

## Strabismus Revealing Ciliary Body Medulloepithelioma: A Case Report and Literature Review

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### Abstract

### Case Report

Ciliary body medulloepithelioma is rare congenital tumor. It develops from the non-pigmented ciliary body epithelium. It occurs mainly in the first decade of life. The clinical examination of the ciliary body is difficult that leads to a significant delay in diagnosis, until enlargement of the tumor, which produces secondary effects. The exact diagnosis is only made on histopathologic examination that found elements to differentiate between medulloepithelioma teratoid and non-teratoid, benign or malignant ones. Enucleation is the preferred method of treatment for advanced intraocular mass. Ciliary body medulloepithelioma have an excellent prognosis. The authors propose to describe a case of this rare tumor revealed by strabismus.

**Keywords:** Medulloepithelioma, ciliary body, strabismus, enucleation.

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## INTRODUCTION

Medulloepithelioma is a rare congenital tumor of the ciliary body. It develops from the non-pigmented ciliary body epithelium [1]. It occurs mainly in children, but can rarely detect in adult [2]. Clinical signs vary and nonspecific, hence the diagnostic delay [3]. The diagnosis is mainly based on the histological examination, which distinguishes two types: non-teratoid medulloepithelioma and medulloepithelioma teratoid, which can be benign or malignant [4].

## OBSERVATION

A 20-month-old girl, with strabismus of the left eye that appeared 3 months ago associated with red eye (Figure1). She had no pathological history, including no notion of eye trauma or fever. The ophthalmologic examination of the left eye found convergent strabismus with conjunctival hyperemia, a non-reactive pupil with 360 synechiae, and total cataract preventing visualization of the posterior pole, intraocular pressure was at 8 mmHg (Figure 2).

The eye examination of the right eye did not show any abnormality. The general examination was normal.

The ocular ultrasound objectified a small eyeball with an intraocular hyperechoic process (Figure3). Computed tomography scan showed an intraocular mass, with heterogeneous enhancement, measuring 15x12mm, irregular, dense, causing deformation of the eyeball, no calcifications were observed (Figure 4). An inflammatory and infectious assessment (toxoplasmosis serology, toxocarosis, syphilis, lyme) were entirely normal. Radiological assessment of extraocular extension has returned to normal.

Due to age and intraocular tumor syndrome the diagnosis of retinoblastoma has not been completely ruled out. Enucleation was performed. The histological analysis revealed a Ciliary Body medulloepithelioma with Ki67 estimated at 2%. The evolution was favorable without any local or general recurrence with a follow-up of 1 year (Figure 5).



Fig-1: Clinical Picture shows a strabismus of the left eye



Fig-2: The ophthalmologic examination shows a non-reactive pupil with 360 synechiae, and total cataract

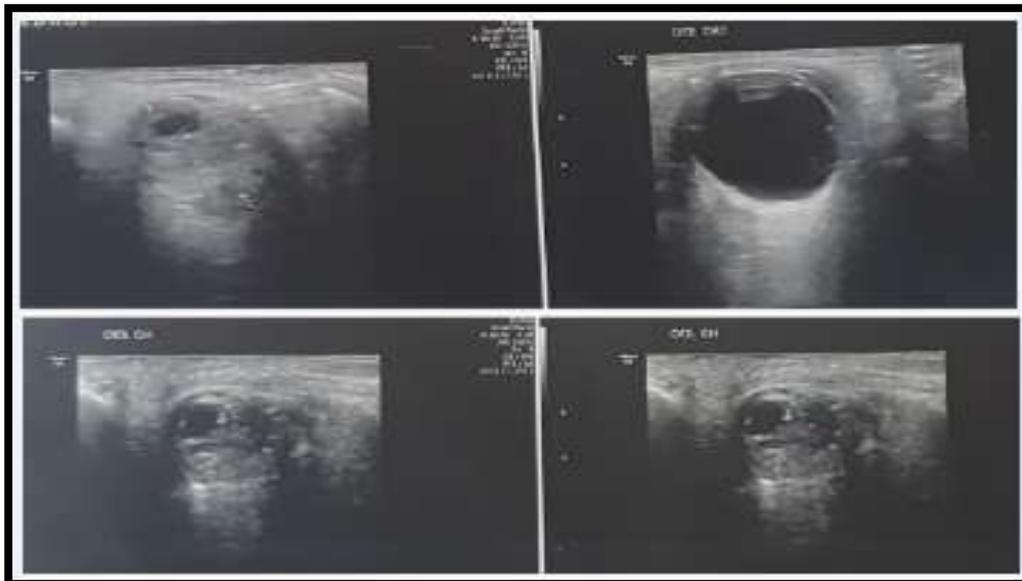


Fig-4: Ocular ultrasound objectified a small eyeball with an intraocular hyperechoic process

