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CASE REPORT

Amelanotic Choroidal Melanoma with Extraocular Extension in a 51 Year-Old Female: A Rare Case

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Abstract

Introduction: Melanoma is a dangerous malignancy primarily involving the choroid, ciliary body, or iris. The most frequent primary intraocular malignant tumor is choroidal melanoma. The tumor most often affects Caucasians of Northern Europe ranging between the ages of 50 and 80. Most choroidal melanomas are pigmented, however, non-pigmented or mixed pigmented and non-pigmented forms can also happen. Compared with other amelanotic choroidal lesions, amelanotic choroidal melanoma showed markedly greater basal diameter, thickness, frequent connection with subretinal fluid, and ultrasonic hollowness. Extrascleral extension is currently detected in 10% to 28% of choroidal melanoma patients, and the mortality rate is much greater than in those without the extension. **Case Presentation:** We reported a rare case of extraocular extension in a 51-year-old female patient with amelanotic melanoma. The primary complaint was a painful and bleeding mass on her left eye that had developed two months prior to admission. On the conjunctiva of the left eye, there was a solid, palpable mass measuring 4 x 3.5 x 1 cm. It was red, well-defined, and had a hard, solid consistency. It was fixed at the base, had minimal bleeding, and pressed on her eyeball from the inferolateral direction. The cornea appears clear, and there is no light perception in visual acuity. The orbital focus computed tomography scan revealed an Enhancing solid mass containing a fat component. The mass was in the left intraocular, extending from intraconal to extraconal. **Conclusions:** Amelanotic melanoma with extraocular extension is a rare condition that can be difficult to detect due to its unclear clinical symptoms and wide range of possible causes. Patients and their families must be educated to receive the appropriate first therapy and prevent the illness from worsening. Melanoma management depends on several factors: tumor size, location, related characteristics, opposite eye status, systemic status, and patient preference. Orbital exenteration is one of the management options for choroidal melanoma with significant extraocular extension. By the time ocular treatment begins, the patient's survival may already be predetermined, and this realization could impact how uveal melanoma is treated in the years to come.

Keywords: choroidal melanoma; amelanotic melanoma; extraocular extension; exenteration

Introduction

Choroidal melanoma is the most frequent adult primary intraocular malignancy. Most patients are adults between 50 and 80 years, with a mean age of 57.[1],[2],[3] It typically has a strong propensity to spread, which raises mortality, and the pace of dissemination is determined by the histological characteristics.[4] The tumor impacted either females (49%) or males (51%).[5] There are three types of choroidal melanoma: pigmented (55%), non-pigmented (15%), and mixed (30%).[6] A study of 4.441 patients with 5.586 amelanotic choroidal tumors revealed that 97% of the patients with amelanotic choroidal melanomas were Caucasians and had a unilateral lesion.[3] A precise diagnosis is essential for an amelanotic choroidal tumor's prognosis and visual outcome, as the differential diagnosis is wide.[3] While some patients may not have any symptoms, choroidal melanoma typically presents as floaters, persistent photopsia, loss of visual field, or vision loss.[7] Patients with choroidal melanoma had impaired visual acuity, visual field

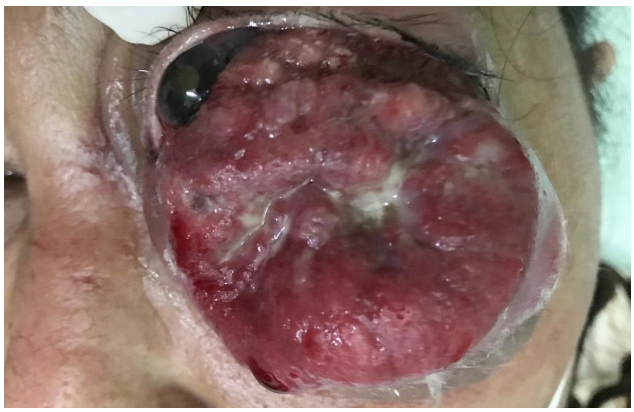

Figure 1. Mass at the left eye showed in clinical appearance.

