

# A Choroidal Melanoma With Ciliary Body Involvement in a Young Female

Yen Sheong Lai <sup>1,2</sup>, Ismail Shatriah <sup>1</sup>, Yee Lin Lo <sup>2</sup>, Koon Ling Koh <sup>2</sup>, Jayaraman Kogilavaani <sup>2</sup>

Review began 03/17/2024

Review ended 03/20/2024

Published 03/26/2024

© Copyright 2024

Lai et al. This is an open access article distributed under the terms of the Creative Commons Attribution License CC-BY 4.0., which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.

1. Department of Ophthalmology and Visual Science, School of Medical Sciences, Universiti Sains Malaysia, Kubang Kerian, MYS 2. Department of Ophthalmology, Hospital Raja Permaisuri Bainun, Ipoh, MYS

**Corresponding author:** Ismail Shatriah, shatriah@usm.my

## Abstract

Choroidal melanoma with ciliary body involvement is rare, especially in young adults and Asians. Here, we report the case of a young, healthy Chinese woman who complained of decreased vision in the left eye for one week. Her ocular examination and imaging were suggestive of choroidal melanoma involving the ciliary body. The patient underwent enucleation of the left eye. Close monitoring was needed, as the involvement of the ciliary body in choroidal melanoma is associated with a high risk of metastasis.

**Categories:** Ophthalmology

**Keywords:** metastasis, asian, female, young, ciliary body, choroidal melanoma

## Introduction

Melanoma is a malignant tumor that originates from melanocytes at various body sites, with the ocular area being the second most common location after the skin [1]. The majority (85%) of ocular melanomas are uveal in origin [1,2], with the choroid being the most frequent site of origin. Ciliary body melanomas are rarely encountered and comprise 10% of all intraocular melanomas [3]. The common presentations of choroidal melanoma include flashes of light, floaters, visual field defects, and reduced vision [4]. Choroidal melanoma typically presents as an elevated, dome-shaped, gray-brown-colored lesion of the choroid [5].

Choroidal melanoma is more common in Caucasians (five to six cases per million) compared to Asians (0.2 cases per million). The mean presenting age is around the fourth to fifth decade of life in Asians, more than a decade earlier than in Caucasians [4]. Choroidal melanoma is found slightly more frequently in men, with a male-to-female ratio of 1.29 [2,4]. Here, we report on a rare presentation of choroidal melanoma with ciliary body involvement in a young Chinese woman.

## Case Presentation

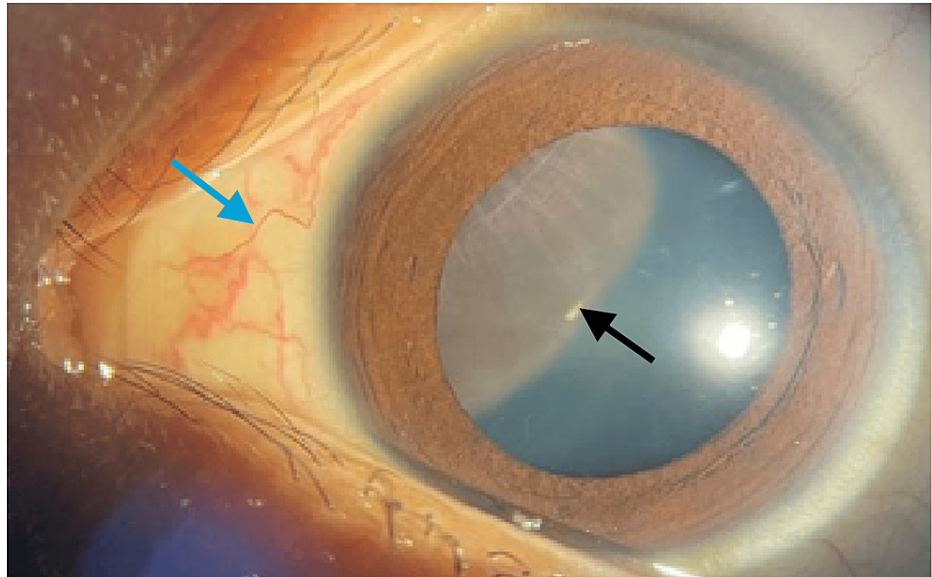
A healthy 23-year-old Chinese woman presented with sudden-onset painless blurred vision in her left eye for one week. She described her symptoms as generalized blurry vision associated with redness of her left eye and two episodes of photopsia. There were no floaters, no eye discharge, no loss of appetite or weight, no history of ocular trauma, and no family history of malignancy.

