Research Paper

Volume : 4 | Issue : 10 | October 2015 • ISSN No 2277 - 8179

MALIGNANT MELANOMA OF CILIARY BODY : A RARE CASE REPORT

Medical Science

KEYWORDS :

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ABSTRACT

Uveal melanoma is the most common primary malignant tumour of the eye in adults, predominantly found in Caucasians, particularly in those with light-colored irides. The lesion is usually single and unilateral, 85% arising from the choroid and 15% from the ciliary body. Ocular malignant melanomas are infrequently seen in Indian patients .The incidence of this tumour is 0.02 - 0.06% in India and most of them involve the choroid. Ciliary body malignant melanoma is rather rare. It is encountered approximately one tenth as often as is choroidal melanoma. This case report illustrates an occurrence of ciliary body melanoma in an Indian male patient.

CASE REPORT

A 42 yrs old army personnel was referred from neuromedicine department to ophthalmology OPD . He was admitted in neurology with a provisional diagnosis of optic neuritis with the chief complaints of gradual progressive diminution of vision in right eye since 3 weeks. On examination he had visual acuity of FC 2 m in the right eye and 6/6 in left eye. Pupillary reaction in RE was ill sustained while in LE was normal. Afferent papillary defect was not elicitable in the right eye. On torch light examination he had episcleral congestion on nasal aspect of RE(figure 1).

Slitlamp examination revealed quiet anterior segment with presence of a dome shaped mass behind the iris occupying superonasal portion of pupillary area which was greyish brown in colour with blood vessels on its surface(figure 2 and 3). Gonioscopy showed increased pigmentation of the trabecular meshwork.

Fundus examination showed optic disc swelling and macular edema with evidence of exudative retinal detachment (Shifting fluid sign was positive) (figure 6). The intraocular tension was normal.

On basis of clinical examination a presumptive diagnosis of ciliary body melanoma was made and MRI and Bscan was done.

MRI reveals well marginated oblong leion in nasal aspect of right eyeball involving region of ciliary boby and choroid. The lesion was hyperintense on T1 images and hypointense in T2w images with no evidence of extrascleral or perineural spread (figure 4 and 5). Ultrasonography Bscan revealed a 16 x 11 mm irregular nodular mass in region of the ciliary body, nasally which with a cystic lesion within the mass which showed low to medium internal reflectivity (figure 7). There was evidence of retinal detachment also.

On colour Doppler increased vascularity in the tumour was evident.

Chest x-ray and ultrasonograph abdomen was normal and liver function test were in normal limit.

Enucleation of the left eye was performed with the patient's consent.

Pathologically, gross examination of the cut section of the eyeball revealed the ciliary body region with adjacent choroid occupied nasally by a 15x10 mm, apparently circumscribed tumor nodule.

Microscopic examination revealed a mixed cell type of malignant melanoma containing predominantly epithelioid cells with few spindle cells interspersed.

The tumour also invaded the adjacent choroid.

The cornea, iris, retina, and optic nerve were not involved.

At the 2-year follow up of this patient, there was no evidence of any local or systemic metastasis

