Chief's Rounds

Sunidhi Ramesh, MD
PGY-2

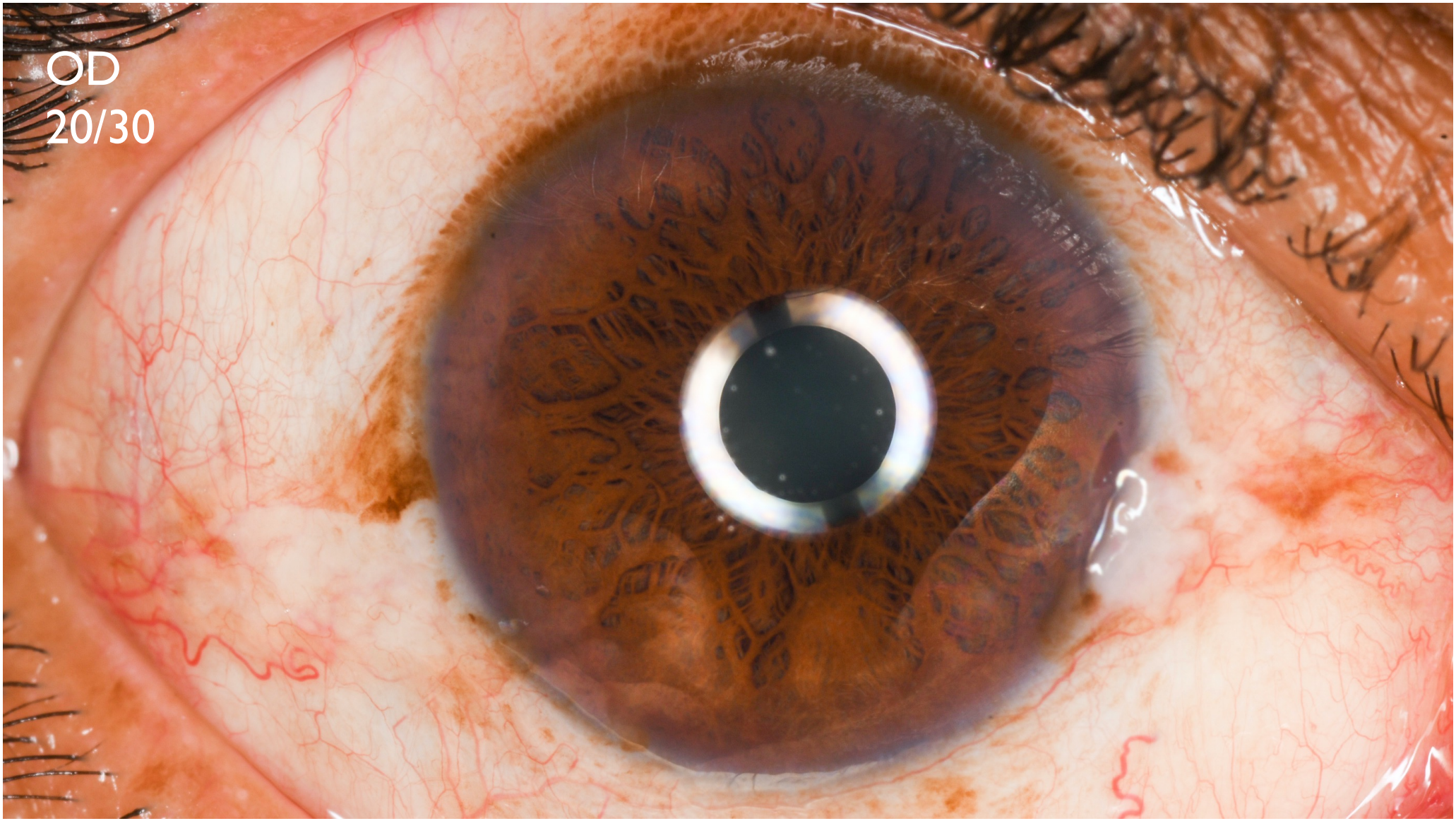
May 3, 2024

I have no financial relationships to disclose. I will not discuss off label use and/or investigational use in my presentation.

52 y.o. male presenting to the Wills Emergency Room with “severe, 10/10 left eye pain” for two months

Past Ocular History	Past Medical History	Medications
• POAG OS	• None • Notably negative for DM, HTN, HLD, malignancy, and stroke	• Simbrinza TID OS • Timolol BID OS
Allergies & Sensitivities	**Social and Family History**	**Review of Systems**
• None	• 30 pack-year smoker; quit ~1 week prior to WER presentation • Choroidal melanoma in maternal uncle with enucleation done in Massachusetts in 2011	• Unremarkable

VA_{sc}	20/30	EOMs	Full, ortho
	20/400		Full, ortho
IOP	15	Pupils	RRL
	34		Sluggish; no APD by reverse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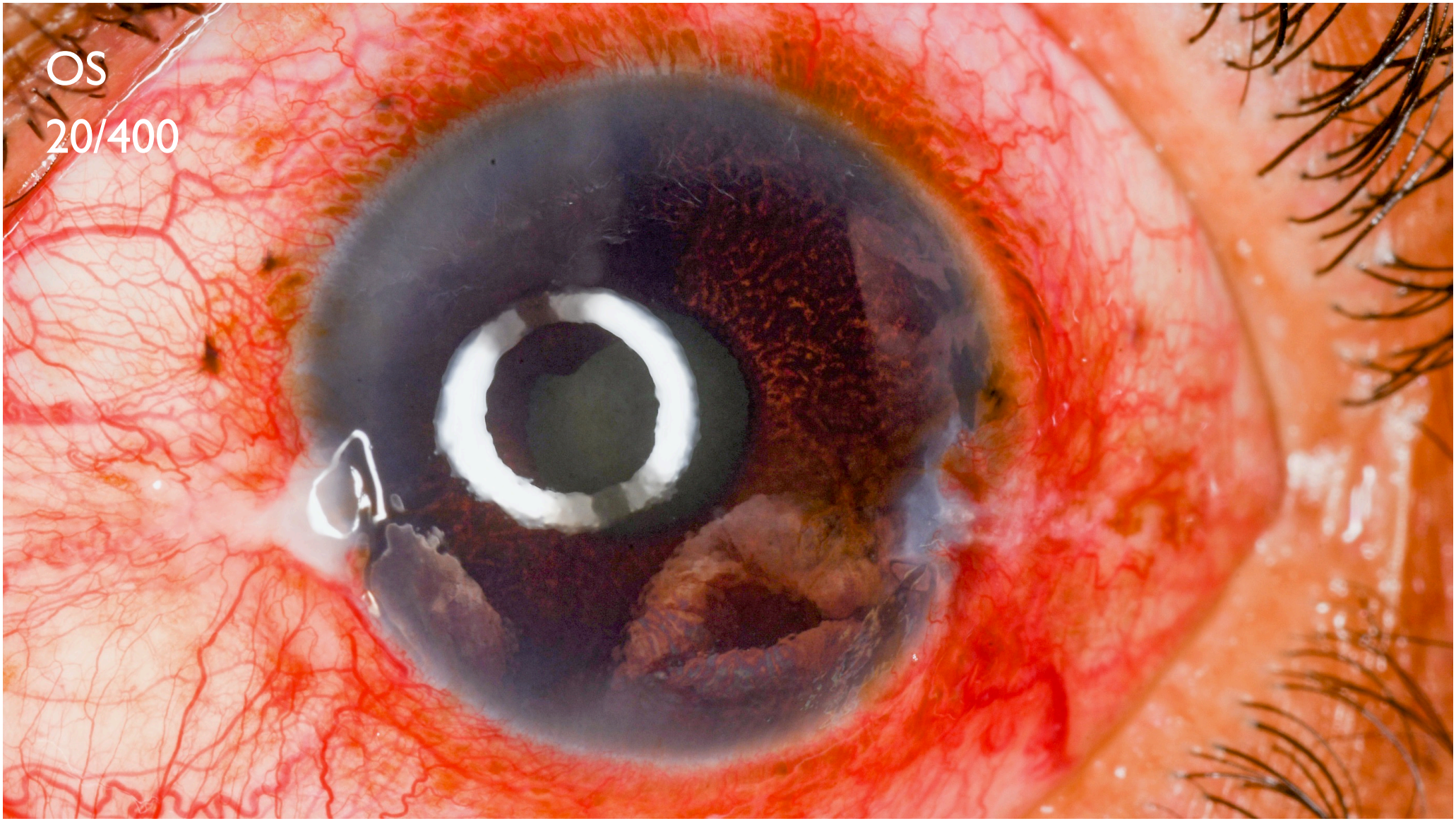
20/30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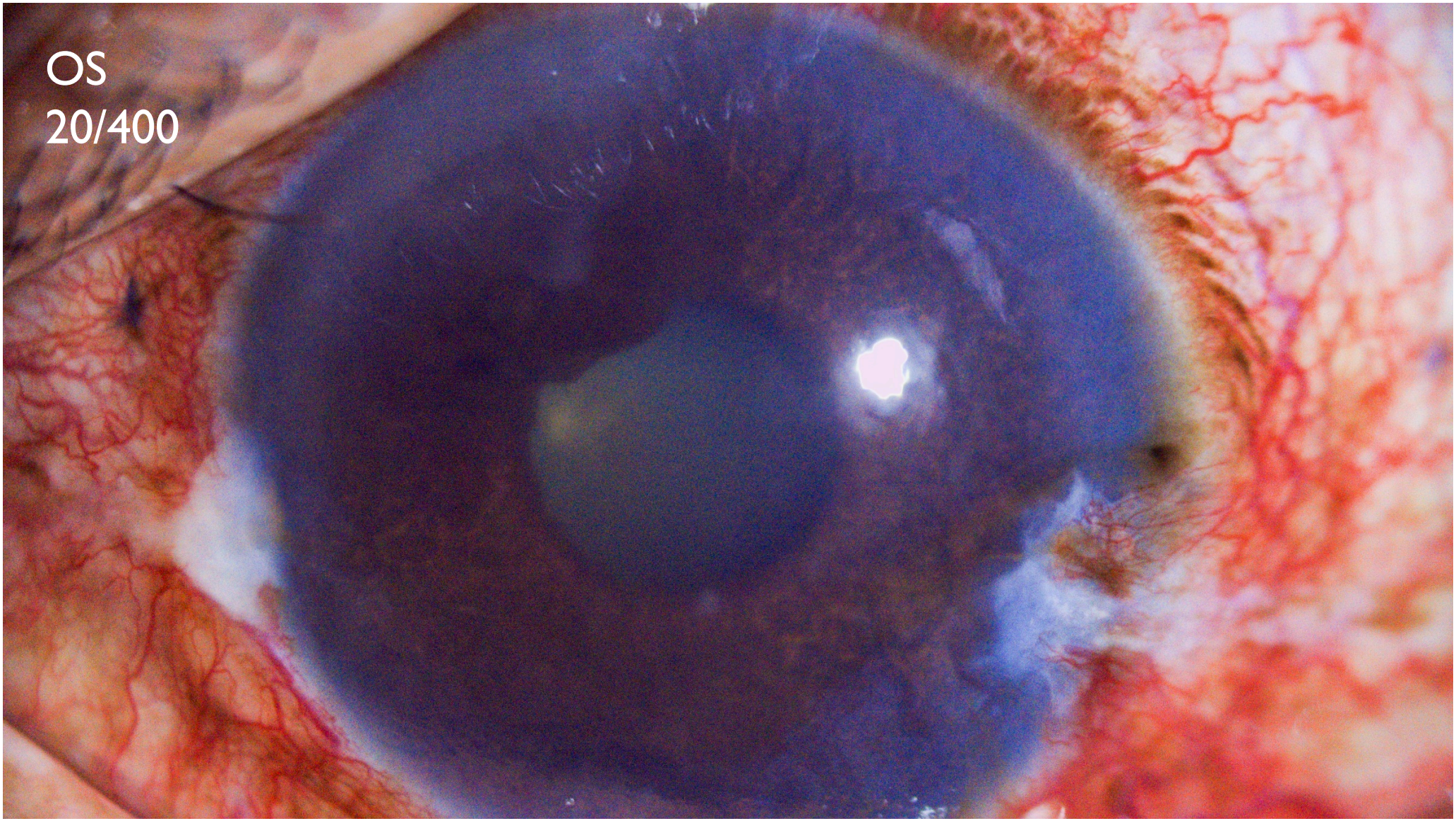
20/400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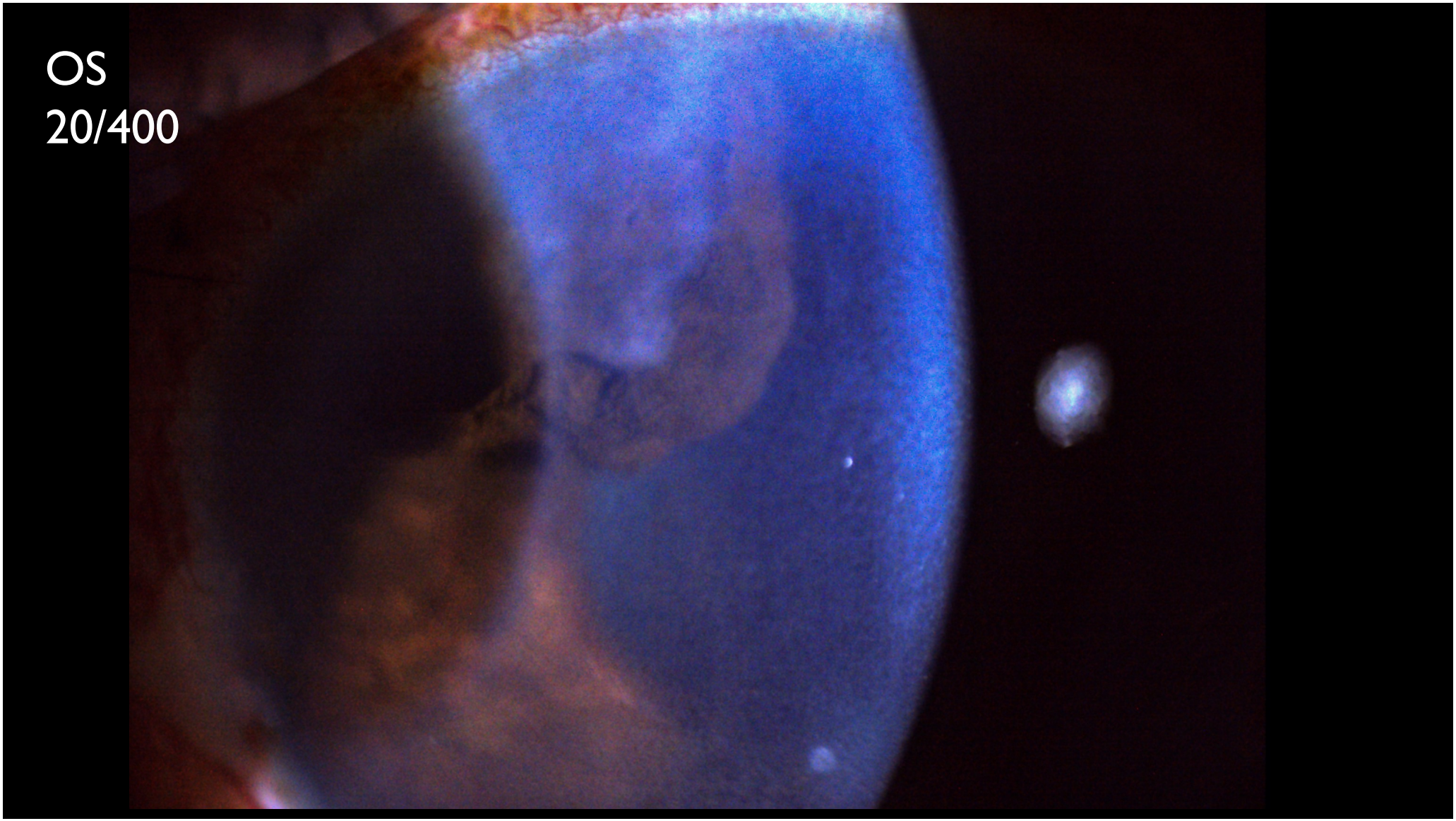
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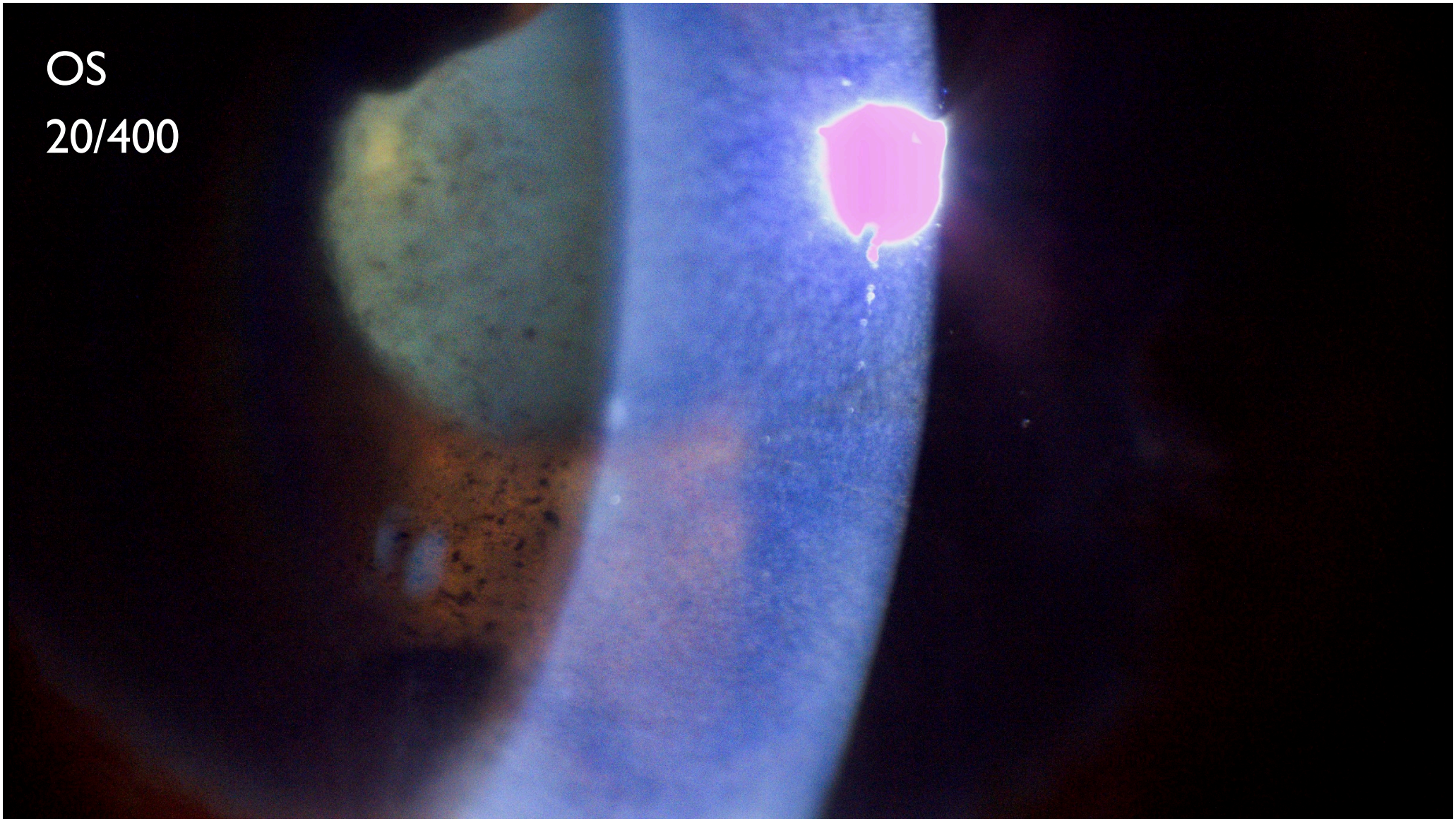
20/400


OS
20/400

OD		OS
Mild blepharitis; no follicles	**Lids**	Mild blepharitis; no follicles
White and quiet	**Sclera & Conjunctiva**	Trace injection
Nasal pinguecula < 1mm onto K surface	**Cornea**	Nasal pinguecula < 1mm onto K surface; diffuse, 2+ SPK; diffuse keratic precipitates; iris-K touch inferiorly and superiorly
Deep and quiet	**Anterior Chamber**	Trace pigmented cell; no flare
Flat; round and reactive	**Iris**	Pigmented iris lesions from 6 to 7 o'clock measuring 3mm(v) by 2.5mm (h) and from 9:30 to 12:00 measuring 4mm(h) by 5.5mm(v); both regions with iris-K touch and extension over mid-dilated pupil
Clear	**Lens**	2+ NS

OD		OS
Clear	**Vitreous**	Clear
Sharp, pink; no disc edema	**Disc**	Sharp, pink; no disc edema
0.3	**C/D Ratio**	0.3
No edema, thickening, or hemorrhage	**Macula**	No edema, thickening, or hemorrhage
Normal in caliber	**Vessels**	Normal in caliber
Flat without breaks, tears, or RD	**Periphery**	Flat without breaks, tears, or RD to the extent visualized

Overview

- 52 y.o. male presenting to the Wills Emergency Room with "severe, 10/10 left eye pain" for two months
  - POHx of POAG; family history of choroidal melanoma s/p enucleation
  - Vasc 20/30 OD, 20/400 OS; IOP 15 OD, 34 OS
  - SLE with diffuse keratic precipitates, iris-K touch inferiorly and superiorly, and several, scattered pigmented iris lesions
  - DFE unremarkable

DDx?