Ocular examination revealed a visual acuity of 6/9 in the right eye and 5/60 in the left eye, pinhole 6/18. A relative afferent pupillary defect was absent. The right eye finding was unremarkable. The anterior segment of the left eye revealed a large ciliary body mass located retrolental at the superonasal quadrant (Figure 1), causing striae at the superonasal of the posterior lens capsule associated with sentinel vessels at the nasal conjunctiva (Figure 2).

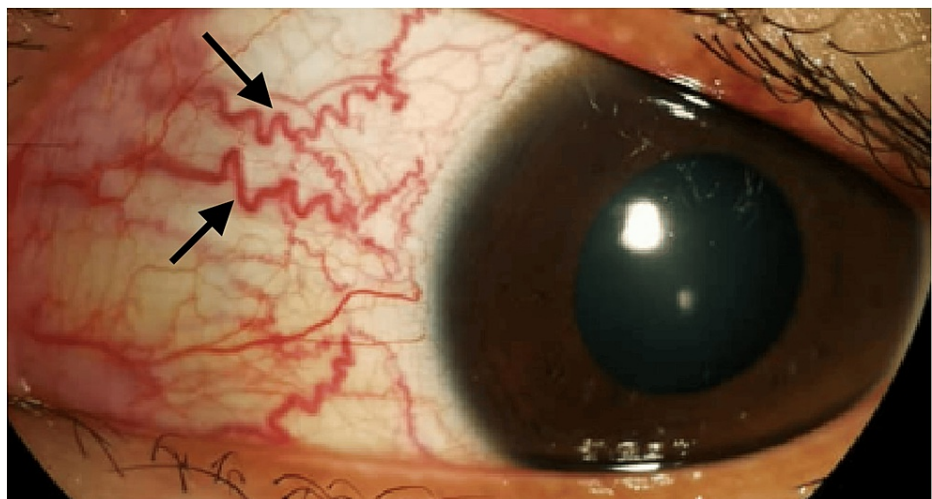
### How to cite this article

Lai Y, Shatriah I, Lo Y, et al. (March 26, 2024) A Choroidal Melanoma With Ciliary Body Involvement in a Young Female. Cureus 16(3): e56955. DOI 10.7759/cureus.56955





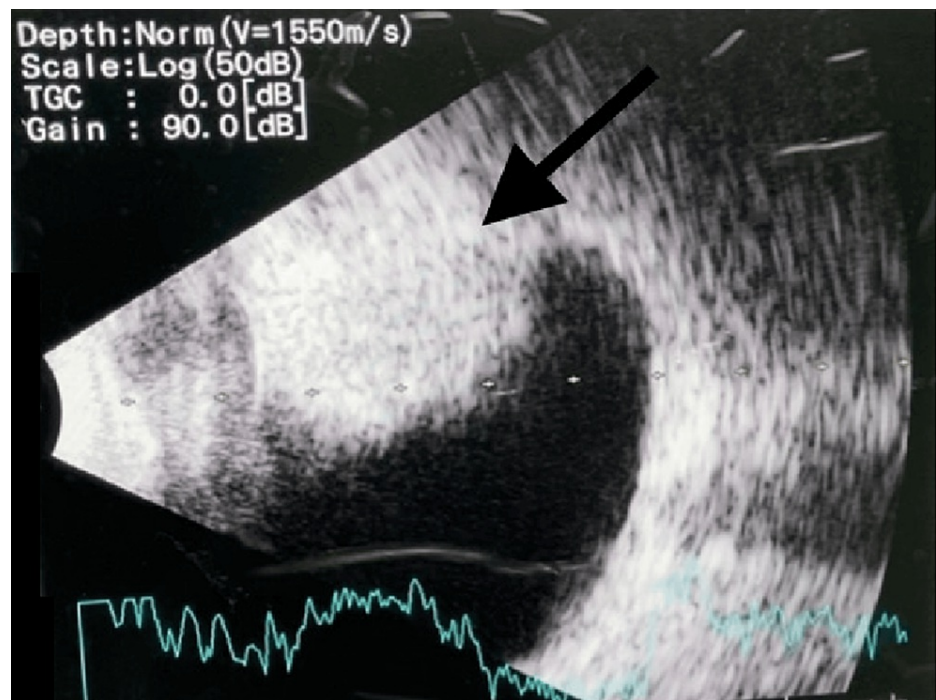
**FIGURE 1: Anterior segment photo of the left eye showed a ciliary body mass (black arrow) with dilated conjunctival vessels (blue arrow)**



**FIGURE 2: Anterior segment photo of the left eye showed sentinel vessels at the nasal conjunctiva (black arrows)**

Gonioscopy showed that all angles were open but unable to appreciate any mass. Fundus examination noted exudative retinal detachment at the inferonasal area of the periphery of the retina. A systemic examination was unremarkable. B-scan ultrasonography revealed a well-circumscribed homogenous mass at the superonasal aspect of the choroid (Figure 3).





