

## Ciliary Body Medulloepithelioma in an Adult

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**Abstract:** Ciliary body medulloepithelioma is predominantly a pediatric tumor. Incidence of ciliary body tumor in adults is very rare. We report a case of ciliary body medulloepithelioma in an adult. This case emphasizes the need to include in the differential diagnosis of ciliary body tumors in adult. A 53-year-old gentleman presented with complaints of poor vision in the left eye of 2 months duration. He underwent posterior vitrectomy for retinal detachment and uncomplicated cataract surgery in the same eye 10 years ago. He had only hand motion vision on affected left eye with an intraocular pressure of 28 mmHg. There were dilated episcleral vessels on superotemporal, whitish spherical cyst inferiorly in anterior chamber. There were also whitish cystic like mass seen at the vitreous cavity posterior to intraocular lens, not permitting the fundus view. Histopathological examination revealed an epithelial tumor arising from non-pigmented ciliary body epithelium consistent with a benign medulloepithelioma.

**Keywords:** adult, ciliary body tumor; medulloepithelioma

### INTRODUCTION

Medulloepithelioma is an embryonal tumor originated from primitive medullary epithelium along inner layer of optic cup [1,2]. It is a rare intraocular tumor of which 78% manifesting in first decade of life with the median age at presentation was 5-year old[3]. Medulloepithelioma in adult is rare with isolated case reports found in literature. Herein, we report a case of an adult with ciliary body medulloepithelioma.

### CASE REPORT

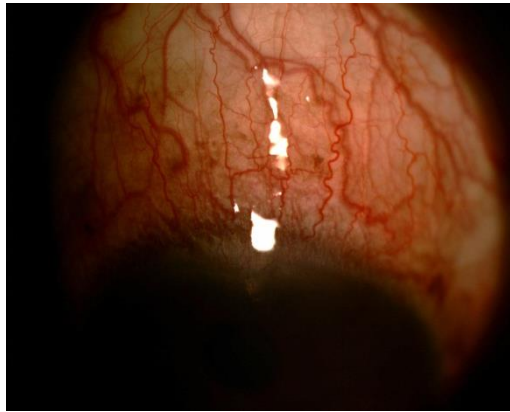
A 53-year-old gentleman presented with complaints of poor vision in the left eye of 2 months duration. He underwent posterior vitrectomy for retinal detachment and uncomplicated cataract surgery in the same eye 10 years ago. He had only hand motion vision on affected left eye with an intraocular pressure of 28 mmHg. There was no eye pain. He was a frequent traveler who used to travel within the country. He did not have history of consuming raw and exotic food. He also reported swelling over right axillary region began 5 years prior to his presentation which slowly increase in size. No other constitutional symptoms reported.

On examination of the left eye, there were dilated episcleral vessels on superotemporal quadrant. Whitish spherical cyst was seen inferiorly in anterior chamber. Anterior chamber depth was shallower on the temporal aspect resulted from anteriorly bulging iris. No iris neovascularization seen. Pupil was round and reactive. No relative afferent pupillary defect. Intraocular lens was stable in the bag however there were whitish cystic like mass seen at the vitreous cavity posterior to intraocular lens, not permitting the fundus view. Right eye was normal.

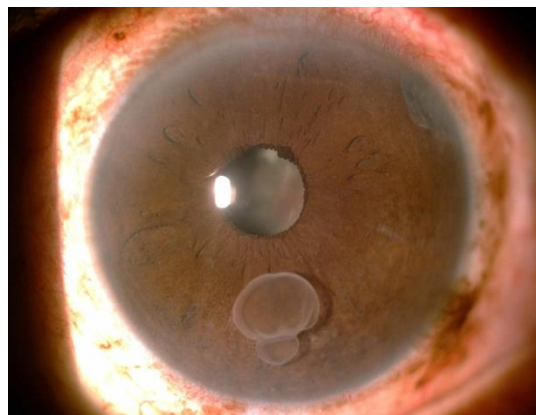
Ultrasonography revealed homogenous mass with moderate reflectivity arising from ciliary body with few free-floating cysts in the vitreal cavity which moves upon movement of the eyeball. There was nevertheless no hyperechoic within the cyst to suggest of a scolex. Systemic examinations revealed a clinically benign cystic mass which was well circumscribed at right axillary region measured about 5 cm in length x 4 cm in width. Other examinations were unremarkable. Our leading diagnosis was ocular cysticercosis. Excision and biopsy of the mass reported as sebaceous cyst. Magnetic resonance imaging of the orbit showed homogeneously enhancing lobulated intra-ocular soft tissue lesion, appear hyperintense on T1 weighted and hypointense on T2 weighted. Magnetic resonance imaging of the brain was normal. Blood investigations and computed tomography did not demonstrate any primary lesion of malignancy. Cysticercosis antibody serology was negative.

