Adenoma of Nonpigmented Ciliary Epithelium With Smooth Muscle Differentiation

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A 35-year-old man underwent successful iridocyclectomy for a ciliary body mass that was subluxating the lens and causing a secondary cataract. Histopathologically the mass proved to be a spindle cell tumor, but leiomyoma and melanoma were initially considered to be diagnostic possibilities. However, further studies and immunohistochemical studies revealed that the tumor probably arose from the nonpigmented ciliary epithelium. The spindle cells represented smooth muscle differentiation within the mass. The final diagnosis was adenoma of the nonpigmented ciliary epithelium with smooth muscle differentiation.

Acquired adenoma of the nonpigmented ciliary epithelium (NPCE) is relatively rare but recent authors have reviewed the literature on this tumor and have defined its clinical and histopathologic features. We describe a patient who had a ciliary body tumor that was difficult to diagnose histopathologically because it was composed mostly of amelanotic spindle cells. Results of more detailed histopathologic and immunohistochemical studies showed that the tumor was an adenoma of the NPCE with smooth muscle differentiation.

REPORT OF A CASE

A 35-year-old African American man was referred for a ciliary body mass in his right eye. Prior to the detection of the tumor, he had been diagnosed as having intraocular inflammation and was treated for 1 month with systemic and topical antibiotics and corticosteroids with no relief of his ocular pain or visual impairment. His ocular and systemic histories were otherwise unremarkable.

Best-corrected visual acuities were 20/60 OD and 20/20 OS. Intraocular pressures were 18 mm Hg in each eye. The affected right eye had fine keratic precipitates and moderate flare and cells in the anterior chamber and anterior vitreous cavity. There was a ciliary body mass superiorly that was subluxating the lens and producing a dense focal cortical cataract that partially obscured the tumor (Figure 1). The posterior fundus was normal. Transillumination showed a shadow measuring 7 mm in diameter extending from the pars plicata to the midportion of the pars plana. Fluorescein angiography revealed early filling and late diffuse staining of the mass. B-scan ultrasonography disclosed a pedunculated mass with medium internal reflectivity.

Pathologic Findings

Grossly the tumor measured $7 \times 5 \times 4$ mm and rested on a base of normal ciliary epithelium.
ary body and sclera 12 mm in diameter (Figure 2). Low-magnification microscopy showed that the peripheral portion of the tumor was composed of viable tumor cells that surrounded a large focus of central necrosis (Figure 3). The viable peripheral region contained plump spindle cells with abundant eosinophilic cytoplasm, fairly uniform nuclei, and moderately prominent nucleoli (Figure 4). Many spindle cells were encompassed by periodic acid–Schiff–positive matrix tissue. Mitoses were not seen. Peripherally, the necrotic material was rimmed by foamy histiocytes.

Although the tumor appeared on initial inspection to be a primary spindle cell neoplasm (Figure 4), it rested on the inner surface of the ciliary body and did not involve the stroma. Furthermore, continuity between the tumor and the NPCE was evident in a few sections. A few bands of polarized epithelium that rested on periodic acid–Schiff–positive basement membrane were observed near the base of the tumor (Figure 5).

The tumor was studied with a battery of immunohistochemical stains. The melanoma-specific antigen (HMB-45) and glial fibrillary acidic protein were negative. The tumor cells displayed impressive immunoreactivity for vimentin. The ciliary epithelium and a few foci of tumor cells also stained positively for S-100 protein. Neither the ciliary epithelium nor the tumor stained positively for 3 keratin markers (903, middle molecular weight; 904, high molecular weight; and CAM 5.2, low molecular weight) (Enzo Manufacturers, New York, NY). The peripheral spindle cells showed intensely positive immunoreactivity for both muscle-specific actin and smooth muscle actin (Figure 6), with the ciliary musculature serving as an internal positive control. The latter observation raised the diagnostic possibility of leiomyoma, which occurs as a spindle cell tumor in the ciliary body and is immunoreactive for smooth muscle actin. However, the aforementioned epithelial features and the absence of ciliary body stromal involvement established that the tumor was an adenoma of the NPCE with smooth muscle differentiation and necrosis.
Acquired neoplasms of the NPCE are relatively rare. Zimmerman emphasized the remarkable polymorphism of tumors of the NPCE and categorized them into solid, papillary, and pleomorphic types. In contrast to other reported cases, the tumor reported here was composed largely of bland spindle cells, raising the possibilities of leiomyoma and melanoma. However, the tumor rested on the inner surface of the ciliary body and spared the uveal stroma, where the aforementioned spindle cell tumors would ordinarily reside. The low-grade spindle cell component of the tumor displayed impressive positive immunoreactivity for the muscle markers, muscle-specific actin, and smooth muscle actin. Although decidedly unusual in an adenoma of the NPCE, smooth muscle differentiation should not be a totally unexpected finding because the iris dilator muscle is a derivative of the neighboring neuroepithelium.

We have recently reviewed our clinicopathologic experience with 9 cases of acquired neoplasms of the NPCE, reviewed the literature, summarized the clinical and histopathologic features of these lesions, and pointed out the features that help differentiate them from melanoma and other ciliary body lesions. Clinically, adenoma of the NPCE generally is nonpigmented, yellow to light tan in color, and often has an irregular, sometimes multilobulated, free surface that directly impinges the vitreous cavity. Histopathologically, adenoma of the NPCE usually is composed of nonpigmented cuboidal or columnar cells that can assume solid, papillary, or pleomorphic patterns. Most tumors of the NPCE show strands of polarized epithelial cells that rest on a prominent periodic acid-Schiff-positive basement membrane. Some tumors contain prominent pools of hyaluronidase-sensitive acid mucopolysaccharide. Tumors that show cytologic atypia exhibit growth and aggressive behavior and local invasion. Despite the malignant classification, these tumors are generally not recognized to undergo distant metastasis. In the case reported here, necrosis was a prominent histopathologic feature. Our other cases of adenoma of the NPCE have not shown appreciable necrosis. The spindle cells in our case exhibited immunohistochemical characteristics of smooth muscle differentiation. Similar spindle cells were not observed in other cases of adenomas of the NPCE.

Historically, most cases of acquired neoplasms of the NPCE were managed by enucleation because they were suspected clinically to be malignant melanoma. More recently, we have had remarkable success in managing these tumors with local resection. Because acquired tumors of the NPCE typically are cytologically benign and the affected eye generally has good vision, it seems reasonable to locally remove the tumor rather than enucleate the affected eye. This is generally accomplished by partial lamellar sclerouvectomy.

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