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Case Series

A series of choroidal melanoma in young patients: insights from a tertiary care facility

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ABSTRACT

Choroidal melanoma is rare in young individuals and often presents at a later stage due to low clinical suspicion. This case series describes a group of young patients, all under the age of 30 years, who presented with advanced choroidal melanoma requiring enucleation. A prospective observational study was conducted from January 2019 to December 2023 at a tertiary care center. Twelve young patients diagnosed with choroidal melanoma were assessed. Patient's demography, clinical presentation, tumor characteristics and histopathological findings were noted. Histopathologically tumors were classified as epithelioid, mixed and spindle cell type. The treatment protocol was devised based on the size, extent of the lesion and any involvement of systemic system. There were 10 females and 02 males. The mean age of patients in the current study was 23±3.5 years. Right eye was involved in 05 patients and left eye was involved in 07 patients. All patients had advanced choroidal melanoma at the time of presentation at our center. The mean basal diameter of the tumor was 21±2 mm and thickness were 11±1.5 mm. Primary enucleation with implant was the most common treatment modality in all patients. Epithelioid cell type choroidal melanoma was the commonest histology pattern was found in the current study. No mortality or death occurred at the end of 02 years follow-up in any patient. This series highlights the aggressive nature of choroidal melanoma in young individuals when diagnosed at an advanced stage. It emphasizes the importance of early clinical suspicion, timely referral, and appropriate imaging in any atypical choroidal lesion in the young population.

Keywords: Young, Choroidal melanoma, Incidence, Large, Enucleation, Epithelioid

INTRODUCTION

The incidence of choroidal melanoma, which constitutes 85% of the uveal melanoma, worldwide is 5 per million and in Asian population is 0.15-0.18 per million.[1,2] As per existing literature, it is considered to be disease of elderly age group (40 to 60 years) and is rarely seen in young population.[3,4] The common risk factors for occurrence of choroidal melanoma are advanced age, fair skin, light colored eyes, choroidal naevus, a family history of uveal melanoma and exposure to ultraviolet light, however the role of exposure to ultraviolet light remains inconclusive in manifestation of choroidal melanoma as per established studies.[5-10] The diagnosis of this entity is done on the basis of clinical, ultrasound and magnetic resonance imaging features. Most of the time it is not diagnosed on time as patients seek ophthalmic consultation when they develop sudden loss of vision. Further, the rate of diagnosis of choroidal melanoma in

young patient is dismal as this diagnosis is not considered presuming that it is a disease of elderly group, which can lead to catastrophic outcome in this subset of patients.[11] Incidence of choroidal melanoma is less common in the Indian population and in younger population it is rarely seen as per various published studies.[12-14] There are very few reports which have commented on occurrence of choroidal melanoma in young Asian population. Hence, the primary aim of presenting this case series is to raise awareness among practicing clinician that this entity can occur in younger age group and due diligence to be observed while dealing with young patient who presents with loss of vision and a choroidal mass.

CASE SERIES

It was a prospective observational study which was conducted from January 2019 to December 2023 at a tertiary care center in the northern part of India. During this period, total 12 patients younger than 30 years were seen during this period. The study was approved by a local ethical committee vide no Army Hospital Research and Referral, Delhi 243. This study adhered to the tenets of the declaration of Helsinki. Informed consent was taken from all the study participants stating that they are part of the study. Demographic characteristics like geographic location gender, age and laterality, onset of symptoms, history of prior systemic condition, details of previous treatment history were noted. All patients underwent assiduous eye exam which included recording of visual acuity and intraocular pressure, slit lamp examination, fundoscopy. Ancillary investigations like fundus fluorescein angiography and optical coherence tomography were advised in all cases. Imaging like B-scan USG and magnetic resonance imaging (MRI) with contrast were done in all cases (Figure 1 A and B). All patients were evaluated by oncophysician and positron emission tomography (PET) scan to detect systemic risk factors and to rule out systemic metastasis. Details of lesion like location, basal diameter, thickness of tumor, color, tumor configuration, associated subretinal fluid and any extraocular extension of tumor were documented.

