What is That Black Spot?

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PGY-4
Grand Rounds
Nov. 11, 2019

Patient Presentation

CC

Decreased vision left eye for several months

HPI

10yo WM who failed a school vision screening, and was then sent to a local optometrist who diagnosed him with histoplasmosis of the left eye. He was then referred to UL Ophthalmology. Denies pain, flashes, floaters, or any recent change in his vision.

History (Hx)

Past Ocular Hx: Bilateral medial rectus recession

Past Medical Hx: Asthma

Meds: albuterol inhaler PRN

Fam Hx: unremarkable

Allergies: NKDA

Social Hx: unremarkable

RoS: 10 pt ROS negative

External Exam

	OD		OS
VA sc	20/20		20/70
Refraction	unable		unable
Pupils	4→2mm	No rAPD	4→2mm
IOP	soft		soft
EOM		Full	
CVF		Full	

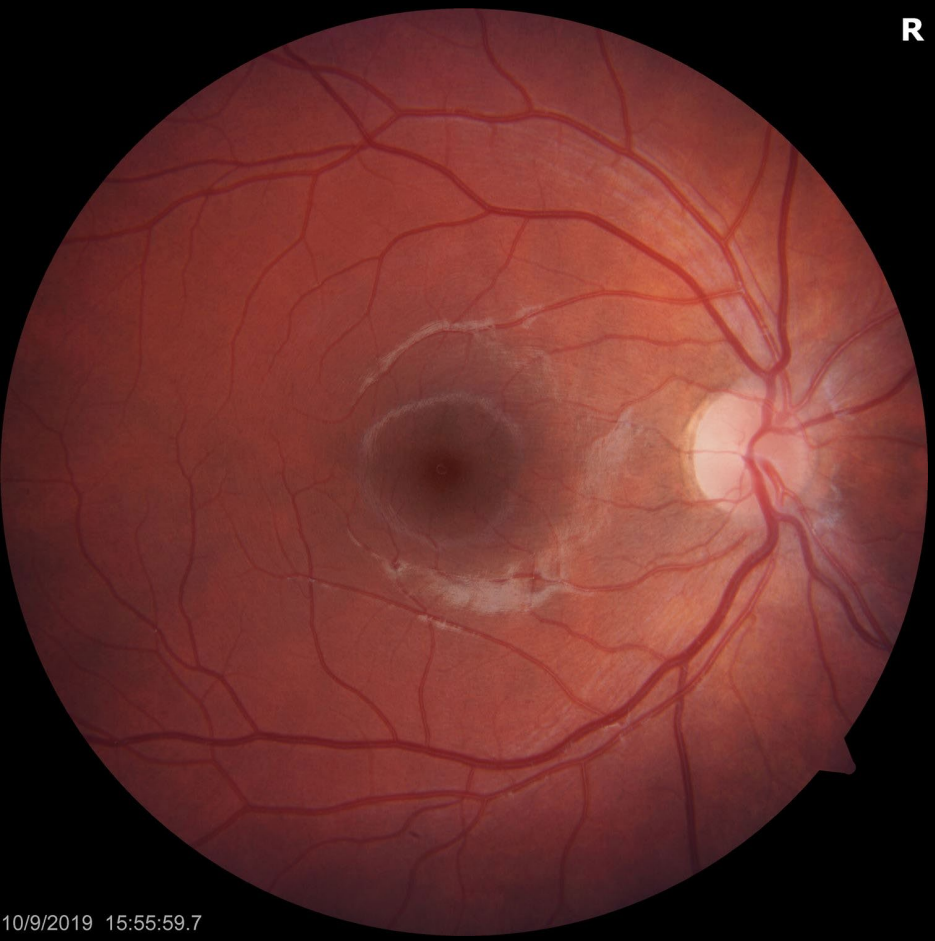
Anterior Exam

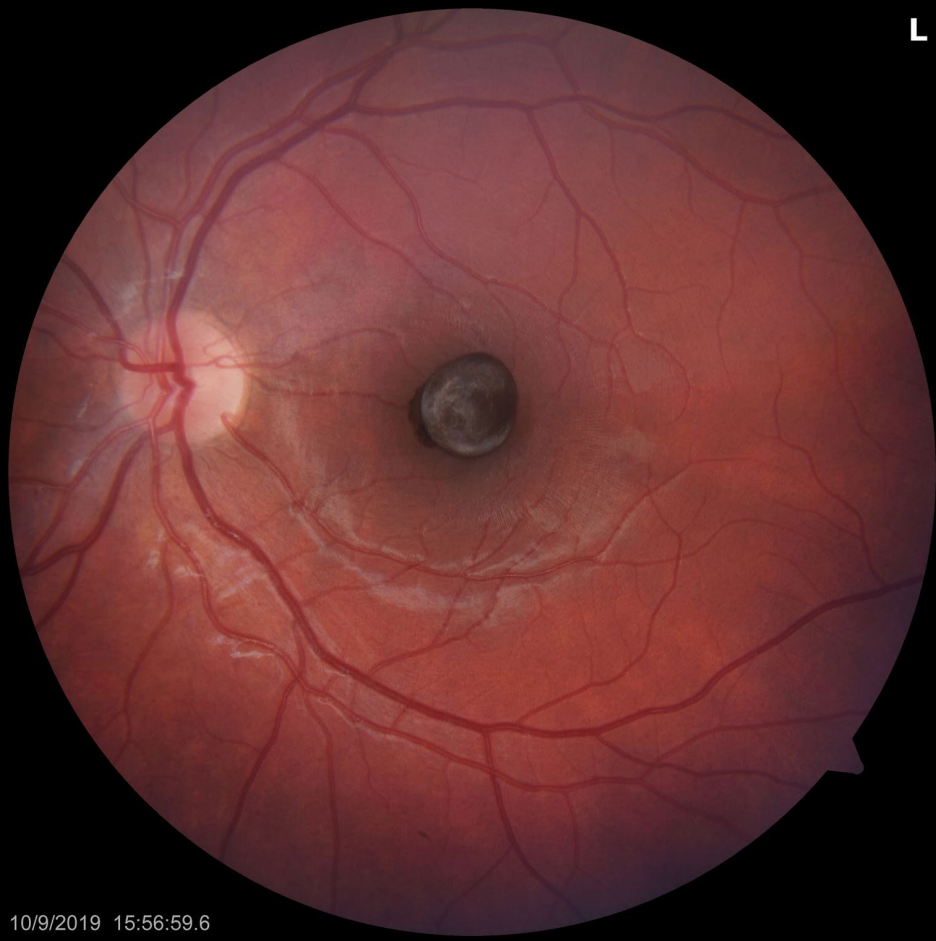
SLE	OD		OS
External/Lids	normal		normal
Conj/Sclera	White & quiet		White & quiet
Cornea	clear		clear
Ant Chamber	Deep & quiet		Deep & quiet
Iris	Flat		Flat
Lens	clear		clear
Vitreous	Clear		Clear

