

The Current Management of Choroidal Hemangioma

BY APARNA RAMASUBRAMANIAN, MD; AND CAROL L. SHIELDS, MD

Choroidal hemangioma is a benign vascular tumor of the choroid and manifests in two subtypes: circumscribed and diffuse. The diffuse choroidal hemangioma occurs frequently in association with Sturge-Weber syndrome and the circumscribed hemangioma has no systemic associations. Timely diagnosis and treatment is critical as these benign tumors predominantly affect the posterior choroid, causing significant visual disturbance. In this review, we discuss the current management of circumscribed and diffuse choroidal hemangioma.

DIAGNOSIS OF CHOROIDAL HEMANGIOMA

The diagnosis of choroidal hemangioma is a combination of clinical features and ancillary tests. Clinically, circumscribed hemangioma appears as a subtle red-orange mass in the posterior choroid (Figure 1A). Diffuse choroidal hemangioma appears as an extensive red-orange thickening of the posterior choroid (Figure 2A).^{1,2} Ancillary testing for choroidal hemangioma includes the diagnostics below.

Ultrasonography. Choroidal hemangioma shows high internal reflectivity (A-scan) and acoustic solidity (B-scan). Diffuse hemangioma shows diffuse marked thickening of choroid whereas circumscribed lesions appear as a placoid or oval mass.¹

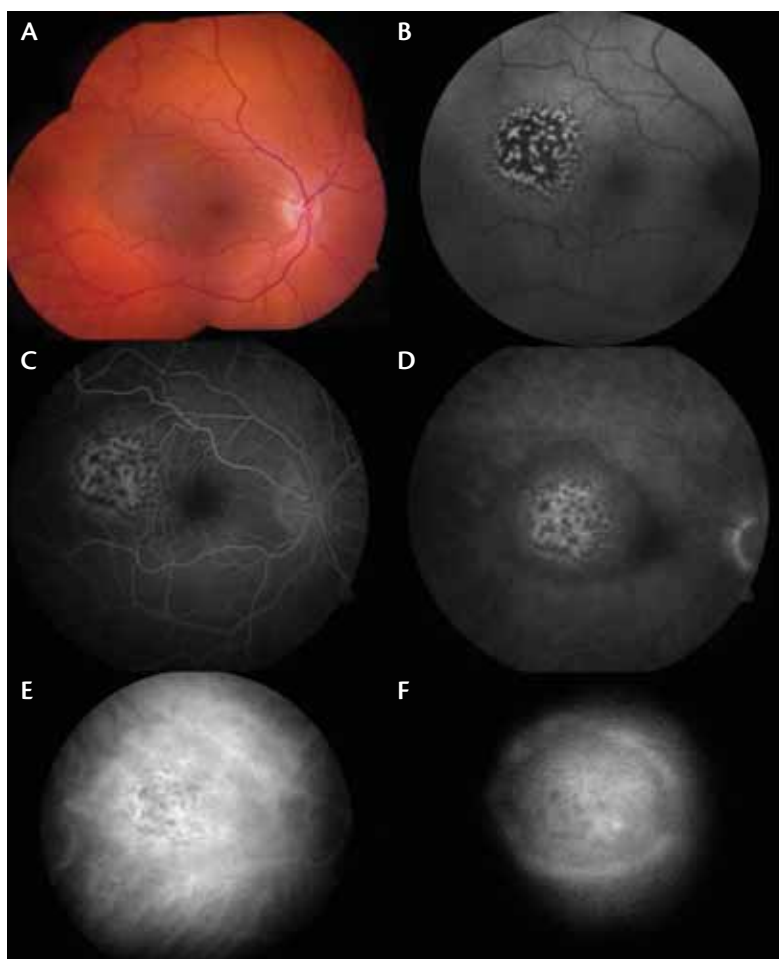


Figure 1. Fundus appearance of circumscribed choroidal hemangioma showing the characteristic orange-red lesion (A). Fundus autofluorescence showing the hypoautofluorescent lesion and hyperautofluorescent overlying orange pigment (B). Fluorescein angiography showing early hyperfluorescence (C) and late diffuse leakage (D). Indocyanine angiography showing early hyperfluorescence (E) and late washout of dye (F).

