

Ciliar Body Intraocular Medulloepithelioma. Case Report

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Abstract

Background: We present the case of a 10-year-old patient who underwent a diagnosis of ciliary body medulloepithelioma with subsequent enucleation.

Objective: To present the main clinical characteristics, diagnostic and therapeutic approach of a patient with a ciliary body medulloepithelioma, carrying out a review of the existing literature of this pathology.

Study Design: Case report.

Case Summary: A 10-year-old male patient presented with loss of visual acuity associated with ocular pain. Ophthalmological examination revealed a mass in the anterior chamber. A biopsy of an iris lesion was made, diagnosing a ciliary body medulloepithelioma with subsequent enucleation.

Conclusion: The case of a child with ciliary body medulloepithelioma was presented, exposing the clinical manifestations, their diagnosis and treatment, as well as the review of the existing literature.

Keywords: Medulloepithelioma; Ciliary Body; Diagnosis; Treatment; Enucleation

Introduction

Intraocular medulloepithelioma is a unilateral, very rare intraocular tumor that arises from the non-pigmented ciliary epithelium [1]. Frequently they have a similarity to the primitive spinal cord epithelium, which is why it is called that. Isolated cases have been described with tumor locations, such as the head of the optic nerve and the retina, which may come from the retinae [2] nerve stratum. In 1931, Grinker [1,3] proposed the term medulloepithelioma. Zimmerman [3] classified intraocular medulloepitheliomas in non-teratoids and teratoids [3]; Teratoid variants contain different elements such as cartilage, glial tissue and/or skeletal muscle, both with benign and malignant [3] variants. This classification shows inconsistencies and does not reflect clinical behavior. A simple and unifying classification has been proposed of progression: tumors of grade I (benign), grade II (pleomorphism, increased mitotic activity and local invasion) and grade III (transformation of the tumor to metastatic potential with extra-scleral extension or metastasis) [1].

It has an incidence of 1 in 17,000 live births. With a 1:30 to 1:50 relationship with retinoblastoma. About 100 cases of this tumor have been published [1]. The average age of onset is 3.8 to 6.8 years [1], although cases have been reported in adulthood [5]. There are no differences in gender or race or other risk factors. There is a laterality of the tumor to the right eye of 8: 2 [1]. The causes of this relationship are unknown.

It has been considered to have a non-hereditary character and without systemic association. However, it has recently been identified as a component of a tumor predisposition syndrome associated with pleuropulmonary blastoma related to the Dicer-1 mutation (trait autosomal dominant located at 14q32) [6]. Patients with this mutation are at risk of cancer of the cervix, colon, brain and others [7]. Cytogenetic abnormalities such as deletion of chromosome 16, 6q and monosomy 15 [5] have also been attributed.

They are slow-growing tumors [8]. Clinically, it presents with a decrease in visual acuity, pain, leucocoria, discoria, intraocular mass in the anterior chamber, behind the iris or ciliary body, cataract and glaucoma [9]. On average, 10 months pass between the onset of the first symptoms and the diagnosis of intraocular medulloepithelioma [1].

It can infiltrate the orbit and extend intracranially through the optic canal. The metastatic potential seems to be low. Hematogenous metastases do not constitute a significant risk while the tumor is limited to the eye. The mortality rate is up to 10% and death is usually caused by intracranial extension due to secondary local recurrence [1].

The clinical differential diagnosis is complex, since they can also be presented as masking syndromes [1].

CT and MRI are useful to differentiate medulloepithelioma from other intraocular tumors and to plan treatment. MRI is the technique of choice to rule out associated malformations or malignancies of the intracranial CNS. MRI shows hyperintensity in T1 and hypointensity in T2 [10]. On CT the non-teratoid variant is shown as a dense mass not calcified in the ciliary body region, while the teratoid appears as an irregular calcified mass [11].

Reference has now been made to the use of immunohistochemistry to establish the diagnosis. Histochemical phenotyping is performed with the streptavidin-biotin method using monoclonal antibodies. There is combined immunoreactivity to vimentin, S100, NSE (neuronal specific enolase) and CD 138 when there is retinoblastic differentiation [12].

