Chiefs' Rounds

WillsEye Hospital

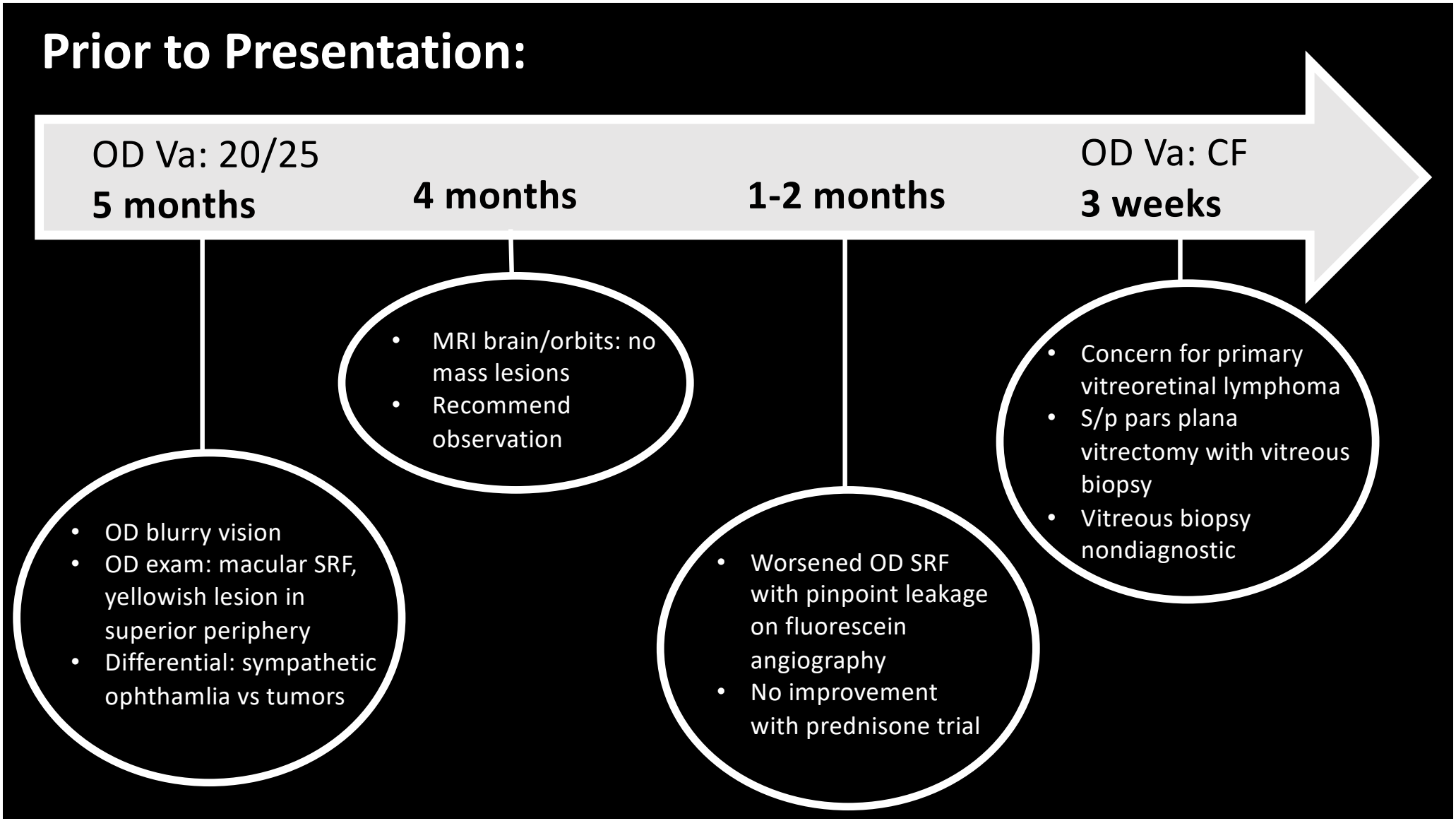
May 7th, 2021

Lucy Cobbs MD, PGY-2

- I have no financial disclosures to report.

Chief Complaint:

55-year-old female referred for second opinion on vision loss in her right eye x 5 months.


Prior to Presentation:
OD Va: 20/25
5 months
4 months
1-2 months
OD Va: CF
3 weeks
• OD blurry vision
• OD exam: macular SRF, yellowish lesion in superior periphery
• Differential: sympathetic ophthamlia vs tumors
• MRI brain/orbits: no mass lesions
• Recommend observation
• Worsened OD SRF with pinpoint leakage on fluorescein angiography
• No improvement with prednisone trial
• Concern for primary vitreoretinal lymphoma
• S/p pars plana vitrectomy with vitreous biopsy
• Vitreous biopsy nondiagnostic

Past Ocular History

- OS traumatic choroidal rupture in 2009
- Glaucoma suspect

Ocular Medications

- None

Complex Past Medical History

1. Intestinal lymphangiectasia (Lymphocytic enterocolitis) →
   - Total parenteral nutrition (TPN)
   - Budesonide
   - Infliximab
   - Octreotide acetate
   - Furosemide
   - Potassium chloride
   - Multivitamins
   - Bactrim
2. Embolic cerebellar stroke →
   - Aspirin
3. Subclavian vein thrombosis →
   - Rivaroxaban
4. Endocarditis
5. Hypertension →
   - Carvedilol
6. Hyperlipidemia →
   - Atorvastatin

Past Surgical History

- Caesarian Section

Social History

- Born in China, immigrated in 1993
- Never smoker
- No EtOH use or IVDU

Review of Systems

+ Lower extremity swelling

Family History

- Diabetes (mother)
- Stroke (maternal grandfather)