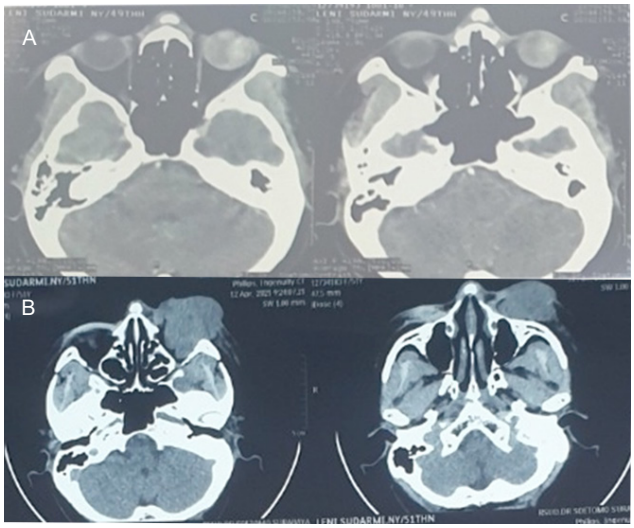

Figure 2. (A) CT scan on 2019 showed strong enhancing solid mass on the left intraocular stick in the sclera lateral side with the left vitreous body hemorrhagic suggestive of intraocular melanoma and (B) CT Scan on 2021 showed an enhancing solid mass measuring 1.3 x 1.2 x 0.74 cm in the left intraocular extends from intraconal to extraconal, and the specifics of the expansion above may indicate a malignant mass.

defect, photopsia, and floaters in 59% of cases, while 41% of patients showed no symptoms.[7]

Extrascleral extension has been found in 10% to 28% of choroidal melanoma patients.[8] The mortality rate among these people is notably more significant than that of patients who do not have the extension. Starr and Zimmerman[2] found that the five-year mortality rate for patients with extrascleral extension was 66%, while the rate for those without extension was 33%. The principal factor determining the orbital expansion of choroidal melanoma was increased intraocular mass, as shown by the growth of the maximum tumor diameter, the rupture of Bruch's membrane, retinal invasion, and vitreous cavity filling.[9] The mixed or epithelioid cell type is more likely to spread throughout the orbital tissues than spindle cell malignancies.[9]

There is still debate over the best course of action for exenteration and additional orbital irradiation.[10] Treatment options for uveal melanoma with macroscopic extrascleral extension often involve ordinary enucleation or orbital exenteration. The prognosis for uveal melanoma depends on extraocular extension, tumor location, size, and configuration, among other factors.[5]

In this case study, a 51-year-old female Southeast Asian who was previously healthy is described as having an amelanotic choroidal melanoma with extraocular extension as a result of a lost cause. This case report aims to review clinical presentation, examination, and treatment options in a patient with extraocular extension of amelanotic choroidal melanoma and attempt to ascertain the correlation between tumor features.

Case presentation

A 51-year-old female presented with a mass on her left eye two months before admission. The patient said the mass had grown more quickly after bursting two months ago; however, it had previously been as little as a peanut. Following that, the mass grew larger and became worse, releasing pus and blood. There was redness on the eyelid and pain in the left eye. There had previously been visual problems. She did not have any past medical history of trauma, swelling in other areas, or any systemic abnormalities. There was no other family member who had disclosed a history of tumors or ocular malignancies. Two years prior, the patient complained of blurry vision in the left eye, so they visited the retinal division. No complaints of pain or edema were made, however, there was redness on the transparent membrane of the left eye.

The general assessment resulted in a normal condition and visual analogue scale (VAS) score of 3–4. The human immunodeficiency virus (HIV) testing is performed on patients, and the results are negative. During the ophthalmology examination, a firm, palpable mass measuring 4 x 3.5 x1 cm and reddish was found on the conjunctiva of her left eye. The mass was well-defined, hard, solid, and fixed at the base. There was minimal bleeding, minimal pus, no necrotic tissue, no

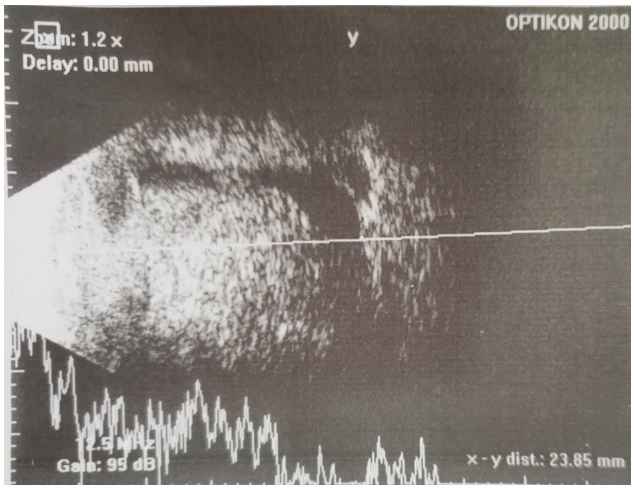

Figure 3. B-scan ultrasonography revealed vitreous echogenic lesion like membrane with echospike between 70-80 retinal choroid scleral complex indicated an intraocular tumor.