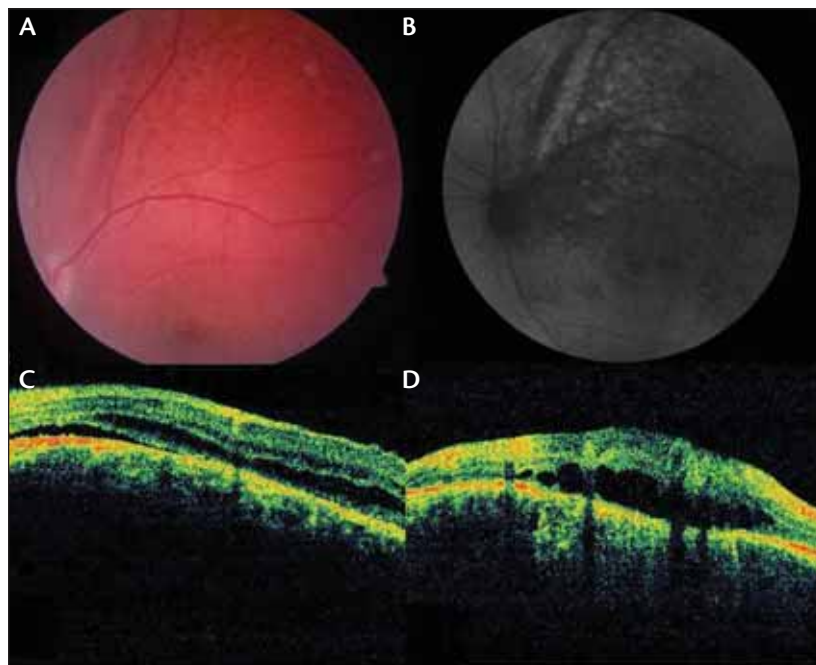


Figure 2. Fundus appearance of diffuse choroidal hemangioma (A). Fundus autofluorescence showing hypoautofluorescence tumor and hyperautofluorescent overlying orange pigment (B). Optical coherence tomography showing subretinal fluid (C) and intraretinal edema (C,D).

Fluorescein angiography. In early arterial phase, hyperfluorescence of the mass is evident (Figure 1C) with late diffuse hyperfluorescence (Figure 1D). Diffuse hemangioma shows diffuse hyperfluorescence in the pre-arterial stage. Often related subretinal fluid is visualized with hyperfluorescence.

Indocyanine green angiography. This test shows early hyperfluorescence by 1 minute (Figure 1E) with late dye washout at 20 minutes, appearing hypofluorescent relative to the surrounding choroid (Figure 1F). The late washout seen in circumscribed hemangioma is not classically visualized in diffuse lesions.

Autofluorescence. Choroidal hemangioma shows little intrinsic autofluorescence. Overlying lipofuscin and fresh subretinal fluid show hyperautofluorescence and RPE hyperplasia and atrophy show hypoautofluorescence (Figure 1B and 2B).³

TREATMENT OF CIRCUMSCRIBED CHOROIDAL HEMANGIOMA

The decision to treat circumscribed hemangiomas is based on the location, size and related ocular symptoms.⁴ Shields and coworkers⁴ reported 200 patients with circumscribed choroidal hemangioma and found that the most common cause for decreased vision was chronic subretinal fluid and chronic macular edema. Optical

coherence tomography can be useful in detecting subtle subretinal fluid (Figure 2C) and retinal edema (Figure 2D). Asymptomatic hemangiomas that demonstrate no related subretinal fluid are managed by observation. Hemangiomas with advanced visual deficit and minimal anticipated visual potential can also be observed but it should be understood that progressive subretinal fluid could lead to neovascular glaucoma and ultimate need for enucleation.⁴ The available treatment modalities are detailed below.

Laser photocoagulation (Xenon or Argon). Laser photocoagulation has been an effective treatment modality for hemangioma for many years. Shields and coworkers⁴ reported 62% resolution of subretinal fluid and 71% stability of vision with argon laser photocoagulation. The main complication of laser photocoagulation is the expansion of RPE atrophy and coexistent scotoma. Other

reported complications include preretinal membrane, choroidal neovascular membrane, vascular occlusion and retinal bleeding. Diode laser photocoagulation has been shown to be equally efficacious with probably lower absorption by the retinal pigment epithelium.⁵ Currently, laser photocoagulation is rarely used to treat hemangiomas as this has been largely replaced by photodynamic therapy.

