AAO 2019 Course 630
Cases with a Point
SEP Case 1

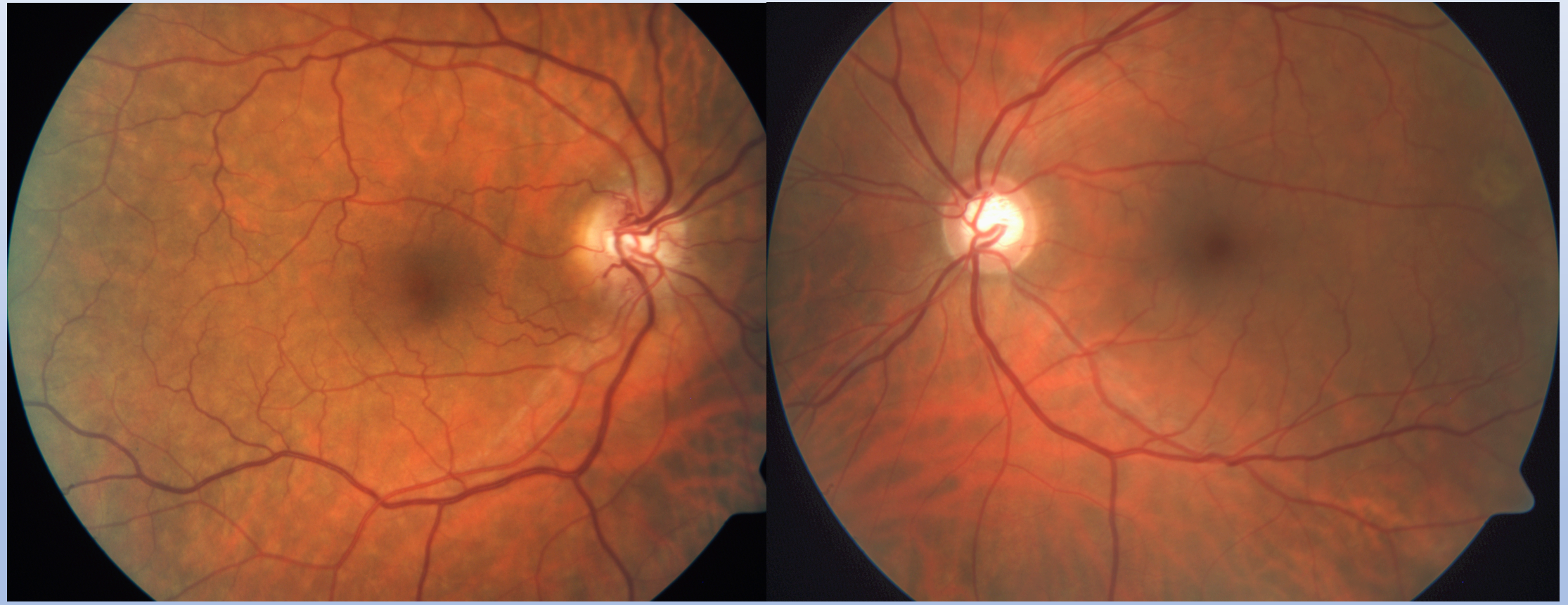
Scott E Pautler, MD

Tampa, FL

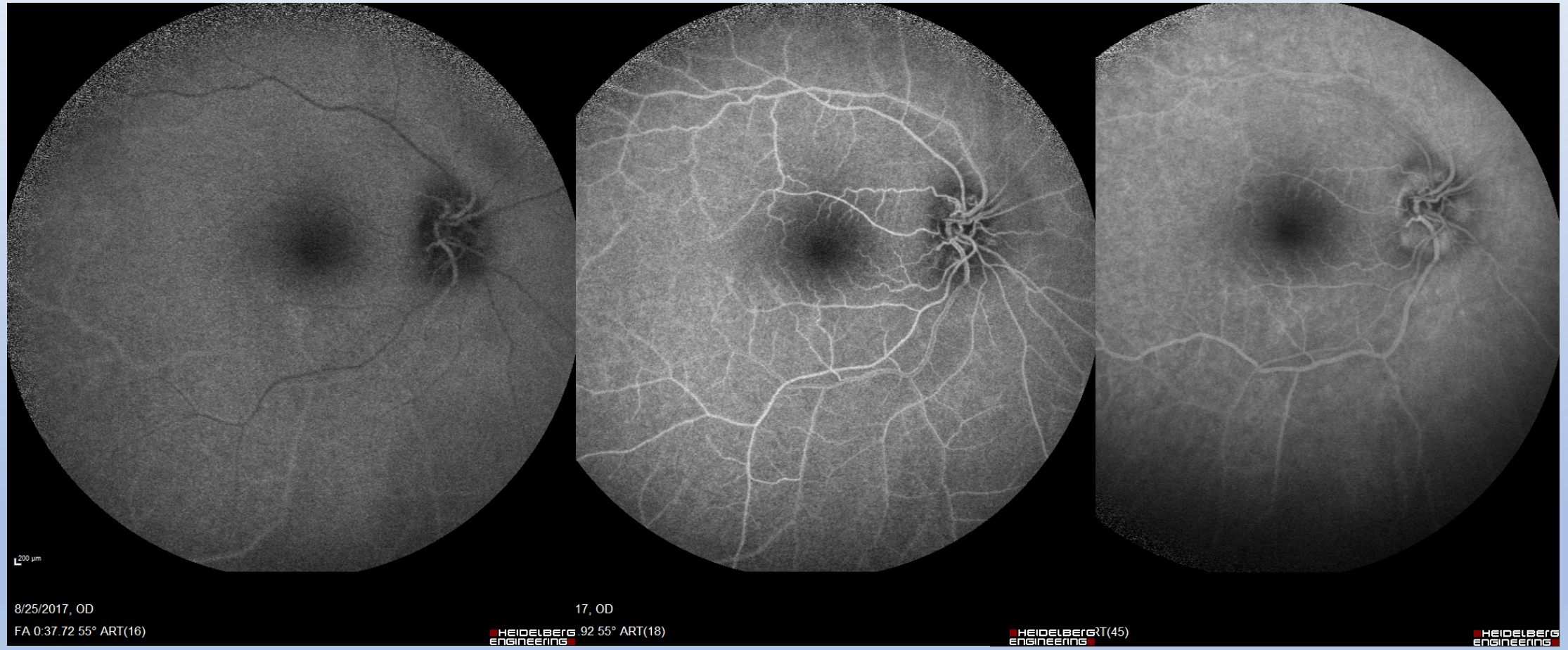
Blurred vision OD

- 54yo WM with blurred vision for a month OD
- Unremarkable PMHx, ROS, FmHx, Soc Hx
- Va 20/25 J1+ OD, 20/20 J1+ OS
- Kruckenberg Spindle and Scheie Stripe OU; no glaucoma dx