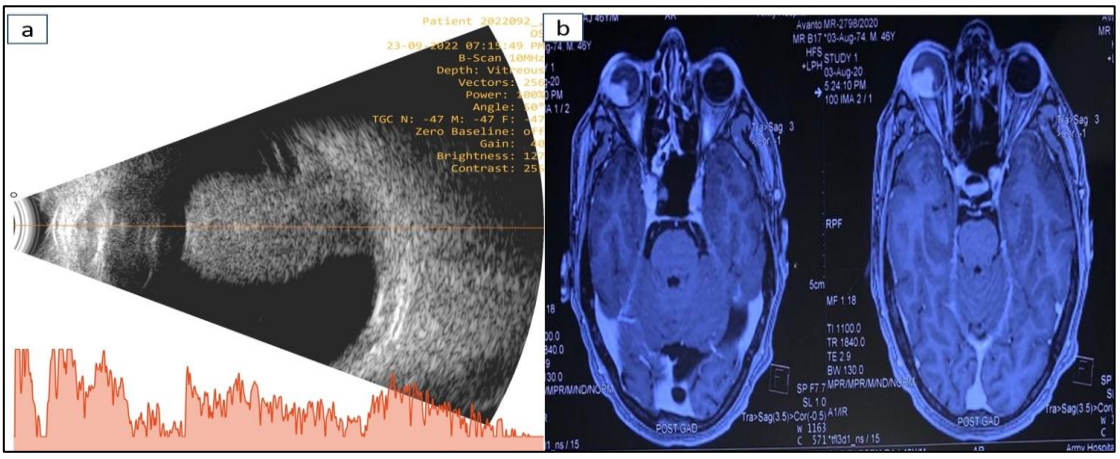

Figure 1 (a and b): Ultrasound B scan of hyperechoic mushroom-shaped intraocular mass. MRI orbit showing intraocular lesion in posterior part of eyeball with no involvement of optic nerve and no sign of extraocular extension.

Treatment plan

At our center, treatment protocol was formulated based on the dimension of the tumor and laterality. Enucleation with implant was recommended when the basal diameter of the tumor was more than 18 mm in size and thickness more than 10 mm in size. Treatment for salvaging globe was advised when basal diameter of tumor was less than 16 mm in size and thickness was less than 8 mm in size.

All patients were followed up for 02 years. Patients were counselled to come for periodic follow-up every 03 months for 1st two visits and then every 6 months. Systemic evaluation was done by medical oncologist at our center to rule out systemic involvement. Following surgery patient was evaluated again twice in a year to detect metastasis at an early stage. Physical examination and liver function test, serum LDH, chest x-ray and ultrasound abdomen were done at each visit.

All 12 patients were found to have advanced choroidal melanoma when they reported at our center for further management. The mean age of the study population was 23.5±3.5 years. Maximum patients were from western and northern part of India and were referred from peripheral hospital. There were 02 male patients and 10 female patients in the present study. Right eye was found in 05 patient and left eye was involved in 07 patients. All patients had unilateral presentation in the present study. The demographic and clinical profile of patient are depicted in Table 1. The common symptom with which patient presented in the current study was sudden loss of vision. The usual sign was detached retina in maximum patients (Figure 2 A and B) The common tumor configuration was an elevated, dome shaped lesion (Figure 2 C). The basal diameter of the tumor in most of the cases was more than 21±2 mm in size and tumor thickness was more than 11±1.5 mm in size in the study population (Table 2). Anterior segment finding showed

quiet eye in most of the patients. Systemic association was not found in any patient in the present study group. Neovascularization of iris and raised intraocular pressure was found in one patient. The most common primarily treatment modality was enucleation with implant (Figure 2 D), as basal diameter of the tumor was more than 18 mm in size and thickness was more than 10 mm in size in maximum patients in the current study all enucleated eyeball were sent for histopathological examination which confirmed the diagnosis of choroidal melanoma. Cytology showed epithelioid cell melanoma in maximum number of enucleated eye ball specimen (Figure 2 E and F). None of the patient was found to have systemic metastasis at the time of presentation at our center and no mortality was reported at end of 1 year. Extra-scleral extension was not found in any patient in the present study population. Mean period between presentation and surgery 7 days at our center. In current study, eye globe could not be salvaged in any patient as all of them had advanced stage of intraocular choroidal melanoma when they sought consultation ay our center. Clinical outcomes in the current study have been shown in Table 3.