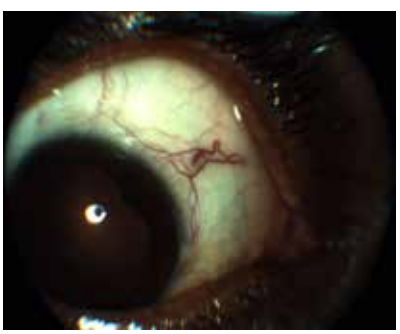

Fig 1 Dilated episcleral 'sentinel' vessel

Fig 2- Dome shaped mass projecting from superonasal area behind iris

Figure 3 -Ciliary body malignant melanoma with blood vessels on its summit

Figure 5 . MRI shows Hyperintensity on T1W images

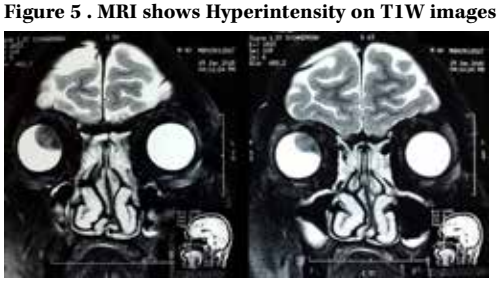

Figure 5 . MRI shows Hypointensity on T2W images

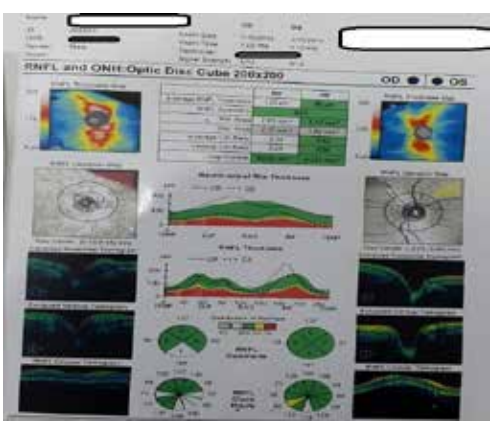

Figure 6 .OCT showing optic disc edema on right side

Figure 7. B scan showing melanoma along with retinal detachment

DISCUSSION

Uveal melanoma, although commonest primary intraocular tumor, is rare and even rarer in the Asian population[1] while people of northern European descent are most often affected.

A ciliary body melanoma is a malignant neoplasm that arises from the melanocytes of the ciliary body[2] .Since it is in a hidden location behind the iris, ciliary body melanoma usually attains a relatively large size before it is detected clinically. . External ocular signs, however, such as dilated episcleral "sentinel" vessels or a focus of transscleral extension of the tumour can alert the clinician to a possible underlying neoplasm.. More typically, however, it is confined to the ciliary body and can be detected only by slit-lamp or fundus examination after wide dilation of the pupil. Ciliary body melanoma can be uniformly pigmented, partially pigmented or amelanotic

Predisposing conditions for uveal melanoma include the following:

- Uveal nevus
- Congenital ocular melanocytosis[3]
- Xeroderma pigmentosum
- Dysplastic nevus syndrome
- Race predilection

Most cases (97.8%) of uveal melanoma occur in the whites, particularly whites of northern European descent. The incidence of ocular melanoma among blacks is extremely rare. Hispanics and Asians are thought to have an intermediate risk compared with whites and blacks.

Sex predilection - Uveal melanoma is slightly more common in men for most age groups; however, from age 20-39 years, a small predilection for women is noted.

Age predilection - Uveal melanoma shows a peak incidence at 55 years. In Asians, a tendency exists for the condition to occur at a younger age.

Sign and symptoms- Ciliary body melanomas can remain asymptomatic until they grow enough to affect neighbouring ocular structures. Patients may present with the following symptoms:

- Blurred vision
- Floaters
- Painless visual field loss
- Severe ocular pain

Blurred vision may occur consequent to growth of the melanoma into the crystalline lens, leading to lenticular astigmatism or cataract; they may also block the visual axis directly or via an intraocular hemorrhage.

Floaters can be reported when areas of necrosis within the tumor or adjacent structures produce vitreous hemorrhage or hyphema.

Painless visual field loss may be present as the melanoma grows centrally and posteriorly.

Severe ocular pain occasionally can be associated with ciliary body melanoma, secondary to high intraocular pressure because of acute angle-closure glaucoma.

A history of weight loss, marked fatigue, cough, or change in bowel or bladder habits should prompt consideration of primary non ocular malignancy with ciliary body metastasis.

