Conservative Surgical Treatment of Medulloepithelioma of the Ciliary Body

Medulloepithelioma of the eye is a rare embryonic tumor. The rare published cases show that eyes with a large tumor are enucleated. Smaller tumors treated by surgery or radiotherapy frequently relapse, often requiring secondary enucleation. We describe a young girl with a large tumor of the ciliary body in which surgical resection followed by iodine 125 plaque brachytherapy allowed conservation of the eye with no recurrence after a follow-up of 7 years.

Report of a Case. A 5-year-old girl was referred for a growing tumor of the left eye. Visual acuity was 20/20 OU. Biomicroscopy showed a nonpigmented vascular mass of the inferior ciliary body with invasion of the iridocorneal angle and iris. The tumor was immediately suggestive of medulloepithelioma owing to the presence of superficial cysts (Figure 1). Staging, including magnetic resonance imaging and high-frequency ultrasonography, showed a tumor extending from the 5-o’clock position to the 7-o’clock position with invasion of the pars plana but sparing the ora serrata. The tumor had doubled in size by the time of staging. An iridocyclectomy was performed, followed by irradiation depending on the results of histological examination. Macroscopically, the ciliary body mass was a well-circumscribed, grayish tumor measuring 10 × 5 mm. Microscopically, this tumor corresponded to a proliferation of immature neuroepithelial cells forming cords and tubules in a loose stroma and displaying cystic spaces, necrotic foci, and glial tissue. The tumor cells exhibited nuclear pleomorphism, and mitotic activity was brisk (10 mitoses per high-power field). There were retinoblastoma-like areas with Flexner-Wintersteiner rosettes. There was no microscopic infiltration of the tumor’s surgical margins. On immunostaining, the neoplastic cells expressed retinoblastoma protein, vimentin, and neural markers such as CD10, CD56, and synaptophysin. They were negative for cytokeratin and S-100 protein. On ultrastructural examination, zonula adherens were found between adjacent neoplastic cells and were localized near the lumen of tubules. Cilia were observed inside the lumens. Microtubules were present in the cytoplasm of the tumor cells (Figure 2).

Owing to the presence of histological criteria of malignancy, complementary iodine 125 plaque brachytherapy was performed. Two 12-mm iodine 125 plaques were placed in the inferior limbus with the following dosimetry: 112 Gy at the tumor base, 42 Gy at 3 mm, and 25 Gy at 5 mm (to convert gray to rad, multiply by 100).

At last review following cataract surgery with a follow-up of 7 years, visual acuity was 20/25 OS and ocular examination did not show any signs of recurrence (Figure 3).

Comment. Malignant medulloepithelioma is a rare tumor in children for which no consensus concerning treatment has been reached. Published series are based on a meta-analysis of the various published cases or retrospective review of cases collected by the Armed Forces Institute of Pathology or the Ophthalmologic Institute of London. The tumor appears as a nonpigmented vascular mass arising from the ciliary body and displacing the iris or lens, which is sometimes subluxed, associated with cystic elements.

In the series reported by Canning et al, 12 patients underwent primarily enucleation and 4 patients were treated by local resection but subsequently underwent enucleation owing to recurrence. In the series by Shields et al, 4 patients initially underwent enucleation and 6 underwent iridocyclectomy. Five of the 6 underwent secondary enucleation for local recurrence. Treatment by exclusive brachytherapy has also been proposed for small tumors, with a second application in the case of recurrence.

In the case reported here, combination of local resection followed by iodine 125 brachytherapy allowed maintenance of visual acuity with no recurrence after a follow-up of 7 years.

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Mixed Tumor of the Choroid

Mixed tumors, often referred to as pleomorphic adenomas, are common tumors of salivary1 and lacrimal2 glands. Rarely these tumors occur elsewhere in the body, most commonly in the soft tissue.3,4 I describe the first reported case, to my knowledge, of an intraocular mixed tumor arising in the choroid.