A Rare Case of Ciliary Body Melanoma Masquerading as Secondary Angle Closure Glaucoma

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INTRODUCTION

Uveal melanoma is the most common primary malignancy of the eye in Caucasian adults, of which choroidal melanoma is the most common subtype.[1] Ciliary body melanoma is a rare tumour seen in 1 out of 10 cases of all intraocular melanomas with an average age of 55 to 62 years.[2, 3] Uveal melanoma arises from atypical melanocytes present in iris, ciliary body and choroid, although each tumour has some particular characteristics depending on its location and structure. Because of the continuous contractions of the ciliary muscle and rich vascularization of the ciliary body, metastasis is faster in ciliary body melanoma.[2] Vascular metastasis leads to impairment of other organs which is the main cause of death among ciliary body melanoma patients with 10 year mortality rate being 30-50%.[2] Ciliary body melanoma has the worst prognosis of all intraocular tumours due to early hematogenous metastasis which are frequently seen in the first year after diagnosis.[3]

PRESENTATION OF CASE

A 55-year-old male patient, farmer by occupation, residing in Bhilawara district of Rajasthan state, India presented with complaint of diminution of vision, pain and redness in right eye since 2 months with aggravation of symptoms since last 3 days. The patient was asymptomatic before 2 months, when he noted gradual diminution of vision and redness in the right eye. This gradually progressed to severe pain, increasing redness and gradual, progressive loss of vision over the last 3 days.

The patient had no significant past medical history or family history. There was no history of trauma, use of prescription glasses, foreign body, previous eye diseases or eye surgery.

On examination, the best corrected visual acuity in right eye was counting finger 3 meters without glasses which was not improving with pin hole or glasses and in left eye 6/12 with -1.00D spherical glasses for far vision and N8 with +2.50 D spherical glasses. On slit-lamp examination, lid and lacrimal apparatus were normal in both eye. Right eye showed conjunctival congestion, generalized corneal oedema with the shallow anterior chamber temporally. Iris chafing with bulge and iridocorneal contact was seen on temporal aspect and hyphema of approximately 2 mm in size was present in the right eye. Pupil was semi dilated, fixed and not reacting to light with multiple posterior synechiae and blood-stained anterior lens capsule with cataractous lens changes were seen in the right eye. Anterior segment of left eye appeared normal.

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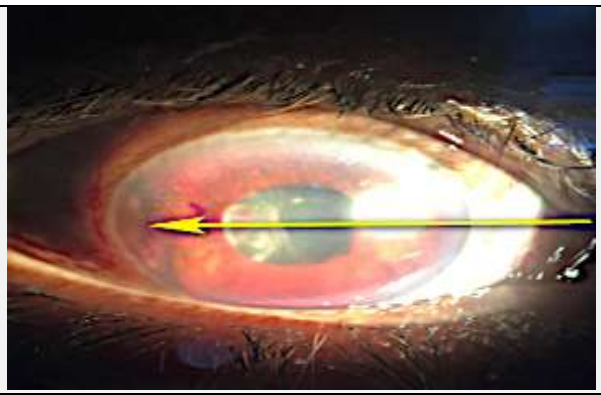
Eye movements were full in both eyes. Intraocular pressure was 38 in right eye while 16 in left eye on applanation tonometry. On indirect ophthalmoscopy, fundus details were not seen in right eye while left eye fundus was within normal limits.

All routine investigations of the patient were done and initial management of lowering the intraocular pressure with brimonidine + timolol eye drops twice a day in right eye along with systemic tablet acetazolamide four time a day with syrup potassium chloride and syrup glycerol 1 ounce three times a day with lime juice was started. Cyclopentolate eye drops at bedtime were initiated to alleviate pain induced by ciliary spasm and treat posterior synechiae. A combination of moxifloxacin and prednisolone acetate eyedrops were also started four times a day.

Investigations

On B-scan ultrasonography and ultra-biomicroscopic sonography of right eye, a small iso to hypo dense inhomogeneous mass of 5.4 mm * 4.1 mm was seen in temporal aspect of right eyeball (adjacent to iris and ciliary body) which showed mild vascularity in it and caused indentation as well as posterior displacement of adjacent part of the lens of right eye, however right eye lens itself appeared normal. Thus, the above findings were suggestive of small soft tissue mass in temporal aspect of right eyeball (adjacent and arising from iris and ciliary body) suggestive of melanoma in nature. Vitreous humour was clear with thick choroid and no evidence of vitreous or retinal detachment was seen along with normal optic nerve head. The ultrasound of left eye showed normal anterior chamber and clear vitreous humour with no evidence of vitreous or retinal detachment present. Ultrasound biomicroscopy (UBM) revealed ciliary body mass in the right eye.

