Choroidal Melanoma: A Rare Case Report

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Abstract

Although choroidal melanoma is the most common primary malignant tumor of adults in western countries, it is rare in the Indian population. We report a case of malignant melanoma of the choroid in a 48 years old female who presented with complete loss of vision in the right eye. On examination, she had no light perception and a dark brown elevated lesion was seen on ophthalmoscopy. USG B-Scan, CT scan and histopathology confirmed the diagnosis. Patient underwent enucleation as all useful vision was lost.

Key Words: *Choroidal Melanoma, Enucleation.*

Introduction

Worldwide, choroidal Melanoma is the most common primary malignant intraocular tumor, having metastatic potential. Global incidence of uveal melanoma ranges from 5.3 to 10.9 cases per million populations. These lesions are very uncommon in the Indian population. It has a predilection for light skinned caucasians and is rare amongst the black population. Average age at which the disease is seen is 50 years, with rising incidence in older population. Sex ratio is equal in both men and women. Etiology of choroidal melanomas remains obscure. Environmental factors like ultra violet light exposure and cigarette smoking may play a role. A rare possibility of genetic predisposition has also been suggested. It is always primary, single and unilateral. It arises from the outer layers of the choroid and is composed of melanocytes; pigment containing and pigment producing cells.

Case report

A 48 year female presented with complete progressive loss of vision in the right eye over a period of one year. On examination visual acuity in the left eye was 6/6. There was absence of light perception in the right eye. Anterior segment did not reveal any abnormality. Intra ocular pressure was 17.3 mm of Hg in both eyes. Distant direct ophthalmoscopy showed multiple floaters behind the lens. Direct ophthalmoscopy revealed a large dark grayish brown mass occupying two thirds of the vitreous cavity in the temporal part of the retina involving the macula. Optic disc was faintly seen and appeared pale. Surface of the mass was convex and rough with tiny multiple red areas. Surrounding retina was detached. Left eye showed no abnormality. B-scan ultrasonography showed a hyperechoic, well defined growth deeply embedded and arising from the choroid on the temporal side of eyeball with a collar button shape configuration and nature. Mass measured 0.95 x 0.76 cm. Retinal detachment was noted along the mass. Colour Doppler showed moderate increase in vascularity. These features were typically suggestive of melanoma of the choroid. CT scan revealed no extra ocular extension. USG abdomen and chest X - ray did not show any evidence of distant metastasis. Considering the present visual status and risk of future complications and metastasis; enucleation of the right eye was performed after informed written consent from the patient. Postoperative histopathological examination of the enucleated eyeball confirmed the clinical and radiological diagnosis. The tumor mass was composed of malignant melanocytes with melanin pigment, eosinophilic nucleoli, pleomorphism and medium mitotic activity with both spindle shaped and epitheloid cells; all indicating an intermediate prognosis.

Discussion

Choroid is the most common site of uveal malignant melanoma. The hallmark of the choroidal melanoma is

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a collar button or mushroom shaped, single, unilateral and primary growth arising from the posterior pole with significant elevation of the lesion, serous detachment of the adjacent retina, pigment epithelial proliferation and foci of orange pigmentation on the surface of the tumor (lipofuscin).

Choroidal nevus, haemangioma and metastatic carcinoma form the differential diagnosis. A Nevus is relatively flat, avascular and has a regular structure. It is found in the first decade of life and shows high internal reflectivity on USG, B-scan. On the other hand metastases are bilateral multiple avascular and have an irregular structure with medium to high internal reflectivity on USG. The primary focus is located elsewhere. Haemangioma shows lack of black pigmentation and has less internal blood flow than melanoma.

Ancillary tests like USG, fundus fluorescein angiography, colour Doppler and CT scan should be applied particularly in patients with hazy media. Microscopic identification of the major cell types is useful in assigning prognosis in a specific case. Liver is by far the most common site of metastasis. It is encountered if primary tumor is composed of epitheloid cells or mixed cell type. Metastases within the liver develop within 15 years of initial diagnosis and treatment and it is therefore mandatory to follow these cases regularly due to the possibility of sub clinical metastasis at the initial presentation which may manifest later. The disease is usually fatal within 5 years, if not eradicated and death occurs within one year of detection of metastasis. Chemotherapy given as a palliative treatment may prolong the lifespan. Uveal melanomas typically metastasize before they become symptomatic. Enucleation probably does not, therefore, influence survival but should be carried out as soon as possible to save those cases in which it has not occurred (Manschot and Van Peperzeel, 1980).

Current clinical management option for uveal melanoma

In the past, observation or enucleation was the only option available to ophthalmologists. Today due to improved understanding of uveal malignant melanoma an ophthalmologist can offer multiple modalities of treatment including brachytherapy, charged partial beam irradiation, local resection or thermal therapy. Immunotherapy and gene therapy are also under research and have significant potential. Which treatment modality is to be applied to which patient depends not only the size of the tumour but also upon the site, extent and apparent activity of the tumor. The state of the fellow eye also needs to be considered.

Enucleation: For a longtime, enucleation remained the mainstay of therapy for uveal melanomas. It is still performed for large tumors i.e. generally those with 8 mm or greater height and in those cases where no useful vision is left. It is also performed in case of insufficient regression after eye salvaging treatment modalities have been tried.

Brachytherapy / Plaque therapy:

It is the treatment of choice for localized tumors <10 mm height and <20 mm basal diameter. It involves suturing a plaque of radioactive isotope to the sclera overlying the tumor. This provides localized radiotherapy. Associated radiation induced side effects like cataract, papillopathy, retinopathy can be minimized and eyeball can be preserved.

Charged Particle beam irradiation:

Tumors unsuitable for brachytherapy either because of size or posterior location and where globe needs to be salvaged, external beam irradiation is a better option. Either proton or helium ions are used for irradiation. Most common associated complications are punctual stenosis, dry eyes, keratitis, loss of eyelashes, cataract and retinal detachment.

Trans-scleral local resection: This is undertaken when tumors measure up to 16 mm in diameter, and are too thick for radiotherapy, but where the globe can be salvaged, this modality can be opted for. It involves excision of a tumor with a rim of healthy choroid under a partial thickness scleral flap. Haemorrhage, retinal detachment and tumor recurrence are the major complications.

Trans pupillary thermotherapy:

It is a technique of tumor heating by infra red irradiation, delivered through the pupil into the tumor. This method causes dramatic tumor necrosis in choroidal melanoma up to 4 mm thickness. Small tumors located near the optic disc or fovea is selected as these tumors are too posterior for brachytherapy. Major side effects are visual field defects.

Both brachytherapy and transpupillary thermotherapy can be used to complement each other. COMS(Collaborative Ocular Melanoma Study) is founded in 1985 by national eye institute, a section of national health institute, Bethesda, USA. It aims to enable

the physician to recommend with more certainty a specific form of treatment in individual cases using randomized clinical trial.

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