
SHORT COMMUNICATION

Adenoma of the nonpigmented ciliary epithelium

J. ELIZALDE, S. UBIA, R.I. BARRAQUER

Ocular Oncology Service, Centro de Oftalmología Barraquer, Barcelona - Spain

PURPOSE. *To report the clinicopathologic features of an adenoma of the nonpigmented ciliary epithelium.*

DESIGN. *Single interventional case report.*

METHODS. *A nonpigmented iris and ciliary body tumor was diagnosed in a 66-year-old woman who complained of blurred vision related to a unilateral cataract. A combined cataract surgery and partial lamellar sclerouvectomy was performed.*

RESULTS. *Histopathologic findings disclosed an adenoma of the nonpigmented ciliary epithelium.*

CONCLUSIONS. *Unilateral cataract in an adult patient can be rarely related to a tumor growing from the ciliary epithelium. Adenoma of the non pigmented ciliary epithelium may mimic an amelanotic melanoma. Partial lamellar sclerouvectomy is an effective method to manage this condition and to confirm the diagnosis. (Eur J Ophthalmol 2006; 16: 630-3)*

KEY WORDS. *Adenoma, Ciliary epithelium tumor, Iridocyclectomy, Iris pseudomelanoma*

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INTRODUCTION

In the clinical context of neoplasms of the nonpigmented ciliary epithelium (NPCE) it is necessary to differentiate the congenital forms (medulloepithelioma) from the acquired presentations (1-3). Among the latter, a distinction is to be made between the cytologically benign forms (adenoma or benign epithelioma) and locally malignant lesions (adenocarcinoma) (4-9) with very little tendency to metastasize.

Adenoma of the NPCE is an amelanotic ciliary body mass that can have variable features. It frequently causes an adjacent cataract or a dislocated lens. The extremely low prevalence of these tumors often causes them to be mistaken for other more common iridociliary tumors such as melanoma or uveal metastases. We present the clinical and histopathologic characteristics of an adenoma of the NPCE diagnosed during routine preoperative exploration for cataract surgery.

Case report

A 66-year-old woman presented with progressive loss of visual acuity for the previous 5 months, a diagnosis of cataracts, and an intraocular lesion in the right eye. Examination of the affected eye revealed an advanced cataract and a certain prominence of the iris towards the anterior chamber in the nasal quadrant (Fig. 1). After dilating the pupil an evident irregular pupillary shape was noted. Visual acuity was 0.06, and the intraocular pressure was normal. Dilated eye examination revealed a bilobular and vascularized minimally pigmented tumor in the ciliary body (Fig. 2A), compressing the lens at its equatorial zone, and invading the ciliary processes. The lesion showed small transilluminable cystic lesions, and did not appear to extend beyond the ciliary band (Fig. 2B). Ultrasound revealed the presence of a solid mass measuring 4.07 mm x 3.10 mm, with moderate internal reflectivity and a negative kappa angle (Fig. 3). We performed

combined cataract surgery involving sono-phaco-aspiration (with implantation of a capsular tensing ring and intraocular lens) and partial sclero-iridocyclectomy. Six years after the operation the visual acuity is 0.6, and there are no signs of tumor relapse (Fig. 4).

The macroscopic study of resection specimen showed a fragment of iris, ciliary body, and deep scleral tissue measuring 7.5 x 5 x 2.5 mm, with a 4 x 3.5 mm whitish-yellow tumor adhered to the internal surface of the pars plicata and posterior aspect of the iris (Fig. 5).

Microscopically, the tumor consisted of columnar and cuboidal cells forming papillae, with an eosinophilic cytoplasm, hyperchromatic nuclei, occasional prominent nucleoli, and no mitotic figures. The tubular distribution of the cells was clearly manifest, even surrounding two clearly differentiated cystic spaces. Cellular staining with vimentin proved positive, with partial positivity for cytokeratin. Staining proved negative with Congo red, Alcian blue, CEA, EMA, HMB-45, NSE, S-100, synaptophysin, and smooth muscle alpha-actin. The stromal tissue component of the tumor was abundant, of myxoid appearance, with PAS-positive material and some foci of chronic inflammatory cell infiltrates. The resection margins were free of tumor cells (Figs. 6 and 7).

