Diffuse Retinal Involvement in Malignant Nonteratoid Medulloepithelioma of Ciliary Body in an Adult

Intraocular medulloepithelioma is a rare embryonal neoplasm that occurs predominantly in children and arises from the primitive medullary epithelium of the optic cup. Medulloepitheliomas that contain heterotopic elements or tissues such as brain, cartilage, or rhabdomyoblasts are classified as teratoid, and tumors lacking these elements are classified as nonteratoid medulloepitheliomas. Both nonteratoid and teratoid medulloepitheliomas may be benign or malignant.

The clinical and histopathological features of intraocular medulloepitheliomas have been described previously in various reports. We herein report a case of a malignant nonteratoid medulloepithelioma of a ciliary body in an adult. To the best of our knowledge, this is the first documented report of a medulloepithelioma of the ciliary body with intraretinal involvement.

Report of a Case. A 27-year-old Hispanic man developed a mass in the left side of the neck in 1976. A histopathological diagnosis of Hodgkin disease was made. The patient was treated with radiation and chemotherapy. Following treatment, he remained free of disease. In October 1984, the patient noted decreased vision in the left eye of 1-week duration. The initial clinical diagnosis was iritis and vitreous hemorrhage of unknown etiology.

The patient underwent uncomplicated phacoemulsification of the cataract in his left eye with implantation of a posterior chamber intraocular lens. In March 1991, the patient had a visual acuity of 20/400 and intraocular pressure of 25 mm Hg, and he developed ruberosis iridis in the left eye. The angle was totally occluded and tumor cells were present in the anterior chamber. A recurrent ciliary body tumor was noticed at the 11-o’clock position.

Pathologic Findings. Grossly, the specimen consisted of a firm left eye measuring 23.5 × 24 × 23 mm with no optic nerve attached. The semitranslucent cornea measured 12 × 11 mm. An elevated yellowish-tan subconjunctival nodule measuring 7 × 6 × 4 mm was present between the 9- and 11:30-o’clock positions. The nodule was located 1 mm behind the limbus. On transillumination, a shadow measuring 8 × 6 mm was noted in the ciliary body nasally adjacent to the epibulbar mass.

The epibulbar mass was connected through a ciliary body coloboma to a 5 × 3-mm yellowish-tan mass involving the ciliary body nasally (Figure 1). The intraocular and epibulbar masses showed a dumbbell-shaped configuration. The vitreous was liquefied and condensed anteriorly. The retina was attached and thickened, and its surface was studied with multiple elevated gelatinous nodules of 1 to 1.5 mm (Figure 1). The optic disc was absent, and a small scleral hole was present in the peripapillary region. A portion of choroid was visible within the scleral defect at the optic disc.

Histopathologically, the tumor displayed cystic structures filled with primitive vitreous and outlined by distended neuroepithelial tubules and cords that resembled the primitive medullary epithelium (Figure 2). Higher magnification of the neuroepithelial tubules showed increased mitotic activity and apoptotic bodies (Figure 3). Sheets of tumor cells were present throughout the posterior chamber and around the remnants of the lens capsule (Figure 4). Diffuse seeding of tumor cells along the inner surface of the retina and vitreous with multiple foci of intraretinal invasion were present (Figure 5). Stains for mucopolysaccharides (colloidal iron before and after hyaluronidase digestion) disclosed abundant pools of hyaluronidase-sensitive mucopolysaccharides (hyaluronic acid) within the neuroepithelial tubules and rosettes (Figure 6). The histopathological diagnosis was a malignant nonteratoid medulloepithelioma of ciliary body with diffuse intraretinal involvement and extracocular (sub-
conjunctival) extension. There were broad anterior synechiae present temporally with ectropion uveae.

Endothelialization and descemetization of anterior and posterior iridic surfaces were present.

Comment. Intraocular medulloepithelioma usually arises from the ciliary body, rarely the optic nerve, reti-

Figure 1. Gross appearance of the enucleated eye depicting a bilobed grayish-white mass at the site of a surgical coloboma of the ciliary body. Notice a posterior chamber intraocular lens partially surrounded posteriorly by a translucent grayish membrane. The vitreous has been condensed anteriorly. The peripheral retina on both sides has thickened and shows prominent cystoid changes.

Figure 2. Solid neuroepithelial tubules, some containing rosettes, are present along the surface of the ciliary body. Foci of necrosis of the ciliary body with melanin-laden macrophages are observed (hematoxylin-eosin, original magnification ×10).

Figure 3. High-power view of the tumor displaying neuroepithelial tubules with increased mitotic activity and apoptotic bodies (hematoxylin-eosin, original magnification ×100).

Figure 4. A monolayer of primitive neuroepithelial cells is growing along the surface of the anterior lens capsule, which has been folded upon itself. The lumen occupied by 1 haptic (H) is identified (hematoxylin-eosin, original magnification ×51).

