Central Retinal Vascular Obstruction Secondary to Melanocytoma of the Optic Disc

Jerry A. Shields, MD; Carol L. Shields, MD; Ralph C. Eagle, Jr, MD; Arun D. Singh, MD; Maria H. Berrocal, MD; Jose A. Berrocal, MD

A 35-year-old black man developed abrupt visual loss in his left eye. Ophthalmic examination revealed a deeply pigmented mass obscuring the optic disc, hemorrhagic retinopathy, and signs of central retinal vascular obstruction. Fluorescein angiography disclosed sluggish filling of the retinal blood vessels; ultrasonography disclosed an acoustically solid mass in the optic nerve head. Cytopathologic findings of a fine needle aspiration biopsy specimen demonstrated probable benign tumor cells, but melanoma could not be excluded. Histopathologic findings in the enucleated eye revealed a large, necrotic melanocytoma of the optic disc and hemorrhagic necrosis of the retina secondary to obstruction of the central retinal artery and vein. Melanocytoma of the optic nerve can undergo spontaneous necrosis and induce central retinal vascular obstruction. Abrupt visual loss in a patient with a melanocytoma does not necessarily imply malignant transformation.

Melanocytoma is a variant of melanocytic nevus that is most often recognized on the optic disc but can occur anywhere in the uveal tract. Most cases that occur on the optic disc are visually asymptomatic, but they can cause an afferent pupillary defect and a visual field defect. Although melanocytoma of the optic nerve rarely can cause visual loss when the tumor becomes necrotic, associated central retinal vascular obstruction is rare. We report a clinicopathologic correlation of a necrotic optic disc melanocytoma that caused a central retinal vascular obstruction and hemorrhagic retinopathy.

REPORT OF A CASE

A healthy 35-year-old black man noticed painless, progressive visual loss in the left eye for 1 week. He was found to have an optic nerve mass and retinal hemorrhages and was referred to the Ocular Oncology Service at Wills Eye Hospital, Philadelphia, Pa, for further evaluation and management of this condition. His medical history was otherwise noncontributory and he did not recall having a prior fundus examination. His visual acuity was 20/20 OD and hand movements OS. The left pupil was slightly dilated and there was a left afferent pupillary defect. Intraocular pressure was 21 mm Hg OU. Anterior segment examination showed scattered cells in the anterior chamber and anterior vitreous of the left eye. There was no iris neovascularization. The fundus of the right eye was normal and the pertinent findings were in the left fundus.

The left optic disc was obscured by a black mass that measured 5 × 3 mm in diameter (Figure 1). The retinal vessels were dilated and tortuous and the entire fundus posterior to the equator contained numerous superficial and deep retinal hemorrhages. A localized hemorrhagic retinal detachment extended from the optic nerve inferonasally. Many of the retinal vessels appeared white.

Fluorescein angiography demonstrated delayed flow through the retinal arteries and veins with hypofluorescence of the retinal hemorrhages. The black mass overlaying the optic disc remained hypofluorescent throughout the angiogram.
There was widespread vascular leakage and diffuse staining of the sensory retina in the late frames. Using ultrasonography, the lesion showed high internal reflectivity with A-scan and acoustic solidity with B-scan and measured 3 mm in thickness (Figure 3). Magnetic resonance imaging of the orbits demonstrated a small, elevated lesion at the level of the optic disc with 2 to 3 mm of increased enhancement in the optic nerve posterior to the globe (Figure 4). The lesion was slightly hyperintense to vitreous on T1-weighted images and markedly hypointense to vitreous on T2-weighted images. There was only minimal evidence of tumor in the optic nerve posterior to the lamina cribrosa. The patient underwent a systemic evaluation for neoplastic, vascular, and inflammatory diseases including a chest x-ray film, angiotensin-converting enzyme levels, and sickle cell preparation, all results of which were normal or negative.

Our clinical diagnoses were melanocytoma of the optic disc, secondary obstruction of the central retinal artery and vein, and hemorrhagic retinopathy. However, less likely possibilities, including a necrotic melanoma of the optic disc or a neoplasm of the retinal pigment epithelium could not be excluded on clinical grounds.

The options of observation, enucleation, or diagnostic fine-needle aspiration biopsy (FNAB) were discussed with the patient and he elected to have FNAB prior to making a definitive therapeutic decision. Fine-needle aspiration biopsy was performed through the pars plana of the ciliary body with a transvitreal route and indirect ophthalmoscopic guidance using a published technique.9 Cytopathologic studies demonstrated numerous benign-appearing cells that contained abundant cytoplasmic melanin (Figure 5). An experienced cytopathologist believed that the cells were more suggestive of adenoma or adenocarcinoma of the retinal pigment epithelium rather than melanocytoma or melanoma. However, melanoma could not be absolutely excluded.