Exam

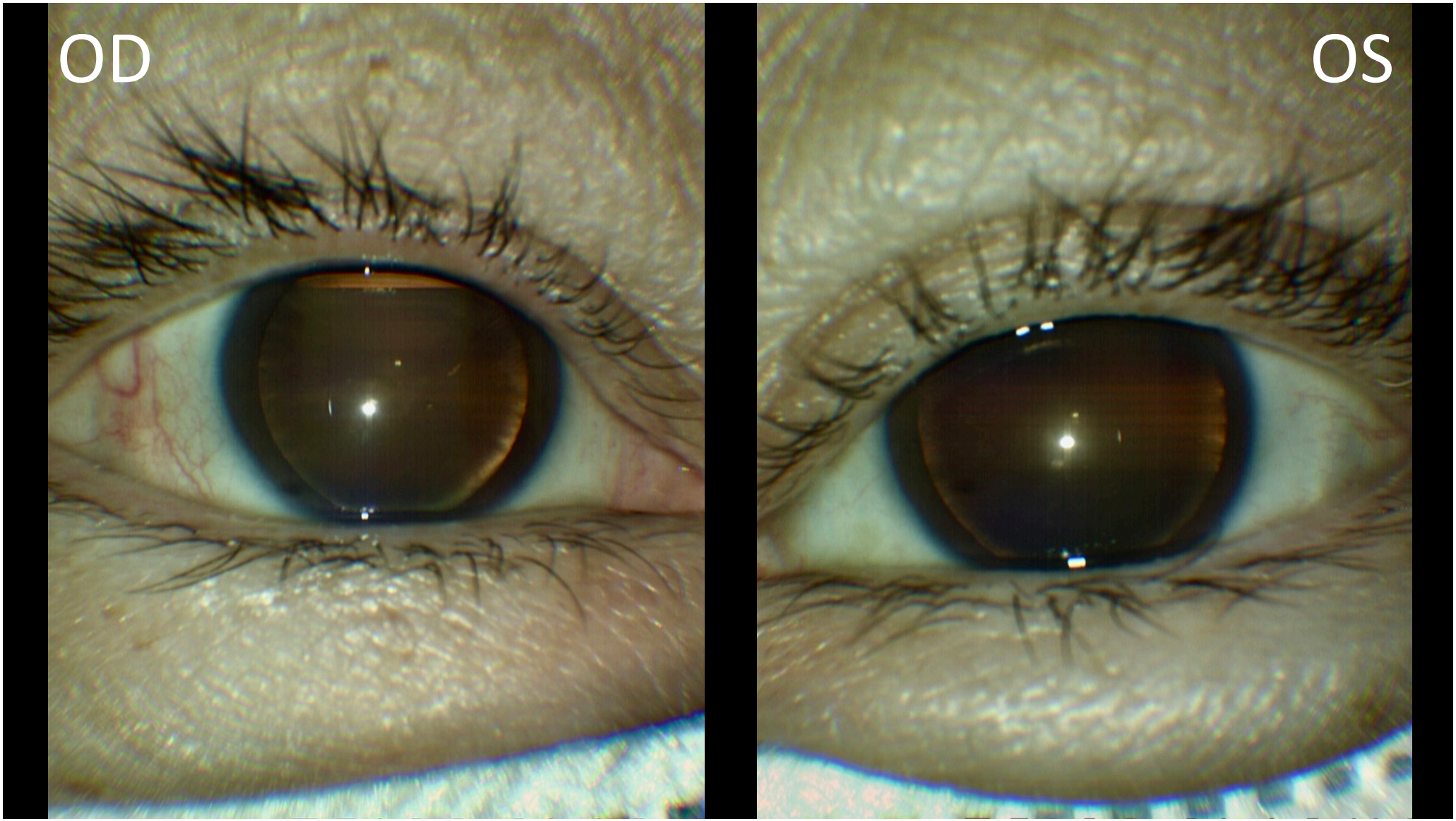
V_{sc} < CF @ 2ft niph
20/400 niph

P < Equal, round, reactive to light
No rAPD

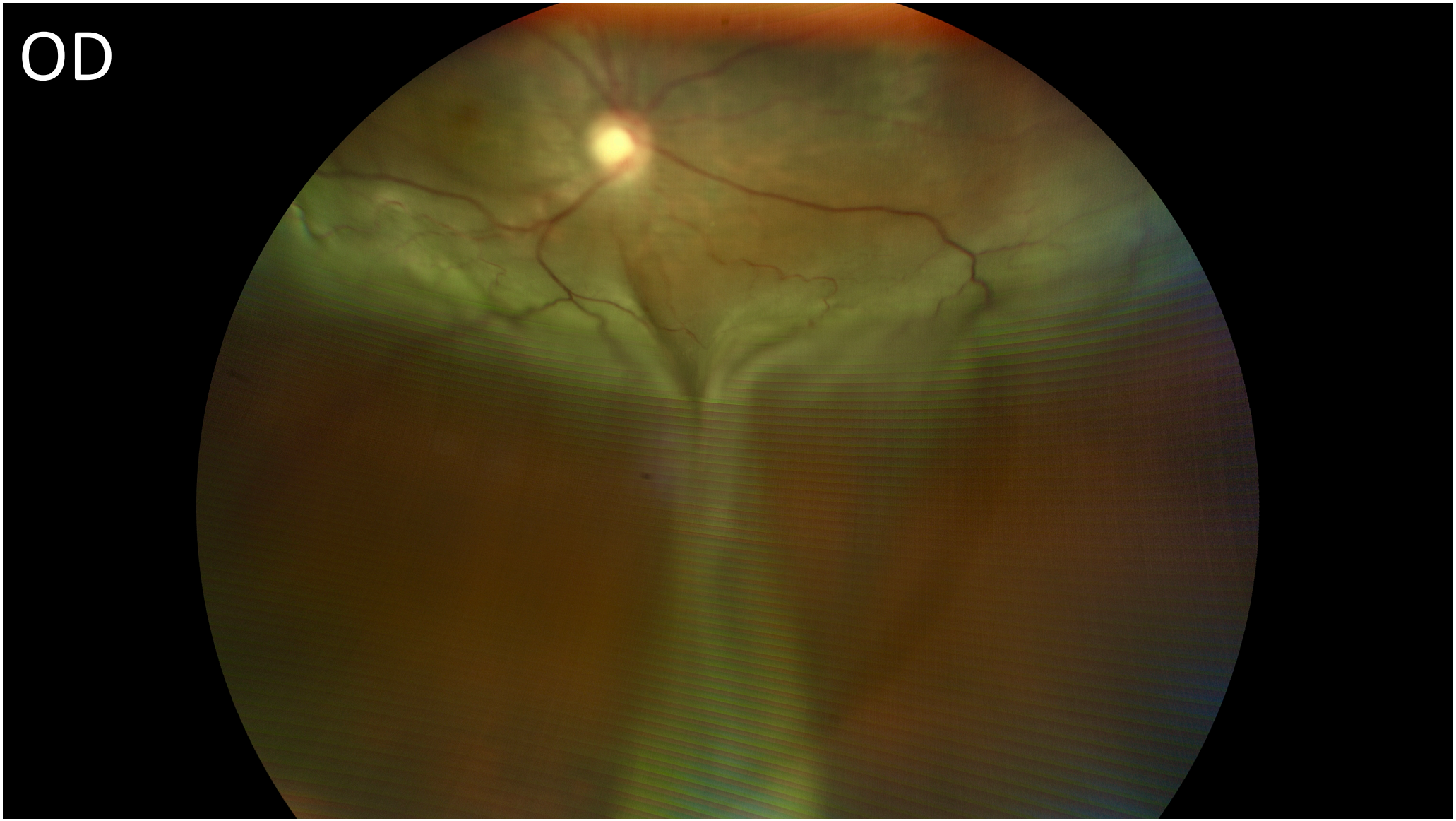
T_{App} < 10
10

CVF < Not performed
Not performed