**FIGURE 3: B-scan ultrasonography of the left eye at initial presentation, showing a well-circumscribed homogenous mass (black arrow)**

Blood investigations and tumor markers were normal, except for raised liver enzymes, which showed transaminitis. An ultrasound of the hepatobiliary system revealed hepatomegaly with hepatic steatosis, but neither a liver mass nor any lesions were noted. An MRI of the brain and orbits was suggestive of an enhancing lentiform-shaped intraocular lesion seen at the superomedial aspect of the left globe involving the sclera and ciliary body and possible infiltration at the attachment of the medial rectus (Figure 4).

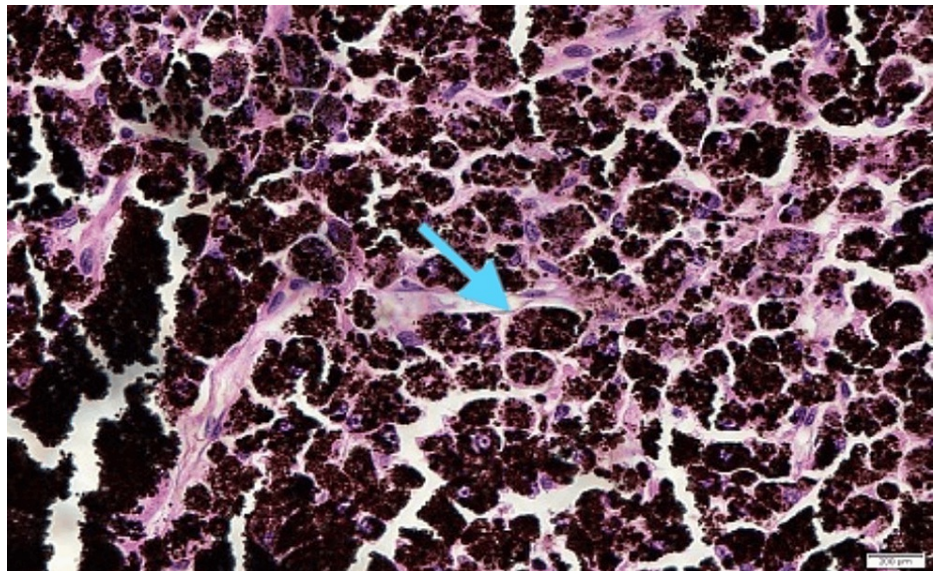




**FIGURE 4: Coronal view T1 post-contrast MRI of the brain and orbits showed an enhancing lentiform-shaped intraocular lesion seen at the superomedial aspect of the left globe (blue arrow)**

The patient was diagnosed with left eye choroidal melanoma involving the ciliary body and sclera. Left eye enucleation was done 20 days after presentation, and intraoperatively, no extrascleral extension was noted. Histopathological examination showed an epithelioid-type choroidal melanoma measuring 14 mm at the largest basal diameter, with ciliary body involvement (Figure 5).





**FIGURE 5: Histopathological examination (x400 magnification) showed epithelioid-appearing neoplastic cells with round to oval nuclei, prominent nucleoli, and abundant eosinophilic cytoplasm (blue arrow)**

Postoperatively, the patient was doing well. Her left eye socket wound was clean and intact, with no pigmented lesions. The patient remained under close follow-up every four months, with no evidence of recurrence or metastasis at one year.

## Discussion

Uveal melanoma is a malignancy of melanocytes that can be found in the uveal tract of the eye, which comprises the iris, ciliary body, and choroid. Choroidal melanoma is the most common of all subtypes of uveal melanoma. Risk factors for uveal melanoma include being Caucasian, elderly, and having ocular melanocytosis and oculodermal melanocytosis (nevus of Ota) [6].