Transpupillary thermotherapy (TTT). TTT utilizes 810 nm infrared light with a large spot size and long exposure time leading to increased temperature and irreversible cytotoxic effect. The use of TTT is limited to extrafoveal tumors. Treatment with TTT successfully causes tumor regression in many patients (42%, partial 50%) complete but carries a risk of cystoid macular edema, preretinal fibrosis, focal iris atrophy and retinal vascular occlusion.⁶

Photodynamic therapy (PDT). PDT involves administration of photosensitizer drug that reaches the target tissue that is irradiated with light of wavelength coinciding with the absorption maximum of the photosensitizer. Cellular injury from PDT is mediated by singlet oxygen. The main advantage of PDT is the selectivity of the treatment and minimal disruption of tissues. In various studies the visual acuity improvement or stabilization after PDT for choroidal hemangioma ranges from 73% to

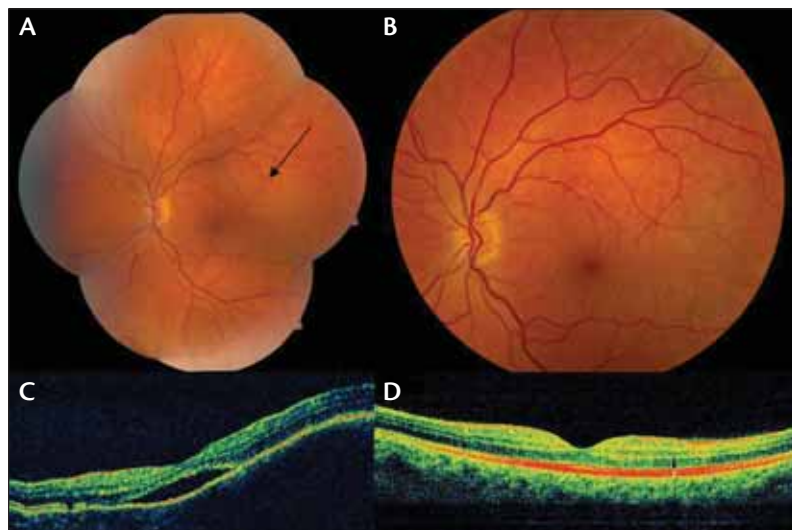


Figure 3. Fundus appearance of a faint choroidal hemangioma (arrow) (A) with overlying subretinal fluid visualized on OCT (C). The lesion showed good response to PDT (B) and the subretinal fluid is completely resolved with good foveal contour on OCT (D).

100%.⁷ Blasi and co-workers reported the five year outcome of 25 patients treated with PDT for circumscribed hemangioma and found that visual acuity improved by two lines in 76% of patients with complete resolution of macular exudation in all cases and no complications were observed.⁸ In our experience with nearly 50 patients treated with PDT, 95% of patients required only one session with complete resolution of the tumor and fluid. A second session was needed in 5% to resolve persistent or recurrent subretinal fluid. Long-term recurrence of subretinal fluid is uncommon (Figure 3).^{9,10}

Antivascular endothelial growth factor (anti-VEGF) agents. Anti-VEGF agents are known to reduce vascular permeability and hasten resolution of subretinal fluid and intraretinal edema in a multitude of ophthalmic pathologies. Sagong and coworkers¹¹ reported beneficial effect of bevacizumab (Avastin, Genentech) for three patients with circumscribed hemangioma. One patient was treated with bevacizumab alone for recurrence following laser photocoagulation and two patients were treated with bevacizumab and PDT as primary treatment. All patients showed improvement in visual acuity with resolution of subretinal fluid and edema. At mean follow up of 8 months, none of the patients showed any evidence of recurrence or adverse effects.¹¹ The role of VEGF agents in treatment of choroidal hemangioma is still uncertain and more reports documenting the benefit would be required.

