Vitrectomy as a treatment modality in vitreous seeding secondary to ciliary body melanocytoma

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Melanocytoma is a locally invasive intraocular tumor usually located in the optic nerve head, iris, ciliary body and choroid. Melanocytoma can undergo necrosis and lead to pigment dispersion. We report a case of melanocytoma of the ciliary body with vitreous seeds filling the vitreous cavity. A sub conjunctival pigmented lesion was seen due to extra scleral extension of the tumor. The diagnosis of melanocytoma was confirmed by biopsy of the sub conjunctival lesion. Pars plana vitrectomy was performed to clear the vitreous cavity with good visual recovery.

Key words: Melanocytoma, pars plana vitrectomy, vitreous seeding

Melanocytoma is a benign, locally invasive pigmented intraocular tumor most commonly located in the optic nerve head. It is also reported to occur anywhere along the uveal tract, including iris, ciliary body and choroid.[1-4] Though melanocytoma has a stable course, it can undergo necrosis or rarely malignant transformation. Melanocytoma of the iris can undergo necrosis and lead to pigment dispersion resulting in melanocytic glaucoma.[5] Vitreous seeding from optic disc melanocytoma can present as floaters.[6] We report a case of melanocytoma of the ciliary body with vitreous seeds completely filling the vitreous cavity that was managed by pars plana vitrectomy.

Case Report

A 60-year-old lady presented with sudden onset diminution of vision in her right eye one month ago. She could appreciate hand movements close to face in the right eye and best corrected visual acuity (BCVA) was 20/80 in the left eye. Intraocular pressure was 20 mm of Hg in her right eye and 16mm of Hg in left eye. Slit-lamp examination of the right eye showed two oval shaped, elevated sub conjunctival lesions at 10 o' clock position 3mm posterior to limbus. The lesions were jet black in color [Fig. 1a]. Conjunctiva over the lesion was freely mobile with associated dilated episcleral vessels. Anterior chamber and iris were normal. Posterior sub capsular cataract (PSC) with fine pigments over the anterior capsule was noted. Gonioscopic examination of the right eye showed pigmented angles. There was no evidence of distortion of the angles or neovascularization. Posterior segment examination revealed dense pigment clumps in vitreous cavity. Fundus details were not seen. Ultrasound biomicroscopy (UBM) showed a heterogenous mass in the ciliary body in superotemporal region extending from 7 to 11 o'clock hour [Fig. 1b]. Ultrasonography of the right eye showed high reflective dot echoes in vitreous cavity [Fig. 1c]. On magnetic resonance imaging (MRI), the lesion was hyperintense on T1, hypointense on T2 and showed contrast enhancement [Fig. 1d]. Abdominal ultrasonography, liver function tests and chest X-ray were normal. Incisional biopsy of the sub conjunctival pigmented lesion was performed. Histopathological examination revealed melanin pigment. Melanin bleach failed to identify cellular elements. 3 weeks

