***Case Report***

# CATCHING A RARE KILLER– CHOROIDAL MELANOMA

Stella Sinnappan[1,2], Jemaima Che Hamzah[2], Chandramalar T Santhirathelagan*[1]

[1]Department of Ophthalmology, Hospital Sungai Buloh, Jalan Hospital, 47000 Sungai Buloh, Selangor, Malaysia.
[2]Department of Ophthalmology, Faculty of Medicine, University Kebangsaan Malaysia Medical Centre, Jalan Yaacob Latif, Bandar Tun Razak, 56000 Kuala Lumpur, Malaysia.

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*Corresponding author:*
Dr. Chandramalar T Santhirathelagan

*Email address:*
chandramalarhsb@gmail.com

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**ABSTRACT**

*Choroid melanoma though rare, is the most common primary adult intraocular malignancy and is more common among Caucasians. We present a case of choroidal melanoma rarely seen in the Malay ethnic group where a male patient who was an active smoker for 16 years with underlying colon adenocarcinoma presented with left blurring of vision and pain associated with headache for a duration of one month. He perceived shadowing of the superior visual field of left eye associated with floaters with no preceding trauma or constitutional symptoms. The best corrected visual acuity( BCVA) was 6/9 in the right eye,and 6/12 in the left eye. Left relative afferent pupillary defect and red desaturation were detected. Anterior segment examination revealed a hyperpigmented lesion involving the iris and anterior chamber angle inferiorly. A brownish, smooth dome shaped mass with presence of sentinel vessels was seen on funduscopy. B-scan confirmed an intraocular mass with exudative retinal detachment. Magnetic resonance imaging of orbit showed left intraocular lesion suggestive of choroidal melanoma with no extraocular extension or intracranial involvement. Choroidal melanoma is very rarely seen among the Malaysian population, more so in Malay race. This case underscores the rarity of melanoma in this population and illustrates the need for awareness in both Caucasian and non-Caucasian populations. Despite effective primary therapy, nearly 50% of patients will develop metastatic disease therefore the need to tailor the treatment to the individual patient's clinical findings.*

## INTRODUCTION

Melanoma arising from choroid is the most common primary intraocular malignancy in adult [1,2]. The mean age-adjusted incidence of uveal melanoma in the United States has remained stable approximately 5.1 per million population since 1970s and it is more common in Caucasians and rarely found in Asian population [2,3]. The incidence of occurrence of choroid melanoma is around 20 per million cases per year globally [4].

This malignancy typically starts as a dome shaped lesion. As they grow and break through the Bruch membrane, they acquire a mushroom or collar-button shape. Extension of choroidal melanoma cause serous detachment of adjacent retina, degenerative changes in outer segment of photoreceptors and may extend through scleral emissary channels to gain access to episcleral surface and orbit. Direct access to anterior chamber may lead to secondary glaucoma.

Choroidal melanoma predominantly appears at 6$^{th}$ decade of life with no gender predilection. The incidence is higher in individual with fair complexion, light eye colour, ocular melanocytosis, dysplastic nevus syndrome, the presence of a germline BRCA1 associated protein 1 (BAP1) mutation, chronic exposure to ultraviolet light and smoking [2,5]. We report a rare case of choroidal melanoma in an ethnic Malay patient.

## CASE REPORT

A 46-year-old Malay gentleman, who was an active smoker for16 years, with underlying colon adeno-carcinoma, presented with left blurring of vision associated with pain and headache for 1 month. He perceived superior visual field defect in the left eye associated with floaters, but denied any preceding trauma or constitutional symptoms. He was diagnosed to have colon carcinoma in 2007 and underwent surgical resection in the same year. There was no evidence of metastasis during follow ups.