EOM < Full
Full


OD
OS


OD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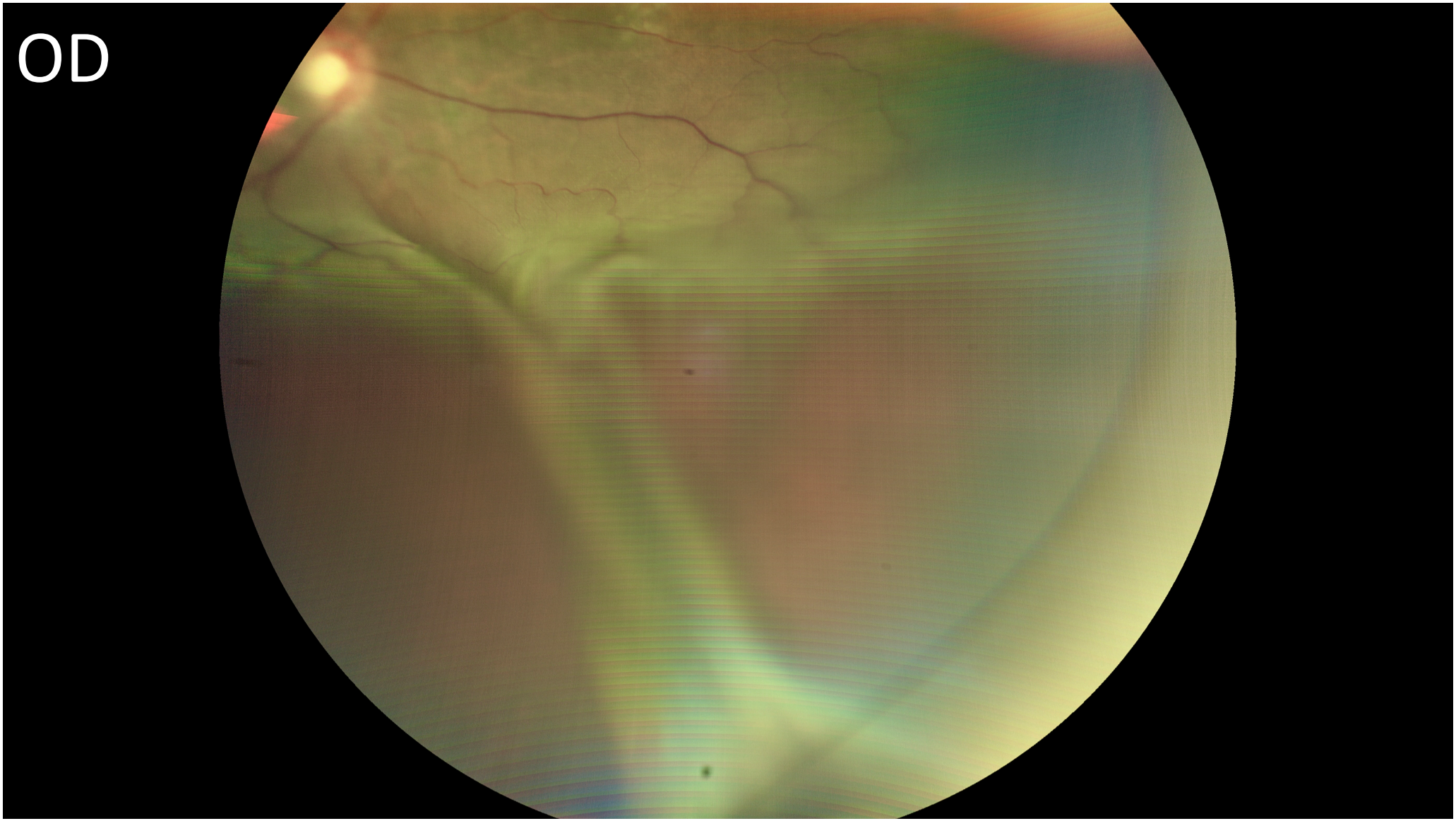
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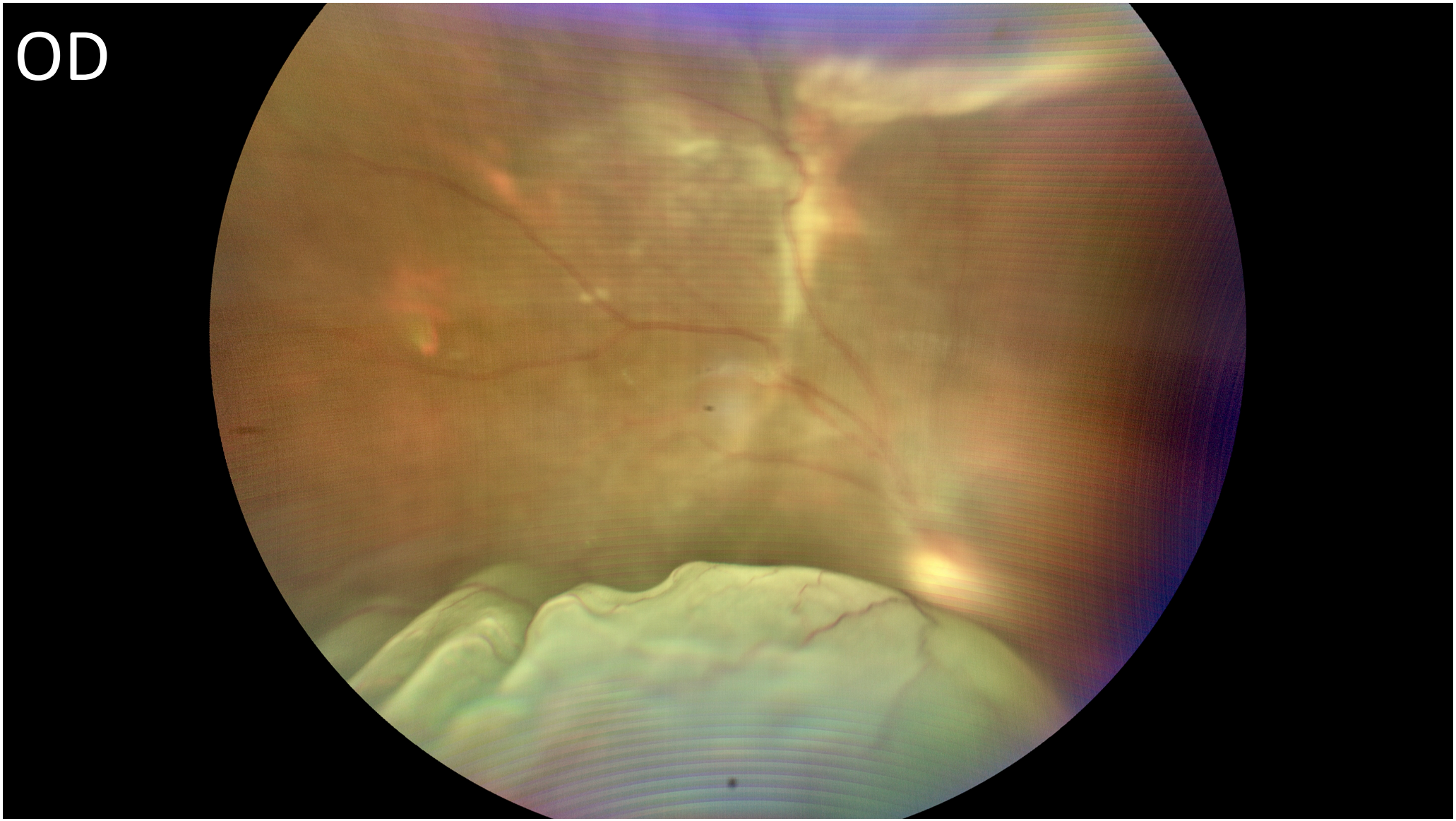
OD


