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Case Report

Acute subconjunctival pigmentation with an underlying ciliary body mass: An unusual case presentation

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Abstract

Intraocular melanocytoma is a rare slowly growing benign tumor that may occur anywhere along the uveal tract including the ciliary body but is more commonly found near the optic disc. It is darkly pigmented and may exhibit fast growth and possibly malignant transformation. Ciliary body (CB) melanocytoma is rare and often misdiagnosed as a melanoma. We are reporting a case of CB melanocytoma in a 74-year old lady with unusual initial presentation of an acute onset of subconjunctival pigmentation and extra-scleral extension of the melanocytoma with coarse melanin granules. The diagnosis of melanocytoma was confirmed by histopathological examination of the prolapsed tissue and by ultrastructural studies. Literature review of similar cases is also summarized.

Keywords: Ciliary body, Melanocytoma, Melanoma, Electron microscopy

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Introduction

Melanocytoma was first described by Zimmerman and Garon in 1962, following which, a larger study including 907 pigmented intraocular tumors was published in 1967.[1] In this study, the authors emphasized on the benign nature of this darkly pigmented lesion, the challenge in its diagnosis and the characteristic histopathological appearance of large tumor cells with abundant cytoplasm and small uniform nuclei seen on bleached sections.[1] They also stressed on the rarity of this tumor in the ciliary body (CB) occurring in 2/89 CB lesions, the close resemblance of malignant melanoma in that location and the possible associated cataract and anterior chamber angle involvement.[1] The original description of this lesion has not changed much over the years, and despite it being classically described at the optic disc, it is known to occur anywhere in the uveal tract including the CB.[2] It's location at the CB has been described to show more irregular corrugated surface in comparison to a melanoma. It is also more likely to cause pigment dispersion causing secondary glaucoma or overlying pigmentation of the sclera and episclera.[3]

We are describing a case of CB melanocytoma that presented initially with an acute onset of sub-conjunctival pigmentation. The diagnosis was confirmed by histopathological examination of the excised CB mass and ultrastructural studies.

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Case report

A 74-year old lady presented to our tertiary care eye hospital with a history of sudden blackish peri-limbal discoloration of her left eye of one-month duration. According to the patient, her symptom started as a small black spot which became diffuse over a short period of 30 minutes. There was no pain or associated ocular symptoms. She was clinically referred from a general hospital as a case of possible Sclero-malacia Perforans with prolapsed uveal tissue. However, the patient was systemically healthy and was re-assessed in our eye hospital, where she was found to have uncorrected visual acuity measuring 20/40 in the right eye and 20/160 in the left eye. Intraocular pressure measured 15 mmHg and 18 mmHg in the right and left eye, respectively. Slit-lamp examination of the right eye was unremarkable. Examination of the left eye revealed diffuse sub-conjunctival dark pigmentation involving the superior, nasal and inferior peri-limbal area and reaching almost to the fornices. The dark pigmentation was obscuring conjunctival blood vessels and the sclera. (Fig. 1a) A small dome shaped conjunctival elevation was appreciated medially at 9 O'clock position about 4 mm from the limbus measuring 3 × 3 mm. The cornea was clear, with normal iris and round central pupil. A pigmented CB mass was noted extending from 8 to 10 O'clock, indenting the lens and causing sectorial cortical cataract. Fundus examination was unremarkable in both eyes.

Magnetic resonance imaging (MRI) was initially suggestive of a scleral mass (Fig. 1b). Ultrasound bio-microscopy (UBM) revealed cilio-choroidal tumor with medium to high internal reflectivity. The mass was invading the sclera (3.83 mm form limbus) with possible extra-scleral extension. The choroidal component of the tumor measured 8.15 × 8.21 × 2.86 mm with medium to high internal reflectivity and uniform regular internal structure. The extension of lesion into the choroid was not seen during fundoscopy as it was located in the extreme fundus periphery (Fig. 1c).

