

MANAGING CIRCUMSCRIBED CHOROIDAL HEMANGIOMA

The case of a patient with a benign vascular tumor that did not respond to PDT but that was ultimately controlled with low-dose plaque radiotherapy.

BY ALEX ALTMAN, BS; KUNAL MALIK, BA; AND CAROL L. SHIELDS, MD



Choroidal hemangioma is a benign vascular tumor of the posterior segment that can manifest as a small circumscribed tumor or an extensive diffuse tumor.¹

Circumscribed choroidal hemangioma is sporadic, unilateral, and commonly overlooked on fundusoscopic examination because the tumor color blends nearly imperceptibly with the normal choroid.² Diffuse choroidal hemangioma appears as an ill-defined orange-colored mass, deep to the retina, often with overlying subretinal fluid (SRF). It is most commonly detected in individuals with Sturge-Weber syndrome.²

CLINICAL FINDINGS AND TREATMENT OPTIONS

Fundusoscopic examination in patients with choroidal hemangioma can be problematic, as the tumor can be difficult to visualize due to its shallow elevation and color similar to background choroidal tissue.² Most hemangiomas remain fairly stable in size over time but occasionally develop nonneoplastic vascular congestion within the tumor that causes the

“Circumscribed choroidal hemangioma classically presents at an older age than diffuse hemangioma and with less severe SRF and cystoid macular edema.

tumor to increase in thickness or to leak fluid into the subretinal space or within the retina.² Ancillary testing with intravenous fluorescein angiography, indocyanine green angiography, ultrasonography, and optical coherence tomography (OCT) has been used to characterize the features of this tumor.²

Circumscribed choroidal hemangioma classically presents at an older age than diffuse hemangioma and with less severe SRF and cystoid macular edema (CME).² This small, benign tumor can lead to visual acuity loss from macular fluid or even ultimately eventuate with total serous retinal detachment and elevated risk for neovascular glaucoma, necessitating enucleation.³

Treatment options for circumscribed choroidal hemangioma include laser photocoagulation, transpupillary thermotherapy (TTT), photodynamic therapy (PDT), external beam radiotherapy, plaque radiotherapy, and proton beam radiotherapy. PDT is the treatment of choice because it has the fewest complications.² Herein, we describe a patient with circumscribed choroidal hemangioma that did not respond to PDT and that was ultimately controlled with low-dose iodine-125 (I-125) plaque brachytherapy.

CASE REPORT

A 45-year-old man noted fluctuating blurred vision in his left eye (OS) for 1 month along with a “black spot” for 2 days. On examination, visual acuity (VA) was 20/20



AT A GLANCE

- Choroidal hemangioma is a benign vascular tumor of the posterior segment.
- Circumscribed choroidal hemangioma is sporadic, unilateral, and commonly overlooked on fundusoscopic examination because the tumor color blends nearly imperceptibly with the normal choroid. It can cause photopsia, floaters, vision loss, and, in some cases, no perceptible symptom.
- The prevalence of this tumor is difficult to estimate because asymptomatic lesions can be difficult to detect.

in his right eye (OD) and 20/25 OS. Intraocular pressure (IOP) was 17 mm Hg in each eye (OU).

Dilated fundusoscopic examination was normal OD. There was an orange-colored choroidal mass temporal to the foveola OS, measuring 6 mm in diameter and 3.2 mm in thickness (Figure, A). The mass was echodense on ultrasonography and demonstrated expansion of the choroidal vascular tissue with shallow overlying SRF on OCT. The fovea was intact. On examination with fundus autofluorescence (FAF), there was a rim of hyperautofluorescence (Figure, B) correlating with the subretinal fluid seen on OCT (Figure, C).

These findings were consistent with circumscribed choroidal hemangioma, and observation was advised due to the patient's good VA. Over the next 18 months the patient's tumor and VA remained stable.

Two years later, although the tumor appearance remained stable, the amount of subfoveal fluid and intraretinal edema had increased (Figure, F), and the patient's VA had decreased to counting fingers (CF) at 2 feet OS. PDT was performed using standard parameters, and an additional intravitreal injection of bevacizumab (Avastin, Genentech) was given adjunctively. One month later, CME was mildly decreased OS, but VA remained poor at CF due to the SRF. Observation was recommended, to allow for a delayed effect of PDT.

At the next follow-up, the patient refused further PDT despite the continued presence of SRF. After a total of 8 months after the initial PDT, with SRF still persistent, the patient agreed to undergo I-125 plaque radiotherapy. This treatment was applied using a 10-mm radiation field with tumor apex dose of 30 Gy over 101 hours.

Two months after the radiotherapy was completed, the hemangioma had regressed to 2.8 mm in thickness (Figure, J) and OCT confirmed resolution of SRF and CME, leaving a flat retina (Figure, L). VA OS improved to 20/200.