On examination, the BCVA were 6/9 and 6/12 in the right and left eye respectively. Left Relative afferent pupillary defect (RAPD) was detected with marked red desaturation and reduced light brightness. Left anterior segment showed hyperpigmented lesion on iris and angle inferiorly with otherwise quiet anterior

chamber (Figure 1: Image A and B). Left funduscopy revealed a brownish, smooth dome shaped mass with sentinel vessels seen temporal to macula, associated with retinal detachment. (Figure 2: Image C) There were no significant optic disc findings (figure2: Image D). Systemic examination was unremarkable with no lymphadenopathy and tumour marker investigation done was normal

B-Scan ultrasonography of the affected eye showed a mushroom shaped intraocular mass with a highly reflective anterior border and exudative retinal detachment (Figure 3). Subsequent fundus fluorescence angiography (FFA) showed patchy areas of hyperflorescence overlying tumour surface with extensive leakage but no obvious double circulation seen (Figure 4).

Magnetic resonance imaging of orbit and brain demonstrated a well-defined intraocular lesion along the inferolateral aspect of the left eye suggestive of choroidal melanoma with no extraocular extension or intracranial involvement (Figure 5). Based on our clinical and imaging findings, a diagnosis of left eye choroidal melanoma was made and patient was referred to an ocular oncologist. Patient was then counselled for enucleation of the left eye.

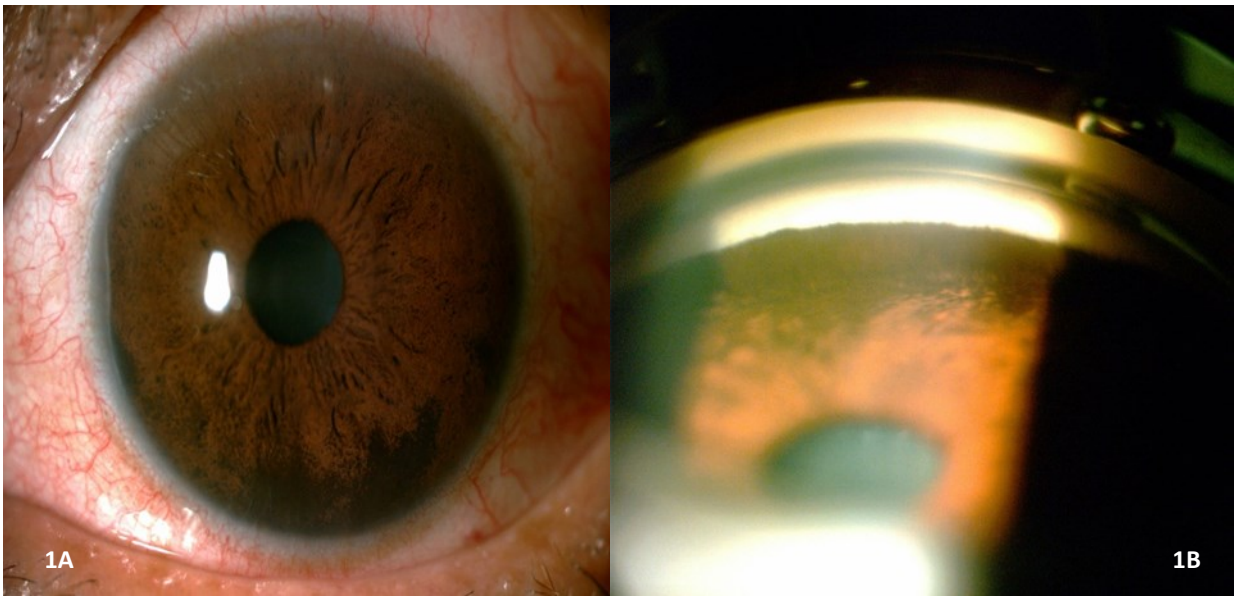


Figure 1: Left anterior segment.(A) Hyperpigmented lesion over the iris. (B) Hyperpigmented lesion involving the angle.