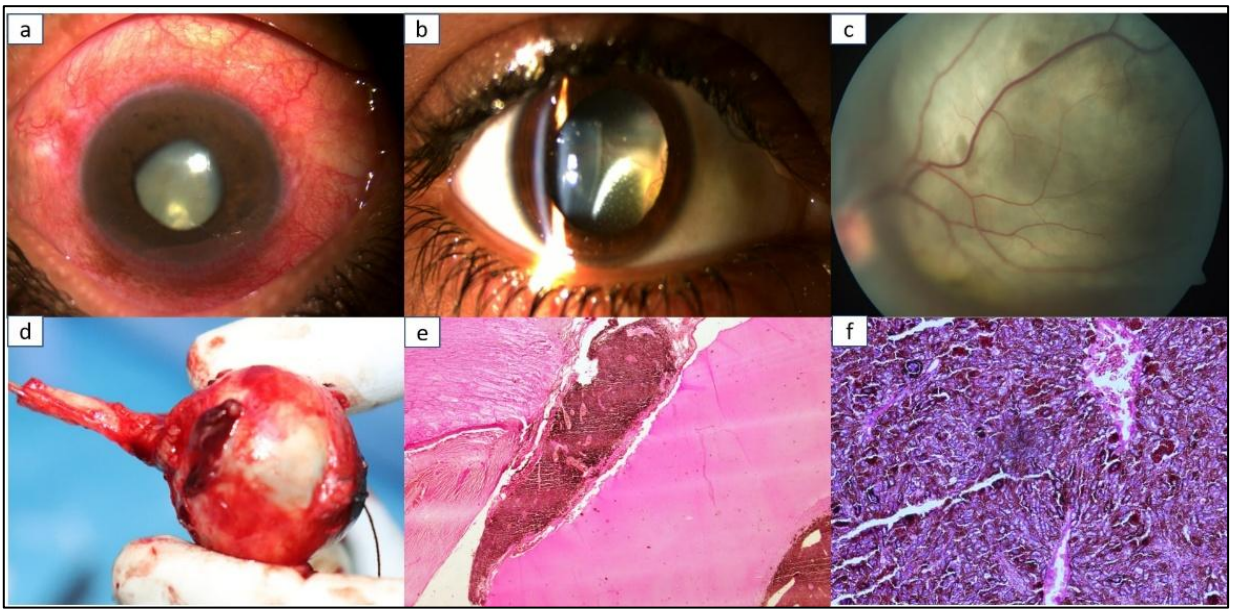

Figure 2 (A-F): Slit lamp photograph of congested conjunctiva, shallow anterior chamber and mass behind lens. Fundus image showing an elevated, dome shaped, pigmented lesion with associated retinal detachment. Clinical image showing enucleated eyeball with optic nerve. Histopathological features 2× and 40× showing epithelioid type choroidal melanoma.

Table 1: Demographic profile and clinical features of choroidal melanoma in young patients in the current study.

Features	N	Percentages (%)
Age at initial presentation (in years), mean±SD		23.5±3.5
Gender		
Male	02	16.6
Female	10	83.3
Laterality		
Unilateral	12	100
Bilateral	0	0
Affected eye		
Right eye	05	41.6
Left eye	07	58.3
Family history of choroidal melanoma		Nil
Common clinical presentation		
Sudden loss of vision	12	100
Redness	03	25
Pain	03	25
Raised Intraocular pressure	02	16.6
Iris neovascularization	01	8.3
Common fundus features		
Elevated lesion with exudative retinal detachment	12	100
Ocular and systemic association	Nil	

Table 2: Tumor characteristics and histopathological features of choroidal melanoma in the study population.

