

Analysis of clinical and pathological features of ciliary body medulloepithelioma

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Abstract

• **AIM:** To analyze and summarize the clinical and pathological features of ciliary body medulloepithelioma.

• **METHODS:** The clinical and pathological data of 11 patients (11 eyes) who were diagnosed with ciliary body medulloepithelioma at Beijing Tongren Hospital, Capital Medical University, from 2007 to 2021 were retrospectively analyzed.

• **RESULTS:** The initial symptoms of 11 patients included vision loss (6 eyes), atrophía bulbi (1 eye), proptosis (2 eyes), and leukocoria (2 eyes). Most patients suffered with corneal opacity, anterior chamber flare and hyphema. Iris neovascularization and synechia, complicated cataract, and secondary glaucoma occurred in several cases. Three patients even had lens subluxation and retinal detachment. B-scan ultrasonography showed vitreous opacity and a medium-high uneven echo mass in the eyeball. Ultrasound biomicroscopy examination showed a spherical or hemispherical ciliary body mass with uneven internal echoes and irregular cystic spaces. All of the 11 patients were diagnosed with malignant ciliary body medulloepithelioma by pathological evidence. In this study, 6 patients had enucleation (2 patients had systemic chemotherapy after surgery), and the other 5 patients had local tumor resection (1 patient had plaque radiotherapy after surgery).

• **CONCLUSION:** Ciliary body medulloepithelioma is a rare intraocular tumor and may be easily confused with retinoblastoma. Analyzing the clinical and pathological features of ciliary body medulloepithelioma is useful to further understand ciliary body medulloepithelioma, and can make an accurate diagnosis and better treatment.

• **KEYWORDS:** medulloepithelioma; ciliary body; intraocular tumor; retrospective study

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INTRODUCTION

Ciliary body medulloepithelioma mainly originates from the nonpigmented ciliary epithelium, but rarely in the retina and optic nerve. Ciliary body medulloepithelioma mostly occurs in children and is unilateral. The initial symptoms often present as vision loss, eye pain and leukocoria, which are similar to other intraocular tumors, such as retinoblastoma. Therefore, the differential diagnosis of ciliary body medulloepithelioma is difficult. Furthermore, due to the occult location of ciliary body medulloepithelioma, it is difficult to detect at the early stage.

Ciliary body medulloepithelioma is rare and only 14 cases have been reported in China in the past 10y^[1-6]. Sufficient analysis of the clinical and pathological features of ciliary body medulloepithelioma is useful for the accurate diagnosis and better treatment of this disease. Therefore, in order to further understand ciliary body medulloepithelioma, we retrospectively analyzed the clinical and pathological data of the 11 patients with ciliary body medulloepithelioma who had received surgery and histopathological examination in Beijing Tongren Hospital, Capital Medical University from 2007 to 2021.

SUBJECTS AND METHODS

Ethical Approval This study was approved by the local Ethical Committee of Beijing Tongren Hospital, Capital Medical University. Written consents were obtained from the parents of each child.

A total of 11 patients (11 eyes) with ciliary body medulloepithelioma were recruited in this study. All the patients underwent surgery and histopathological examination at the Department of Ophthalmology, Beijing Tongren Hospital, Capital Medical University from January 2007 to December 2021. The clinical and pathological data of the 11 patients were collected and analyzed retrospectively in terms of their symptoms and signs, imaging examinations, treatments and histopathological results. The recorded general data included age, gender, affected eye side, course of the disease, initial symptoms and history of treatments. The best corrected visual acuity (BCVA) of patients over 3 years old was obtained by using the international standard visual acuity chart and subjective refraction. Using a non-contact tonometer or finger-pressing method to measure intraocular pressure (IOP).