DISCUSSION

Circumscribed choroidal hemangioma is a benign tumor that can cause photopsia, floaters, vision loss, and, in some cases, no perceptible symptom.² The prevalence of this tumor is difficult to estimate because symptomatic circumscribed choroidal hemangiomas are brought to medical attention, whereas asymptomatic lesions can be difficult to

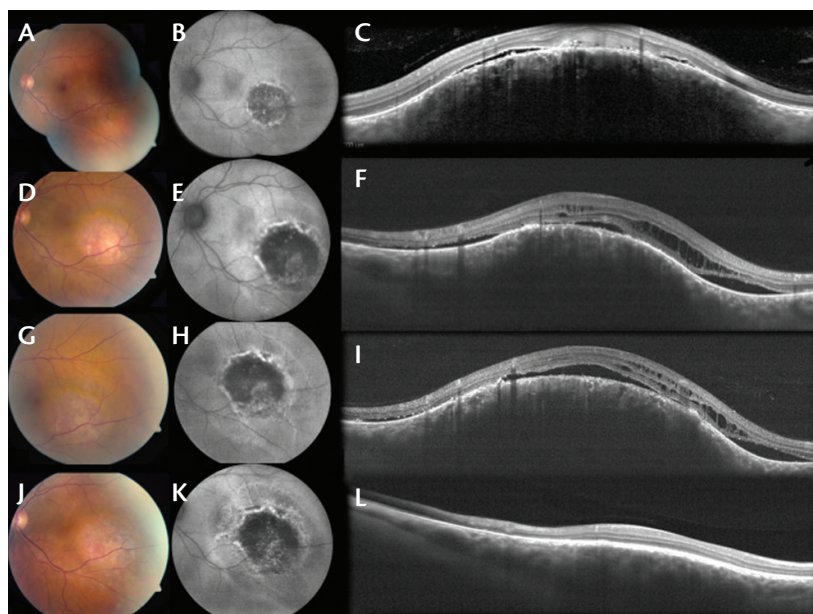


Figure. A 48-year old man with circumscribed choroidal hemangioma OS. At initial fundus examination, subtle circumscribed choroidal hemangioma was noted inferotemporal to the foveola (A), better visualized on FAF as hypoautofluorescent with surrounding hyperautofluorescence (B). OCT documented the elevated choroidal mass with overlying subretinal fluid and deposits (C). Two years later, the tumor's appearance (D) and FAF (E) were stable, but OCT documented increasing SRF and CME (F). After PDT, the mass appeared stable (G) and FAF unchanged (H), but OCT confirmed persistent SRF and CME (I). After plaque brachytherapy was administered, the mass regressed (J) and FAF was stable (K). Tumor response was seen on OCT, with complete resolution of SRF and CME leaving a flat retina with outer retinal thinning (L).

detect on funduscopy.³

The median age at onset of circumscribed choroidal hemangioma is between 45 and 50 years.² Over time, this vascular lesion can increase in size, leading to damage to the overlying retinal pigment epithelium and retina with resulting VA loss. Common goals for management of circumscribed choroidal hemangioma include preservation of VA and prevention of visual field loss, as well as prevention of total retinal detachment with secondary neovascular glaucoma.²

Symptoms related to circumscribed hemangioma include vision loss due to SRF, CME, tumor tilting of the foveola, and amblyopia. Management options include refraction with improvement of refractive error (usually hyperopia), treatment of possible amblyopia, and consideration of PDT.

PDT is the treatment of choice for circumscribed choroidal hemangioma, especially those with serous retinal detachment.⁴ PDT is primarily used to induce tumor fibrosis, with subsequent resolution of SRF and CME.⁵ Blasi et al followed 25 patients with circumscribed choroidal hemangioma who received standard PDT and reported that 76% showed improvement of VA by 2 lines on Snellen chart.⁶ All eyes treated with PDT

“ Treatment of circumscribed choroidal hemangioma is reserved for eyes with visual compromise. PDT is the treatment of choice.

demonstrated reduction in tumor size and complete resolution of macular SRF.⁶ Risks included occasional necessity for multiple treatments, a factor that could increase the risk of neurosensory retinal degeneration and choroidal atrophy.⁵

From the patient's perspective, PDT has a higher potential out-of-pocket cost compared with alternative treatments such as TTT and plaque radiotherapy.⁵ When PDT is ineffective, plaque brachytherapy is typically considered.

Plaque radiotherapy is most often used for large choroidal hemangioma, especially those with near total or total retinal detachment with SRF.⁷ Additionally, plaque radiotherapy can be used as a rescue treatment in the event of continuing tumor activity after failed PDT. The main concern regarding plaque radiotherapy relates to the ultimate complications of retinal ischemia and vision loss.

