Update on the Management of Diffuse Choroidal Hemangioma

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Disclosures

- None

Introduction

- Diffuse choroidal hemangioma is an uncommon and benign type of vascular hamartoma
- Oftentimes associated with Sturge-Weber syndrome
- Characterized by diffuse choroidal thickening with possible overlying non-rhegmatogenous retinal detachment
- Cystoid macula edema (CME) may also be present

Patients and Methods

- Retrospective review of consecutive series of 11 patients (15 eyes) with diffuse choroidal hemangioma
- All patients had clinical features compatible with Sturge-Weber syndrome
- Patients evaluated between 08-2018 and 01-2020, and followed for 6 to 12 months
- Diagnosis established via history, external examination, indirect ophthalmoscopy, echography, and SD-OCT

Clinical Features

- Mean age 19.3 years
- Eleven patients (4 females, 7 males)
- Secondary glaucoma in eight patients
- Four patients had bilateral diffuse choroidal hemangiomas
- Two patients (2 eyes) had total exudative retinal detachment (RD) to the back of their lens, while a total of seven patients (10 eyes) had varying degrees of exudative RD
- VA variable ranging from 20/20 to HM

Treatment Options

Options

- Observation
- Focal thermal photocoagulation
- Anti-VEGF therapy
- Photodynamic therapy (PDT)
- External beam radiotherapy (EBRT), plaque radiotherapy, charged-particle radiation
- Oral propranolol

Treatment Employed

- Photodynamic therapy (PDT) employed in six patients (6 eyes)
- External beam radiotherapy (EBRT) was utilized in two patients (2 eyes) followed by PDT
- Two patients (2 eyes) had EBRT alone