RESULTS

General Data Seven males and four females were included in the research. Besides, 5 cases occurred in the right eye and 6 cases in the left eye. The median age was 9y (range from 2 to 56y). Most patients were adolescents and children, with 10 patients under 18y and 8 patients under 12y. The course of the disease ranged from 1mo to 5y. Some patients were treated primarily at the Beijing Tongren Hospital (cases No.1, 2, 3, 6, 8, 10, and 11), while others had been treated elsewhere and then referred (cases No.4, 5, 7, and 9). Three patients (cases No.1, 2, and 7) had been treated with cyclophotocoagulation for secondary glaucoma. In addition, case No.2 also had received lens extraction and ciliary body mass resection. Case No.7 had cataract extraction for a complicated cataract before the visit. Case No.4 had local resection of ciliary body mass 3 months ago. Case No.5 had undergone ciliary body tumor resection 2 years ago, but relapsed 2mo before the visit. Plaque radiotherapy was performed for case No.9 before 3y (Table 1).

Clinical Features All of the 11 patients had a unilateral tumor. The initial symptom was vision loss in 6 cases, eyeball size change in 3 cases, and leukocoria in 2 cases. All the patients had poor visual acuity and even decreased to no light perception (Table 1). Results of ophthalmic examinations showed most patients had conjunctival congestion and corneal opacity. When the tumor occupies part or all of the anterior chamber, the anterior chamber becomes shallower or even disappears, even with anterior chamber exudation and hyphema, which increase the IOP and eventually lead to secondary glaucoma. Most patients presented with localized iris bombe, unclear iris texture or iris atrophy, exudative membranes, neovascularization on the iris surface, and anterior or posterior synechia of iris. After mydriasis, we found that cataract occurred in the majority of patients, and there was one patient who had lens subluxation (case No.8). The ocular fundus could be seen in two patients with retinal detachment

Table 1 General data of the 11 patients with ciliary body medulloepithelioma

Case	Age (y)	Gender	Affected eye side	Course of disease (mo)	BCVA	IOP (mmHg)	Initial symptoms	Treatments	History of treatments
1	4	M	OS	1	LP	T ₁	Conjunctival tumor/proptosis	Enucleation, systemic chemotherapy	Cyclophotocoagulation
2	5	M	OS	3	HM	7	Atrophia bulbi	Enucleation, systemic chemotherapy	Cyclophotocoagulation (2 times), lens extraction, ciliary body mass resection
3	9	M	OD	12	0.1	24	Vision loss	Enucleation	None
4	17	F	OD	60	NLP	8	Vision loss	Enucleation	Ciliary body mass resection
5	12	F	OS	24	LP	38	Vision loss	Enucleation	Ciliary body mass resection
6	4	M	OS	1	0.05	14	Vision loss	Local resection, plaque radiotherapy	None
7	12	F	OD	60	HM	25	Leukocoria	Local resection	Cyclophotocoagulation, cataract extraction
8	2	M	OS	2	Uncooperative	T _n	Leukocoria	Local resection	None
9	56	F	OS	36	NLP	48	Vision loss	Enucleation	Plaque radiotherapy
10	2	M	OD	3	Uncooperative	T _n	Proptosis	Local resection	None
11	13	M	OD	12	0.01	T _n	Vision loss	Local resection	None

M: Male; F: Female; OS: Left eye; OD: Right eye; LP: Light perception; NLP: No light perception; HM: Hand motions; BCVA: Best corrected visual acuity; IOP: Intraocular pressure; T₁: Moderately elevated intraocular pressure; T_n: Normal intraocular pressure.

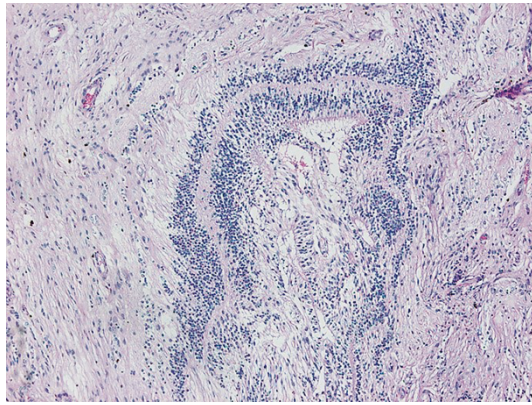


Figure 1 Histopathologic image of ciliary body medulloepithelioma by hematoxylin-eosin staining.