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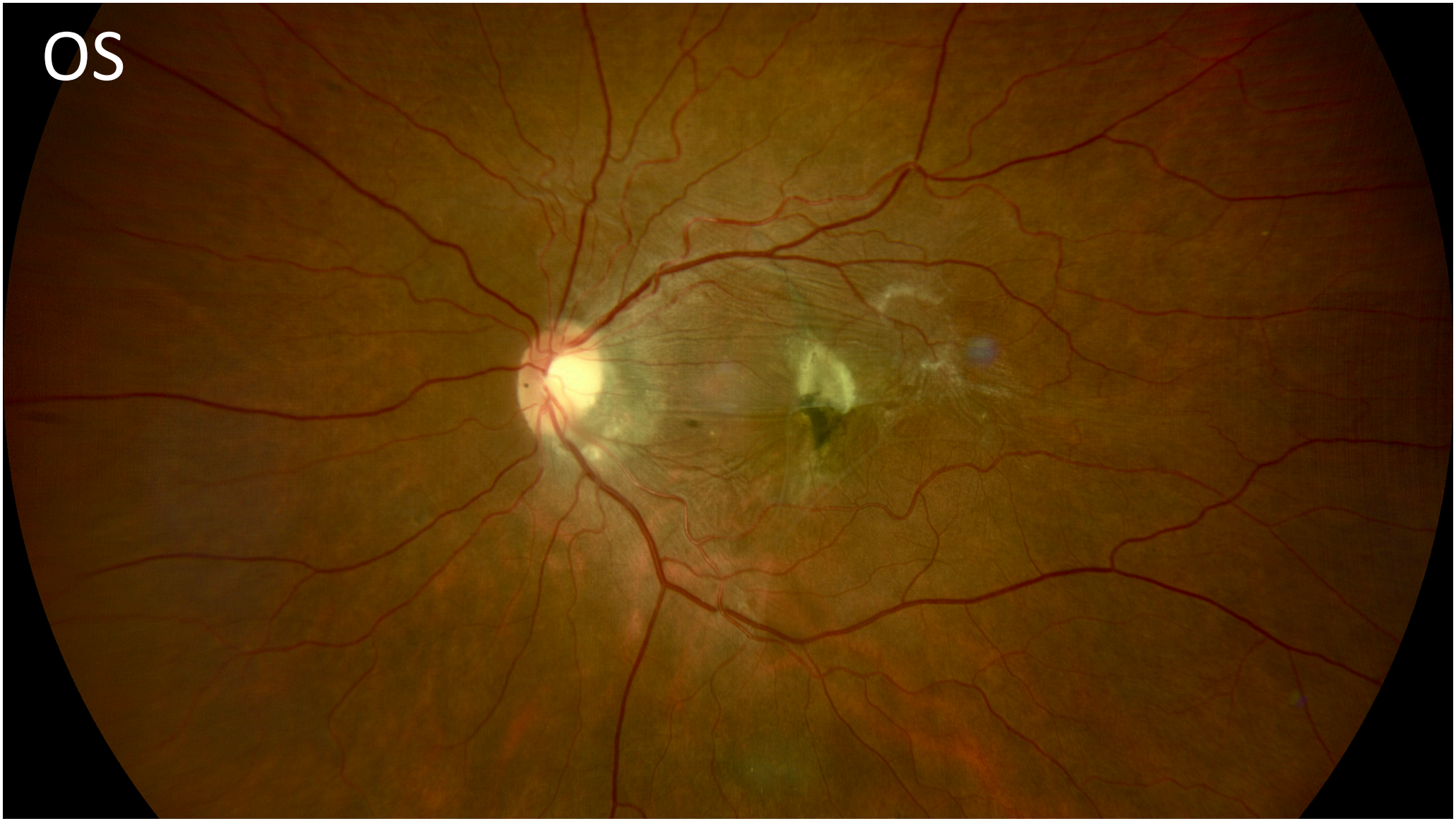

OS

Differential Diagnosis of Unilateral Serous Retinal Detachment with ?subretinal lesion

Infectious
Tuberculosis granuloma, syphilis, choroidal infiltration from systemic infection

Inflammatory
Sympathetic ophthalmia, Vogt-Koyanagi-Harada disease, posterior scleritis, **c**hronic inflammatory process

Neoplastic
Metastasis, choroidal melanoma, primary vitreoretinal lymphoma, choroidal hemangioma, multiple myeloma, retinal capillary hemangioblastoma