Features	N	Percentage (%)
Tumor basal diameter		
Mean±SD	21±2 mm	
20 mm	12	100
<20 mm	0	
Tumor thickness		
Mean±SD	11±1.5 mm	
10 mm	12	100
<10 mm	0	
Tumor configuration		
Dome shaped	10	83.3
Mushroom shaped	02	16.6
Tumor color		
Pigmented	11	91.6
Non pigmented	01	8.3
Associated features		
Subretinal fluid	12	100
Vitreous hemorrhage	01	8.3
Extraocular extension	Nil	
AJCC clinical staging, 8th edition		
T1	Nil	
T2	Nil	
T3	Nil	
T4	12	
T4a	12	
T4b	Nil	
T4c	Nil	
T4d	Nil	
Histopathological features		
Epithelioid type	10	83.3
Mixed cell type	02	16.6
Spindle cell type	Nil	

Table 3: Clinical outcomes in our study.

Outcome parameters	N	Percentages (%)
Primary treatment modality		
Enucleation with implant	12	100
A primary outcome		
Globe salvage rate	Nil	
B secondary outcome		
Complications		
Implant exposure	02	16.6
Recurrence	Nil	
Systemic metastasis	Nil	
Death	Nil	

DISCUSSION

Incidence of intraocular choroidal melanoma increases steadily with the age, but their incidence among younger population is rare.[15-20] Choroidal melanoma in younger population manifest when they attain pubertal age.[21-23] The possible reason for this is that around puberty, there is a spurt in growth hormone which leads to occurrence of this entity in this age group. Choroidal melanoma in younger patient is of smaller size and the incidence of iris melanoma is higher in this group. Females are found affected more in younger patients diagnosed with choroidal melanoma. The common association which is found in this age group is oculo (dermal) melanocytosis. The survival rate is higher in younger patients affected with choroidal melanoma as lesion is limited to the iris and mortality risk factors are fewer in this group. But, young patients with choroidal melanoma face a lot of hurdles in getting appropriate treatment as this entity is normally not contemplated in this age group by most of clinical practitioners leading to poor prognosis in these patients in terms of grim globe salvage rate and poor survival rate. Hence, sensitization of the environment regarding increased rate of choroidal melanoma in younger population in current era is warranted which will help in prompt diagnosis of this entity in this age group. The disease should be treated as aggressively in younger population as we treat elderly age group as prognosis is bad in all age groups if size of the tumor is massive and in presence of extraocular extension.[24-30]

Our case series has demonstrated that frequency of choroidal melanoma in young Indian population has mounted. The mean age of presentation was 23±3.5 years and all patients were below 30 years of age. This is in discordance with shield et al and the collaborative ocular melanoma study which reported mean age of the patient was 60 years and inferred that choroidal melanoma is a disease of elderly age group.[20,31] The current study's finding is in conformity with Biswas et al which inferred that Asian patients present with choroidal melanoma at a younger age than Caucasians and Americans.[13,14] In their study the mean age at presentation was 45.94±14.85 years (range 2-76 years). Among these, 58 (51.3%) patients were in the 40-60 years age group, while 6 (5.3%) patients were less than 20 years old. Egan et al reported that in Asians it occurs more in young and middle-aged people.[30]

The disease was found more common among females in the current study. This is in conformity with Singh et al, Shield et al and Matthew et al which also reported female predominance in their studies.[11,20,27] As per Conte et al and the collaborative ocular melanoma study, choroidal melanoma occurred more in men compared to female.[16,31]

In our series all cases had unilateral advanced choroidal melanoma and bilateral choroidal melanoma was not found in any patient. This is in conformity with Singh et al, Dhupper et al and Shield et al as they also found choroidal melanoma in only 1 eye in their study.[1,4,7,11,12,20,21]

The commonest symptom in the current series was sudden loss of vision for which they sought consultation at our center. This is in discordance with various studies like Dhupper et al and Shield et al which reported

blurring of vision as the common symptom with which patient presented in their studies.[12,20,21] The common sign was subretinal mass associated with retinal detachment. This is in concordance with Dhupper et al as they noticed presence of an elevated mass with exudative retinal detachment in maximum patients.[12]