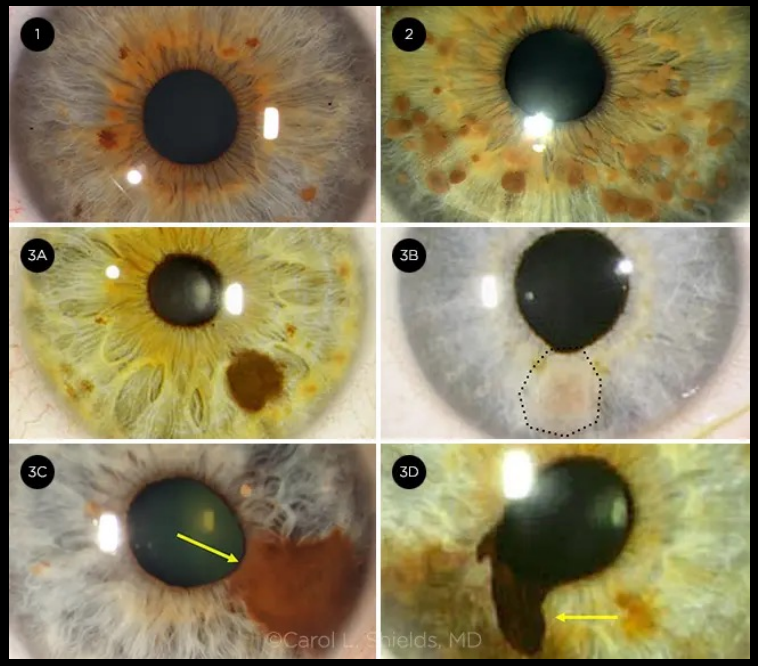
Differential Diagnosis for Iris Lesions

Benign lesions

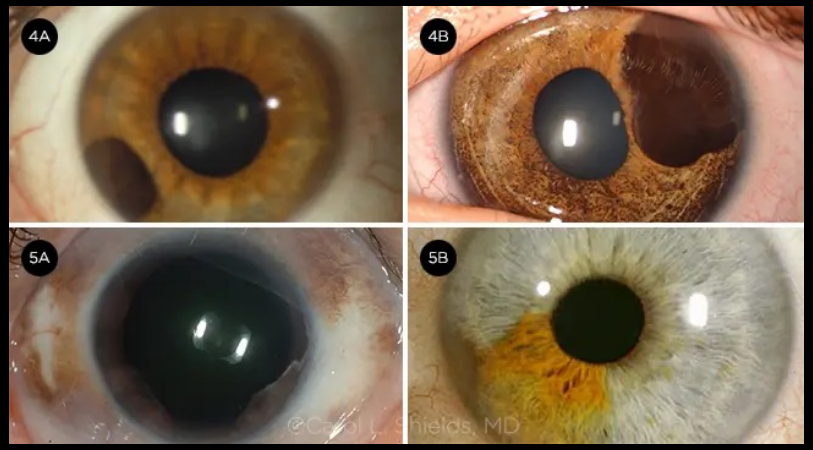
Iris cysts
Iris nevus
Iris freckles
Iris pigment epithelium adenoma
Iris leiomyoma
Iris nodules
Iris melanocytoma
Iris melanocytosis

Malignant lesions

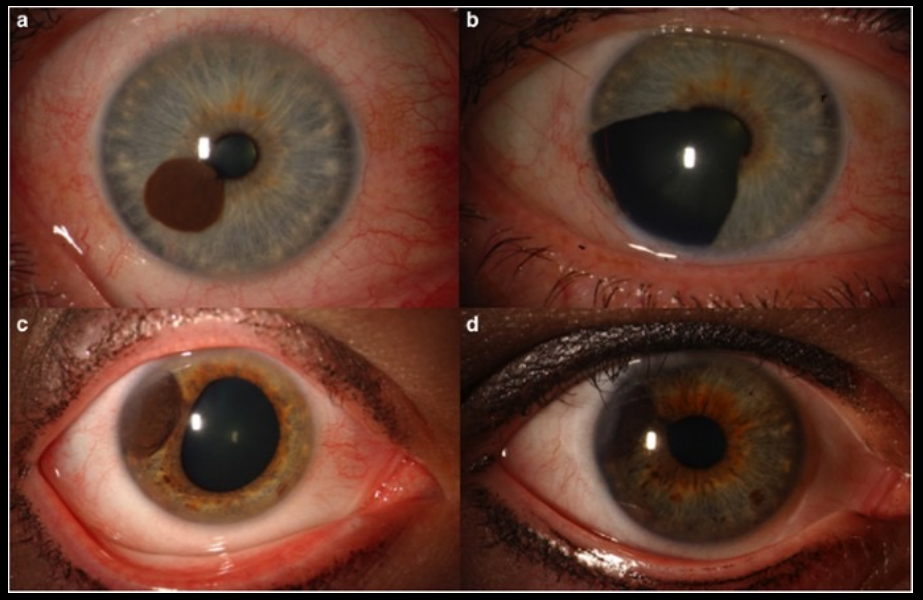
Iris melanoma
Iris pigment epithelium adenocarcinoma
Metastasis to the iris



(1) Iris freckles, (2) Lisch nodules, (3A) pigmented nevus, (3B) nonpigmented nevus, (3C) corectopia, and (3D) ectropion



(4A, B) Iris melanocytoma
(5A, B) Iris melanocytosis



(a-d) Uveal melanoma

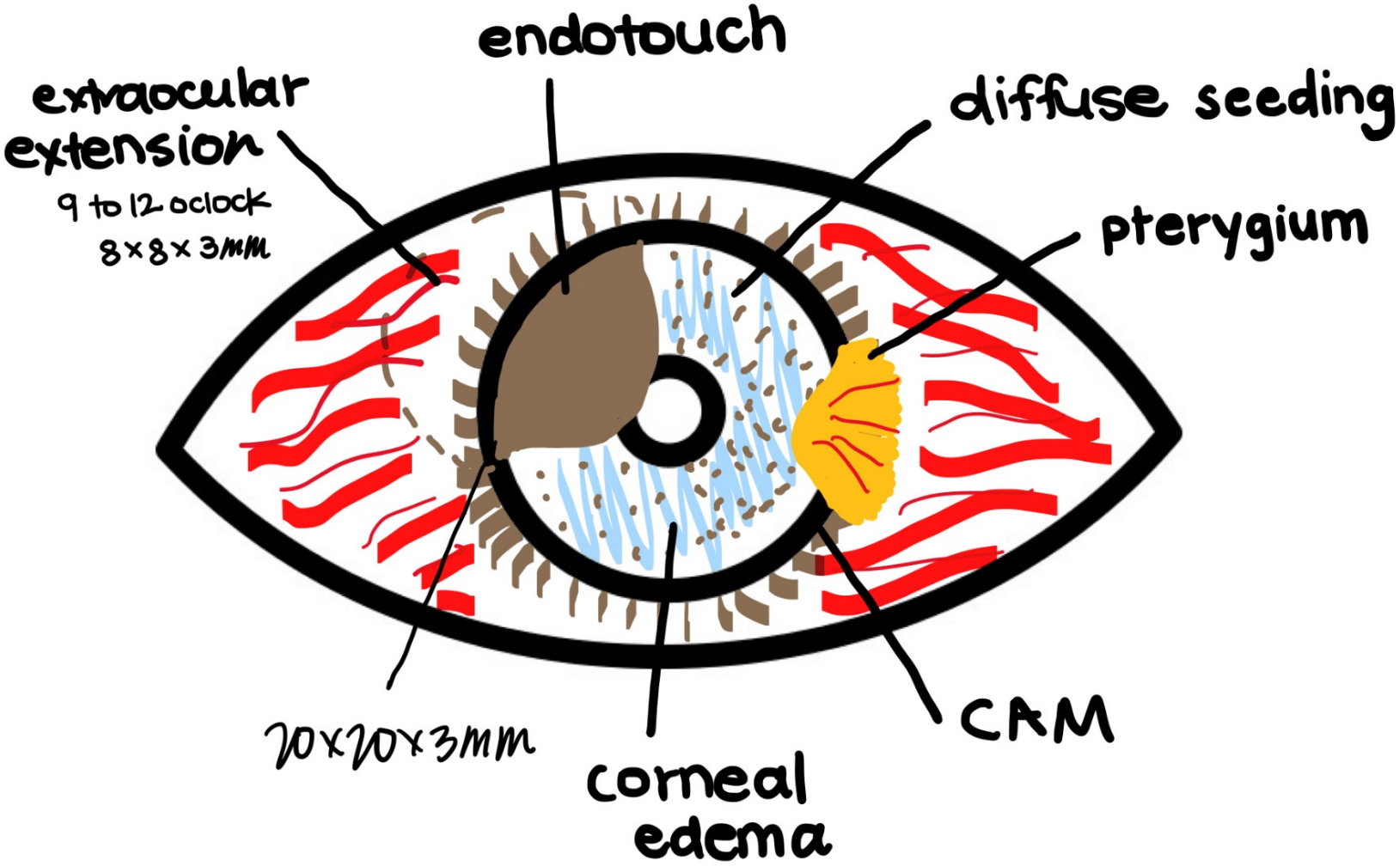
In the Wills ER, IOP OS was lowered from 34 to 26 with 3 rounds of brimonidine, dorzolamide, and timolol. His pain improved, and the patient was discharged to follow up in the Wills Ocular Oncology Clinic.

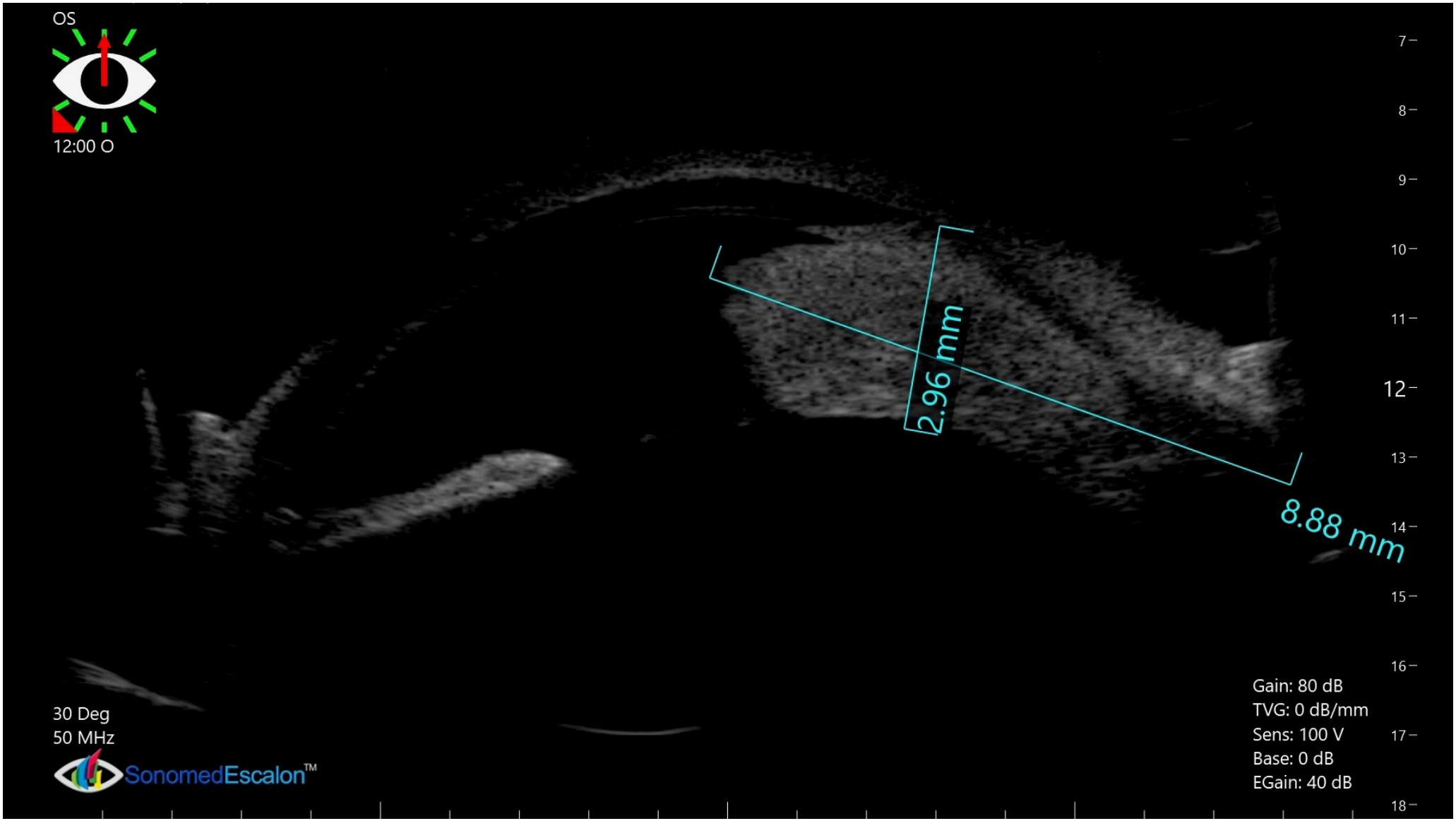
Several weeks later…

VA_{sc}	20/25	EOMs	Full, ortho
	CF @ 2ft		Full, ortho
IOP	12	Pupils	RRL
	34		Sluggish; no APD by reverse