He underwent fine needle aspiration biopsy of the lesion. The cyst in the anterior chamber was also sent for histopathology test. Histopathological examination revealed an epithelial tumor arising from non-pigmented ciliary body epithelium consistent with a benign medulloepithelioma. He was informed

regarding the diagnosis and was called for discussion on the treatment plan however he defaulted.



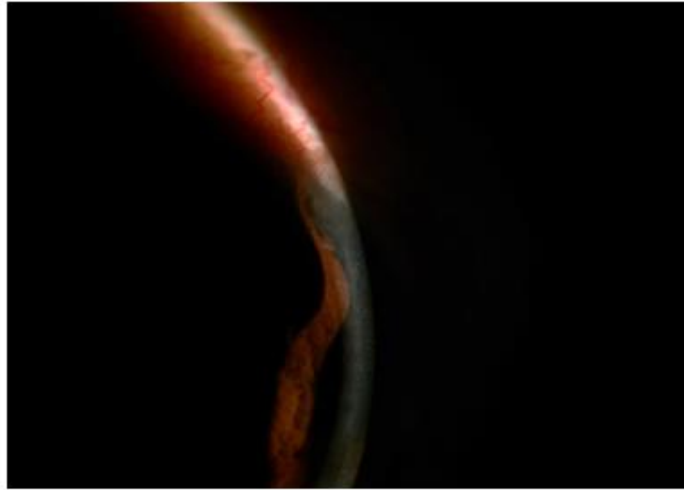
**Fig-1: Prominent episcleral vessels at superotemporal quadrant**



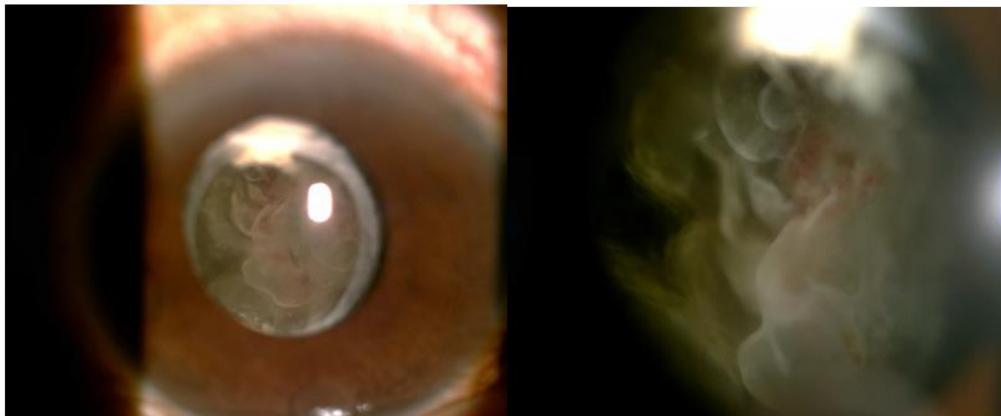
**Fig-2: Free floating cyst in anterior chamber**



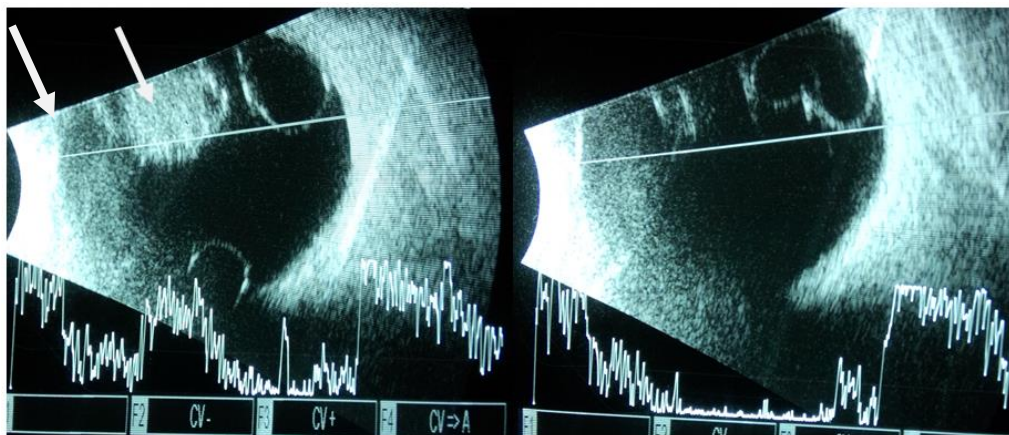
**Fig-3: Magnified image of free floating cyst in anterior chamber**



