Brachytherapy, A viable option of globe salvage in treatment of large ciliary body melanocytoma

Mahesh P. Shanmugam, Manish Saxena, Rajesh Ramanjulu, Pradeep Tekwani

We report a case of large histopathologically proven melanocytoma of the ciliary body in a 15-year-old male, presented with rapid extraocular growth following incisional biopsy with scleral patch graft. We chose brachytherapy with Ruthenium 106 plaque over enucleation as the later was refused by the parents. The initial apical height of the tumor was 14.2 mm on ultrasonography. Two weeks after brachytherapy, the mass regressed to a size of 8.1 mm and 1 year later to 6.7 mm. This is the first case report showing the response of brachytherapy to ciliary body melanocytoma, which results in ocular and visual acuity salvation with considerable decreased in size of the tumor. The authors conclude that brachytherapy is an option in the management of non-resectable melanocytoma of the ciliary body.

Key words: Brachytherapy, ciliary body melanocytoma, plaque radiotherapy, ruthenium 106

Intraocular melanocytoma is an uncommon benign tumor accounting for about 0.6% of all intraocular tumors. These tumor are slow growing but can grow in one week as reported by Zimmermen. It has never been shown to metastasize. Melanocytoma of the ciliary body (CBM) is rare and may be circumscribed or diffused.

CBM can be resected, if confined to 3 clock hours of the ciliary body. Larger tumors may need enucleation. We herein report the use of brachytherapy to treat a large CBM. To the best of our knowledge, this is the first case report showing considerable reduction in size of the tumor after brachytherapy.

Case Report

A 15-year-old male presented with history of blurred vision of 6 months duration in his left eye. He had a BCVA of 20/20 and 20/120 in his right and left eyes respectively. IOP was normal in both eyes. Anterior segment of the left eye showed dilated episcleral vessel, shallow anterior chamber, peripheral anterior synchiae and heterochromia of the inferior iris with clumps of black pigment over the iris surface. A focal cataract involving the inferior part of the lens was present. A solid dark brown mass abutting the lens was seen behind the iris covering the inferior half of pupil [Fig. 1]. Fundus of left eye was normal; view of the inferior periphery, however, being blocked by the tumor.

Ultrasound bio microscopy of the left eye showed a homogeneous mass arising from ciliary body involving 6 clock hours inferiorly. Ultrasonography showed a homogeneous mass of 14.2 mm apical height with moderate to high amplitude on A scan, arising from the inferior half of the ciliary body and extending into the anterior vitreous cavity [Fig. 2]. There was no evidence of acoustic hollowness or any cystic changes. On magnetic resonance imaging (MRI) scan, the mass appeared hypo/isointense on T1- weighted images and hypo intense on T2- weighted images with minimal contrast related enhancement. Systemic workups comprising of liver function test, ultrasonography of the abdomen and x-ray of the chest were normal.

An incisional biopsy was performed as transscleral fine-needle aspiration biopsy (FNAB) using 30-gauge needle was inconclusive. In brief, a posteriorly hinged, partial thickness scleral flap of 5 mm/5 mm was fashioned over the tumor at 6’ o clock ciliary body region. An incisional biopsy of 3 mm/3 mm involving the inner sclera and partial thickness of the mass was performed. The hinged scleral flap was used to secure the biopsy site with multiple sutures.

Histopathological examination of the gross specimen showed black pigmented pieces of tissue, which on microscopy, appeared as heavily pigmented polygonal cells. Bleached sections showed bland nuclear morphology without increase in mitotic activity [Fig. 3], features that are suggestive of melanocytoma. Despite a water tight closure, pigmented tumor cells were seen invading the episcleral space after 1 month of biopsy. The scleral flap was reinforced with a scleral...
patch. On subsequent follow-up, the patient showed thinning of the patch graft with protrusion of the pigmented mass.

Considering the rapid extra ocular growth of the mass, we offered brachytherapy or enucleation of the eye as further management. The patient and his parents preferred eye salvage and brachytherapy with ruthenium 106; the source delivering 7500 cGy to the apex of the tumor was performed. The plaque was placed episclerally primarily over ciliary body but extended partly over the corneo scleral limbus to encompass the anterior chamber component of the tumor.

Two weeks after brachytherapy, the mass regressed to a size of 8.1 mm and 2 months later to 6.9 mm.

On last follow at 1 year after brachytherapy, his vision had decreased to counting finger due to progression of cataract. Anterior segment examination showed well integrated scleral patch graft and a considerably regressed melanocytoma measuring 6.7 mm on ultrasonography. [Fig. 4].