OS

CF @ 2ft


endotouch
extraocular extension
9 to 12 oclock
8x8x3mm
diffuse seeding
pterygium
20x20x3mm
corneal edema
CAM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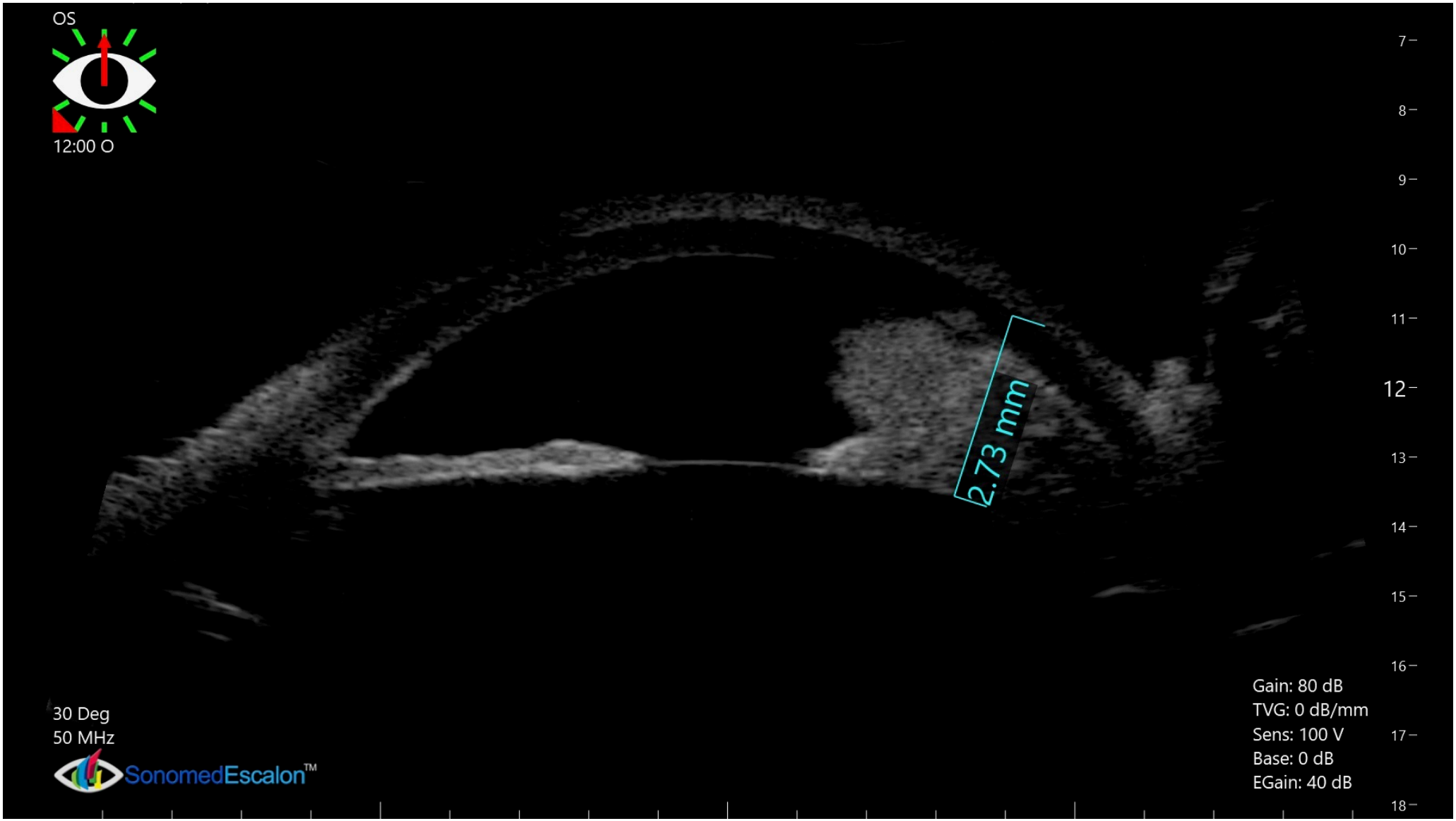
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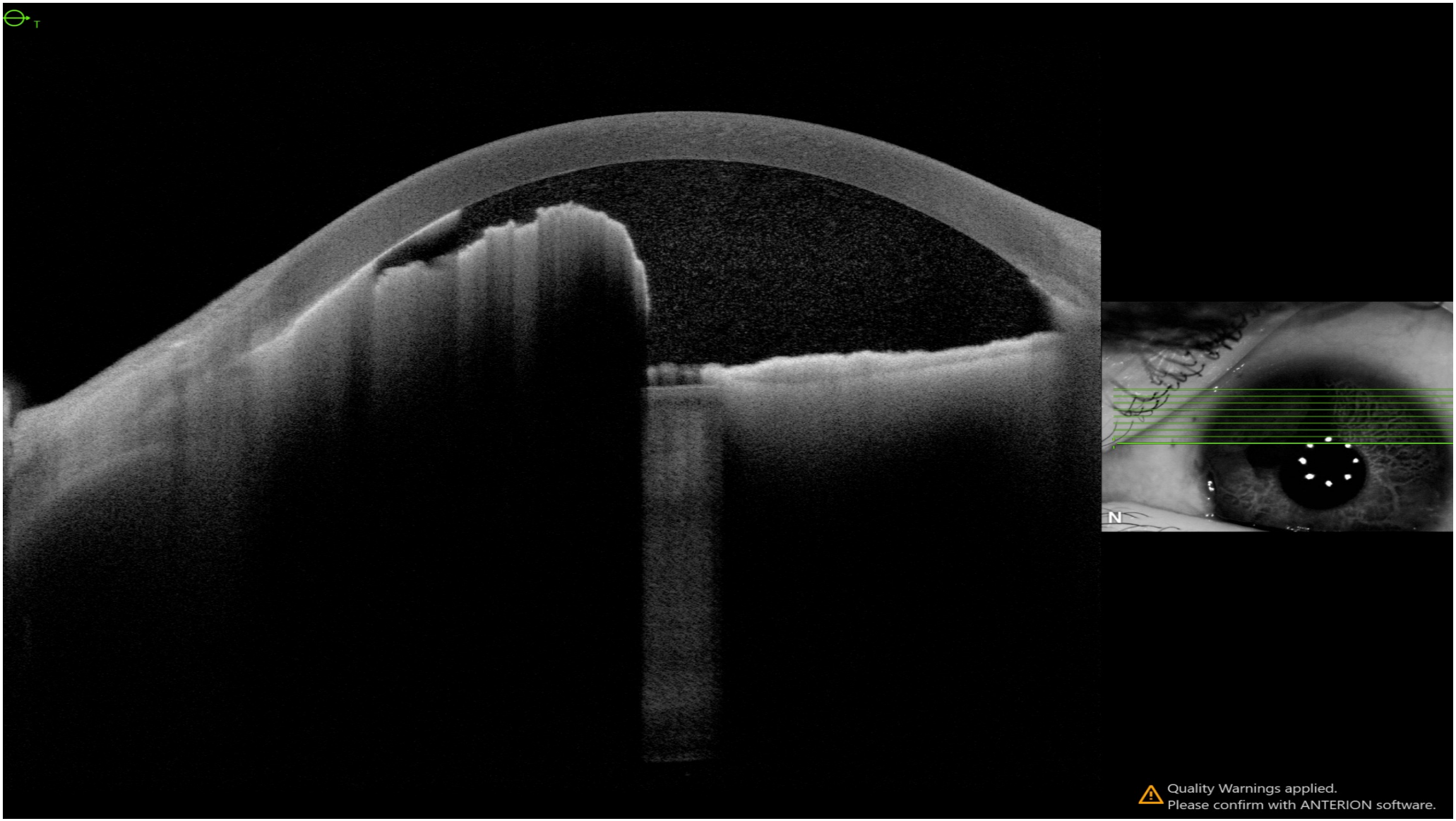
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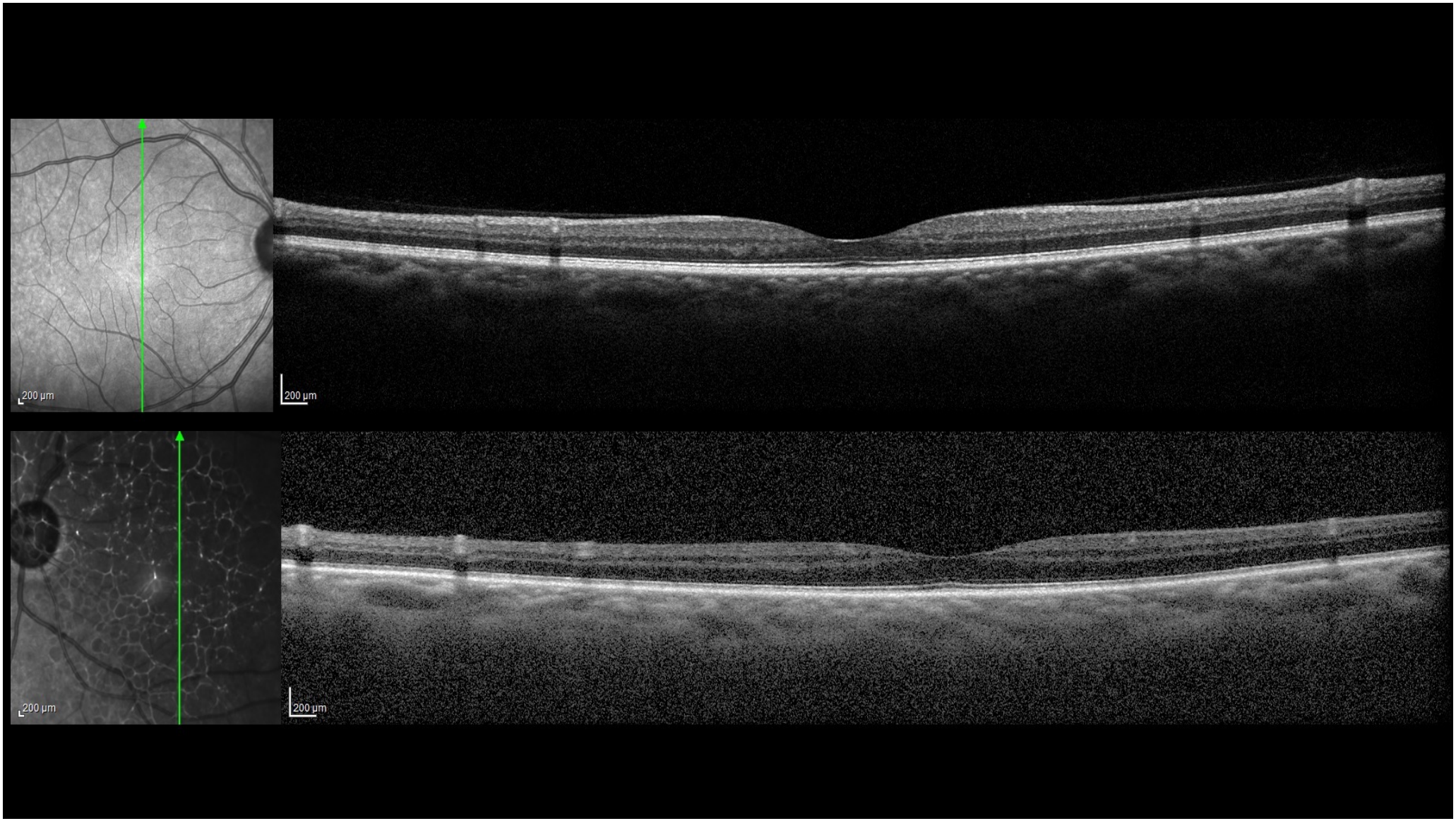
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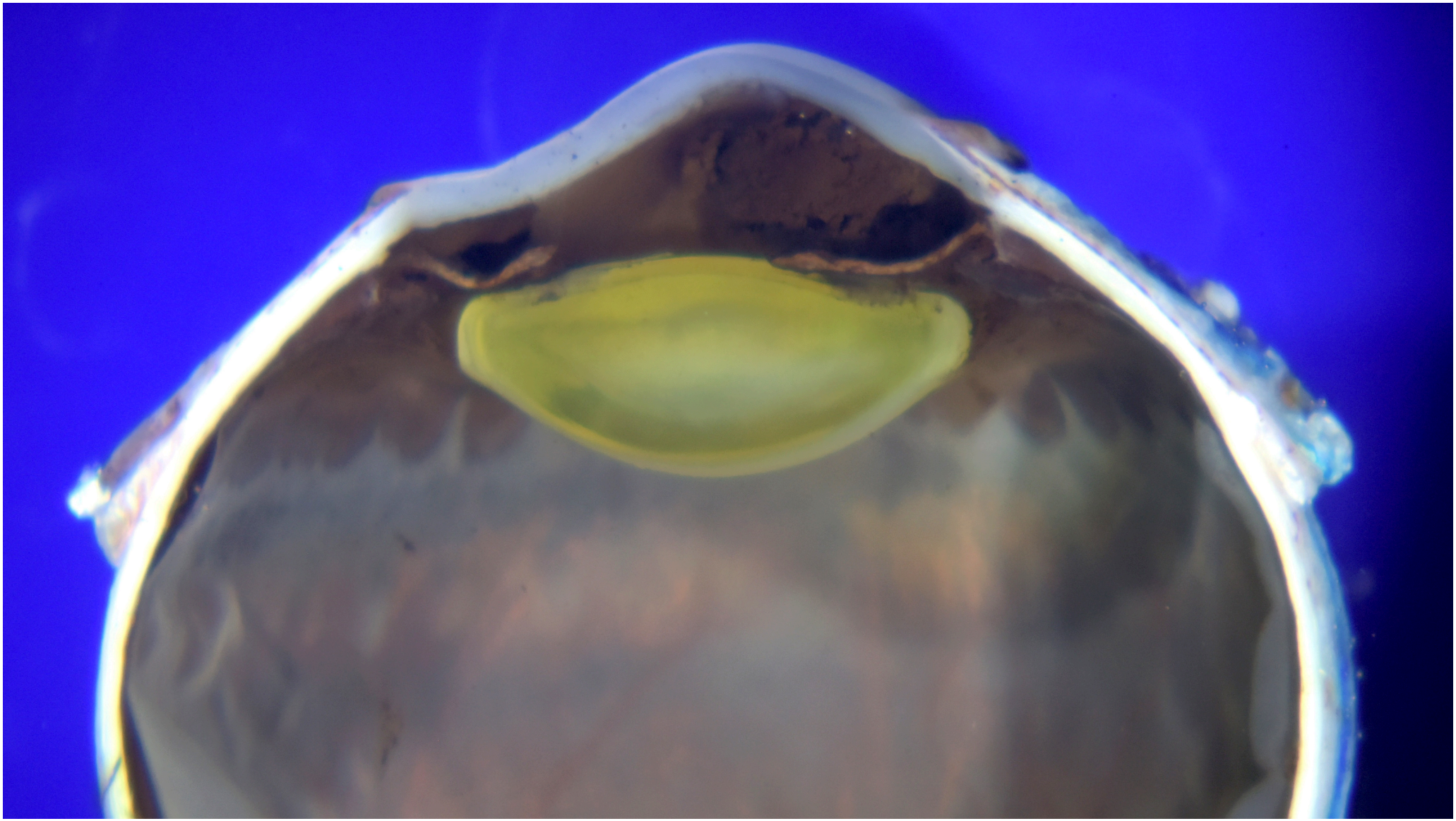
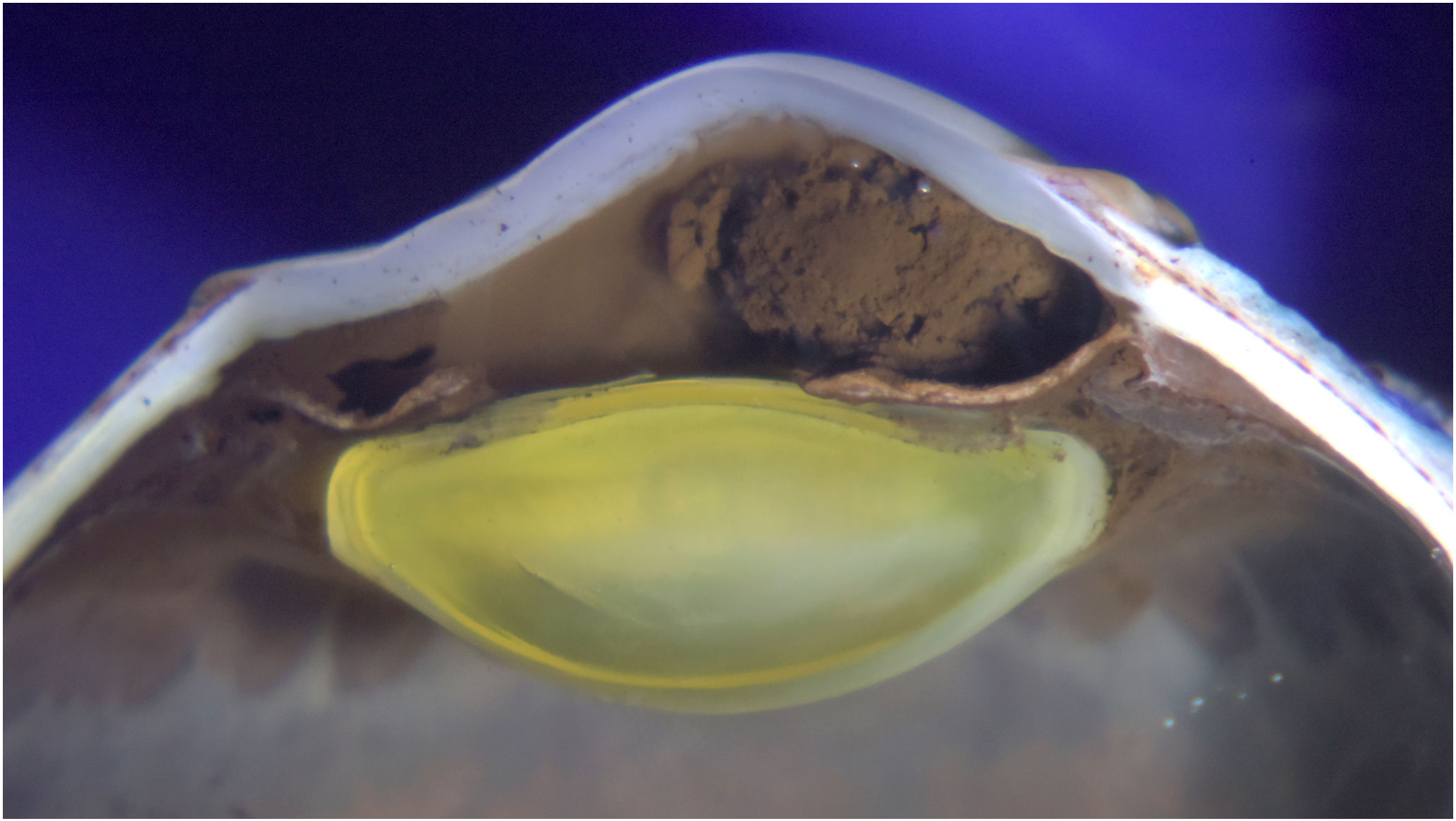
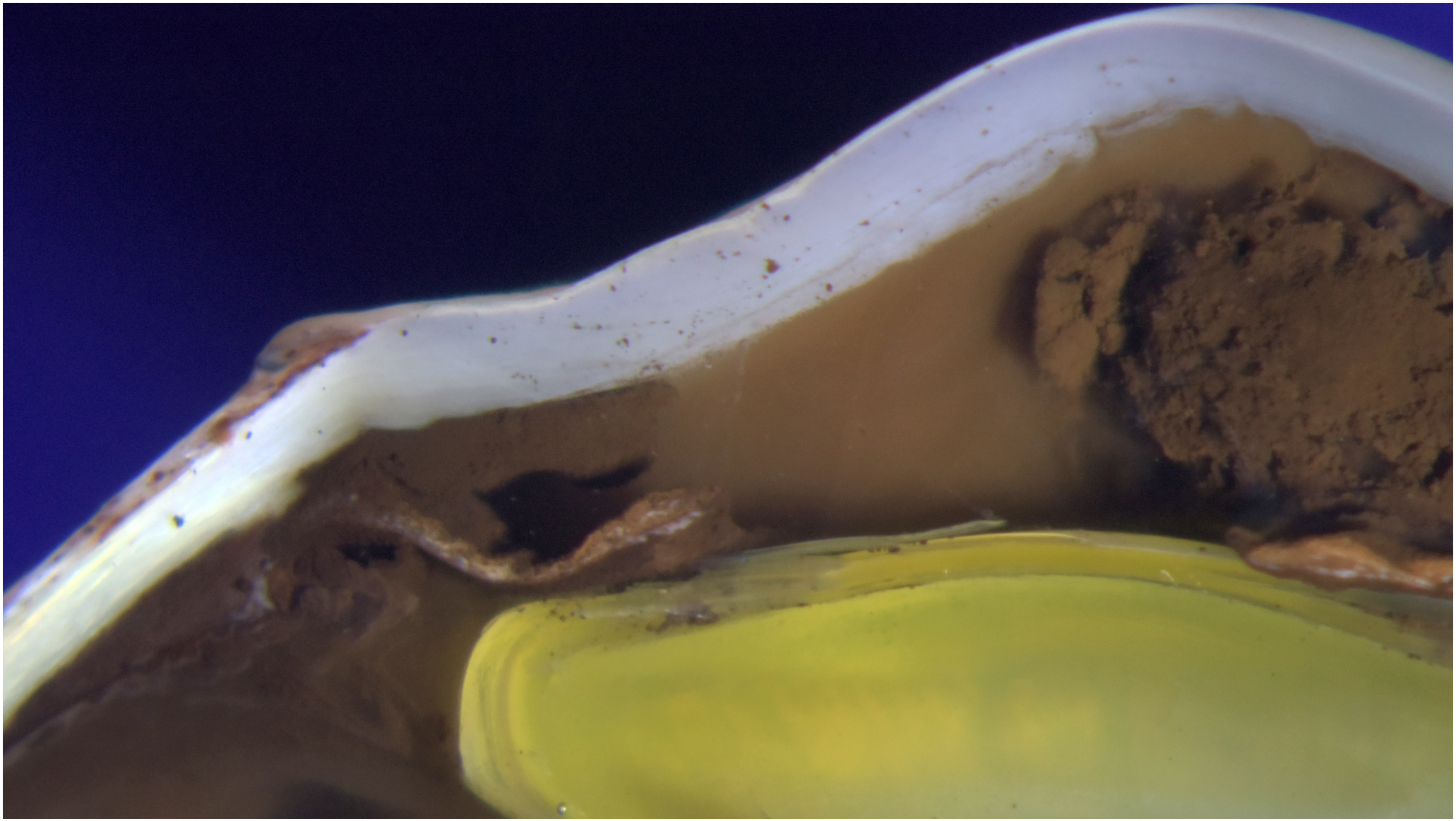
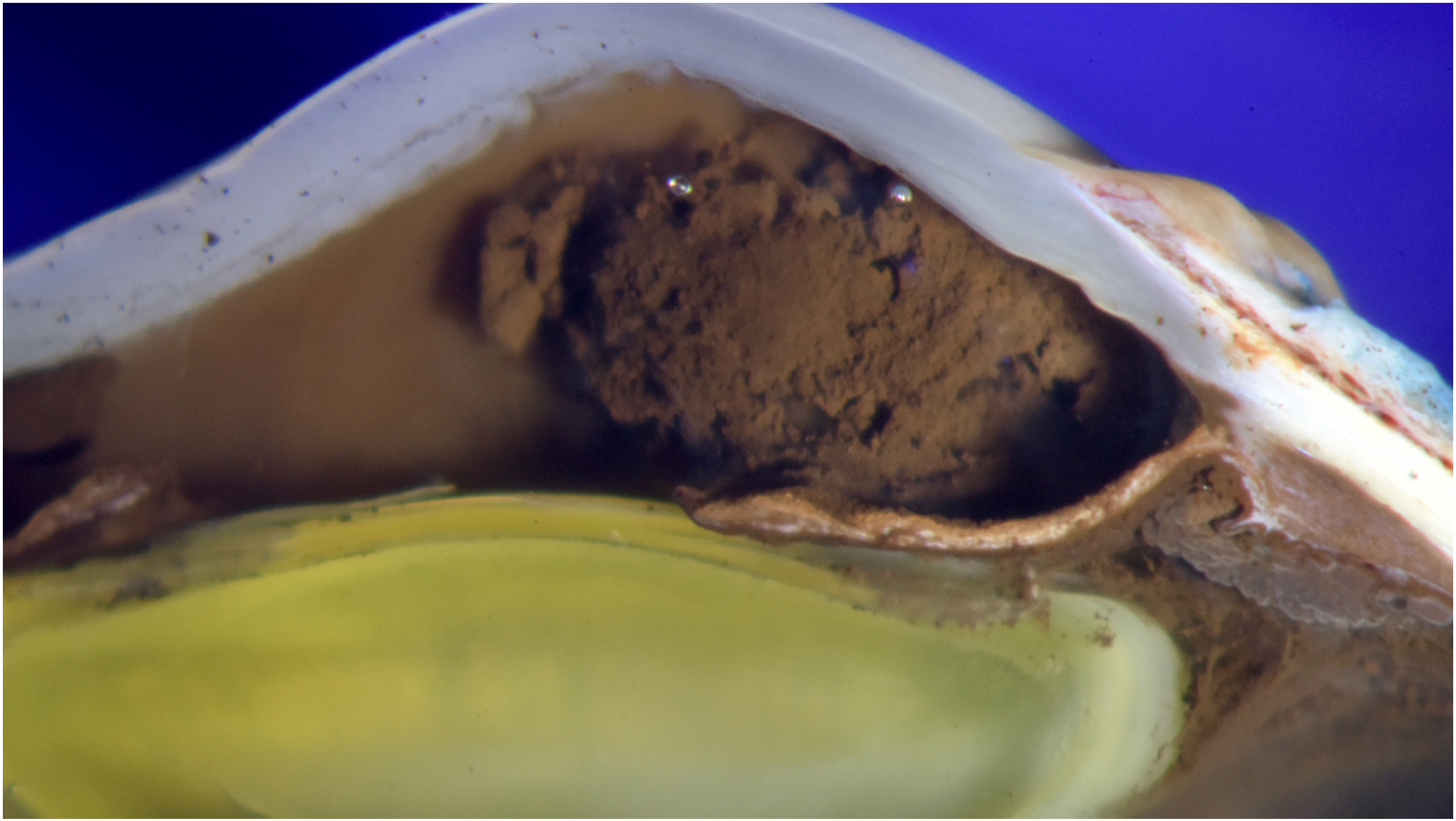
DDx?