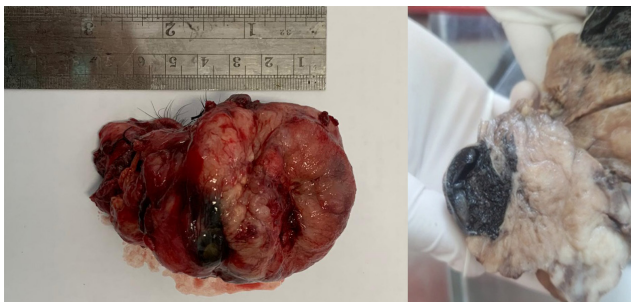

Figure 4. (A) Gross macroscopic mass after exenterated; (B) A pigmented mass fills the intraocular space as seen in the gross macroscopic image.

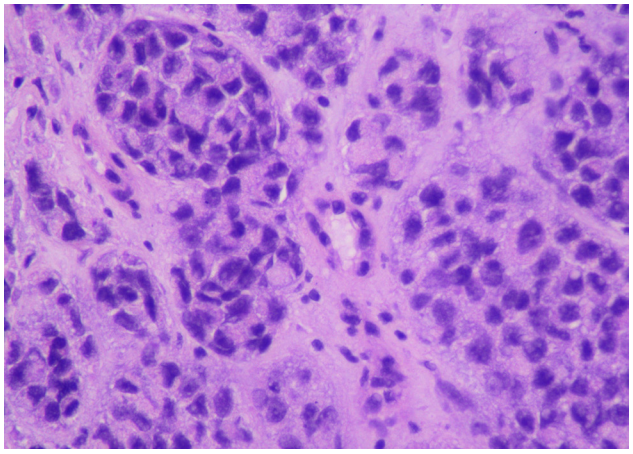

Figure 5. Magnification 40x showing a tissue slice with tumor growth organized in a nest pattern, containing a proliferation of round-oval nucleated cells that are pleomorphic, hyperchromatic, and adequately cytoplasmic; the tumor invades the connective tissue as it grows (haematoxylin and eosin; magnification 40x).

crusting, and the mass was pressing on her eyeball from the inferolateral direction. The cornea appears clear, whereas the other anterior segment is more challenging to assess. Evaluation of the left eye's movement proved challenging. In the right eye, nothing unusual was discovered (Figure 1).

Ancillary test performed two years ago included B scan ultrasonography, which revealed a vitreous echogenic lesion-like membrane with echospike between 70-80 retinal choroid scleral complex indicated an intraocular tumor (Figure 3). A 2019 computed tomography (CT) scan showed no tumor, infarction, hemorrhage, or infectious process in the brain parenchyma. Instead, the scan showed a strong enhancing solid mass on the left intraocular stick in the sclera lateral side with the left vitreous body hemorrhagic, suggestive of intraocular melanoma. A CT scan with an orbital focus in 2021 revealed an enhanced solid mass that included fat. There was left mastoiditis and a mass measuring 1.3 x 1.2 x 0.74 cm in the left intraocular. The mass extends from intraconal to extraconal, and the specifics of the expansion above may indicate a malignant mass. There was no bone degeneration and the right eye showed no abnormalities (Figure 2). An ultrasound examination of the upper and lower abdomen revealed a normal liver with a sharp angle, a flat edge, and a homogeneous parenchyma. There was no enlargement of the para-aortic lymph nodes or a metastatic process in the liver. No anomalies are present in the liver, Gall Bladder, spleen, pancreas, right left kidney, bladder, uterus, or right left adnexa.

In 2019, the patient was first identified with a left ocular intraocular tumor, with melanoma being considered a differential diagnosis. She was supposed to have her left eye enucleated under general anesthesia, however, unfortunately, she did not return for around a year. Because she had seen another therapist and was being mistreated by her previous one, the patient returned two years later in a worse condition. She was subsequently diagnosed with orbital inflammation and squamous cell carcinoma conjunctiva in her left eye. Analgesics and oral antibiotics were prescribed in her treatment.

Based on intraoperative findings, a solid ridge bleeds easily in the inferior orbital area, superior lateral to the apex, with 6 x 4 x 3 cm in size (Figure 3). Erosion of the orbital roof and orbital median wall destruction was discovered intraoperative. Following surgery, the patient was given an oral analgesic, an antibiotic, and a bandage for five days.

The histology report showed that it was malignant melanoma. An intraocular paraffin coupe showed amelanotic melanoma, malignant tumor, and carcinoma with poor differentiation. An abdominal ultrasound did not show any abnormalities. The patient visited the oncology division two weeks following surgery with no eye symptoms. The left eye socket is exenterated, the suture is in good condition, and there is no pus or bleeding. It is planned for the patient to get radiation therapy or chemotherapy (Figure 4-5).

