

Choroidal Hemangioma Masquerading as Central Serous Chorioretinopathy

BY YASMIN POUSTCHI, BA; SWATHI KALIKI, MD; AND CAROL L. SHIELDS, MD

Choroidal hemangioma is a relatively rare, benign vascular hamartoma. The circumscribed type is classically unilateral and is believed to be congenital in origin.^{1,2} Circumscribed choroidal hemangioma typically appears as a slightly oval or round, red-orange mass in the macular or perimacular region of the choroid, often with clinically indistinct borders.¹⁻³ Although choroidal hemangioma is benign, this tumor can manifest progressive retinal detachment with poor visual acuity; rarely does this condition necessitate enucleation. In a review of 200 cases of choroidal hemangioma by Shields et al,¹ 54% of patients were found to have a visual acuity of 20/200 or worse at initial presentation, and enucleation was necessary in 3 cases. Thus, timely recognition and appropriate management of this tumor is important for visual outcome.

Choroidal hemangioma can be misdiagnosed as amelanotic melanoma, choroidal metastasis, central serous chorioretinopathy (CSC), and, in some instances, it remains clinically undetected despite visual loss.¹ Herein, we report a case of choroidal hemangioma that was clinically undetected despite visual loss for more than 1 year and later treated as CSC until an underlying subtle hemangioma was detected.

CASE

A 46-year-old healthy white woman with no history of systemic cancer presented with a 1-year history of mild decreased visual acuity in the right eye (OD) to 20/30. The affected fundus displayed serous macular detachment. The referring ophthalmologist considered a diagnosis of CSC and administered 1 intravitreal bevacizumab (Avastin, Genentech) injection. Postinjection, there was neither improvement in vision nor reduction in subretinal fluid. The patient was referred to the Ocular Oncology Service at Wills Eye Institute.

On our initial examination, visual acuity was 20/30

Although choroidal hemangioma is benign, this tumor can manifest progressive retinal detachment with poor visual acuity.

OD and 20/25 in the left eye (OS). Anterior segment examination of both eyes was unremarkable. Fundus examination OS was unremarkable. Fundus examination OD revealed shallow subretinal fluid and a subtle orange-colored choroidal mass in the peripapillary area 4 x 3 mm in diameter (Figure 1A). Fundus autofluorescence revealed hypoautofluorescence of the tumor and hyperautofluorescence of overlying lipofuscin and subretinal fluid (Figure 1B). Fluorescein angiography (FA) displayed early lacy hyperfluorescence and diffuse, late hyperfluorescence (Figure 1C, 1D). Indocyanine green (ICG) angiography showed slight early tumor hypercyanescence and late washout isocyanescence (Figure 1E, 1F). A-scan ultrasonography OD revealed high internal reflectivity, and B-scan ultrasonography revealed a solid, acoustically dense, dome-shaped, 2.3 mm thick choroidal mass (Figure 1G). Enhanced depth imaging optical coherence tomography (OCT) revealed compression of choriocapillaris, thinning of retinal pigment epithelium, atrophy of photoreceptors, loss of inner segment-outer segment junction, and irregularity of external limiting membrane (Figure 1H), and confirmed the presence of perifoveal subretinal fluid (Figure 1I). The features were consistent with choroidal hemangioma. Photodynamic therapy (PDT) was performed using standard parameters. One year following PDT, the tumor appeared stable and subretinal fluid nearly resolved (Figure 2). Visual acuity was stable at 20/30, likely related to chronic outer retinal changes from previous fluid.