In this case series, all tumors were large-sized lesion as basal diameter of the tumor was more than, 18 mm and thickness was more than 10 mm. However, Shield at al found smaller size of the tumor in younger population, which was further from foveola and optic disc. They reported mean basal diameter of the tumor was 9.8 mm and mean tumor thickness was 5 mm.[20,21] Dhupper et al reported in their study that maximum number of patients had medium-sized tumor. The mean basal diameter of the tumor in their study was 12. 41±1.41 mm while the mean height was 8.91±4.16 mm.[12]

None of the patient in the current series were found to have any risk factor and any systemic association.

Singh et al reported the only associated ocular finding was oculo (dermal) melanocytosis in 7 patients (11%). They also revealed that the oculo (dermal) melanocytosis was diffuse, involving all quadrants of the uvea, in 6 cases and sectoral in 1 case, with choroidal melanoma occurring in the sector with melanocytosis. The same study has observed that compared with the general population of patients with uveal melanoma, oculo (dermal) melanocytosis was 9 times more common in young patients with uveal melanoma. The associated systemic findings included familial uveal melanoma (1 case), dysplastic nevus syndrome (2 cases), and history of cutaneous melanoma (1 case) was found in their study.[6,7] None of the patients had a personal history of prior or subsequent development of a second primary malignant neoplasm. Gunduz et al and Seddon et al reported common risk factor associated with choroidal melanoma was ocular melanocytosis.[9,10]

Shield et al found common association in younger population was ocular melanocytosis as it was found in 4 patients affected with choroidal melanoma in their study.[21]

Histopathological examination in the current case series has shown epithelioid cell type cell melanoma in maximum number of patients and in 02 patients mixed cell tumor type was found. This is in contrast to Dhupper et al as they reported mixed cell type of choroidal melanoma followed by spindle cell type.[12] Singh et al divulged spindle cell melanoma as commonest type followed by mixed cell and epithelioid type melanoma.[11]

The common treatment modality in our case series was enucleation with implant as size of the tumour was more than 18 mm and thickness more than 10 mm in size. Globe salvage treatment was not attempted in the current study due to massive size of the tumour. This finding was in discordance with Singh et al, Dhupper et al and Shield et al as they offered various treatment modality like laser photocoagulation, transpupillary therapy, local tumor resection, plaque brachytherapy or enucleation as different sizes of the tumor was found in their studies.[4,7,11,12,20,21]

The various treatments which have been formulated over the period of time for treating choroidal melanoma is based on the size of the tumor and at what stage of the disease patient seek ophthalmic treatment. The treatment options are limited when tumour attains a massive size. It is better to enucleate when tumor is of large size as any attempt to salvage globe at this stage will endanger the life of patient as risk of metastasis and rate of recurrence increases with incomplete regression of the tumor which is bound to happen with globe salvage treatment in this subset of patients.

This case series has shown that choroidal melanoma should not considered as a disease of elderly population as its frequency has increased in younger patient in Indian subcontinent. We speculate the cause of rise in occurrence in younger generation most probably is due to excessive exposure to UV light. The other factors responsible for the significant increase in choroidal melanoma in younger population could not be ascertained by the authors in the current study.

The strength of the current series is that it is the first series from Indian subcontinent which has commented exclusively on choroidal melanoma in younger population. Though the outcome was not heartening as we had to enucleate all eyes but still, we are reporting this finding with an aim to share the outcomes to alert clinicians about increased rate of the occurrence of choroidal melanoma in younger population.

CONCLUSION

Choroidal melanoma should not be reckoned as a disease of elderly people as increase in occurrence of uveal melanoma among young Indian population has been found in the current series. The prognosis is abysmal in choroidal melanoma regardless of the age group in which it occurs if the lesion attains a huge size. A high index of clinical suspicion is warranted to diagnose choroidal melanoma in younger population for early diagnosis and to achieve better outcomes in this age subgroup.

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