The patient was informed of the poor visual prognosis and the possibility of malignancy and he elected to undergo enucleation. This was performed without complication, taking care to obtain a long section of optic nerve with the globe. Gross histopathologic examination of the eye before sectioning showed a normal globe with 18 mm of optic nerve attached. The globe did not permit transillumination, due to racial pigmentation. Gross examination of the sectioned globe revealed a pigmented epipapillary mass measuring $4 \times 4 \times 3$ mm and causing elevation of the circumpapillary retina. There were numerous superficial and deep retinal hemorrhages from the posterior fundus to the equatorial region.

Microscopic examination revealed scattered polymorphonuclear leukocytes, some of which had been ingested by macrophages, in the anterior chamber inferiorly. The lens was normal. The vitreous was detached posteriorly and contained small foci of blood. The optic disc and anterior portion of the optic nerve were replaced by an intensely pigmented and largely
necrotic tumor that extended into the optic nerve posterior to the lamina cribrosa (Figure 6). Bleached sections revealed that the residual viable tumor cells had a low nuclear-cytoplasmic ratio and bland nuclei (Figure 7). Surprisingly, some cells contained a prominent population of large round melanosomes, including some macromelanosomes, that persisted in the bleached sections. The tumor extended slightly into the peripapillary choroid and sensory retina. The adjacent choroid and retina were necrotic and contained extensive hemorrhage. The retinal blood vessels were markedly dilated and congested. Polymorphonuclear leukocytes filled the lumen of a large thick-walled blood vessel in the optic nerve head, presumably the central retinal artery. The optic nerve posterior to the tumor showed widening of the pial septae and increased cellularity consistent with gliosis. Cross sections of the surgical margin of the optic nerve showed no evidence of tumor.

The final diagnoses were as follows: (1) necrotic melanocytoma; (2) central retinal artery and vein obstruction secondary to necrotic melanocytoma; (3) hemorrhagic infarction of the retina secondary to central retinal artery and vein obstructions; (4) serosanguineous retinal detachment; (5) juxtapapillary choroidal necrosis and inflammation; (6) optic nerve atrophy; and (7) small hypopyon.

**COMMENT**

Melanocytoma of the optic disc is a benign melanocytic tumor that rarely causes visual impairment. However, this tumor has a tendency to undergo necrosis, particularly when located in the iris, where necrotic tumor cells and macrophages can cause reduction of aqueous outflow and secondary glaucoma. Although the visual loss is usually permanent, it has rarely been known to improve after the necrotic episode.

When a pigmented fundus lesion is associated with severe visual loss, it is tempting to attribute the visual impairment to malignant transformation. However, well-documented cases of
malignant transformation of optic disc melanocytoma are rare.

When visual loss occurs in an eye with a melanocytoma, it is more likely to be due to necrosis of the benign tumor rather than malignant transformation. The necrosis of the tumor within the optic nerve causes loss of optic nerve function and visual impairment. Although the sequence of events is not entirely clear, it is possible that the tumor causes vascular obstruction which, in turn, leads to necrosis in the tumor.

In rare instances, a necrotic optic disc melanocytoma can also cause obstruction of the central retinal vessels, as occurred in our patient. We are aware of 2 other cases of retinal vascular obstruction secondary to a melanocytoma of the optic nerve. In his series of optic nerve melanocytomas, Zimmerman reported the histopathologic findings in a case in which a necrotic tumor had caused a central retinal artery obstruction. Croxatto et al described a patient with a melanocytoma of the optic disc that caused retinal vaso-occlusive disease that led to neovascular glaucoma, requiring enucleation.

In our patient, it appears that the highly necrotic melanocytoma caused a combined central retinal artery and vein obstruction, as demonstrated with fluorescein angiography that disclosed minimal blood flow through the retinal vessels. It is quite possible that our patient also would have developed neovascular glaucoma had the eye not been enucleated.

Fine-needle aspiration biopsy is a generally reliable method for diagnosing intraocular tumors and pseudotumors in difficult cases. However, in some instances, it may be difficult to differentiate cytopathologically a melanoma, melanocytoma, and adenoma of the retinal pigment epithelium. In this case, the diagnosis of melanoma could not be excluded on the basis of cytopathologic findings from the FNAB, underscoring the limitations of that technique. Enucleation was justified in this case because melanoma was a possibility and the affected eye had minimal visual potential. This case demonstrates that an optic nerve melanocytoma can undergo necrosis and cause obstruction of the central retinal vessels and severe visual loss. Visual loss in a patient with an optic nerve melanocytoma does not necessarily mean that the tumor has undergone malignant transformation.

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