Malignant Medulloepithelioma of the Ciliary Body Revealed by Neovascular Glaucoma in a 13-Year-Old Child

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ABSTRACT
Medulloepithelioma of the ciliary body is a rare congenital intraocular tumor occurring more frequently in children. It develops from the unpigmented epithelium of the ciliary body. Positive diagnosis relies essentially on anatomopathological examination. In our article, we report a case of malignant medulloepithelioma of the ciliary body in a 13-year-old child revealed by severe panuveitis complicated by neovascular glaucoma and non-axial exophthalmos.

Keywords: Child, Malignant medulloepithelioma, neovascular glaucoma.

1. INTRODUCTION
Ciliary body medulloepithelioma is a rare congenital intraocular tumor. It occurs mainly in children but does not spare adults. It develops from the unpigmented epithelium of the ciliary body. Clinical signs are varied and non-specific, leading to delayed diagnosis. Positive diagnosis relies essentially on anatomicopathological examination, which distinguishes two types: non-teratoid medulloepithelioma and teratoid medulloepithelioma, which may be malignant or benign. We report a case of malignant medulloepithelioma of the ciliary body in a 13-year-old boy whose initial presentation was atypical and whose diagnosis was confirmed histologically.

2. CASE REPORT
The case concerned a 13-year-old child, I.F., who had initially been treated for 4 years for chronic unilateral synechial granulomatous panuveitis of the right eye, complicated by neovascular glaucoma. The patient was initially put on immunosuppressants but with no clinical improvement (Fig. 1). Neovascular glaucoma was treated by retinal pan photocoagulation with intravitreal injection of anti-VEGF agents combined with oral and local hypnotizing treatment.

During his illness, the patient presented with painful non-axial exophthalmos of the right eye. Clinical examination revealed negative light perception visual acuity in the right eye, elevated ocular tone in the fingers, and irreducible, painful non-axial grade III exophthalmos on palpation, with the appearance of a subconjunctival tumour infiltrate and vascular tortuosity opposite (Figs. 2 and 3).

Examination of the anterior segment revealed corneal dystrophy with neovascular callosities and pupillary seclusion without fundus. Examination of the left eye was normal, with visual acuity of 10/10. Ocular ultrasound of the right eye revealed a heterogeneous vitreous and a partial temporal retinal detachment (Fig. 4).

A cranio-orbital CT scan was performed, showing a tumoral process measuring 33 mm in anteroposterior long axis, 15 mm in maximum transverse thickness and 11 mm in height, depending on the superior-external wall of the anterior and posterior segments, infiltrating the intraconical fat and the superior, external and inferior oculomotor muscles, in favor of a suspicious lesion with the presence of a distant medial choroidal nodule (Fig. 5).

A tumor biopsy confirmed the diagnosis of a medulloepithelioma of the ciliary body. Histological and immunohistochemical studies revealed a locally invasive tumor of the anterior ciliary body with undifferentiated cells with high mitosis. The extension work-up was negative. In view of this tumor of the ciliary body with invasion of the oculomotor muscles, exenteration was decided upon, together with courses of chemotherapy.
3. Discussion

Ciliary body medulloepithelioma is rare. It’s a congenital intraocular tumor, with fewer than 200 cases reported in the literature [1]. The disease occurs most frequently in children aged 2–5 years, with 75%–90% of cases in children under 10 [2]. It is a unilateral, mono-focal tumor with no gender or racial predominance, and no hereditary predisposition [3]. Medulloepithelioma of the ciliary body usually presents in early childhood with leukocoria and a mass arising from the ciliary body. Cataracts, ectopia, secondary angle closure, neovascular glaucoma, uveitis, uveal ectropion or vitreous hemorrhage can occur [4]. Long before a ciliary body mass becomes clinically apparent, which can cause a delay in diagnosis, as in our case. Broughton and Zimmerman [2] reviewed 56 cases and found that up to 20% were misdiagnosed and surgically treated for other conditions with procedures such as cataract surgery or filtering surgery procedures for glaucoma [5]. Immunohistochemistry is not necessary. Glial differentiation is considered a common feature, and some cells express GFAP [5]. Histologically, medulloepithelioma is made up of elements that resemble the primitive medullary epithelium of the embryonic retina [5], [6]. The presence of heteroplastic elements differentiates teratoid medulloepitheliomas from non-teratoid medulloepitheliomas [5], [6]. B-ultrasound results are often negative at the onset of the disease. It only becomes positive when masses increase and appear hyperechoic detected in the ciliary body, with clear boundaries, different shapes, irregular echo and cystic areas. Ultrasound biomicroscopy is of great importance in the diagnosis of ciliary body medulloepithelioma, as it can show the location, size and extent of tumour infiltration. However, other differential diagnoses of a ciliary mass in children, such as retinoblastoma with ciliary body infiltration, malignant melanoma, juvenile xanthogranuloma or an ocular metastatic lesion, should also be considered.

Histological study and immunohistochemistry are essential to confirm the diagnosis. Histological features of medulloepithelioma of the ciliary body include a grey or white mass with a clear boundary and variable sizes with cystic degeneration, hemorrhage or necrosis. Microscopically, the tumor mainly appears as a tube, cord or reticule. Tumor cells are cylindrical, with hyperchromatic nuclei, round or oval in appearance, and arranged in one or more layers. Tumor cells are labelled with specific antibodies and express a variety of neuroendocrine markers, such as NSE, S-100, CD56 and vimentin [7]. Tumors are classified into non-teratomatous and teratomatous medulloepitheliomas on the basis of tumor tissue composition and growth pattern. Teratomatous medulloepitheliomas predominantly contain heteroplastic foci, including brain tissue, hyaline cartilage, ependyma, ganglia, rhabdomyoblasts or striated muscle, etc. Hyaline cartilage is the most common tumor type. Hyaline cartilage is the most common type of tumor. Hyaline cartilage is the most common type of tumour [8]. Both types of medulloepithelioma can be classified as benign or malignant. The treatment of medulloepithelioma of the ciliary body is not standardized but is mainly surgical. Enucleation remains the most common treatment for most intraocular tumours; in the case of extra-scleral extension, exenteration is required [9]. Progression is generally very slow, with only local extension. Rare cases of metastasis have been described in malignant forms [10]. The visual prognosis is poor. Vital prognosis is excellent for endo-ocular forms. It may, however, be at stake when there is an orbital extension [10].
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Fig. 4. Heterogeneous vitreous with partial temporal retinal detachment.

Fig. 5. CT scan of a tumor process of the superior-external wall of the anterior and posterior segments infiltrating the intraconal fat and oculomotor muscles.

4. CONCLUSION

Malignant medulloepithelioma of the ciliary body is the most common primary malignant intraocular tumor in pediatrics, after retinoblastoma. Clinical diagnosis and treatment remain difficult, hence the importance of evoking this diagnosis in children presenting with unexplained unilateral glaucoma, uveitis, endophthalmitis, cataract or retinal detachment, with B ultrasound and/or ultrasound biomicroscopy (UBM) performed in search of an intraocular mass, notably of the ciliary body.

CONFLICT OF INTEREST

Authors declare that they do not have any conflict of interest.

REFERENCES

