An Unusual Presentation of Uveal Melanoma; Bilateral Corneal Perforation

Sıradışı Bir Uveal Melanoma Prezentasyonu, Bilateral Korneal Perforasyon

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ABSTRACT

We present here a case of bilateral choroidal melanoma protruding from perforated central corneal zones. A86-year old female presented to our emergency department twice with masses protruding from her eyes throughout the corneal zones. These lesions were massively pigmented with punctate hemorrhages. Computed tomography scans showed that lesions filled the globes completely,extending anteriorly, destructing cornal tissues and protruding out. Abdominal CT revealed liver metastasis. She did not accept any treatment. The most common primary malignant intraocular tumor in adult is uveal melanoma. Choroidal melanomas remain asymptomatic for prolonged periods of time; on a routine outpatient examination tumors are detected incidentally. If the patient is unaware of vision loss or neglects this, choroidal melanomas may grow and show exaggerated presentation. Early detection is critical to prevent extraocular extension of tumor. Choroidal melanoma causes death secondary to distant metastases.

Key Words: Choroidal melanoma, corneal perforation.

ÖZ

Bu vakamızda ileri seviyede bir koroidal melanomun her iki kornayı perfore ederek göz dışına protrüzyonunu sunduk. Seksen altı yaşında bayan hasta gözlerinden dışarıya doğru uzanan ve kanayan kitleler nedeniyle 2 sefer acil servisimize başvurdu. Koyu pigmentli ve hemorajik bu kitleler bilgisayarlı tomografi ile incelendiğinde globu tamamen doldurdukları, korneal dokuyu destrükte ederek dışarıya doğru uzandıkları görüldü. Abdominal BT karaciğer metastazı gösterdi. Hasta herhangi bir tedavi seçeneğini kabul etmedi. Yetişkinlerde en sık görülen primer malign intraoküler tümör uveal melanoma'dır. Uzun süre asemptomatik kalabilen koroidal melanomlar çoğu zaman rutin oftalmolojik muayenede rastlantısal olarak teşhis edilirler. Eğer hasta en sık ilk semptom olan görme azlığını inkar eder, farkında olmaz ve ya ihmal ederse koroidal melanomalar bazen ileri seviyelere ulaşmış haldeyken başvurabilirler. Ekstraoküler yayılımı önleme açısından erken teşhis önemlidir. Ölüm sebebi genellikle uzak metastazdır.

Anahtar Kelimeler: Koroidal melanoma, korneal perforasyon.

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INTRODUCTION

Uveal melanoma is the most common primary malignant intraocular tumor in adults. Clinical presentation depends on the location and extension of tumor. Usually on a routine outpatient examination tumors are detected incidentally. Painless visual loss as a result of retinal detachment or mechanical obstruction of visual axis by growing mass is the leading symptom. Choroidal melanomas may show an infrequent presantation like a mass protrusion from perforated eye depending on patient mental status,awareness of vision or negligence such as in our case.

CASE REPORT

A 86-year old female presented to our emergency department with a mass protruding from her right eye and bleeding. She had severe ocular pain. At presentation ophthalmologic examination revealed that there was no light perception in right eye neither in left. Slit lamp ophthalmoscopic evaluation showed a dark brown-black mass with punctate hemorrhages on it protruding throughout the perforated corneal zone in right side (Figure 1). No further detail was observed. Central corneal opacity,mature cataract were biomicroscopic findings on left eye without any symptom. Fundus examination can not be performed because of lens opacity. B-scan ultrasonography showed intraocular mass in right side and normal findings in left side. Computed tomography scan imaging of orbits showed a mass lesion that completely filled the bulbus oculi and protroding from perforated corneal zone in her right eye and normal findings in her left eye (Figure 2).

The patient underwent systemic evaluation due to the possibility of distant metastasis. Liver enzyme levels were above the normal limits and abdominal computed tomography revealed liver metastasis. Patient and her family rejected further investigation and thus histopathological evaluation could not been done. The patient was referred to a multidisciplinary clinic including chemotherapy unit. Then 16 days later she presented to our ophthalmology department with the same complaint but now on left side. Massive pigmented brown tumor protruding throughout the perforated corneal zone was observed (Figure 3). There was irregular ocular surface with no detectable anterior segment detail on right side. Computed tomography scan imaging showed globe deformity with a posteriorly loacated hyperdense lesion, most propably the residual mass after spontaneous resolution of intraocular hemorrhage, in right side and mass lesion that completely filled the bulbus oculi protruding from perforated corneal zone in her left eye (Figure 4). Axial T2 fat supressed MRI scan showed that mass lesion filled the left bulbus oculi and protruding from central corneal zone. Considering all these findings surgical intervention or chemotherapy were suggested as possible treatment options to the patient. She refused it. The patient was referred to the oncologist and was lost from follow up.