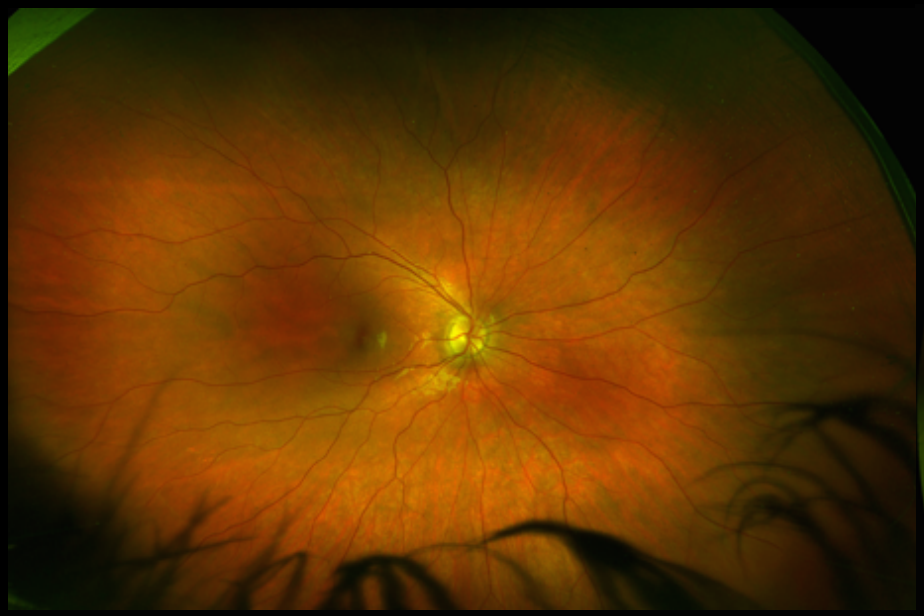
Representative Case

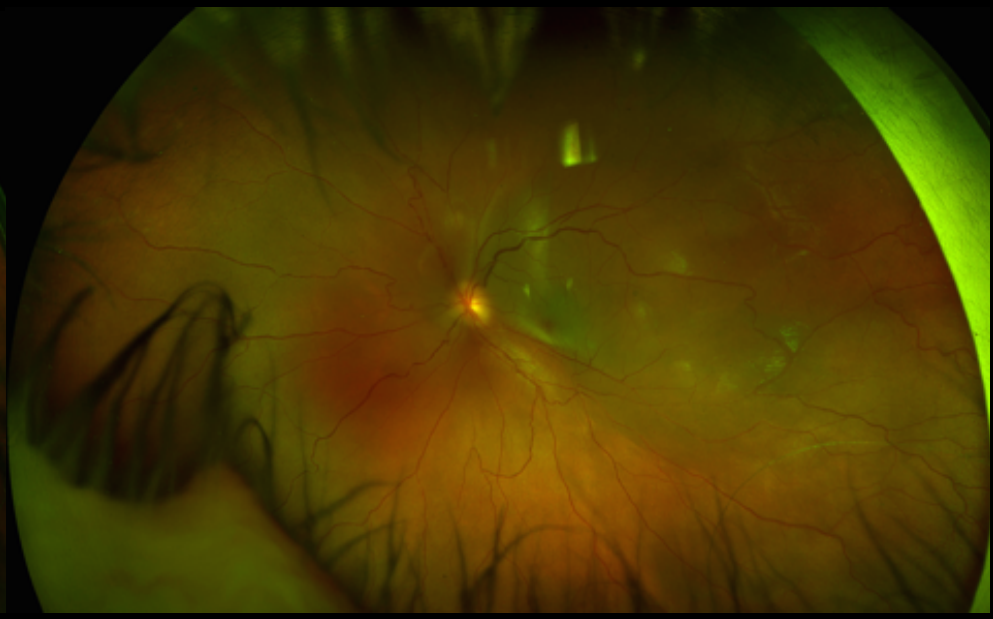
	OD	OS
VA	20/20	6/200
IOP	16	19
CRx (8/19)	-0.75 +0.50 x 090	+4.00 sph
EOM	Full	Full