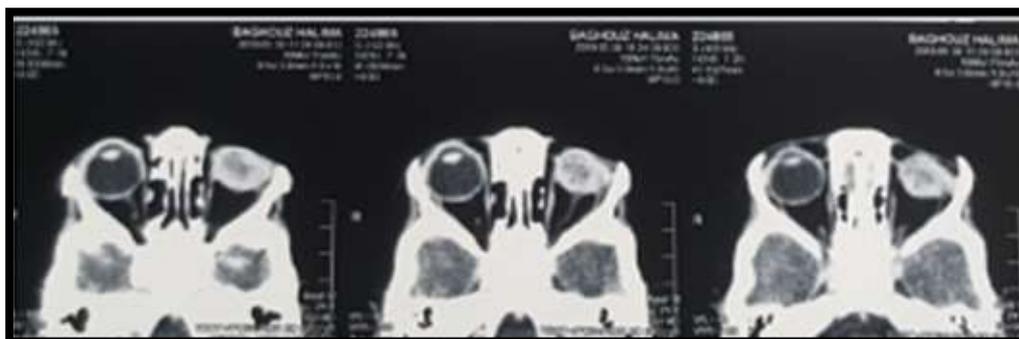


Fig-4: CT scan objectified an intraocular mass, causing deformation of the eyeball, with no calcifications



**Fig-5: Prosthetic rehabilitation of the ocular defect after enucleation**

## DISCUSSION

First report of medulloepithelioma was in 1892 by Badel and Lagrange, it was named carcinoma primitif [5]. Verhoeff coined the term of Teratoneuroma in 1904 due to the histologic detail of this uncommon tumor [6]. Later Fuchs in 1908 renamed it diktyoma [7]. The name of medulloepithelioma was proposed by Grinker in 1931 [8] considering the histological resemblance of the tumor to the neuroepithelium of the embryonic neural tube [9]. This was based on the work on a rarer form of medulloepithelioma of the brain described by Bailey and Cushing [10, 11].

According to a large series by Broughton and Zimmerman [2], the median age at the time of initial clinical manifestation was 3.8 years. But, the median age at the time of surgery and histopathologic diagnosis was 5 years. The peak incidence occurs in the first decade of life. However, it may occasionally present in adults [12-14]. There is no predisposition, race, sex or inheritance [2, 15].

The difficulty in the direct visualizing of the ciliary body often delays the diagnosis of small tumors, until enlargement which produces secondary effects. The most common clinical presentation includes unilateral glaucoma and cataract with or without lens subluxation. In a study of 41 cases, glaucoma was present in 44% and cataract was present in 46% [15]. Which give the following symptoms: loss of vision, leukocoria, pain, or red eye. Other unusual presentations include strabismus [11].

On clinical examination, the mass of the ciliary body is usually appears irregular grey-white to pink [11]. Characteristic clear cysts can be evident in the mass on slit-lamp biomicroscopy in 60%, such cysts can break free from the main tumor and float into the vitreous and aqueous [16]. It can rarely occur as a pigmented solid tumor [17]. The globe can be buphtalm or microphtalm [18]. Some vessels running the surface of the tumor can be shown. Other presentations include uveitis, hyphema, retinal detachment, vitreous hemorrhage, invasion of the optic nerve, and rarely extraocular extension of the tumor [11].

Ultrasound and computed tomography scan confirm the tumor diagnosis, but cannot differentiate neither between different tumors of the ciliary body nor with the anterior retinoblastoma. Magnetic resonance imaging eliminates non-tumor causes, in particular coats disease and persistent hyperplastic primary vitreous, the medulloepithelioma gives a T2 hypersignal and a T1 hyposignal. The exact diagnosis is only made on histopathological examination [3].

Histologically, medulloepithelioma is made up of elements which resemble the primitive medullary epithelium of the embryonic retina. The presence of heteroplastic elements helps to differentiate medulloepitheliomas teratoids and non-teratoid medulloepithelioma. Nonteratoid medulloepithelioma consists of a pure primitive medullary epithelium proliferation [19]. Whereas teratoid medulloepithelioma harbors heteroplastic elements, including hyaline cartilage, rhabdomyoblasts or striated muscle, or brain-like tissue in addition to the proliferation of the neuroepithelial elements. The criteria for malignancy include: [15].

- The presence of areas of poorly differentiated neuroblastic cells resembling those of retinoblastoma with or without rosettes
- Sarcomatous areas resembling chondrosarcoma, rhabdomyosarcoma, or embryonal sarcoma
- Nuclear pleomorphism, and high mitotic activity
- Invasion of the uvea, cornea, or sclera with or without extrascleral extension.

Management depends on the size of the tumor. The treatment options for small tumors include cryotherapy, local resection, plaque radiotherapy, external beam radiotherapy.

Local resection can be used for small circumscribed tumors, but it is reported to have up to 50% recurrence rate [15]. Enucleation is the preferred method of treatment for advanced intraocular medulloepithelioma with large tumors and neovascular glaucoma [11]. The role of systemic chemotherapy in the treatment of ciliary body medulloepithelioma is not well established [15]. Ciliary body medulloepithelioma have an excellent prognosis, with a 5-year survival of 90-95% after enucleation, unless there is extraocular extension or central nervous system involvement [11].

## CONCLUSION

The medulloepithelioma of ciliary body is a rare tumor, often diagnosed at an advanced stage of progression. Some clinical presentations in children should drive to a total eye examination especially the ciliary body, in order to enable an earlier detection and avoid the morbidity of enucleation.

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