Fundus Exam

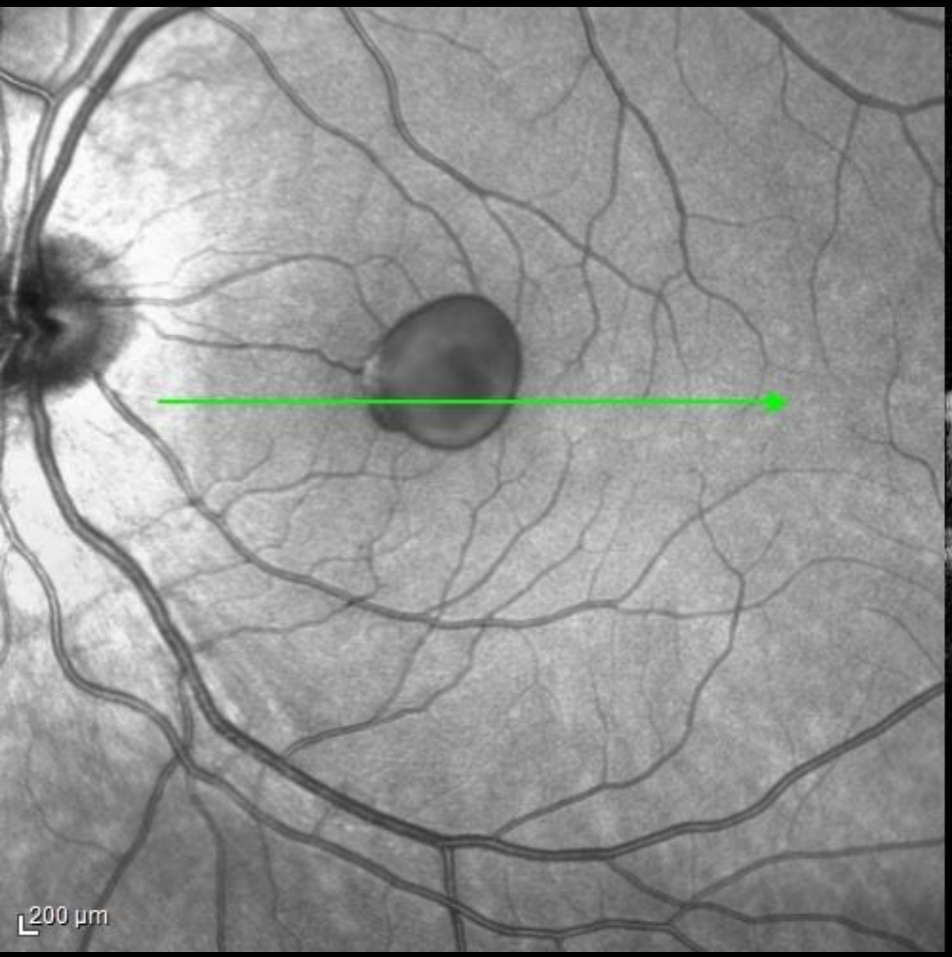
Fundus 20 D	OD		OS
Optic Nerve	Pink/sharp, C/D 0.4		Pink/sharp, C/D 0.4
Macula	flat		Elevated, round pigmented lesion 1DD; juxtafoveal, no organge pigment, no fluid, no heme
Vessels	Normal caliber		Normal caliber
Periphery	Flat, no holes/tears		Flat, no holes/tears