(cases No.9 and 11), and the other patients' fundus were invisible (Table 2).

Imaging Examinations All the patients underwent B-scan ultrasonography and ultrasound biomicroscopy (UBM) examinations. However, we only collected the notes from Beijing Tongren Hospital, and a few patients' original notes from other hospitals were lost, leading to incomplete imaging information. The results of B-scan ultrasonography showed vitreous opacity and a medium-high uneven echo mass in the eyeball. Besides, the secondary retinal detachment was found in cases No.9 and 11. UBM examination showed a spherical or hemispherical ciliary body mass with uneven internal echoes and irregular cystic spaces. Furthermore, three patients who took a computed tomography (CT) scan showed a high-density tumor in the eyeball, and 5 patients who took a magnetic resonance imaging (MRI) scan found that the tumor was moderately or slightly high signal intensity on the T1 signal.

Operation and Post-operation Treatments Six patients had enucleation and two of them (cases No.1 and 2) had systemic chemotherapy after surgery. The other 5 patients had local resection of ciliary body tumor and 1 patient (case No.6) had plaque radiotherapy after surgery.

Histopathological Data Tumors of 11 patients were examined by histopathology, and the results of all patients suggested malignant ciliary body medulloepithelioma. Especially, case No.2 was accompanied by retinoblastoma. Microphotograph hematoxylin-eosin staining (H&E) revealed tumor tissues mostly arranged in tubular, cordlike, or reticular patterns in a myxoid stroma. Observed under high magnification, the tumor cells have a columnar or polygonal, round or oval appearance, with hyperchromatic nuclei, and some pathological mitotic figures could be seen (Figure 1). Furthermore, we detected the expression of neuron specific enolase (NSE), glial fibrillary acidic protein (GFAP), epithelial membrane antigen (EMA), S-100, Bcl-2, vimentin, synaptophysin (Syn), CD56, CD99, cytokeratin (CK), CD38, CD20, CD45,

Table 2 Clinical and pathological data of the 11 patients with ciliary body medulloepithelioma

Case	Symptoms and signs							Imaging examinations (B-scan/UBM/CT/MRI)	Pathology	Positive immunohistochemistry markers
	Conjunctiva	Cornea	Anterior chamber	Iris	Pupil	Lens	Retina			
1	5x5 mm ² mass	Opacity	Exudation	Exudative membrane, neovascularization, synechia	Synechia	Cataract	-	Medium-high uneven echo, ciliary body cystic mass	Malignant medulloepithelioma (invasion of conjunctiva)	NSE, S100, GFAP, Bcl-2, Ki67(+)
2	Normal	Opacity	Shallow, flare	Iris atrophy	Mydriasis	Lens coloboma	-	Ciliary body mass	Malignant medulloepithelioma, retinoblastoma	Vimentin, CD56, Syn, Ki67 (40%)
3	Normal	Opacity	Shallow, cystic mass	Neovascularization, iris bombe	Corectopia	Cataract	-	Uneven echoes, ciliary body cystic mass, 8.7x7.6x7.3 mm ³	Malignant medulloepithelioma	NSE, S100, vimentin, CD56, Ki67 (40%)
4	Congestion	Normal	Shallow, flare	Iris atrophy, synechia	Synechia	Cataract	-	-	Malignant medulloepithelioma	NSE, S100, GFAP, Bcl-2, vimentin, CD56, Syn, Ki67 (70%)
5	Congestion	Opacity	Shallow, flare, hyphema	Neovascularization, synechia	Synechia, mydriasis	Cataract	-	Uneven echoes, ciliary body cystic mass, 5x7 mm ²	Malignant medulloepithelioma	NSE, GFAP, vimentin, Syn, Ki67 (50%)
6	Normal	Opacity	Shallow	Neovascularization, iris bombe	Normal	Cataract	-	Ciliary body cystic mass	Malignant medulloepithelioma	NSE, S100, GFAP, CD56, CD99, Ki67 (40%)
7	Normal	Opacity	Shallow	Synechia	Synechia	Pseudophakia	-	Medium-high uneven echo, ciliary body cystic mass, 14.48x10.77x4.36 mm ³	Malignant medulloepithelioma	NSE, S100, Bcl-2, vimentin, CD56, Syn, CK, Ki67 (50%)
8	Normal	Opacity	Shallow, cystic mass	Neovascularization, iris bombe, synechia	Synechia	Lens subluxation	-	Medium-high uneven echo, ciliary body cystic mass, lens subluxation	Malignant medulloepithelioma	NSE, S100, GFAP, vimentin, CD56, Syn, CD99, EMA, Ki67 (30%)
9	Congestion	Opacity	Shallow, flare, hyphema	Iris atrophy, synechia	Synechia	Cataract	Retinal detachment	Ciliary body mass, retinal detachment	Malignant medulloepithelioma	NSE, S100, vimentin, CD56, Ki67 (80%)
10	Congestion	Normal	Filled with cystic mass	-	-	-	-	Ciliary body mass	Malignant medulloepithelioma	Vimentin, CD56, CD99, Ki67 (40%)
11	Normal	Normal	Shallow	Neovascularization, iris bombe, synechia	Synechia	Cataract	Retinal detachment	Ciliary body cystic mass, retinal detachment	Malignant medulloepithelioma	Vimentin, Syn, CD56, CD99, CK, EMA, Ki67 (80%)

