Age-Related Hyperplasia of the Nonpigmented Ciliary Body Epithelium (Fuchs Adenoma) Simulating a Ciliary Body Malignant Neoplasm

Fuchs adenoma (coronal adenoma) is a peculiar mass that develops in the pars plicata of the ciliary body. It is believed to be a reactive proliferation of the nonpigmented ciliary epithelium (NPCE) that is associated with aging. Hence, we prefer to use the noneponymic, more descriptive term age-related hyperplasia (ARH) of the NPCE. Although extremely common, ARH-NPCE generally remains undetected clinically because of its occult location in the ciliary body. We report an unusual case of ARH-NPCE that grew through the iris root, was removed by iridocyclectomy, and was confirmed histopathologically.

Report of a Case. A 54-year-old woman was referred because of a suspected ciliary body melanoma. Her corrected visual acuity was 6/6 OU and intraocular pressures were normal. The left eye was healthy. Slit-lamp biomicroscopy of the right eye disclosed a tan ciliary body mass that had eroded through the iris root and was barely visible in the anterior chamber angle (Figure, A). There was a cortical cataract nasally, which obscured a view of the tumor (Figure, B). Ultrasound biomicroscopy revealed a well-defined ciliary body mass 3 mm in thickness (Figure, C). The differential diagnosis included ciliary body melanoma, acquired neoplasm of the NPCE, tumor of the pigment epithelium, and leiomyoma; ARH-NPCE was not considered. The tumor was removed by iridocyclectomy (Figure, D). Three years after surgery, visual acuity in the affected eye was 6/15 owing to continued enlargement of the cataract into the visual axis.

Gross examination revealed a well-circumscribed white mass arising from the ciliary body measuring 3 × 3 mm in diameter. Histopathologic evaluation disclosed linear segments of nonpigmented, benign, ciliary epithelial cells encompassing large amounts of avascular, amorphous, periodic acid–Schiff–positive extracellular matrix (Figure, E and F). There was no evidence of malignancy. The final diagnosis was ARH-NPCE.

Comment. Age-related hyperplasia of the NPCE (Fuchs adenoma or coronal adenoma) occurs in an occult location in the pars plicata of the ciliary body. Although it is extremely common in enucleated eyes, it generally is unapparent clinically because it is small and asymptomatic. It is believed to represent a proliferation of the NPCE that is associated with aging. In their study of eyes mostly at autopsy, Iliff and Green found 7 cases of ARH-NPCE among 11 patients older than 80 years compared with 1 lesion in 17 eyes from patients younger than 50 years. Bateman and Foos, in a similar series, also found a relationship with age. Age-related hyperplasia of the NPCE can be solitary or multiple and unilateral or bilateral, and usually it is confined to 1 ciliary process. When it is larger, it can simulate a cyst or a solid neoplasm. Single lesions removed by iridocyclectomy were included in the series by Iliff and Green and Bateman and Foos. Another lesion removed by iridocyclectomy was reported by Zaidman et al in 1983. Our literature search disclosed no additional cases that were recognized clinically. Rarely, ARH-NPCE can assume tumorous proportions as occurred in our case.

In summary, we report a case of ARH-NPCE that enlarged, caused a secondary cataract, and grew through the iris root to appear as a mass in the anterior chamber angle. It is important to realize that ARH-NPCE rarely can grow to sufficient size to simulate a malignant neoplasm of the ciliary body.

Correspondence: Dr J. A. Shields, Ocular Oncology Service, Wills Eye Institute, 840 Walnut St, Philadelphia, PA 19107 (jerry.shields@shieldsoncology.com).

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Contrast Sensitivity Following Amblyopia Treatment in Children

Prior studies have found a reduction in contrast sensitivity in eyes with amblyopia using sinusoidal gratings, whereas minimal loss has been reported with Pelli-Robson charts. Most studies have evaluated contrast sensitivity at the time of diagnosis of amblyopia or after short-term treatment. A follow-up study of an earlier randomized trial provided us the opportunity to evaluate contrast sensitivity using Pelli-Robson low-contrast letter charts at age 10 years, several years after treatment of amblyopia.

Figure. Clinical, ultrasonographic, and histopathologic findings. A, Focus of the tan tumor extending through the iris root and appearing in the anterior chamber angle (arrow). B, Right gaze, showing a cortical cataract overlying the ciliary body mass. The underlying tumor cannot be clearly photographed because of the cataract. C, Ultrasound biomicroscopy showing the ciliary body mass (arrow). D, View of the exposed tan tumor at the time of iridocyclectomy. E, Photomicrograph showing the mass arising from the pars plicata of the ciliary body. The linear basophilic areas represent cells of the ciliary epithelium, and the pink area represents basement membrane material elaborated by the nonpigmented ciliary epithelium (hematoxylin-eosin, original magnification ×10). F, Another section through the tumor showing more typical cords of nonpigmented ciliary epithelium cells that are separated by basement membrane material (hematoxylin-eosin, original magnification ×75).