Multifocal choroidal melanoma in oculodermal melanocytosis in an Asian Indian

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A 51-year-old Asian Indian male with right oculodermal melanocytosis presented with a blurred visual acuity of 20/40. Upon fundus examination, he was discovered to have two independent pigmented choroidal melanomas in the temporal juxtapapillary region and inferiorly, in the region of trace clinical sectoral choroidal melanocytosis. The patient underwent enucleation, and on histopathology the two tumors were found to be in the area of choroidal melanocytosis, in continuity with each other. Multifocal choroidal melanoma is an exceedingly rare diagnosis. Ocular melanocytosis is a known predisposing factor for both unifocal and multifocal melanomas. Lifelong monitoring for uveal melanomas must be done in all patients with ocular melanocytosis.

Key words: Choroid, eye, melanocytosis, melanoma, multifocal, multinodular, uvea

Choroidal melanoma is a serious, life-threatening malignancy and generally manifests as a unilateral, unifocal tumor.[1] Rarely, this malignancy can be bilateral or multifocal.[1-6] In such cases, a search for causes of multifocal tumors should be performed including undetected primary cutaneous melanoma with choroidal metastasis or underlying ocular melanocytosis, a condition known to predispose to unifocal melanoma and occasionally multifocal tumors.[1] Ocular melanocytosis can be diffuse, affecting the entire uvea and sclera, or it can be sectoral, affecting only a limited portion of the uvea, with or without complementary sectoral scleral pigmentation.[7] Herein, we report a patient with ocular melanocytosis presenting with a multifocal melanoma which was histopathologically confirmed to be in continuity with each other.

Access this article online

Quick Response Code:

Website: www.ijo.in

DOI: 10.4103/ijo.IJO_1773_18

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Received: 24-Oct-2018 Revision: 04-May-2019
Accepted: 11-Nov-2019 Published: 22-Nov-2019

Case Report

A 51-year-old Asian Indian man noted 1 month of blurred vision with photopsia and floaters. There was no other significant medical or family history. External examination showed the presence of periocular melanocytosis [Fig. 1a]. The visual acuity was 20/40 in the right eye and 20/20 in the left eye. Intraocular pressure was 13 mmHg in both eyes. The anterior segment was unremarkable in the left eye, while the right eye revealed sectoral scleral melanocytosis [Fig. 1b]. There was no temporal fossa, palate, or iris melanocytosis.

Funduscopically, the left eye was normal. Evaluation of the right eye fundus revealed two separate pigmented choroidal tumors [Figs. 1c and d]. The smaller tumor was juxtapapillary and temporal in location and measured 4 × 4 mm in basal diameter and 2.8 mm in thickness. The larger inferior tumor measured 13 × 11 mm in base and 9.2 mm in thickness. Both tumors demonstrated signs of tumor activity including prominent overlying orange pigment and subretinal fluid. Though quite subtle, trace sectoral choroidal melanocytosis was noted surrounding the two tumors. B-scan ultrasonography documented both tumors with acoustic hollowness and without extrascleral extension.

Thorough systemic evaluation with imaging of the chest and abdomen and blood tests revealed no evidence of any other primary or secondary malignancy. The findings were consistent with unilateral primary multifocal choroidal melanomas arising in sectoral choroidal melanocytosis of the right eye. The treatment options included enucleation or plaque radiotherapy. After appropriate informed consent, the patient underwent enucleation with a primary silicone orbital implant.

Gross examination of the enucleated eye revealed patchy grey pigmentation of the episclera. On histopathology, the choroid temporal to the optic disk [Fig. 2a] and in the inferior quadrant was thickened by a dense population of darkly pigmented plump and benign dendritic melanocytes, characteristic of choroidal melanocytosis [Fig. 2b]. There was no evidence of melanocytosis in the remainder of the choroid, iris, and ciliary body stroma. There were two elevated choroidal tumors [Fig. 2c and d], both of which were composed of pigmented epithelioid melanoma cells and neither tumor showed scleral or retinal invasion. On careful serial sections, both tumors were confirmed to be within the area of flat choroidal melanocytosis with a steep indentation and base continuity in the transition between the two [Fig. 2d].

Discussion

Choroidal melanoma generally appears as a solitary lesion in a dome-shaped or mushroom-shaped configuration.[1]

Cite this article as: Rao R, Honavar SG, Mulay K. Multifocal choroidal melanoma in oculodermal melanocytosis in an Asian Indian. Indian J Ophthalmol 2019;67:2089-91.

