Case Report

Retinal vasoproliferative tumor with associated cystoid macular edema treated with cryotherapy and intravitreal triamcinolone.

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CASE PRESENTATION

A 43-year-old white woman was referred with a gradual decrease in vision in the right eye over 3 months. Fifteen years previously she had been treated elsewhere for bilateral pars planitis with oral steroids. She was on no medications for systemic hypercholesterolemia.

At presentation, the best corrected visual acuities were 20/70 in the right eye and 20/30 in the left eye. Slit-lamp examination showed bilateral posterior subcapsular cataracts, and intraocular pressures were normal in both eyes. Funduscopy of the right eye disclosed a red, vascularized mass in the inferotemporal quadrant, with minimally dilated feeding vessels and measuring 8 x 8 x 4 mm (Figure 1). Extensive retinal exudation was found affecting all four quadrants of the fundus. B-scan ultrasonography disclosed a solid mass with high internal reflectivity and a secondary shallow retinal detachment. Fluorescein angiography showed hyperfluorescence of the lesion throughout all phases. There was cystoid macular edema (CME) on angiography, measuring 519 µm in thickness by optical coherence tomography (OCT) (Figure 1).

> Vasoproliferative tumors of the ocular fundus, previously named "presumed acquired retinal hemangiomas" or "retinal hemangioma-like lesions," are a distinct clinical entity.

The diagnosis of secondary retinal vasoproliferative tumor was made. Treatment with double-freeze thaw cryotherapy to the vasoproliferative tumor combined with intravitreal triamcinolone (4 mg/0.1 mL) injection was performed. Over 6 months, there was gradual resolution of the exudation, subretinal fluid and CME (Figure 2) . Macular thickness by OCT measured 171 µm. The patient underwent cataract surgery in the right eye, but the chronic damage from CME left her with visual acuity of 20/400. The vasoproliferative tumor appeared atrophic and measured 2.3 mm in thickness.

DISCUSSION

Vasoproliferative tumors of the ocular fundus, previously named "presumed acquired retinal hemangiomas"

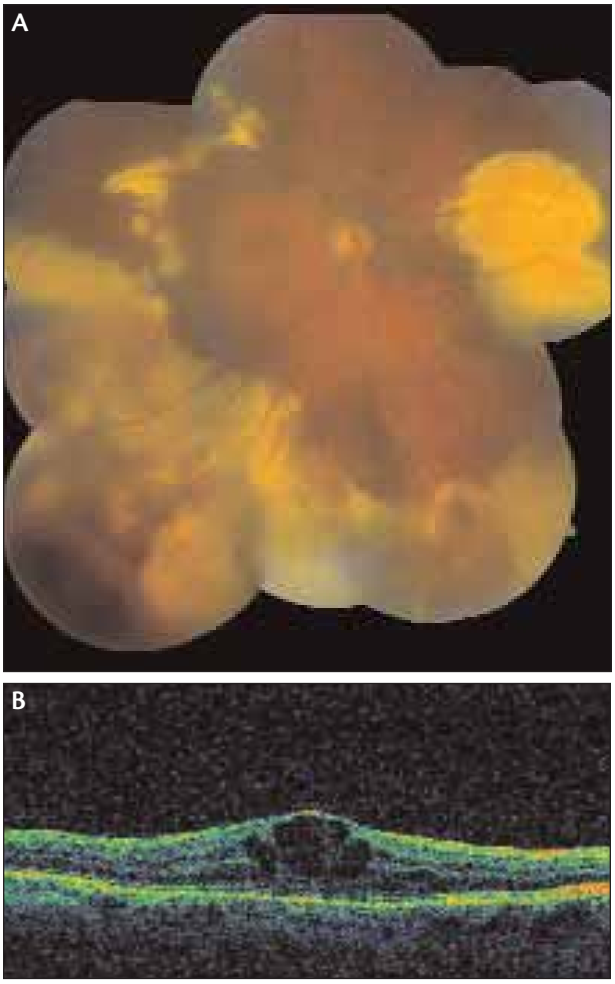

Figure 1. Highly vascularized mass located inferotemporally in the periphery of the right eye with extensive exudation and subretinal fluid (A). OCT of the right eye showing moderate CME (B).

