Content available at: https://www.ipinnovative.com/open-access-journals

IP International Journal of Ocular Oncology and Oculoplasty

Journal homepage: https://ijooo.org/

Case Report

The pigmented puzzle: A case of cilio-choroidal melanocytoma

Sourabh Madanrao Jadhav[1], Krishnakumar Subramanian[2], Suganeswari Ganesan[1]*

[1]*Dept. of Vitreo Retina, Shri Bhagvan Mahavir, Vitreoretinal Services, Sankara Nethralaya, Chennai, Tamil Nadu, India*
[2]*Dept. of Ocular Pathology, Sankara Nethralaya, Chennai, Tamil Nadu, India*

ARTICLE INFO

Article history:
Received 06-09-2023
Accepted 08-11-2023
Available online 01-02-2024

Keywords:
Ciliary body
Melanocytoma
Melanoma
Enucleation

ABSTRACT

A 5-year-old female patient presented with complaints of an inability to identify objects at home. Upon examination, a large retro-pupillary pigmented mass was discovered in the right eye. B-scan ultrasonography and magnetic resonance imaging (MRI) suggested a diagnosis of cilio-choroidal melanoma, leading to the decision to perform an enucleation. However, the histopathological analysis revealed the mass to be a cilio-choroidal melanocytoma, a rare benign pigmented tumor commonly associated with the optic nerve head. This case is unique due to the patient's young age, the unusual tumor location, and its larger size compared to previous reports. This case underscores the importance of considering melanocytoma in the differential diagnosis of pigmented intraocular tumors, especially given its clinical resemblance to malignant melanoma.

1. Introduction

Melanocytoma, a variant of melanocytic naevus,[1] was first described by Zimmerman and Garron in 1962.[2,3] Although it accounts for approximately 0.6% of all intraocular tumors,[4] making it a relatively rare occurrence, its importance cannot be understated. This benign tumor has the capacity for local invasion and on rare occasions, can undergo malignant transformation.[5,6] Typically, melanocytomas are associated with the optic disc area, but in rare instances, they can occur in extra-papillary locations within the uveal tract (iris, ciliary body, choroid).[6] These uncommon locations can pose a diagnostic challenge due to their resemblance to uveal melanoma and naevus.[6] Histopathology remains the gold standard for diagnosing melanocytoma and differentiating it from melanoma.[7] We report an atypical case of a five-year-old female with a large cilio-choroidal mass in her right eye, initially diagnosed clinically and radiologically as melanoma."

2. Case Report

We saw a five-year-old girl brought by her parents due to difficulties identifying objects at home, despite a normal birth and developmental history. Her visual acuity was reduced to 20/50 in her right eye (OD), while the left (OS) was near normal at 20/25.

A detailed ocular examination was conducted. The right eye's slit-lamp biomicroscopy revealed no abnormalities in the conjunctiva, cornea, and anterior chamber. However, iris bowing was noted inferiorly (Figure 1, Yellow arrowhead) and a large brown pigmented mass was observed retro-pupillary, obscuring fundus details. The OS was within normal limits, and intraocular pressure was normal in both eyes.

An ultrasound of the OD revealed a large dome-shaped mass located in the ciliary body region, its characteristics arousing suspicion for melanoma (Figure 2,

* *Corresponding author*.
E-mail address: suganes@gmail.com (S. Ganesan).

https://doi.org/10.18231/j.ijooo.2023.044

White arrowhead). Supporting this, an MRI of the orbit revealed a similarly sized tumor that exhibited hyper-intense signals on T1-weighted images (Figure 3a), hypo-intense signals on T2-weighted images (Figure 3b,c), and mild enhancement on contrast features consistent with a cilio-choroidal melanoma in the OD.

Systemic evaluations, including an ultrasound of the abdomen, hematological evaluation, and chest x-ray, found no evidence of metastasis, keeping the suspected diagnosis localized to the eye.

Given these findings, we considered two options: fine needle aspiration biopsy for histopathological evaluation or enucleation with subsequent histopathology. The patient's parents opted for enucleation, which was performed on the right eye.