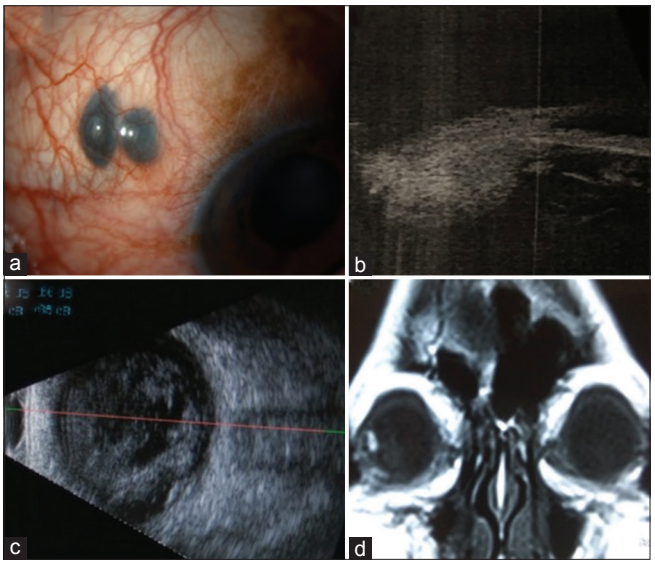

Figure 1: (a): Slit lamp examination showing 2 darkly pigmented nodular lesions with dilated episcleral vessels. Overlying conjunctiva was freely mobile. (b): Ultrasound biomicroscopy (UBM) showing a heterogenous mass in the ciliary body in supero-temporal region. (c): Ultrasonography of right eye showing high reflective dot echoes in vitreous cavity. (d): Magnetic resonance imaging (MRI) coronal section of the orbit through the lesion showing contrast enhancement

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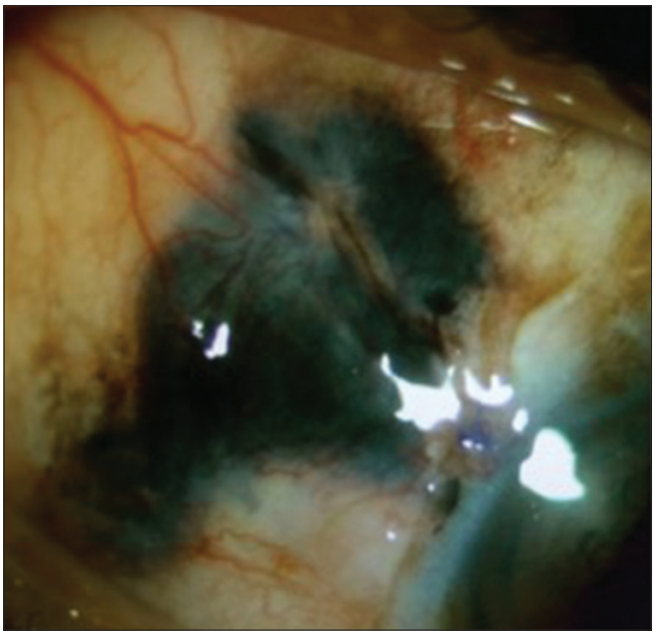

Figure 2: Color photograph 20 days after incisional biopsy, showing increase in size of sub conjunctival pigmented lesion with pigment dispersion

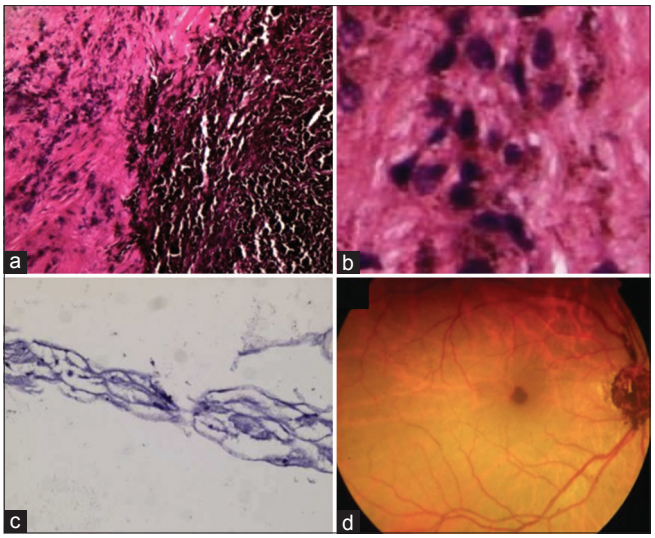

Figure 3: (a): Haematoxylin and Eosin staining of the extrascleral pigmented lesion showing sheets of densely pigmented cells in an eosinophilic stromal background. (b): High resolution image. (c): Melanin bleach of the vitreous sample showing ghosts of cells without prominent nucleoli. (d): Fundus photo at 3 month post-operative follow-up showing pigment clump over the disc and fovea