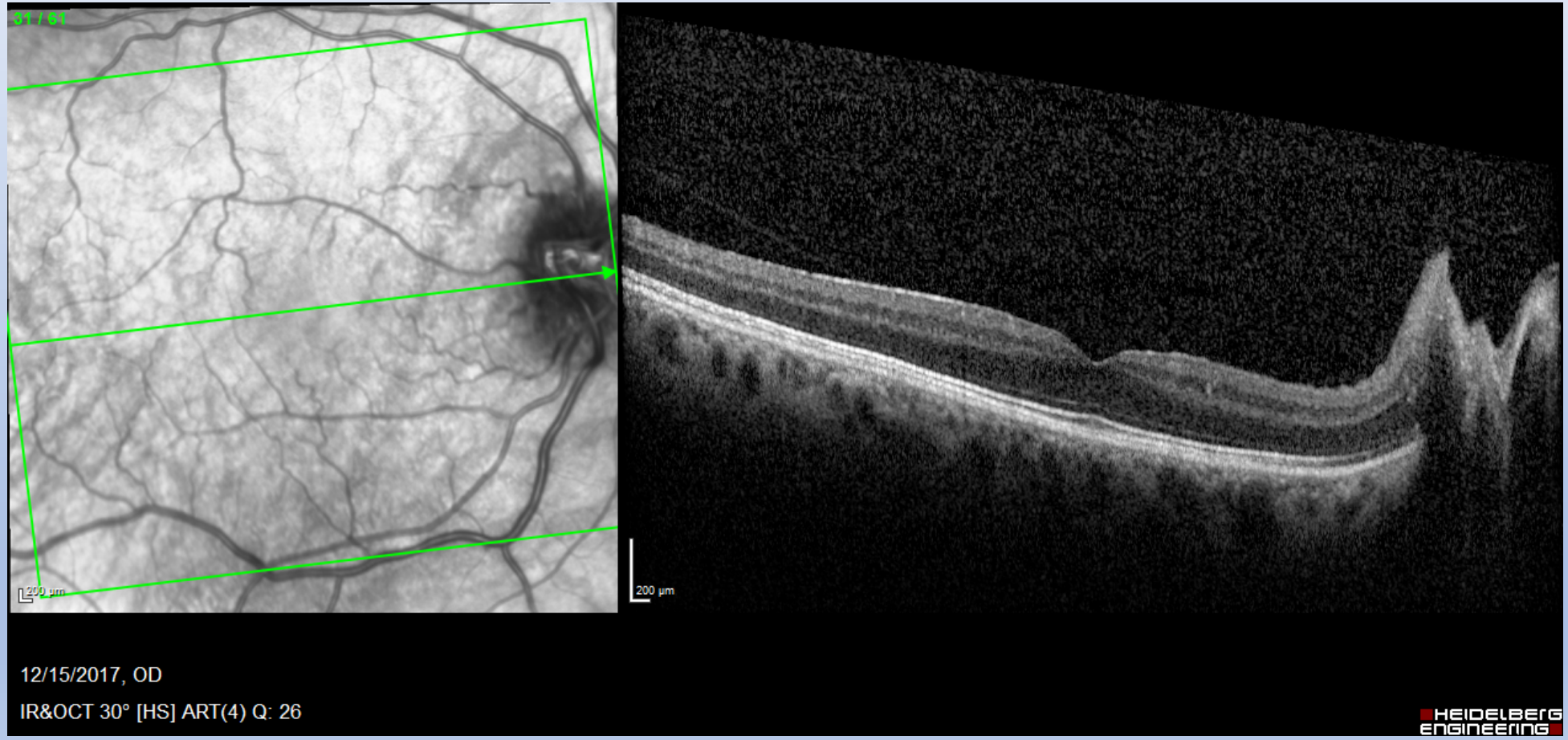
Fundus Photos



Fluorescein Angiography

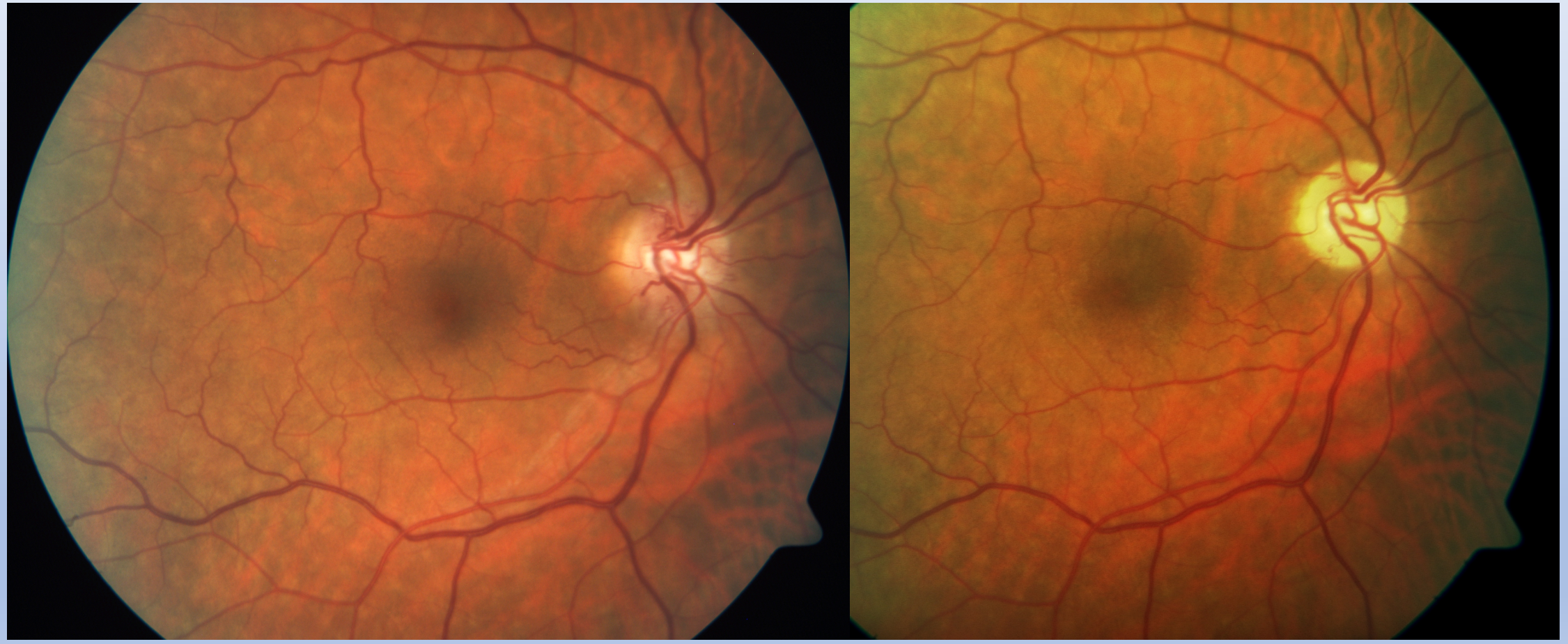


SD-OCT


31 / 61
200 µm
200 µm
12/15/2017, OD
IR&OCT 30° [HS] ART(4) Q: 26
HEIDELBERG ENGINEERING

Diagnosis and management?