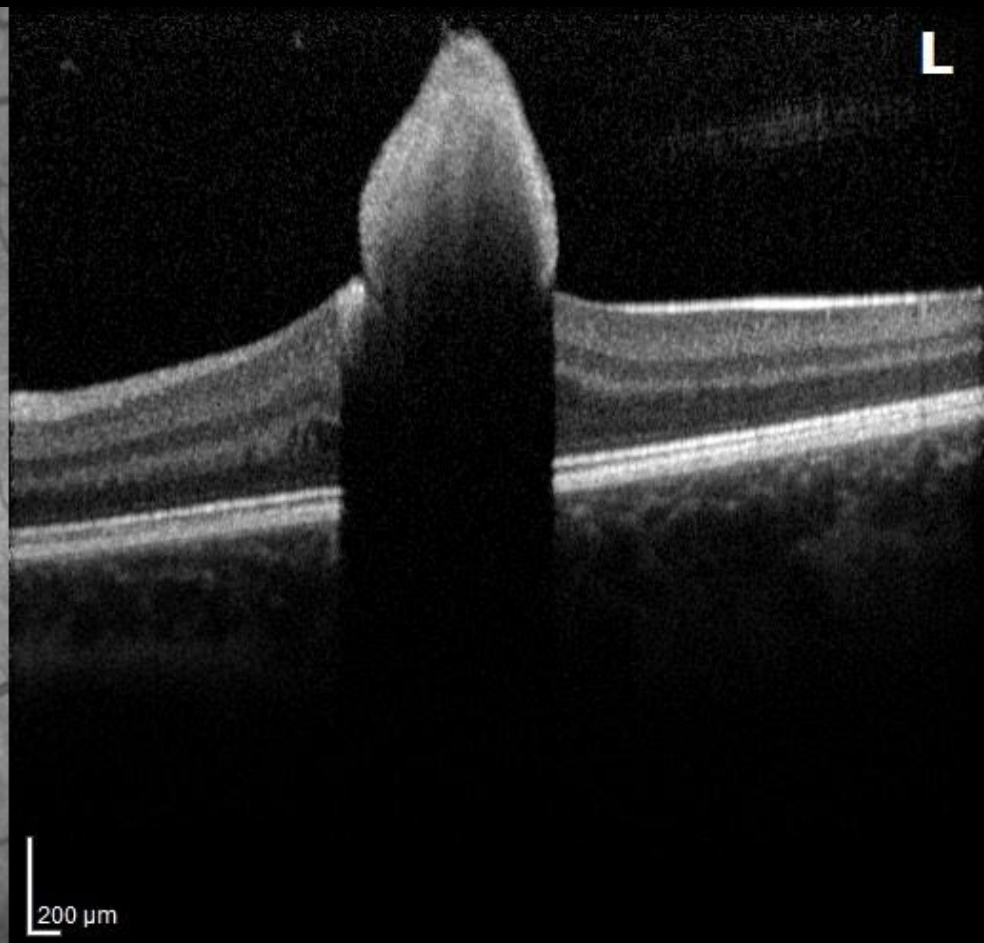
Fundus Photo



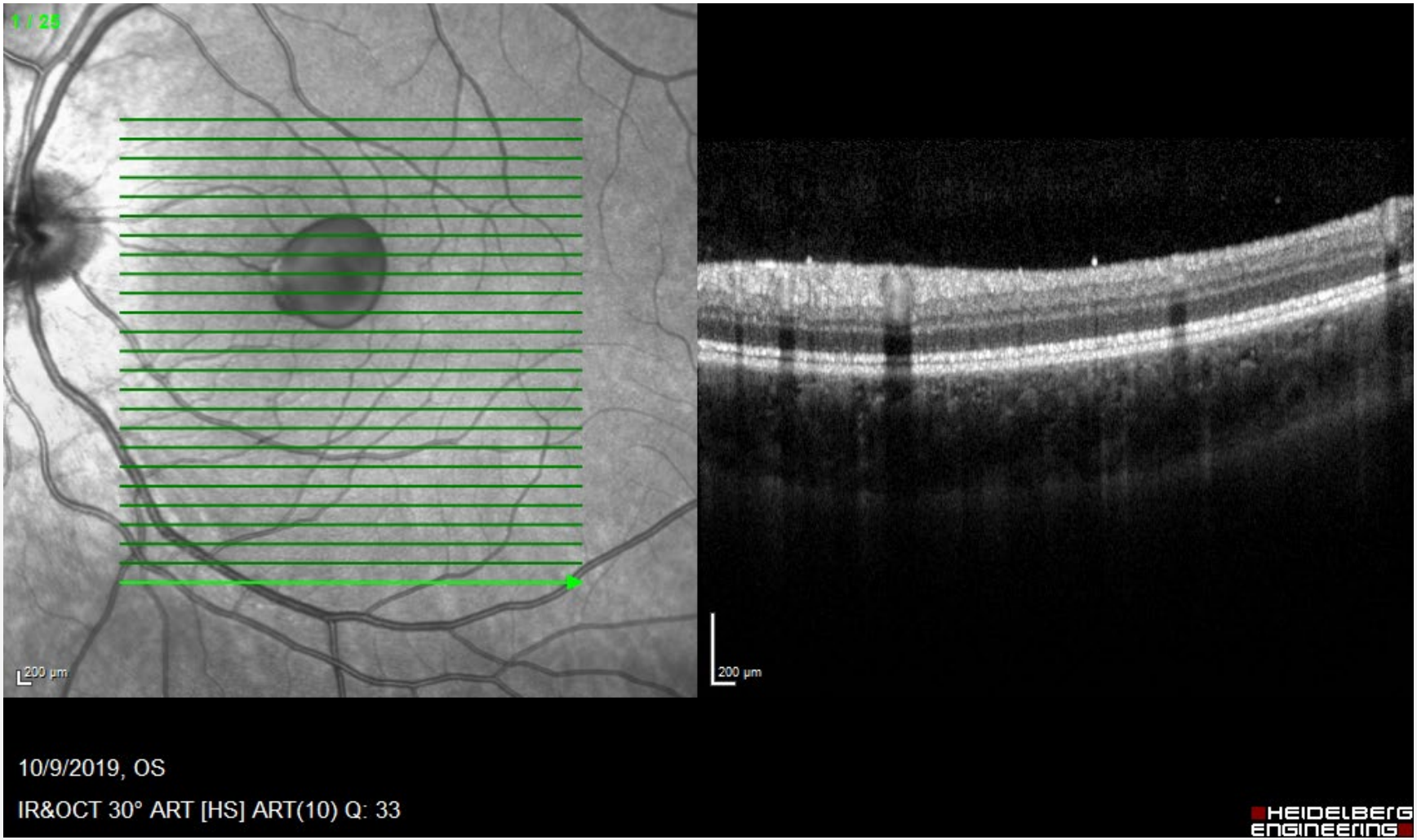


OCT

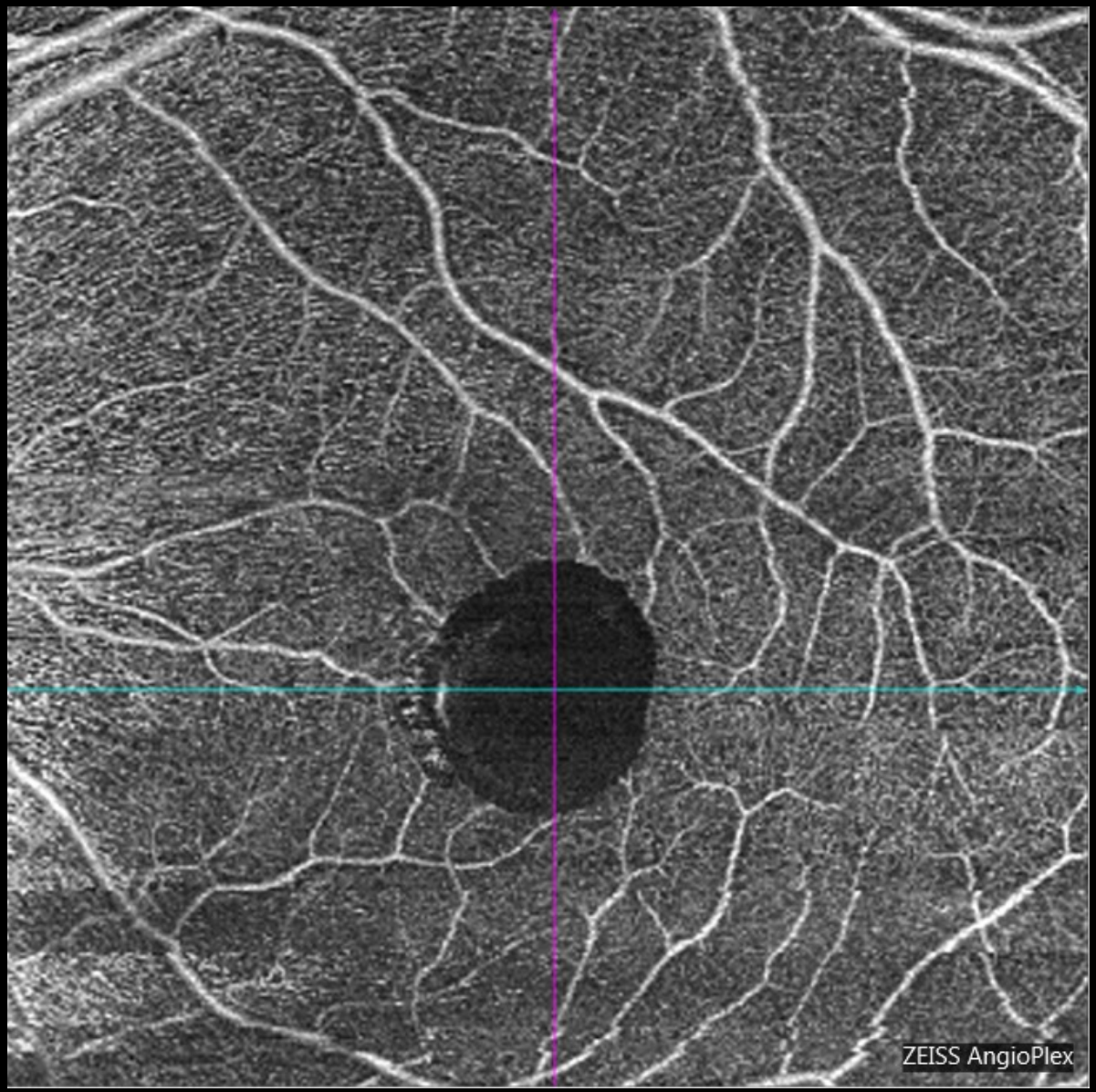




OCT Movie

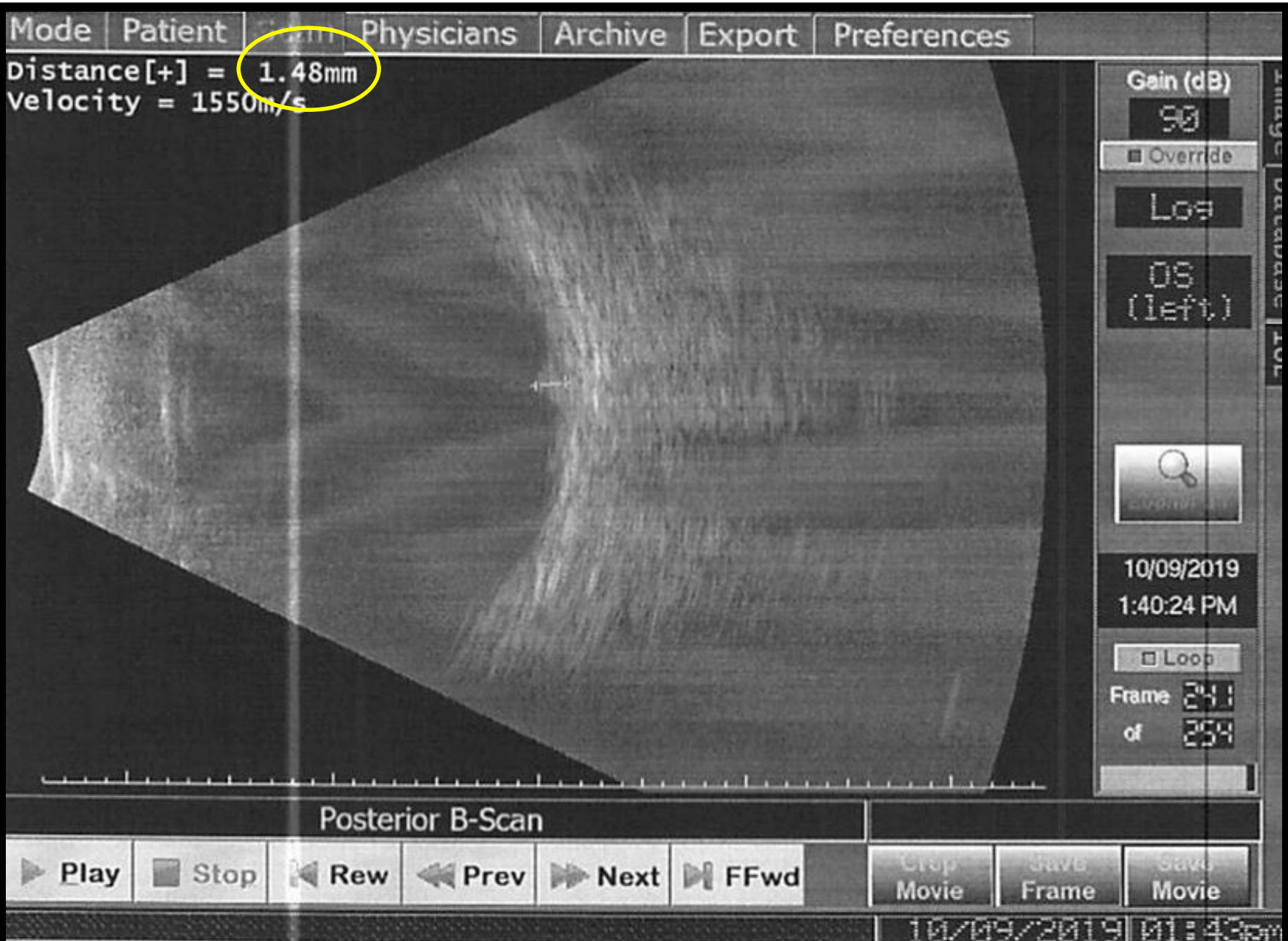

1 / 25
200 µm
200 µm
10/9/2019, OS
IR&OCT 30° ART [HS] ART(10) Q: 33
HEIDELBERG ENGINEERING

OCTA Superficial



OCTA Retina Depth Encoded

B Scan Ultrasound



Assessment

- 10yo with left, pigmented macular mass of unknown duration, otherwise healthy, with h/o strabismus surgery OU

