European Journal of Ophthalmology / Vol. 19 no. 1, 2009 / pp. 159-162

Polypoidal choroidal vasculopathy following photodynamic therapy for choroidal hemangioma

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PURPOSE. *To report a case of circumscribed choroidal hemangioma (CCH) that responded to photodynamic therapy (PDT) but 3 years later developed polypoidal choroidal vasculopathy (PCV) with exudative retinopathy.*

METHODS. *Case report.*

RESULTS. *A 59-year-old woman with a juxtapapillary CCH in her left eye was treated with a single 83-second, 7.5 mm PDT laser spot at 689 nm (50 J/cm^2) 15 minutes after the injection of intravenous verteporfin (6 mg/m^2). Three years later, the patient presented with photopsia in her left eye. Fundus examination of the left eye showed CCH regressed completely to a flat atrophic scar. There was diffuse macular edema and exudative retinopathy along the inferotemporal vascular arcade. On indocyanine green angiography, there were hyperfluorescent dilated choroidal vessels inferior to the foveola with late staining and leakage consistent with PCV. Hypofluorescence superior and nasal to the optic disc at the site of the treated hemangioma, consistent with choroidal ischemia, was observed. She was treated with 1.25 mg (0.05 cc) intravitreal bevacizumab. After 21 months of follow-up, the exudative retinopathy and macular edema completely regressed.*

CONCLUSIONS. *PDT is an effective treatment for CCH. Side effects of PDT for CCH are rare but include PCV. (Eur J Ophthalmol 2009; 19: 159-62)*

KEY WORDS. *Choroid, Eye, Hemangioma, Photodynamic therapy, Polypoidal choroidal vasculopathy*

Accepted: August 14, 2008

INTRODUCTION

Circumscribed choroidal hemangioma (CCH) is a benign vascular tumor of the choroid. Treatment is generally reserved for symptomatic patients with decreased visual acuity or metamorphopsia from cystoid macular edema (CME) or exudative foveal detachment (1). The therapeutic options include argon laser photocoagulation, transpupillary thermotherapy, episcleral radioactive plaque therapy, and recently, photodynamic therapy (2).

Photodynamic therapy (PDT) has become the treatment of choice for CCH (2). Michels and coauthors (2) reviewed 15 patients with CCH who were treated with PDT. They found complete tumor regression with no recurrence of tumor or subretinal fluid in all patients after a mean of 37 months. The only side effect was residual relative scotoma corresponding to the area of PDT in 12 patients with extrafoveal peripapillary CCH. They reported that none of the three treated subfoveal choroidal hemangiomas showed residual central visual field defects or choroidal atrophy. Other side effects following PDT have been few and include transient choroidal effusion, perifoveal retinal hemorrhage, retinal neovascularization, and transient visual disturbances (3-5). There have been no previous reports of polypoidal choroidal vasculopathy (PCV) following PDT of CCH. We report a case of CCH that responded to PDT but 3 years later developed PCV.

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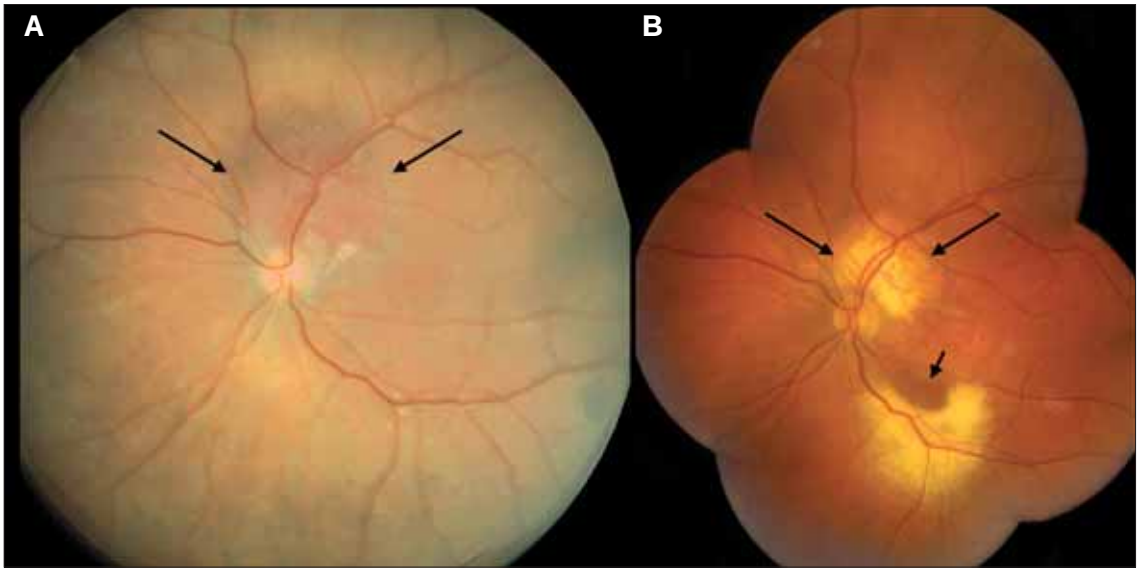