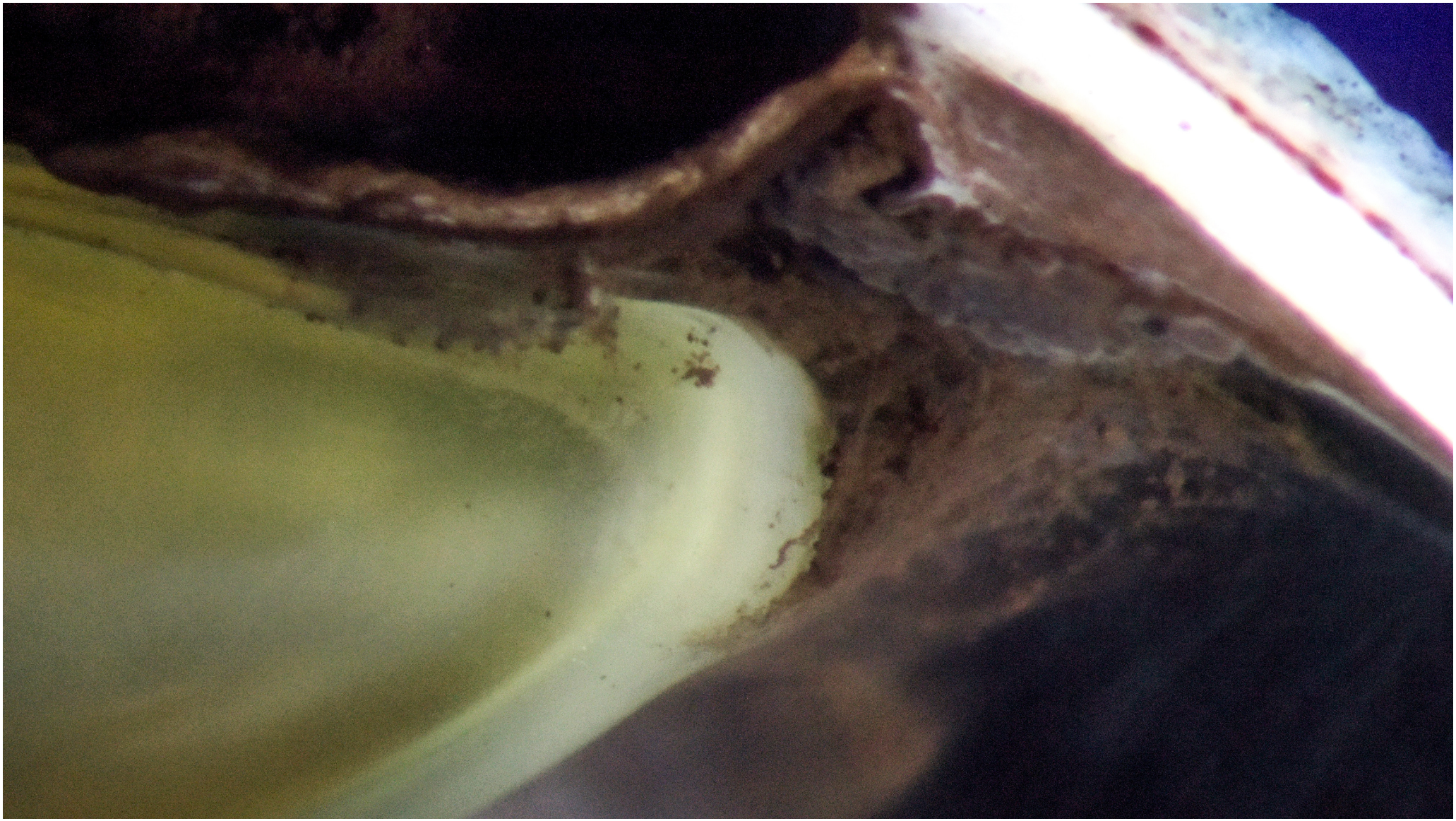
Next steps?