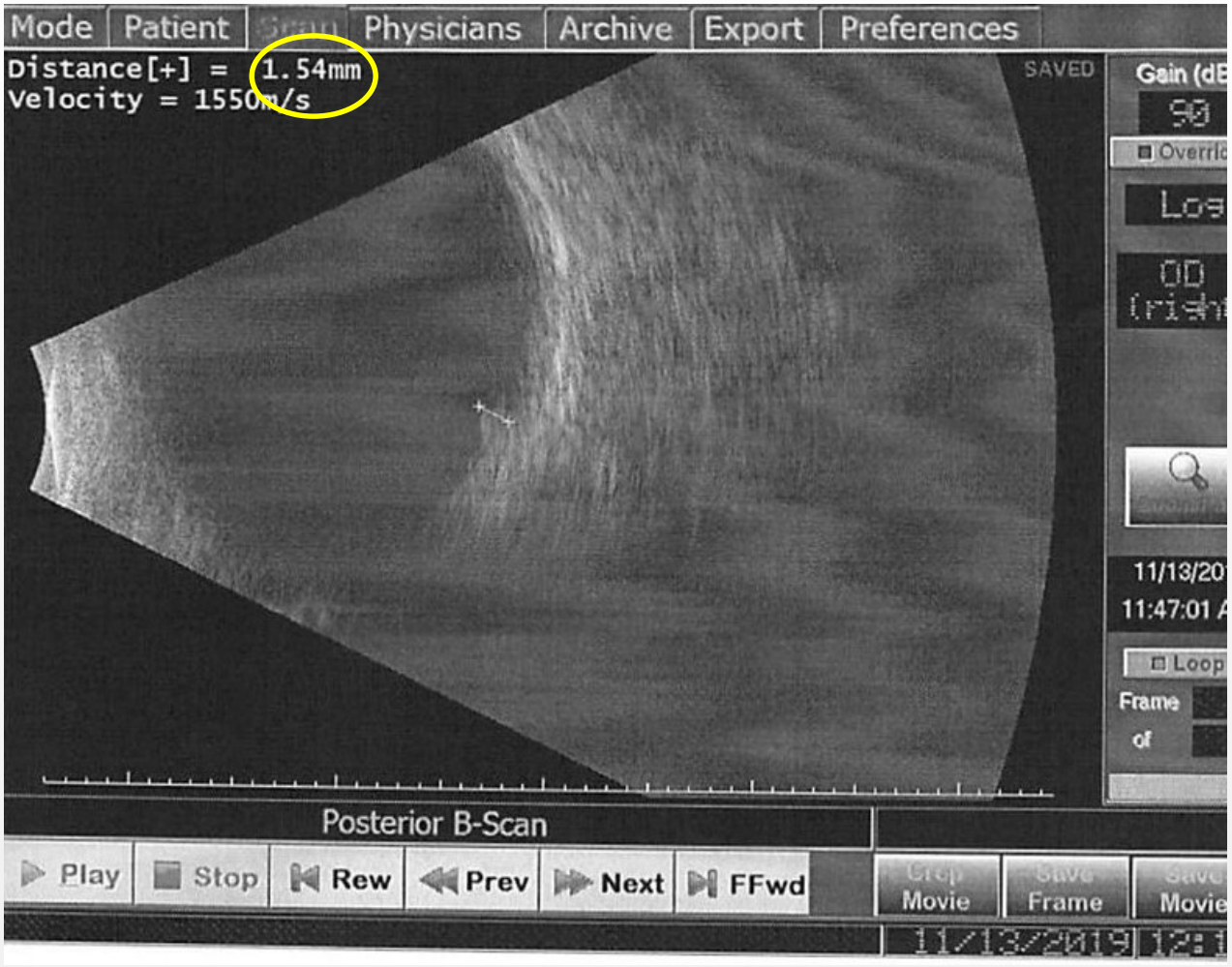
- DDX:
  - Simple Hamartoma of RPE
  - Combined harmatoma of retina & RPE
  - Congenital hypertrophy of RPE
  - RPE adenocarcinoma
  - RPE hyperplasia
  - choroidal melanoma, retinoblastoma
  - Trauma with old hemorrhage