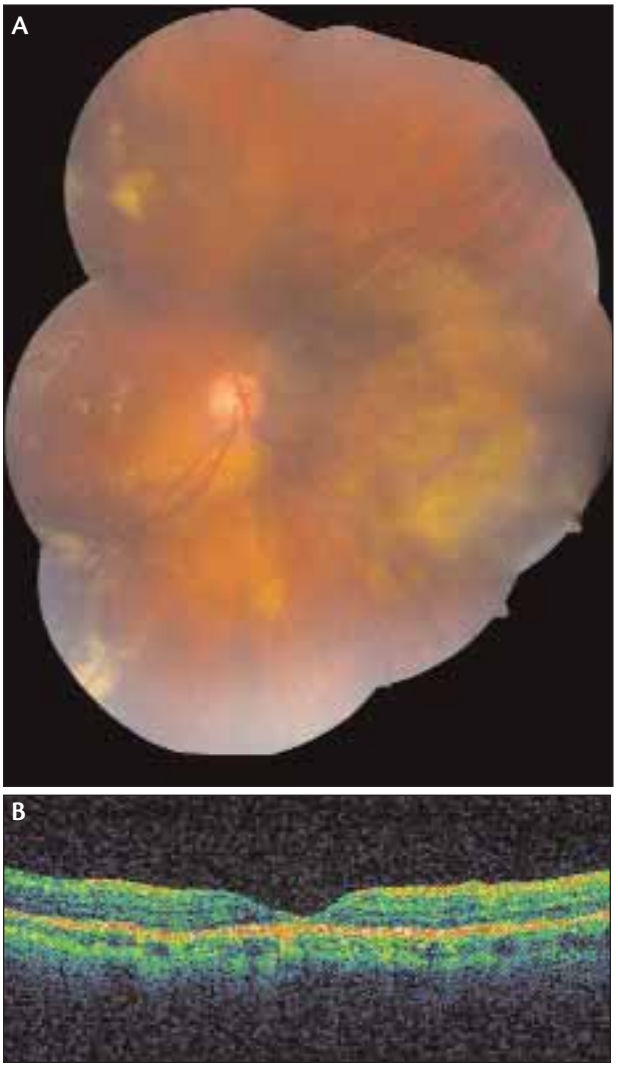

Figure 2. Six months after treatment, dramatic regression of the mass and secondary exudation and subretinal fluid is noted (A). OCT image disclosing resolution of the macular edema and foveolar thinning 6 months after treatment (B).

or "retinal hemangioma-like lesions," are a distinct clinical entity. In an analysis of 103 patients with this tumor, it was noted that this lesion could occur as a primary lesion or secondary to other conditions.[1] The primary tumors are idiopathic and represent 74% of all cases. The secondary tumors are associated with several conditions including pars planitis, retinitis pigmentosa, toxocariasis, and Coats' disease. The most common location for vasoproliferative tumors is the inferotemporal quadrant. On histopathologic exam, they have been shown to consist mostly of glial tissue proliferation with vascular elements.[2]

Our case showed dramatic response with complete resolution of the exudation and macular edema within 6 months following treatment with cryotherapy and intravitreal steroids. Other treatment options for vasoproliferative tumors include observation, laser photocoagulation, photodynamic therapy, plaque radiotherapy, and intravitreal injection of anti-vascular endothelial growth factor medications. In general, small tumors without exudation or subretinal fluid are observed, whereas those with the above findings are treated. The treatment choice depends on the tumor size and location. Smaller tumors are usually treated with non-radiotherapeutic methods while larger tumors, particularly those with extensive subretinal fluid, are treated with radiotherapy. ■

The treatment choice depends on the tumor size and location.

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1. Shields CL, Shields JA, Barrett J, et al. Vasoproliferative tumors of the ocular fundus. Classification and clinical manifestations in 103 patients. *Arch Ophthalmol.* 1995;113:615-623.
2. Irvine F, O'Donnell N, Kemp E, Lee WR. Retinal vasoproliferative tumors: surgical management and histological findings. *Arch Ophthalmol.* 2000;118:563-569.

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