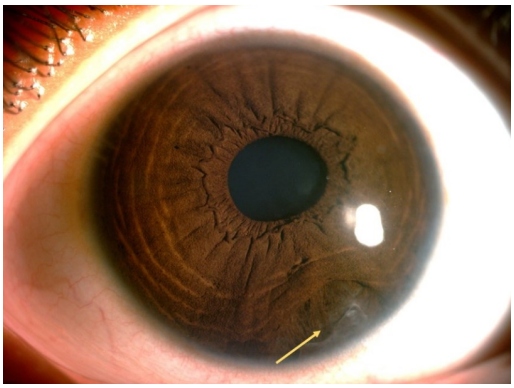

Figure 1: Slit lamp image of the right eye showing inferior iris bowing with an atrophic patch.

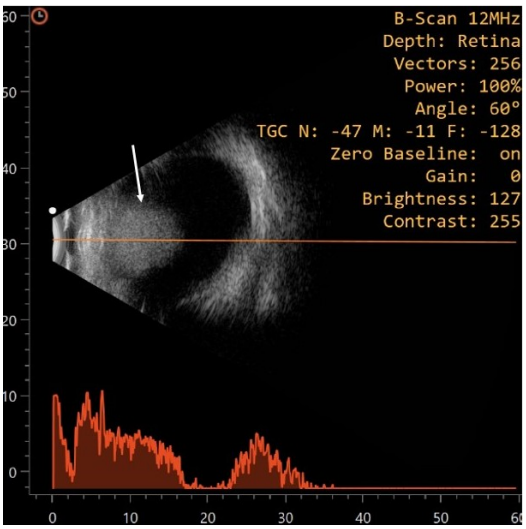

Figure 2: Ultrasonography showing a retro-pupillary mass lesion occupying the ciliary body area.

The histopathology of the eye showed a dark brown pigmented mass in the region of the ciliary body and

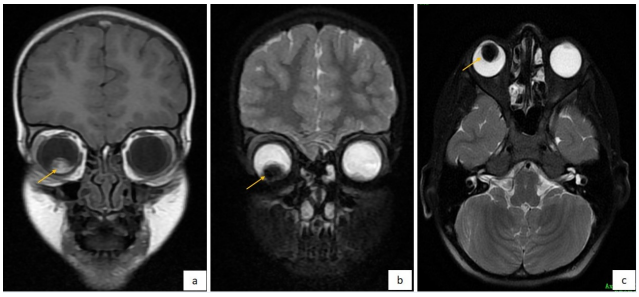

Figure 3: Magnetic resonance imaging showing, ahyperintense mass lesion on T1 weighted image in the coronal section (**a**) and hypointense mass on T2 weighted images in the coronal and axial sections (**b,c**).

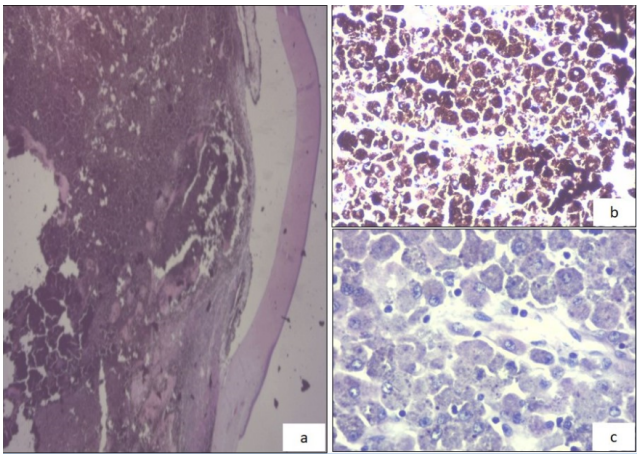

Figure 4: Histopathology of the enucleated eye showing a pigmented mass in theciliary body and anterior choroid (**a**), Hematoxylin and Eosin staining showing deeply pigmented polygonal cells (**b**) and bleach preparation showing type 1 melanocyte cells (**c**).

anterior choroid (Figure 4a). The heavy pigmentation of the tumor cells obscured details on Hematoxylin–Eosin (H&E) staining (Figure 4b), but a bleach preparation with potassium permanganate revealed magno-cellular cells with small round nuclei, characteristic of type 1 melanocytes (Figure 4c). No mitosis, atypia, or scleral invasion was observed, and a diagnosis of cilio-choroidal melanocytoma was made.