The magnetic resonance imaging (MRI) showed small well-defined altered signal intensity area of approximately 4.5*2.8*5.5 mm in the lateral ciliary body and iris region adjacent to the lens on the lateral aspect in the right eyeball which appeared hyper intense on T1W images and hypo intense on T2W images causing slight posterior displacement of the lens, thus above features suggested possibility of neoplastic mass lesion involving right lateral ciliary body and iris region - ? Melanoma.



Figures 1. Slit-Lamp Examination of the Right Eye - Iris Chaffing with Irido-Corneal Contact with Hyphema with Sentinel Vessel

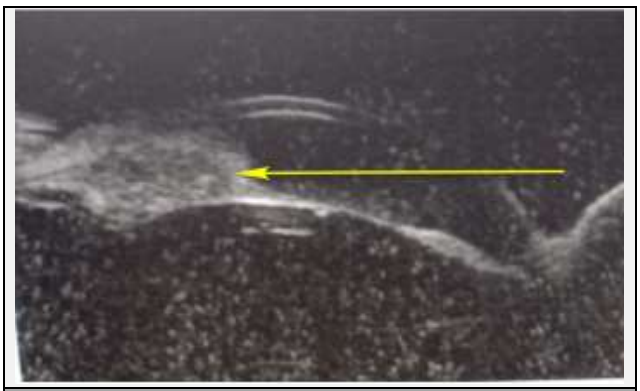


Figure 2. Ciliary Body Mass on Ultrasound Biomicroscopy

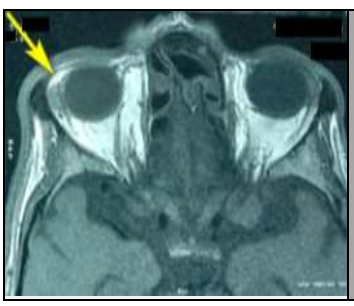


Figure 3. T1 Weighted MRI Showing Hyperintense Mass in the Right Eye

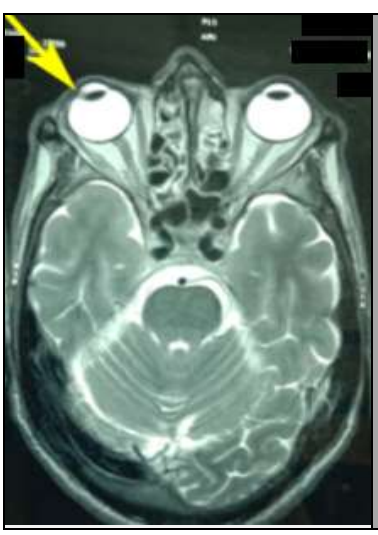


Figure 4. T2 Weighted MRI Showing Hypointense Mass in the Right Eye

Chest x-ray was within normal limits and no any abnormality was detected on ultrasound of the abdomen with normal size and appearance of liver.

CLINICAL DIAGNOSIS

Ciliary Body Melanoma

DIFFERENTIAL DIAGNOSIS

1. Iris cyst-which is hollow in the center on UBM and does not metastasize
2. Diktyoma or ciliary body medulloepithelioma or teratoneuroma which arises from ciliary body epithelium and is very similar to ciliary body melanoma. Only

histopathological sections can differentiate between the two.

DISCUSSION OF MANAGEMENT

Enucleation with gentle handling of the affected eye was done in consultation with oncology department as the radiological investigations suggested the diagnosis of ciliary body melanoma with less possibility of metastasis to the liver and lung.

The enucleated specimen of the right eyeball was 2.5 * 2.5 * 2.5 cms in size with 0.5 cm of the optic nerve stump and on cutting, small firm mass of 0.5*0.5 cm was found in the anterior segment of the globe.

Histopathological examination of the enucleated right eyeball specimen confirmed diagnosis of ciliary body melanoma as mixed type melanoma (epitheloid + spindle cells) melanoma with G2 histological grade on microscopic examination. The size of the tumour reported was 0.5*0.5 cm with atypical mitosis seen. Tumour necrosis and pigmentation was not seen and tumour infiltrating lymphocytes were also not seen. Extra scleral extension was not seen, and tumour cells had not involved the optic nerve head and cut margin of optic nerve.