Plan

- Discussed with Dr. Ramasubramanian
- Will observe and return in 1 mo

1 Month Follow-up

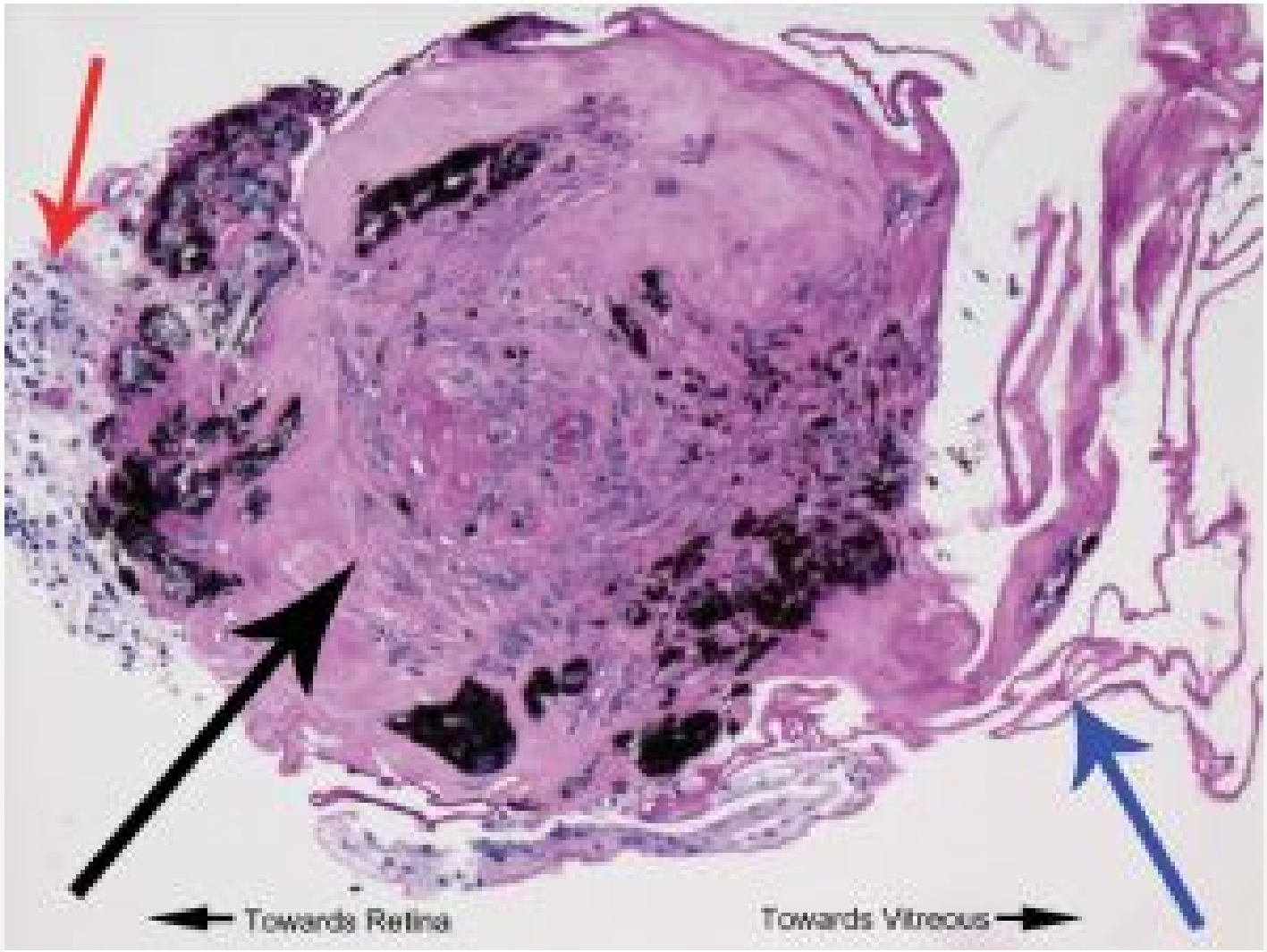
	OD		OS
VA sc	20/20		20/50



Congenital Simple Hamartoma of the RPE

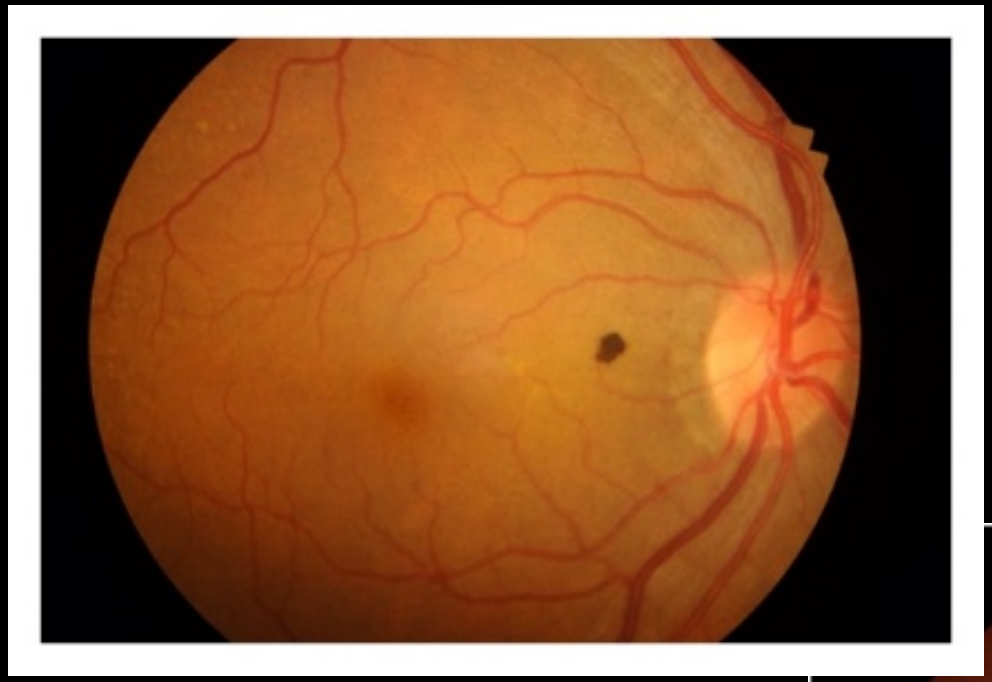
- Very rare, benign
  - Only a handful of reports worldwide
- First described in 1981 by Laqua, who recognized that this lesion belonged to the spectrum of RPE tumors
- 1989, Gass described the clinical features of this tumor using fundus photos and diagrams
- C. Shields et al. published case series in 2003 of 5 cases