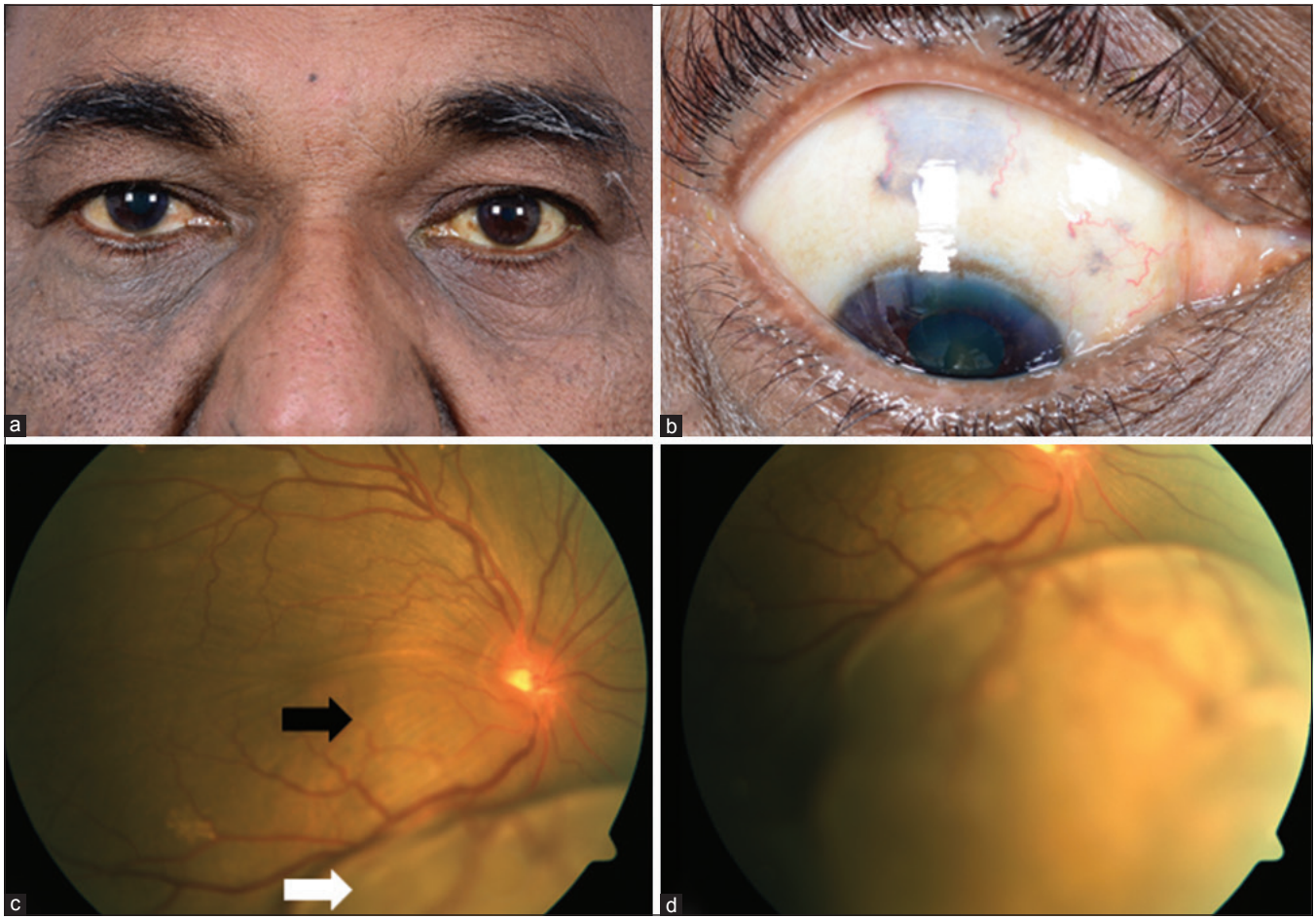

Figure 1: External photographs of a 51-year-old male show (a) right periocular melanocytosis with (b) blue-grey episcleral pigmentation. Fundus photographs reveal (c) two separate pigmented choroidal melanomas (black and white arrows) with overlying orange pigment and subtle sectoral choroidal melanocytosis, with the smaller juxtapapillary tumor measuring 4 × 4 mm in basal diameter and (d) larger inferior tumor measuring 13 × 11 mm in basal diameter

Bilateral choroidal melanoma is an extremely rare occurrence.[1] Multifocal choroidal melanoma is also exceedingly rare. Honavar *et al*. reported two discrete choroidal melanomas in a patient with ocular melanocytosis and estimated that 1 in 160,000 patients with unilateral ocular melanocytosis might develop two melanomas.[1] Furthermore, on the basis of chance, they calculated that patients with two melanomas in the same eye have approximately 1000-fold greater likelihood of demonstrating related melanocytosis, compared with the general Caucasian population.[1]

