

Choroid Melanoma – A Rare Case Report

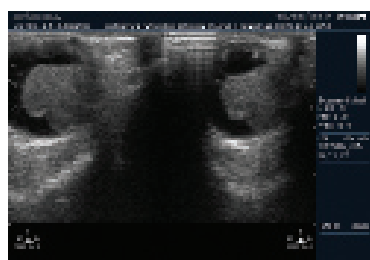
SAMARTH SHUKLA¹, SOURYA ACHARYA², MANISHA DULANI³

ABSTRACT

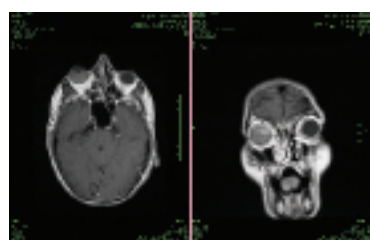
Uveal tract encompass melanomas of the iris, ciliary body and the choroid. The choroid melanomas are more frequent to occur in comparison to iris and ciliary body melanomas. Choroid melanoma though rare, is the most common occurring intraocular malignancy amongst the adults. Incidence of occurrence of choroid melanoma is around 20 per million cases per year globally. The uveal tract pigment producing melanocytes are the origin for the melanoma in the eye. Melanomas tend to commonly arise from sixth decade of age with increasing incidence with progressive age. We present a rare case choroidal melanoma, in a 65-year-old adult, who presented with three months history of headache and gradual diminution of vision, his investigations revealed clinical diagnosis of choroid melanoma, an enucleation was carried.

CASE REPORT

A 65-year-old male presented to the medicine OPD with chronic headache, right eye pain and photopsia along with gradual diminution of vision in the right eye of three months duration. His general and systemic examination was normal. Ophthalmic examination was carried out PL+, PR+ both eyes, however the Right eye vision on presentation was limited to counting fingers at 15 cm. The Intraocular pressure (IOP) of both eyes was within normal limits, the left side at 12mm Hg and the right eye IOP at 20mm Hg (close to upper limit). Binocular Indirect ophthalmoscopy of the right eye showed solid dark gray mass in the posterior segment (choroid) with intense brown pigmentation, occupying posterior third of the vitreous chamber along with mild retinal detachment observed at the peripheral rim of the nodular choroidal mass. Perimetric evaluation of left eye was within normal limits; however the right eye revealed visual field defects. 'B scan' ultrasound of right eye [Table/Fig-1] was carried to confirm the finding and assess the size of the mass and extent of intraocular involvement, there was low internal tumour reflectivity and significant change in angle kappa. The mass turned out to be arising in the Choroid with a dome shaped nodular growth of size 13 mm in elevation with a diameter of 20 mm occupying more than a third of the posterior segment of right eye. Further to evaluate the extent of tumour mass involvement as well as exact localization a MRI scan (with and without contrast) was carried out [Table/Fig-2,3]. MRI confirmed the mass to be choroidal growth



[Table/Fig-1]: Ultrasound B scan showing gray opaque mass in the choroid. Uveal excavation in posterior choroid melanoma

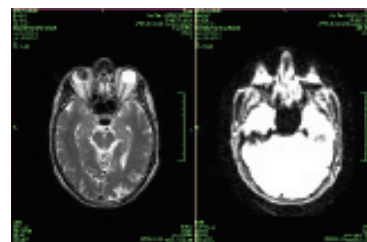


[Table/Fig-2]: MRI without contrast showing right side intraocular choroidal mass

Keywords: Ciliary body, Iris, Malignancy, Uveal

limited within the posterior chamber arising from the postero medial side of choroid of the right eye. Considering the imaging diagnosis and clinical findings a diagnosis of choroid melanoma was made categorized as T3a which was stage IIB according to the American Joint committee on cancer.

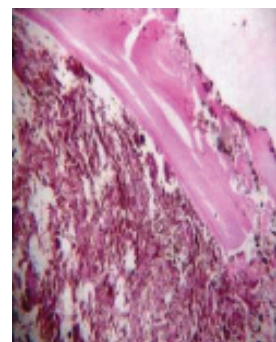
Modality of management involved enucleation of right eye [Table/Fig-4] and the enucleated eye and its mass were subjected to histopathological examination. Histopathological report confirmed Choroid Melanoma of (Mixed Spindle A and B type) [Table/Fig-5]. After enucleation the patient became asymptomatic for associated



[Table/Fig-3]: MRI with contrast showing right side intraocular mass arising from the posteromedial choroid, seen as a high-density image in T1 and as a low-density image in T2



[Table/Fig-4]: Enucleated Specimen of right eye, horizontal plane of section showing massive intraocular choroidal (pigmented) growth



[Table/Fig-5]: Shows H&E stained slide 10x (low power view) showing fascicular arranged cohesive spindle cells of A and B type (mixed variety), with evidence of brownish pigmentation, suggestive of choroidal melanoma (spindle mixed type)

symptoms like headache and pain. A follow up of 12 months was kept where the general and systemic examination was normal with no evidence of metastasis.

