

CONJUNCTIVAL MELANOMA – A CASE REPORT

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SUMMARY – Conjunctival melanoma is a relatively rare malignancy. It is usually a pigmented, unilateral lesion, often in the perilimbal interpalpebral bulbar conjunctiva, mostly arising from primary acquired melanosis. An 81-year-old male, otherwise healthy, presented with a 2-year history of a slowly growing protruding massive pigmented tumor of the right eye. Biomicroscopy showed a massive pigmented brown tumor protruding from 3 mm of the temporal sclera and 4/5 of the temporal cornea causing mechanical ectropion with keratinized conjunctiva of the lower eyelid. The extent of the tumor was 10 o'clock with only the limbus from 2 to 4 o'clock being tumor-free. The surface of the tumor of 10 mm in size at the base and 7 mm in height was rough, necrotic, with rich feeder vessels and spotting bleedings. Cytology of the tumor surface revealed only inflammatory cells. Melanoma cell staining was negative for both HMB 45 and BerEp4(EA). Since the tumor surface was highly necrotic, a fragment was obtained and referred for histopathology. The diagnosis was: melanoma. Enucleation with no primary orbital implant was performed as a less invasive palliative surgery. An additional 4-mm fragment of the healthy-looking marginal conjunctiva was also excised. Histopathology of the tumor indicated conjunctival melanoma.

Key words: *Melanoma; Conjunctival neoplasms; Melanosis; Ophthalmology*

Introduction

Conjunctival melanoma is a rare clinical entity (the incidence is 0.2-0.8 *per* million in Caucasian population)^{1,2}, with potentially life-threatening implications (10-year mortality from 13% to 30%)^{1,3}. It is a relatively rare condition that occurs 500 times less frequently than cutaneous melanoma and 40 times less frequently than choroidal melanoma¹. As many as 75% of conjunctival melanoma develop from primary acquired melanosis with atypia and almost 50% of patients with primary acquired melanosis with atypia will develop invasive melanoma¹. It is usually a pigmented, unilateral conjunctival lesion, often in the perilimbal interpalpebral bulbar conjunctiva. Conjunctival melanoma is diagnosed by the presence of atypical tumor cells invading the conjunctival substantia propria.

The aim of our paper is to present an unusually large conjunctival melanoma protruding from the eye that was

diagnosed rather late being wrongly considered as a neglected spontaneous corneal perforation.

Case Report

The 81-year-old male, otherwise healthy, presented to our Department with a 2-year history of the slowly growing protruding massive pigmented tumor of the right eye (Fig. 1). At the age of 16, he had sustained a right eye injury inflicted with a large piece of wood, without documented follow up.

At presentation complete ophthalmologic examination revealed the right eye BCVA counting fingers nasally and the left eye 0.7 on Snellen charts. Biomicroscopy showed a massive pigmented brown tumor protruding from 3 mm of the temporal sclera and 4/5 of the temporal cornea causing mechanical ectropion with keratinized conjunctiva of the right eye lower eyelid (Figs. 1 and 2). No data on prior primary acquired melanosis or nevus were available. The location of the tumor epicenter was bulbar. The extent of conjunctival melanoma was 10 o'clock with only the limbus from 2 to 4 o'clock

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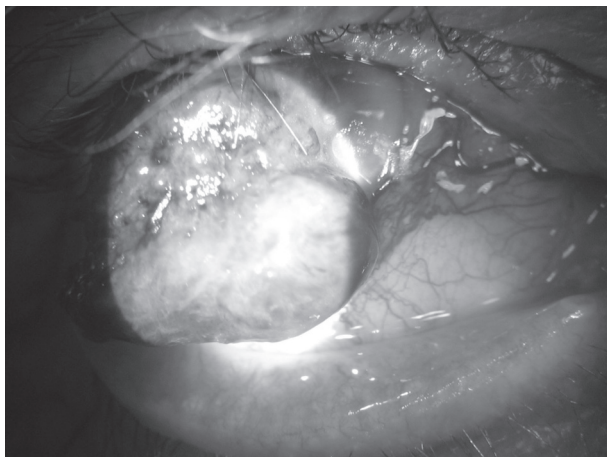


Fig. 1. Massive protruding lesion of the right eye.