Pt had testing and treatment



What happened

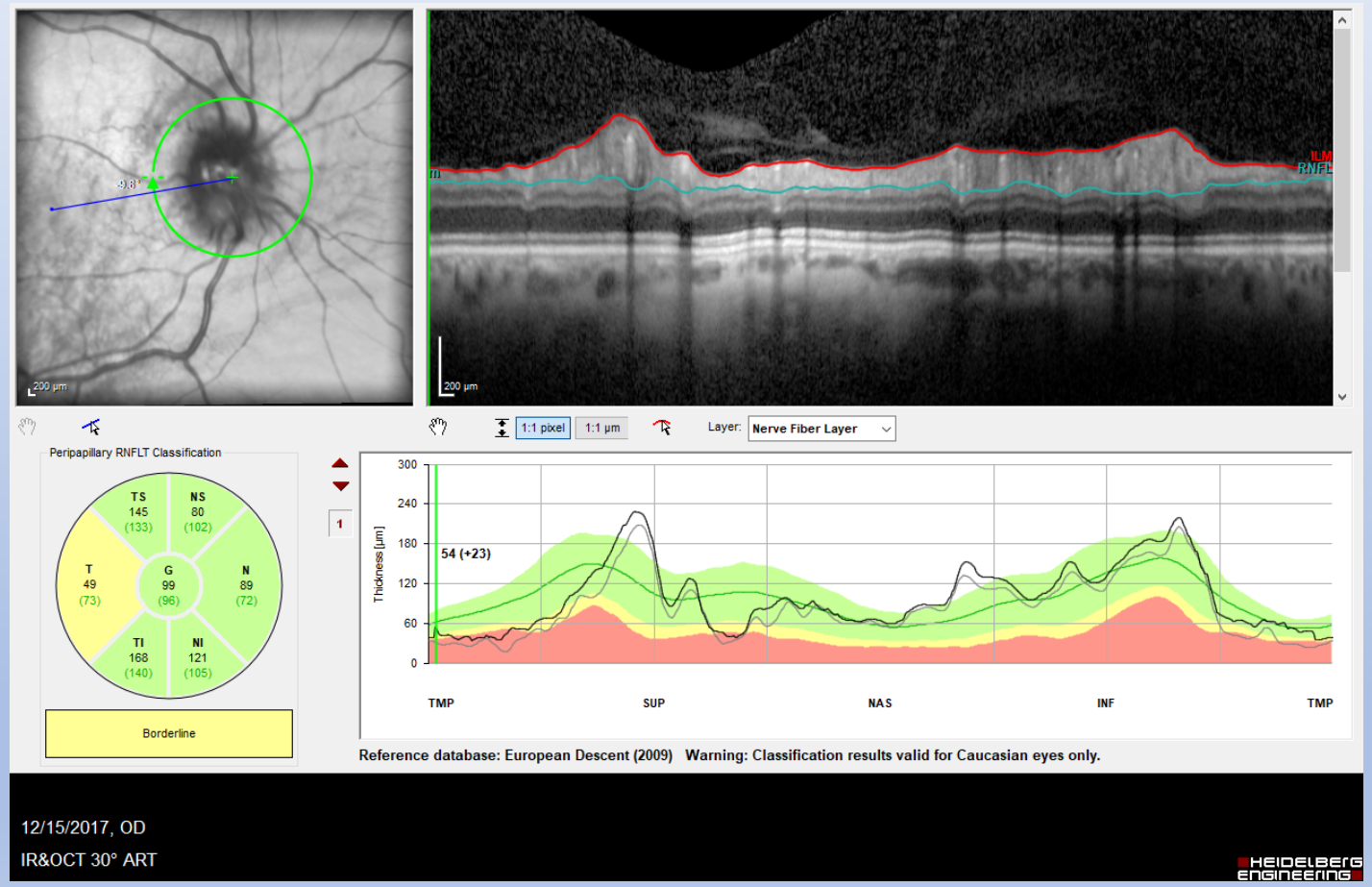
- I thought he had a resolving CRVO associated with glaucoma suspect owing to pigment dispersion syndrome
- Pt moved away and lost to follow up after initial visit.
- He had progressive symptoms with new headaches
- Testing elsewhere revealed intracranial meningioma (resected)
- Disc pallor with 6/200 vision