Vascular
Choroidal neovascularization, disseminated intravascular coagulopathy

Idiopathic uveal effusion syndrome

Central Serous Retinopathy

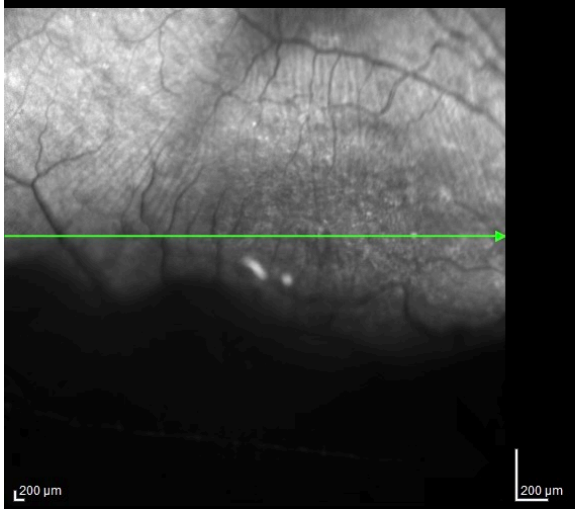
Malignant hypertension

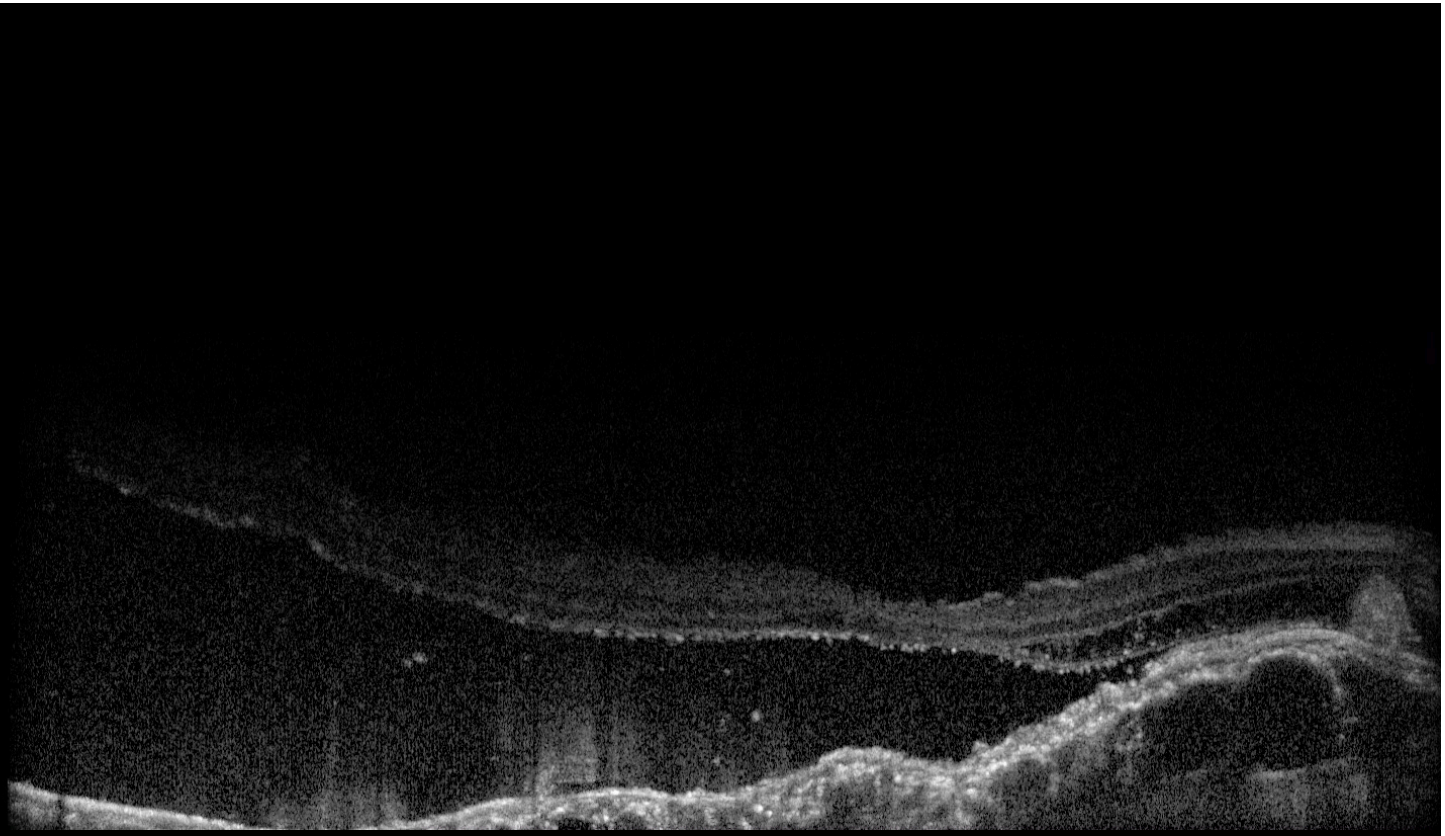
Next Steps?

→ Optical coherence tomography

→ B-scan ultrasonography

OD





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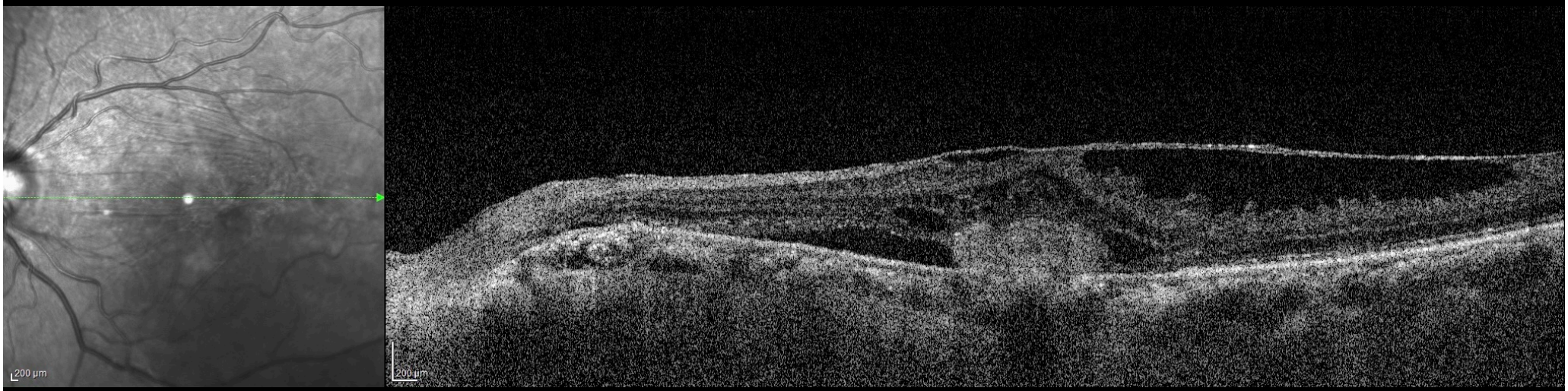