DISCUSSION

The most common primary malignant intraocular tumor in adult is uveal melanoma. Uveal melanomas can be divided into two categories. Anterior uveal melanomas in which the tumor arises in iris and posterior uveal melanomas in which tumor arises in ciliary body or choroid. Data from collaborative Ocular Melanoma Study (COMS) indicates that uveal melanoma affects older people and both sexes equally.[1] Race is one of the most important predisposing factor. Other risk factors to develop uveal malignant melanoma are congenital ocular or oculodermal melanocytosis,exposure to ultraviolet light.[2,3]

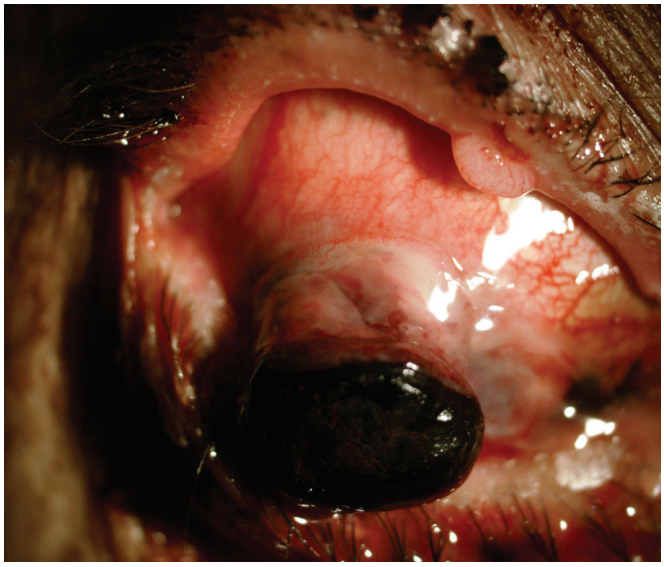

Figure 1: *Dark brown-black mass with punctate hemorrhages protruding throughout the perforated corneal zone in right side.*

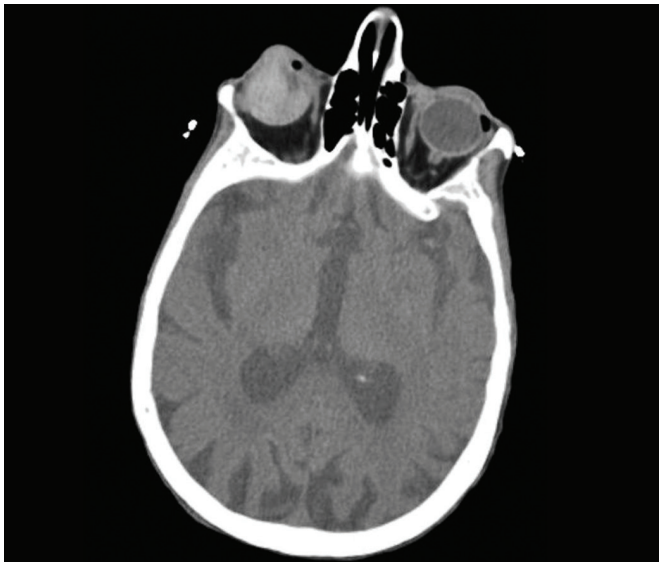

Figure 2: *Computed tomography scan showed a mass lesion that completely filled the bulbus oculi and protroding from perforated corneal in right eye.*