Fig. 1 - (A) *Fundus photograph at presentation showing juxtapapillary circumscribed choroidal hemangioma superior to the optic disc (arrows).* **(B)** *Three years after photodynamic therapy, the tumor was regressed, leaving an atrophic scar (long arrows), but exudative retinopathy inferior to the fovea was detected (small arrow).*

Case report

A 59-year-old white woman presented with decreased visual acuity in her left eye. She was diagnosed with a juxtapapillary CCH located superior to the optic disc in the left eye based on clinic examination and indocyanine green angiography (ICGA). Eleven years prior to referral, she was treated with marginal delimiting argon laser photocoagulation.

On our examination, her visual acuity was 20/20 in the right eye and counting fingers in the left eye. Fundus examination of the left eye revealed a juxtapapillary CCH located superior to the optic disc, measuring 5 x 5 x 2.7 mm (Fig. 1A). The mass was acoustically solid on B-scan ultrasonography and showed high internal reflectivity on A-scan. CME and subfoveal photoreceptor loss was noted on optical coherence tomography (OCT). After discussion of the guarded prognosis for visual recovery, the patient was treated with PDT. A single 83-second, 7.5 mm PDT laser spot at 689 nm (50 J/cm^2), covering the entire lesion, was applied 15 minutes after the injection of intravenous verteporfin (6 mg/m^2). The patient was followed by her local ophthalmologist and the subretinal fluid resolved.

Three years later, she returned to us with photopsia in her left eye. On examination, visual acuity was 20/20 in the right eye and counting fingers in the left eye. Fundus examination of the left eye showed complete regression of the choroidal hemangioma to a flat atrophic scar. There was diffuse macular edema and exudative retinopathy along the inferotemporal vascular arcade (Fig. 1B). Fluorescein angiography showed progressively increasing macular hyperfluorescence consistent with edema as well as dilated choroidal vessels inferior to the fovea. ICGA revealed hypofluorescence superior and nasal to the optic disc at the site of the treated hemangioma, consistent with choroidal ischemia. There were hyperfluorescent dilated choroidal vessels inferior to the foveola with late staining and leakage consistent with PCV (Fig. 2, A and B). Treatment with 1.25 mg (0.05 cc) intravitreal bevacizumab (Avastin; Genentech, Inc., South San Francisco, CA) was performed. Six weeks later, the visual acuity and fundus findings were stable and OCT showed less foveal edema. The exudative retinopathy and macular edema slowly improved to complete regression by 21 months with no change in visual acuity.

DISCUSSION

Adverse effects following PDT for CCH have been rarely reported. Leys and coworkers (3) observed retinal neovascularization in three untreated eyes with CCH. After PDT, the retinal neovascularization worsened in all three eyes. They hypothesized that the triggers for retinal neovascularization in eyes with CCH include subtle inflammation, chronic retinal detachment with ischemia, and release of angiogenic factors from PDT-induced necrosis of CCH.