Pupils	4 to 3 mm	Peaked pupil

Anterior Segment OS

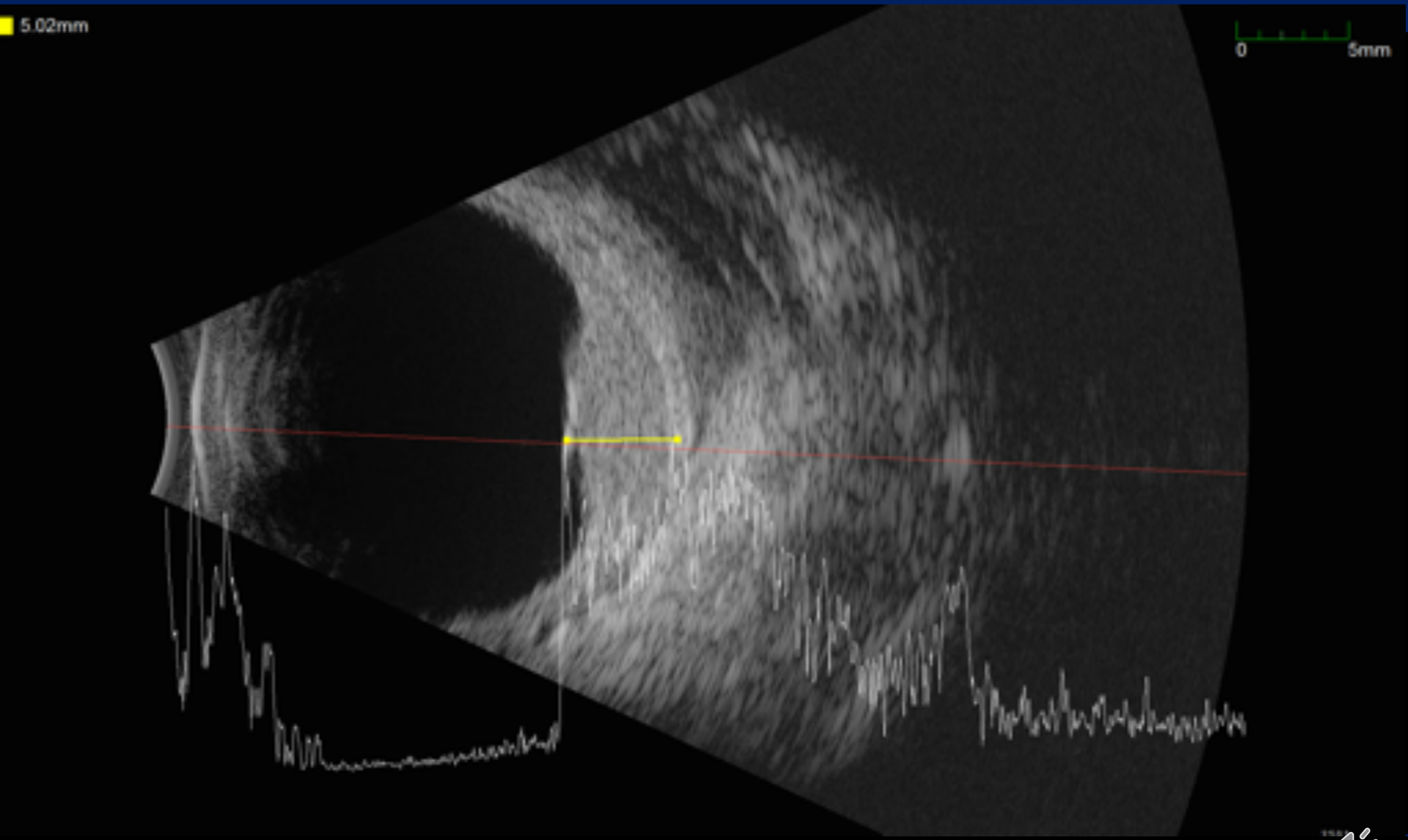
Tube in AC, unobstructed, no tube-cornea touch
Positive posterior synechiae
Anterior capsular pigment

