Ocular Melanoma: A Rare Entity

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ABSTRACT

Malignant melanoma of eyeball is a rare entity. The overall incidence of malignant melanoma is 3%–7% per year worldwide. Early detection and meticulous treatment planning and its execution are challenge to surgeons. A 38-year-old male presented with loss of vision in one eye, and after investigating, it turned out to be malignant melanoma of the eyeball. He was treated surgically by enucleation of the eyeball followed by prosthetic rehabilitation.

Key words: Enucleation, malignant, melanoma, uvea

INTRODUCTION

Malignant melanoma of the eyeball is a rare entity. The overall incidence of malignant melanoma is 3%–7% per year worldwide.¹ Among various types of malignant melanoma, ocular melanoma is the second most common type after cutaneous melanoma, with incidence being only 3.7%. In spite of being the second most common, it is still rare.² Uvea is the most common site for this malignancy. Various sites of origin of ocular melanoma are ciliary body, choroid, uvea, iris, and retina. Diagnosis of such condition is very challenging, because there are no obvious signs and symptoms at early stage. Early diagnosis and management is a key factor in the treatment of malignant melanoma to avoid distant spread and grave prognosis owing to the systemic spread of disease.³ Here, we have presented a rare case of uveal melanoma, which was treated surgically and further prosthetic rehabilitation of the eyeball was done with regard to psychosocial well-being of the patient.

CASE REPORT

A 38-year-old gentlemen presented with a chief complaint of loss of vision in the right eye for 2 months. To start with, there was initially blurring of vision in the right eye, which was neglected by the patient and later he experienced complete loss of vision in that eye. There was no associated pain, swelling, or any other significant signs and symptoms. On physical examination, there was no proptosis or enophthalmos. Intraocular pressure (IOP) of both eyes was within normal limits with right eye pressure being 20 mm of Hg, which is near to the highest limit of IOP. There was no perception of light in the right eye. The left eye was essentially normal with 6/6 vision. Magnetic resonance imaging (MRI) of orbit revealed a well-defined sharply demarcated soft-tissue lesion measuring 1.2 cm × 1.1 cm × 1 cm arising from the right lateral margin of uveal coating of the right eye globe. It showed bright signal on T1 and low signal on T2 with avid enhancement in postcontrast scan [Figures 1 and 2]. No extraocular extension of the lesion with clear retro-orbital fat was noted. Fine-needle aspiration cytology was suggestive of malignant melanoma. Positron emission tomography–computed tomography (PET-CT) scan revealed no distant metastasis and focal uptake near chorioretinal

Figure 1: Axial view magnetic resonance imaging eye
membrane measuring 1.2 cm × 1.1 cm × 1 cm with maximum standard uptake value of 4.8. Retinal detachment was also noted. Final diagnosis of malignant melanoma of the eyeball–uveal origin was made and surgical enucleation of the eyeball under local anesthesia was planned. Temporary obturator was given to the patient immediate postoperatively, and 15 days later, he was fitted with eyeball prosthesis. Final histopathology revealed malignant melanoma of the eyeball with conjunctiva and optic nerve being free of tumor. The patient received local radiotherapy following enucleation and he had no recurrence up to follow-up period of 1 year.

**DISCUSSION**

Ocular melanoma is a rare variety of neoplasm found in the Indian population (3.7%). Among various types of ocular melanoma, the incidence of uveal melanoma is 87.3%.\(^4\) We have reported a rare case of ocular melanoma of uveal origin. Etiology of such neoplasm remains unclear. It usually occurs after the age of 50 years; our patient was 38 years of age. Usually, in 30% of cases, it is detected incidentally on ophthalmic examination.\(^5\) Ocular melanoma is said to remain asymptomatic in many of patients unless it becomes large enough to cause visual disturbances. The most common and first symptom is blurring of vision. Other common symptoms are visual field defects, photopsia, irritation, and pain,\(^6\) while our patient presented with complete loss of vision in the right eye. Melanomas are said to arise either from anterior compartment or posterior compartment. Anterior compartment lesions arise from iris whereas posterior compartment lesions arise from choroid and ciliary body. Our case presented with anterior compartment melanoma–uveal melanoma. MRI is an important imaging tool in the diagnosis of such conditions depicting the location and extent of the tumor. Other diagnostic imaging tools are ultrasound of the eye, B scan, and CT scan, but are of minimal diagnostic value. We performed MRI of the eye, which depicted exact location of the lesion and gave clear image about confinement of tumor to eyeball and also described its origin from the uveal membrane. By the time disease is diagnosed, many of patients are metastatic. Liver (89%) is the most common site of metastasis from ocular melanomas, followed by lung (29%) and then brain (17%).\(^7\) PET-CT scan was done to rule out distant metastasis and the patient had no evidence of distant metastasis. The incidence of cervical nodal metastasis is also rare in such conditions. Various available treatment modalities include enucleation of the eyeball, orbital exenteration, charged particle beam irradiation, brachytherapy, transscleral local resection, and transpupillary thermotherapy. Prognosis of ocular melanoma is poor after metastasis. The overall survival is only 6–8 months after detection of metastasis.\(^8\) Selection of appropriate treatment depends on site size and extent of the tumor, condition of opposite eye, age, and general condition of the patient. As the size of the lesion was large, it was in confines of the eyeball and there was no extraconal spread. Furthermore, there was no significant

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**Figure 2:** Coronal view magnetic resonance imaging eye

**Figure 3:** Eye speculum in situ for lid retraction

**Figure 4:** Eyeball specimen
vision left in that eye. Considering the prognosis of ocular melanomas, age of the patient, status of vision, and extension of tumor,\(^9\) we chose surgical enucleation of the eyeball as a choice of treatment [Figures 3 and 4]. Therapeutic radiation of 3DCRT 60 Gy/30 # was given to local region. Restoration of surgical defect is usually ignored and it provides mental trauma to the patient [Figure 5]. Prosthetic rehabilitation of the patient was done which restored the major facial defect and contributed to psychosocial well-being of the patient.

**CONCLUSION**

Early diagnosis, ruling out of distant metastasis, selection of proper treatment modality, and rehabilitation of patient are key in the successful management of patients to provide functional, social, and psychological rehabilitation with such rare conditions of ocular melanoma.

**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.

**Disclosure**

This material has never been published and is not currently under evaluation in any other peer reviewed publication.

**Ethical approval**

The permission was taken from Institutional Ethics Committee prior to starting the project. All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

**Informed consent**

Informed consent was obtained from all individual participants included in the study.

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