SURVEY OF OPHTHALMOLOGY VOLUME 56 • NUMBER 1 • JANUARY–FEBRUARY 2011

ELSEVIER

THERAPEUTIC REVIEWS

JOEL MINDEL, EDITOR

Photodynamic Therapy of Choroidal Hemangioma in Sturge-Weber Syndrome, with a Review of Treatments for Diffuse and Circumscribed Choroidal Hemangiomas

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Abstract. We report three new cases of patients with Sturge-Weber Syndrome and symptomatic retinal detachments from diffuse choroidal hemangiomas successfully treated with photodynamic therapy (PDT) and review medical literature on the available treatment options for choroidal hemangiomas. All patients were treated with a single session of PDT with verteporfin infused at a concentration of 6 mg/m^2 and treated for 83 seconds with 689-nm Zeiss laser that was delivered with total energy level of 50 J/cm^2 with an intensity of 600 mW/cm^2. The exudative retinal detachment (RD) and macular edema completely resolved in all cases by 1–4 months after PDT treatment. Visual acuity improved in all three cases with diminished tumor size in the areas of treatment. One case was followed for 5 months, another for 2 years, and the third case for 6 years, with no recurrence of exudative RD. PDT is an effective treatment option for visual deterioration from exudative retinal detachment in patients with diffuse choroidal hemangiomas. (**Surv Ophthalmol 56**:68–85, 2011. © 2011 Elsevier Inc. All rights reserved.)

Key words. age-related macular degeneration • circumscribed and diffuse choroidal hemangioma • cystoid macular edema • laser photocoagulation • PDT • photodynamic therapy • preretinal fibrosis • proton beam radiotherapy • radiation therapy • stereotactic radiotherapy • transpupillary thermotherapy • verteporfin

Choroidal hemangioma is a benign vascular tumor that can occur as a circumscribed mass, generally without systemic associations, or as a diffuse variant that is often associated with facial nevus flammeus or variations of Sturge-Weber syndrome (SWS).[15,51,52] Circumscribed choroidal hemangioma is almost always unilateral. Its pathogenesis is unknown. The tumor is often overlooked on routine eye examination or it is misdiagnosed as a malignant lesion such as choroidal melanoma or metastasis.[53] These lesions have been treated with photocoagulation,[3,26,28,47] external beam therapy,[28,38,41,47] stereotactic radiotherapy,[23,24,40] radiation therapy,[17,40] proton beam radiotherapy,[18,27,68] plaque radiotherapy,[8,28,66,68] transpupillary thermotherapy,[17] and more recently photodynamic therapy with verteporfin (Visudyne; Novartis Ophthalmics, Basel, Switzerland).[54]

There are notable differences between circumscribed hemangiomas and SWS–associated diffuse choroidal hemangiomas. In SWS, a nonhereditary

0039-6257/$ - see front matter
doi:10.1016/j.survophthal.2010.08.002

phakomatosis, hemangiomas predominately manifest as diffuse bodies in the leptomeninges and facial skin as well as the choroid. These hemangiomas are usually unilateral and ipsilateral to the angiomatous malformation of the skin. The diffuse choroidal hemangiomas of SWS may have localized areas of excessive thickening simulating circumscribed choroidal hemangiomas.[45] These patients are most likely to develop secondary retinal detachment with shifting of the subretinal fluid, either spontaneously or after filtering operations for glaucoma.[15] Diffuse choroidal hemangiomas have been treated with radiotherapy,[17,40] proton beam,[16,28,38,40,44,68] stereotactic radiotherapy,[24] plaque radiatiotherapy,[31,66] anti-vascular endothelial growth factor (VEGF),[67] and photodynamic therapy.[2,5,18,53]

Our Experience with Treatment of Diffuse Choroidal Hemangioma with Photodynamic Therapy

All three of our cases received the same standard protocol for treatment: verteporfin (Visudyne; Novartis AG, Basel, Switzerland), was infused at a concentration of 6 mg/m^2 over 10 minutes. Fifteen (15) minutes after start of the infusion the lesion was treated for 83 seconds with a Zeiss laser wavelength beam of 689 nm (Carl Zeiss Meditec Inc, Dublin, California, USA) that was delivered with total energy density of 50 J/cm^2 and intensity of 600 mW/cm^2.