Discussion

CBMs are rare and in a review of 40 patients, the average age of diagnosis of the tumor was 47 years with a range of 3 months to 90 years. CBM may present as diffuse melanotic lesion of the iris resulting in heterochromia, the lesion representing either diffuse extension of the tumor into the iris or deposition of pigment released by the tumor on the iris surface. The diagnosis in our patient was a solitary melanocytoma with the differential being a melanoma. However, young age, presence of heterochromia, clusters of dark black pigment over the iris with a deeply pigmented mass were suggestive of a melanocytoma rather than a melanoma. Ultrasonography of the mass showed a solid mass but without the acoustic solidity of a melanoma as indicated by the absence of angle kappa or shadowing. However, ultrasonographic features in a melanoma may be variable, particularly a diffuse “ring” melanoma where typical ultrasound features such as angle kappa and shadowing may be absent. Hence an ultrasound examination cannot conclusively differentiate a melanoma from a melanocytoma. Similarly, an ultrasound bio microscopic examination will also show the presence of a solid mass but unlikely to differentiate between a melanoma and a melanocytoma.

Similarly, MRI cannot differentiate a melanoma from a melanocytoma with certainty and the MRI features of a melanocytoma vary depending on the degree of melanin pigmentation. Histopathological confirmation may rarely be necessary. Grossly, melanocytomas are heavily pigmented, ranging from dark brown to jet black. They are composed of large polyhedral cells with abundant cytoplasm, containing densely packed, large melanin granules. Bleached preparations can reveal a low nuclear-to-cytoplasmic ratio, central or para central nucleus with small or inconspicuous nucleoli, an distinct cellular outlines. These features and absence of mitoses were seen in the incisional biopsy specimen of our patient.

The main complications of large melanocytoma are cataract due to compression of the lens by the mass, as seen in our case. Secondary glaucoma may occur due to invasion of

Figure 2: Ultrasonography showing homogeneous mass of 14.2 mm apical height with moderate to high amplitude on A scan, arising from the inferior half of the ciliary body and extending into the anterior vitreous cavity

Figure 3: Histopathological examination revealed sheets of heavily pigmented polygonal cells, which showed bland cytological features after bleaching with KMnO4 without increase in mitotic activity
the dispersed melanin in the angle or in Schlemm’s canal, occlusion of the angle by the large tumor or melanin laden macrophages. CBM has also been associated with overlying scleral pigmentation, which represents passage of pigment laden melanophages released by necrotic tumor cells.

The management of CBM essentially requires observation, if there is no growth, secondary glaucoma or vision-threatening complications.

Plaque brachytherapy has not been employed as a primary mode of treatment for melanocytoma. There are however reports of melanocytoma being misdiagnosed as melanoma and treated with (125I) brachytherapy. These were choroidal melanocytomas that were ultimately enucleated due to non-response to brachytherapy. Considering the circumstances, we offered brachytherapy and treated the tumor with 7500 cGy to the apex considering the heavy pigmented nature of the tumor. As per the experimental study on human melanoma cell lines, degree of pigmentation has been shown to be inversely related to radiosensitivity of melanoma cells. Considering the high degree of pigmentation, we decided to treat it with the therapeutic radiation dose employed for a melanoma, despite the melanocytoma being a benign tumor. The encouraging response prompts us to consider brachytherapy as another viable option in the management of non-resectable melanocytoma of the ciliary body.

References
1. Howard GM, Forrest AW. Incidence and location of melanocytomas. Arch Ophthalmol 1967;77:61-6.
2. Zimmerman LE. Melanocytes, melanocytic nevi and melanocytomas. Invest Ophthalmol 1965;4:11-41.
3. Raichand M, Peyman GA, Juarez CP, Seetner AA, Sugar J, Goldberg MF. Resection of uveal melanocytoma: Clinicopathological correlation. Br J Ophthalmol 1983;67:236-43.
4. Biswas J, D’Souza C, Shanmugam MP. Diffuse melanotic lesion of the iris as a presenting feature of ciliary body melanocytoma: Report of a case and review of the literature. Surv Ophthalmol 1998;42:378-82.
5. 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.
6. Shammas HJ, Minckler DS, Hulquist R, Sherins RS. Melanocytoma of the ciliary body. Ann Ophthalmol 1981;13:1381-3.
7. Shields JA, Augsburger JJ, Bernardino V Jr, Eller AW, Kulczycki E. Melanocytoma of the ciliary body and iris. Am J Ophthalmol 1980;89:632-5.
8. Shetlar DJ, Folberg R, Gass JD. Choroidal malignant melanoma associated with a melanocytoma. Retina 1999;19;346-9.
9. Kinnarat E, Morandini R, Simon S, Hill HZ, Ghanem G, Van Houtte P. The degree of pigmentation modulates the radiosensitivity of human melanoma cells. Radiat Res 2000;154:497-502.

Cite this article as: 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.

Source of Support: Nil. Conflict of Interest: None declared.