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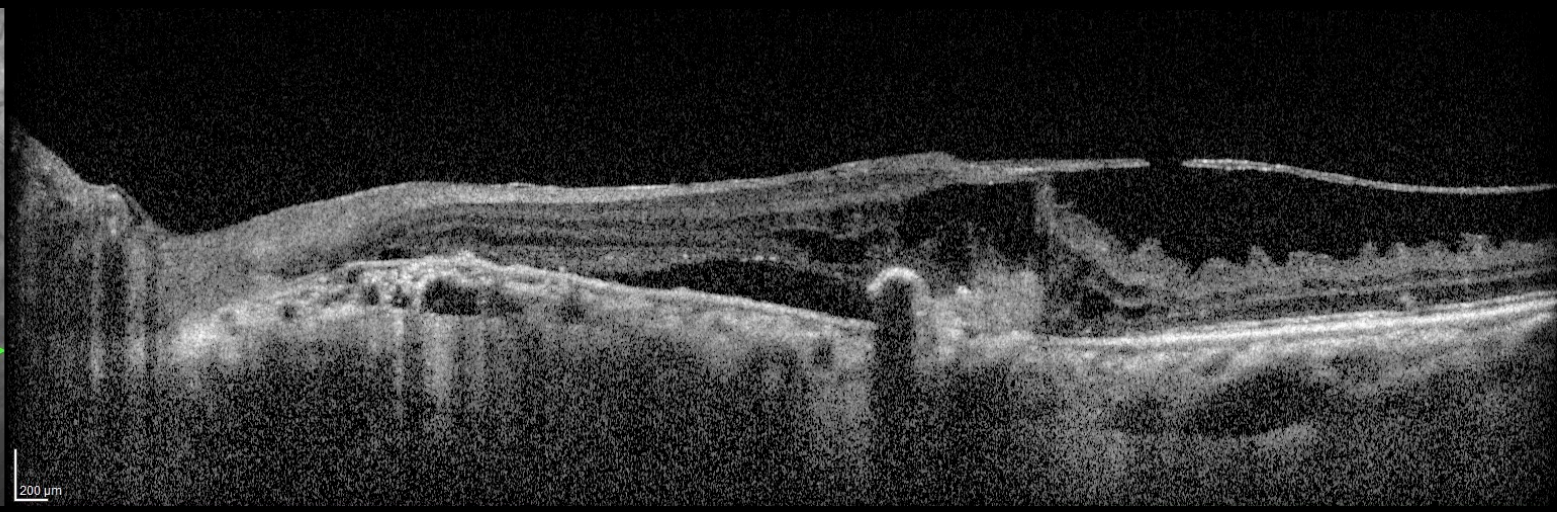

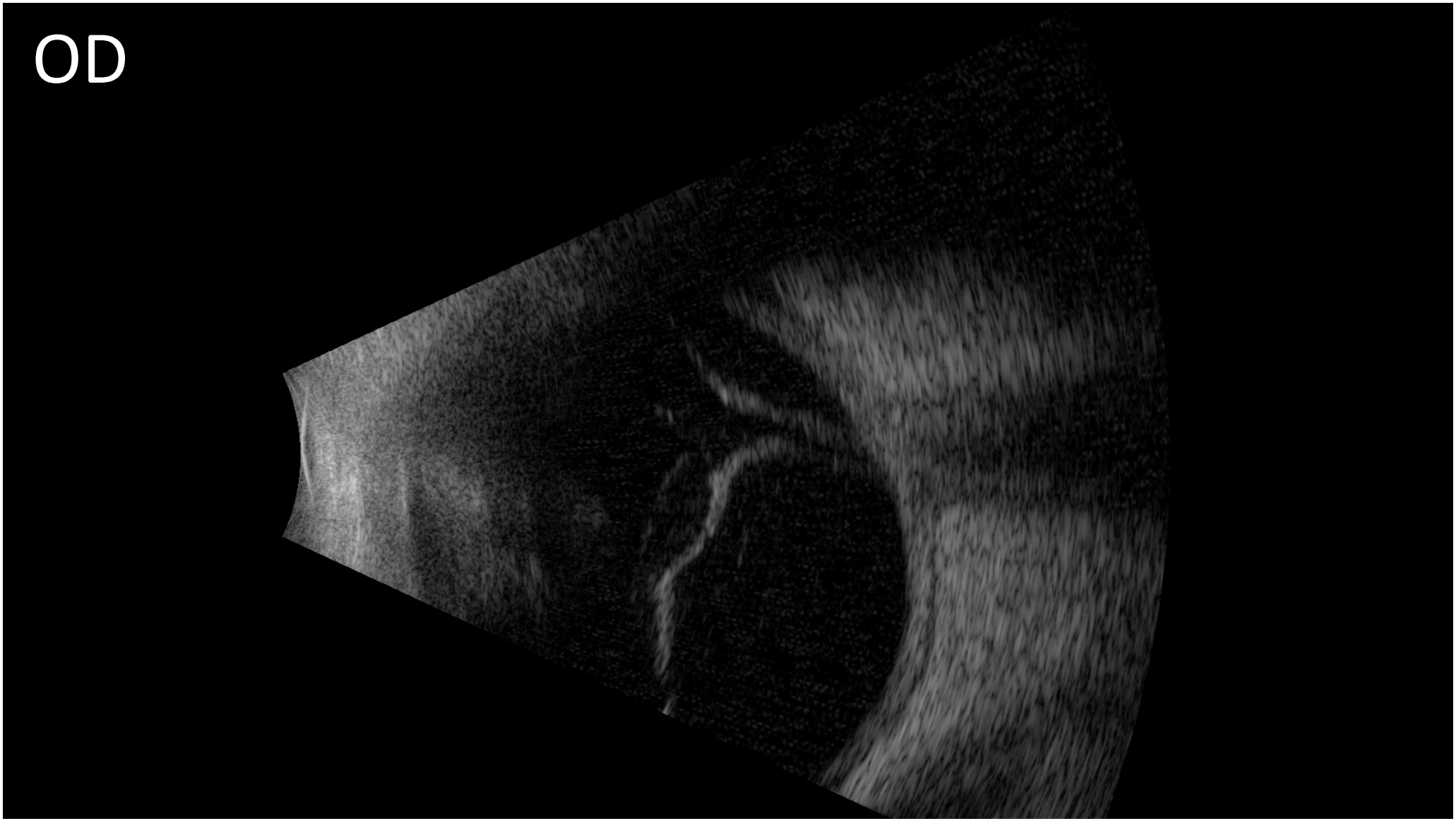
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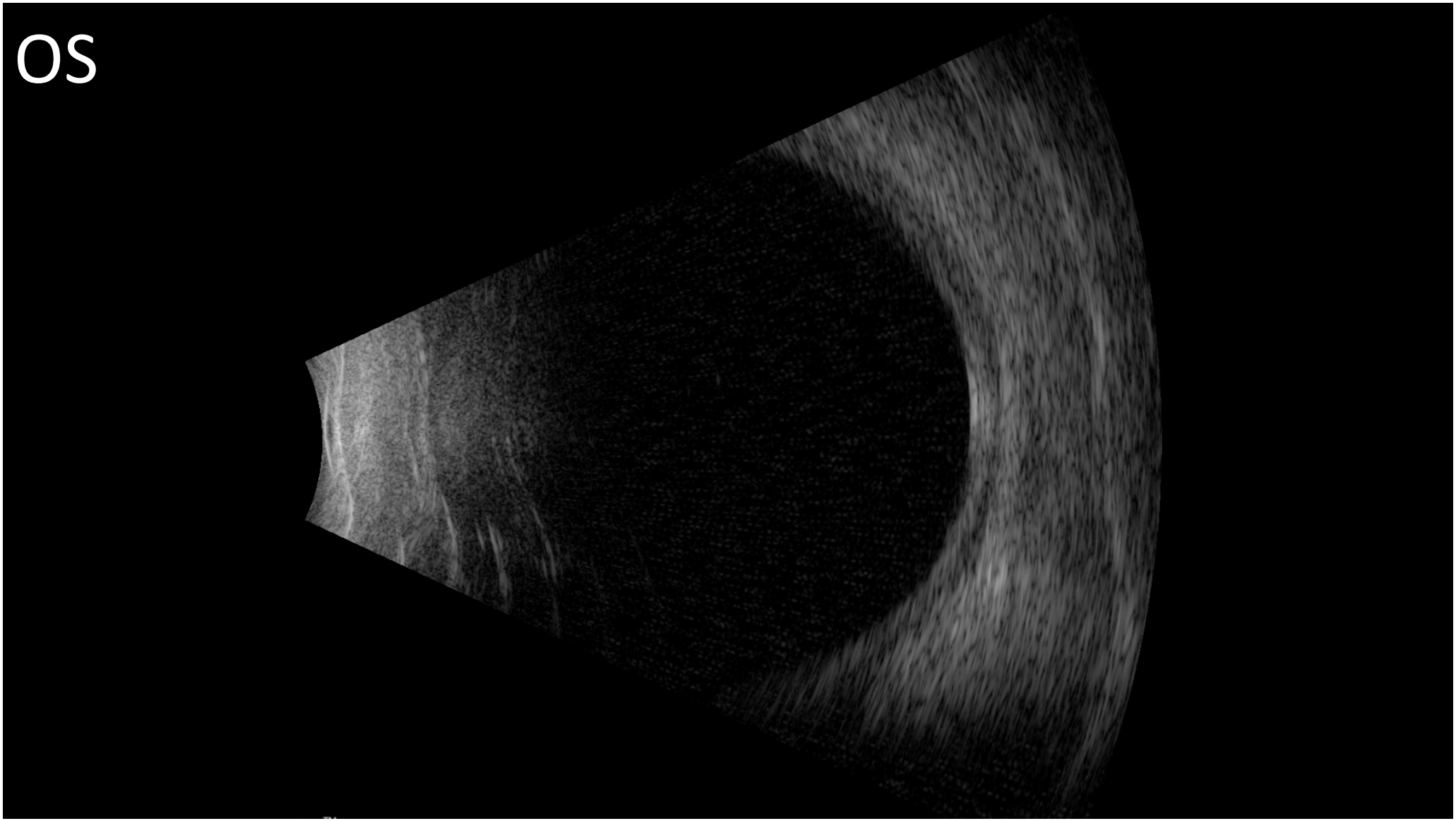
OS