The initial clinical diagnosis was CB melanoma with choroidal involvement, extra-scleral extension, and pigment dispersion in the sub-conjunctival space. Given this unusual clinical picture, a conjunctival biopsy from the area of suspected extension was obtained to reach the correct diagnosis. The excisional biopsy of the conjunctival and episcleral tissue was performed with bare underlying sclera, which showed an area of extreme thinning with no visible protruding CB tissue. This biopsy was received at our Pathology department as a tissue block for consultation to rule out melanoma. The initial histopathological findings included the presence of heavy coarse melanin pigments, which were mostly seen within CD-68 positive macrophages and absent expression of Melan-A & HMB45 (Fig. 1d). The impression was suspicious of an underlying melanocytoma of the CB rather than a melanoma.

Since the histopathological results were not conclusive for CB melanoma, a deeper excisional biopsy or en-block excision was planned under peri-bulbar anesthesia, via a fornix-based scleral flap nasally. The medical rectus was isolated and dis-inserted for adequate exposure. Dissection of the sclera revealed a pigmented prolapsed mass, which was shaved with radiofrequency diathermy needle and blade. A cryotherapy was applied over the scleral defect on a double freeze-thaw technique. A scleral graft was used to close the defect and was sutured in place using non-absorbable sutures. The excisional biopsy was sent to us for routine tissue processing and Electron Microscopy (EM). The CB tumor mass showed large polyhedral non-cohesive cells with large heavy melanin pigment granules and uniform nuclei consistent with type I cells of a melanocytoma, which was further confirmed by ultra-structural studies (Fig. 1e & f).

