Additional Contributions: Ed Trager, MS, created the pedigree in Figure 2.


Acquired Vasoproliferative Retinal Tumor: A Late Sequela of Retinopathy of Prematurity

Late sequelae of retinopathy of prematurity (ROP) in the Multicenter Trial of Cryotherapy for Retinopathy of Prematurity included retinal detachment, retinoschisis, retinal fold, and retrolental membrane. Examination of these eyes is often difficult, and mechanisms of failure are not thoroughly studied. We report a case of vasoproliferative retinal tumor (VPRT) in a teenager who had been born prematurely. We recommend that VPRT be added to the sequelae of chronic ROP that require examination of the peripheral retina beyond childhood.

Report of a Case. A 16-year-old boy visited for routine ROP follow-up. He was born at 27 weeks’ gestation, weighing 1134 g. He developed severe ROP leading to tractional retinal detachment despite laser photocoagulation. Subsequently, he underwent scleral buckle placement and multiple vitrectomies in each eye. In 2009, his visual acuity was hand motions OD and 20/300 OS. Dilated examination of the right eye showed a falciform fold extending from the disc to the periphery. The macula of the left eye showed pigmentary disruption and heterotropia. Inferonasally, there was an elevated mass 2 disc diameters in size with minimally dilated and tortuous feeder vessels. Fluorescein angiography showed leakage, consistent with a hemangioma-like lesion, and peripheral nonperfusion. A photographic survey of the periphery 3 years earlier documented absence of the lesion (Figure).

The feeder vessels and the tumor were treated with green laser, and additional laser was applied to the nonperfused area. Bevacizumab, 1.25 mg, was injected intravitreously. One month later, the tumor completely regressed with no leakage on fluorescein angiography.

Comment. We report a case of VPRT in a patient with ROP with residual nonperfusion. This tumor developed within 12 months of the previous examination and was successfully treated with laser photocoagulation and bevacizumab.

There is 1 previous report of an angioma-like mass in a patient with ROP. However, the diagnosis of ROP in that case was presumptive, and the timing of lesion development was unclear. Our case clearly documents the development of a VPRT during a 12-month interval. It is unique in that the patient was followed up photographically and we provided definitive proof that the lesion was not preexisting.

Exudative retinopathy has been described as a late sequela of ROP in adults. In 2 cases, mass lesions were noted, which may have been VPRT. Those cases were diagnosed as exudative retinopathy on the basis of the presence of lipid exudates, which were not present in our case. A recent article reported bilateral massive retinal gliosis in enucleated eyes after ROP. We suspect that exudative retinopathy and massive gliosis result from vasoproliferative stimulation from chronic ischemia.

Acquired VPRT has been associated with different ocular conditions, including uveitis, retinitis pigmentosa, and familial exudative vitreoretinopathy, although they remain idiopathic in most cases. These lesions are different.

Figure. Photographs of the left eye. A, Retcam color photograph in 2006. Arrow indicates the corresponding area at which the hemangioma-like lesion developed in 2009. B, Color photograph in 2009 showing a well-circumscribed, elevated, yellow lesion inferonasally anterior to the equator with associated tortuous feeder vessels. Scars from previous laser photocoagulation for retinopathy of prematurity are seen next to the tumor. The 2 white opacities seen in the middle of the photograph are posterior cortical opacities. C, Close-up of the hemangioma-like lesion. Early (D) and late (E) frames of fluorescein angiography showing leakage of the tumor as well as areas of nonperfused peripheral retina adjacent to the tumor. F, Immediately after laser photocoagulation of the tumor. Intravitreous bevacizumab was also given on the same day. G, One month after intravitreous bevacizumab treatment and laser photocoagulation of the tumor and peripheral nonperfused retina, showing complete regression of the tumor. H, Fluorescein angiogram showing no leakage.
from those associated with von Hippel-Lindau disease by unilocularity, negative family history, and the absence of large dilated tortuous feeder vessels. The exact cause is unknown, although it has been proposed that they could be expressions of vascularization of pigment epithelial proliferation or reactive gliosis. We postulate that in the setting of chronic ROP and familial exudative vitreoretinopathy, there is retinal ischemia that produces a microenvironment conducive to vascular proliferation.

Ophthalmologists who care for patients with a history of ROP should be aware of this late sequela because it is responsive to treatment and may cause additional vision loss. This lesion may be underreported because it may be difficult to identify in the setting of fibrotic residua and tractional elements. We recommend fluorescein angiography if new retinal thickening or exudates are seen in order to identify treatable lesions. In this case, we were able to identify the lesion before massive exudation occurred. Treatment options include laser photocoagulation, cryotherapy, photodynamic therapy, and, more recently, antivascular endothelial growth factor such as bevacizumab.7,8

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Bilateral Diffuse Uveal Melanocytic Proliferation With a Positive Ophthalmoscopic and Visual Response to Plasmapheresis

Bilateral diffuse uveal melanocytic proliferation (BDUMP) is a paraneoplastic syndrome resulting in profound bilateral vision loss, with approximately 30 cases reported in the world’s literature. In 1990, Gass et al1 described 5 characteristic signs of the disease: multiple subretinal round red patches in the retinal pigment epithelium (RPE), early fluorescence of these lesions on fluorescein angiography, multiple elevated pigmented and nonpigmented uveal melanocytic tumors with diffuse uveal tract thickening, exudative retinal detachments, and rapid cataract development. The histopathologic findings include diffuse uveal infiltration by benign hypopigmented spindle cells and occasional epithelioid cells. There is focal infiltration of the choroid by heavily pigmented melanocytes with sparing of the choriocapillaris.2-4 Destruction of the RPE occurs in areas overlying the infiltrate.5

Treatment for BDUMP has been largely unsuccessful. Modalities have included corticosteroids, ocular surgery, ocular radiation, and treatment of the underlying malignant neoplasm.1,2,5 While some have shown transient vision improvement or stabilization, we describe a new treatment for this visually devastating condition that resulted in vision improvement and stability with continued treatment until the patient’s death.

Report of a Case. A 72-year-old man had bilateral decreased, dim vision for 1 month. Four months prior, he was diagnosed as having metastatic bronchogenic carcinoma, for which he was taking sorafenib. Best-corrected visual acuities were 20/40+2 OD and 20/50+2 OS. Anterior segment examination results were normal. Dilated examination revealed clear media, a normal disc, and attenuated arterioles in each eye. In the right eye, there was a small pigmented lesion, a localized exudative retinal detachment, and an area of orange-brown giraffe-type pigmentation. In the left eye, 7 slightly elevated pigmented lesions with extensive giraffe-type pigmentation were present. The oval spots within the giraffe-type pigmentation appeared mildly hyperpigmented and were hypoautofluorescent on fundus autofluorescence photography and hyperfluorescent on fluorescein angiography in both eyes. The pigmented tumors appeared dark on indocyanine green angiography. Optical coherence tomography revealed subretinal fluid in the area of the inferotemporal retinal detachment, which extended to the fovea in the right eye. There was also subfoveal fluid in the left eye (Figure 1). B-scan ultrasonography showed diffuse choroidal thickening and discrete nodules with medium to high internal reflectivity. Electroretinographic results were normal. Goldmann visual fields revealed scotomas corresponding to the pigmented tumors and a generalized decreased peripheral visual field.

The findings were diagnostic of BDUMP. Treatment with sorafenib continued. Because we believed that a cir-