Medulloepithelioma of the Ciliary Body
Ultrasonographic Biomicroscopic Findings

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Intraocular medulloepithelioma is an uncommon embryogenic neoplasm of neuroepithelial origin that usually develops from the nonpigmented epithelium of the ciliary body, retina, and optic nerve. Clinically, medulloepithelioma usually appears as a tumor of the ciliary body; only rarely does it arise in the optic nerve and retina. It frequently occurs on the ciliary body of a young child as a fleshy gray or pink mass, which, on examination using slit lamp biomicroscopy, often reveals cysts. These cysts impart an irregular shape to the tumor surface. The tumor tends to be locally aggressive and can invade adjacent intraocular structures, but it rarely metastasizes. The utility of conventional echography in the diagnosis of medulloepithelioma has been established, but, to our knowledge, no reports have been published describing the ultrasonographic biomicroscopic features of medulloepithelioma. Ultrasonographic biomicroscopy (UBM) is an ideal method to analyze the anterior segment; it allows an accurate assessment of iris and ciliary body tumors, including size, tumor margins, and internal characteristics. Moreover, UBM can help with the differential diagnosis of ocular tumors. In this report, we describe the UBM characteristics of medulloepithelioma.

Case Report

A 16-year-old male patient was referred to our department with a history of redness in his right eye during the previous month. Best corrected visual acuity was 20/40 in the right eye (−1.00 × 180°) and 20/20 in the left eye. Intraocular pressure was 18 mm Hg in both eyes. Slit lamp examination of his right eye revealed temporal hyperemia and a superotemporal ciliary body brown-gray solid mass. The anterior part of this mass had a cyst-like appearance. The tumor indented the iris and touched the crystalline lens, where there appeared to be a coloboma. Fundus examination revealed a mass with an irregular surface, which reached the pars plana. Ophthalmic examination of the left eye was unremarkable.
Although the patient was in his mid teens, the clinical features were suggestive of medulloepithelioma. The clinical differential diagnosis included melanoma and leiomyoma. A UBM examination was performed to confirm the cystic component, to determine the internal characteristics and extension, and to assist with the differential diagnosis. The UBM was carried out with the patient in the supine decubitus position under topical anesthesia according to the technique described by García-Feijoó et al with the use of an orbital cup and a lid speculum. The UBM showed the existence of a highly heterogeneous reflective mass in contact with the crystalline lens and with areas of small cysts and pseudocysts in its interior (Figures 1 and 2). Stromal infiltration of the ciliary body and the iris was found, and disruption of the hyperechoic line that represents the normal pigmented epithelium of the iris and the ciliary body was seen in some areas (Figure 3). Moreover, a large oval cyst (2194 × 2416 µm in diameter) was found in the anterior area of the tumor (Figure 4). In the posterior area, the tumor had extended over the pigmented epithelium, forming a mass with offshoots extending toward the pars plana and peripheral retina (Figures 5 and 6).

The parents were offered the options of enucleation or excision of the tumor. They chose to have the tumor excised. An iridocyclochoroidectomy and a localized vitrectomy were carried out. The entire tumor was removed, and the part of the tumor that had adhered to the crystalline was detached with ease and without apparently leaving any remnants on the capsule; because of this, it was decided not to extract the crystalline. Histopathologic evaluation confirmed the tumor to be a malignant teratoid medulloepithelioma.

Postoperatively, the lens coloboma was confirmed. Best corrected visual acuity 1 month postoperatively was 20/40 in the right eye, and the situation remained stable during early-follow up. Eleven months after surgery, a severe inflammation of the vitreous occurred, causing the suspicion that the tumor had spread. Enucleation of the eye was advised.

Figure 1. Central area of the tumor. A, Radial section. B, Transverse section. Note its irregular internal reflectivity, with multiple hypoechoic areas, which correspond to cystic cavities. The small arrows delimit the tumor. CONJ indicates conjunctiva; PE, pigmented epithelium (large arrows); and SC, sclera.

Figure 2. A and B, Anterior border of the tumor, radial sections. Detail of the area of the tumor that had adhered to the crystalline lens (CRIST, arrows) is shown.

Figure 3. Central area of the tumor, oblique section. In this area, the disruption of the hyperechoic line that represents the normal pigmented epithelium is produced by a medium-to-low echogenic tumoral area. The arrows delimit the internal border of the tumor.
Medulloepithelioma is a neoplasm composed largely or in part of epithelium resembling that of the embryonic neural tube. The neuroepithelium in medulloepithelioma may differentiate toward the retinal pigment epithelium, nonpigmented and pigmented ciliary epithelia, neurons, and neuroglia. Medulloepitheliomas that contain heterologous tissues (cartilage, skeletal muscle, and brain) are classified as teratoid.\textsuperscript{1–3} Intraocular medulloepithelioma generally occurs in the first decade of life as a nonpigmented ciliary body mass. In this sense, in the series reported by Shields et al,\textsuperscript{1} the mean age at referral to their department was 4 years. Although it is usually an amelanotic, fleshy mass with an intraradial cystic component, it can occur as a pigmented solid tumor that may resemble a melanoma or a neoplasm of the pigment epithelium.\textsuperscript{2} The clinical differential diagnosis includes ciliary body melanoma, a neoplasm of the pigment epithelium of the ciliary body, melanocytoma, and an iridociliary cyst.\textsuperscript{1,2} The fact that an ultrasonographic description of this type of tumor has not been possible until now may be due to its rarity. In this particular case, the patient was older than usual and had a brown-gray solid mass; for these reasons, UBM was a very useful diagnostic tool to rule out the diagnosis of other anterior segment tumors, especially melanoma.

The findings discussed here may help in the differential diagnosis of other types of intraocular masses. The echogenic heterogeneity of the tumor, together with the notable presence of multiple cysts in the tumor, is not altogether usual in melanomas\textsuperscript{5,10} and has also been detected by conventional echography.\textsuperscript{4} Moreover, in this case, the tumor did not assume the more common spherical or regular surface form found in other tumors; rather, it was found to have stalklike prolongations. In this case, UBM not only allowed us to determine the ocular structures affected by the tumor, thereby assisting in the surgical decisions adopted, but also helped in the differential diagnosis with a ciliary body melanoma.

Figure 4. Anterior border of the tumor. A large cyst located in the area determined by slit lamp examination is shown (right arrow). A solid area of the tumor is indicated by the left arrow. AC indicates anterior chamber.

Figure 5. Posterior area of the tumor. Detail of tumoral expansion that follows the oribulociliary fibers and reaches the ora serrata is shown (white arrow). CONJ indicates conjunctiva; and EP, pigmented epithelium (black arrow).

Figure 6. A and B. Posterior area of the tumor, radial scans of the pars plana. The tumor had extended over the pigmentary epithelium. Note the offshoots extending toward the ora serrata (arrows). The internal hyporeflective cavities correspond to cysts and pseudozysts. CONJ indicates conjunctiva; OS, ora serrata; and SCL, sclera.
Naturally, the resolution and characteristics of UBM do not allow an absolutely precise or reliable diagnosis. The definitive diagnosis for any tumor should be made by a histopathologic study. However, UBM can offer us certain information, which, as in this case, when combined with an ophthalmologic examination, can lead us to the true diagnosis.

References