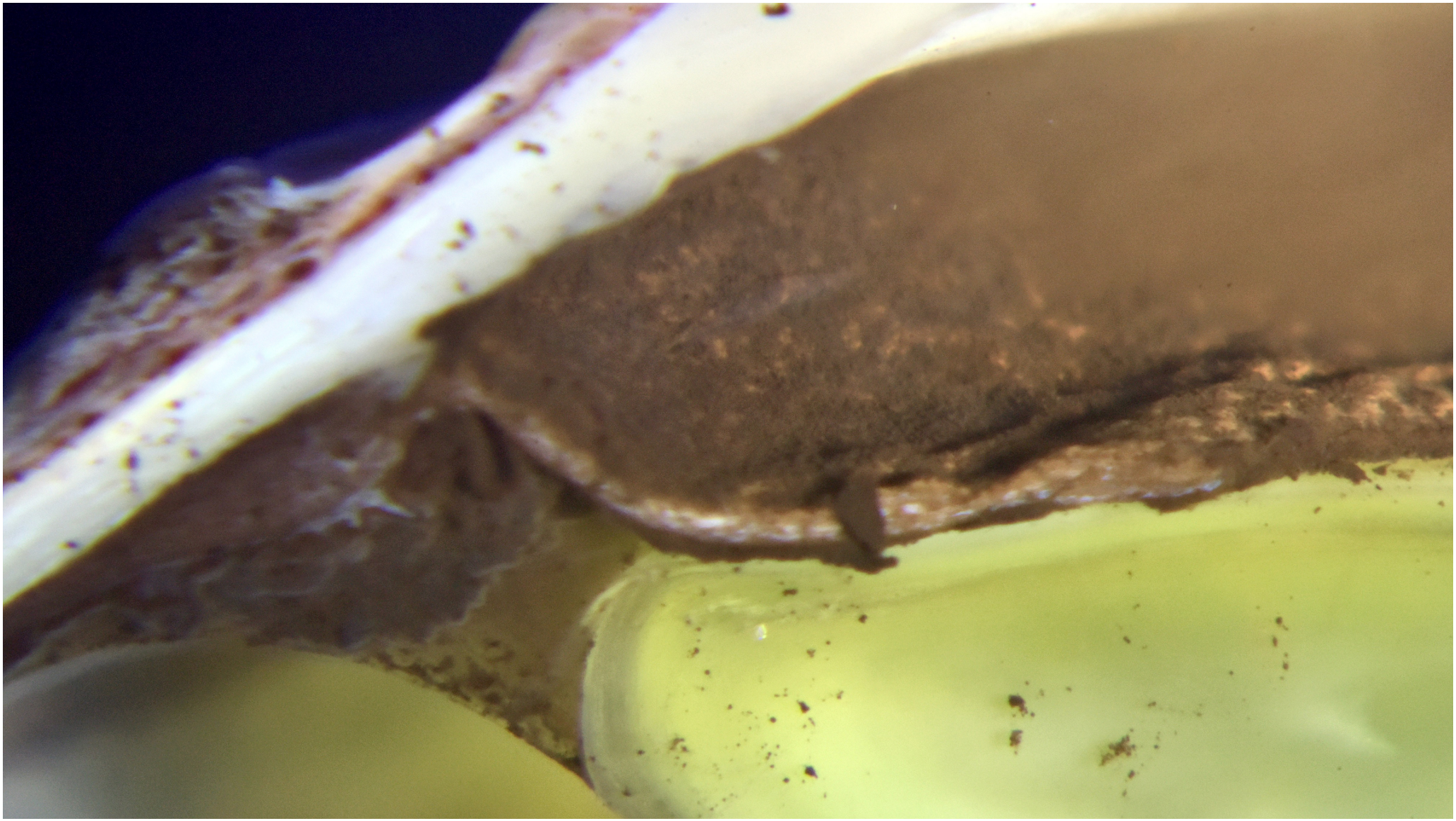


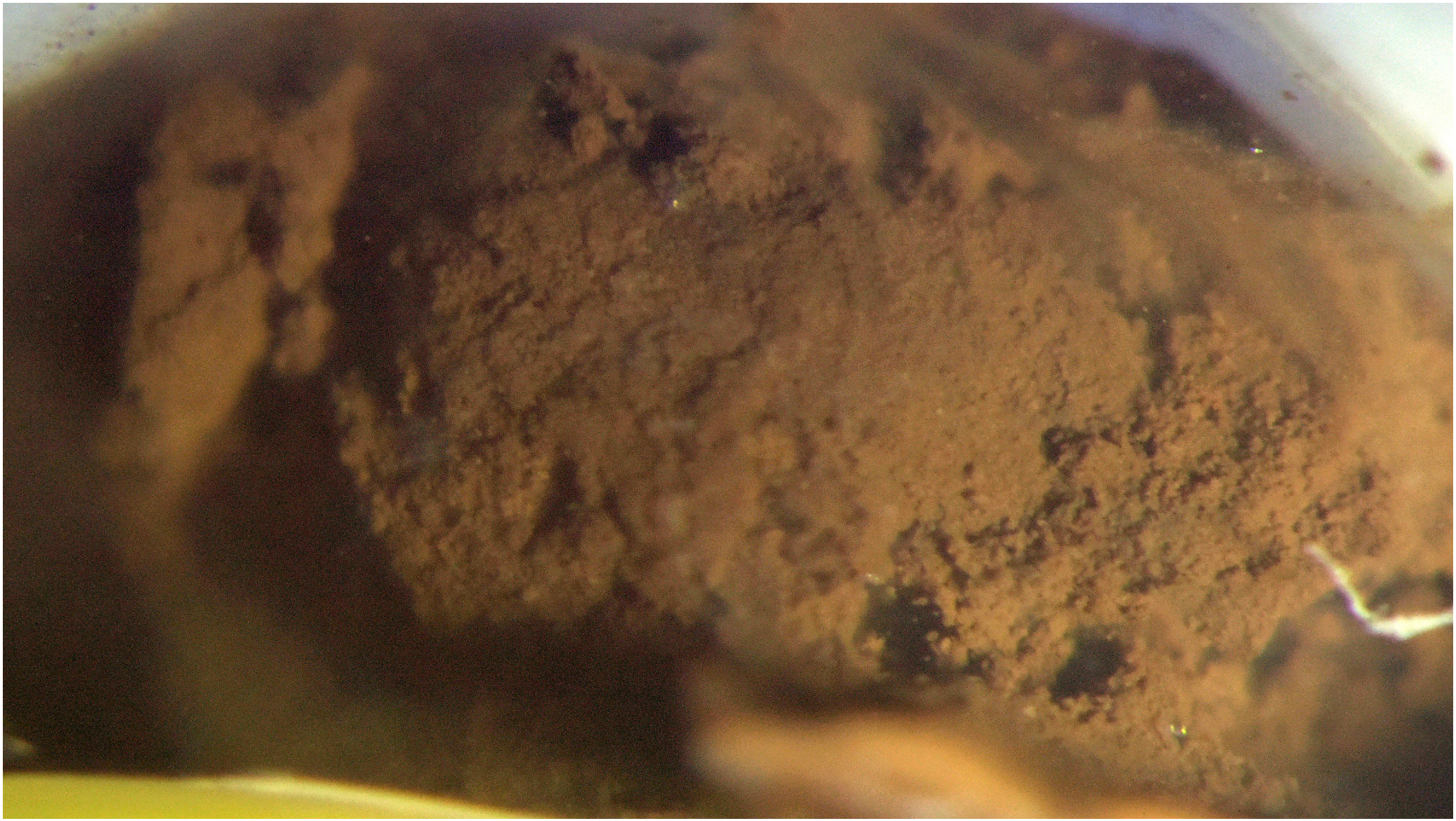


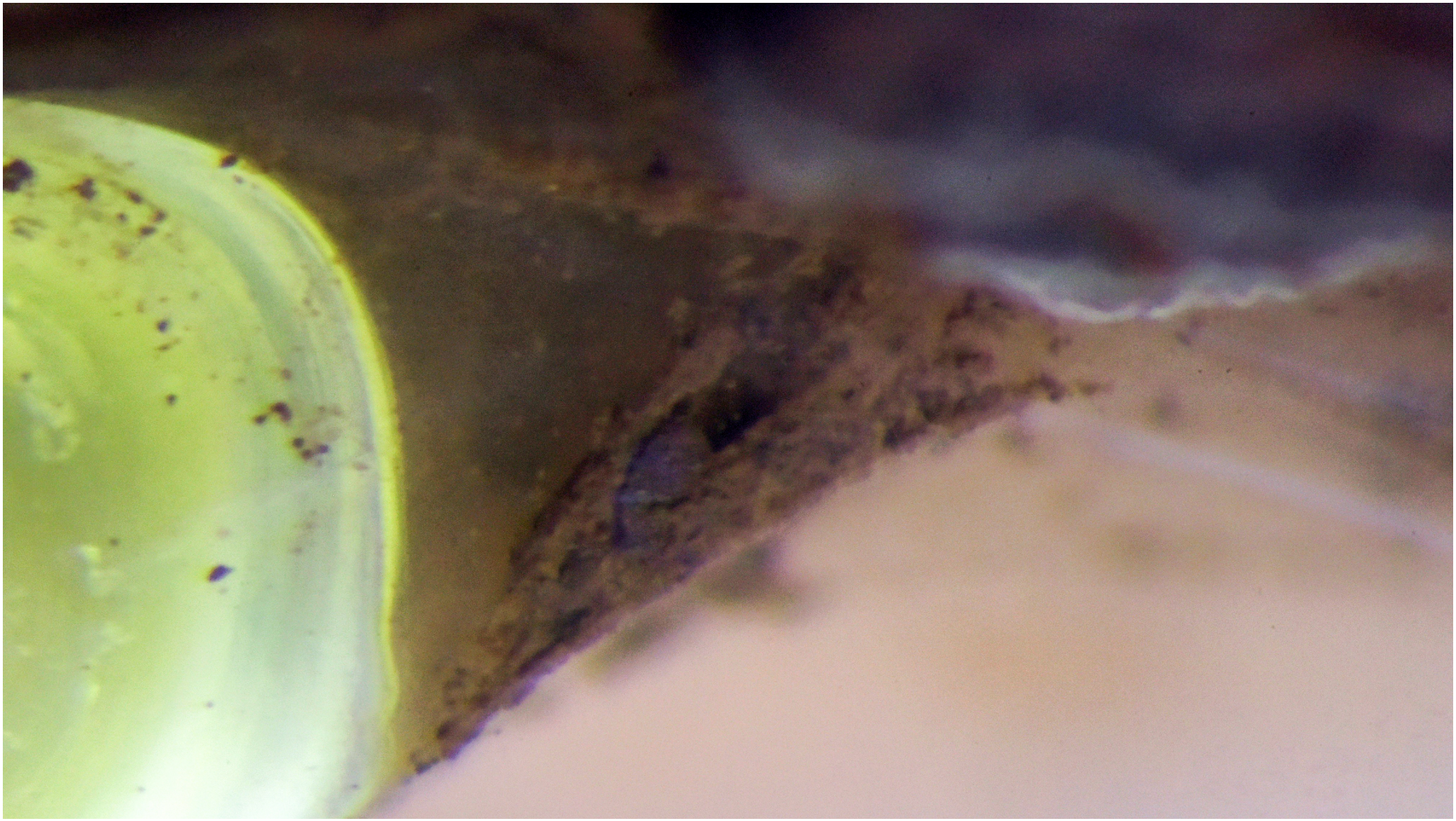


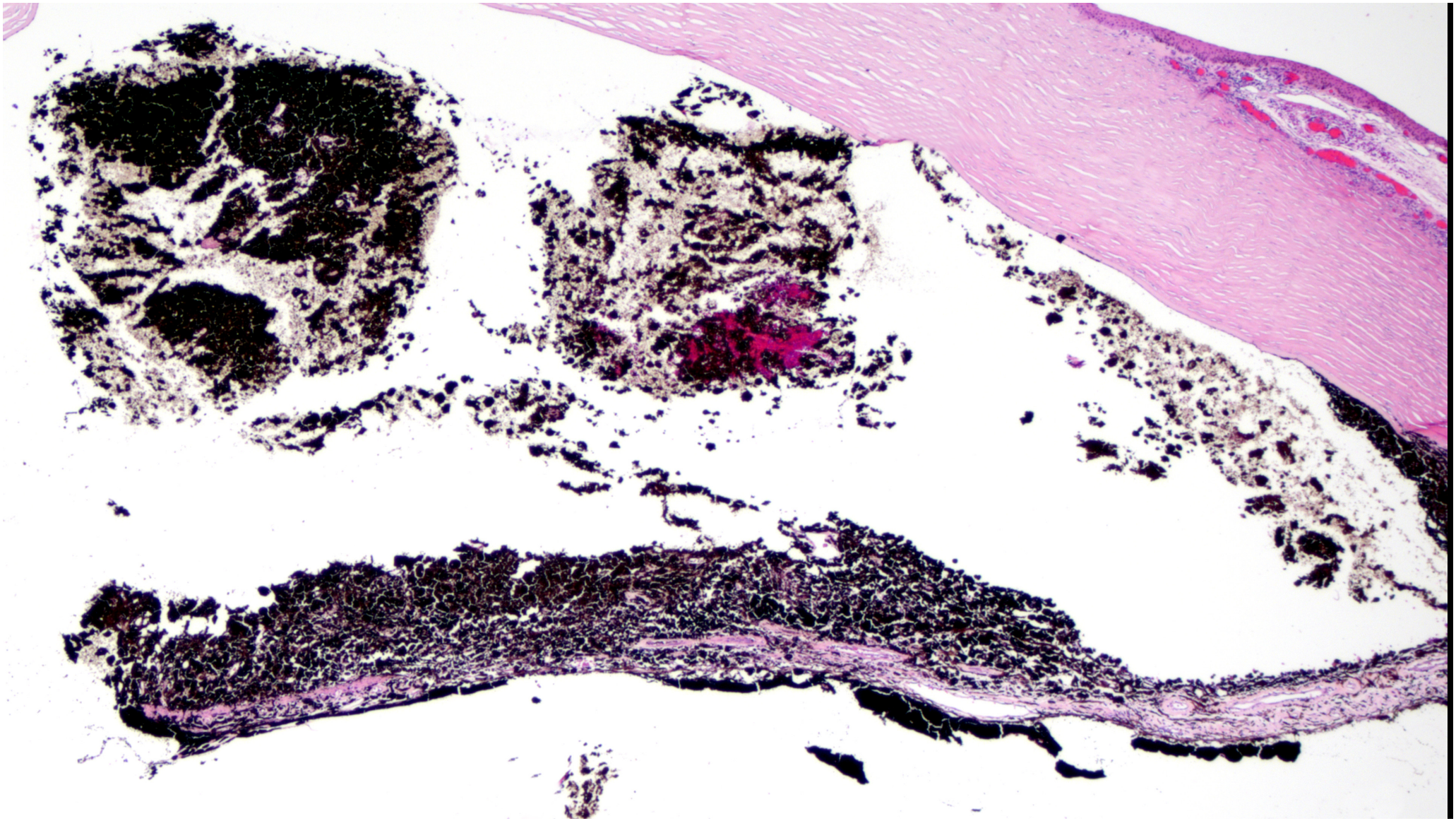


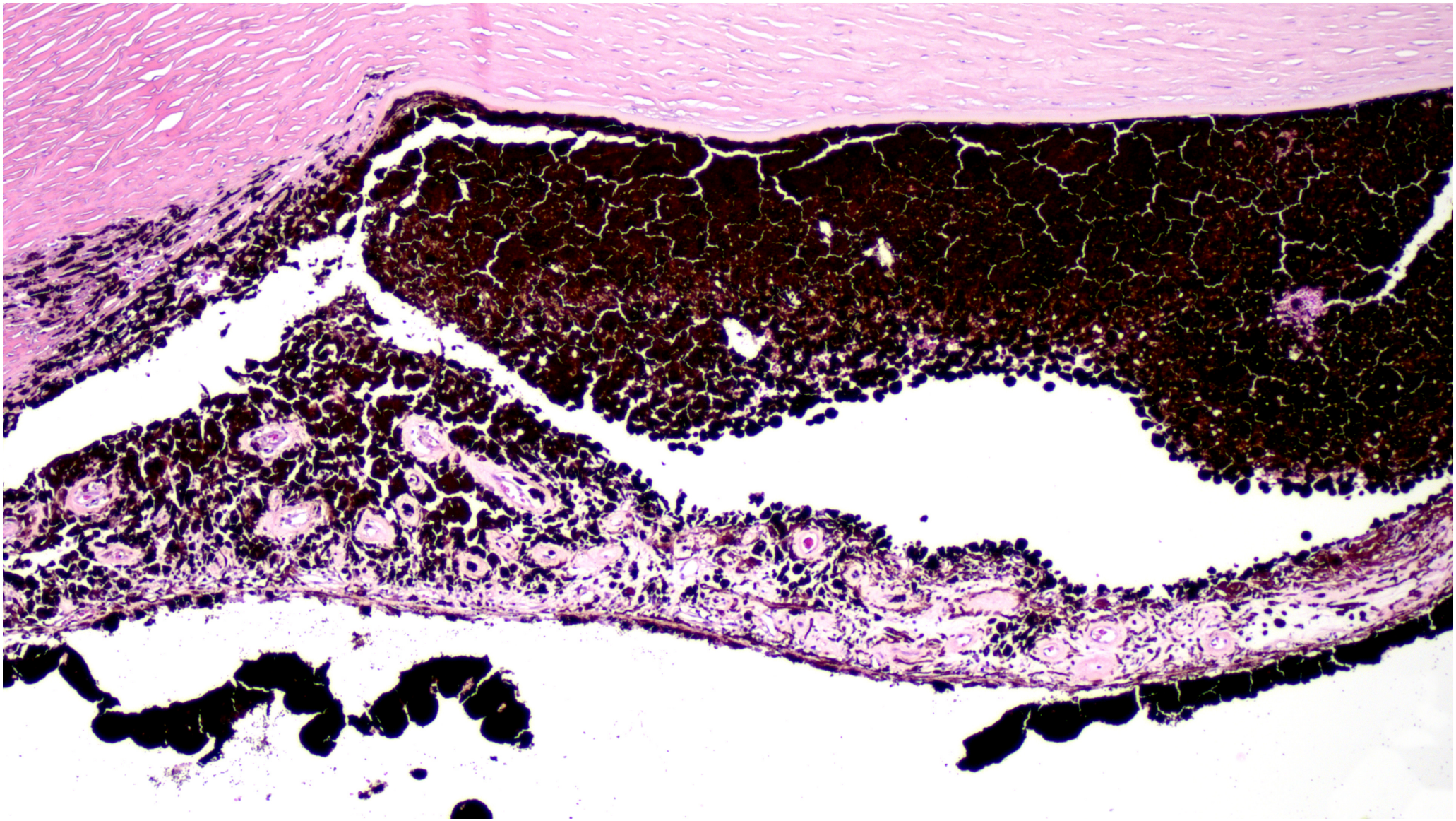


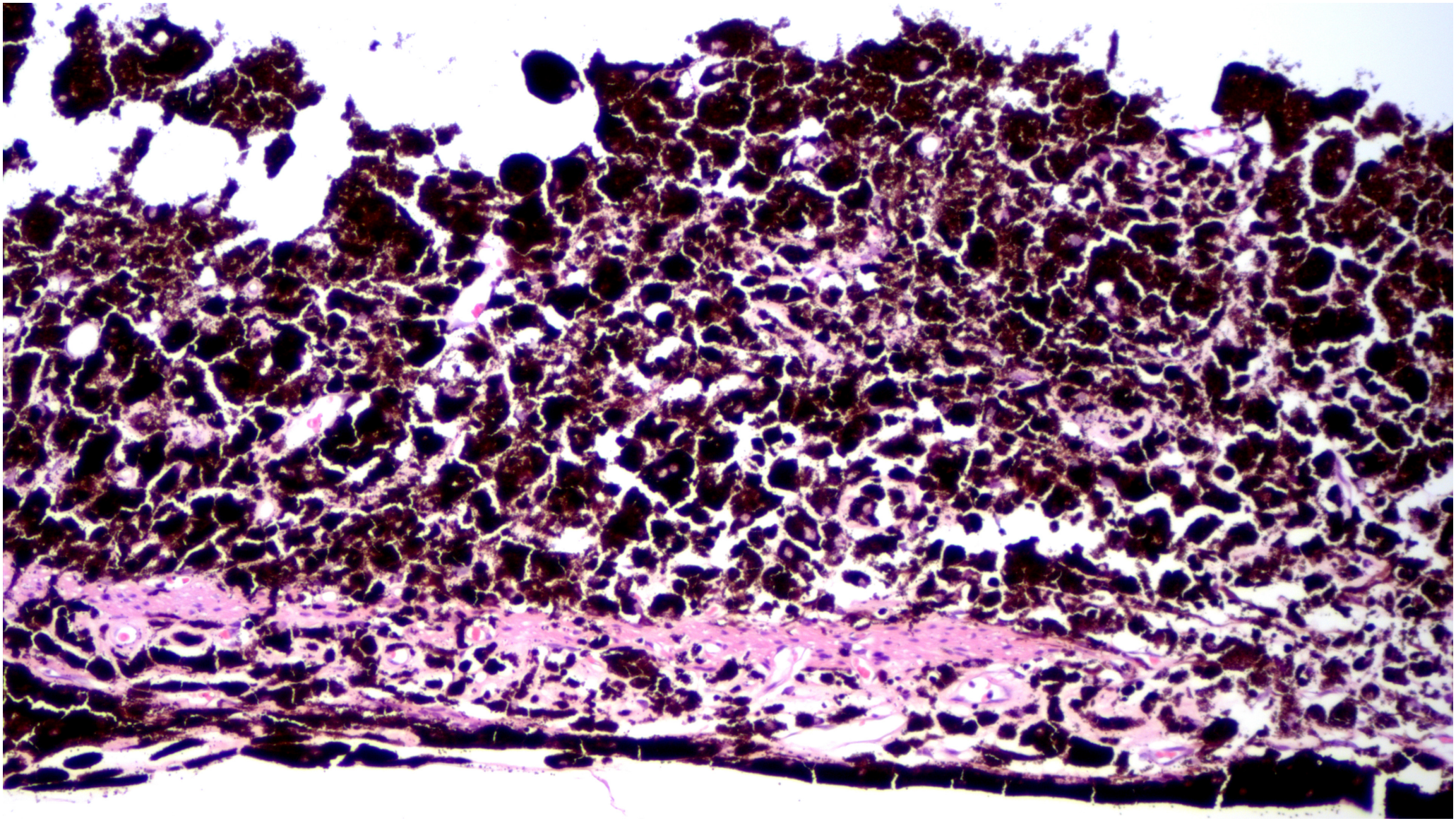


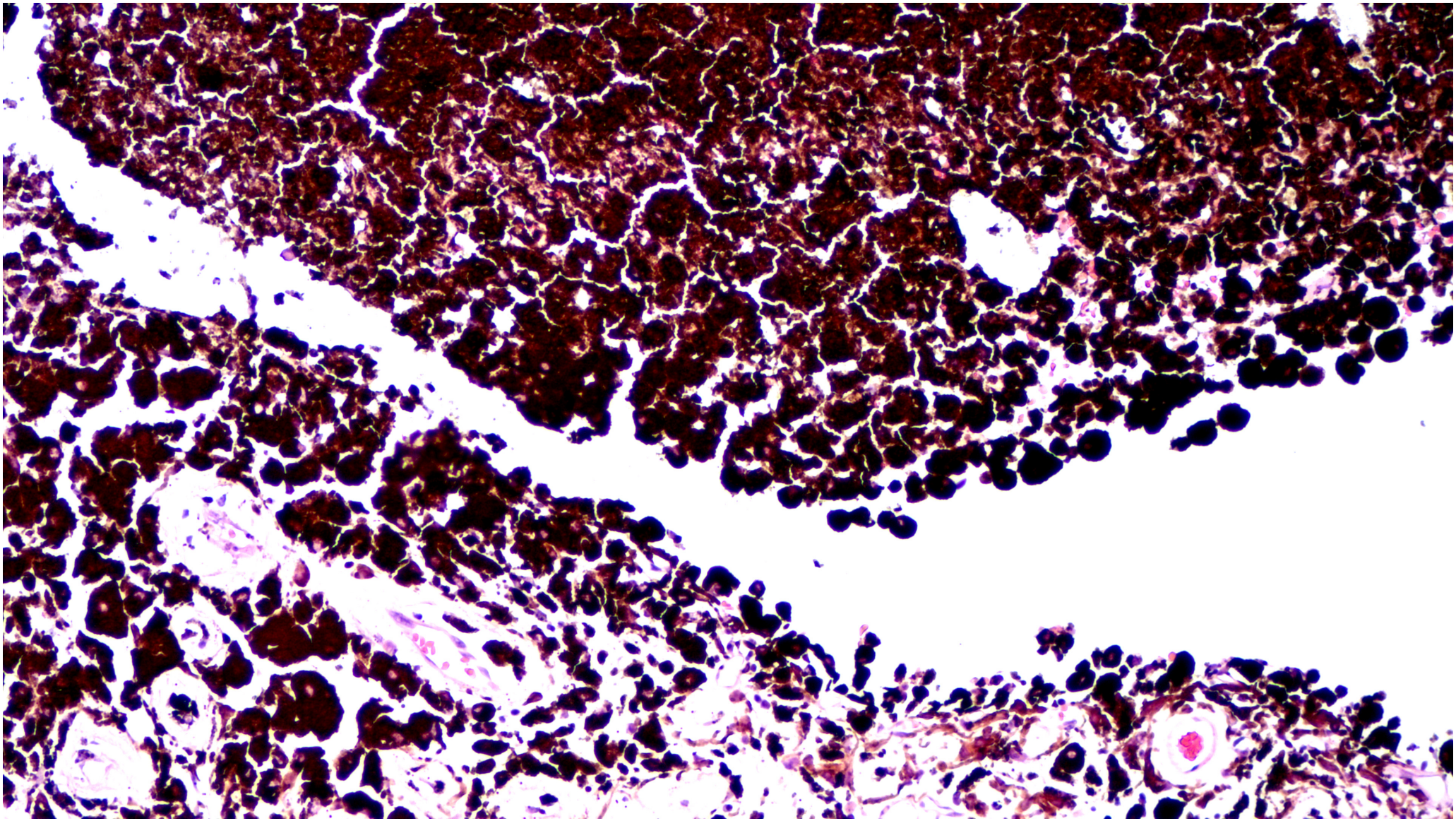


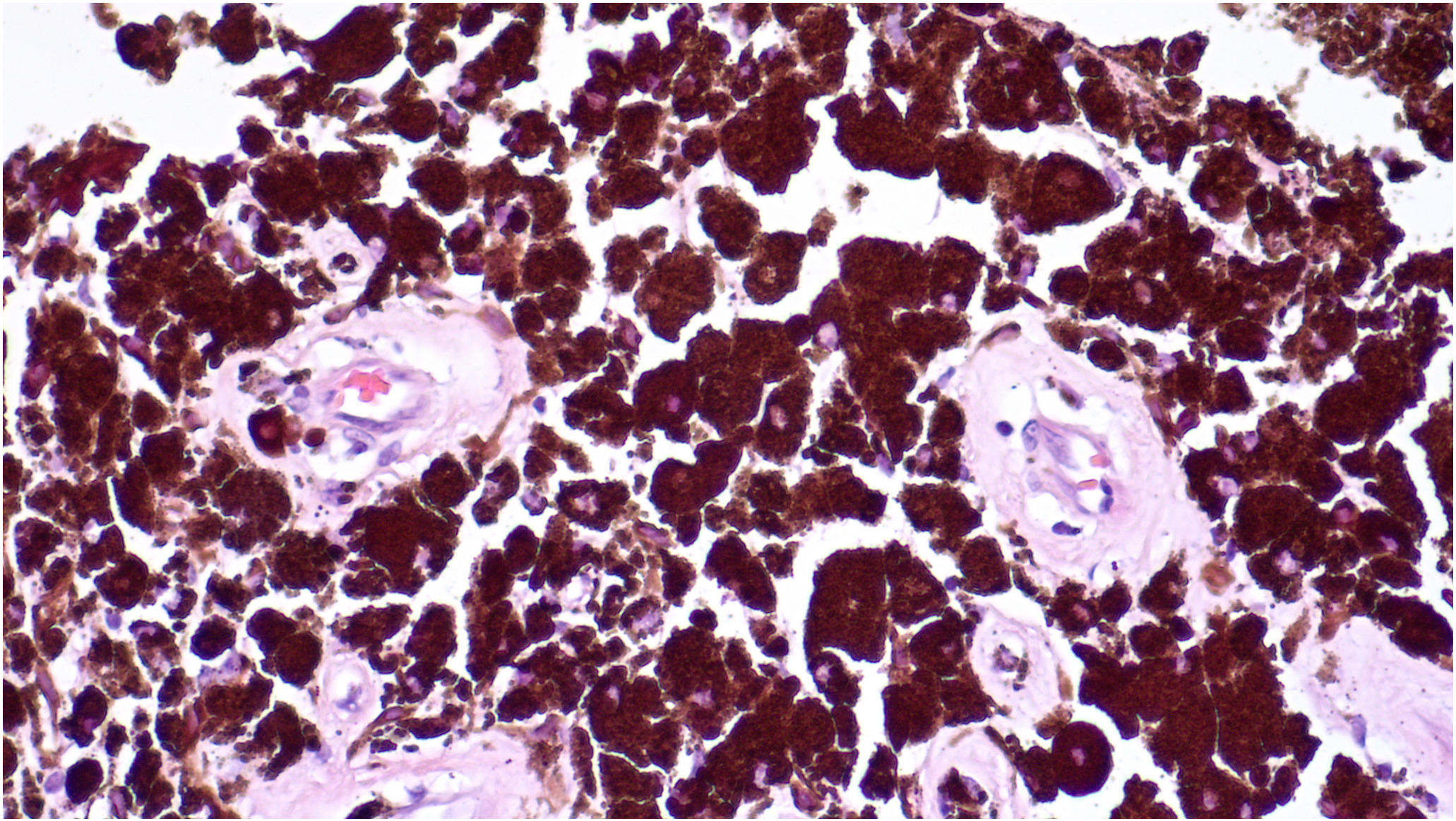


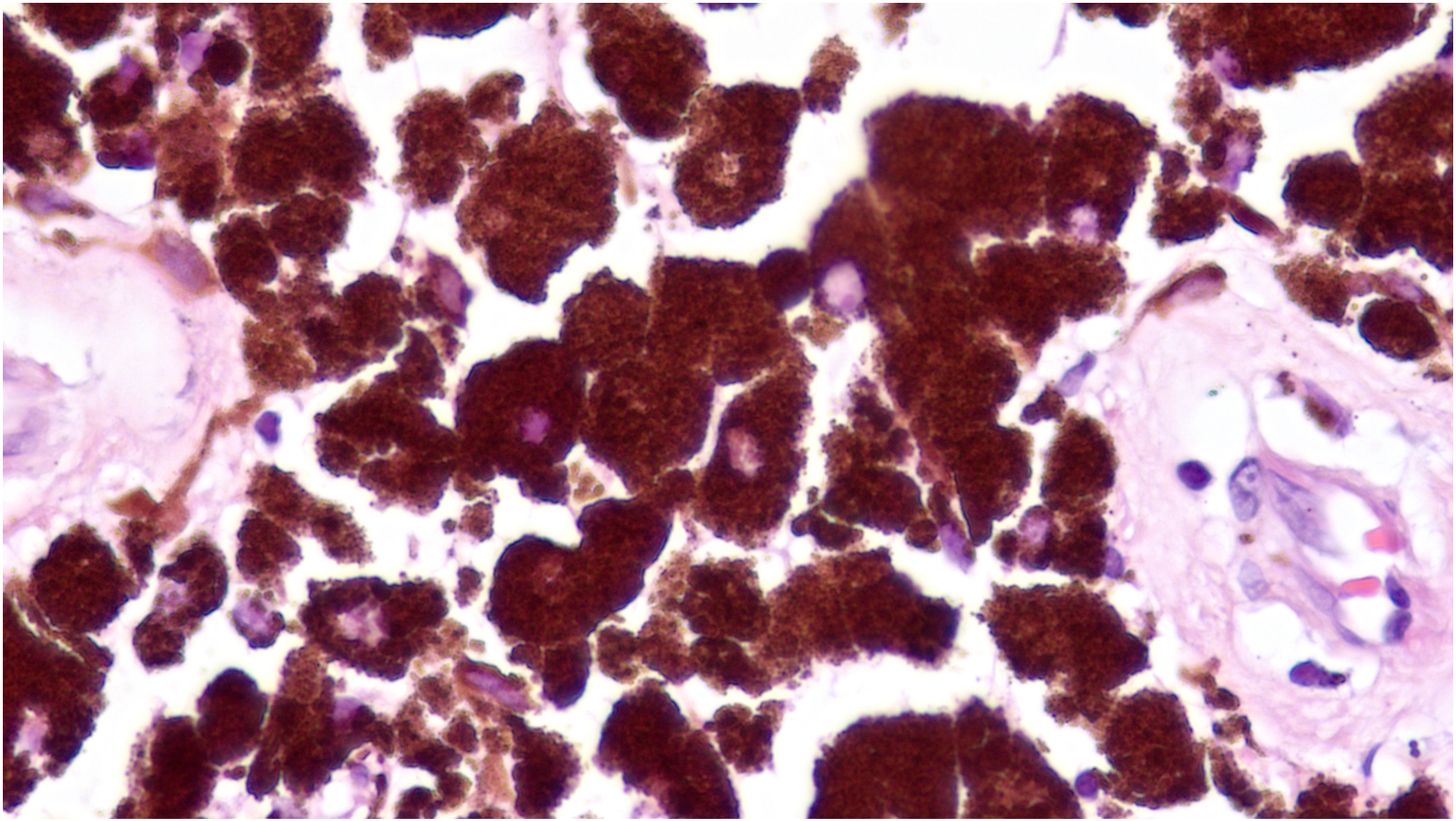


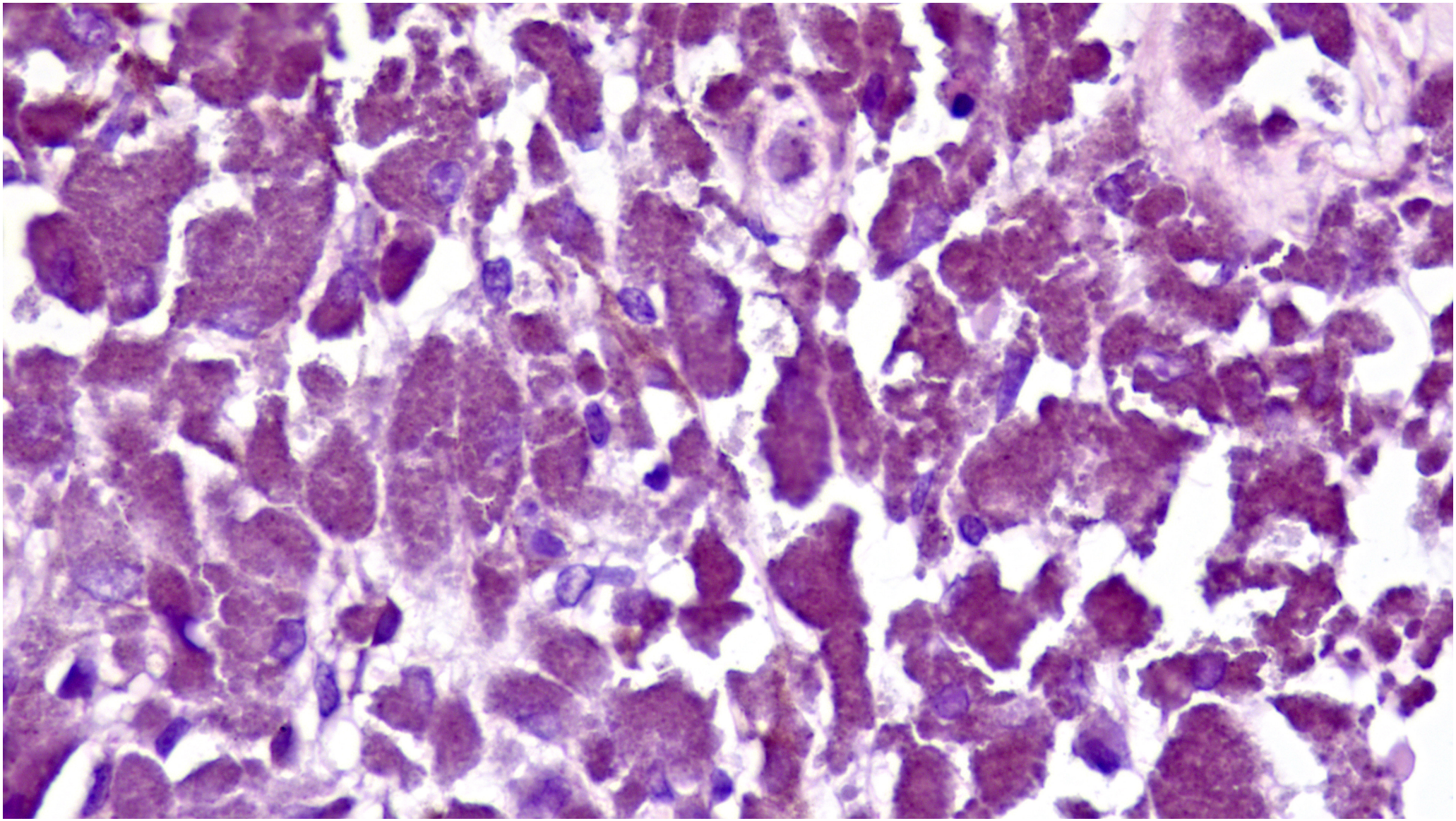


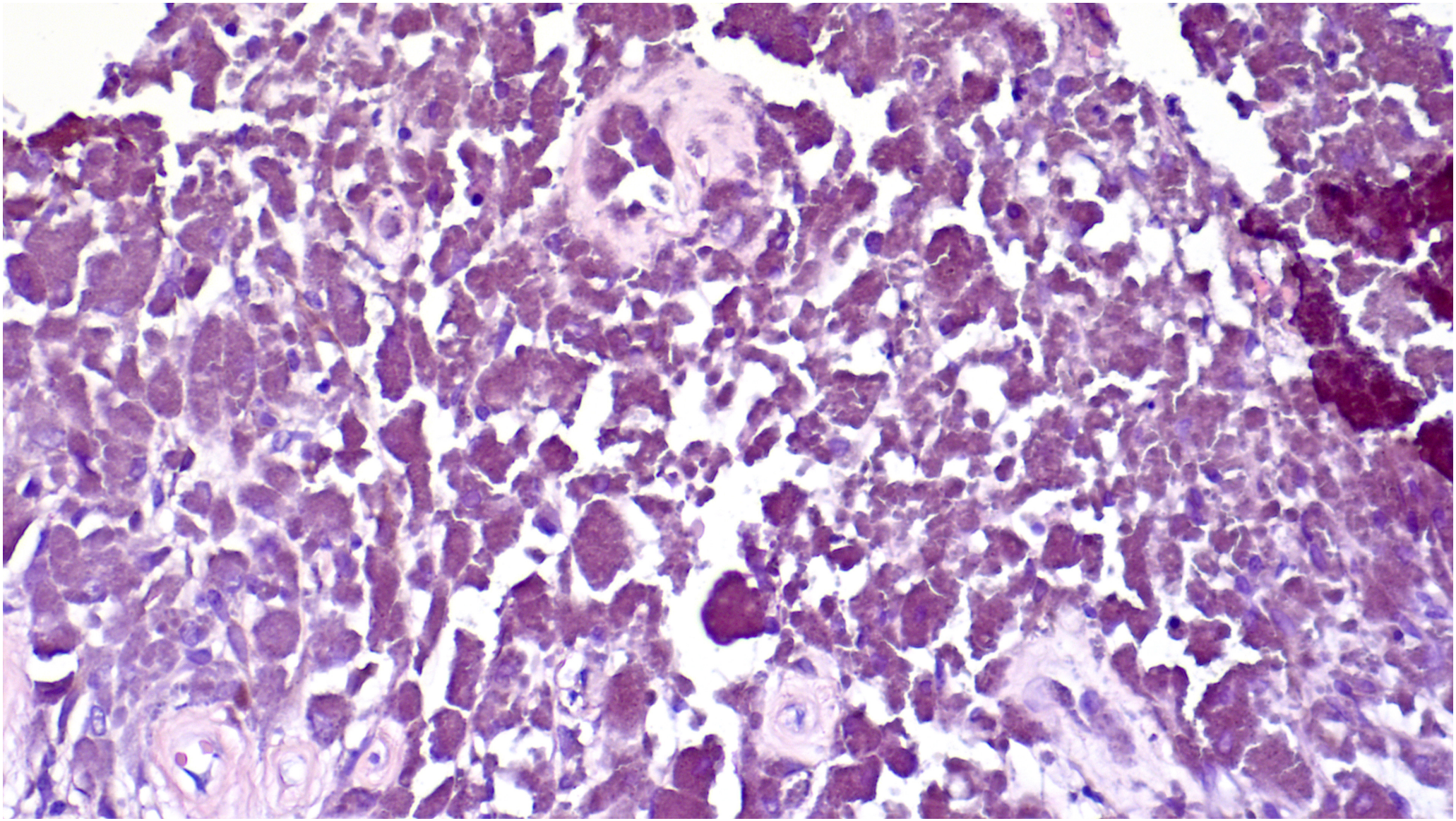