Proton beam radiation. Proton beam radiation involves delivery of a precise dose of radiation to a target

tissue. Protons unlike other rays deposit high energy when they slow down reducing the scattering effect on surrounding tissue. In a retrospective review of 71 patients with choroidal hemangioma treated with proton beam radiation, 52% showed improvement in visual acuity, 100% showed resolution of subretinal fluid although cataract developed in 28% and radiation maculopathy developed in 8%.¹²

Plaque radiotherapy. Plaque radiotherapy (brachytherapy) has been employed in multiple ocular disorders most common being choroidal melanoma. Aizman and coworkers¹³ reported five patients treated with palladium 103 plaque for circumscribed hemangioma. All patients showed complete resolution of subretinal fluid with the tumor height decreasing by a mean of 50%. López-Caballero and coworkers¹⁴

reported use of Iodine 125 plaque in the treatment of large circumscribed hemangioma with retinal detachment. Tumor regression and resolution of subretinal fluid was noted in all patients with signs of radiation retinopathy in 38%. We reserve plaque radiotherapy for choroidal hemangioma with extensive subretinal fluid where PDT would not be advised, but plaque radiotherapy could be performed. Low-dose treatment is sufficient using 20 Gy apex dose. We have had experience with a patient who manifested iris neovascularization from extensive subretinal fluid from a small choroidal hemangioma that showed total response with plaque radiotherapy. The NVI resolved and the total detachment settled.¹⁵

External beam radiation (EBRT). EBRT has predominantly been used for diffuse choroidal hemangioma with a dose range of 20 Gy to 25 Gy in some cases and 35 Gy to 40 Gy in others.¹⁶ More precise radiation in a single session has been performed for circumscribed hemangioma with gamma knife radiosurgery. Kong and coworkers reported three patients treated with a maximal dose of 10 Gy with good anatomical and functional outcome with no side effects noted in the follow up period of 18 to 36 months.¹⁷

TREATMENT OF DIFFUSE CHOROIDAL HEMANGIOMA

The management of diffuse choroidal hemangioma can be challenging. Diffuse hemangioma can be asymptomatic but visual loss can be secondary to hyperopic amblyopia, foveal distortion and secondary retinal

Though benign, choroidal hemangioma can cause visual impairment from subretinal fluid, refractive error, intraretinal edema, and amblyopia.

detachment.¹ In addition to choroidal hemangioma, patients with Sturge-Weber syndrome also have congenital glaucoma in 70% of patients. The mechanism of raised intraocular pressure is angle anomaly and raised episcleral pressure.¹⁸ Treatment options for diffuse choroidal hemangioma include observation, amblyopic therapy, laser photocoagulation, irradiation, photodynamic therapy, retinal detachment surgery or even enucleation in advanced cases with neovascular glaucoma.¹

Photodynamic therapy (PDT). Multispot photodynamic therapy has been used successfully in patients with diffuse hemangioma. Reported cases in the literature document resolution of subretinal fluid, decrease in thickness of the tumor and improvement in visual acuity.^{19,20}

External beam radiation (EBRT). EBRT is effective in decreasing tumor thickness and resolving subretinal fluid. Our preference is to treat these patients with 20 Gy (low dose) or 40 Gy (standard dose) EBRT to the posterior segment of the eye and our results have been successful in most cases with subretinal fluid resolution and tumor involution. Recurrence of subretinal fluid following radiotherapy is rare. Schilling and coworkers²¹ reported 15 patients with diffuse hemangioma treated with low dose radiation (20 Gy). All patients showed resolution of subretinal fluid but the poor functional outcome was attributable to secondary glaucoma. Isolated case reports have also reported beneficial effect with gamma surgery, brachytherapy and proton beam radiation.

Management of glaucoma. Medical therapy is ineffective in most cases. Surgical treatment options include trabeculectomy, trabeculectomy and implant devices.

SUMMARY

In summary, choroidal hemangioma has a typical clinical appearance. The diagnosis can be aided by ancillary tests like fluorescein angiography and ICG angiography. Though benign, this tumor can cause visual impairment from subretinal fluid, refractive error, intraretinal edema, and amblyopia. Treatment options are several and have to be altered to suit the individual tumor clinical characteristics. Diffuse choroidal hemangiomas can be associated with systemic Sturge-Weber syndrome and have a poorer long-term visual prognosis due to coexistent amblyopia and glaucoma. Photodynamic therapy is the

most promising treatment for circumscribed choroidal hemangioma and selected diffuse hemangiomas as it causes minimal damage to surrounding tissues. ■

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Aparna Ramasubramanian, MD, is fellow at the Ocular Oncology Service at Wills Eye Hospital, Thomas Jefferson University. She can be reached via e-mail at aparna.r.krishna@gmail.com.

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



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