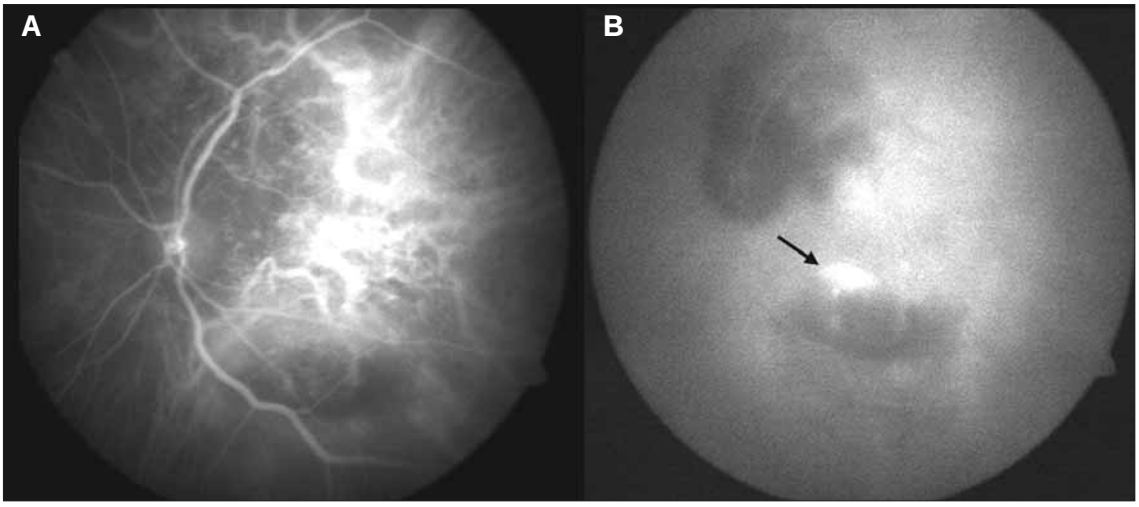

Fig. 2 - (A) *Early phase indocyanine green angiography revealing hypofluorescence superior to the optic disc at the site of the previous hemangioma. Note the dilated ectatic polypoidal choroidal vessels (PCV) in the macular region.* **(B)** *Late phase indocyanine green angiography showing persistent hypofluorescent area above the optic disc and actively leaking PCV inferior to the fovea (arrow).*

Vicuna-Kojchen and associates (4) reviewed the results of PDT for CCH in 9 patients. Two (22%) had transient visual disturbances and 1 (11%) developed choroidal neovascularization which was thought to be secondary to age-related macular degeneration rather than PDT. In a review of 8 patients with CCH treated with PDT, Landau and coworkers (5) found 1 (12.5%) who developed transient choroidal effusion and perifoveal retinal hemorrhage with subsequent visual loss.

Idiopathic PCV is a primary abnormality of the inner choroidal circulation with an unknown pathogenesis. Potential risk factors that may be linked to PCV include systemic hypertension, chorioretinal inflammation, or radiation retinopathy after plaque radiotherapy of a choroidal melanoma (6, 7). Li et al (6) reported the concurrent presence of PCV and CCH in an 80-year-old Chinese man. They proposed these two choroidal conditions could be related pathophysiologically. Spaide and associates (7) have found a related case of PCV following I_{125} plaque radiotherapy of a choroidal melanoma. They speculated that the choroidal neovascularization grew in response to choroidal ischemia caused by radiation therapy. In our case, we did not observe the secondary lipid exudate or PCV at the edge of PDT-treated CCH. Additionally, PCV developed 3 years after the PDT. However, considering the ethnic background, gender, and age of the patient, we speculate that PDT-induced choroidal ischemia could have contributed to the development of PCV.

In conclusion, PDT is a safe and effective treatment for CCH. Adverse reactions like exudative retinopathy or PCV can develop.

ACKNOWLEDGEMENTS

Support provided by the Scientific and Technical Research Council of the Turkish Republic (TUBITAK), Ankara, Turkey (S.T.); the Retina Research Foundation of the Retina Society, Capetown, South Africa (C.L.S.); the Paul Kayser International Award of Merit in Retina Research, Houston, TX (J.A.S.); a donation from Michael, Bruce, and Ellen Ratner, New York, NY (J.A.S., C.L.S.); Mellon Charitable Giving from the Martha W. Rogers Charitable Trust, Philadelphia, PA (C.L.S.); the LuEsther Mertz Retina Research Foundation, New York, NY (C.L.S.); and the Eye Tumor Research Foundation, Philadelphia, PA (C.L.S., J.A.S.).

None of the authors has a financial or proprietary interest in any method or material mentioned in the text.

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