Choroidal melanoma in a young patient.

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Background: *To report a case of choroidal melanoma in a young Asian lady.*

Case Report: *A 36 year-old healthy Malay lady presented with left eye progressive, painless, decreasing vision over a year with new onset of excruciating eye pain. Left eye examination showed no light perception, conjunctival chemosis, cornea edema and total hyphema with no fundus view. Intraocular pressure was 42mmHg. B scan ultrasonography revealed retinal detachment with subretinal mass. Right eye was normal. Systemic examination and investigations were unremarkable. Blood investigations including tumour markers were normal. Magnetic resonance imaging of orbits was suggestive of choroidal melanoma with retrobulbar extension. Left eye enucleation was performed and histopathology was consistent with choroidal melanoma. The patient remains under close follow-up with no evidence of recurrence or metastasis.*

Conclusion: *Although predominantly a disease in elderly, choroidal melanoma needs to be suspected in younger patients with intraocular lesion.*

Conflict of interest: *none.*

Keywords: *Choroidal melanoma; enucleation.*

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Introduction

Uveal melanoma is the most common primary ocular malignancy in adults, accounting for 3-5% of all melanomas.[1] It usually affects elderly Caucasians of northern European descent during the fifth to sixth decade of life. Its incidence in Asians (0.39 per million) and blacks (0.31 per million) is significantly less than that of Caucasians (6.02 per million).[1]

Objective

To report a case of choroidal melanoma in a young Asian lady.

Method

Case report

Results

A 36 year-old Malay lady was referred for left eye acute glaucoma. She complained of left eye progressive, painless, decreasing vision which was preceded by flashes over a year with new onset of excruciating eye pain for two days. She had no known medical illness.

Ocular examination of the left eye showed no light perception, conjunctival chemosis, cornea edema and total hyphema secondary to neovascular glaucoma with no fundus view. Intraocular pressure was 42mmHg. B-scan ultrasonography revealed retinal detachment with subretinal mass (Figure 1). Right eye was normal. Magnetic resonance imaging of orbits was suggestive of choroidal melanoma with retrobulbar extension (Figures 2a-b).

Figure 1 : B-scan ultrasonography of the left eye at initial presentation, showing retinal detachment with subretinal mass (*)

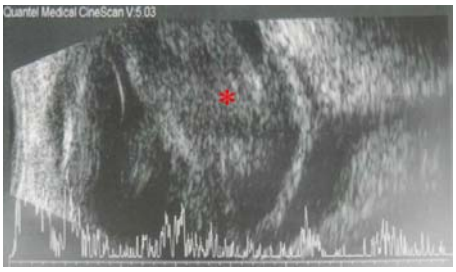


Figure 2a: Axial T1W FS pre-gadolinium contrast.

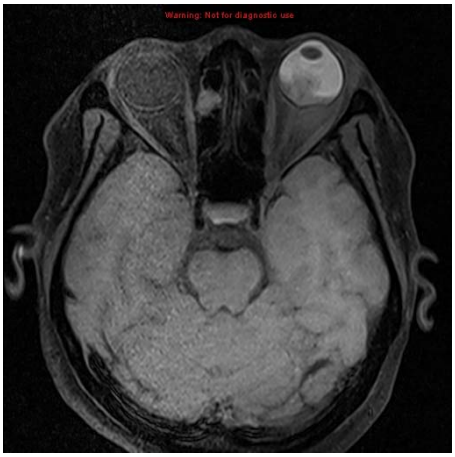
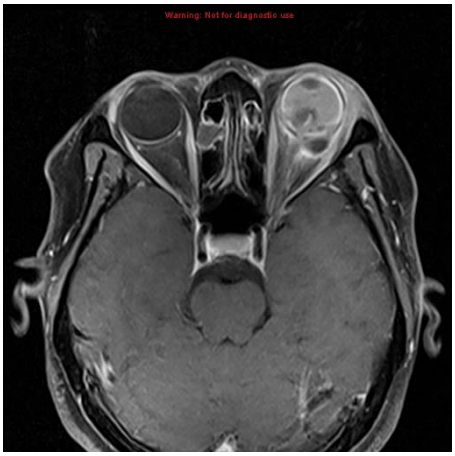


Figure 2b: Axial T1W FS post-gadolinium contrast.



Systemic examination was unremarkable. Metastasis work-up including tumour markers, oesophagogastroduodenoscopy, colonoscopy, chest X-ray, computed thomography (CT) scan of the thorax, abdomen and pelvis were normal.

Left eye enucleation was performed. Cut section showed a solid yellow green tumour arising from the choroid, protruding into vitreous body and extending to the posterior extraocular area. The tumour measured 24mm in basal diameter and 7mm in thickness. Surgical margins were negative. Optic nerve and extraocular muscles were spared. Histopathology confirmed a mixed cell variety of choroidal melanoma (Figures 3a-d) with a pathological staging of pT4d Nx.

Figure 3a-d: Histopathology confirmed a mixed cell variety of choroidal melanoma.