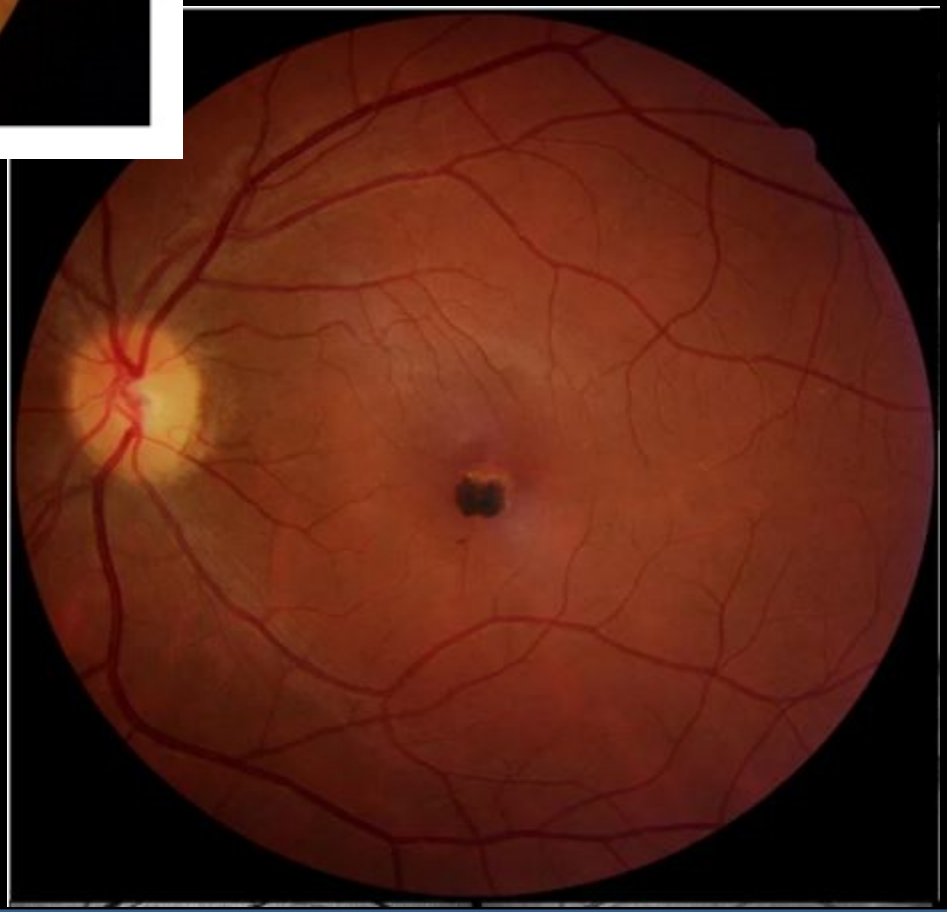
Pathology



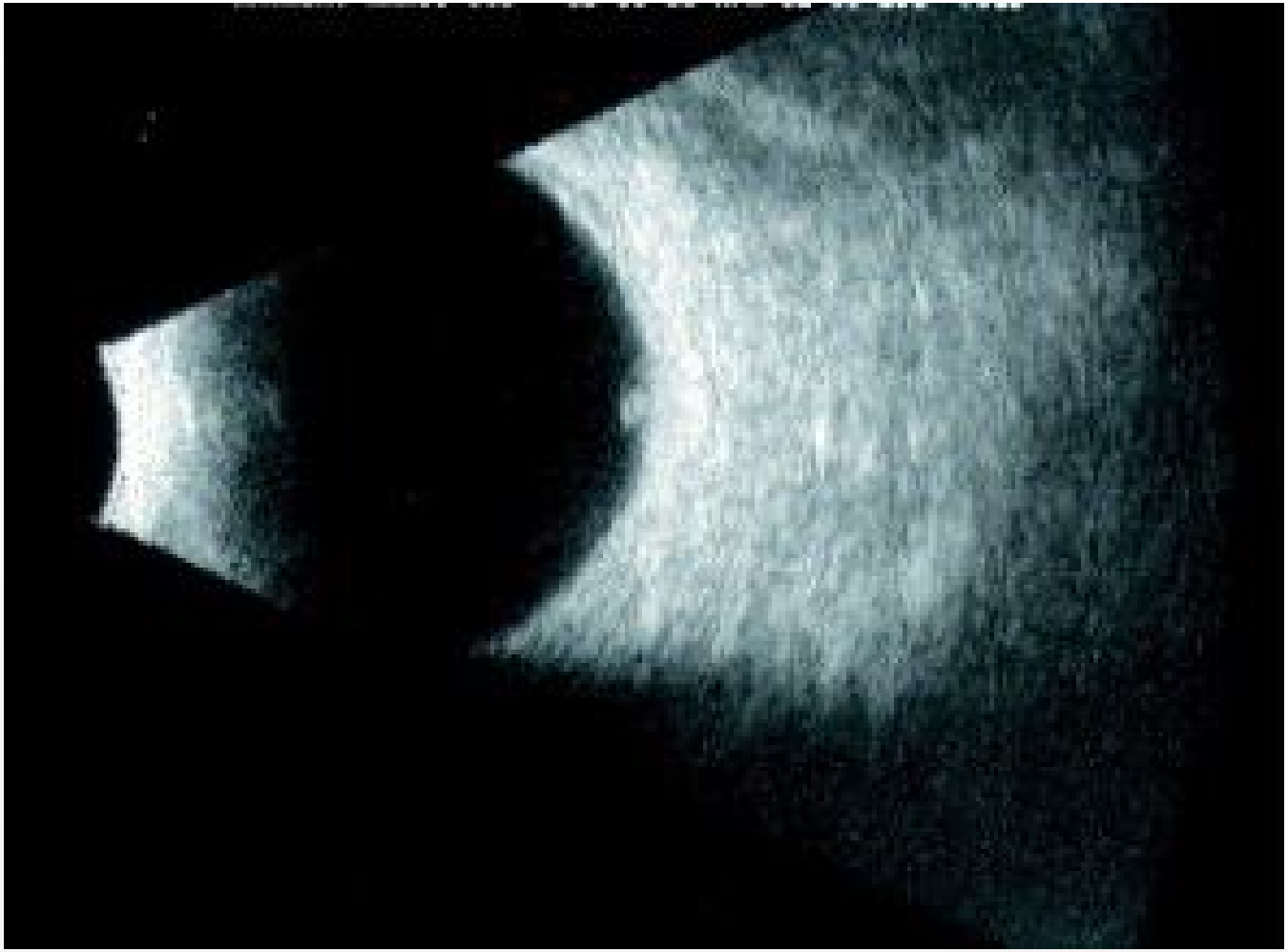
Clinical Features

- Most asymptomatic, incidental finding
- Can cause decreased vision
- Age varies depending upon visual interference: 10-80 years old