-: Invisible or missed; UBM: Ultrasound biomicroscopy; CT: Computed tomography; MRI: Magnetic resonance imaging; NSE: Neuron specific enolase; GFAP: Glial fibrillary acidic protein; Syn: Synaptophysin; CK: Cytokeratin; EMA: Epithelial membrane antigen.

and other immunohistochemistry markers in tumor cells. Immunohistochemical examination showed that all specimens expressed a variety of positive proteins. Vimentin, CD56, NSE, S-100 protein, and Syn were positive in most cases (Table 2). Neuronal markers such as NSE, GFAP, and S-100 protein can confirm that the tumor contains neuronal and glial components, which further prove that the tumor originates from nonpigmented ciliary epithelium.

DISCUSSION

Ciliary body medulloepithelioma is a rare non-hereditary neuroepithelial tumor which is mainly originated from the medullary epithelial cells of the primitive optic cup inner layer, so the tumor mostly occurs in the ciliary body. Occasionally, it occurs in the optic nerve, retina, or iris^[7]. Ciliary body medulloepithelioma was first reported and termed 'carcinoma primitif' by Badel and Lagrange^[8] in 1892. Finally, with further research and understanding of the disease, Grinker^[9] of the Department of Neurology found that its pathological presentations were similar to those of the central nervous system medulloepithelioma, so he named it 'Ciliary body medulloepithelioma' in 1931 and it is still in use. However, ciliary body medulloepithelioma is different from medulloepithelioma which occurs in the central nervous system, its malignancy is relatively low and the mortality rate is about 10%^[10].

The disease most commonly occurs in children aged 2 to 5y, with 75% to 90% of cases under 10 years of age^[10]. It is the most frequent pediatric primary malignant intraocular tumor second only to retinoblastoma^[11-12]. In our series, the median age was 9y, and 10 patients (90.9%) were children or adolescents (aged less than 18y). While ciliary body medulloepithelioma is rare in adults. Only 1 patient was an adult (aged over 18y) in our study. Most of the tumors are unilateral and patients often present with vision loss, eye pain, leukocoria, and the discovery of eyeball masses. In our research, 6 patients had vision loss and 2 patients had leukocoria. Intraocular tumors often present with proptosis or enlarged eyeballs, cases No.1 and 10 appeared proptosis. However, case No.2 presented with atrophied bulbi, which we considered may be associated with the combination of retinoblastoma and the low IOP.