Figure 3 a: Overview of the left eye harbouring choroidal melanoma, optic nerve spared (arrow), S-100 stained, VLP

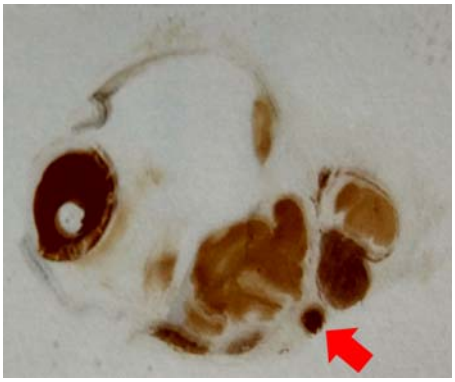


Figure 3b: Staining with S-100, diagnostic marker of uveal melanoma, 40x

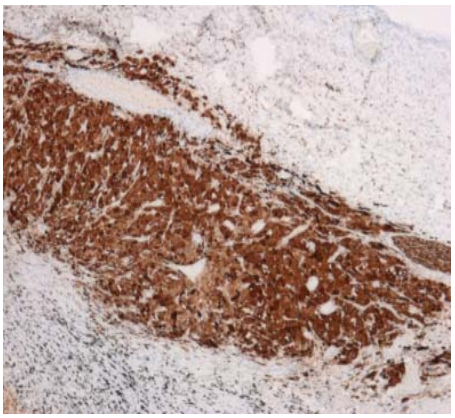


Figure 3 c: Presence of epithelioid cells, H&E stained, 200x

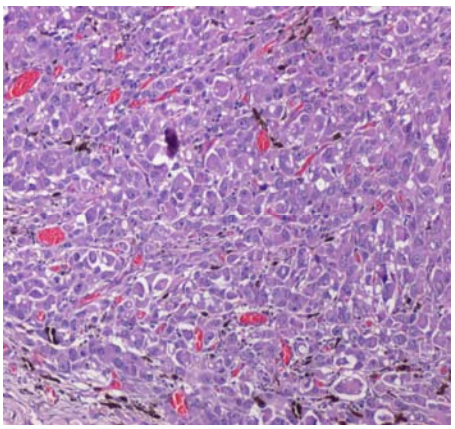
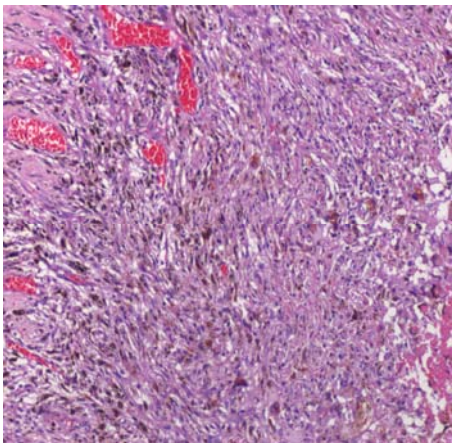


Figure 3 d: Presence of spindle cells, H&E stained, 200x



She was subsequently referred to a visiting oncologist. Chemotherapy or radiotherapy was not required as magnetic resonance imaging of orbits post enucleation revealed no sign of tumour recurrence. Prosthetic eye was offered to her. She remains under close follow-up with no evidence of recurrence or metastasis for the past one year.

Discussion

Melanoma arises from melanocytes at various anatomic locations of the body, ocular being the second most common site after skin. 83% of ocular melanoma arises from the uveal tract,[2] with choroid being the most frequent site of origin and comprises 90% of cases.[3]

The overall mean age-adjusted incidence of primary uveal melanoma is 5.1 cases per million inhabitants per year in the United States and significantly lower in Asia countries.[1] Its incidence is highest among White population, and rarely occurs among Asian, Black, or Hispanic population.[4] The median age of diagnosis is 62 years, with a peak incidence at the 7th decade of life.[1] Shields et al. compared the mean age of presentation among different races and concluded that Asians present with uveal melanoma at a younger age.[5] Studies also show a

slight male predilection with male to female rate ratio of 1.29.[1] Risk factors include White race, ocular melanocytosis, and presence of cutaneous melanocytic lesions.[6] Our patient was a relatively young Asian lady with no co-morbidities or predisposing risk factors.

Presentation may vary from asymptomatic to visual loss in the affected eye, depending on the size and location of the tumour. Choroidal melanoma can cause Bruch's membrane rupture and retinal detachment whereas iris involvement may lead to hyphema and secondary glaucoma,[5] all of which were observed in our patient. Dilated fundus examination is mandatory and B-scan ultrasonography should be carried out if opaque media precludes clinical examination. Computed tomography (CT) and magnetic resonance imaging (MRI) are useful to assess the extent of the tumour and rule out metastases if present.

Management of uveal melanoma depends on the tumour location, size, local extension and systemic status. The main treatment options include enucleation, plaque radiotherapy, proton beam ratiotherapy, transpupillary thermotherapy (TTT), and orbital exenteration for tumours with orbital tumour extension.[4,6] Our patient underwent enucleation even though there was extraocular extension as she refused for orbital exenteration. Some of the novel therapeutics that have emerged include ICON-1 and AU-011 for primary disease as well as immune-based therapy IMCgp100 for metastatic uveal melanoma.[7,8] Based on the Collaborative Ocular Melanoma Study (COMS), the 5-year and 10-year rates of metastatic disease were 25% and 34%, respectively.[4] Thus, long term regular systemic follow-up is necessary as prognosis is poor once metastases have developed.

Conclusion

Although predominantly a disease in elderly Caucasians, the possibility of uveal melanoma should not be overlooked in younger patients of other races with intraocular lesion. Uveal melanoma is a known masquerader and may perplex the clinician. Misdiagnosis is common if not properly evaluated and investigated, causing delay in treatment and development of metastatic disease.

Declarations of interest: none.

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