DISCUSSION

Melanomas of the uveal tract can be divided into the lesions of the anterior and the posterior tract, the anterior tract melanoma involve the iris whereas the posterior tract melanomas involve the ciliary body and the choroid layer. Malignant melanomas of the uvea are frequented more often in the choroid and the ciliary body in comparison to the iris. Melanomas are highly malignant epithelial cancers [1]. Melanomas tend to commonly arise from 70 years of age, however no age is spared, melanomas can occur in adolescents, children and rarely even in neonates [2,3]. Clinically the presentation of choroidal melanomas is variable. In general anterior choroidal melanomas have a delayed presentation because of slow growth however clinical signs and symptoms can present earlier. Patients of choroidal melanoma usually present with blurring of vision. Patient may experience painless and progressive visual field loss as the peripheral melanoma enlarges. Floaters and at times 'balls of light' are experienced by subjects in case of necrosis of tumour or haemorrhage in the adjoining areas. Severe pain may be experienced with impingement of tumour mass on ciliary nerves or due to acute angle closure glaucoma. Not infrequently the patient remains asymptomatic until the tumour has grown sufficiently to become necrotic and produce complications such as endophthalmitis, massive intraocular haemorrhage, and/or secondary glaucoma. Choroid layer being devoid of lymphatics hence majority of the choroidal melanomas spread by haematogenous route mainly to the liver [4].

Choroidal melanomas have variable pigmentation from being highly pigmented to being amelanotic in appearance. The choroidal melanomas irregular, slate-gray, solid, initially present as dome shaped subretinal nodular circumscribed mass. With more expansive behaviour they tend to form irregular bilobular to multilobular configurations, some of the melanomas tend to become mushroom shaped after breaching the Bruch's membrane. Though rarely but uveal melanomas can spread diffusely through the sclera canals into the orbit. Traditionally on histology the choroid melanoma has been categorized into three divisions namely spindle type A, spindle type B and epitheloid type.

The modified Callender's classification of uveal melanomas has four categories [5]:

1. Spindle cell type tumours comprising 45% of all choroidal melanomas.
2. Pure epitheloid cell Melanomas 5% (rare occurrence).
3. Mixed cell melanoma 45% (comprising of spindle cell and epitheloid cell types).
4. Necrotic melanoma 5% (predominant cell type unrecognizable).

Based on an extensive study carried on 400 cases at Armed Forces Institute of Pathology by Shields et al., enlisted the following lesions which may clinically resemble melanoma of the posterior uveal (pseudomelanoma), they include choroid nevus (26.55), disciform degeneration (12.5%), peripheral disciform degeneration (11%), congenital hypertrophy of retinal pigment (9.5%), choroidal haemangiomas (8%) [6]. Diagnosis of nevi has to be particularly kept in mind, as they may be at times be propagators giving rise

to malignant melanomas. Microscopically certain differentials for malignant melanoma of the choroid have to be kept into mind for evaluation, foremost is the spindle cell nevus which is extremely difficult to differentiate from pure spindle type A cell choroidal melanoma. Metastatic carcinoma and metastatic melanoma have to be differentiated from primary intraocular melanoma specially the epitheloid variant. Besides certain spindle cell variants (amelanotic types) may be difficult to distinguish from uveal neurofibroma and schwannomas. Immunohistochemically, malignant melanomas are reactive for S-100 protein, HMB-45, and Mart-1 (Melan-A) [7].

Iris melanomas have the best prognosis whereas the ciliary body melanomas have the worst prognosis. Iris melanomas are the least common. The anterior uveal tract melanomas have better prognosis which has been attributed to their dormant behaviour with slow growth and restricted field of spread as compared to their posterior tract counterparts which are more aggressive and metastasize early and are detected much later. The overall prognostication of malignant melanoma of the uvea is based on several factors; however, the malignant melanoma can be said have an intermediate prognosis, mortality being close to 50%, 15 years after enucleation [8].

Present day management trend generally centers on cautious conservative maximum vision sparing approach. Tumours of the posterior segment not readily visible of size less than 2.0-2.5mm in elevation and <10mm in diameter should be left for observation, sequential photograph and ultrasounds should be carried out to keep follow up on size. Brachy therapy for tumour less than <10mm elevation and <20mm in diameter supplemented with transpupillary thermotherapy, external beam irradiation for tumours unsuitable for brachytherapy, segmental iridectomy can be used for iris melanomas, a difficult approach but conservative can be transscleral local resection as well as stereotactic radiosurgery and finally larger tumours involving the entire globe require enucleation. Exenteration is carried out in case of wide spread orbital involvement. Systemic chemotherapy are only advocated in case of positive distant metastasis [9].

CONCLUSION

Malignant melanomas of the uvea are frequented more often in the choroid and the ciliary body in comparison to the iris. Symptomatology is extremely variable with a choroid mass. They can arise de novo or may arise in existing precursor lesion. Treatment in current scenario is preferably conservative and should be tailored to the individual patient's symptomatology.

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PARTICULARS OF CONTRIBUTORS:

1. Professor, Department of Pathology, J N Medical College, DMIMS Univ, Sawangi (Meghe), Wardha, Maharashtra, India.
2. Professor, Department of Medicine, J N Medical College, DMIMS Univ, Sawangi (Meghe), Wardha, Maharashtra, India.
3. Professor, Department of Ophthalmology, J N Medical College, DMIMS Univ, Sawangi (Meghe), Wardha, Maharashtra, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:

Dr. Samarth Shukla,

Professor, Department of Pathology, J N Medical College, DMIMS Univ, Sawangi (Meghe), Wardha-442004, Maharashtra, India.

Email: samarth21174@gmail.com

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