Our patient was a Chinese woman in her early 20s with no comorbidities or predisposing risk factors. Thus, it was unusual for her to have choroidal melanoma. To date, there have been five published cases of uveal melanoma affecting people in their 20s, and most were Caucasian, irrespective of gender. All five cases had uveal melanoma in different clinical presentations, but none involved the ciliary body. Our patient's Chinese ethnicity and presentation of a choroidal mass involving the ciliary body made her different from the published case reports. Ciliary body involvement is associated with a high risk of metastasis owing to the rich blood supply [3]. The diagnosis of uveal melanoma is mainly clinical; thus, a high level of clinical suspicion is needed. Table 1 summarizes the published reports on uveal melanoma diagnosed in patients in their 20s [7-11], including our patient.



Authors/year	Age/gender/race or country	Risk factor	Presentation	Examination	Treatment	Outcome
Pomeranz et al. (1981) [7]	29/female/White	Ocular melanocytosis	Blurred vision	Two separate choroidal masses with shallow retinal detachment	Enucleation	Not available
de A Silva et al. (2011) [8]	28/female/Brazilian	History of right eye enucleation 10 years ago for choroidal melanoma	Intense headache	The left frontal lobe, single metastatic melanoma	Whole-brain radiation therapy	No recurrence for 24 months post-radiation or death from pulmonary embolism
Nair et al. (2014) [9]	24/male/Asian Indian	Nil	Sudden loss of vision, preceded by painful, progressive proptosis of the right eye (orbital cellulitis)	A B-scan showed a large dome-shaped mass filling the posterior segment	Three days of intravenous dexamethasone, intravenous antibiotics, topical steroids, and cycloplegic eye drops, followed by enucleation	There is no evidence of systemic metastasis
Gupta et al. (2016) [10]	26/male/United States	Nil	Headache and a red, painful eye with nausea and vomiting	Neovascular glaucoma, necrotizing scleritis, and exudative retinal detachment	Enucleation	Not available
Hackett et al. (2018) [11]	21/female/United States	History of right optic nerve melanocytoma	Transient right-sided visual field deficits, floaters, unilateral headache, and photopsia	Dark-pigmented vascular mass obscuring the optic nerve with no extension to the surrounding orbit	Referred to a retina specialist for further management	Not available
Our patient (2024)	23/female/Asian (Chinese)	Nil	Blurred vision and eye redness	Choroidal mass involving the ciliary body	Enucleation	No systemic metastasis

TABLE 1: Summary of published case reports of uveal melanoma in patients in their 20s

Computed tomography and MRI are important for identifying tumor extension and ruling out metastases, if present. The main treatment options include enucleation, brachytherapy, transpupillary thermotherapy, and orbital exenteration for tumors with orbital invasion [12]. In our case, enucleation was performed because there was no extrascleral extension.

A better prognosis for choroidal melanoma is observed in Asian populations due to the younger age of onset, with a five-year survival rate of more than 70% [6]. Large basal tumor diameters, ciliary body involvement, a non-spindle cell type, and extrascleral extension are worse prognostic indicators [6,13]. Tumor sizes are classified into small (11 mm diameter), medium (11-15 mm diameter), or large (15 mm diameter), with a 10-year survival rate of 81% for small melanomas, 60% for medium melanomas, and 35% for large melanomas [13]. Our patient had a medium-sized melanoma. Therefore, long-term, regular systemic follow-up is necessary, as the prognosis is poor once metastases have developed.

Conclusions

Uveal melanoma is a disease that usually affects elderly Caucasians. However, the possibility of its occurrence in younger patients of other races should not be overlooked. Misdiagnosis is common when patients are not properly evaluated and investigated, leading to delays in treatment and, consequently, the development of metastatic disease.

Additional Information

Author Contributions

All authors have reviewed the final version to be published and agreed to be accountable for all aspects of the work.



**Concept and design:** Yen Sheong Lai, Ismail Shatriah

**Acquisition, analysis, or interpretation of data:** Yen Sheong Lai, Ismail Shatriah, Yee Lin Lo, Koon Ling Koh, Jayaraman Kogilavaani

**Drafting of the manuscript:** Yen Sheong Lai

**Critical review of the manuscript for important intellectual content:** Yen Sheong Lai, Ismail Shatriah, Yee Lin Lo, Koon Ling Koh, Jayaraman Kogilavaani

**Supervision:** Ismail Shatriah

## Disclosures

**Human subjects:** Consent was obtained or waived by all participants in this study. **Conflicts of interest:** In compliance with the ICMJE uniform disclosure form, all authors declare the following: **Payment/services info:** All authors have declared that no financial support was received from any organization for the submitted work. **Financial relationships:** All authors have declared that they have no financial relationships at present or within the previous three years with any organizations that might have an interest in the submitted work. **Other relationships:** All authors have declared that there are no other relationships or activities that could appear to have influenced the submitted work.