later, slight increase in size of the sub conjunctival pigmented lesion was noted [Fig. 2]. On repeat incisional biopsy, sheets of densely pigmented cells were seen in an eosinophilic stromal background [Fig. 3a and b]. On melanin bleach, cells were small, round to polygonal and bland appearing with scant cytoplasm, without prominent nucleoli. No malignant cells were seen. Clinical evaluation, imaging and histopathological examination were suggestive of a ciliary body melanocytoma with extra scleral extension and vitreous seeding. In view of PSC and dense vitreous seeding, phacoemulsification, intraocular lens implantation and 23 gauge pars plana vitrectomy was performed. On scleral indentation, a darkly pigmented ciliary body mass was noted in the superotemporal quadrant. Histopathological examination of the vitreous sample revealed pigmented cells arranged in clusters on a clean background. On melanin bleach features were consistent with melanocytoma and negative for atypical cells [Fig. 3c]. 3 months post-operatively, patient had a BCVA of 20/40 in right eye. Pigment clump was seen over the disc and fovea [Fig. 3d]. No change in the size of the lesion was noted.

Discussion

Melanocytoma is reported to occur most commonly at the optic nerve head,[1-3] but can be encountered less frequently in the ciliary body.[4,7] The diagnosis of ciliary body melanocytoma is difficult until it is large enough to be visible through the pupil or shows extrascleral extension. These changes may also suggest malignant transformation of the tumor.[8] Elevated intraocular pressure, changes in refractive error, increasing astigmatism, or development of cataract are other tell-tale signs. Melanocytoma is friable, discohesive tumor and can undergo spontaneous necrosis with pigment dispersion. Extra scleral extension of iridociliary melanocytoma represents passage of pigment-laden macrophages released by necrotic tumor through the emissary channel.[3] So, it should not necessarily be considered a sign of malignant transformation. Anterior uveal melanomas can also present as sub conjunctival pigmented lesions. So, a multidisciplinary approach including imaging and histopathological examination would be required to differentiate these locally invasive benign lesions from melanoma.

Conclusion

Seeding of cells from a necrotic tumor onto the iris surface and angle, results in increased intraocular pressure.[1,5,9] But vitreous seeding severe enough to limit visualization of fundus with normal intraocular pressure is rare and not reported in literature. Pars plana vitrectomy is a valuable option in clearing the media in such cases.

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.

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Conflicts of interest

There are no conflicts of interest.

References

1. Demirci H, Mashayekhi A, Shields CL, Eagle RC Jr, Shields JA. Iris melanocytoma: Clinical features and natural course in 47 cases. Am J Ophthalmol 2005;139:468-75.
2. Brownstein S, Dorey MW, Mathew B, Little JM, Lindley JI. Melanocytoma of the choroid: Atypical presentation and review of the literature. Can J Ophthalmol 2002;37:247-52.

3. Shields JA, Shields CL, Eagle RC Jr. Melanocytoma (hyperpigmented magnocellular nevus) of the uveal tract: The 34th G. Victor Simpson lecture. Retina 2007;27:730-9.
4. LoRusso FJ, Boniuk M, Font RL. Melanocytoma (magnocellular nevus) of the ciliary body: Report of 10 cases and review of the literature. Ophthalmology 2000;107:795-800.
5. Fineman M, Eagle RC Jr, Shields JA, Shields CL, De Potter P. Melanocytomalytic glaucoma in eyes with necrotic iris melanocytoma. Ophthalmology 1998;105:492-6.
6. Guo H, Li Y, Chen Z, Guo X. Vitreous seeding from a large optic disc melanocytoma. J Neuroophthalmol 2014;34:276-7.
7. Shammas HJ, Minckler DS, Hulquist R, Sherins RS. Melanocytoma of the ciliary body. Ann Ophthalmol 1981;13:1381-3.
8. Cialdini AP, Sahel JA, Jalkh AE, Weiter JJ, Zakka K, Albert DM. Malignant transformation of an iris melanocytoma. A case report. Graefes Arch Clin Exp Ophthalmol 1989;227:348-54.
9. Radcliffe NM, Finger PT. Eye cancer related glaucoma: Current concepts. Surv Ophthalmol 2009;54:47-73.