Widefield fundus photo OU





B-Scan OS



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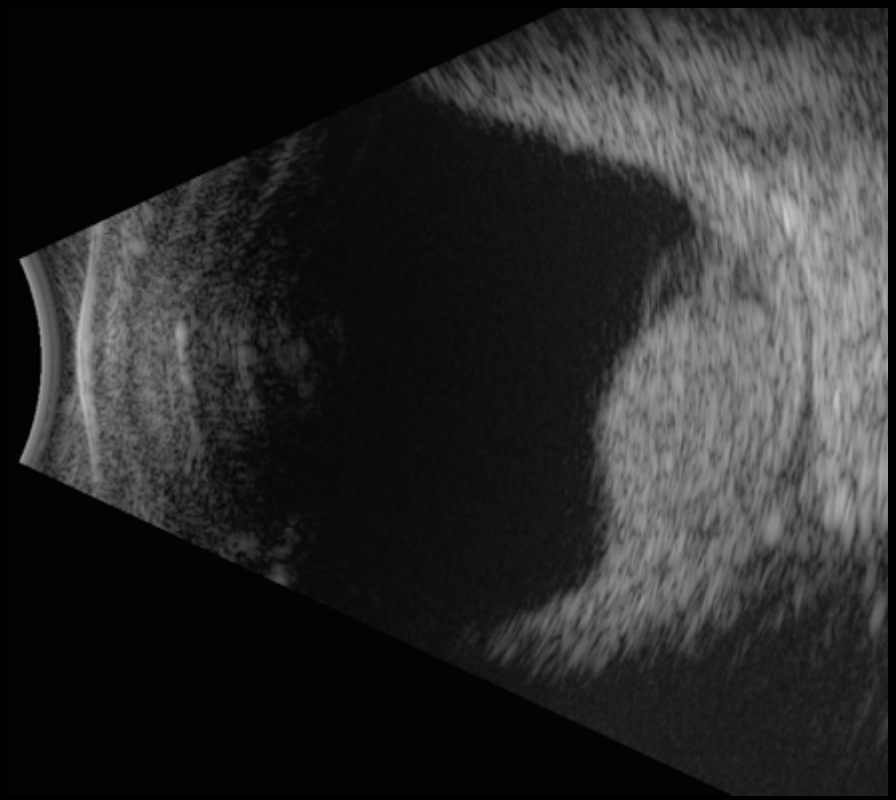
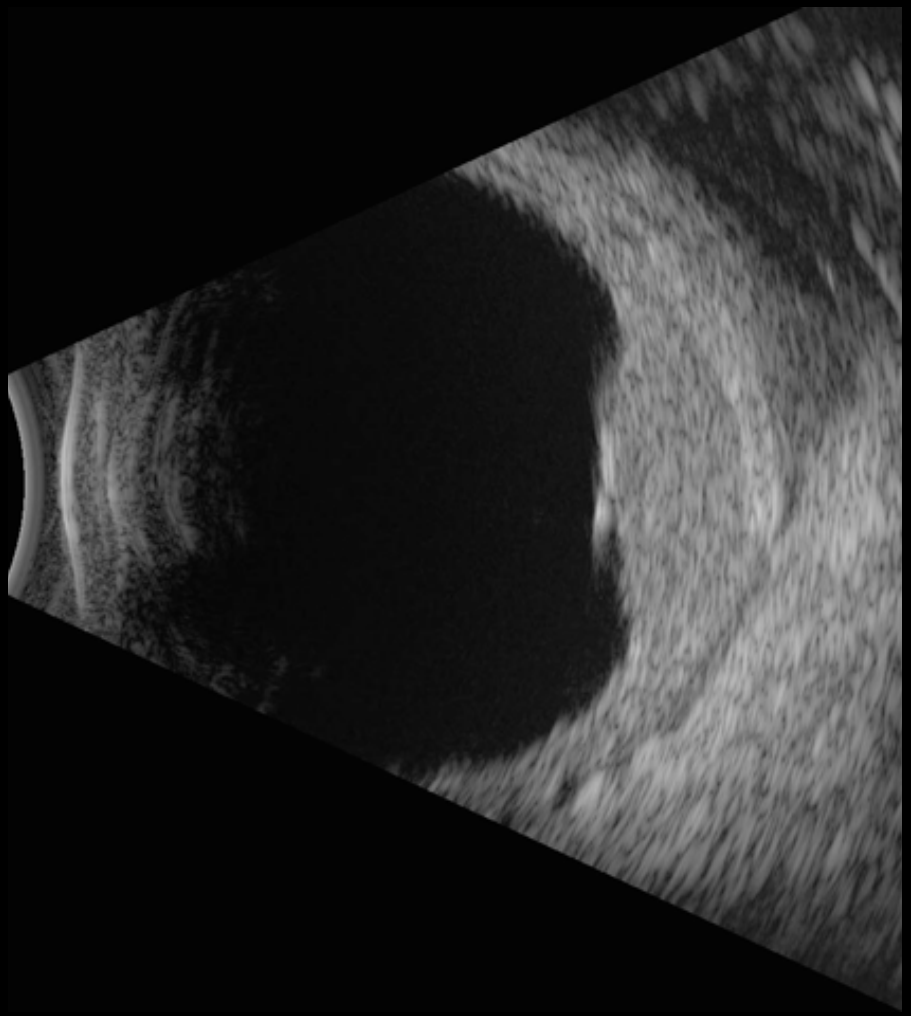
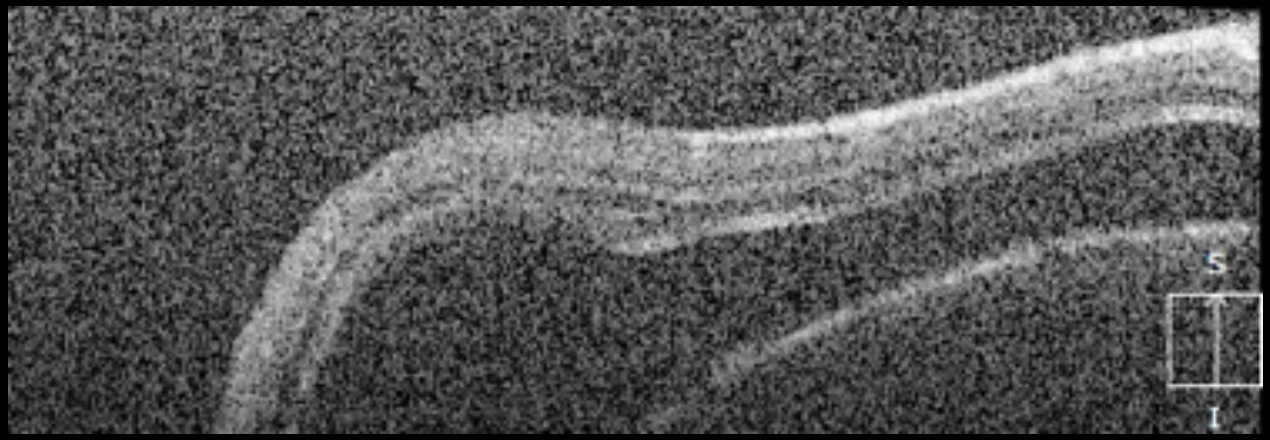