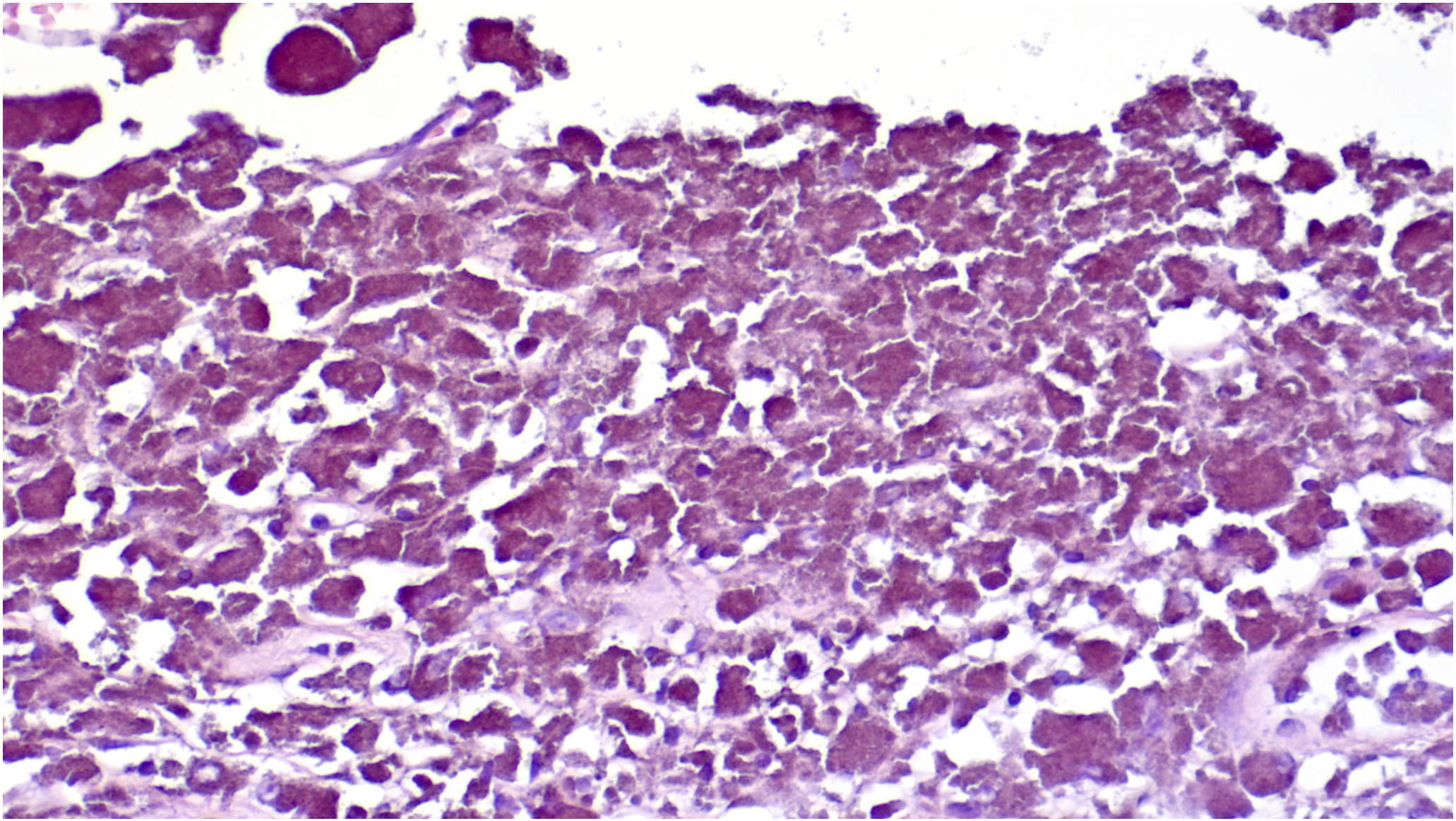


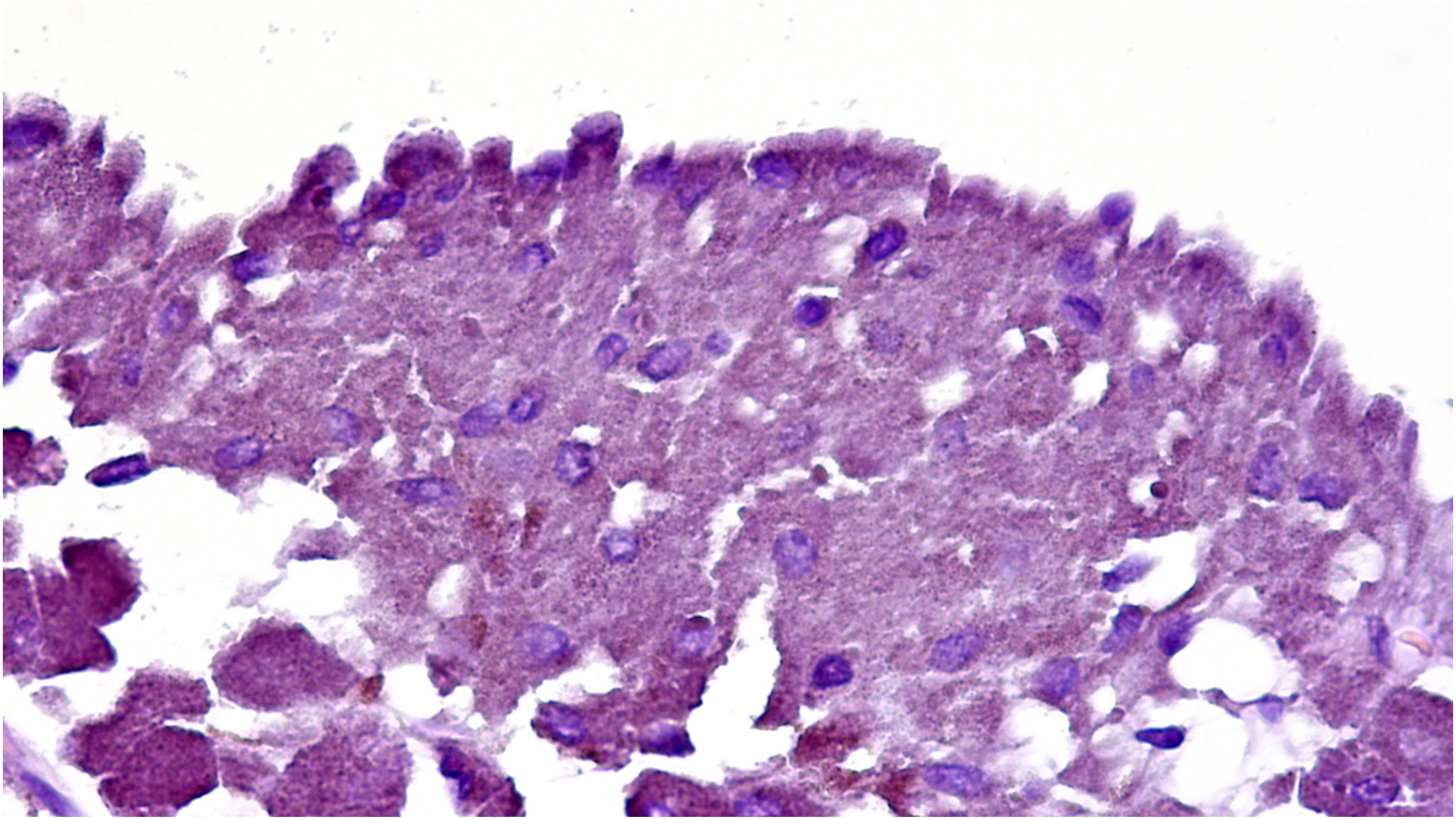


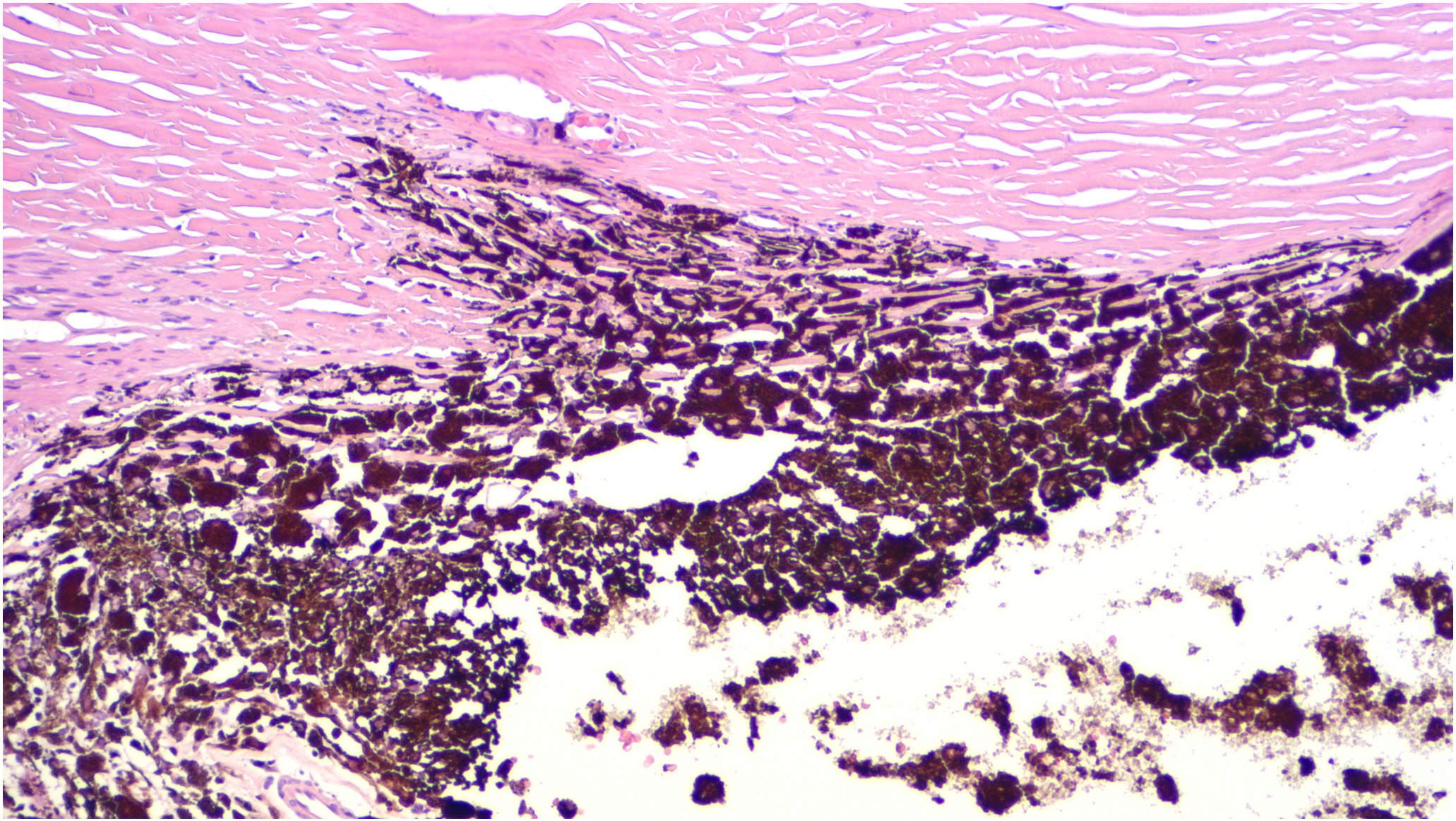


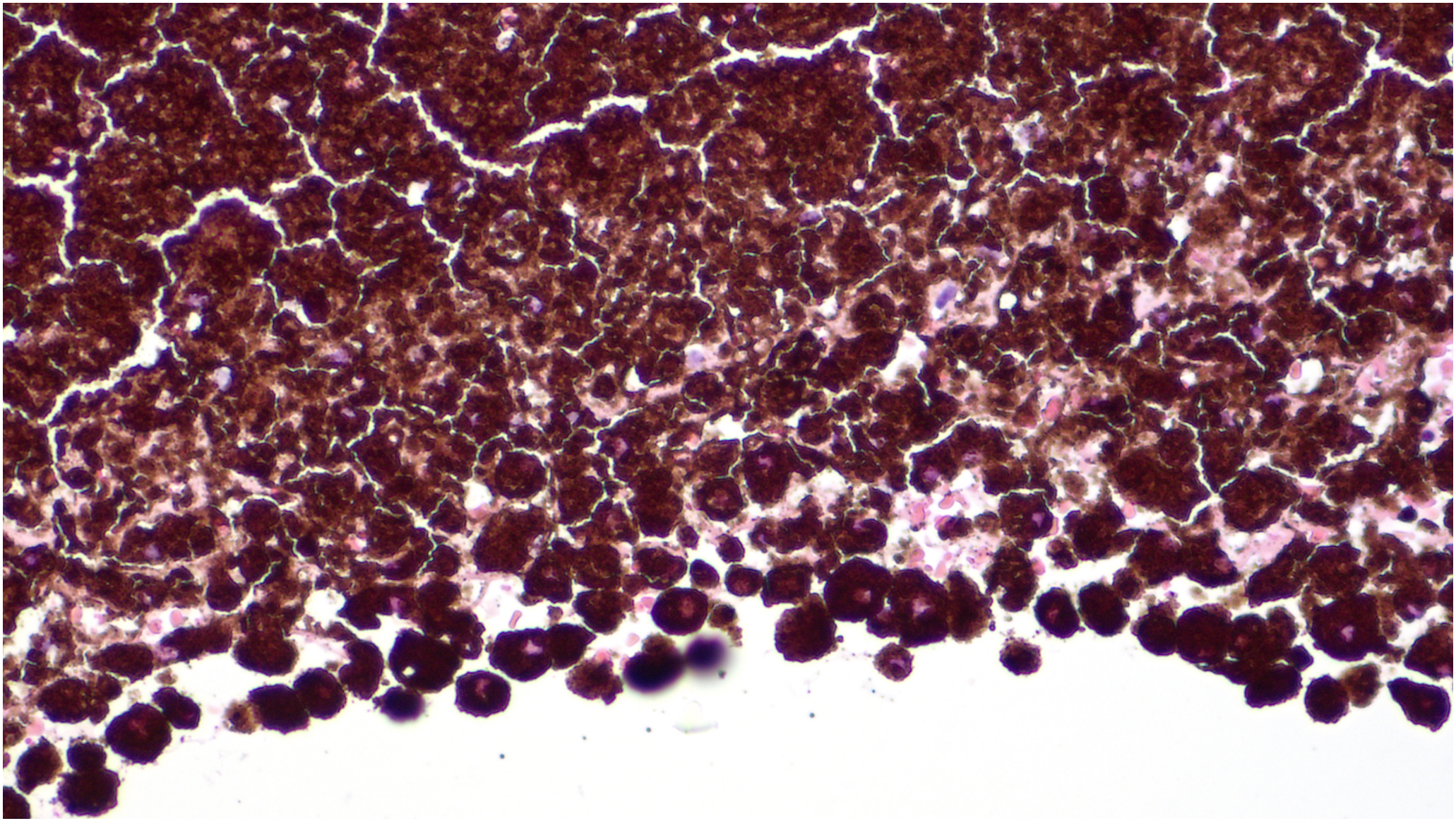


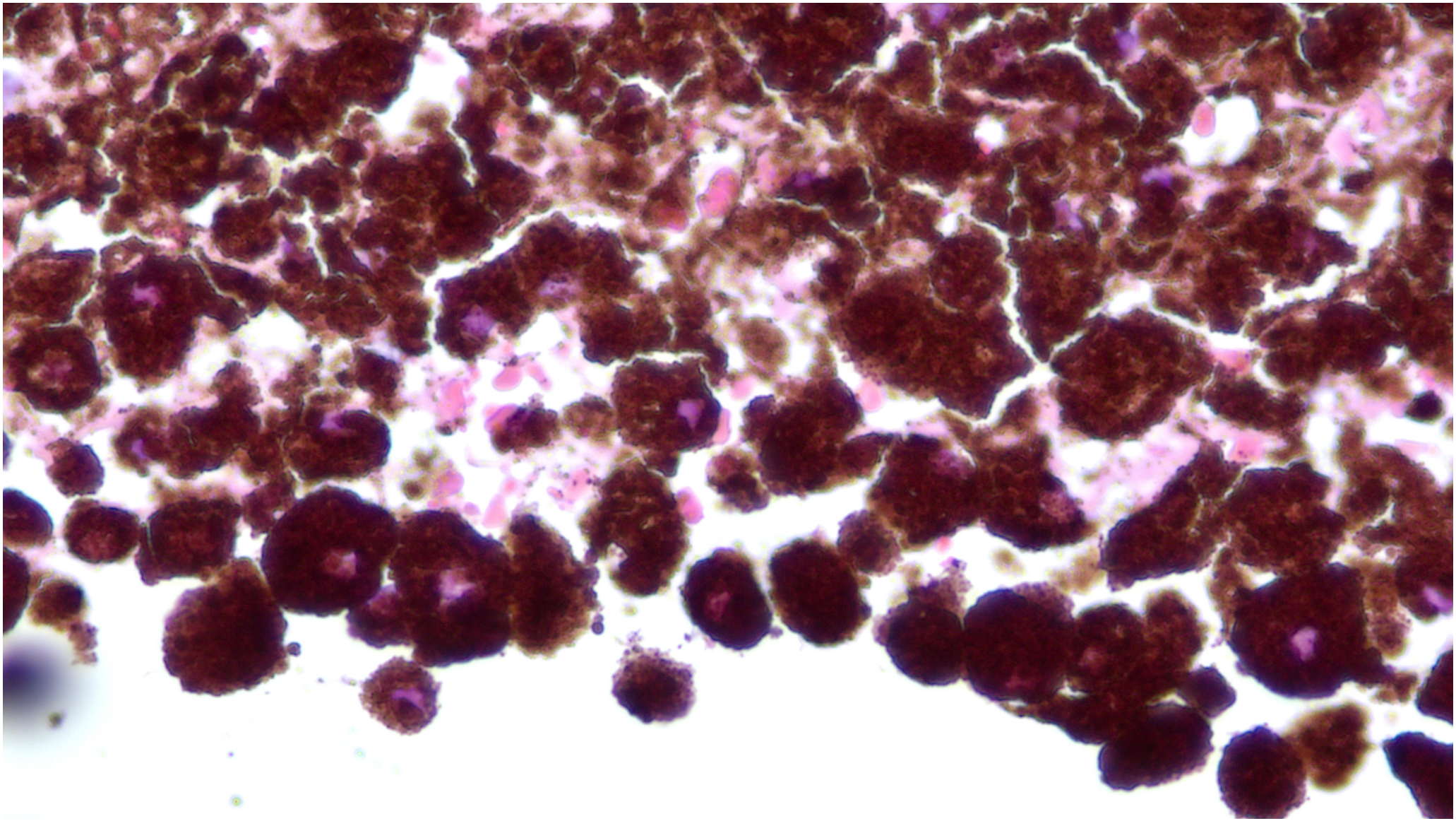


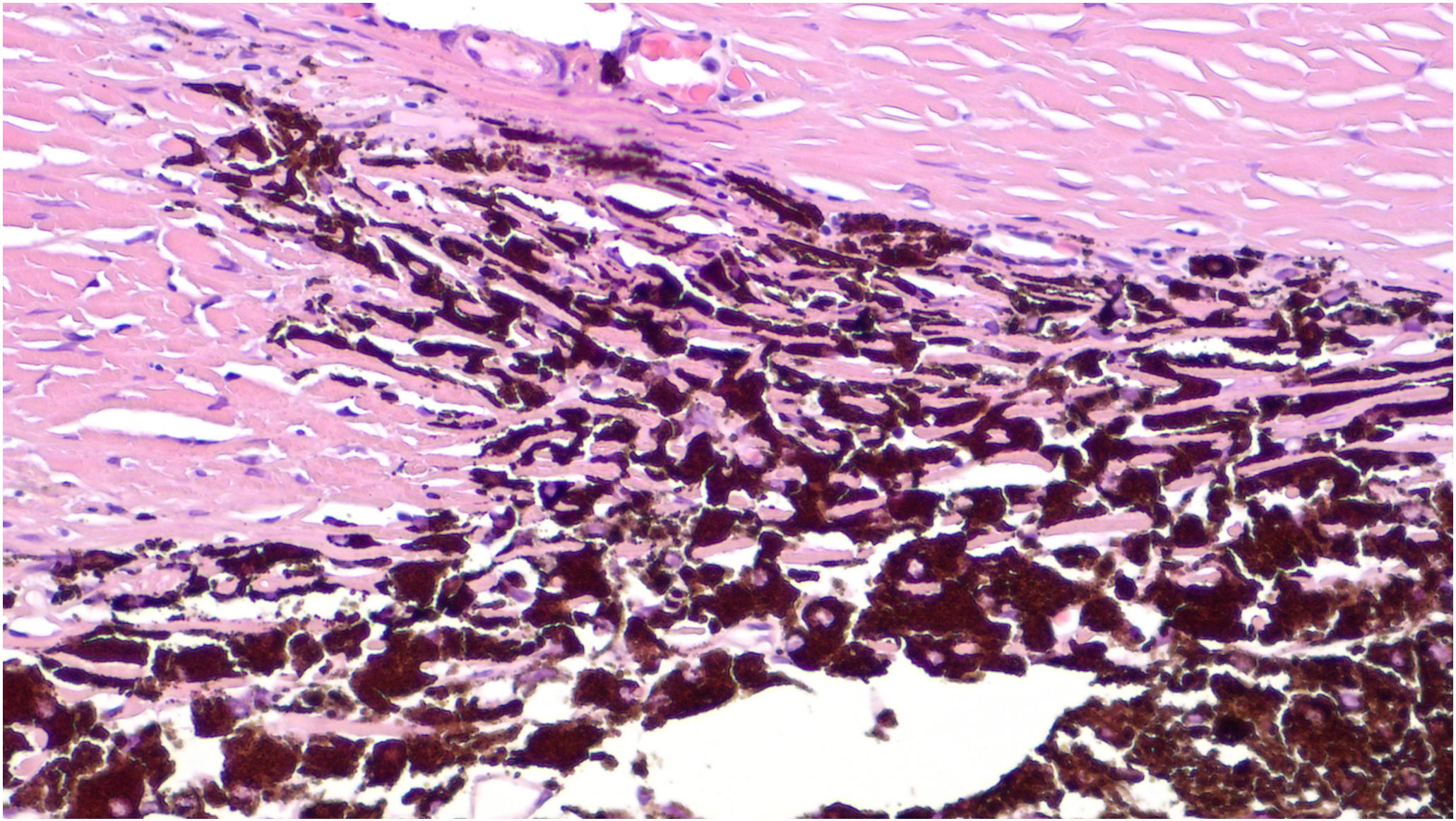


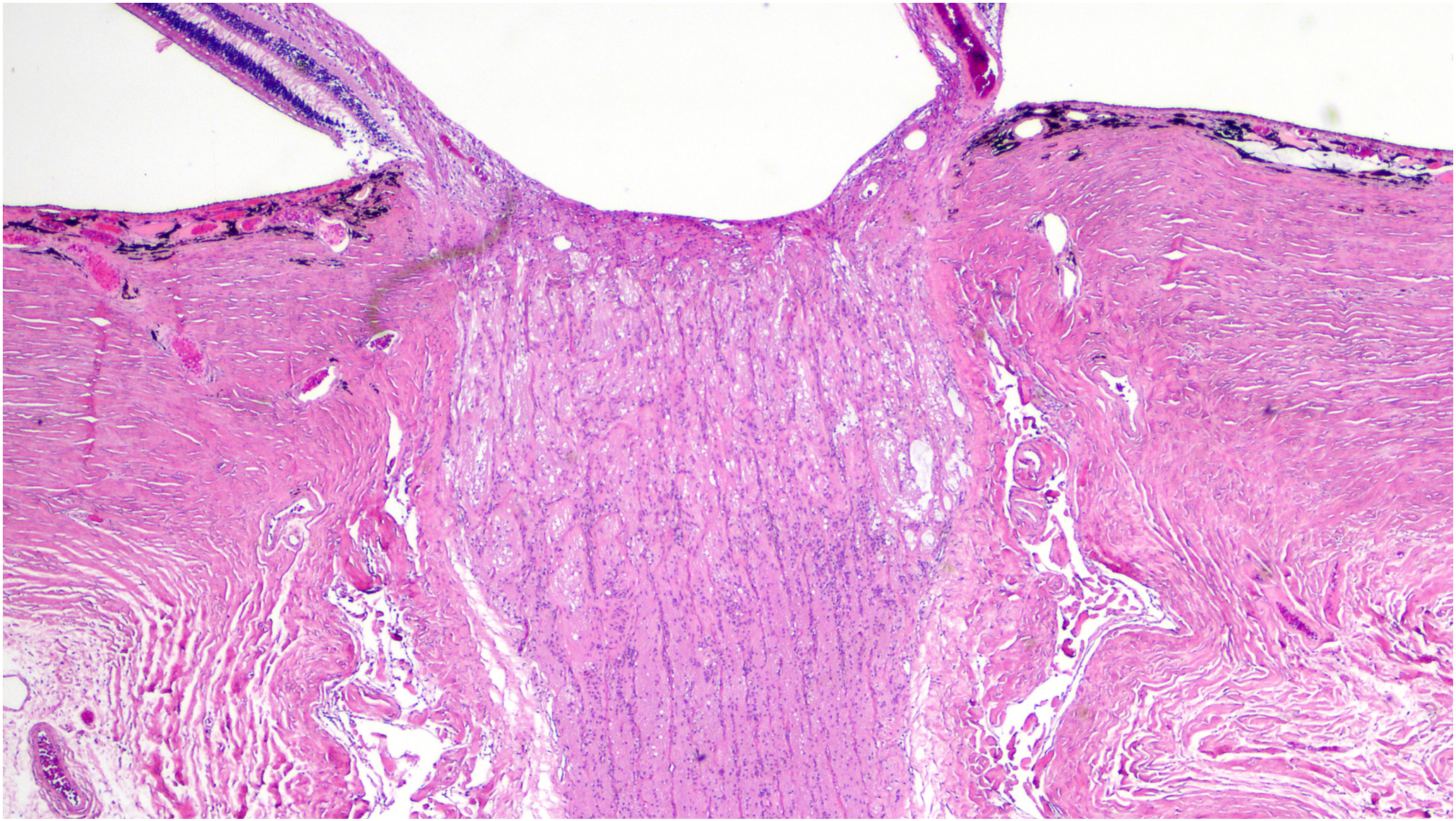


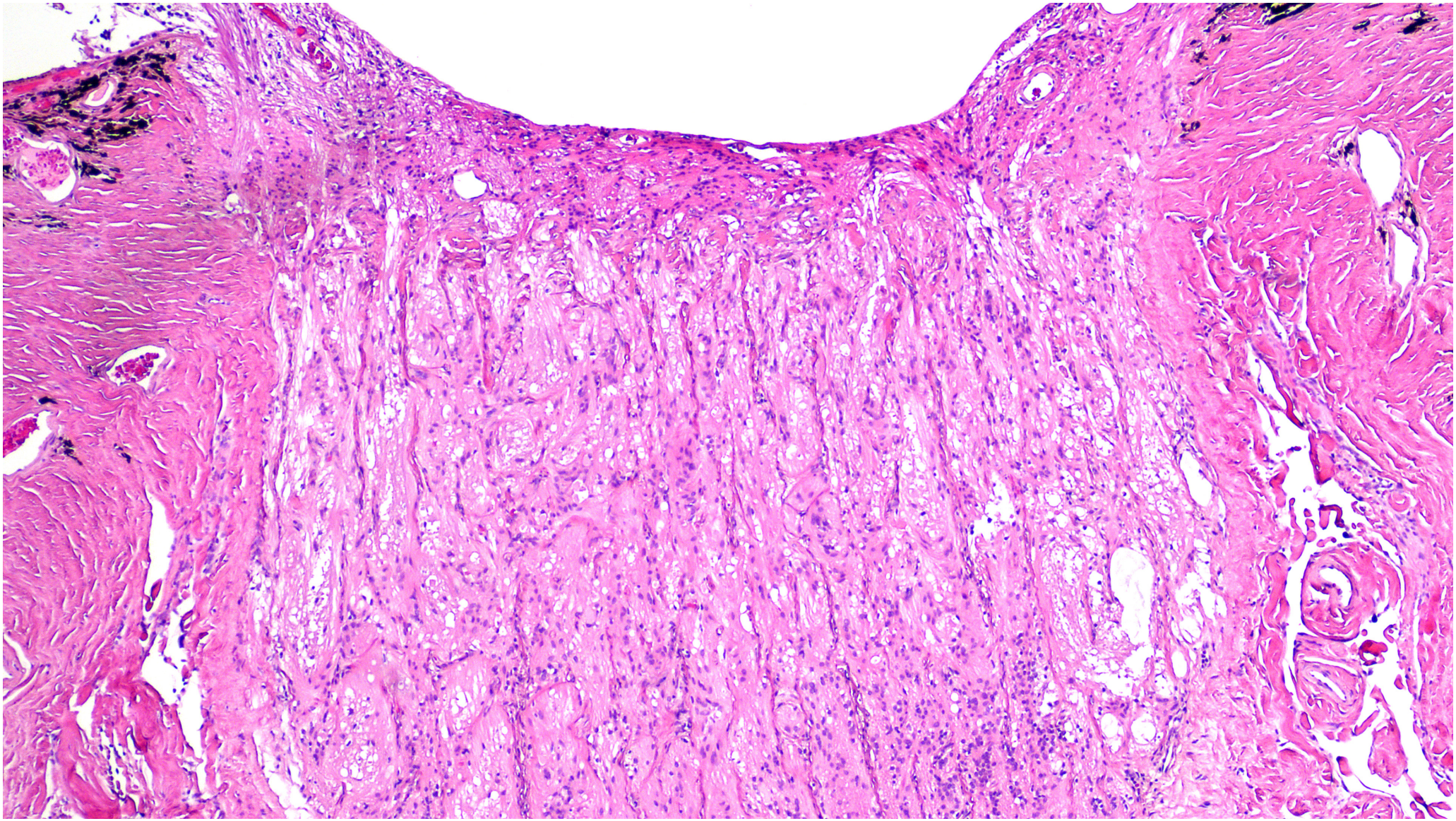


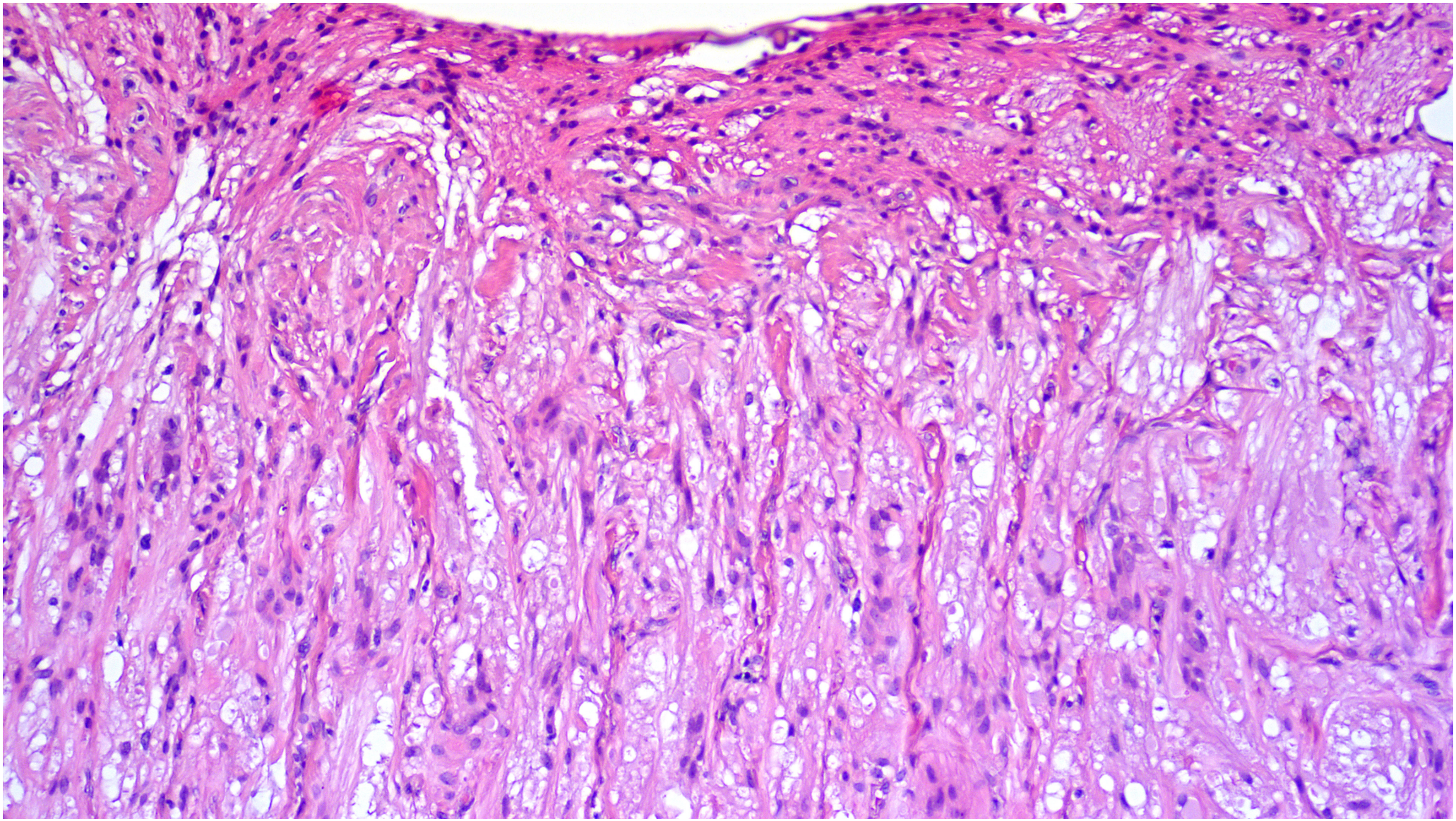












1965