Generally the affected eye must be removed; the main indications of enucleation are the growth of the intraocular tumor, seeding of the vitreous and drainage metastases in the posterior pole, extraocular growth with infiltration of the orbit and the appearance of a non-therapeutic secondary glaucoma. Some cases have been addressed with conservative eyeball therapy. Brachytherapy was used in some cases but this did not prevent secondary enucleation after 2.5 years [1]. Davidorf [13] published a successful case when treated with an iodine applicator 125. It has been performed brachytherapy with an applicator of Ruthenium 106, with complete remissions of the tumor, avoiding enucleation [14].

This article presents the case of a 10-year-old male patient who was diagnosed with ciliary body medulloepithelioma with subsequent enucleation, which is described with the purpose of presenting the main clinical characteristics and management of a patient with ciliary body medulloepithelioma, reviewing the existing literature on this entity.

Case Description

10-year-old male patient who consults due to a 15-day decrease in vision and pain in the right eye. To the ophthalmological examination the uncorrected visual acuity in right eye was 20/400, PH: 20/200; Biomicroscopy: Initial nasal and temporal band keratopathy, lower thick retrokeratic deposits; anterior chamber mass of inferior 50% occupying location, whitish, irregular with highly pigmented areas; posterior iris synechia, pupil in mydriasis (Figure 1), the rest of the anterior segment was not possible to assess; Anterior uveitis vs malignant tumor is diagnosed in right eye. IgG and IgM are ordered for toxoplasma, simple and contrasted skull and orbit CT and UBM. The paraclinical report and CT scan of the skull and orbit were normal. In the UBM, mass from a ciliary body extended towards the anterior chamber was evident (Figure 2). A biopsy of the lesion was reported, reporting medulloepithelioma (Figure 3). Subsequently, the patient underwent enucleation. No extraocular extension of the tumor was found. Therefore, systemic chemotherapy and/or orbital irradiation were not indicated after enucleation.

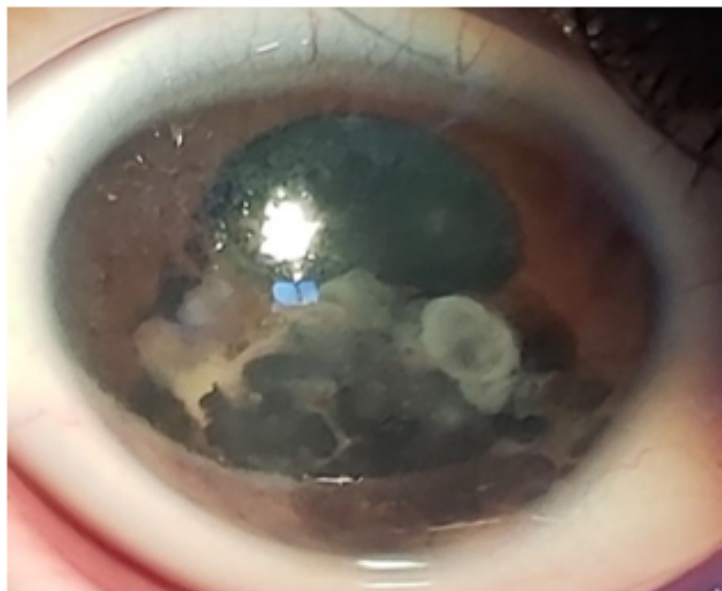


Figure 1: Anterior chamber mass of 50% lower occupying location, irregular with hypo and hyperpigmented areas.

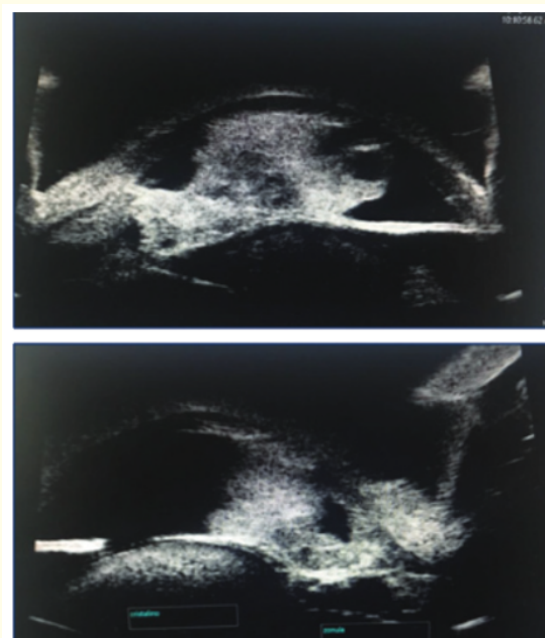


Figure 2: The UBM shows cavitated mass that comes from the ciliary body extended into anterior chamber, in contact with corneal endothelium, commitment of iridian stroma is observed.