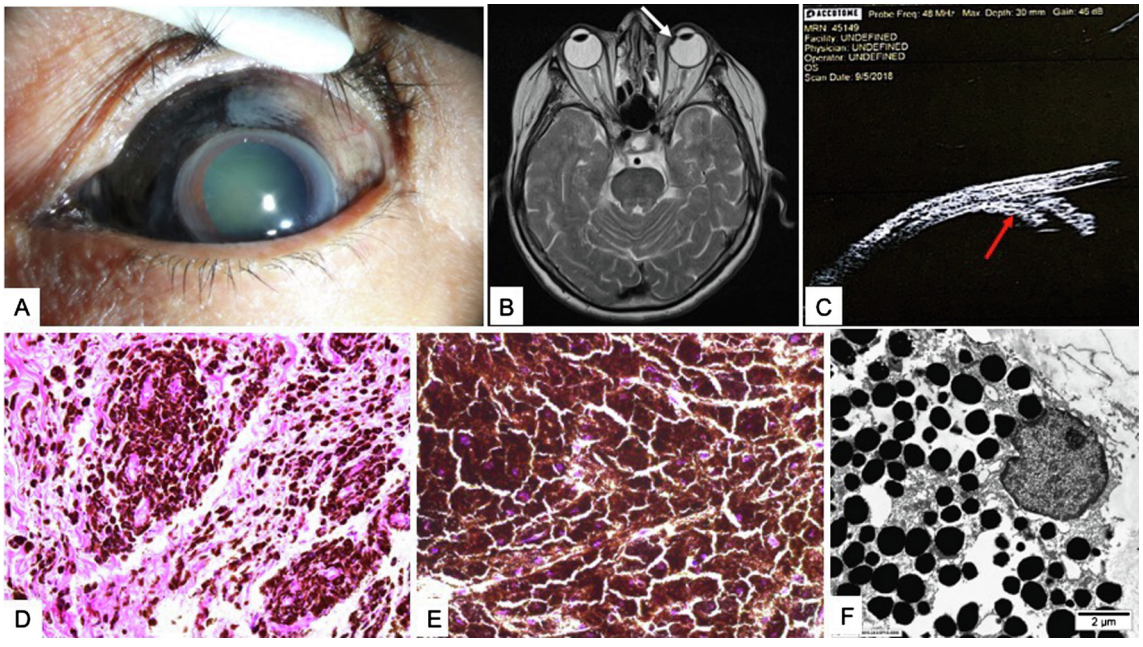

Fig. 1. (**A**) The clinical appearance of the left eye peri-limbal subconjunctival heavily pigmented area nasally at initial presentation. (**B**) Axial T2-weighted image showing a mass in the ciliary body area interpreted as a possible scleral tumor. (**C**) The cilio-choroidal tumor as it appears by ultrasound. (**D**) Episcleral tissue in the first biopsy with numerous coarse pigment laden macrophages overlying the ciliary body mass (Original magnification × 200 Hematoxylin and Eosin). (**E**) Melanocytoma type I cells with uniform small nuclei and heavy pigment in the second excisional biopsy (Original magnification × 400 Hematoxylin and Eosin). (**F**) The electron microscopic appearance of the melanocytoma cells with giant mega-melanosomes and slightly indented nucleus (Original magnification × 12,000).

Discussion

CB melanocytoma is a rare slowly growing tumor that is considered to be benign but may exhibit fast growth, may resemble melanoma, and can be associated with ocular morbidity such as melanocytomalytic glaucoma, cataract, extrascleral extension, and rarely necrosis with bone formation as well as iridodialysis.[4–7]

The clinical diagnosis of CB melanocytoma can be challenging and often resulting in enucleation with the presumed diagnosis of a melanoma.[8] However, nowadays better diagnostic techniques may help in reaching the proper diagnosis and planning for better management such as UBM, anterior segment optical coherence tomography (OCT), MRI and fine needle aspiration biopsy (FNAB).[5,9,10] For anterior segment tumors, UBM has been found to be more informative and thus superior to anterior segment OCT especially for pigmented larger tumors involving the CB such as melanocytoma.[10] In our case, the clinical impression and the UBM were highly suggestive of a melanoma with an extra-scleral extension. The tissue diagnosis of the conjunctival tissue initially excised has aided in the diagnosis of a melanocytoma in contrary to the MRI, which was supportive of a malignant scleral tumor.

Histopathologically, the tumor is composed of either Type I or Type II. Type I cells are typically large, polyhedral with abundant cytoplasm, giant melanosomes and small round nuclei as in our case.[11] Type II cells are more elongated with smaller melanin pigments. Necrosis due to the high metabolism and poor vascularity has been described in addition to calcification and cavitary cystic changes.[6,12] Regarding the use of immunohistochemical (IHC) staining, the tumor cells exhibit positive staining with melanocytic markers (Melan-A and HMB45) and show variable number of pigment-laden macrophages, which were predominant in the initial scleral tissue biopsy of our case. EM findings also differ between the 2 types with a corresponding smaller cells, less coarse melanin, and prominent indentations of the nuclear membranes in Type II cells versus giant melanosomes in Type I, which was appreciated in our case by EM thus supporting our diagnosis.[13]

Demographically, Lo Russo and co-authors summarized a total of 40 cases of CB melanocytoma including 10 cases of their own and concluded an average age of 47 years, 65% prevalence in females, and higher occurrence in white patients (80%) compared to black (10%).[13] Following their review in 1999, several sporadic cases have been reported in the English-written literature, the majority of which are describing the challenging clinical diagnosis and the impact of this on treatment modalities other than enucleation which includes local excision, iridocyclectomy with lamellar sclera flap and brachytherapy.[14,15,16,17] Our patient was older than the age described, however the lesion being in the CB might indicate a long-standing lesion that had a recent rapid episode of growth leading to her presenting symptoms. She underwent superficial debulking of the tumor and is currently being followed up. She has been stable since her surgery; however, we do not have access to her most recent clinical photos.

In terms of prognosis, the tumor is not expected to metastasize. However, in other locations (other than the CB), the tumor has been reported to be associated with or to transform into malignant melanoma, thus such patients should be closely followed up.[2,18]

In conclusion, CB melanocytoma has variable presentation, and has often been misdiagnosed as a melanoma. Diagnostic methods including UBM, biopsy, and EM should be utilized to differentiate between the two and too plan the proper treatment aiming to save the globe as in our case.

