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. Results of an ophthalmologic examination of the right eye were entirely normal.

After treatment with steroids, the iritis improved and the vitreous hemorrhage cleared. A mass compatible with a ciliary body tumor was visible superonasally. Computed tomographic scans showed an intraocular mass in the left eye. No evidence of extraocular involvement was noticed. In February 1985, the patient underwent cyelectomy for a presumptive clinical diagnosis of malignant melanoma of the ciliary body. Histopathological examination showed large deposits of hyaline material and foci of hemorrhage containing only a few neuroepithelial structures. In our opinion, the material was insufficient to establish an unequivocal diagnosis.

In August 1987, 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. In September 1991, a staphyloma was noticed on the superonasal quadrant. In August 1992, the patient underwent enucleation of his left eye, which was sent to the Ophthalmic Pathology Laboratory of the Cullen Eye Institute (Houston, Tex) for histopathological examination. The patient was alive and well when last examined in March 1998 (6 years after the enucleation). Unfortunately, the patient was lost to follow-up.

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 eye was opened horizontally, with the superior calotte removed first. The anterior chamber was totally obliterated. A posterior chamber intraocular lens was present.

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 extraocular (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-
nal stalk, or retina. It is a neoplasm of childhood that usually becomes clinically symptomatic during the first decade of life (mean age, 5 years). However, there are well-documented cases in which the tumor had become symptomatic in adulthood. The most frequent clinical signs are leukocoria; nystagmus; strabismus; and a mass in the iris, ciliary body, or anterior chamber. Almost all tumors are unilateral. There is no predilection of this tumor for race, sex, and laterality. It also has a strong tendency to induce secondary glaucoma due to iris neovascularization.

Broughton and Zimmerman 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. Metastases are very rare and usually occur in cases with extraocular extension.

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. Ultrasonography and cystic examination of vitreous aspirate have led to correct preenucleation diagnosis of medulloepithelioma in a few cases.

Shields et al 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). Carillo and Streeter 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 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 cyclectomy 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 zoophi has been recovered from 2 patients.

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. Ocular manifestations of protothecosis have never been reported in a human; however, several cases have been reported in animals (primarily in dogs).

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-