Adenoma of the Nonpigmented Ciliary Epithelium Mimicking a Malignant Melanoma of the Iris

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Adenomas of the nonpigmented ciliary epithelium are rare tumors and are difficult to differentiate from amelanotic malignant melanomas of the ciliary body. We describe a 62-year-old woman with a nonpigmented, vascularized iris tumor with small satellites involving the anterior chamber angle. Clinically, an amelanotic, tapioca, malignant melanoma of the iris was suggested although the blood-aqueous barrier was intact. A block excision of 7.5 mm, a tectonic corneoscleral graft, and a simultaneous extracapsular cataract extraction with implantation of a posterior chamber lens were performed. Histopathologic and electron microscopic studies revealed an adenoma of the nonpigmented ciliary epithelium. Postoperative visual acuity was 20/40 OD after 6 weeks. An adenoma of the nonpigmented ciliary epithelium may mimic an amelanotic malignant melanoma of the iris and should be considered in its differential diagnosis.

Adenomas of the nonpigmented ciliary epithelium (NPCE) are rare tumors that may cause subluxation of the lens, segmental cataract, or secondary glaucoma. It is difficult to differentiate tumors of the NPCE from malignant melanomas or metastatic tumors of the ciliary body. In contrast with malignant melanomas, adenomas of the NPCE are more likely to display an irregular surface, to transmit light well on transillumination, and to show high internal reflectivity on ultrasonography. We describe a patient with an adenoma of the NPCE appearing as an ill-defined nonpigmented iris mass with small satellite lesions that mimicked a tapioca malignant melanoma of the iris.

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A 62-year-old woman had an asymptomatic, nonpigmented iris tumor of the right eye between the 11- and 1-o’clock positions that showed prominent blood vessels, an irregular surface, and several small satellite lesions anterior to the iris surface in the aqueous (Figure 1). The mass measured 4 mm in diameter. Gonioscopic examination showed that the tumor covered the anterior chamber angle between the 11- and 1-o’clock positions (Figure 1). A segmental cataract was present. Best-corrected visual acuity was 20/20 OU and the intraocular pressure was 12 mm Hg. Otherwise, the ophthalmologic examination showed no abnormalities. The results of flare and cell counts showed no abnormalities. Ultrasound examination revealed high internal reflectivity of the mass and ultrasound biomicroscopy disclosed partial involvement of adjacent ciliary body. The tumor did not block transillumination. Clinically, a nonpigmented malignant melanoma of tapioca type of the iris was suspected. Despite tumor cell seeding, the blood-aqueous barrier was intact and local excision was advised. A (7.5-mm) block excision, tectonic corneoscleral graft simultaneous with an extracapsular cataract extraction (phacoemulsification), and implantation of a posterior chamber lens were performed by one of us (G.O.H.N.). Six weeks postoperative visual acuity was 20/40 OD, and there is no evidence of recurrence (Figure 2).
Histopathologic examination revealed a predominantly unpigmented tumor emanating from the nonpigmented epithelium of the ciliary body and extending through the iris stroma into the anterior chamber (Figure 3). The tumor consisted of loosely arranged, round to polygonal, well-differentiated, epithelial cells located in an abundant hyaluronidase-sensitive, mucoid, extracellular material (Figure 3). Capillaries with perivascular inflammatory cells (lymphocytes and plasma cells) and many large cells with a vacuolated cytoplasm were present (Figure 3). Fontana staining was negative. Acid mucopolysaccharides sensitive to hyaluronidase (hyaluronic acid) were abundant.

Using an immunoperoxidase technique, vascular endothelial growth factor, vimentin, and S-100 protein were detectable in the tumor cells; however, HMB-45 was not found.

Electron microscopically, tumor cells did not show intracellular melanin granules, but displayed intercellular interdigitations, gap junctions, desmosomes, microvillous processes, basement membrane material, rough endoplasmic reticulum, polysomes, cytoplasmic filaments, and irregularly outlined nuclei with evenly dispersed chromatin (Figure 4). Part of the cells were dilated with intracellular mucoid material (Figure 4). Electron microscopic demonstration of basement membranelike and mucoid extracellular material confirmed the histopathologic diagnosis of an adenoma of the nonpigmented ciliary body epithelium.

**COMMENT**

Adenomas of the NPCE are clinically often indistinguishable from amelanotic malignant melanomas of the ciliary body or metastatic carcinomas. The described adenoma of NPCE in our study is unusual because it seems to be an amelanotic malignant melanoma of the iris with small satellite lesions on the iris surface and extension into the anterior chamber. To our knowledge, these features of an adenoma of the NPCE have not been reported before, although retroiridic location, anterior distortion, and thinning of iris due to tumors of NPCE have been noted before. The development of small satellite lesions on the iris and the irregular tumor surface may be due to disruption of the epithelial lining of the tumor because of excessive production of hyaluronic acid; abundance of mucus was described in a previous article about an adenoma of NPCE. The location of the tumor in the anterior chamber is due to extension of the mucus-producing mass through the iris stroma. The tumor seems highly vascularized, but on histologic examination only a few capillaries were observed. The nonpigmented transparent features of the adenoma allowed observation of the capillar-
Figure 3. A, Light microscopy shows a large adenoma of the nonpigmented ciliary body epithelium extending into the anterior chamber. Discrete reactive hyperplasia of adjacent pigmented ciliary epithelium (hematoxylin-eosin, original magnification ×16). B, Note abundant hyaluronidase-sensitive mucus (hyaluronic acid) and scattered epithelial tumor cells in between (acid mucopolysaccharides, original magnification ×40). Alcian blue was positive at pH 2.5 and negative at pH 1. Sulfated mucopolysaccharides could not be detected.

Figure 4. A, Electron microscopy shows tumor cells from the adenoma of the nonpigmented ciliary epithelium with extracellular accumulation of mucoid material (*) and basement membrane material (bm; scale bar=5 µm). B, A desmosome between epithelial cells of the adenoma of the nonpigmented ciliary body epithelium (arrow; scale bar=1 µm). C, Tumor cell with prominent rough endoplasmic reticulum surrounded by basement membrane material (bm; scale bar=5 µm). D, A tumor cell with dilated cisterns of rough endoplasmic reticulum and beginning accumulation of mucoid material (scale bar=5 µm). E, A vacuolated tumor cell with intracellular accumulation of abundant mucoid material traversed by cytoplasmic strands (scale bar=10 µm).
ies. Immunohistochemistry for vascular endothelial growth factor was positive within tumor cells; but is it unclear whether this is related to tumor angiogenesis or to low constitutive expression of vascular endothelial growth factors in normal adult ocular tissue. Our findings demonstrate that an adenoma of the NPCE with small satellite lesions on the iris surface can mimic an amelanotic malignant melanoma of the iris and the chamber angle. An adenoma of the NPCE has to be considered in the differential diagnosis of malignant melanomas of the iris.

Block excision with a tectonic corneoscleral graft is indicated to confirm the diagnosis, avoid enucleation, and maintain satisfactory visual acuity.¹⁻⁴

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REFERENCES


From the Archives of the Archives

Hortz reports a number of cases in which he has filled in the space left after the excision of large pterygia by grafts from the skin behind the ear. He makes the graft a little smaller than the denuded space, in the horizontal direction. Both eyes are bandaged for 48 hours. In four reported cases there was but one failure.