Prior Workup

- Labwork: infectious / inflammatory tests nondiagnostic
- Vitreous biopsy: nondiagnostic
- MRI brain / orbit: no masses
- Fluorescein angiography

Prior Workup

POSITIVE:

- Anti-dsDNA antibodies
- ANA

NEGATIVE

- Syphilis, rheumatoid factor, ACE, C-ANCA, P-ANCA, hepatitis C virus, hepatitis B virus, T-spot, proteinase 3, MPO, CMV viral load, lupus anticoagulant, blood cultures (drawn on initial presentation at outside hospital)

VITREOUS BIOPSY

- Few cells, mostly mature T cells, absent B cells

MRI Brain and Orbit

1. **Moderate enlargement and edema of optic chiasm, prechiasmatic optic nerves extending to the optic tracts bilaterally with no associated enhancement.** Intraorbital and prechiasmatic optic nerves appear normal. Findings are nonspecific. Given lack of associated enhancement, inflammatory changes or neoplasm such as optic pathway glioma are less likely. No pituitary mass is seen.

2. No discrete enhancing mass is seen involving the choroids. **Slight edema without enhancement is seen in the intraorbital fat predominantly surrounding the bilateral sclera right slightly worse than left.**

3. No enhancing brain lesion to suggest brain metastasis. No acute intracranial abnormalities. Mild chronic small vessel ischemic changes and small old appearing lacunar infarcts of the cerebellar hemispheres