The occurrence of choroidal melanoma in Asian Indians and other pigmented races is very rare, with the Caucasians being three times more prone to the disease than Asian Indians.[6,7] In a review of 103 eyes of 103 Asian Indian patients with uveal melanoma over 14 years, Biswas *et al*. found only two eyes (2%) with oculodermal melanocytosis.[6] There were no eyes with multifocal melanoma in this large series.[6] In a yet another clinicopathologic study of Asian Indians with uveal melanoma, there was not a single case with multifocal melanoma.[7] In addition, none of the 76 eyes in the study was reported to have ocular melanocytosis.[7] In the current case, the eye demonstrated a clinically multifocal melanoma in the area of subtle melanocytosis in the choroid.

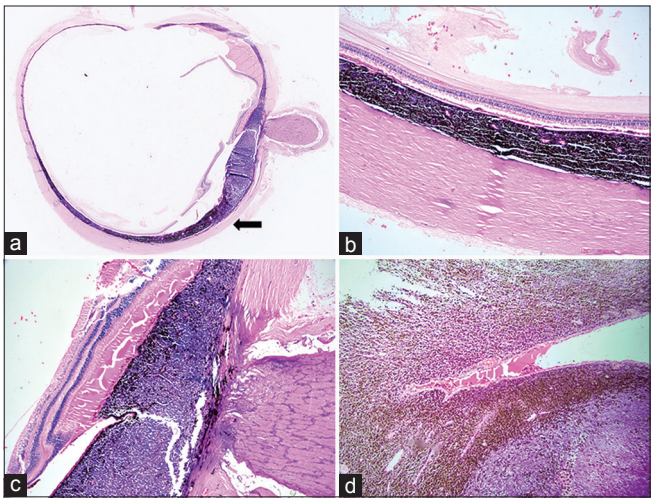

Figure 2: Histopathology of the enucleated right eyeball demonstrates (a) the flat thickened choroid (black arrow) due to melanocytosis. (b) Melanocytosis component on HE ×400. Two distinct melanomas composed of pigmented epithelioid melanoma cells with (c) the larger inferior tumor arising in the area of choroidal melanocytosis on HE ×400 and (d) the base continuity with a steep indentation between the two tumors on HE ×200

Multifocal uveal melanoma with ocular melanocytosis has been described in a spectrum of patient ages with the youngest reported child at age 6 years.[1-5] Oculodermal melanocytosis is a congenital pigmentary condition that can affect the periocular skin, sclera, uvea, orbit, palate, and meninges.[1,8] This condition can manifest as a diffuse pigmentary abnormality, affecting all quadrants of the eye in a homogeneous fashion, or it can affect only a sector, displaying pigment in a limited portion of the eye.[8] Oculodermal melanocytosis is known to carry a serious risk for melanoma, particularly uveal melanoma and, less commonly, orbital and meningeal melanoma.

Regarding multifocal uveal melanoma, there are reports with documented underlying ocular melanocytosis.[1,3-5] Shields *et al*. reported subtle melanocytosis in a child with two separate melanomas, seen only as a slight grey patch in one clock hour on the sclera.[4] Sectoral melanocytosis can be far more subtle than its diffuse counterpart but it should be routinely monitored as it poses a risk for uveal melanoma.[8] Rao *et al*. reported a case of multifocal melanoma where the patient was detected to have BAP-1 mutation and the authors suggested that BAP-1 mutation could result in multifocal choroidal melanoma.[9] The authors treated both the tumors with a single plaque.[9] In our case, although clinically there were two distinct choroidal melanomas, histopathologically there was continuity between the two tumors. Hence, if plaque brachytherapy is being planned for a patient with multifocal tumor, it might be pertinent to cover the intervening area in the radiation field to prevent future recurrences.

Patients with uveal melanoma arising from ocular melanocytosis carry an elevated risk for metastasis.[10,11] In an analysis of 7872 eyes with uveal melanoma, the authors found a 1.6 times greater risk for metastasis compared with those without melanocytosis.[10] This finding is further reinforced by a matched study of patients with uveal melanoma associated with ocular melanocytosis which demonstrated a two times higher rate of metastasis in those with melanocytosis.[11] Specifically, the 10-year rate of melanoma metastasis was 48% in eyes with melanoma and melanocytosis compared with 24% in eyes lacking melanocytosis.[10]

Conclusion

In summary, we describe a rare entity of unilateral multifocal melanoma arising in an eye with oculodermal melanocytosis in an Asian Indian. Ocular melanocytosis can be subtle and sectoral, often difficult to visualize. Ophthalmologists should be aware of the association of unifocal and multifocal melanoma with ocular melanocytosis.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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