3. Discussion

Melanocytomas are rare benign pigmented tumours1, which are a variant of melanocytic nevus.[1] Classically melanocytoma is a tumor of the optic nerve head, which appears as a pigmented homogeneous mass with a total absence of autofluorescence.[2] It can be confined to the disk in 15% of cases. The inferotemporal quadrant is the most common site (33%) and the nasal side is the least common (12%)2. It is usually isolated but can be associated with

neurofibromatosis type 2, basal cell carcinoma, and vitiligo. But there are case reports of uveal tract locations. In such situations, it is clinically very challenging to differentiate uveal melanocytomas from uveal melanomas.

In previous studies by Howard et al, a study of 907 pigmented intraocular tumors, found only 5 melanocytomas (an incidence of 0.6%).[8] Jakobiec and Silbert et al in a review of 189 iris or ciliary body lesions, identified 10 of 189 (5%) as melanocytomas.[9]

In the literature, the most common age of presentation of cilio-choroidal melanocytoma is in the range of 30-40 years.[10] Few case reports of younger age groups are also reported.[10] To our knowledge, this is the youngest reported case of cilio-choroidal melanocytoma (5 years). Classical melanocytomas rarely exceed 2-disc diameter 11 in size, the tumour size of our case was unusually large measuring 11.0 mm in height and 11.0mm in base, which corresponds to the average size of the reported cases in literature, varying from 3.0mm to 15 mm.[9]

On direct clinical examinations, melanocytes are jet-black homogenous tumors, and rarely cystic and necrotic changes can be seen. Whereas melanomas are brown with non-uniform distribution and show surface irregularities and orange pigmentation.[10,11] Both melanomas and melanocytomas are slow-growing tumours so clinically very tough to distinguish from each other.[10,12]

Histopathology is the diagnostic modality.[7] Melanocytomas can present two types of cells, type I and type II. Type I cells are composed of large and polyhedral round nuclei and cytoplasm with giant melanosomes seen on electron microscopy. Type II cells present elongated cytoplasm and small melanin granules.[7,13]

Melanocytomas are highly reflective on ultrasound, but this is not true for all tumors, case reports of extra-papillary melanocytomas are shown to have low internal reflectivity.[14] Our case had a large mass in the cilio-choroidal area with moderate surface and uniform low internal reflectivity. Findings are equivocal with ultrasound findings of uveal melanoma.

MRI orbit is not considered as superior to other diagnostic modalities due to equivocal features of uveal melanoma and uveal melanocytomas.[11,12]

Diagnostic fine needle aspiration biopsy is usually preferred in ciliary body mass to differentiate melanoma from other pigmented masses. Can be done either by trans scleral or trans-vitreal route. In melanoma, the cytology should reveal melanoma cells, if it reveals melanocytoma cells it is possible that sampling error or foci of tumor mass is missed.[11,15]

Melanocytomas are slow-growing rarely affect the vision and have rare malignant transformations seen in 1-2% of cases.[4,7] So, diagnosed cases of melanocytoma can be observed at 3 monthly intervals and thereafter 1 yearly.[16] When less than 3 clock hours of ciliary body are involved local tumor resection is possible.[10] which can be done in the form of irido-cyclectomy with or without corneo-scleral resection or scleral patch graft.

LoRusso et al. advised surgical removal rather than observation for management of pigmented lesions of the ciliary body due to both the difficulty in clinically distinguishing a melanocytoma from malignant melanoma and the frequent occurrence of necrosis (36%) seen in ciliary body melanocytomas.[10,16]

Saxena et al have advocated the use of plaque brachytherapy for ciliary melanocytomas with intra-ocular and extra-ocular growth of the tumor and have found it effective in such cases.[17]

Melanocytoma can undergo spontaneous necrosis, vaso-occlusion, pigment dispersion glaucoma, and neovascular glaucoma resulting in painful eye.[10,16] Painful blind eyes, cases with large tumor size, malignant transformation of the tumor, and patient conscious willingness are indicated for enucleation.[10,16]

Though our case did not show any gross signs of malignancy. However, tumor size and radiological imaging did not suggest melanocytoma, instead gave a suspicion of cilio-choroidal melanoma. Thus, an enucleation was performed.