Spectral domain OCT OS



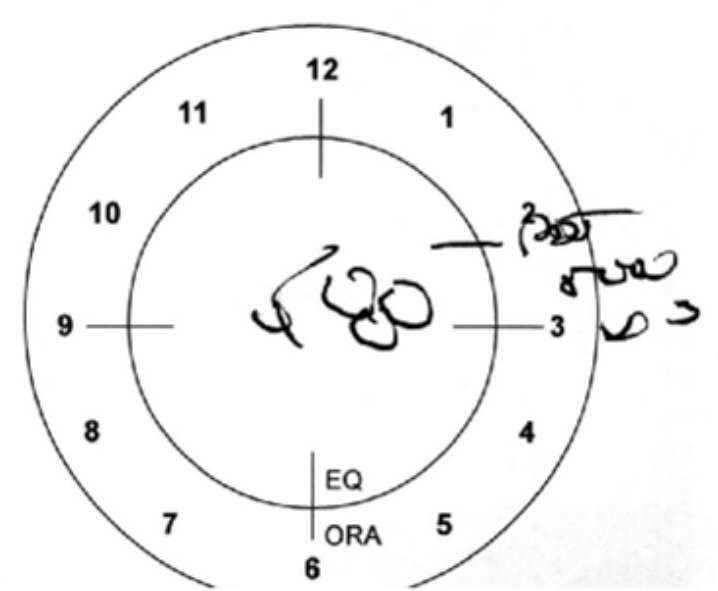
Treatment

Opted to employ

Photodynamic therapy (PDT)