An early sign of an occult ciliary body melanoma is a sentinel vessel, which is 1 or more dilated episcleral blood vessels feeding

the metabolically active tumor and is visible through the conjunctiva overlying it. This finding should prompt the physician to dilate and carefully examine the anterior-posterior segment. Another early physical sign of an occult ciliary body melanoma is unexplained, unilateral low intraocular pressure, as compared with the healthy fellow eye. A difference of 5 mm Hg or more may be the only initially detectable external sign of a tumor affecting the ciliary body

Most ciliary body melanomas can be observed as a darkly pigmented mass that is posterior to the pupil. Nevertheless, pigmentation ranges from inapparent to dark brown. These tumors may have a diffuse, nodular, or mixed pattern. They usually are solid, but they can be cystic..

The location of the melanoma in the ciliary body makes diagnosis difficult, because pupillary dilation and indirect ophthalmoscopy or a 3-mirror contact lens are needed to visualize the lesion.

Transillumination is helpful in localizing the tumor. Its accuracy depends on melanin content, dark pigmentation, and whether or not a hemorrhage is present in the tumor.

A-scan ultrasonography of the eye
In tumors more than 3 mm thick, standardized ultrasonography has a diagnostic accuracy of over 95%, and it is helpful in distinguishing melanomas from ciliary body cysts.

B-scan ultrasonography of the eye
B-scan ultrasonography is especially useful for imaging the eye in patients with media opacity and as a means of estimating tumor size. The ciliary body is a difficult ocular region to evaluate with B-scan. Immersion technique and comparison with the contralateral healthy eye often are needed to detect a small mass in the ciliary body.

Intraocular melanomas have several distinctive features, as follows[10,11]:
- Low-to-medium reflectivity
- Excavation of underlying uveal tissue
- Shadowing of subjacent soft tissues
- Internal vascularity
- Ultrasonographic biomicroscopy

Ultrasonographic biomicroscopy (UBM) has high resolution for ciliary body abnormalities, including melanomas. It can help to differentiate tumors of the ciliary body from those of choroidal origin and help to define the anterior border. It also is helpful in assessing angle closure and localized narrowing.

MRI is more sensitive in detecting extra-ocular spread. It is the procedure of choice for evaluation of transscleral and perineural spread. The tumor appears hyperintense on T1W images and hypointense on T2W images due to the paramagnetic effects of melanin pigment[1011,12.] Intravenous gadolinium highlights the melanoma in the ciliary body. Color Doppler reveals vascularity of the lesion and helps to differentiate it from other benign pathologies like subretinal effusion, macular lesions, etc. It helps in the localization of the tumor and also helps to assess the extraocular spread and optic nerve involvement[18.]

Liver Enzyme Levels
Since the most common site of metastasis is the liver, liver enzyme levels should always be evaluated in any patient with ciliary body melanoma.

The most sensitive hepatic function tests are serum levels of gamma-glutamyl transpeptidase, lactic dehydrogenase, and glutamic-oxaloacetic transaminase.

Chest radiography should be done to rule out the possibility of lung metastasis.

Histology and prognostic factors
Tumor cell type is an important prognostic factor. Callenderinitially proposed a classification system for uveal melanoma including spindle A, spindle B, epithelioid, mixed, fascicular, and necrotic types. This was later modified to include spindle A, spindle B, epithelioid, and mixed tumors. The modified Callender classification showed an improved correlation between the cell type and the mortality[8].

Epithelioid cells are associated with more aggressive behavior and carry a poorer prognosis for the patient's survival

Uveal melanoma can arise in the iris, ciliary body, or choroid. Iris melanoma has a better prognosis and ciliary body melanoma has the worst prognosis[4,5]