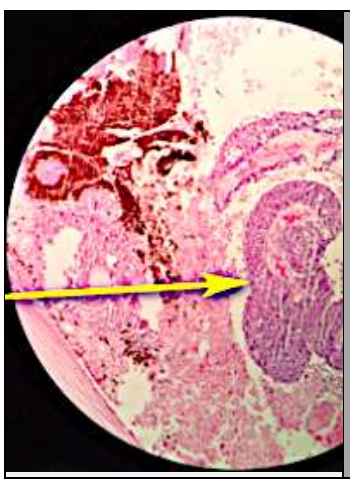


Figure 5. Microscopic Examination of Ciliary Body Melanoma Showing Non-Pigmented Mixed (Epitheloid + Spindle Cells) Tumour

PATHOLOGICAL DISCUSSION

Amongst the subtypes of uveal melanoma, choroidal melanoma is the most common whereas ciliary body melanoma as an isolated entity is rarely seen. Due to metastasis and local extension, ciliary body melanoma is associated with choroidal melanoma or iris melanoma and rarely diagnosed as a single entity.[1]

It commonly remains asymptomatic or may have local signs such as blurring of vision because of lens dislocation, cataractous changes, astigmatism, floaters or pain because of the raised intraocular pressure due to acute angle-closure glaucoma or may have general signs like weight loss, marked fatigue, cough and gastrointestinal or urinary symptoms due to metastasis.[2]

As the tumour grows, it can be seen as a variable pigmented mass with diffuse, nodular or mixed pattern seen behind the pupil or the tumour can invade the anterior chamber and affect the iris or can grow into the posterior pole affecting the choroid.[2] The first ophthalmological sign of this tumour is the presence of "sentinel vessels" which are the dilated episcleral vessels and an unexplained low intraocular pressure of 5 mm Hg.[2]

The ophthalmological investigations like ultrasound biomicroscopy (UBM), CT or MRI are helpful in the diagnosis whereas histopathological examination can confirm diagnosis of ciliary body melanoma.[1] The UBM precisely delineate anterior and posterior margins of the tumour and helps to choose the management option amongst biopsy, resection or plaque radiotherapy.[4]

The Callender modified classification.[5] for uveal melanoma divided the ciliary body melanoma into four histopathological subtypes: spindle cell melanoma, mixed cell melanoma, epithelioid cell melanoma and necrotic melanoma. Spindle A and B type melanoma has the best prognosis as type A spindle cells has small, fusiform nuclei with rare mitoses whereas type B spindle cells are bigger, more pleomorphic, with prominent nucleoli and mitotic activity.[5] The necrotic type of melanoma has undistinguishable cell type due to necrosis and has poor prognosis, whereas the epithelioid cells melanoma has the worst prognosis due to presence of an anaplastic appearance and intense nuclear pleomorphism with frequent mitoses.

The radiotherapy such as plaque brachytherapy or proton beam therapy is most commonly preferred treatment option for ciliary body melanoma, amongst which external beam therapy or brachytherapy can be used for medium-size tumour (less than 15 mm in diameter).[2,6] The surgical treatment such as enucleation of the eye, block excision or sclerouvectomy are done in advanced cases, depending on the size of the tumour, cellular type of the melanoma and the local extension.[2]

The clinical prognostic factors are the presence of metastasis, the presence of the dysplastic nevi, local extension, the age of the patient and the occurrence of local and general signs but the most important prognostic factor is the macroscopic size of the tumour.[1] The 5 year survival rate is 86% in "small"-type tumour with the biggest diameter of less than 11 mm, 66% in "medium" tumour having a diameter between 11 to 15 mm and 56% survival rate in "large" tumours having more than 15 mm in diameter.[7] Correlation of molecular and cytogenetic profile of primary Uveal melanoma with patient survival is well established.

CONCLUSIONS

The ciliary body melanoma is a rare intraocular tumour. It was masquerading as secondary angle closure glaucoma. So, a good clinical examination is of utmost importance. As it can have early hematogenous metastasis, aggressive and

multimodal diagnostic and treatment approach can save the patient's life.

Financial and Competing Interests - None

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