Case Report

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Malignant choroidal melanoma with suspected vitreous seeding - A case report

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Abstract:

Retinal perforation with vitreous seeding is an uncommon condition of treatment-naïve choroidal malignant melanoma. We reported a 52-year-old male who had a pigmented tumor protruding from choroid of his right eye. He had only black shadow sensation for 4 months then a rapid deterioration of vision. Fundus examination showed vitreous haze with many pigmented materials. B-scan ultrasonography revealed a mass with low internal reflectivity and vitreous opacity. The eyeball was enucleated and a stage IIIA melanoma was confirmed by the pathologist. No local recurrences or metastases were found during 31-month follow-up. Although vitreous seeding may indicate rapid tumor growth, early enucleation may insure a better prognosis.

Keywords:

Choroid neoplasms, melanoma, neoplasm seeding

Introduction

Uveal melanoma is the most common primary intraocular tumor in adults. It is also the main fatal intraocular disease.[1,2] Most uveal melanoma may invade into the sclera or continuous horizontal growth into the adjacent ciliary body. Although the tumor may invade into the retina once Bruch's membrane has been broken through, it rarely gains access to the vitreous cavity with vitreous seeding.[3,4] Till date, there have been only eight cases reported. Whether vitreous seeding is a poor prognosis remains further investigation and observation of more cases. We report a case of treatment-naïve medium-sized choroid melanoma with rapidly decreased visual acuity due to vitreous seeding during regular follow-up.

Case Report

A 52-year-old male with glaucoma presented with photopsia and black shadow of the right eye for 1 year. His vision was 20/20. Fundus examination showed a dark-pigmented elevated tumor, arose from the choroid, and protruded into the vitreous cavity, over superonasal quadrant [Figure 1]. Fluorescein angiography demonstrated fluorescence blockage by heavy pigmentation of the tumor [Figure 2]. As malignancy was suspected, Gamma Knife radiotherapy or enucleation of eyeball was recommended, but the patient hesitated. The tumor size and vision remained stationary for 4 months until a sudden onset of blurred vision with rapid deterioration in 1 week to 20/250. On follow-up fundus examination, vitreous cavity was hazy with many pigmented materials [Figure 3]. B-scan ultrasonography showed a protruding mass with low internal reflectivity and vitreous opacity [Figure 4]. Subsequent orbital computed tomography (CT) revealed a solid tumor protruding from choroid [Figure 5].

The eyeball was enucleated and a 22 mm bioceramic implant was inserted. Pathology showed a solid tumor measuring 1.2 cm × 0.8 cm × 0.3 cm arises from the

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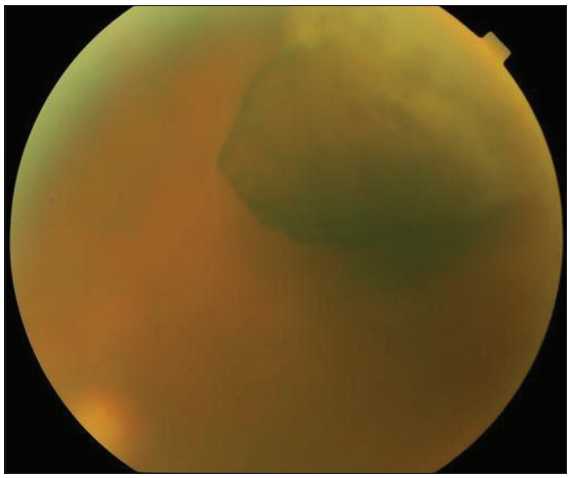

Figure 1: A protruding pigmented tumor at initial presentation

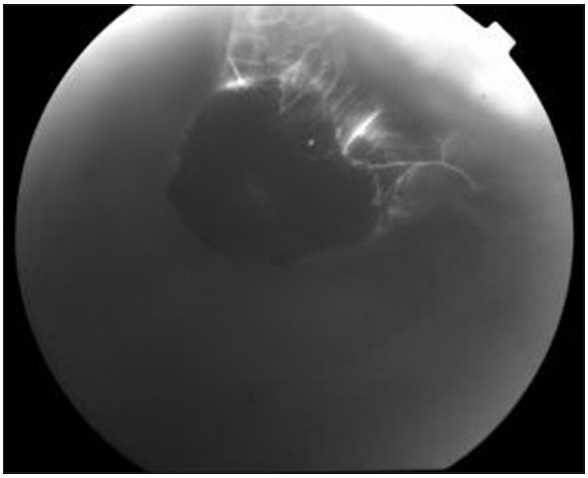

Figure 2: Fluorescein angiography showed hypofluorescence of tumor due to blocked fluorescence by heavy pigmentation