4. Conclusion

This case presents a unique occurrence of a cilio-choroidal melanocytoma in a very young patient, with a tumor size larger than typically reported. It underscores the importance of including melanocytoma in the differential diagnosis of uveal tract tumors, even in young patients. Despite the availability of eye salvage options, the potential for malignancy and large tumor size often lead to enucleation. Clinicians should carefully weigh these factors when managing such cases. Further research is needed to develop strategies for early differentiation between melanocytoma and melanoma, potentially preserving vision in affected individuals.

5. Source of Funding

None.

6. Conflict of Interest

None.

References

1. Reidy JJ, Apple DJ, Steinmetz RL. Melanocytoma: nomenclature, pathogenesis, natural history and treatment. *Survey Ophthalmol*. 1985;29(5):319–27.
2. Lee E, Sanjay S. Optic Disc Melanocytoma Report of 5 Patients from Singapore with a Review of the Literature. *Asia Pac J Ophthalmol (Phila)*. 2015;4(5):273–8.
3. Zimmerman LE, Garron LK. Melanocytoma of the optic disc. *Int Ophthalmol Clin*. 1962;2:431–40.

4. Howard GM, Forrest AW. Incidence and location of melanocytomas. *Arch Ophthalmol*. 1967;77(1):61–6.
5. Raichand M, Peyman GA, Juarez CP, Seetner AA, Sugar J, Goldberg MF, et al. Resection of uveal melanocytoma: clinicopathological correlation. *Br J Ophthalmol*. 1983;67(4):236–43.
6. Shields JA, Shields CL, Eagle RC. Melanocytoma (hyperpigmented magnocellular nevus) of the uveal tract: The 34th G. Victor Simpson lecture. *Retina*. 2007;27(6):730–9.
7. Odashiro M, Odashiro A, Leite L, Melo M, Odashiro P, Miiji L, et al. Melanocytoma of ciliary body and choroids simulating melanoma. *Pathol Res Pract*. 2010;206(2):130–3.
8. Howard GM, Forrest AW. Incidence and location of melanocytomas. *Arch Ophthalmol*. 1967;77(1):61–6.
9. Jakobiec FA, Silbert G. Are most iris "melanomas' really nevi? A clinicopathologic study of 189 lesions. *Arch Ophthalmol*. 1981;99(12):2117–32.
10. Lorusso FJ, Boniuk M, Font RL. Melanocytoma (magnocellular nevus) of the ciliary body: report of 10 cases and review of the literature. *Ophthalmology*. 2000;107(4):795–800.
11. Abdellaoui T. Large Choroidal Melanocytoma Simulating Choroidal Melanoma: A Difficult Differential Diagnosis and an Inevitable Enucleation. *Case Rep Ophthalmol Med*. 2020;p. 1–5. doi:10.1155/2020/8890857.
12. Shields JA, Augsburger JJ, Bernardino V, Eller AW, Kulczycki E. Melanocytoma of the ciliary body and iris. *Am J Ophthalmol*. 1980;89(5):632–5.
13. Al-Hinai A, Edelstein C, Burnier MN. Unusual case of melanocytoma. *Can J Ophthalmol*. 2004;39(4):461–3.
14. Robertson DM, Campbell RJ, Salomao DR. Mushroom-shaped choroidal melanocytoma mimicking malignant melanoma. *Arch Ophthalmol*. 2002;120(1):82–5.
15. Tang PH, Shields RA, Schefler AC, Mruthyunjaya P. Biopsy of a choroidal melanoma using transvitreal pars plana vitrectomy. *Ophthalmic Surg Lasers Imaging Retina*. 2018;49(8):645–7.
16. Ahmad SS, Lad L, Ghani SA. A case of choroidal melanocytoma mimicking a choroidal melanoma. *Saudi J Ophthalmol*. 2015;29(3):242–5.
17. Shanmugam M, Saxena M, Ramanjulu R, Tekwani P. Brachytherapy, a viable option of globe salvage in treatment of large ciliary body melanocytoma. *Indian J Ophthalmol*. 2014;62(9):966–8.

Author biography

Sourabh Madanrao Jadhav, Fellow

Krishnakumar Subramanian, Senior Consultant

Suganeswari Ganesan, Senior Consultant

Cite this article: Jadhav SM, Subramanian K, Ganesan S. The pigmented puzzle: A case of cilio-choroidal melanocytoma. *IP Int J Ocul Oncol Oculoplasty* 2023;9(4):205-208.