"I have used the name "melanocytoma" to reflect the peculiar cytologic feature of this tumor— its composition of cells that look so much like those that thicken the uvea diffusely in congenital melanosis oculi… I believe that "melanocytoma" is an appropriate designation for a tumorlike accumulation of rather normal-appearing uveal melanocytes…"

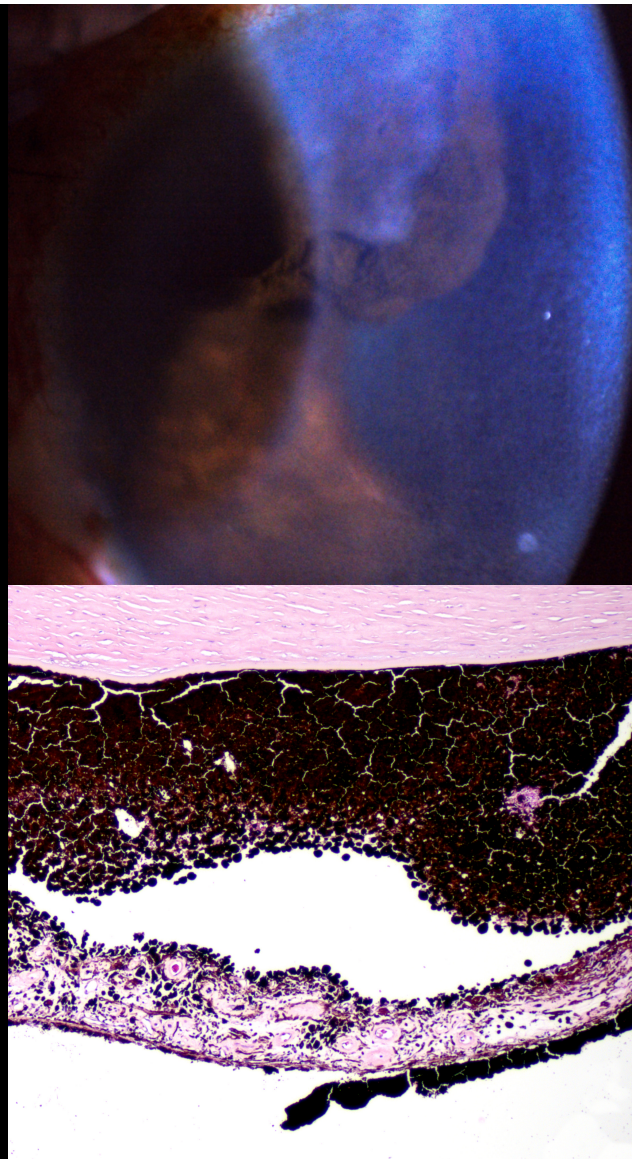
Zimmerman LE. Melanocytes, melanocytic nevi and melanocytomas. The Jonas Friedenwald Memorial Lecture. Invest Ophthalmol 1965;4:11–41.