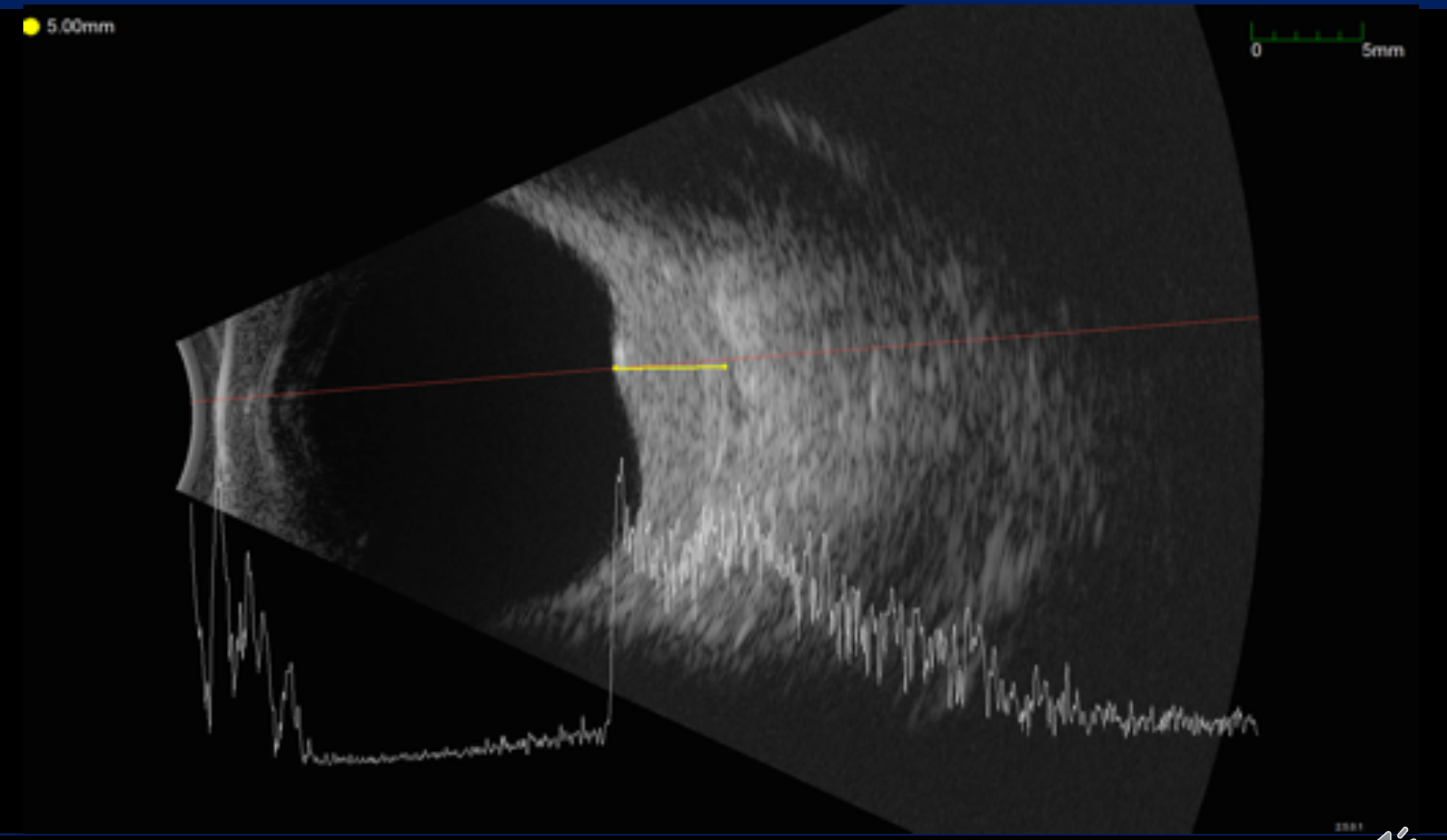
Treatment Response

PDT parameters
Spot size: 5000 µm x 3 minimally overlapping spots
Duration: 83 sec

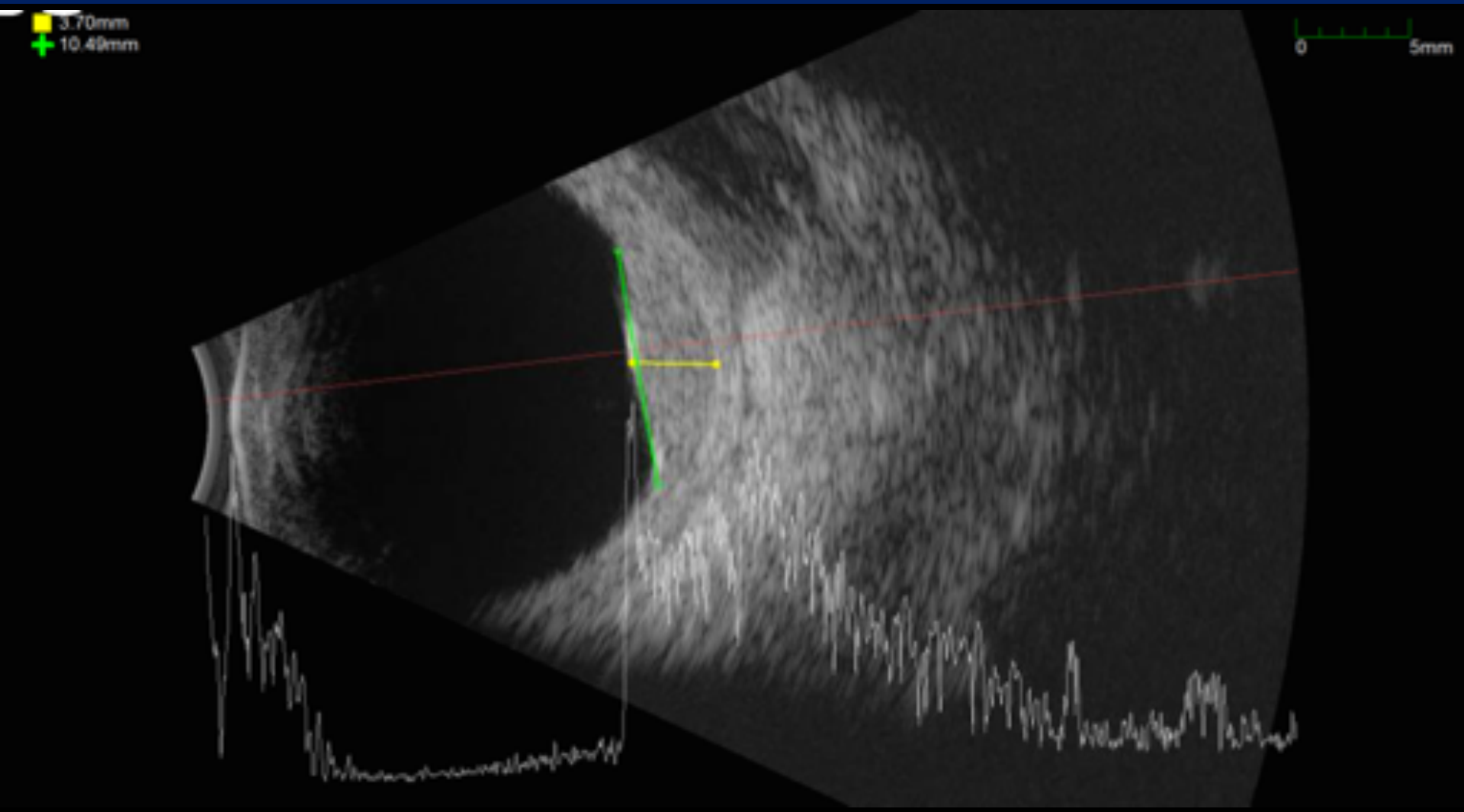


	OS (pre-PDT)	OS (1 month post-PDT)	OS (3 months post-PDT)
VA	6/200	20/70	20/50
IOP	19	19	17