DISCUSSION

Adenoma of the NPCE is an amelanotic tumor presenting variable characteristics and which, in the same way as melanoma of the ciliary body, tends to manifest in adults

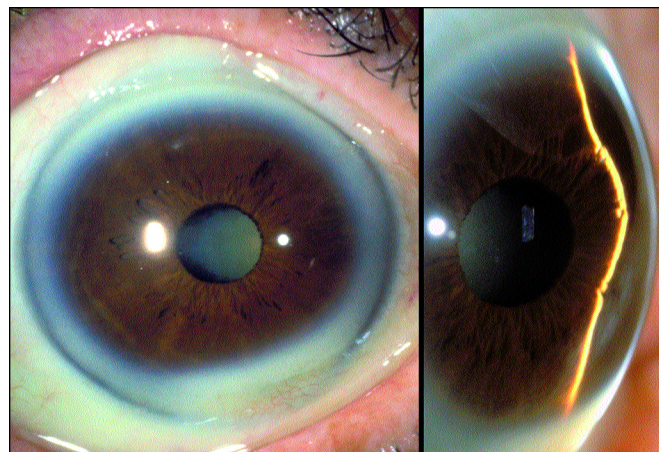


Fig. 1 - Advanced cataract and irregular papillary shape secondary to the presence of a tumor lesion in the posterior aspect of the iris and ciliary body.

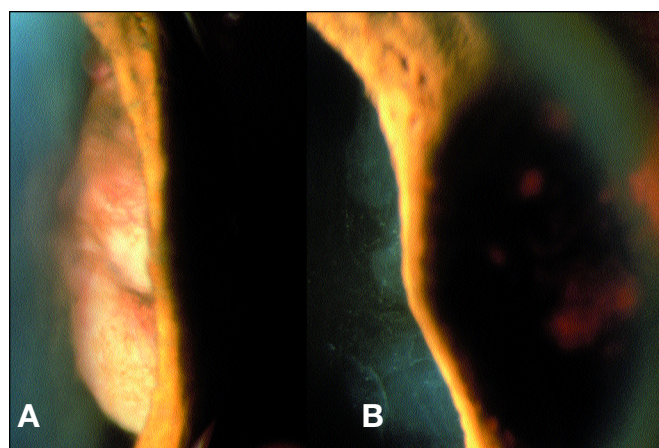


Fig. 2 - (A) (Left) Clinical appearance of the hypopigmented iridociliary tumor. (B) (Right) Tissue defects within the mass facilitating transillumination.

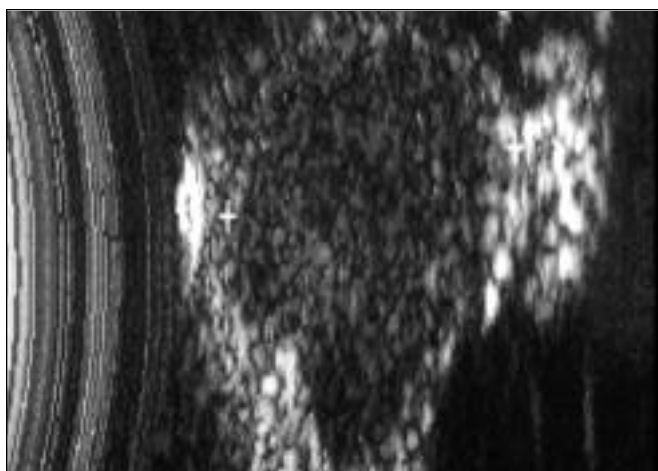


Fig. 3 - (A) B-scan showing the solidity of the ciliary body mass.