*AFIP Acc. 618807.** In September, 1953, a 60-year-old white man sought examination for a change in his spectacles. He had no other complaints, and his visual acuity was 20/20, O.U. (corrected). On examination a densely pigmented tumor was observed on the left iris inferonasally. The tumor was markedly elevated and lay almost in contact with the cornea, completely blocking the chamber angle from 7 to 9 o'clock. The pupil dilated poorly in the sector containing the tumor, and a sector opacity was present in the lens corresponding to the position of the mass. The intraocular pressures were recorded as 24, O.D., and 28, O.S. Malignant melanoma was suspected, a consultant agreed, and the eye was enucleated on Oct. 29, 1953.

Examination of the enucleated eye confirmed the presence of a heavily pigmented tumor protruding from the iris in the lower nasal sector. In addition, the tumor was found to involve the pars plicata of the ciliary body, where a nodule measuring 2 by 3 by 3 mm. presented in the posterior chamber. On microscopic examination the tumor was found to be so heavily pigmented that bleached sections were necessary to make out cytologic detail (Fig. 39). I prepared the microscopic description of this tumor in 1954, before I was familiar with the melanocytomas, and I would like to quote verbatim from my report: "With bleaching, the tumor cells are seen to have a rather uniform appearance. They are

Iris Melanocytoma

- Rare variant of iris nevus; presents as a darkly-pigmented iris nodule with mossy, irregular surface
- Generally located on or adjacent to the optic disc but can rarely arise from the iris, choroid, ciliary body, or conjunctiva
- Natural course is variable
  - Undergoes (infrequently) spontaneous necrosis (possibly related to related to lack of circulation in a highly metabolic tumor)
  - Transforms to iris melanoma in rare cases



Iris Melanocytoma

- 1400 patients seen on the Oncology Service with iris nevus; 47 of these patients (3%) had iris melanocytoma
  - Color: darkly pigmented (94%)
  - Configuration: nodular (85%), diffuse (15%)
  - Involved quadrant: inferior (45%), lateral (38%), nasal (9%), superior (9%)

Iris Melanocytoma: Clinical Features and Natural Course in 47 Cases

HAKAN DEMIRCI, MD, ARMAN MASHAYEKHI, MD, CAROL L. SHIELDS, MD, RALPH C. EAGLE, JR., MD, AND JERRY A. SHIELDS, MD

• PURPOSE: To describe the clinical features, natural course, management and histopathologic features of iris melanocytoma.
• DESIGN: Single-center retrospective case series.
• METHODS: PATIENT POPULATION: Forty-seven consecutive patients (47 eyes) with iris melanocytoma. INTERVENTION PROCEDURE: Data regarding patient and tumor features were analyzed for their impact on the main outcome measures using univariate and multivariate regression models. Kaplan-Meier estimates were used to analyze the main outcomes as a function of time. MAIN OUTCOME MEASURES: Increased intraocular pressure (IOP), tumor seeding, and tumor growth.
• RESULTS: Associated findings at initial presentation included iris stromal seeds in 20 patients (43%), and anterior chamber angle seeds in 12 (26%). Intrinsic vascularization and sector cataract were not seen in any eyes. The management at presentation included observation in 39 patients (83%), tumor removal by sector iridectomy/iridocyclectomy in 7 (15%), and enucleation for blind painful eye with secondary increased IOP in 1 (2%). The diagnosis was confirmed by histopathologic examination in 11 patients (23%). The mean follow-up was 58 months. Using Kaplan-Meier estimates, clinical evidence of growth was observed in 23% at 5 years, 48% at 10 years, and 74% at 15 years. New tumor seeds developed in 34% at 5 years, 63% at 10 years, and 75% at 15 years. Increased IOP was observed in 11% at 5 years, 11% at 10 years, and 55% at 15 years.
• CONCLUSIONS: Iris melanocytoma represented only 3% of all iris nevi. Related iris stromal and anterior chamber angle seeds were common, and secondary glaucoma occurred in 11% at 5 years. Growth was observed in 23% at 5 years but no malignant transformation was found. (Am J Ophthalmol 2005;139:468–475. © 2005 by Elsevier Inc. All rights reserved.)

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From the Oncology Service (H.D., C.L.S., A.M., J.A.S.) and Department of Pathology (R.C.E., Jr.), Wills Eye Hospital, Thomas Jefferson University, Philadelphia, Pennsylvania.
Presented in part at the annual meeting of The Association for Research in Vision and Ophthalmology, Ft. Lauderdale, Florida, May 2003 (H.D.) and at the annual meeting of American Academy of Ophthalmology, New Orleans, Louisiana, October, 2004 (H.D.).
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MELANOCYTOMA IS A VARIANT OF MELANOCYTIC nevus with distinctive clinical and pathologic features.[1] Clinically, it appears as a deeply pigmented tumor that is usually located on or adjacent to the optic disk.[2,3] In rare instances, it can arise in iris, ciliary body, choroid, or conjunctiva.[4] In 1965, Zimmerman suggested the term "melanocytoma" to describe this tumor and reported two patients with iris melanocytoma, a 60-year-old white man with a deeply pigmented iris mass and a 34-year-old white man with recurrent iritis and pigmented iris mass. Iris melanoma was suspected in both cases, but both lesions proved to be iris melanocytoma after enucleation.[1] Since then, a few other reports of iris melanocytoma have been published.[5–14]

The natural course of iris melanocytoma is variable. Iris melanocytoma can undergo spontaneous necrosis with resultant pigment dispersion, causing secondary glaucoma and heterochromia.[5,6,8–13] In some cases, iris or ciliary body melanocytoma can show progressive growth or even extrascleral involvement making clinical differentiation from malignant melanoma difficult.[7,15,16] In rare cases, it can transform into iris melanoma.[8,14] Failure to recognize iris melanocytoma could lead to misdirected therapy, including enucleation. To further expand our understanding of this unique tumor, we report our experience with 47 patients with iris melanocytoma and describe its clinical and histopathologic features, management, and natural course.

PATIENTS AND METHODS

THE MEDICAL RECORDS OF ALL PATIENTS DIAGNOSED WITH iris nevus at the Oncology Service at Wills Eye Hospital between January 1974 and February 2003 were reviewed

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Iris Melanocytoma

- Presenting features: dark spot on the iris (15%), blurred vision (11%), and conjunctival hyperemia (4%); 33 patients (70%) were asymptomatic
  - Common (>10%) features included: heterochromia, sentinel vessels, iris stromal seeds and satellite tumors
  - Less common (<10%) features included: ectropion irides, AC inflammation, and pigmented keratic precipitates

TABLE 2. Related Anterior Segment Findings at Presentation of 47 Patients with Iris Melanocytoma

Clinical finding	No of patients (%)
Heterochromia	6 (13%)
Sentinel vessel	7 (15%)
Ectropion irides	3 (6%)
Satellite tumors	12 (26%)
Number of satellite tumors	
Mean	4
Median	3
Range	1–15
Iris stromal seeds	20 (43%)
Extent of iris stromal seeds (clock hours)	
Mean	5
Median	3
Range	1–12
AC angle seeds	12 (26%)
Extent of AC angle seeds (clock hours)	
Mean	8
Median	7
Range	3–12
AC inflammation	2 (4%)
Hyphema	0 (0%)
Pigmented keratic precipitate	1 (2%)
Intrinsic vascularization	0 (0%)
Sector cataract	0 (0%)

AC: Anterior chamber

- Managed typically with cautious observation (q6m); can use fine-needle aspiration biopsy or local resection using iridectomy, iridogoniectomy, or iridogoniocyclectomy to confirm diagnosis
- Enucleation is usually reserved for blind painful eyes

TABLE 5. Clinical Findings in 13 Well Documented Cases of Iris Melanocytoma in the Literature*

Author (Year)	Age/Race/Sex	Eye	Symptoms or signs	Color	Location	ACA seeds	Iris stromal seeds	Glaucoma	Growth	Association with melanoma	Management
Zimmerman (1965)	62 WM	OS	Asymptomatic	Black	Inferonasal	NA	NA	NA	NA	Absent	Enuc
Zimmerman (1965)	34 WM	OS	Recc. iritis	Black	Inferotemp	Present	NA	Absent	Absent	Absent	Enuc
Thomas (1969)	20 AAF	OS	Spot on iris, pain, redness	Black	Superior	Present	NA	Absent	Absent	Present	Enuc
Shields (1977)	23 WM	OS	Spot on iris	Black	Inferior	Present	Present	Present	Absent	Absent	Sector iridectomy
Nakazawa (1984)	46 AF	OD	Discomfort	Black	Diffuse	Present	Present	Present	Absent	Absent	Trabeculectomy and Inc Bx
Cialdini (1989)	34 WF	OS	Spot on iris, pain, redness	Black	Temporal	Present	Absent	Present	Present	Present	Sector iridectomy
Fountain (1992)	18 AAF	OS	Asymptomatic	Black	Inferior	Present	NA	Present	Absent	Absent	Sector iridectomy
Teichmann (1995)	28 ArF	OS	Spot on iris, pain, redness	Brown	Nasal	Present	Absent	Present	Absent	Absent	Sector iridectomy
Green (1996)	6 F	OS	Spot on iris	Black	Superior	Present	NA	Present	Absent	Absent	Enuc
Fineman (1998)	61 WM	OS	Spot on iris	Black	Inferonasal	Present	Present	Present	Absent	Absent	Enuc
Fineman (1998)	48 WF	OD	Spot on iris	Brown	Inferior	Present	Present	Present	Present	Absent	Sector iridectomy
Kiratli (2001)	27 WF	OS	Spot on iris	Black	Inferior	Present	Absent	Present	Absent	Absent	Sector iridectomy
Shields (2002)	9 WM	OS	Spot on iris	Brown	Inferotemp	Present	Absent	Absent	Present	Absent	Sector iridectomy

ACA = Anterior chamber angle; W = White, AA = African-American; A = Asian; Ar = Arabic; M = Male; F = Female; OD = Right eye; OS = Left eye; Enuc = Enucleation; Recc = Recurrent; Inferotemp = Inferotemporal; Inc Bx = Incisional biopsy

*. Well-documented cases published in English literature.

Elevated Intraocular Pressure

- Can undergo spontaneous necrosis resulting in iris stromal and angle seeding, dispersion of melanophages, and secondary elevated IOP
  - Pigmented KPs were a risk factor for development of AC and/or iris seeds
- Of note, the association of glaucoma and acquired hyperchromic iris heterochromia from a pigmented iris mass usually suggests a high-grade malignant melanoma; complicates diagnostic certainty

TABLE 4. Univariate and Multivariate Analysis of Clinical Factors Correlated with Increased IOP, Development of New Iris Stromal and Anterior Chamber Angle Seeds and Growth in 47 Patients with Iris Melanocytoma*

Factor	P	Relative Risk (95% Confidence Interval)
Increased IOP		
Univariate analysis		
Race (Non-Caucasian** vs Caucasian)	0.008	27.5 (2.3–323.4)
Pigmented keratic precipitates (Present** vs Absent)	0.04	11.5 (1.1–129.1)
Anterior chamber inflammation (Present** vs Absent)	0.04	18.4 (1.1–295.4)
New iris stromal and ACA seeds		
Univariate analysis		
Race (Non-Caucasian** vs Caucasian)	0.04	18.5 (1.2–295.7)
Pigment keratic precipitates (Present** vs Absent)	0.04	18.5 (1.2–295.7)
Growth		
Univariate analysis		
Anterior chamber angle seeds (Present** vs Absent)	0.04	5.5 (1.1–26.3)

*Multivariate analysis was performed for each main outcome, but there were no factor found to be a significant predictor for any of the outcome measures.
**Reference variable

Melanocytomalytic Glaucoma

- Three patients who had an iris melanocytoma that underwent necrosis and caused secondary open-angle glaucoma from necrotic debris in the angle

	Patient 1	Patient 2	Patient 3
VA	CF	20/20	20/20
IOP	45 mmHg	27 mmHg	29 mmHg
Exam	darkly pigmented lesion in the inferonasal iris and the angle between 6 and 8 o'clock; hyperchromic heterochromia iridum	uniformly pigmented dark brown mass in the inferior iris between the 5 and 7 o'clock positions with a central crater suggestive of necrosis	pigmented tumor measuring about 3.0 mm in diameter and 1.0 mm high extended from the pupillary margin to the midportion of the iris inferonasally; two other pigmented lesions with angle pigmentation
Tx	enucleation	partial lamellar scleroiridogonio-cyclectomy	inferonasal sector iridectomy after which intraocular pressure normalized without the use of medication

Melanocytomalytic Glaucoma in Eyes with Necrotic Iris Melanocytoma

Mitchell S. Fineman, MD,[1] Ralph C. Eagle, Jr, MD,[2] Jerry A. Shields, MD,[1] Carol L. Shields, MD,[1] Patrick De Potter, MD[1]

Objective: Iris melanocytoma, although histologically benign, may undergo spontaneous necrosis with the resultant pigment dispersion causing secondary open-angle glaucoma. The authors describe the clinical findings in three patients with this syndrome and review the current literature.

Design: The study design was a small case series.

Participants: Three patients with secondary glaucoma caused by a necrotic iris melanocytoma participated.

Intervention: Clinical records and pathologic preparations were reviewed. The diagnosis of iris melanocytoma was confirmed by histopathologic examination in all three cases.

Results: All patients had intrinsically pigmented localized iris tumors and pigmentation of the angle. Hyperchromic heterochromia iridum was noticeable in all patients. In two instances, a central crater in the lesion signified tumor necrosis. Two patients were treated successfully by surgical resection of the iris tumor and the glaucoma resolved. One patient was treated with enucleation. In all patients, histopathologic analysis disclosed extensively necrotic tumor comprised of maximally pigmented nevus cells with bland nuclei and infiltration of the trabecular meshwork by melanophages.

Conclusion: Secondary glaucoma may result from obstruction of the trabecular meshwork with necrotic iris melanocytoma. Resection of the tumor may relieve the glaucoma. *Ophthalmology 1998;105:492–496*

Melanocytoma or magnocellular nevus is a relatively uncommon intraocular tumor that appears clinically as a deeply pigmented mass. Although melanocytoma classically is associated with the optic disc, extrapapillary locations throughout the uveal tract, particularly the iris, can be found.[1] Histopathologically, a melanocytoma is composed of large, uniform, round-to-polygonal cells that contain small, bland nuclei and copious amounts of intensely pigmented cytoplasm.[2] Although histologically benign, this tumor may spontaneously undergo necrosis with the resultant pigment dispersion causing secondary glaucoma and heterochromia.[3] However, the association of glaucoma and acquired hyperchromic iris heterochromia from a pigmented iris mass usually suggests a high-grade malignant melanoma.[4] Indeed, in a retrospective review, 7 of 102 eyes (7%) with iris melanoma had secondary intraocular pressure elevation.[5] Therefore, the clinical differentiation of secondary glaucoma caused by a necrotic melanocytoma versus that caused by an aggressive malignant melanoma may have significant prognostic and therapeutic implications. We describe three patients who had an iris melanocytoma that underwent necrosis and caused secondary open-angle glaucoma from necrotic debris in the anterior chamber angle. A review of the current literature is provided.

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Presented in part at the Combined Meeting of the Verhoeff and the European Ophthalmic Pathology Societies, Nürnberg, Germany, May 8–12, 1991.

Supported by the Eye Tumor Research Foundation, Philadelphia, Pennsylvania.

Reprint requests to Jerry A. Shields, MD, Ocular Oncology Service, Wills Eye Hospital, 900 Walnut Street, Philadelphia, PA 19107.

Case 1

A 61-year-old white man was referred to the Oncology Service, Wills Eye Hospital, after a pigmented iris mass was noted during a routine eye examination. The visual acuity was 6/5 in the right eye and finger counting in the left eye. A left relative afferent pupillary defect was present. Applanation tonometry showed 14 mmHg in the right eye and 45 mmHg in the left eye. There was no evidence of congenital ocular or oculodermal melanocytosis. A darkly pigmented lesion was noted in the inferonasal iris and the angle of the left eye between the 6:30- and 8-o'clock positions. Diffuse shedding of pigment onto the anterior iridic surface had produced a noticeable hyperchromic heterochromia iridum. Gonioscopy disclosed diffuse circumferential pigmentation of the trabecular meshwork and the zonule, and a prominent central crater in the tumor (Fig 1A). With the exception of an inferonasal sector cataract, the media were clear. There was advanced glaucomatous excavation of the left optic nerve (cup-to-disc ratio of 0.9). The remaining results of the fundus examination were normal. The right eye was normal with a healthy, uncupped optic nerve.

Based on these findings, the patient was thought to have an iridociliary malignant melanoma of the left eye that had infiltrated the trabecular meshwork, causing secondary glaucoma.

Melanocytomalytic Glaucoma

- Necrosis of the tumor is typically observed both clinically or histopathologically in association with melanocytomalytic glaucoma
  - Increased frequency of secondary glaucoma seen with necrotic **iris** melanocytoma simply is a function of the proximity to the trabecular meshwork and aqueous outflow pathway
- Treatments vary but must be preceded by diagnostic confirmation
  - Changes induced by the melanocytoma on the trabecular meshwork may be reversible after removal of the tumor

Melanocytomalytic Glaucoma in Eyes with Necrotic Iris Melanocytoma

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Summary

- 52 y.o. male presenting to the Wills Emergency Room with "severe, 10/10 left eye pain" for two months; family history of choroidal melanoma s/p enucleation
- Follow-up exam OS with VA CF @ 2ft & IOP 35; 20x20x3mm SN mass with ?EOE & diffuse seeding
  - Patient underwent enucleation due to concern for melanoma
  - Pathologic specimen consistent with iris melanocytoma
- Iris melanocytoma is a rare variant of iris nevus; presents as a darkly-pigmented iris nodule with mossy, irregular surface
  - Can undergo spontaneous necrosis resulting in iris stromal and angle seeding, dispersion of melanophages, and secondary elevated IOP
  - Enucleation is usually reserved for blind painful eyes

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A Big Thank You

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Wills Ocular Oncology Service

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