CASE 1

This case was previously reported in 2004, and we now include the 5-year follow-up.[5] We described a 10-year-old white girl with a history of SWS and port-wine stain involving the right upper lid who was diagnosed with anisometropic amblyopia involving the right eye at age 2.5 years. The patient underwent treatment for right eye amblyopia with a stabilized visual acuity (VA) of 20/40. One month prior to the presentation, VA decreased to 20/400 (pinhole 20/100) with a hyperopic shift from +4.00 to +8.50. Ophthalmoscopic evaluation revealed a tomato catsup red appearance with nodular, more elevated area centered along the superotemporal arcade and an exudative retinal detachment (RD) involving the macula (Fig. 1). The patient underwent photodynamic therapy (PDT). Two partially overlapping, maximally large spots (6400 μm) were centered upon the elevated area of the diffuse choroidal hemangioma under the superotemporal arcade. Two weeks after treatment, the treated area of the tumor appeared purple (Fig. 2A,B), apparently due to thrombosis of the lesion. There was a mild decrease in the amount of subretinal fluid. A dramatic improvement was noted 10 weeks after treatment with a visible decrease in both the thickness of the choroidal tumor in the treated area, as well as the subretinal fluid. Four months after PDT, the subretinal fluid was completely resorbed and residual mottling of the overlying retinal pigment epithelium was noted in the area of treatment. Best-corrected visual acuity had improved to baseline of 20/40. At 5-year follow-up, the patient has retained the same visual acuity with no recurrence of the exudative detachment and no change in the noted pigmentary changes (Fig. 2C,D).

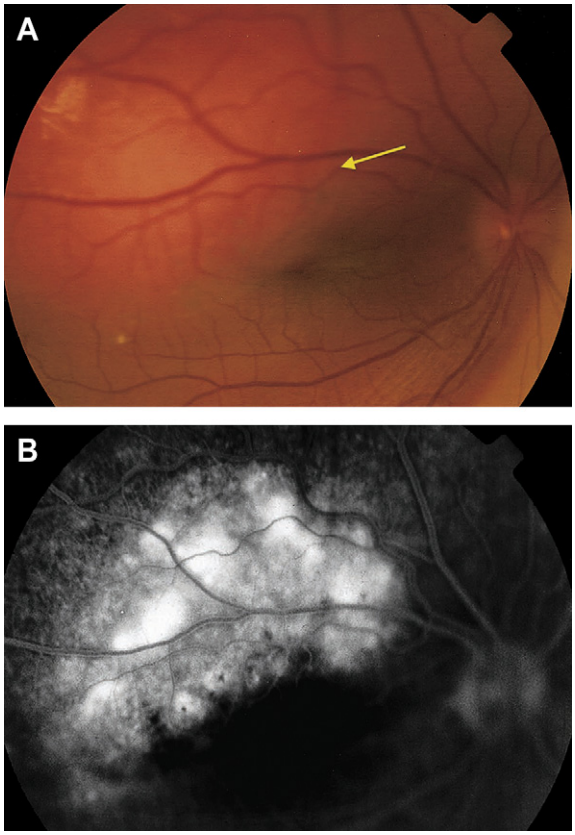

Fig. 1. Case 1. *A:* Color fundus photograph of the diffuse choroidal hemangioma with serous retinal detachment. *B:* Fluorescein angiogram showing retinal pigment epithelium degeneration and leakage. (Figure reprinted from Bains et al[5] with permission of *Retina*.)

CASE 2

A 20-year-old woman with known SWS and a diffuse choroidal hemangioma in the left eye complained of gradual reduction in vision with occasional photopsia. Her past ocular history was significant for successful trabeculectomy in the left