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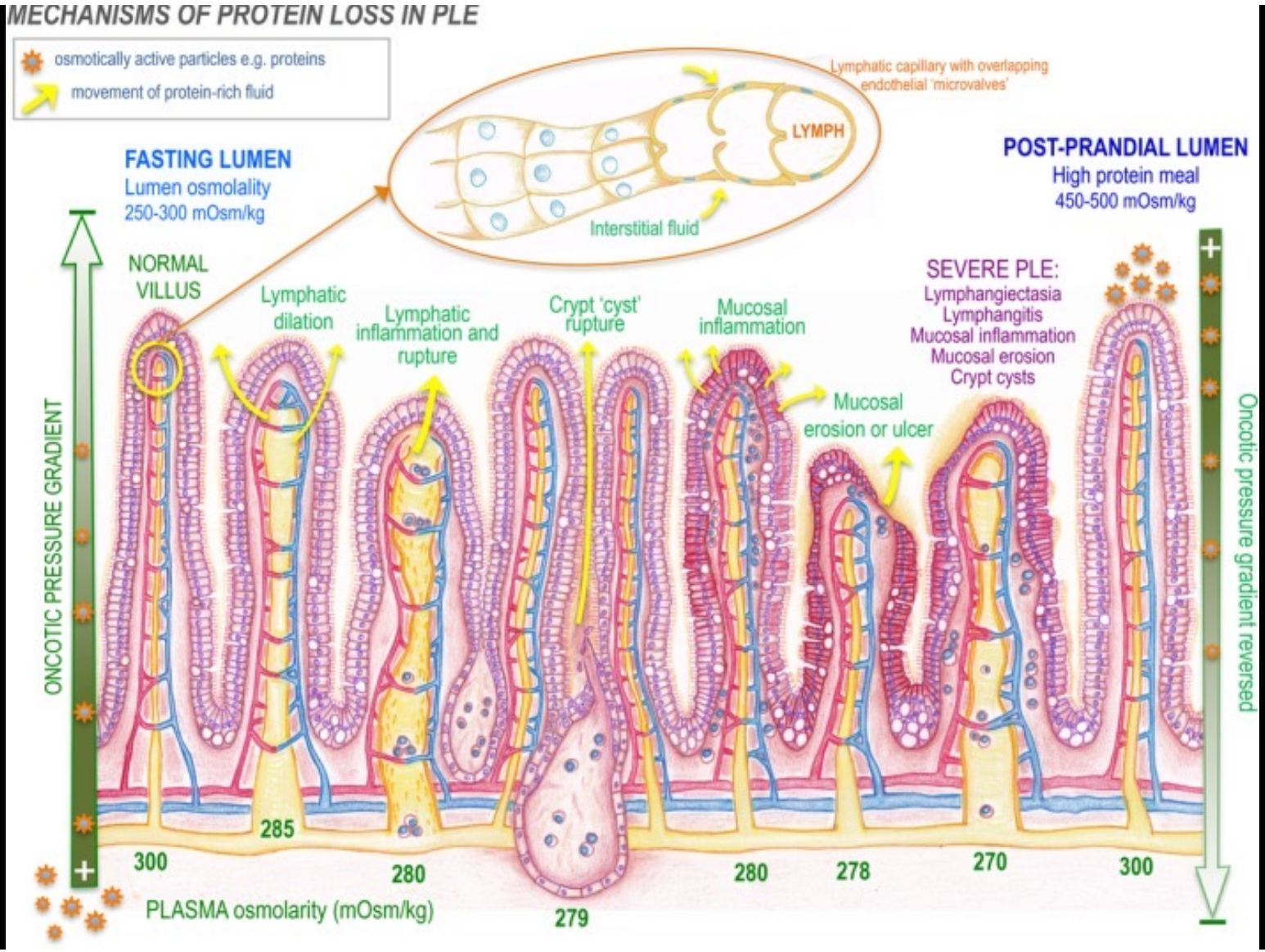
MECHANISMS OF PROTEIN LOSS IN PLE
osmotically active particles e.g. proteins
movement of protein-rich fluid
Lymphatic capillary with overlapping endothelial 'microvalves'
LYMPH
Interstitial fluid
FASTING LUMEN
Lumen osmolality
250-300 mOsm/kg
POST-PRANDIAL LUMEN
High protein meal
450-500 mOsm/kg
NORMAL VILLUS
Lymphatic dilation
Lymphatic inflammation and rupture
Crypt 'cyst' rupture
Mucosal inflammation
Mucosal erosion or ulcer
SEVERE PLE:
Lymphangiectasia
Lymphangitis
Mucosal inflammation
Mucosal erosion
Crypt cysts
ONCOTIC PRESSURE GRADIENT
Oncotic pressure gradient reversed
300
285
280
279
280
278
270
300
PLASMA osmolarity (mOsm/kg)

Protein-Losing Enteropathy: Case Illustrations and Clinical Review

2010; 105:43–49;
Sarah B. Umar, MD[1] and John K. DiBaise, MD, FACG[1]
The American Journal of GASTROENTEROLOGY

- Protein-losing enteropathy (PLE) is a rare condition characterized by a loss of serum protein into the gastrointestinal tract resulting in hypoproteinemia, which can be complicated by edema, ascites, pleural and pericardial effusions, and malnutrition

- Gastrointestinal protein loss in PLE has been reported to involve up to 60 % of the total albumin pool

July 2004 *Arch Ophthalmol.* 2004;122(7):1067-1070.

Bilateral Serous Retinal Detachment Due to Protein-Losing Enteropathy

Jayant Venkatramani, MBBS; Justin L. Gottlieb, MD; Thom S. Thomassen, MD; et al

- We describe an unusual patient with a corticosteroid-responsive PLE who developed bilateral serous retinal detachments (RDs) coincident with a flare of her enteropathy.
- With appropriate treatment of the enteropathy, there was resolution of the serous detachments.
- To the authors' knowledge, this is the first case reported of bilateral serous RD secondary to a PLE.

Effects of Subretinal and Systemic Osmolality on the Rate of Subretinal Fluid Resorption

Akira Negi and Michael F. Marmor

INVESTIGATIVE OPHTHALMOLOGY & VISUAL SCIENCE / May 1984

Oncotic pressure is important for maintaining an attached retina.

Next Steps

- Sub-tenon triamcinolone acetonide
- Discuss systemic treatments for protein losing enteropathy with gastroenterologists
- Recommend head-up position to shift SRF to bottom of eye
- If no improvement with corticosteroids, consider externally draining the fluid or performing internal drainage with vitrectomy and applying tamponade

Summary

- 55-year-old female referred for second opinion on OD serous RD and subretinal lesion x 5 months.
- Initially, there was a broad differential including inflammatory conditions (SO, VKH), neoplastic (choroidal tumors, PVRL), and other causes.
- Extensive workup ruled out malignancy and clinical course was not consistent with inflammatory etiology.
- Upon closer review of medical history, her presentation is most consistent with a rare manifestation of protein-losing enteropathy as serous RD.
- This case demonstrates the importance of considering ocular pathology in the context of the whole patient and their medical history.

References

1. Alshikho, M. J., et al. (2016). "Intestinal Lymphangiectasia: Insights on Management and Literature Review." Am J Case Rep **17**: 512-522.
2. Craven, M. D. and R. J. Washabau (2019). "Comparative pathophysiology and management of protein-losing enteropathy." J Vet Intern Med **33**(2): 383-402.
3. Nagra N, Dang S. Protein Losing Enteropathy. StatPearls Publishing; 2020. Available at: [Accessed April 25, 2021].
4. Negi, A. and M. F. Marmor (1984). "Effects of subretinal and systemic osmolality on the rate of subretinal fluid resorption." Invest Ophthalmol Vis Sci **25**(5): 616-620.
5. Umar, S. B. and J. K. DiBaise (2010). "Protein-losing enteropathy: case illustrations and clinical review." Am J Gastroenterol **105**(1): 43-49; quiz 50.
6. Venkatramani, J., et al. (2004). "Bilateral Serous Retinal Detachment Due to Protein-Losing Enteropathy." Archives of Ophthalmology **122**(7): 1067-1070.

Thank You

- Dr. Tatyana Milman
- Dr. Carol L. Shields

Photo credit: Roger Barone