View in hindsight

Color plates:

OD 8/11

OS 11/11



Optic Disc Collaterals (Opto-Ciliary Shunts)

- Central retinal vein occlusion (acute obstruction by thrombosis)
- Chronic obstruction of central retinal vein:
  - Chronic glaucoma (deformation of lamina cribrosa vs CRVO?)
  - Spheno-orbital meningioma
  - Optic nerve glioma
  - Chronic papilledema
  - Optic disc drusen
  - Intracranial aneurysm
  - Meningocele of optic nerve

Optic disc collaterals: when to order MRI

- No clear-cut evidence of previous CRVO?
  - Required evidence (retinal heme, FA capillary ectasia)?
  - Asymptomatic vs Hx acute past loss of vision?
- Unexplained reduced acuity or color plates?
- Atypical findings:
  - collaterals with disc edema but no retinal heme?
  - Bilateral disc collaterals?
  - Decreased vision with normal macular OCT and/or signs of optic neuropathy?

Points of this case

- Although CRVO is the most common cause of optic disc collaterals, remain vigilant for less common causes
- Order neuroradiological studies in atypical cases

Thank you

Scott E Pautler, MD

Tampa, FL

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Scott E. Pautler, MD
Tampa, FL

Case 1
Optic Disc Collaterals (Opto-Ciliary Shunts)

Optic disc (venous) collaterals (ODC) represent a dilation of pre-existing vascular channels that connect the retinal venous circulation on the optic disc with the peripapillary choroidal circulation. They develop when there is increased resistance to retinal venous outflow. The most common cause of ODC is central retinal vein occlusion (CRVO) in which thrombosis occurs in the central retinal vein at the lamina cribrosa. Other less common causes of ODC include chronic glaucoma (due to chronic compression of the central retinal vein from a posterior displacement of the lamina cribrosa or possibly an epiphenominal risk factor of prior CRVO?), spheno-orbital meningioma, optic nerve glioma, chronic papilledema of any cause, optic disc drusen, intracranial aneurysm, meningocele of optic nerve.[1,2] There is no specific treatment for optic disc collaterals; management is centered around the underlying cause.

In many cases of optic disc collaterals, there is evidence of previous CRVO. Acute CRVO may have been documented in the past. Residual blot hemorrhage may be present after acute onset as they take months to resolve. Fluorescein angiography may show capillary damage from CRVO in the distant past. In these situations, further investigation into the cause of optic disc collaterals may not be necessary.

In some cases, the cause of optic disc collaterals may not be obvious. There may be no predisposing risk factors (table 1) or evidence of previous central retinal vein occlusion. The clinical course may be atypical for CRVO (table 2).

Table 1: <u>Absence of risk factors for CRVO</u>:
Young patient
No vascular risk factors (HTN, lipids)
No glaucoma
No hypercoagulability

Table 2: <u>Atypical clinical course:</u>
No acute loss of vision from CRVO
No retinal hemorrhage or macular edema
Unexplained headache or neurological symptoms
Disc collaterals and disc edema/pallor without retinal findings

When there is no evidence of CRVO, other causes of optic disc collaterals must be considered as listed above. Optic disc collaterals have been described in chronic glaucoma. In some of these cases, there may have been past CRVO (as glaucoma is a risk factor for CRVO). Optic drusen

may be detected on clinical exam or by fundus autofluorescence photography. Optical coherence tomography and B-scan echography are helpful to detect buried drusen.

Magnetic resonance imaging (MRI) of the brain and orbit may be necessary to rule out compressive lesions. Glioma (#1 optic nerve tumor) is usually diagnosed in childhood and is sometimes seen with neurofibromatosis type 1. Meningioma (#2 optic nerve tumor) usually presents in midlife, sometimes associated with neurofibromatosis type 2.[3] Intracranial aneurysms and congenital abnormalities are rare causes of optic disc collaterals. Following MRI brain scan, a spinal tap may be needed to rule-out elevated intracranial pressure in evaluation of papilledema if present on exam.

References:

1.) Masuyama et al. Clinical studies on the occurrence and the pathogenesis of optociliary veins. J Clin Neuroophthalmol 1990; 10:1-8.
2.) Miller et al. Retinochoroidal (optociliary) shunt veins, blindness and optic atrophy: a non-specific sign on chronic optic nerve compression. Aust N Z J Ophthalmol 1991; 19:105-9.
3.) Dutton JJ. Optic nerve sheath meningiomas. Surv Ophthalmol 1992; 37:167-83.