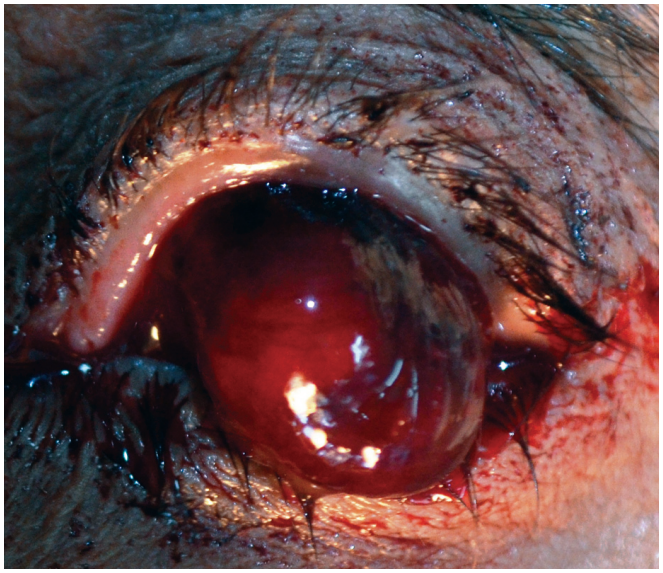

Figure 3: *Massive pigmented brown tumor protruding throughout the perforated corneal zone in left side.*

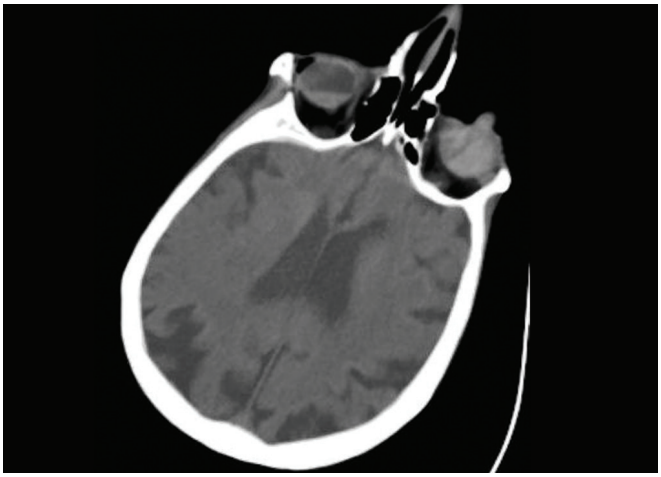

Figure 4: *Computed tomography scan showed globe deformity with a posteriorly loacated hyperdense lesion most propably the residual mass after spontaneous resolution of intraocular hemorrhage in right side and mass lesion that completely filled the bulbus oculi protruding from perforated corneal zone in left eye.*

The location of tumor and extent of the disease determine the clinical presentation. Choroidal melanomas remain asymptomatic for prolonged periods of time; on a routine outpatient examination tumors are detected incidentally, but mostly painless visual loss as a result of retinal detachment or mechanical obstruction of visual axis by growing mass is the leading symptom. Other symptoms are paracentral scotoma, floaters and sometimes severe ocular pain. A dome-shaped or sessile, pigmented (or sometimes totally amelanotic) mass on fundoscopic examination is the typical first finding. When tumor breaks through Bruch's membrane, mushroom or collar button shape is the configuration. Serous retinal detachment, secondary glaucoma, sentinel vessels, cataract may exist depending on size and location of tumor. Ultrasonography, computed tomography, magnetic resonance imaging, fluorescein angiography, fine needle aspiration biopsy are the common ancillary studies used to support the diagnosis especially in eyes with opaque media ,otherwise the diagnosis can be made by fundoscopic examination. According to the report published by COMS, the clinical diagnosis of choroidal melanomas has an accuracy of more than 99%.[4]

Rarely patients with unrecognized choroidal melanoma present with extraocular extension of their tumors.[5] These ways are aqueous channels, ciliary arteries, vortex veins,ciliary nerves,optic nerve and a variety of rare combinations of these routes.[6] Depending on patient mental status,awareness of vision or negligence, choroidal melanomas grow and may show an infrequent presantation like a mass protrusion from perforated eye such as in our case. Because of no known previos history, follow up and no medical record, we could not be able to sure about the progress of the disease. According to history our patient has been blind for nearly 4 years and nothing else. She had been living in a rural area in eastern Turkey and had not admitted to hospital before. This neglected condition shows what an uncontrolled choroidal malignant melanoma results in clinically.

Choroidal melanoma causes death secondary to distant metastases rather than local spread. The most common site of metastasis is the liver (95%), followed by lung (24%), bones (16%), and skin (11%).[7] Once patients develop metastases, the prognosis is generally poor and treatment of the metastatic disease does not seem to improve the overall survival time.[8,9] In cases which distant metastases are found during systemic examination, treatment of intraocular tumour becomes palliative. Systemic chemotherapy is the primary treatment in such cases.[10]

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