

CHOROIDAL MELANOMA- A CASE REPORT

Shivakumar Mallappa¹, Mamatha Nagaraj², Vandana Parasar³

¹Professor, Department of Ophthalmology, Rajarajeswari Medical College and Hospital, Bangalore.

²Postgraduate Student, Department of Ophthalmology, Rajarajeswari Medical College and Hospital, Bangalore.

³Postgraduate Student, Department of Ophthalmology, Rajarajeswari Medical College and Hospital, Bangalore.

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PRESENTATION OF CASE

A 48-year-old female presented to our institute with complaints of diminution of vision in right eye since 3 months. No other ocular complaints. Past history and family history was not significant.

Ocular examination revealed visual acuity of 6/60 in right eye and 6/18 in left eye. Anterior segment was normal in both eyes. On dilated fundus evaluation, distant direct ophthalmoscopy showed a mass projecting behind the lens in inferotemporal quadrant from 6 to 9 o'clock position (Figure 1). Indirect ophthalmoscopy revealed a large elevated pigmented lesion approximately 8 DD in size present at posterior pole from 5-8 o'clock hours, approximately 1 DD below the disc with vascularisation seen over the mass (Figure 2). It was associated with retinal detachment from 4-8 o'clock position in inferior quadrant, macula was spared.

Clinical observation was correlated on B-scan showing 11 x 11 mm pedunculated mushroom like choroidal mass in inferior quadrant with RD. The lesion had regular round margin with initial prominent spikes followed by internal spikes of moderate-to-low reflectivity (Figure 3). Extrascleral or optic nerve involvement not seen. OCT showed serous RD with normal retinal thickness (Figure 4).

With clinical diagnosis of choroidal melanoma, patient was advised for routine blood investigation (including LFT), chest x-ray, CT abdomen, MRI brain and orbit to rule out any metastatic changes. She was referred to higher centre for oncologist and oculoplasty opinion on further management.

DIFFERENTIAL DIAGNOSIS

There are a plethora of differential diagnoses for melanotic and amelanotic choroidal melanoma. Our case presented more with changes of melanotic melanoma. Various differential diagnosis include-

1. Choroidal naevi- Choroidal nevi are common incidental findings upon ophthalmologic examination and are usually benign. However, they have been associated

with increased risk of developing uveal melanoma, and when subfoveal, may affect vision. They may also cause anatomic changes in the overlying retina.¹

2. Choroidal metastasis- The choroid is the most common ophthalmic site for metastatic disease and it is postulated that haematogenous dissemination of metastasis from remote major sites typically leads to the high flow choroidal vasculature with metastatic disease. In contrast to choroidal melanomas, which display medium-to-low reflectivity on A-scan and are acoustically hollow on B-scan, choroidal metastases have a higher reflectivity on A-scan and appear echodense on B-scan with a significantly lower height to base ratio compared to melanomas.²
3. Congenital hypertrophy of RPE- It is a darkly pigmented lesion with a depigmented halo in the retina. It may be single or multiple, unilateral or bilateral. The size of a CHRPE lesion is variable, but most are similar in diameter to the optic disc. CHRPE has no malignant potential.³
4. Melanocytomas- Intraocular melanocytomas are relatively rare, benign tumours. They are usually diagnosed in patients between the ages of 30 and 50 years. Commonly occurring near or on the optic disk, they have also been reported to develop in the choroid, ciliary body, iris, conjunctiva or sclera. It can also form a part of paraneoplastic syndromes (super nevus syndrome or bilateral diffuse uveal melanocytic proliferation syndrome), in such a condition, they tend to be very dark, multiple, occur bilaterally and may grow rapidly. This is unlike melanocytomas in other situations where mild growth is seen in 10-15% cases. Choroidal melanocytomas are also reported not to exceed 1 disk diameter in size. Hence, proper diagnosis of benign intraocular tumours is important for more conservative approach. This can avoid unnecessary enucleations and thereby protect the quality of life in a young individual.⁴

Additional differentials for amelanotic or melanotic choroidal melanoma includes choroidal osteomas, choroidal haemangiomas, choroidal detachment, lymphoma, metastatic carcinoma, subretinal haematoma, localised suprachoroidal haematoma, nodular posterior scleritis or massive gliosis of the retina.

CLINICAL DIAGNOSIS

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.⁵ Ocular

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Corresponding Author:

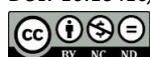
Dr. Mamatha Nagaraj,

No. 1950, 6th Cross, 2nd Railway Parallel Road,

Kengeri, Upanagara, Bangalore-560060, Karnataka.