1 month Post-PDT B-scan OS



3 month Post-PDT B-scan OS

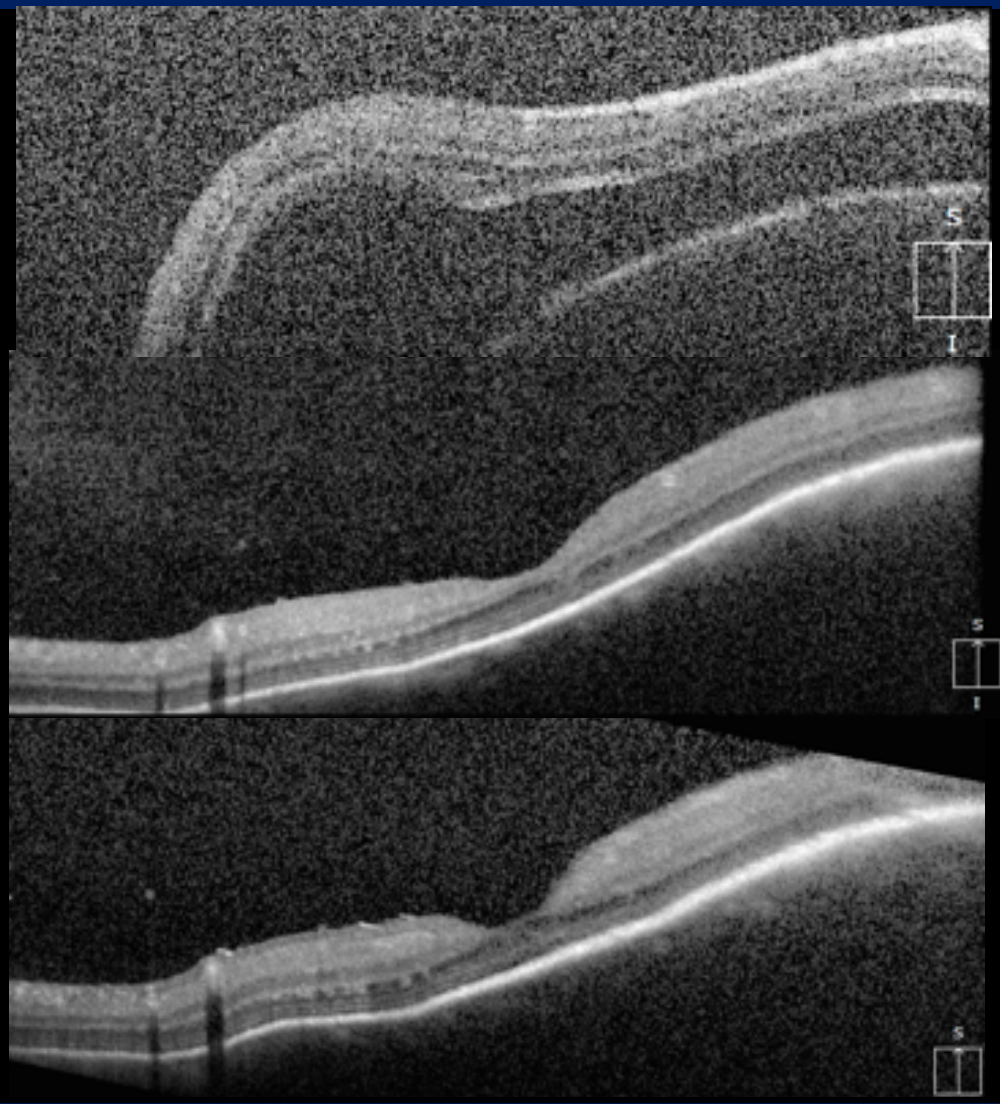


Treatment response on SD-OCT

Pre-treatment

1 month

3 months

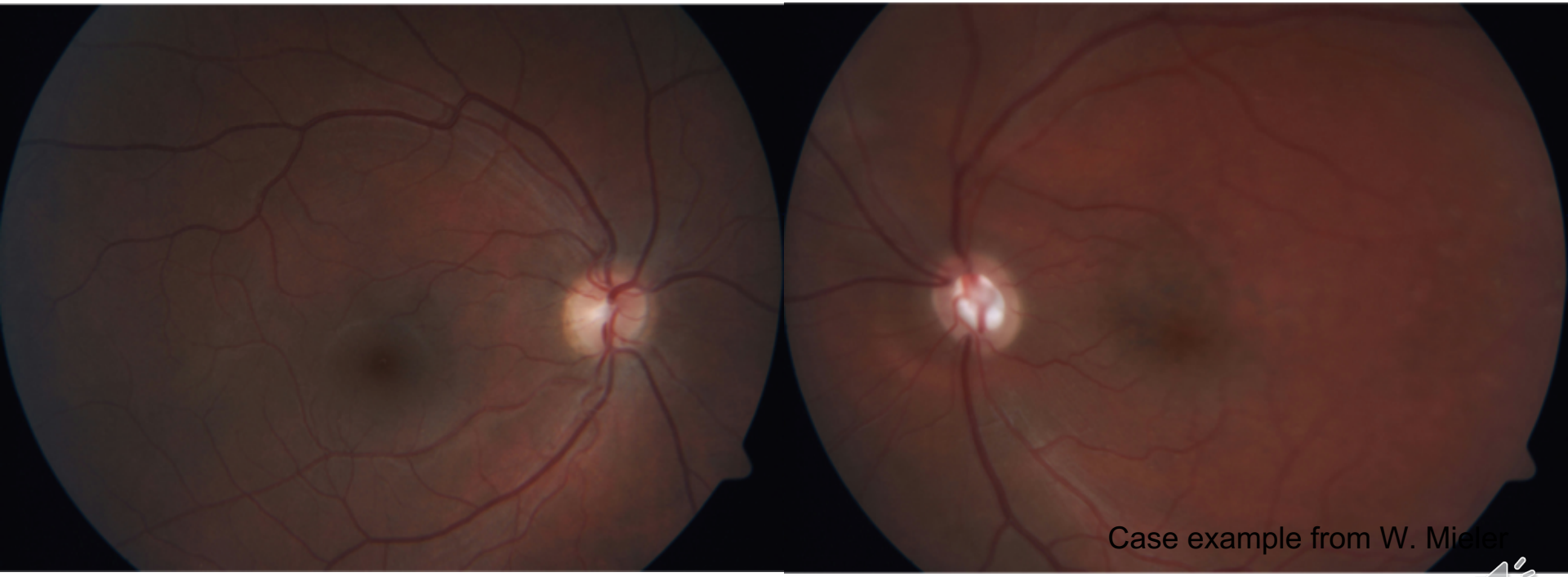


Response to Treatment

- All hemangiomas responded to the PDT and/or EBRT
- Two patients required a second treatment 5-6 months following the initial therapy
- Post-treatment VA varied widely, though all patients showed improvement
- Echographic measurements of choroidal thickness in the macular region dropped on average from 4.3 mm to 2.9 mm

Diffuse Choroidal Hemangioma
Background

- Usually ipsilateral to angiomatous malformation of skin (though may be bilateral)
- More likely to develop secondary RD spontaneously or following glaucoma filtering surgery
- Observed in 50% of patients with Sturge-Weber syndrome



Case example from W. Mieler

Diffuse Choroidal Hemangioma
Treatment Options

1. Photodynamic therapy (PDT)
2. Anti-VEGF therapy
3. External beam radiotherapy
4. Proton beam or stereotactic radiotherapy
5. Plaque brachytherapy (Iodine-125, Ruthenium-106, Cobalt-60)
6. Oral propranolol

Diffuse Choroidal Hemangioma
PDT

- Standard dose of verteporfin (6 mg/m^2), followed by application of 689 nm red laser light to lesion
- Variation in number of spots, spot size (2,500-7,700 µm), duration
- Avoiding extensive overlapping PDT spots → theoretical risk of increased fibrosis in area of overlap
- Generally treat the nodular area of the hemangioma
- There can be slow reabsorption of fluid, and possible initial worsening of exudative RD following treatment

Summary

- Treatment is individualized on a case-by-case basis
- In the presence of an overlying non-rhegmatogenous retinal detachment, PDT is generally deployed
- The entire extent of the choroidal hemangioma generally does not need to be treated
- Visual and anatomic outcomes tend to be favorable (though quite variable)

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Thank You