ophthalmologists", "For Practice Management", "For Clinical Teams", and "For Public & Patients". A dark blue navigation bar contains several menu items: "Journals", "Education" (with a dropdown arrow), "Guidelines" (with a dropdown arrow), "Multimedia" (with a dropdown arrow), "News" (with a dropdown arrow), "CME" (with a dropdown arrow), and "Residents" (with a dropdown arrow).



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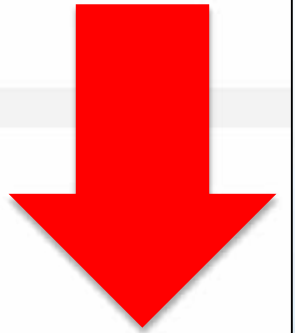
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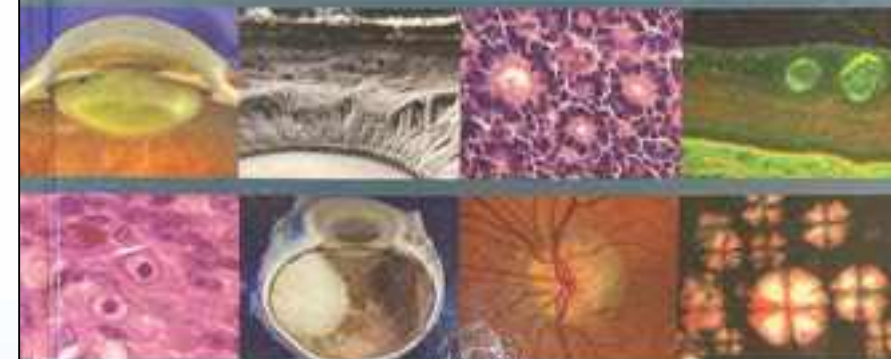




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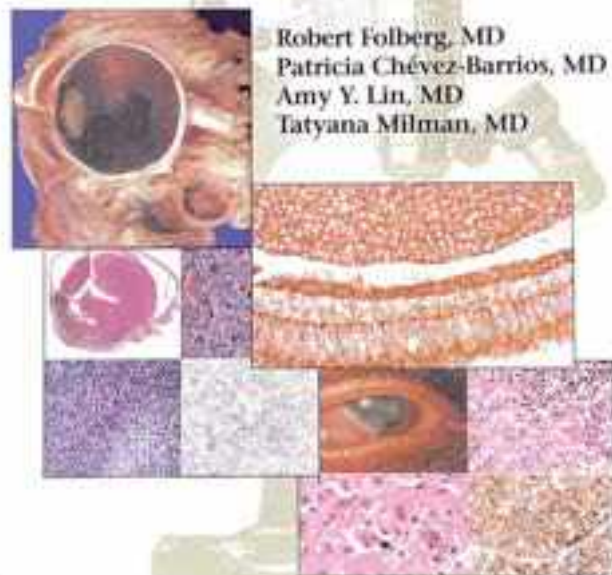
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