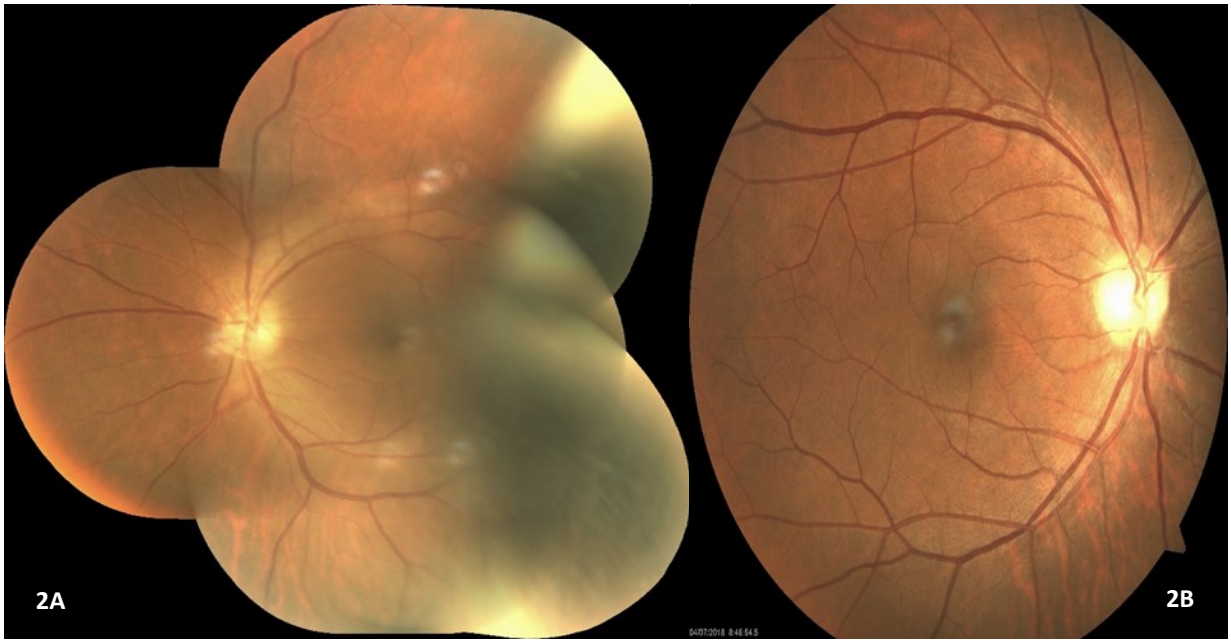


Figure 2: (A) Left fundus shows a brownish, smooth dome shaped mass at temporal aspect.
(B) Normal right fundus

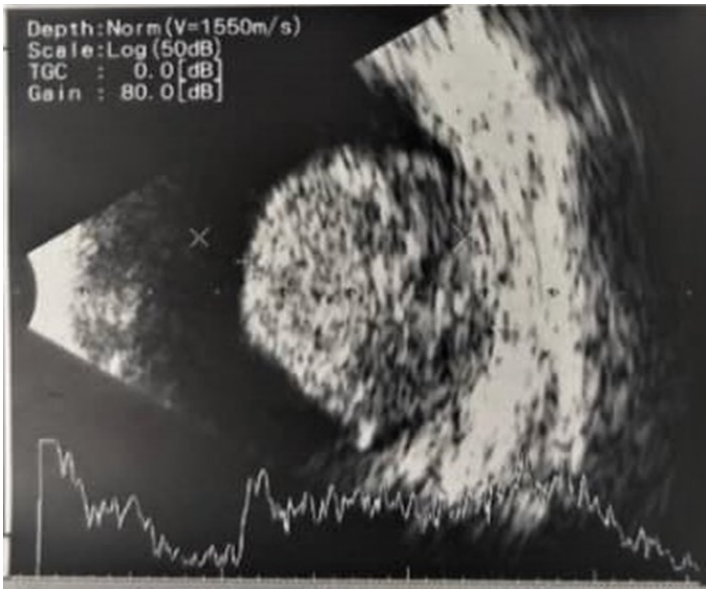


Figure 3: B-Scan shows a mushroom shaped intraocular mass with a highly reflective anterior border and exudative retinal detachment.

