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OPHTHALMOLOGY
Case Report

Malignant Melanoma of the Choroid: A Case Report

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Received: November 25, 2015; **Published:** December 22, 2015

Abstract

A case of malignant melanoma of the choroid in a 55 year old female is reported because of not so common occurrence of this malignancy in Indian population. The Tumour had typical clinical and investigative features. The basal tumour diameter was 12 mm and height 10 mm. It was situated in the posterior pole underlying macula and nasal margin was overhanging the optic disc. The tumour was enucleated and orbital implant and prosthesis were fitted. Histopathologically tumour was of spindle cell type.

Keywords: *Choroid; Enucleation; Fundus Flourescein Angiography; Malignant Melanoma; PET Scan; Retinal Detachment; Ultrasonography*

Introduction

Malignant melanoma of the choroid is the most common primary intraocular malignancy present in adults. It usually presents in the 4th - 6th decades of life. The incidence of this tumour is 0.02 - 0.06% [1]. Its incidence is more in Caucasians and is much lower in Asians and Africans. Even though this tumour is relatively rare and is less often reported in Indian subcontinent, the tumour has to be borne in mind in eyes of adults with secondary/exudative detachment. Clinical examination with indirect ophthalmoscopy and FFA in clear media and imaging technique like USG, CT and MRI in both clear and opaque media are the mainstay of diagnosis and assessing tumour features. Recently, PET scan is an added advantage in scanning the whole body for metastasis and further confirming the tumour activity. As such, malignant melanoma of the choroid is not very often reported in Indian population [2-5]. To the best of our knowledge there are only few reports of malignant melanoma of choroid from India and moreover many have been misdiagnosed as Glaucoma [6].

Case Report

A fifty five year old female patient presented with a history of painless and gradually progressive diminution of vision in her left eye of one and a half months duration. She was diagnosed to have retinal detachment. On examination, the anterior segments of both eyes were normal. Visual acuity in the left eye was hand movements and in the right eye 6/9, N8. Intra ocular pressure was 16 mm of Hg in both eyes. Fundus examination with the indirect ophthalmoscope showed a subretinal, well demarcated, dark brown solid looking mass lesion with prominent surface vessels involving the macular area. Its nasal margins were overhanging and covering the optic disc.

There was no abnormality of the iris and ciliary body. Fluorescein angiography showed intrinsic tumour vascularity (double circulation) in the arterio-venous phase, multiple punctate hyperfluorescent patches over the tumour in the mid phase and diffuse hyperfluorescence in the late phase. Ultrasonography B-scan showed circumscribed dome shaped solid mass with areas of acoustically hollowness, choroidal excavation and orbital shadowing .The basal tumour diameter was 12 mm and height 10 mm. There was overlying RD with scanty sub retinal fluid (SRF). A-scan USG showed a tall anterior wall spike, low to medium intralesional echoes and sound attenuation forming angle kappa.