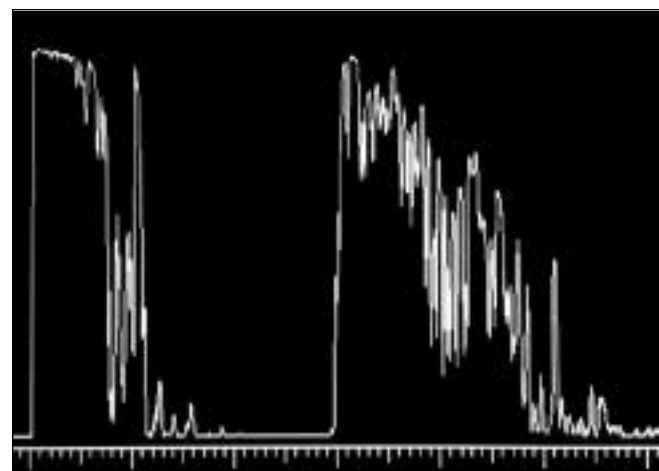


Fig. 3 - (B) A-scan ultrasound showing moderate internal reflectivity within the tumor.

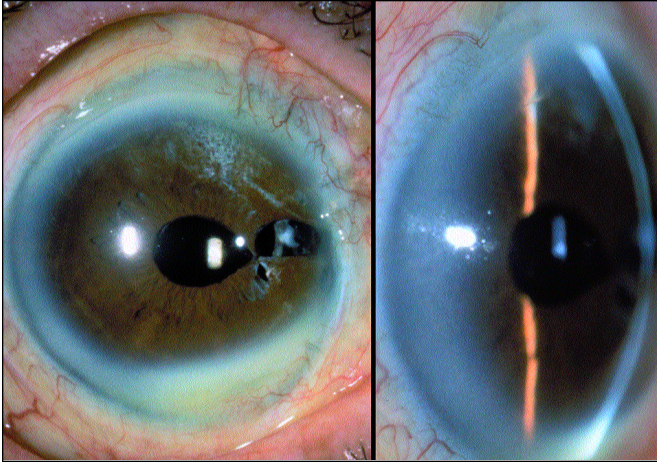


Fig. 4 - Postoperative photograph 6 years after surgery. Note the iridoplasty, some iris atrophy, and no clinical signs of tumor relapse.

(3). The course is either asymptomatic or involves a painless reduction in visual acuity, almost always related to an adjacent cataract. Tumors located at ciliary body level are pale white, with an irregular and sometimes multilobulated surface (2, 3, 8, 9). Uveal melanoma tends to be more pigmented, with a smooth surface and presenting a mushroom-like growth pattern. Some cases may present cellularity within the anterior chamber, with the presence of a sentinel vessel in the overlying episclera, though this finding is more characteristic and evident in the case of uveal melanoma (10). It is not uncommon to observe an eccentrically shaped pupil and secondary cataract formation induced by tumor compression (as in our case), and secondary lens subluxation may even occur. Unlike amelanotic melanoma, the appearance of lacunar tissue defects in the intrinsic structure of adenomas of the NPCE is quite characteristic, due to the presence of cystic spaces that facilitate transillumination during exploration (2, 10).

From the histologic perspective, adenoma of the NPCE is a solid tumor with cellular proliferation in the form of papillae and clusters that grow on the internal surface of the ciliary body, without invading the stroma (unlike in the case of melanoma) (2, 5, 10). Positivity of the tumor cells to vimentin confirms the nonpigmented ciliary epithelial origin (10). In our case we also observed immunopositivity with antibodies targeted to different cytokeratins, though this pattern tends to be highly variable. Immunoreactivity to HMB-45, which is typical of melanoma, proved negative (2).