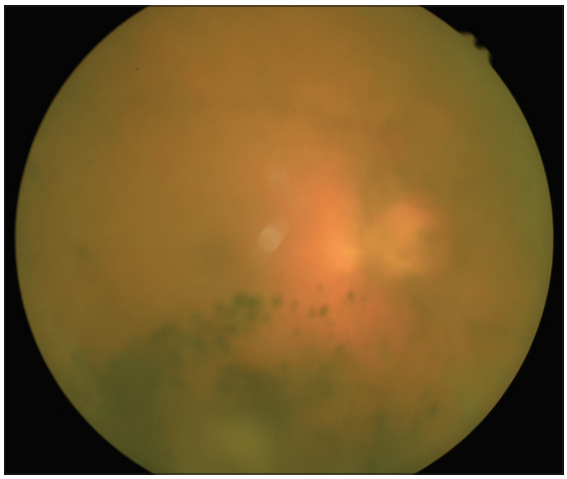

Figure 3: Fundus examination showed vitreous haze with pigmented material in vitreous cavity after rapidly decreased visual acuity

superonasal quadrant of globe. It formed nodulation along the retina with massive involvement of the ciliary body and protruding into the vitreous cavity with intrascleral involvement. Histopathologic findings confirmed a heavy pigmented mixed cell melanoma (pT3bNx) [Figure 6a-c]. Optic nerve was not involved, and there is no extrusion of tumor out of eyeball. Positron emission tomography and abdominal ultrasonography did not show metastasis. The patient is still alive without tumor recurrence or metastasis at the recent follow-up for 31 months (February 2016).

Discussion

Vitreous seeding is rarely seen in choroid melanoma, especially in treatment-naïve choroid melanoma without previous intervention. So far, there were four case reports of vitreous seeding in treatment-naïve choroidal melanoma and one case series of vitreous seeding either primarily or after local intervention.[3-7] Totally, there were only nine reported cases of treatment-naïve choroidal melanoma with vitreous seeding. Of these cases, vitreous seeding with malignant cells was confirmed by vitreous aspiration and cytology study in five cases.[5,7] The other four cases underwent enucleation without cytology proof.[3,4,6]

Vitreous opacity on fundus examination is the first sign to suspect vitreous seeding of choroidal melanoma.[1] B-scan ultrasonography is a method to detect vitreous seeding as in our case. High-resolution spectral-domain optical coherence tomography is another method to suspect vitreous seeding.[3] Orbital CT scan was not sensitive enough to detect the vitreous seeding.

Histopathological results of these nine cases showed three cases of mixed cell type choroidal melanoma, two cases spindle cell type choroidal melanoma, and the rest four did not reveal the pathological types of melanoma.[3-7] Pathological types of treatment-naïve choroid melanoma did not have a relation to vitreous seeding.

All the previous cases with treatment-naïve vitreous cloudiness in choroidal melanoma were later known to have vitreous seeding of melanoma cells on histological study.[3-7] We enucleate the melanoma eyes immediately after vitreous opacity in our case without vitreous aspiration for cytology study. All the previous treatment-naïve cases received enucleation once vitreous seeding was suspected except one case who received transretinal endoresection after adjuvant Gamma Knife radiation.[3-7] Enucleation might be necessary when vitreous seeding in treatment-naïve choroidal melanoma is suspected.[7] Melanoma cells seeding to vitreous might lead to extraocular extension through aqueous channels, resulting in a significant risk for liver metastases and an increased local recurrence rate.[8]

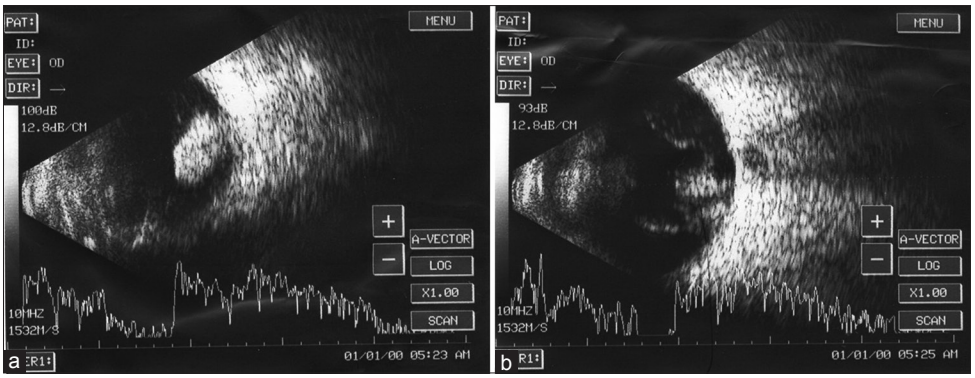

Figure 4: (a) B-scan showed a protruding mass arising from choroid with low internal reflectivity and (b) Vitreous opacity