In the study of 41 patients reported by Kaliki *et al*^[11], corneal opacity, anterior chamber exudation and hyphema, iris neovascularization, cataract and lens subluxation are the most frequent signs, and a few patients with large tumors can cause retinal detachment. Most of the patients in our study had the above clinical features. In a few cases, the initial lesion is disguised as anterior uveitis^[1,13]. In addition to the above clinical manifestation, as tumor cells can produce acidic mucopolysaccharide substances, and cysts are easily formed

in the tumor, some cases can find cystic tissue floating in the anterior chamber or vitreous. In this study, cystic substances appeared in the anterior chamber in 3 cases. Compared with other intraocular tumors, ciliary body medulloepithelioma is more prone to secondary glaucoma. Kaliki *et al*'s^[11] research showed that about 44% to 60% of cases had secondary glaucoma and most of them were neovascular glaucoma. Glaucoma occurred in 6 cases of 11 patients (54.5%) in this study, of which 3 patients (cases No.1, 3, and 5) were neovascular glaucoma. The tumor is insidious, the early tumor is small or hidden behind the ciliary body, and routine ophthalmic examinations are often difficult to detect, which may easily lead to misdiagnosis or underdiagnosis. Therefore, imaging examinations are particularly important for the diagnosis of this disease. B-scan ultrasonography results are often negative in the early stage of the disease, but as the tumor size increases, hyperechoic masses in the ciliary body can be detected, with clear boundaries, different shapes, uneven echo and cystic areas. UBM is of great significance for the diagnosis of ciliary body medulloepithelioma, which can show the location, size, and infiltration extent of the tumor. Although CT and MRI scans can detect ciliary body tumors, the imaging changes are similar to those of retinoblastoma, they are often used to detect recurrence and metastasis during follow-up.

The definitive diagnosis of ciliary body medulloepithelioma still depends on histopathological examination. The tumor type and character were examined by immunohistochemistry. Histological features of ciliary body medulloepithelioma include a gray or white mass with a clear boundary and different sizes. Cystic degeneration, hemorrhage and necrosis can be found in the tumors. Microscopically, the tumor is mainly arranged in tubular, cordlike or reticular patterns. The tumor cells are columnar, with hyperchromatic nuclei, round or oval appearance, and arranged in single or multiple layers. Tumor cells are labeled by related antibodies and express a variety of neuroendocrine markers, such as NSE, S-100, CD56, and vimentin^[14]. The tumors are classified into non-teratomatous and teratomatous medulloepithelioma on the basis of tumor tissue composition and growth pattern. Teratomatoid medulloepithelioma mostly contains heteroplastic foci, including brain tissue, hyaline cartilage, ependymal, ganglion, rhabdomyoblasts or striated muscle, *etc*. The most common is hyaline cartilage^[11]. Both types of medulloepithelioma can be categorized as benign and malignant. According to the criteria of Broughton and Zimmerman^[10], the diagnostic basis of malignant medulloepithelioma includes 1) areas composed of poorly differentiated primitive neuroblastoma cells resembling retinoblastoma; 2) tumor cells have obvious heteroplastic elements and pathological mitotic figures; 3) sarcomatous areas in the tumor; 4) invasion of the adjacent cornea, sclera, lens,

uvea, optic nerve or orbit. In our research, all the patients were diagnosed with malignant ciliary body medulloepithelioma. On the contrary, a benign tumor is smaller, usually confined to the inner surface of the ciliary body, posterior chamber, and intraocular region, mainly composed of sheets and cords of medullary epithelial cells, similar to the embryonic retina and ciliary epithelium. Compared with a malignant tumor, it is rare to find neuroblastoma clumps similar to retinoblastoma, and tumor cell atypia, sarcomatous lesions, distant invasion and metastasis are infrequent. According to Broughton and Zimmerman^[10], typical pediatric ciliary medulloepithelioma originated from embryonic or incompletely differentiated ciliary epithelium, whereas in adults it is considered that arises from fully differentiated ciliary epithelium after undergoing a non-tumor-reactive proliferative stage. Fuchs^[15] considered that adult medulloepithelioma usually occurs in inflamed or traumatized eyes. In our study, there was 1 case of an adult with no history of ocular inflammation or trauma. This is the same as the 6 adult medulloepithelioma cases reported by Kaliki *et al*^[11]. We suggest that adult medulloepithelioma may originate from an occult embryonic neoplasm that grows from childhood and slowly proliferates into malignant tumors. Therefore, adult medulloepithelioma is mostly malignant.