Since adenoma of the NPCE is a slow-growing tumor, with benign cytologic characteristics and usually good patient vision, conservative management with local resec-

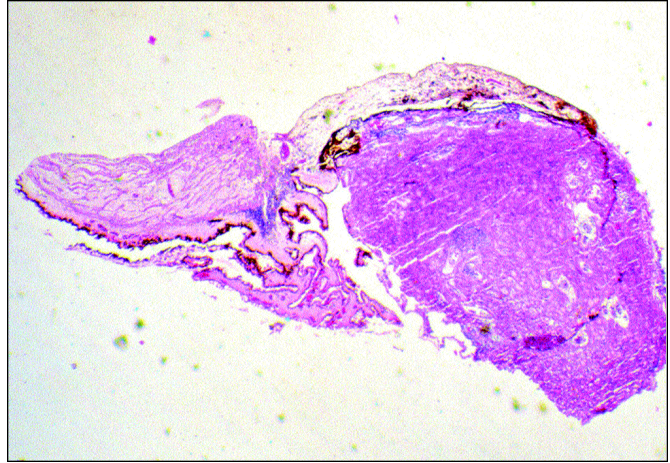


Fig. 5 - Adenoma of the nonpigmented ciliary epithelium growing from the posterior aspect of the iris and more anterior portion of the internal surface of the ciliary body (hematoxylin-eosin, 5x).

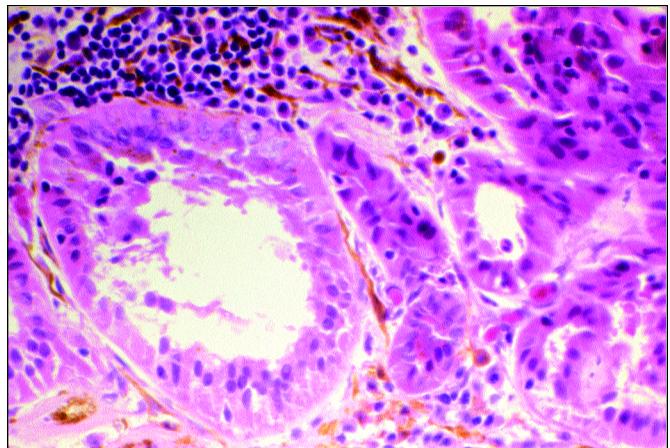


Fig. 6 - Distribution of the neoplastic cells forming papillae and clusters, and occasionally surrounding vacant cystic spaces. Note the associated chronic inflammatory infiltrate (hematoxylin-eosin, 100x).

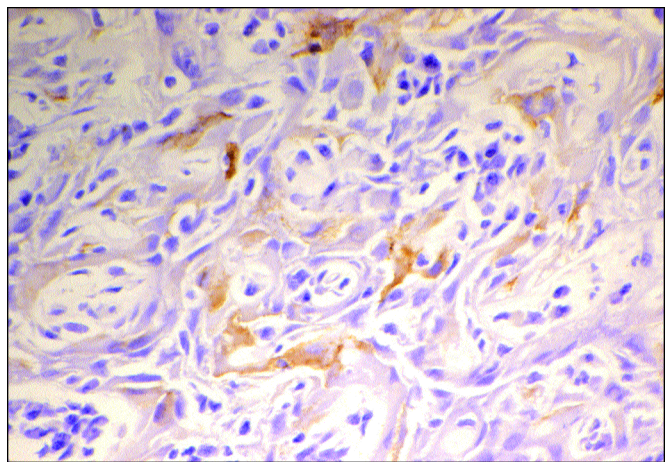


Fig. 7 - Partial positivity for cytokeratin.

tion via a partial sclero-iridocyclectomy is advised, also for diagnostic purposes. In the event of lens opacification, this procedure may be combined with small-incision cataract surgery. Usually, when the tumor is located in the ciliary body, with little or no iris involvement, it is easier to conserve most of the entire iris by commencing the resection posteriorly and then dissecting anteriorly until the anterior tumor margin appears (11). However, as we did not know exactly the posterior extension of the tumor, in our surgical procedure the flap was hinged posteriorly

and a reconstructive iridoplasty was performed at the end of the surgery.

No authors have any proprietary interest.

Reprint requests to:
J. Elizalde, MD
Ocular Oncology Service
Centro de Oftalmología Barraquer
Muntaner 314
08021 Barcelona, Spain
javierelizalde@barraquer.es

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