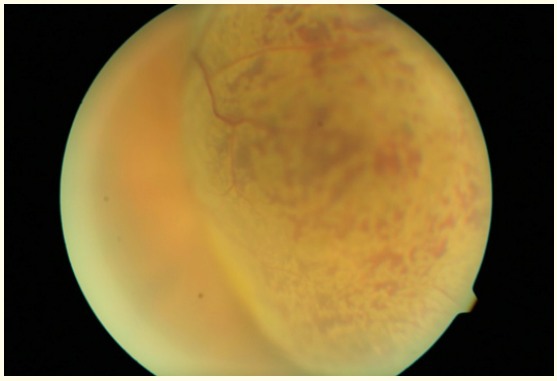

Figure 1: *showing solid retinal detachment with surface blood vessels.*

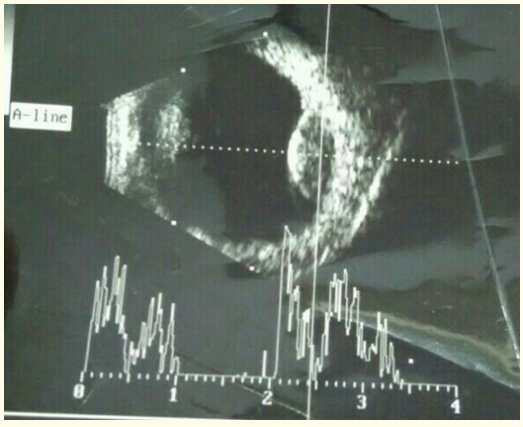

Figure 2: *B-scan and A-scan USG of the lesion.*

The transillumination test was positive. MRI of the eye showed hyperintense choroidal lesion on T1 weighted image and hypointense on T2 weighted image. The tumour was confined to the eye without any extra ocular extension and there was no optic nerve involvement. On general physical examination, there was no hepatomegaly and Liver function tests were normal. USG of the liver showed normal echotexture. A whole body PET scan showed metabolically active lesion in eye only.

Based on these findings the diagnosis of primary intraocular malignant melanoma of the choroid was made. The Patient was counselled on management options including Brachytherapy, Enucleation, Charged particle radiation therapy and Observation. As the tumour was located at the posterior pole, so the feasibility of placing radioactive plaque was not possible and moreover the tumour was too large for treatment with brachytherapy. Further the likelihood of development of profound vision loss due to external beam radio therapy (EBRT) causing radiation optic neuropathy and hence poor visual outcome due to location of the tumour under the macula were discussed with the patient. Further, keeping in view the absence of extraocular extension and hence, the likelihood of total cure following enucleation was explained to the patient. The patient elected to undergo enucleation with orbital prosthesis. Enucleation was performed least traumatically. The patient tolerated the surgery well and later received an orbital prosthesis. Due to the clear margins indicating lack of extra ocular spread and lack of evidences indicating distant metastasis, no further treatment was considered necessary. The patient was counselled to undergo yearly eye examination with annual tumour surveillance consisting of a physical examination, liver function tests and imaging of the liver with ultrasonography or MRI with contrast.

Pathologically, gross examination of the cut section of the enucleated eyeball showed a greyish-brown mass measuring 12 mm in basal diameter, attached to the posterior surface of eyeball and 1 mm away from innermost surface of posterior wall. The representative microsections examined showed malignant melanoma of choroid of spindle cell type. The posteriormost margin of eyeball and Optic Nerve were free from tumour infiltration. On Immunohistochemistry, the tumour cells were found to be HMB-45 positive, Melan-A positive.

Discussion

Malignant melanoma of the choroid is relatively rare in our country. Because of the relative rarity of the condi¬tion, this is often not suspected till the tumour is large enough to cause secondary glaucoma and/or distant metastasis. In the present case the tumour was detected early due to involvement of macula resulting in early visual loss. It is important to distinguish between melanoma and pseudo-melanomas like choroidal nevus, choroidal metastasis, melanocytoma or hyperplasia of retinal pigment epithelium. Nevi are usually flat, < 6 mm in diameter and may have overlying drusen. Choroidal metastases are typically amelanotic, lobulated and associated with more SRF than melanoma of similar size. Melanocytomas are black and often extend onto the optic nerve and invade the nerve fibre layer, giving a feathered appearance. The congenital hyperplasia of the RPE (CHRPE) is typically a sharply demarcated pigmented lesion with scalloped margin. In the present case, the tumour was dark brown, elevated, dome shaped with scanty SRF without involving the optic nerve. Positive transillumination test ruled out haemorrhagic retinal detachment. Further, the signal characteristic on ultrasonography and characteristic findings on MRI were in favour of malignant melanoma of the choroid. Moreover, the whole body PET scan confirmed the activity of the tumour in eye and showed absence of similar lesion elsewhere in the body pointing to a primary ocular metabolically active/proliferating tumour.

Regarding various treatment options of malignant melanoma of choroid such as brachytherapy (in small to medium sized anteriorly located tumours), Charged particle radiation therapy (can cause radiation optic neuropathy and other radiation related complications), Trans pupillary thermotherapy (TPT) for small posteriorly located tumours, Sclerouvectomy (in selected cases) and Enucleation were considered [7]. Keeping in view the large size of the tumour and posterior location under macula, poor potential visual outcome following other treatment measures and tumour confinement to the eyeball without extrascleral extension and optic nerve involvement, the enucleation was found to be the best option in this case as this could provide a total cure and confirm the diagnosis.

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Volume 3 Issue 1 December 2015