Currently, the treatments for ciliary body medulloepithelioma include plaque radiotherapy, local resection, cryotherapy, chemotherapy, and enucleation. However, there is a lack of established guidelines for optimal treatments. Small localized tumors are often treated with local tumor resection^[15], but usually have a high recurrence rate and require secondary surgery. Such as case No.5 had undergone ciliary body tumor resection 2 years ago but relapsed 2mo before the visit. ¹²⁵I or ¹⁰⁶Ru plaque radiotherapy is attempted as the primary treatment for smaller tumors. For medium-sized and well-defined tumors, local resection combined with plaque radiotherapy can be performed, and the recurrence rate is significantly lower than that of local resection alone^[16-17]. Enucleation is the most common surgery for large or aggressive tumors, and in cases involving the orbit, combined with radiotherapy or chemotherapy as needed. In our series, case No.1 presented with an extraocular extension of the tumor in the conjunctiva, and case No.2 was accompanied by retinoblastoma, so they underwent chemotherapy after enucleation. In recent years, the anterior chamber and vitreous injection of melphalan chemotherapy have been reported in the literature and achieved satisfactory effects^[18]. It is generally considered that the tumor may have extraocular invasion but rarely distant metastasis. Recently, Yazıcıoğlu *et al*^[19] reported a case of submandibular lymphnode metastasis, surgically verified by the otolaryngologist, this merits to be taken into account for the patient's follow-up. When the tumor is confined to the eyeball

without distant invasion and metastasis, the prognosis is better after treatment, and tumor-related death is mainly seen in cases of orbital and intracranial metastasis.

Due to the rarity of ciliary body medulloepithelioma, and the lack of characteristic clinical symptoms and signs, the tumor cannot be detected by the auxiliary examination in time at the early stage. Patients are often diagnosed with 'secondary glaucoma', 'congenital cataract', and 'endophthalmitis', failing to make an early diagnosis and giving correct treatment in time, or even giving unnecessary surgery which makes the disease continue to develop and deteriorate. Eventually, enucleation is performed due to tumor invasion, metastasis, and severe eye pain. In our study, 5 patients had been diagnosed with the above diseases, and some of them underwent cyclophotocoagulation or cataract extraction. Pediatric ciliary medulloepithelioma should be differentiated from other diseases that can cause leukocoria, such as retinoblastoma, congenital cataract, Coats' disease, and persistent hyperplastic primary vitreous. Retinoblastoma and medulloepithelioma have similar clinical manifestations, making them difficult to distinguish. Neuroendocrine markers of both retinoblastoma and medulloepithelioma can be positive, but the former has more numerous mitotic figures, earlier age of onset, higher degree of malignancy, and stronger invasiveness. According to the different papers collected on SIDUO acta books, no calcifications are described in the medulloepithelioma, against the presence of retinoblastoma. It has been reported in the literature^[4] that this disease was misdiagnosed as persistent hyperplastic primary vitreous by ultrasonography, but this disease is generally not accompanied by short axial, which can be used as the key point of identification. Adult medulloepithelioma should be distinguished from adult acquired epithelioma of the nonpigmented ciliary epithelium, which does not have embryonal histopathologic features.

In conclusion, the clinical diagnosis and treatment of ciliary body medulloepithelioma are still difficult. Through the summary and analysis in this article, we hope to make the clinical and pathological characteristics of ciliary body medulloepithelioma clear. In the clinic, we should consider or exclude the possibility of this disease for children with unexplained unilateral glaucoma, uveitis, endophthalmitis, cataract, and retinal detachment. Combined with UBM and other imaging examinations, timely and correct diagnosis and treatment should be made to reduce the occurrence of misdiagnosis.

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