



CASE REPORT

MALIGNANT MELANOMA OF CHOROID: A CASE REPORT

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ABSTRACT

Choroid melanoma has an overall incidence of about 20 per million per year globally. It is the most common occurring intraocular malignancy in adults and accounts for 90% of all uveal melanoma's. 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 increased incidence with progressive age. We report a case of 65 year old female, presenting with chronic headache, mild pain along with gradual diminution of vision in the left eye of three months duration. Investigations revealed clinical diagnosis of choroidal melanoma, the eyeball was enucleated.

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INTRODUCTION

Case Report

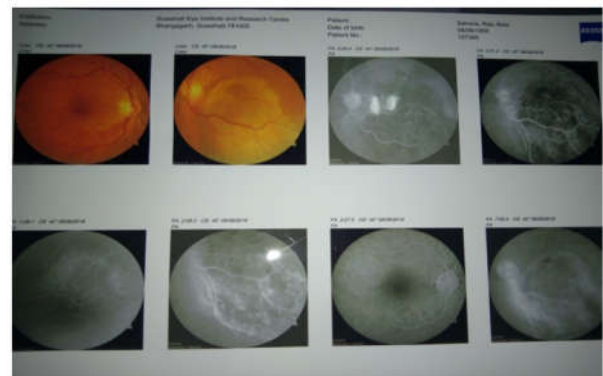
A 65 year old female presented to eye opd with chief complaints of chronic headache, mild pain along with gradual diminution of vision in the left eye of three months duration. Her general and systemic examination was normal. Ocular examination was carried out. Her right eye vision was 6/18 and left eye was finger counting at 2m. The IOP (intraocular pressure) of both eyes was 10 and 14 mmhg. Binocular indirect ophthalmoscopy of the left eye showed solid gray mass in the posterior segment (choroid). Perimetric evaluation of right eye was normal while left eye showed visual defects. USG B scan of left eye was carried out to confirm the findings and it showed well defined hyperechoic lesion in the left eye adjacent to optic nerve in its temporal aspect. Further to evaluate the extent of tumor mass involvement as well as exact location a MRI scan carried out. MRI Orbit confirmed the diagnosis as well defined plaque like T1 hyperintense intraglobular lesion noted in the posterior aspect of left globe suggestive of choroidal melanoma. Considering the imaging diagnosis and clinical findings a diagnosis of choroidal melanoma was made.

MRI ORBIT well defined plaque like T1 hyperintense intraglobular lesion noted in the posterior aspect of left globe. A consultation with an ocular oncologist confirmed the diagnosis. The patient was advised about the pros and cons of brachytherapy, radiotherapy and enucleation.

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Fig shows different modalities of the investigations being done.



FFA showing choroidal melanoma

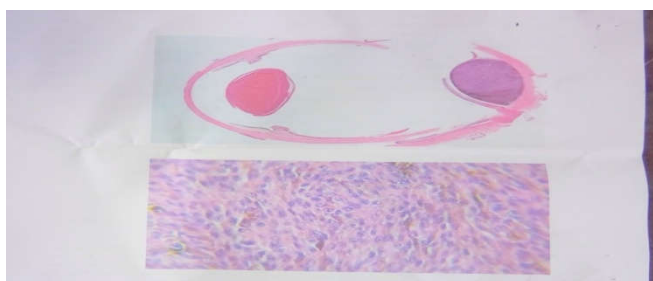


The patient opted for enucleation as the modality of management. The eye was enucleated and sent for histopathological examination. The histopathological report was consistent with choroidal melanoma (spindle cell melanoma) with tumor size (maximum basal diameter 8mm and maximum apical height 4mm. PTNM staging pT1Anx. Optic nerve cut end, vortex veins are free of tumor.

Usg whole abdomen was within normal limit with no signs of metastasis. Patient was followed up after two weeks and is kept under regular follow up.



The Enucleated Specimen of Eyeball



Histopathology Shows Spindle Cell A Variety

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 cancers¹ with incidence of 6 cases / 1 million in U.S.A. The risk factor Increases with age with most of the patients in 6th decade of life. Melanomas tend to commonly arise from 60 years of age, however no age is spared, melanomas can occur in adolescents, children and rarely even in neonates^{2,3}. Clinically the risk factors also includes preexisting lesions like melanosis oculi, ocular melanocytosis, uveal nevi, congenital melanosis, light iris and associated tumors-visceral malignancies [bowel, gall bladder, pancreas, ovary Most patients are asymptomatic.

While others have Photopsia (Ball of light travelling across visual field) Blurring of vision, Field defects, Floaters, Metamorphosia and Pain due to secondary glaucoma, spontaneous tumour necrosis.