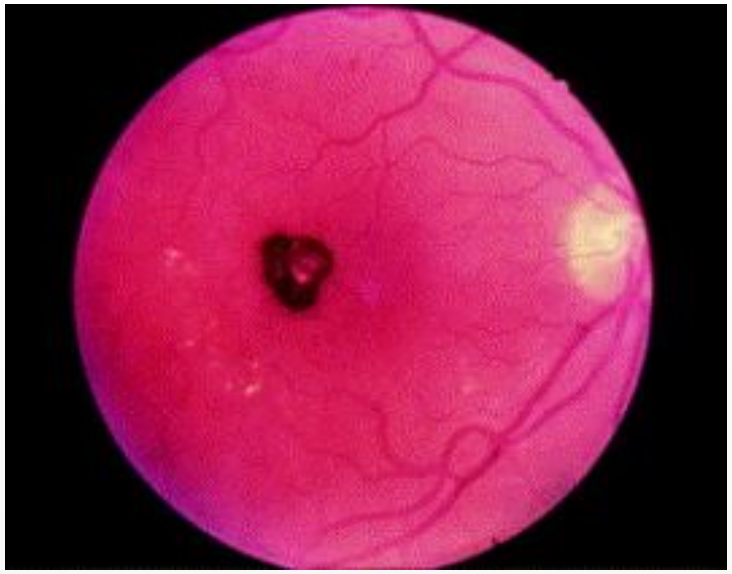


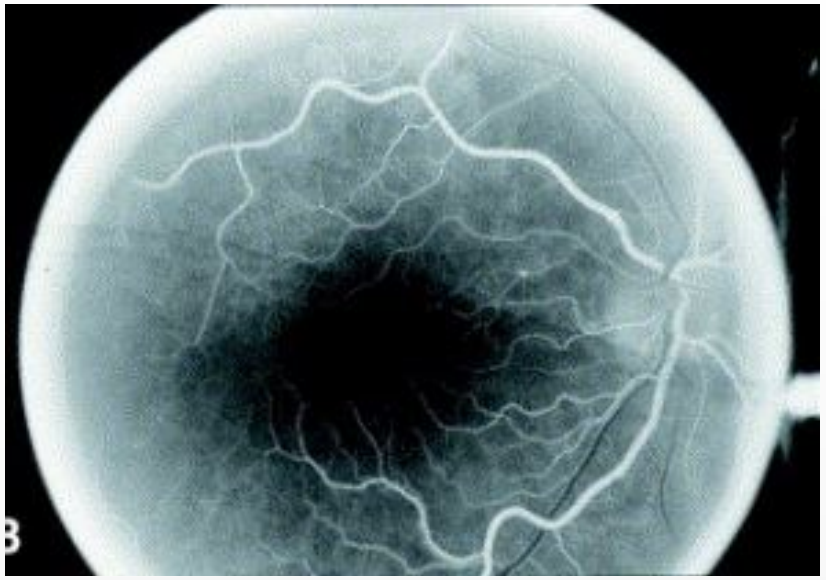


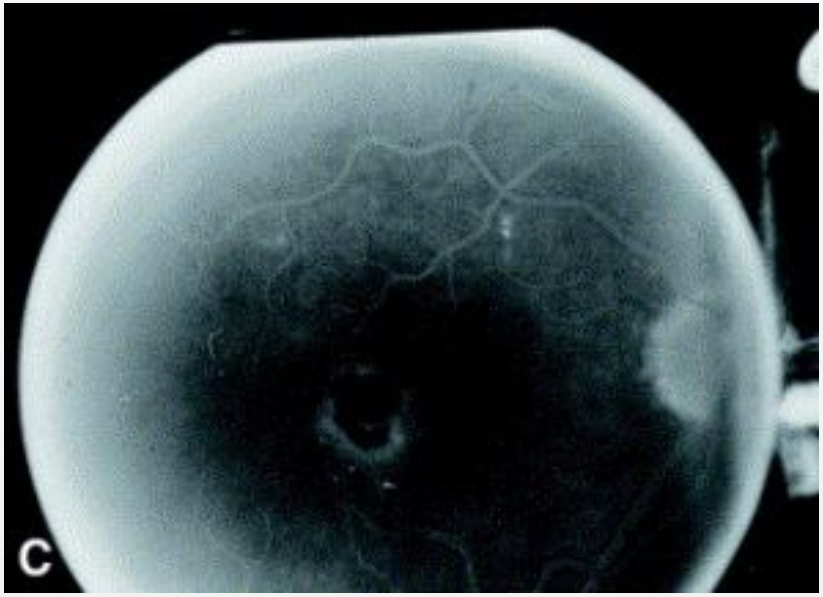
B-scan Ultrasound Features



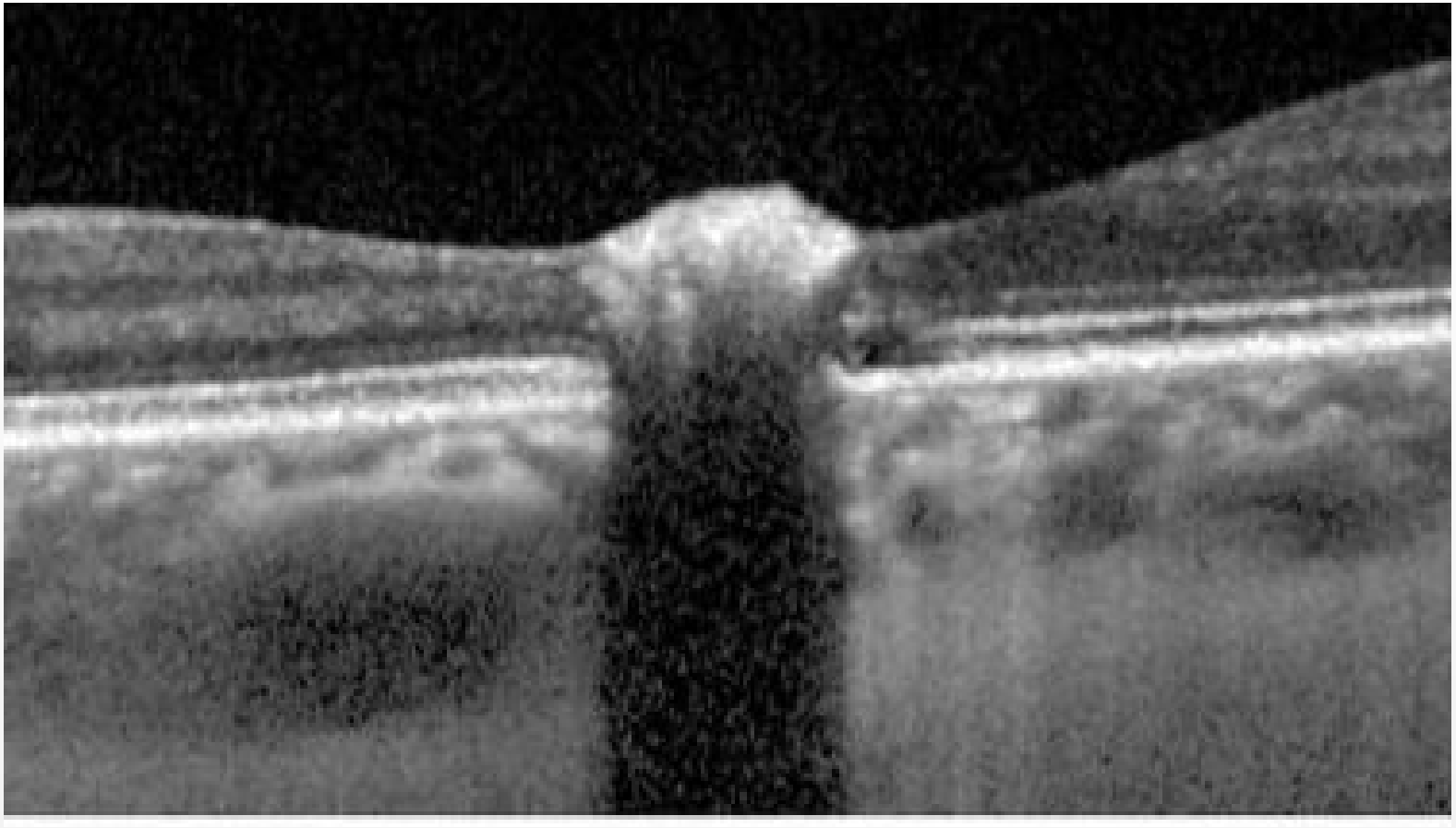
Fluorescein Angiography







OCT Features



OCTA Features

- Arjamand & Shields described in 2019

- “extensive haphazard intratumoral vascular network with fairly large-caliber (100–200 microns) vessels occupying full-thickness tumor, demonstrating more vascular details than visualized on fluorescein angiography”

Associated Complications

- Surrounding retinal/foveal traction
- Vitreous pigmented cells
- Retinal exudation

Prognosis

- Generally good with variable, low grade vision loss
- No report of growth in all cases reported
- Should always see back, as vision could decrease or lesion could grow

Conclusions

- Rare benign tumor or RPE
- Associated vision threatening complications are small
- Management is primarily observation with good prognosis

References

- Ito Y, Ohji M. Long-term follow up of congenital simple hamartoma of the retinal pigment epithelium: A case report. Case Rep Ophthalmol. 2018 Mar 22;9(1):215-220.
- Teke MY, et al. Congenital simple hamartoma of reintal pigment epithelium: clinical and imaging findings. Case Rep Ophthalmol Med. 2012;2012:654502.
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- Barnes AC, et al. Congenital simple hamartoma of the retinal pigment epithelium: clinical, optical coherence tomography, and histopathological correlation. Eye (Lond). 2014 Jun;28(6):765-6.
- Rodrigues MW, et al. Retinal sensivity and photoreceptor arrangement changes secondary to congenital simple hamartoma of retinal pigment epithelium. Int J Retina Vitreous. 2019 Jan 15;5:5.

Thank you