Patients with ciliary body melanoma greater than 7 mm in thickness are at 2.5 times greater risk than patients with thinner tumors for metastatic disease and melanoma-related death[6]. According to Li *etal*., there is a significant relation between degree of ciliary body involvement (% of tumor base within the ciliary body) and melanoma-related metastasis[7]. According to this study, a melanoma of presumed ciliary body origin (>50% of the tumor base within the ciliary body) had 1.6-2.3 times higher chance of metastasis than a choroidal (<50% of the tumor base within the ciliary body) tumor. Tumors with 100% ciliary body involvement had 3.6 times higher chance of metastasis than choroidal melanoma[7]. The poor prognosis of ciliary body melanoma has been related to larger tumor size, predilection for monosomy 3 and 8q gain, and tumor microvascular patterns[8]. Tumor cell type is an important prognostic factor. Callender initially proposed a classification system for uveal melanoma including spindle A, spindle B, epithelioid, mixed, fascicular, and necrotic types[12.] This was later modified to include spindle A, spindle B, epithelioid, and mixed tumors[9.] In a study of 2652 enucleated eyes with uveal melanoma by Paul *et al*., the 15-year mortality for spindle A tumor was 19%, spindle B was 26%, mixed spindle B and epithelioid was 59%, and epithelioid tumor was 72%.

Differential diagnosis
A number of ocular lesions or pseudomelanomas can clinically simulate malignant melanomas of the ciliary body. These include aphakic cyst of ciliary body, melanocytoma of ciliary body, tumours of non-pigmented ciliary epithelium, conjunctival nevi and melanoma and ciliary staphyloma.

Treatment Medical and Surgical Management
Enucleation
Multiple modes of treatment are available for ciliary body and other uveal melanomas. In determining an approach, multiple factors need to be considered, such as visual acuity of the affected eye and of the contralateral eye, intraocular pressure, ocular structures involved, size of the tumor, age and general health of the patient, and presence of metastases.

Enucleation is the classic approach to posterior ciliary body melanomas.. Because of the potential release of malignant cells into the bloodstream and orbital soft tissues during the surgical procedure, keep manipulation of the globe to a minimum[13]. Some physicians advocate for preenucleation radiation therapy to hypothetically reduce local and hematogenous dissemination.[14]

External beam irradiation
External beam irradiation with either protons or helium ions is a frequently used alternative method to treat medium-sized tumors (< 10 mm in height and <15 mm in diameter). Radiopaque

tantalum rings are usually sutured to the sclera to serve as reference markers for alignment of the radiation beam. By causing irradiation-induced vessel damage, the tumor necroses and regresses

Plaque brachytherapy

Plaque brachytherapy is a widely accepted alternative to enucleation for medium-size posterior uveal melanomas (< 10 mm in height and 15 mm in diameter)[15]. It has similar indications and success rates as external beam irradiation.

Plaques containing radioactive isotopes of iodine-125 are attached temporarily to the sclera and limbus underlying the melanoma.

Block excision or sclerouvectomy-

It is an alternative treatment method for ciliary body melanomas covering less than 4 clock hours of the circumference[16].It consists of a full-thickness excision, with in-block removal of the ciliary body, cornea, iris, and sclera, with a 2-mm to 3-mm margin of healthy tissue around the tumor, followed by the grafting of banked sclera and cornea to close the defect.

Retinal detachment, vitreous hemorrhage, and cataract are common complications. These risks are improved by a modified approach, lamellar sclerouvectomy, which uses a scleral flap and minimizes altering of the retina and vitreous. Other procedures

Laser photocoagulation and transpupillary thermotherapy are used in selected small choroidal melanomas, but it is not usually useful for ciliary body melanomas.

Orbital exenteration is a radical treatment reserved for cases with extensive orbital extension. The usefulness of such disfiguring surgery is not established. Patients with such advanced melanomas are likely to have extensive distant metastases and poor prognosis for survival, with or without orbital exenteration surgery.

Conclusion

Ciliary body melanoma is a rare tumor in the Indian population; however, its . Since it is in a hidden location behind the iris, ciliary body melanoma usually attains a relatively large size before it is detected clinically. It can be easily misdiagnosed and has a poor prognosis if detected late. Other differential diagnoses should be excluded and kept in mind. Currently, the most effective measure to minimize poor prognosis is early detection of melanoma at a time when the tumor is small and at least risk for metastatis.

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