Malignant melanoma is always primary, single and unilateral. May be elevated, subretinal, dome shaped brown or grey mass. In 20 % breaks bruchs membrane and acquires mushroom or collar-stud appearance. Clumps of lipofuscin [orange colour] are seen in R.P.E overlying the tumour. Amelanotic tumors may be light or medium grey. And diffuse tumors are flat grey brown, irregular discoloration. Extraocular extension is common

Pathology

A.F.I.P [Modified Callender] Classification⁴

- According to histopathology

Spindle Cell Melanoma [44%]

- Composed of spindle cells with pallasiding or ribbon like arrangement in parallel rows.
- Spindle A-5%-Best prognosis.5 yr mortality 5%
- Spindle B-39%-

Epitheloid Cell Melanoma [3%]

- Poor prognosis, 5yr mortality 69%

Mixed Cell Melanoma:[45%]

- Mixture of epitheloid and spindle cells.
- Intermediate prognosis.

Necrotic - cell type can not be assessed,

Immuno histo chemical markers-S-100 and HMB-45

Poor Prognostic Factors

- Large tumor size
- Histopathology
- Epitheloid cells worst and spindle cells best prognosis.
- High mitotic index
- Greater pigmentation
- fibrovascular loops within tumour
- Extra scleral extension
- loss in chromosome 3 and gain in chromosome 8
- anterior tumours involving ciliary body

SITEs of Metastasis includes Liver [56%], Subcutaneous nodules[24%] Lungs[7%],Vertebra[7%],CNS[2%

Management

Depends upon

1. Visual acuity-salvagable vision-conservative treatment.
2. Size of tumour-small[2-3 mm]- observation medium[3-5 mm]-irradiation / local resection large[10mm]-enucleation
3. Location- close to optic nv - Enucleation-close to equator-Irradiation/resection
4. growth pattern - diffuse pattern -Radical surgery
5. Activity of tumour
6. Status of opposite eye.

7. Age & general health of patient.

Increasing trend towards

Multimodal Therapy

Adiotherapy

1. A.Brachytherapy-Treatment of choice ^{60}Co , ^{125}I
Indication-small tumours
2. External Beam Radiotherapy-
3. Indication-posterior located large size tumour.
4. Transpupillary Thermootherapy-
5. For small tumours located near optic disc or fovea.
6. Trans Scleral Local Resection-
7. Tumours too thick for radiotherapy.
8. Stereotactic Radiosurgery-for localised tumour.
9. Enucleation-Large tumours when all vision is lost.
10. Palliative Treatment-Chemotherapy and/or Immunotherapy to prolong life in a patient with metastasis.

Prognosis and Survival

Uveal melanoma size is the most important clinical factor related to prognosis. Coupland *et al.*⁵ evaluated 847 patients with uveal melanoma for metastatic death and found clinical and histopathologic predictive factors of largest basal tumor diameter, closed loops, epithelioid cells, mitotic rate, and extraocular spread. Damato *et al.*⁶ included genetic testing in their analysis for factors predictive of metastatic death and found the most important independent predictors to be basal tumor diameter, chromosome 3 loss, and epithelioid cell histopathology. Eskelin⁷ explored tumor doubling times and speculated that most metastases initiate 5 years before primary treatment.

Despite the availability of alternative treatment modalities, the survival rates of patients with uveal tract melanoma have not changed in 30 years. Cumulative rates of metastases in the Collaborative Ocular Melanoma Study at 5 and 10 years after treatment were 25% and 34%, respectively. Common sites of metastases include liver (90%), lung (24%), and bone (16%)^{8,9} Patients with metastases confined to extrahepatic locations have longer survival (19–28 months).The median survival for a hepatic metastasis is 6 months with an estimated survival of 15–20% at 1 year and 10% at 2 years, irrespective of treatment.¹⁰ Asymptomatic patients at the time of diagnosis of metastases have a slightly longer survival in relation to symptomatic patients.¹

CONCLUSION

Choroidal melanoma can be diagnosed quite accurately with non-invasive techniques like IO, A- and B-ultrasonography scans, FFA, and transillumination, preventing complications of FNAC like seeding of tumor cells in the needle track. The role of CT, magnetic resonance imaging, and nuclear magnetic resonance spectrography in the diagnosis of melanoma is still uncertain. Radioactive phosphorus uptake is no longer used because the rates of false negatives and false positives are quite high. Treatment for a small choroidal melanoma in the posterior fundus ranges from observation to several treatment options, including laser photocoagulation, plaque radiation therapy, external beam charged particle radiation therapy, transpupillary thermootherapy, location tumor resection, and enucleation. For medium and large lesions, enucleation is

considered primarily if there is a diffuse melanoma or if there is extraocular extension. Plaque brachytherapy is the most frequently used eye-sparing treatment for these lesions. External beam (charged particle) either helium ions or protons may have several theoretical advantages over plaque therapy. Patients treated with proton beam irradiation had a 64% incidence of maculopathy for tumors within four disc diameters of the macula. But, radiation therapy is about twice as expensive as enucleation and there appear to be no significant quality of life differences between patients treated with radiation or enucleation. Other radiation therapy technique that has shown potential in preliminary studies is Gamma knife surgery. Considering the gamut of treatment options available, the treatment of choroidal melanoma needs to be individualized, keeping in mind various tumor characteristics and patient factors.

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