Short communication

Macrogenoma of the non-pigmented ciliary epithelium

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ABSTRACT

Case report: We report the clinical features and surgery of a patient with an adenoma of the non-pigmented ciliary epithelium. The adenoma measured 5 mm × 7 mm. The patient underwent radical ocular surgery consisting of partial iridociclectomy associated to lamellar sclerouvectomy.

Discussion: Adenomas of ciliary body can mimic clinically amelanotic melanomas. We present details of the patient’s medical records and review the literature. Clinically, adenoma in ciliary body can mimic amelanotic melanomas. Conservative surgery of the eye allows diagnosis and treatment, maintaining visual function.

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Macrogenoma del epitelio no pigmentado del cuerpo ciliar

RESUMEN

Caso clínico: Se describen los hallazgos clínicos y la cirugía conservadora de un paciente con un adenoma no pigmentado del cuerpo ciliar. El adenoma presentaba un tamaño de 5 × 7 mm. El paciente fue intervenido con una cirugía conservadora mediante iridociclectomía parcial asociada a esclerouvectomía lamelar.

Discusión: Los adenomas del cuerpo ciliar clínicamente pueden imitar a los melanomas amelanóticos. La cirugía conservadora del globo ocular permite realizar un diagnóstico y un tratamiento del paciente manteniendo la función visual.

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Introduction

Ciliary body adenoma derived either from the pigmented or non-pigmented epithelium (NPE) is a highly infrequent, slow growing benign tumor which usually appears in middle-aged patients without preference for either sex, although congenital cases have been described. A definitive diagnostic is through histology as clinically said adenoma can be confused with other ciliary body tumors, particularly with melanomas. We present the case of ciliary body NPE adenoma treated and diagnosed with conservative surgery, allowing adequate visual function.

Case report

Male, 71, referred for asymptomatic mass in the inferior temporal quadrant of the right eye ciliary body. Visual acuity was of 0.7 in said eye and 0.8 in the contralateral eye. The patient referred surgical bilateral aphakia dating 21 years back. Examination evidenced a grayish mass through the pupil axis with non-ingurgitated superficial vessels (Fig. 1). Gonioscopy did not reveal iridocorneal angle invasion. The lesion had high internal reflectiveness in echography and a size of 5 mm × 7 mm. Differential diagnostic was established between adenoma, adenocarcinoma, epitheloid leiomyma, granular cell tumor, epithelial reactive hyperplasia, melanoma and metastasis. A general study was made to discard primary tumor with negative result. For this reason, conservative surgery was decided for diagnostic purposes and treating the ciliary mass. Partial iridocyclectomy with lamellar sclerouvectomy was performed. Initially, the base of the tumor was defined by means of transillumination, leaving a safety margin of 5 mm around the lesion, carving a posterior base superficial scleral flap and the outlined area, increasing it anteriorly toward the corneoscleral limbus. The flap must have a width of about 80% of the sclera as it will be subsequently used for covering the defect created in the ocular wall after extracting the tumor. Subsequently, the neoplasia was resected in one piece together with the deeper sclera (Fig. 2). Thereafter, the ocular wall was closed, to which end the superficial scleral flap was placed and sutured with loose stitches to the adjacent sclera. Surgery was completed with scleral cerclage and pars plana vitrectomy, applying argon laser in the retina near the resected area together with low molecular weight silicon tamponade.

Histopathological analysis revealed that the tumor was mainly constituted by polygonal and cubic cells with abundant eosinophilic cytoplasm (Fig. 3) organized in the form of laces and separated by extracellular matrix with positive staining for periodic acid Schiff (PAS). Tissue necrosis or nuclear pleomorphism was not detected, and a low proliferative index was evidenced (Ki-76). By means of immunological techniques, tumor cells were positive for protein S-100, vimentin, and absence of positivity for HMB-45. For this reason the anatomopathological diagnostic was ciliary body EPN adenoma. Six months later additional surgery was decided to remove the silicone, obtaining a corrected visual acuity of 0.7 (+12 –4a164°). The retina remains applied to this date (24 months).

Discussion

Ciliary body NPE adenoma generally appears as a solitary, grayish and blackish unilateral mass. Normally, this adenoma courses without showing symptoms and is discovered casually. On other occasions it produce diminished visual acuity due to inducing secondary cataracts, lens dislocation and exudative retina detachment. The main problem of these lesions is that frequently they cannot be clinically distinguished from other malign tumors such as adenocarcinoma, melanocytoma and particularly ciliary body melanoma. However, a number of characteristics can assist in differentiating ciliary body adenoma from melanoma (Table 1). In this case the lesion exhibited the typical characteristics of ciliary body NPE adenoma, i.e., standalone tumor, gray color, abrupt edges, high echography reflectiveness and absence of prominent tumor vessels.

In what concerns therapeutic management, observation is recommended if the patient is asymptomatic and the adenoma diagnostic is of high certainty. However, if the patient exhibits symptoms the most adequate surgical treatment

Fig. 1 – Grayish tumor mass located in the inferior temporal with superficial vessels and iris root anterior displacement.

Fig. 2 – Surgical resection: fragments of sclera containing iris and ciliary body showing rounded grayish mass measuring 5 mm × 7 mm. The mass was resected entirely together with a broad safety margin.
Fig. 3 – (A) Histopathology of the tumor (hematoxylin–eosine stain (200×): polygonal cells or grouped in laces separated by PAS+ panels. Typically, cells are positive for S-100, vimentin and negative for HMB-45. Nuclei do not exhibit mitosis or pleomorphism. (B) Immunohistochemical staining with Ki-64 (40×) showing a very low cell proliferative index.

Table 1 – Main clinical differences between ciliary body non-pigmented epithelium adenoma and melanoma.

<table>
<thead>
<tr>
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<th>NPE adenoma</th>
<th>Melanoma</th>
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<tbody>
<tr>
<td>Color</td>
<td>Gray-black</td>
<td>Brown</td>
</tr>
<tr>
<td>Morphology</td>
<td>Abruptly elevated edges from ciliary body</td>
<td>Mushroom or pearl necklace shape</td>
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<tr>
<td>Prominent blood vessels</td>
<td>Absent</td>
<td>Present</td>
</tr>
<tr>
<td>Pigment circle surrounding the base</td>
<td>Absent</td>
<td>Frequently present</td>
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<td>Ocular echography</td>
<td>High reflectiveness and dome-shaped appearance</td>
<td>Low or medium reflectiveness and acoustic cavities</td>
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<td>Fluorescein angiography</td>
<td>Absence of double circulation</td>
<td>Typical double choroidal circulation</td>
</tr>
<tr>
<td>Transillumination</td>
<td>Dense shadow</td>
<td>Dense shadow</td>
</tr>
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</table>

NPE: non-pigmented epithelium.

is local resection with lamellar iridocyclectomy. In the case reported herein, the large size and the visual axis invasion prompted us to opt for surgical treatment. On the basis of our tumor surgery experience, we generally associate lamellar iridocyclectomy with pars plana vitrectomy with scleral cerclage and silicon tamponade in order to reduce post surgery complications (retina detachment, vitreous hemorrhage).

Conflict of interest

No conflict of interest has been declared by the authors.

REFERENCES