In a series of 15 eyes with circumscribed choroidal hemangioma managed with low-dose plaque brachytherapy with target doses of 30 Gy to 35 Gy, resolution of SRF was seen in all cases (100%) and VA was stable or improved in eight (53%).¹ The limited effectiveness regarding VA restoration was probably primarily a result of chronic, highly elevated retinal detachment, which in itself can lead to retinal ischemia and permanent visual acuity loss, retinal atrophy, RPE alterations, and minor radiation maculopathy.¹ In a later study including five patients with diffuse choroidal hemangioma with SRF managed with plaque radiotherapy, all patients showed complete regression of SRF and tumor over a mean follow-up of 32 months.⁸ However, only two patients demonstrated improved VA.

In addition to I-125, there are other isotopes for ophthalmic radiotherapy delivery, including ruthenium-106 (Ru-106)⁹ and palladium-103 (Pd-103).⁷ Lopez-Caballero et al evaluated eight eyes with circumscribed choroidal hemangioma treated with high dose I-125 plaque therapy with target dose of 48 Gy and noted reduction of tumor thickness from a mean of 4.4 mm to a mean of 1 mm, resolution of retinal detachment in all eyes, and stable VA in six eyes.¹⁰ Aizman et al reported on five eyes with circumscribed choroidal hemangioma treated with 29 Gy of Pd-103 plaque radiotherapy; all demonstrated tumor regression with resolution of SRF.⁷ As in reports by Shields et al¹ and Arepalli et al,⁸ post-treatment VA

in the Aizman et al paper varied depending on the chronicity of the macular detachment, with three patients showing VA improvement, one patient with stable vision, and one patient with decreased VA due to macular ischemia.

SUMMARY

Treatment of circumscribed choroidal hemangioma is reserved for eyes with visual compromise. PDT is the treatment of choice. However, if PDT fails to resolve SRF, then I-125 plaque radiotherapy is an option. In this case, the radiation induced tumor regression, prompt SRF resolution, and modest visual improvement from CF at 2 feet to 20/200. ■

1. Shields CL, Honavar SG, Shields JA, et al. Circumscribed choroidal hemangioma clinical manifestations and factors predictive of visual outcome in 200 consecutive cases. *Ophthalmology*. 2001;108(12):2237-2248.
2. Shields JA, Shields CL. Circumscribed choroidal hemangioma. In: *Intraocular Tumors: An Atlas and Textbook*. 3rd ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2016:247-263.
3. Shields JA, Stephens RF, Eagle RC Jr, et al. Progressive enlargement of a circumscribed choroidal hemangioma: a clinicopathologic correlation. *Arch Ophthalmol*. 1992;110(9):1276-1278.
4. Porri G, Giovannini A, Amato G, et al. Photodynamic therapy of circumscribed choroidal hemangioma. *Ophthalmology*. 2003;110(4):674-680.
5. Schmidt-Erfurth UM, Michels S, Kusserow C, et al. Photodynamic therapy for symptomatic choroidal hemangioma: visual and anatomic results. *Ophthalmology*. 2002;109(12):2284-2294.
6. Blasi MA, Tiberti AC, Scupola A, et al. Photodynamic therapy with verteporfin for symptomatic circumscribed choroidal hemangioma: five-year outcomes. *Ophthalmology*. 2010;117(8):1630-1637.
7. Aizman A, Finger P, Shabto U, et al. Palladium 103 (103Pd) plaque radiation therapy for circumscribed choroidal hemangioma with retinal detachment. *Arch Ophthalmol*. 2004;122(11):1652-1656.
8. Arepalli S, Shields C, Kaliki S, Emrich J, Komarnicky L, Shields JA. Diffuse choroidal hemangioma management with plaque radiotherapy in 5 cases. *Ophthalmology*. 2013;120(11):2358-2359.
9. Naseripour M, Maleki A, Astaraki A, et al. Ruthenium-106 brachytherapy in the treatment of circumscribed choroidal hemangioma [published online ahead of print March 23, 2017]. *Retina*.
10. López-Caballero C, Saornil MA, De Frutos J, et al. High-dose iodine-125 episcleral brachytherapy for circumscribed choroidal hemangioma. *Br J Ophthalmol*. 2010;94(4):470-473.

Alex Altman, BS

- medical student at Drexel University College of Medicine, Philadelphia, Pa.
- namtlaa@gmail.com

Kunal Malik, BA

- medical student at the Lewis Katz School of Medicine, Temple University, Philadelphia, Pa.
- kunalmalik50@gmail.com

Carol L. Shields, MD

- director of the Ocular Oncology Service, Wills Eye Hospital, Thomas Jefferson University, Philadelphia, Pa.
- member of the *Retina Today* editorial advisory board
- carolshields@gmail.com

No conflicting relationship exists for any author.

Support provided by the Eye Tumor Research Foundation, Philadelphia, Pa. (CLS). The funders had no role in the design and conduct of the study, in the collection, analysis, and interpretation of the data, or in the preparation, review, or approval of the manuscript. Carol L. Shields, MD, has had full access to all the data in the study and takes responsibility for the integrity of the data and the accuracy of the data analysis.