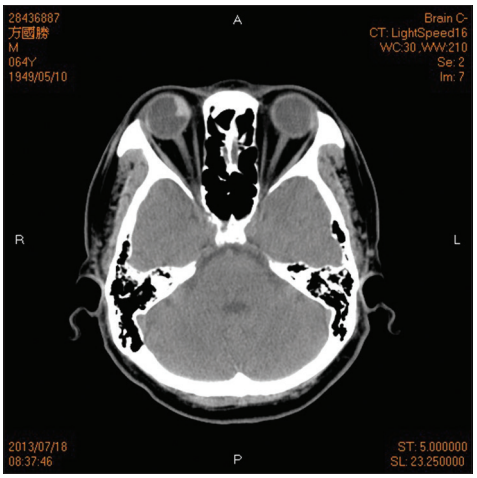

Figure 5: Orbital computed tomography revealed a solid tumor protruding from choroid

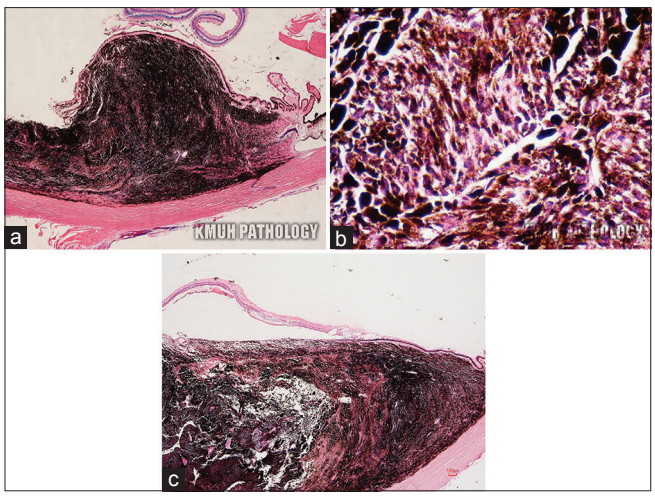

Figure 6: (a) In low-power fields, the tumor arises from the ciliary body and forms nodulation along the retina and protruding into the vitreous cavity (H and E, ×40). (b) In high-power fields, the tumor is composed of epithelioid and spindle neoplastic cells with scant cytoplasm, prominent nucleoli, and abundant melanin pigments (H and E, ×40). (c) There is some focally detached retina that covered on the tumor

Early enucleation when vitreous seeding is suspected in treatment-naïve choroidal melanoma might prolong the survival. In Metz's report, tumor recurrence was free after long-term follow-up.[7] Our case has already survived for 31 months without any recurrence or metastasis.

In the recently published case series, enucleation was performed only in cytology-proved vitreous seeding.[7] They found that vitreous cloudiness may be pigmented melanophage instead of vital tumor cells, and shedding may be the results of tumor necrosis after local treatment.[7] Vitreous cloudiness can occur in choroidal melanoma secondary to brachytherapy and other local treatments. Not all cytological studies of these secondary cases by vitreous aspiration reveal malignant cells.

The location of the tumors is not a key factor for vitreous seeding as these nine cases have different locations. In our case, melanoma is in the ciliary body while location of the rest tumor was juxtapapillary in two cases, the mid-periphery in one case, peripheral of the equator in two other cases, superonasal quadrant in two cases, and no mention of tumor location in one case.[3-7]

One astonishing characteristic of our case is heavy pigmentation of the tumor which totally blocked background fluorescence on fluorescein angiography. However, contribution to vitreous seeding by this special feature remains dubious. Dunn *et al*. had reported a case of amelanotic choroidal melanoma with vitreous seeding.[6]

To the best of our knowledge, this is the ninth reported case of treatment-naïve choroidal melanoma with suspected vitreous seeding. Vitreous cloudiness in treatment-naïve choroidal melanoma all leads to vitreous seeding of melanoma cells. Enucleation is thus unpreventable in such cases. Although vitreous seeding seems to be a poor prognostic factor, our case has already survived for 31 months without any recurrence or metastasis and so were the previous reported cases. Choroidal melanoma with vitreous seeding may not lead to poor prognosis if the patient had early enucleation.

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Conflicts of interest
There are no conflicts of interest.

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