## Acknowledgements

We would like to express our sincere gratitude to Dr. Amizatul Aini binti Salleh, a pathologist based at Hospital Serdang, Kajang, Selangor, for contributing the histopathological examination slide and interpretation.

## References

1. Chang AE, Karnell LH, Menck HR: The National Cancer Data Base report on cutaneous and noncutaneous melanoma: a summary of 84,836 cases from the past decade. *Cancer*. 1998, 83:1664-78. [10.1002/\(sici\)1097-0142\(19981015\)83:8%3C1664::aid-cnrcr23%3E3.0.co;2-g](https://doi.org/10.1002/(sici)1097-0142(19981015)83:8%3C1664::aid-cnrcr23%3E3.0.co;2-g)
2. Singh AD, Turell ME, Topham AK: Uveal melanoma: trends in incidence, treatment, and survival. *Ophthalmology*. 2011, 118:1881-5. [10.1016/j.ophtha.2011.01.040](https://doi.org/10.1016/j.ophtha.2011.01.040)
3. Costache M, Patrascu OM, Adrian D, Costache D, Sajin M, Ungureanu E, Simionescu O: Ciliary body melanoma - a particularly rare type of ocular tumor. Case report and general considerations. *Maedica (Bucur)*. 2013, 8:360-4.
4. Manchegowda P, Singh AD, Shields C, Kaliki S, Shah P, Gopal L, Rishi P: Uveal melanoma in Asians: a review. *Ocul Oncol Pathol*. 2021, 7:159-67. [10.1159/000512738](https://doi.org/10.1159/000512738)
5. Rodríguez A, Dueñas-Gonzalez A, Delgado-Pelayo S: Clinical presentation and management of uveal melanoma. *Mol Clin Oncol*. 2016, 5:675-7. [10.3892/mco.2016.1037](https://doi.org/10.3892/mco.2016.1037)
6. Yue H, Qian J, Yuan Y, Zhang R, Bi Y, Meng F, Xuan Y: Clinicopathological characteristics and prognosis for survival after enucleation of uveal melanoma in Chinese patients: long-term follow-up. *Curr Eye Res*. 2017, 42:759-65. [10.1080/02713683.2016.1245422](https://doi.org/10.1080/02713683.2016.1245422)
7. Pomeranz GA, Bunt AH, Kalina RE: Multifocal choroidal melanoma in ocular melanocytosis. *Arch Ophthalmol*. 1981, 99:857-63. [10.1001/archophth.1981.03930010857014](https://doi.org/10.1001/archophth.1981.03930010857014)
8. de A Silva DO, Matis GK, Costa LF, et al.: Ocular melanoma: keep your eyes open for late brain metastases. *Surg Neurol Int*. 2011, 2:144.
9. Nair AG, Kaliki S, Ali MJ, Naik MN, Vemuganti GK: Intraocular malignant melanoma of the choroid presenting as orbital cellulitis. *Int Ophthalmol*. 2014, 34:647-50. [10.1007/s10792-013-9836-1](https://doi.org/10.1007/s10792-013-9836-1)
10. Gupta N, Terrell W, Schoenfield L, Kirsch C, Cebulla CM: Uveal melanoma mimicking advanced coats' disease in a young patient. *Ocul Oncol Pathol*. 2016, 2:140-3.
11. Hackett A, Stuart J, Blessing N, Borden N: Headache with aura: a case report of ocular melanoma. *Am J Emerg Med*. 2018, 36:2135.e7-8. [10.1016/j.ajem.2018.08.047](https://doi.org/10.1016/j.ajem.2018.08.047)
12. Wong W, Sundar G, Chee C, Zhao PS, Rajagopalan R, Gopal L: Clinical spectrum, treatment and outcomes of uveal melanoma in a tertiary centre. *Singapore Med J*. 2019, 60:474-8. [10.11622/smedj.2019054](https://doi.org/10.11622/smedj.2019054)
13. Shields CL, Furuta M, Thangappan A, et al.: Metastasis of uveal melanoma millimeter-by-millimeter in 8033 consecutive eyes. *Arch Ophthalmol*. 2009, 127:989-98. [10.1001/archophthalmol.2009.208](https://doi.org/10.1001/archophthalmol.2009.208)