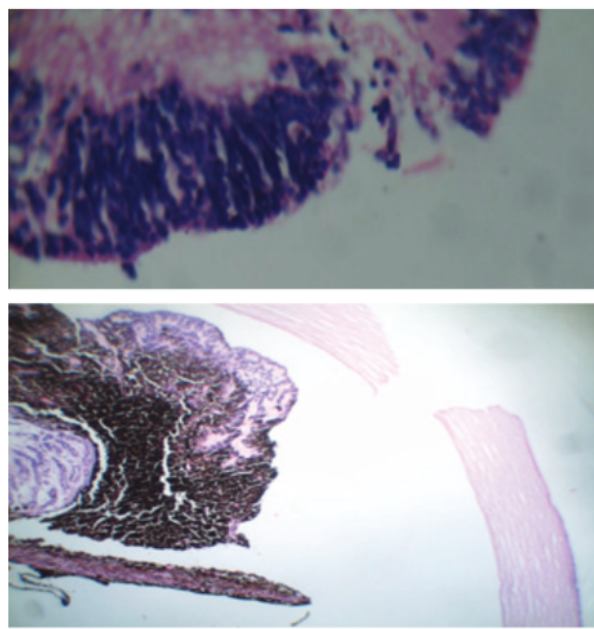


Figure 3: Biopsy of pre-enucleation lesion where tumor of the iris and ciliary body formed by tubules, cords and rosettes of polarized neuroectodermal cells that resemble spinal cord epithelium.

Discussion

Intraocular medulloepithelioma is a rare, unilateral, congenital tumor that appears in childhood. It originates from the non-pigmented epithelium of the ciliary body. The patient in the reported clinical case has 10 years old, its presentation was unilateral, which fits with the publications found. There is greater laterality for the right eye, as observed in this patient.

Although benign and malignant variants are recognized independently of teratoid and non-teratoid forms, all medulloepitheliomas should be considered as potentially malignant tumors that would have been more clearly reflected in a classification scheme than in a dichotomy in two types. This patient was approached with that concept clear.

The diagnosis is usually made only after a long latency, after the appearance of the first symptoms, although in this patient the diagnosis was early suspected and confirmed histopathologically. They can manifest as pseudo-uveitis, within masking syndromes, as happened in this case.

Computed tomography, magnetic resonance imaging and UBM are useful for diagnosis. In our case, the CT scan of the skull and simple and contrasted orbit were normal, showing the mass from the ciliary body extended towards the anterior chamber through the UBM.

Currently, immunohistochemistry of the lesion is also used, which was not practiced in this patient.

Metastases are rare; in this case no extraocular extension of the tumor was found. Therefore, systemic chemotherapy and/or orbital irradiation were not indicated after surgical treatment.

As for treatment, sometimes the eyeball can be preserved with brachytherapy and Ruthenium, however, most patients require enucleation, as happened in this case.

Conclusion

In conclusion, intraocular medulloepithelioma is an extremely rare congenital tumor with unilateral involvement that has its origin in the non-pigmented epithelium of the ciliary body. Its diagnosis is usually delayed, therefore it should be suspected in a patient with pseudo-uveitis associated with possible intraocular tumor. The UBM is useful to identify the tumor, as well as the biopsy of the lesion to characterize it. Generally, the treatment consists of enucleation.

Recommendations

Although intraocular medulloepithelioma is an extremely rare tumor, it should be suspected in a patient who presents findings compatible with pseudo-uveitis associated with possible intraocular tumor of unilateral involvement, thus avoiding delaying diagnosis and subsequent treatment. The UBM, CT and MRI are useful to identify the tumor, as well as the biopsy of the lesion and immunohistochemistry to characterize it and offer the required treatment.

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