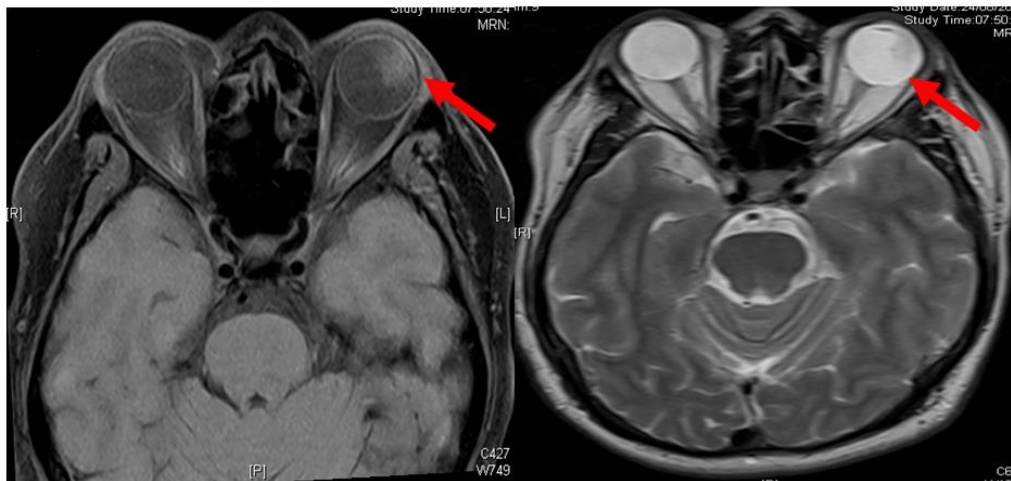
**Fig-4:** anterior bulging of the iris, shallower anterior chamber depth



**Fig-5a:** Cystic mass posterior to the intraocular lens b) grayish cystic mass



**Fig-6:** Ultrasonography showed few free floating cyst moves with eye movement, arrow showing hyperechoic well circumscribed mass posterior to the lens



**Fig-7: MRI T1 weighted showed hyperintense mass Fig-8: MRI T2 weighted hypointense**

## DISCUSSION

Medulloepithelioma is very rare in adult. The first histological description was by Verhoeff in 1904[1]. He described the findings as ‘terato-neuroma’ although he did not illustrate features of teratoma. Later in 1908, Fuchs introduced the term ‘diktyoma’ originated from Greek word to describe net-like pattern of poorly differentiated neuroepithelial cells [2]. Medulloepithelioma was coined by Grinker in 1931 which gain its popularity and being used till today[3]. Histopathologically, medulloepithelioma can be categorised as teratoid and nonteratoid [4,7,8]. Both categories are further subclassified into malignant or benign. Nonteratoid type contains well differentiated cells of non-pigmented ciliary epithelium, whereas teratoid variety featured heterologous elements such as brain, cartilage and skeletal muscle. Features of malignant medulloepithelioma include presence of areas with poorly differentiated neuroblastic cells, increase mitotic figures or nuclear pleomorphism, sarcomatous area and invasion of other ocular tissue with or without extraocular extension. In consonance with the criteria described by Broughton and Zimmerman, our case can be classified as nonteratoid benign medulloepithelioma. We identified 15 adults’ cases in the literature. All except 3 were malignant type.

Most commonly the presentation includes poor vision, pain and leucocoria [7,10]. Other clinical findings were mass in the iris, ciliary body or anterior chamber with variable sizes. Generally mass appear grayish white to fleshy pink. Other predominant clinical features are glaucoma and cataract. Pigmented intraocular mass especially when occurs in adult may closely resemble malignant melanoma for which patients underwent enucleation [9, 11, 12]. Classical and very rarely cyst in anterior and posterior chamber predominates the clinical findings which can be mistaken as parasitic infection as exhibited by our patient [7,10]. A histopathology result of our patient was also unexpected.

MRI findings demonstrated hyperintense lesion on T1 weighted and hypointense in T2 weighted which mimic the radiological findings seen in malignant melanoma. Ali et al reported an elderly woman with surprising histological diagnosis of medulloepithelioma whom clinical presentation suggested of ciliary body metastasis. Other clinical findings include non-pigmentary ciliary body mass, secondary neovascular glaucoma, lens notch or subluxation and neoplastic cyclitic membrane [4].

Local resection, enucleation and radiotherapy are among the management options for medulloepithelioma. Local resection by means of iridocyclectomy can be done in a small tumor less than 3 clock hours. However, it has been found that recurrence after local resection is common with ultimately enucleation is required [8, 10, 13, 14].

## CONCLUSION

Ciliary body medulloepithelioma is predominantly a paediatric tumor. This case emphasizes the need to include in the differential diagnosis of ciliary body tumors in adult.

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