Conflict of interest

The authors declared that there is no conflict of interest.

References

1. Howard GM, Forrest AW. Incidence and Location of Melanocytomas. *Arch Ophthalmol* 1967;**77**():61–6.
2. Shields JA, Shields CL, Eagle RC. Melanocytoma (Hyperpigmented Magnocellular Nevus) of the Uveal Tract. *Retina* 2017;**27**:730–9.
3. Zimmerman LE. Melanocytes, melanocytic nevi and melanocytomas. *Invest Ophthalmol* 1965;**4**:11–41.
4. Muiños Diaz Y, Saornil Alvarez MA, Mẽndez Diaz MC, Blanco Mateos E, Becerra Aguiar E, Esteban Casado MR, Pastor Jimeno JC. Symptomatic ciliary body melanocytoma: melanocytomalytic glaucoma. *Arch Soc Esp Oftalmol* 2004;**79**(10):511–4.
5. Mohamed MD, Gupta M, Parsons A, Rennie IG. Ultrasound biomicroscopy in the management of melanocytoma of the ciliary body with extrascleral extension. *Br J Ophthalmol* 2005;**89**:14–6.
6. Hiscott P, Jean Campbell R, Robertson DM, Damato B. Intraocular melanocytoma in association with bone formation. *Arch Ophthalmol* 2003;**121**:1791–4.
7. Kim M, Lee SJ. Melanocytoma of the ciliary body misdiagnosed as iridodialysis. *Clin Ophthalmol* 2014;**8**:1051–3.
8. Odashiro M, Odashiro A, Leite L, Melo M, Odashiro P, Miiji L, Odashiro D, Fernades P, Burnier Jr M. Melanocytoma of ciliary body and choroids simulating melanoma. *Pathol Res Pract* 2010;**206**:130–3
9. El-Harazi SM, Kellaway J, Font RL. Melanocytoma of the ciliary body diagnosed by fine-needle aspiration biopsy. *Diagn Cytopathol* 2000;**22**(6):394–7.
10. Bianciotto C, Shields CL, Guzman JM, et al. Assessment of anterior segment tumors with ultrasound biomicroscopy versus anterir segment optical coherence tomography in 200 cases. *Ophthalmol* 2011;**118**:1297–302.
11. Al-Hinai A, Edelstein C, Burnier Jr MN. Unusual case of melanocytoma. *Can J Ophthalmol* 2004;**39**:461–3.
12. Capeans C, Pineiro A, Blanco MJ, et al. Ultrasound biomicroscopic findings in a cavitary melanocytoma of the ciliary body. *Can J Ophthalmol* 2003;**38**(6):501–3.
13. LoRusso FL, Boniuk M, Font RL. Melanocytoma (Magnocellular Nevus) of the ciliary body: report of 10 cases and review of the literature. *Ophthalmology* 1999;**107**:795–800.
14. Wen-bin W, Wen-li Y, Shi-min HU, Bin LI. Local excision of ciliary body tumors: a long-term prospective cohort study in China. *Chin Med J* 2008;**121**(21):2152–6.
15. Lee CS, Kim DK, Lee SC. A case of ciliary body melanocytoma presenting as a painful iris mass. *Korean J Ophthalmol* 2010;**24**(1):44–6. https://doi.org/10.3341/kjo.2010.24.1.44.
16. Haruta M, Miyamoto K, Horii T, Hirata Y, Yoshimura N. Iridociliary melanocytoma with suspected pulmonary metastasis. *Jpn J Ophthalmol* 2005;**49**(6):545–6. https://doi.org/10.1007/s10384-005-0235-7.
17. Shanmugam MP, 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**:966–8.
18. Alkatan HM, Al-Rashed S. Choroidal melanoma in association with juxtapapillary melanocytoma. *Can J Ophthalmol* 2008;**43**(2):250–1. https://doi.org/10.3129/i08-019.