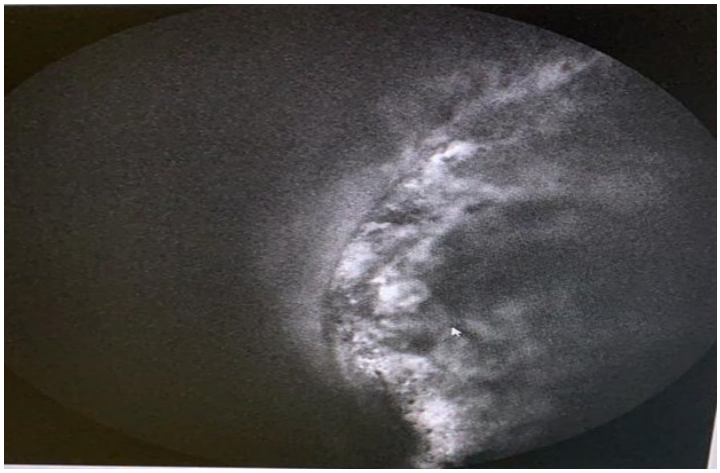

Figure 4: Left FFA shows patchy areas of hyperfluorescence overlying tumour surface with no obvious double circulation seen.

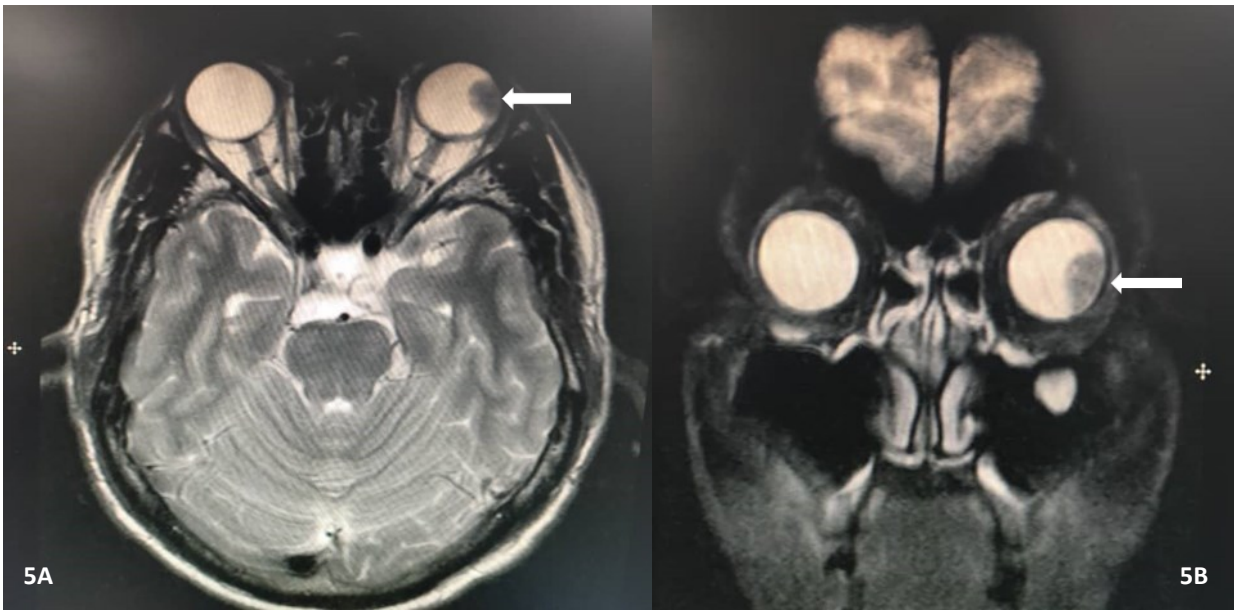


Figure 5: MRI of the orbit on axial view (5A) and (5B) coronal view T2-weighted image shows a well-defined intraocular lesion in the inferolateral aspect of the left eye (white arrow)

## DISCUSSION

Uveal melanoma is the commonest primary malignant intraocular tumour in Caucasian adult with a lifetime incidence of about 0.05% [6]. The incidence of Choroidal melanoma in the United States is 5.1 per million population [2]. The risk of choroidal melanoma among Caucasians was 8-fold that of African Americans [6]. To date, many studies have focused on the Caucasian population due to lack of data in other population.