Figure 5. Sections of sensory retina showing papillary fronds and sheets of tumor cells growing along the wrinkled internal limiting membrane of the retina. The intraretinal tumor, which reaches the inner nuclear layer, shows cystic and tubular structures that stain positively with colloidal iron. This material is sensitive to hyaluronidase digestion (hematoxylin-eosin, original magnification ×40).

Figure 6. Adjacent section to that shown in Figure 5 displaying the presence of mucopolysaccharides along the internal limiting membrane of the retina as well as in cystic structures and neuroepithelial tubules in the inner nuclear layer (colloidal iron, original magnification ×40).
nal stalk, or retina.\textsuperscript{2,3} It is a neoplasm of childhood that usually becomes clinically symptomatic during the first decade of life (mean age, 5 years).\textsuperscript{1} However, there are well-documented cases in which the tumor had become symptomatic in adulthood.\textsuperscript{4,5} The most frequent clinical signs are leukocoria; notching or subluxation of the lens; cataract; and a mass in the iris, ciliary body, or anterior chamber. Almost all tumors are unilateral. There is no predilection for this tumor for race, sex, and laterality.\textsuperscript{6} It also has a strong tendency to induce secondary glaucoma due to iris neovascularization.

Broughton and Zimmerman\textsuperscript{1} established the histopathological criteria for malignancy that includes the presence of poorly differentiated neuroblastic cells, numerous mitoses, pronounced pleomorphism, sarcomatous areas, or invasion of other ocular structures with or without extraocular extension. Mostly these tumors are nonpigmented; however, a few cases of clinically pigmented medulloepithelioma of ciliary body have been reported.\textsuperscript{10} Metastases are very rare and usually occur in cases with extraocular extension.\textsuperscript{6}

Immunohistochemically, the neuroblastic cells are positive for neuron-specific enolase and synaptophysin while the spindle cells in the stroma stain positively for vimentin, glial fibrillary acidic protein, and S100 protein.\textsuperscript{7} Ultrasonography and cytologic examination of vitreous aspirate have led to correct preenucleation diagnosis of medulloepithelioma in a few cases.\textsuperscript{8}

Shields et al\textsuperscript{10} concluded that local surgical resection (iridocyclectomy) of the tumor is usually insufficient and enucleation ultimately will be necessary because of tumor recurrence. Six of the 10 patients in their series were managed by local resection, and 5 eventually required enucleation, 4 because of local tumor recurrence. One of the 10 cases in the same series had intraretinal involvement (J. A. Shields, oral communication, April 2004).\textsuperscript{9} Carrillo and Streeter\textsuperscript{2} reported a case of malignant teratoid medulloepithelioma in an adult in which the tumor had extended over the inner retinal surface (in the macular region), producing large retinal con traction folds, but no evidence of intraretinal involvement was present.

Our patient had an unusual mass that disclosed extensive seeding of tumor cells along the internal limiting membrane of the retina with foci of intraretinal involvement. In addition, seedings of tumor cells were present along the anterior segment structures, surrounding the remnants of the anterior lens capsule. We believe it is quite unlikely that the pattern of spread of the tumor to the retinal surface and onto the lens capsule is related to the prior cycloectomy specimen. To our knowledge, this is a unique case of medulloepithelioma of ciliary body with diffuse intraretinal involvement occurring in a young adult.

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**Bilateral Choroiditis From Prototheca wickerhamii**

**Algaemia**

Algae are a diverse group of eukaryotic, photosynthetic microbes. They are not plant, animal, or fungus but are—along with molds and mildews—members of the kingdom Protista. Though generally considered noninfectious agents in mammals, a few algae are pathogenic. One example is Prototheca, a nonphotosynthetic alga. Prototheca organisms exist worldwide and are readily isolated from rivers, lakes, ponds, and soil. While protothecosis in canine and bovine species is common, infection in humans is rare. In the past 25 years, approximately 100 human cases of protothecosis have been reported. Most of these cases have been caused by Prototheca wickerhamii, although Prototheca zopfi has been recovered from 2 patients.\textsuperscript{1}

The most common site of protothecosis is the skin, and this may manifest as erythema, papulonodules, plaques, and ulcers. Immuno-compromised patients tend to have more severe lesions that may be vesiculobullous with purulent discharge. Disseminated infection such as peritonitis, endocarditis, urinary tract infection, or meningitis may also occur less commonly.\textsuperscript{1} Ocular manifestations of protothecosis have never been reported in a human; however, several cases have been reported in animals (primarily in dogs).\textsuperscript{2-10}

We report what is to our knowledge the first case in the literature of bilateral choroiditis due to algae in a human.

**Report of a Case.** Three years prior, a 58-year-old white man was diag-