E-mail: drmk1982@gmail.com

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melanoma arises from the melanocytes situated in conjunctival membrane and uveal tract of the eye. Although, second most common type of melanoma after cutaneous ocular melanoma is still rare and amounts 3.7% of all melanoma cases.⁶ Choroidal melanoma is the most common primary intraocular malignant tumour.⁷

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.⁵ Presentation of choroidal melanoma mainly depends on size and location of the tumour and can vary from asymptomatic, detected incidentally on eye examination over various visual disturbances to visual loss in the affected eye.⁶ In general, anterior choroidal melanomas have a delayed presentation because of slow growth, however, clinical signs and symptoms can present earlier.⁵ Whereas, the most common symptoms for posterior choroidal melanoma are blurred vision, visual field defect, photopsias, irritation and pain, but symptoms as metamorphopsia, floaters, redness and pressure can also occur.⁶ Choroidal melanoma presents with dome-shaped subretinal mass with secondary retinal detachment, colour can vary from typically brown pigmented to amelanotic.⁶

Diagnosis of choroidal melanoma is mostly established by ophthalmic examination including slit lamp biomicroscopy, indirect ophthalmoscopy and ancillary diagnostic testing such as ultrasonography, fluorescein angiography and optical coherence tomography.⁵ Invasive studies such as Fine-Needle Aspiration Cytology (FNAC) can have significant morbidity and should only be considered if therapeutic intervention is indicated and diagnosis cannot be established by any other means.⁷

Treatment

Several modes of treatment are available for choroidal melanoma. Multiple factors are taken into account when deciding one approach over other approaches such as visual acuity of the affected eye, visual acuity of the contralateral eye, tumour size, location, ocular structures involved and presence of metastases.⁷ Management of uveal melanoma varies from observation to orbital exenteration.⁶

A small and medium-sized choroidal melanoma in the posterior fundus is amenable to several treatment options, including laser photocoagulation, photodynamic therapy, plaque radiation therapy, external beam charged particle radiation therapy, transpupillary thermotherapy, tumour resection and enucleation,⁷ while large tumours, especially if locally advanced are still mostly treated by enucleation or orbital exenteration.⁶

Enucleation has been the time honoured method of management of the large ocular melanoma and melanomas that cause severe glaucoma or invade the optic nerve.⁷

Local treatment of choroidal melanoma has improved a lot with increased use of conservative treatment and preservation of the eye. However, improvement in local

treatment did not provide significant increase in survival rates and metastatic disease is remaining a leading cause of death among the patients with choroidal melanoma.⁶

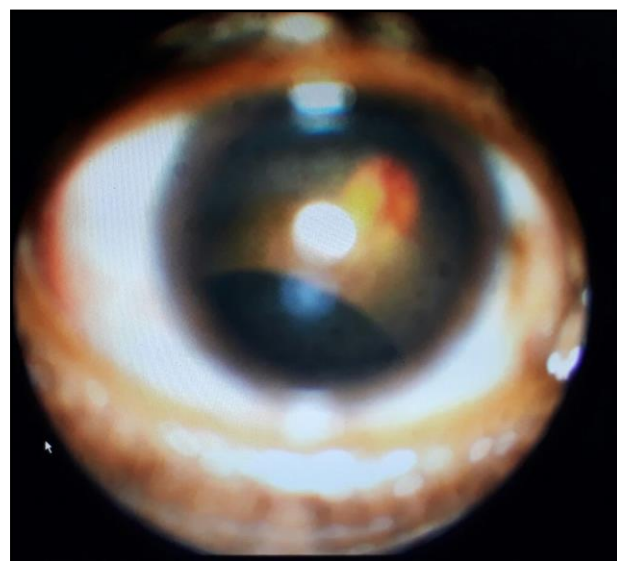


Figure 1. Anterior Segment View Showing Retrolenticular Mass in Inferior Quadrant



Figure 2. Fundus Picture Showing Melanotic Choroidal Melanoma with Vessels Over it Associated with Retinal Detachment

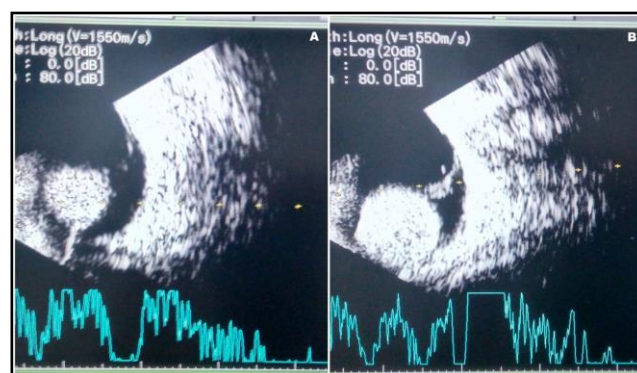


Figure 3. B-scan. (A) A Dome-Shaped Polypoidal Mass from Choroid. (B) Associated Retinal Detachment

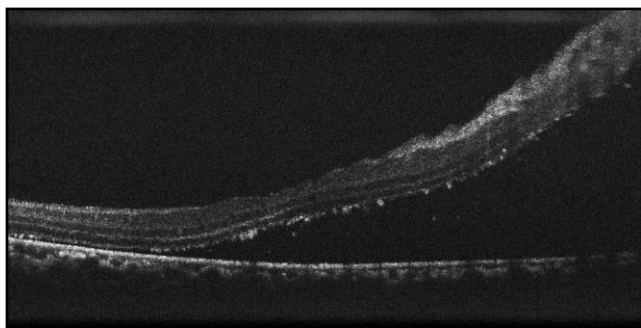


Figure 4. OCT Picture Showing Serous Retinal Detachment

FINAL DIAGNOSIS

Based on the clinical presentation, slit lamp biomicroscopy and fundus evaluation, our patient was diagnosed with choroidal melanoma. On further evaluation with B-scan and OCT, reports strongly suggested choroidal melanoma with presence of hyperechoic mass in posterior segment with low-to-medium internal reflectivity associated with significant subretinal fluid.

Our final diagnosis was right eye large size melanotic choroidal melanoma with exudative retinal detachment sparing macula.

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