The incidence of choroidal melanoma is so rare in Malaysia that only 8 cases reported in a retrospective study from 2012 till 2016, in a tertiary referral center in Malaysia. Among that 8 cases, 5 patients were Chinese female, 2 Malay female and 1 Malay male who was an ex-smoker with underlying renal carcinoma [4]. Again, it shows how rare for our patient to get it as a Malay male who also has smoking as a risk factor. In this retrospective study 4 patients underwent enucleation, 1 patient subjected for lid sparing exenteration and 2 of them treated with plaque brachytherapy and 1 patient underwent stereotactic radiotherapy.

Nicholas A *et al* documented a case of uveal melanoma in a 65-year-old woman of African American descent and with moderately light skin pigmentation in Philadelphia [5]. This patient also had a history of pulmonary large cell lymphoma, which was successfully treated with chemotherapy and radiotherapy 25 years prior with no history of recurrence. She presented with superonasal choroidal mass of 12 mm in basal diameter and 5 mm in thickness. Given the location and size of the tumour, plaque radiotherapy using iodine-125 isotope was provided to this patient. The author have concluded that, this patient could have represented some Caucasian lineage over the past generations in view of her skin colour which was lighter than most African American complexion.

CL Shields *et al* in their retrospective, nonrandomized, interventional case series, reviewed the clinical features and prognosis of uveal melanoma based on race in 8100 patients in the Caucasian versus non-Caucasian populations [6]. They found that Caucasians presented with uveal melanoma at an older age (58 years) compared to non-Caucasians (44–52 years; $P<0.001$) and clinically, Caucasians showed melanoma with significantly more posterior location, closer to the foveola, and optic disc, with smaller basal dimension and thickness, and with less Bruch's membrane rupture and less intraocular hemorrhage, compared to non-Caucasians but the outcomes of metastasis were similar between them. They also noted that most reports on prevalence data and outcomes focus on Caucasians with little data on other races.

Currently, the changing demographic of certain population in many countries illustrates the need for a more comprehensive understanding of this disease across all racial groups and that racial differences in the incidence of uveal melanoma should be well recognized. Dan-Ning Hu *et al* have investigated the racial/ethnic differences in the development of uveal malignant melanoma in a large population-based study (Black. Asian, Pacific Islander, Hispanic and non-Hispanic White) [7]. The result shows that the difference in the incidence between each racial/ ethnic group was highly statistically significant with the exception of the Black versus Asian population. They stated that the relative risk of uveal melanoma was 1.2 for Asian and Pacific Islander patients, 5.4 for Hispanic patients, and 19.2 for non-Hispanic white patients as compared with the black patients. This finding is consistent with known data in cutaneous melanoma. Therefore, they have concluded that the amount of racial pigmentation may not be the only risk factor that is associated with uveal melanoma.

Apart from rare ethnicity of my patient, he also had an history of previous malignancy. Collaborative Ocular Melanoma Study (COMS) have reported that 5% of deceased individuals with uveal melanoma had a history of malignancy as seen in our patient, however few other studies concluded that there was no or only a weak association between previous malignancies and increased risk of uveal melanoma [8].

The primary goal of treatment for uveal melanomas is to prevent metastasis. Nearly 50% of patients will develop metastatic disease, despite effective primary therapy; with the most common initial site being the liver [8]. The management of choroidal melanoma depends on patient's age, general health, preference, status of fellow eye, tumour size and location.

## CONCLUSION

Choroidal melanoma is a rare tumor but associated with high mortality rates. Even though it is typically found in Caucasians with light-colored eyes and fair skin with a propensity to burn when exposed to ultraviolet light, high index of suspicion is needed when typical presentation is seen in a non-Caucasian person. This case underscores the rarity of melanoma in this population and illustrates the need for full ocular examination in both Caucasian and non-Caucasian populations.

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