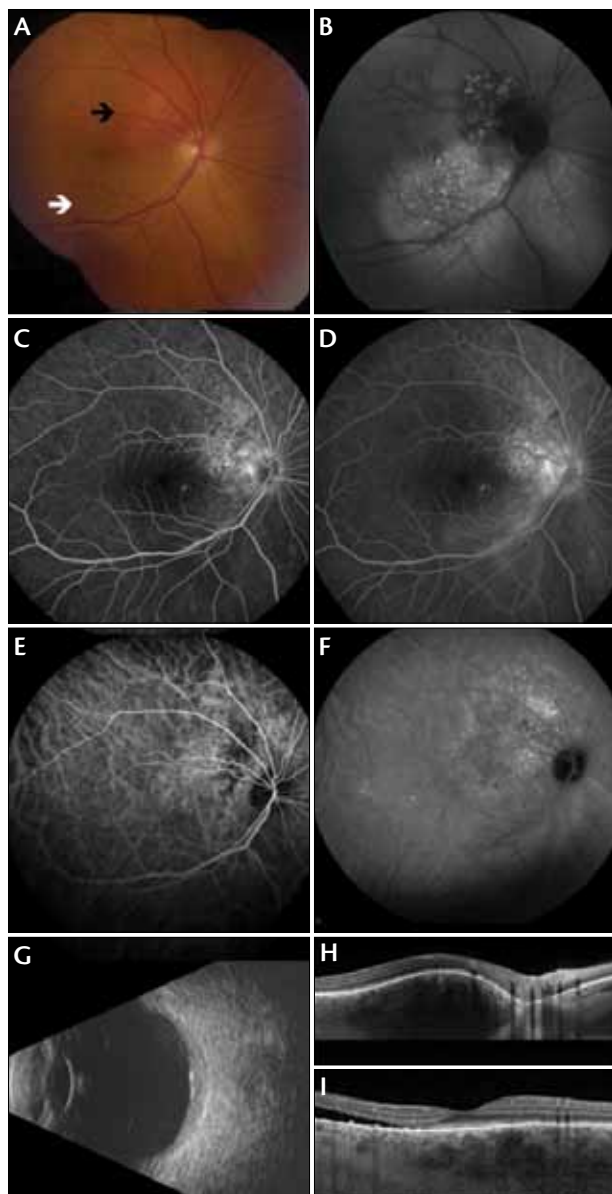


Figure 1. Choroidal hemangioma at presentation showing orange-colored choroidal hemangioma (black arrow) associated with subretinal fluid (white arrow) (A), with hypoautofluorescence of tumor and with hyperautofluorescence of overlying lipofuscin and subretinal fluid (B). Fluorescein angiography demonstrates mottled hyperfluorescence of mass at 28 seconds (C) and diffuse late staining at 17 minutes (D). Indocyanine green angiography shows early tumor hypercyanescence at 83 seconds (E) and late “washout” isocyanescence at 26 minutes (F). B-scan ultrasonography reveals acoustic solidity of choroidal hemangioma (G) and optical coherence tomography (OCT) demonstrates elevated choroidal mass with compression of overlying choriocapillaris (H) and shallow subfoveal fluid (I).

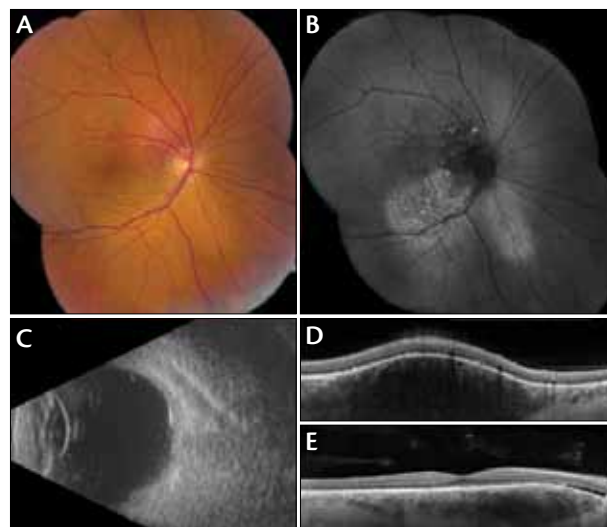


Figure 2. Choroidal hemangioma after photodynamic therapy. Fundus photograph of choroidal hemangioma with minimal residual subretinal fluid (A) confirmed on auto-fluorescence (B). B-scan ultrasonography reveals decreased thickness of choroidal hemangioma (C), and OCT demonstrates minimal residual subretinal fluid (D, E).

DISCUSSION

Choroidal hemangioma is a benign vascular tumor that can cause permanent loss of vision due to progressive leakage from intrinsic tumor vessels, leading to serous retinal detachment and cystoid macular edema.⁴ Prompt diagnosis of symptomatic choroidal hemangioma is achieved by ophthalmoscopy with recognition of classic clinical features and diagnostic testing with ultrasonography, OCT, FA, and ICG angiography. Misdiagnosis of choroidal hemangioma can be problematic, as this tumor can remain hidden within the similar-hued choroid, often camouflaged by shallow serous retinal detachment. In a review by Shields and associates¹ of 200 consecutive patients with choroidal hemangioma, temporary misdiagnosis prior to referral was noted in 71% of cases and included choroidal melanoma (29%), choroidal metastasis (9%), retinal detachment (6%), CSC (5%), macular edema (3%), choroidal nevus (2%), choroidal granuloma (1%), vasoproliferative retinal tumor (1%), optic neuritis (1%), retinoblastoma (1%), choroidal osteoma (1%), age-related macular degeneration (1%), high hypermetropia (1%), and no diagnosis (14%).

Choroidal hemangioma manifests as an orange-red colored lesion, similar to the color of the surrounding choroid, but hemangioma can compress the surrounding choroid imparting a slight brown ring around its margin. Amelanotic melanoma displays a more yellow-tan color, often with features of drusen and retinal pigment epithelial (RPE) alterations on the surface.^{1,2} However, the differentia-

tion is occasionally not so obvious. For example, overlying chronic fibrous metaplasia of the RPE over long-standing hemangioma can obscure the underlying orange-red tumor, leading to mistaken diagnosis of choroidal melanoma, metastasis, or other condition.⁵ Ultrasonography is an important tool in differentiation as choroidal hemangioma displays high internal reflectivity on A-scan and acoustic solidity on B-scan, whereas choroidal melanoma shows medium to low internal reflectivity and acoustic hollowness.^{1,2} To further confuse the issue, certain choroidal metastases can closely simulate hemangioma, such as those that originate from primary renal cell carcinoma, carcinoid tumor, and thyroid carcinoma, as these can also impart an orange hue.²