Discussion and conclusions

Amelanotic choroidal melanoma is a relatively rare type with an incidence of about 15%.[3] Melanoma, nevus, metastasis, hemangioma, peripheral exudative hemorrhagic chorioretinopathy, scleral calcification, osteoma, lymphoma, isolated idiopathic choroiditis, and choroidal effusion are just a few of the manifestations that amelanotic choroidal tumors can take.[3],[11] Compared to the pigmented (55%) and mixed pigmented or non-pigmented (30%) cases, this occurs less frequently.[6] The multiplication of melanocytes that lose their capacity to produce the pigment melanin is the root cause of non-pigmented choroidal melanoma.[9] When the patient first arrived, their complaint was blurred vision in the left eye. Among the common symptoms of melanoma patients include persistent photopsia (flash of light), loss of vision, and floaters. Not all individuals have symptoms, however, some have silent tumors that were accidentally found during routine eye exams.[5],[6] Choroidal melanomas have

elevated tumors that resemble domes or mushrooms. Most choroidal melanomas have a thickness of more than two millimeters, while smaller melanomas should not be confused with choroidal nevi.[12]

This patient had a large extraocular mass on arrival for the second time, which probably confused the diagnosis. The patient presented with an extraocular tumor that appeared to be quite similar to conjunctival squamous cell carcinoma (SCC). From the history taking and ancillary test, the condition does not support the etiology and risk factors for conjunctival SCC, such as age, history of human papillomavirus (HPV) infection, exposure to ultraviolet B (UVB) radiation, HIV/AIDS, and other types of immunodeficiency like medical immunosuppression, so the diagnosis of conjunctival SCC can be ruled out.[13] Amelanotic melanoma was discovered on histological examination of the removal eyeball, confirming the intraocular melanoma that had been detected by the patient's first CT scan upon admission.

It has been observed that 10% to 28% of patients with choroidal melanoma experience extraocular extension.[12] Only 3-5.8% of patients present with uveal melanoma with extraocular extension at the time of diagnosis.[8] The mass size is unknown based on the preliminary CT scan results, although the extraocular extension is more common in big or medium-sized tumors and located in the juxta papillary area, the ciliary body, or the vortex veins.[8] This patient was a missing case, meaning that extraocular extension would not have happened with the first therapy. The two most commonly used therapies for choroidal melanoma that do not include extraocular extension are enucleation or localized radiotherapy using either plaque radiotherapy or proton beam radiotherapy. An eye with a sizable extraocular extension field is used for orbital exenteration.[14] There may be pus and blood when the patients arrive, exacerbating their symptoms. Because people in rural areas still have faith in non-medical persons and alternative remedies, the usage of these drugs raises questions about whether the infection is being treated incorrectly. By providing appropriate wound care, the appropriate medical personnel can reduce the incidence of these infections.

Various options for management are talked about. Transpupillary thermotherapy, charged particle irradiation, plaque radiotherapy, local resection, enucleation, or orbital exenteration are among the management options for choroidal melanoma.[5] Numerous publications have questioned the efficacy of exenteration as a treatment for ocular melanomas. The exenteration in this patient was carried out following the guidelines provided by Rendahl and Henderson.[2] These guidelines state that regardless of the size or encapsulation of the extension, orbital tissue exenteration should be carried out as soon as the diagnosis of orbital extension is confirmed histopathologically.[10] In addition, consideration for exenteration is associated with an infection that is already severe enough to make people interested.

Liver (89%), lung (29%), bone (17%), skin and subcutaneous tissue (12%), and lymph nodes (11%) were the most frequently metastasized sites.[13] Nonetheless, an abdominal ultrasound scan ruled out the potential of metastases in this patient. The prognosis for choroidal melanoma without metastases and extraocular extension appears favorable. Although extraocular extension may not significantly worsen the prognosis for life on its own, it may be a symptom of a more malignant tumor. The highest percentages of survival are seen in early exenteration.[2]

Patients should have routine systemic evaluations following surgical treatment for uveal melanoma. Past medical history should be interpreted carefully since it could indicate a non-ocular malignancy, suggesting a metastatic lesion. The ocular oncologist needs to keep an eye out for side effects from treatment and tumor regression in the uveal scar. Radiation retinopathy, cataracts, papillopathy, scleral necrosis, glaucoma, and discomfort are among the side effects of irradiation. Since these cancers frequently spread to the liver, lungs, and skin, a particular examination of these areas needs to be done. It is advised that a physical examination, liver function tests, liver magnetic resonance imaging, and chest radiography be carried out twice a year for monitoring purposes.[14]

The case report's conclusion discusses amelanotic melanoma with extraocular extension, which is a rare condition that can be difficult to detect due to its unclear clinical symptoms and wide range of possible causes. Patients and their families must be educated to receive the appropriate first therapy and prevent the illness from worsening. Melanoma management depends on several factors: tumor size, location, related characteristics, opposite eye status, systemic status, and patient preference. Orbital exenteration is one of the management options for choroidal melanoma with significant extraocular extension. By the time ocular treatment begins, the patient's survival may already be predetermined, and this realization could impact how uveal melanoma is treated in the years to come.

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