In this case, choroidal hemangioma was misdiagnosed as CSC. CSC leads to similar neurosensory retinal detachment classically in the macular region due to hyperpermeability of the choroid. Both CSC and choroidal hemangioma typically present in middle-aged adults and both manifest features of serous macular detachment.^{6,7} The difficulty in differentiation of the conditions stems from their clinical similarities and the fact that choroidal hemangioma can remain relatively subtle or hidden under the fluid.⁷ The most important testing utilized to differentiate choroidal hemangioma from CSC includes ultrasonography to demonstrate the underlying mass or OCT to show the choroidal mass and overlying subretinal fluid. Enhanced depth imaging OCT allows further description of the hemangioma, but it should be realized that CSC also leads to choroidal thickening that is generally diffuse and of approximately 500 μm ,⁶ whereas the thickening with hemangioma is sloped, discrete, and more than 1000-2000 μm .

Other useful diagnostic tests, such as FA and ICG angiography, help differentiate these conditions. FA of choroidal hemangioma shows linear, lacy hyperfluorescence within the mass in the prearterial or early arterial phase and progressive hyperfluorescence and staining in the venous and recirculation phases.^{7,8} CSC shows a different picture with focal leaks in the RPE that appear almost pinpoint and evolve into leakage in a smokestack, ink blot, or mushroom pattern.⁶ Additionally, other sites of RPE damage are noted. ICG angiography of choroidal hemangioma shows reticular hyperfluorescence of the mass in the early phases that peaks at approximately 1 minute and later “washes out” with iso- or hypofluorescence surrounded by annular staining.^{7,8} ICG angiography of CSC shows multifocal choroidal points of hyperpermeability with intermittent hypofluorescent areas suggestive of focal choroidal vascular compromise.⁶

Treatment for choroidal hemangioma is focused on improvement of visual acuity by inducing resolution of

retinal detachment or reduction of hyperopia-related symptoms from the tumor. Current preferred treatments include photodynamic therapy for smaller tumors and plaque radiotherapy for more extensive tumors or those with extensive retinal detachment.^{2,5} Photodynamic therapy (PDT) is currently regarded as the treatment of choice for choroidal hemangioma. In 1 report, visual acuity improved from 20/200 to 20/25 following PDT.⁹ In a study by Blasi and associates of 25 cases of choroidal hemangioma treated with PDT,⁴ there was complete resolution of fluorescein leakage and decreased tumor thickness in 100% of the eyes. No complications were observed in the 5-year follow-up period, and visual acuity improved within the first 3 months and remained stable at the 5-year follow-up.⁴ We anticipate that visual acuity will remain at 20/30 in our patient as OCT and autofluorescence studies showed minor RPE abnormalities in the foveal region. ■

Yasmin Poustchi, BA, is a medical student at Robert Wood Johnson Medical School in New Brunswick, NJ.

Swathi Kaliki MD, is an ocular oncology fellow at Wills Eye Institute in Philadelphia.

Carol L. Shields, MD, is the Co-Director of the Ocular Oncology Service, Wills Eye Institute, Thomas Jefferson University. She is a Retina Today Editorial Board member. Dr. Shields can be reached at +1 215 928 3105; fax: +1 215 928 1140; or via email at carol.shields@shieldsoncology.com.

Support provided by 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, and 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.

No conflicting relationship exists for any author.



1. Shields CL, Honavar SG, Shields JA, Cater J, Demirci H. 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: Shields JA, Shields CL. *Intraocular Tumors: An Atlas and Textbook*, 2nd ed. Philadelphia, PA: Lippincott Williams and Wilkins, 2008:230-245.
3. Boixadera A, Garcia-Arumi J, Martinez-Castillo V, et al. Prospective clinical trial evaluating the efficacy of photodynamic therapy for symptomatic circumscribed choroidal hemangioma. *Ophthalmology*. 2009;116(1):100-105.
4. 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.
5. Tsipursky MS, Golchet PR, Jampol LM. Photodynamic therapy of choroidal hemangioma in Sturge-Weber syndrome, with a review of treatments for diffuse and circumscribed choroidal hemangiomas. *Surv Ophthalmol*. 2011;56(1):68-85.
6. Ross A, Ross AH, Mohamed Q. Review and update of central serous chorioretinopathy. *Curr Opin Ophthalmol*. 2011;22(3):166-173.
7. Mashayekhi A, Shields CL. Circumscribed choroidal hemangioma. *Curr Opin Ophthalmol*. 2003;14:142-149.
8. Arevalo JF, Shields CL, Shields JA, Hykin PG, De Potter P. Circumscribed choroidal hemangioma: characteristic features with indocyanine green videoangiography. *Ophthalmology*. 2000;107(2):344-350.
9. Shields CL, Materin MA, Marr BP, Mashayekhi A, Shields JA. Resolution of advanced cystoid macular edema following photodynamic therapy of choroidal hemangioma. *Ophthalmic Surg Lasers Imaging*. 2005;36:237-239.