being tumor-free. The tumor set on the limbus. The surface of the tumor of 10 mm in size at the base and 7 mm in height was rough, necrotic, with rich feeder vessels and spotting hemorrhages. Episcleral blood vessels were wide and tortuous in the temporal and nasal part of the eye globe. The anterior chamber of normal depth and clarity as well as a part of the pupil and corticonuclear opacities in the lens and red reflex could only be seen through the nasal 1/4 of the cornea. There was no proptosis, with full motor range of the eye. Regional lymph nodes were not palpable. Left eye biomicroscopy revealed corticonuclear lens opacities with no further pathology. Digitally examined right eye pressure was normal. Left eye pressure was 15.6 mm Hg. B-scan axial immersion ultrasonography of the right eye showed a massive mushroom-like epibulbar lesion of 7 mm in size at the base and 7 mm in height, internal echogenicity of 30%-50%. The anterior chamber was visualized in the nasal part. More detailed anatomic relationships toward the iris and pars plana of the ciliary body could not be imaged. Transverse and longitudinal sections of the ciliary body were without pathology. Retinal, choroidal and orbital echograms were normal. Microbiological examination of the right eye conjunctiva showed $\hat{\alpha}$ -lactamasis negative *Staphylococcus epidermidis*, so the patient was treated with local antibiotics. Computed tomography scan imaging of the brain and orbits showed bulging of the right eye globe

with sharp homogeneous contours sized about 10 mm in craniocaudal direction, 8 mm in laterolateral dimension and 5-6 mm in anteroposterior direction. No significant pathology of the vitreous body, orbits and brain were found. Magnetic resonance imaging of the brain and orbits showed expansive tumor in the central part of the right eye cornea, sized approximately 11 mm in laterolateral direction and 5-6 mm in anteroposterior direction, with a higher signal on T1 images. Cytology of the tumor surface revealed only inflammatory cells. Melanoma cell staining was negative for both HMB 45 and BerEp4(EA). Since the tumor surface was highly necrotic, a fragment was obtained and referred for histopathology. The diagnosis was: melanoma.

Having this diagnosis in mind, tumor staging was performed. All standard blood tests were within the normal limits except for total bilirubin that was slightly increased (23.8 μ mol/L). X-ray of the heart and lungs showed no significant pathology. Electrocardiography

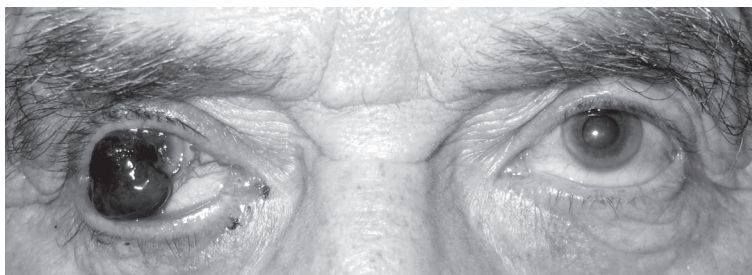


Fig. 2. Biomicroscopic appearance of the right eye.

(ECG) showed arterial fibrillation with heart rate of 80/minute. Liver ultrasonography showed two hyperechoic lesions sized 6 mm and 7 mm in the right part of the

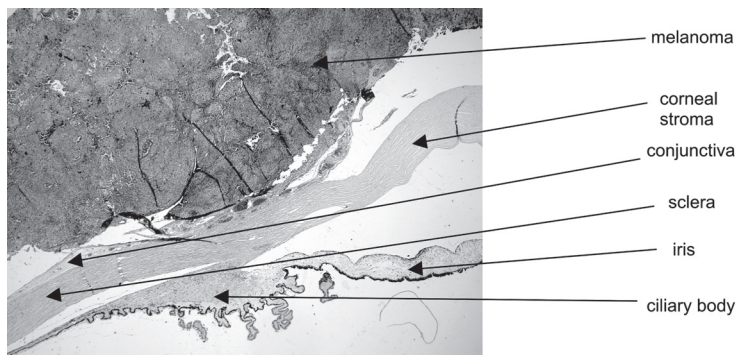


Fig. 3. Histopathologic specimen showing massive melanoma with shallow infiltration of the underlying corneal stroma.

liver, close to the hepatic veins, and additional two micro subdiaphragm lesions close to the Glisson's capsule were interpreted as metastases from the melanoma. The spleen, bile sac, lymph nodes and other visible parts of the abdominal cavity were found clear from ultrasonographically visible lesions. Whole body ^{99m}Tc MBq 740 scintigraphy showed no specific findings for metastases.

Considering all these findings, exenteration of the right orbit was suggested to the patient. Being aware of his condition, he refused it. Enucleation with no primary orbital implant was performed as a less invasive palliative surgery. Additional 4 mm of healthy-looking marginal conjunctiva was also excised. Histopathology of the tumor revealed conjunctival melanoma (Fig. 3). Infiltration of the corneal stroma and the intact optical nerve were described. The patient was referred to the oncologist and unfortunately was lost from follow up.

Discussion

Differential diagnosis of conjunctival melanoma relies on presentation of a pigmented tumor, usually in the interpalpebral conjunctiva. Our patient was referred to us after having been followed-up for one year as a healed spontaneous corneal perforation due to an old injury. Only when the lesion started to grow rapidly within a month he was urgently referred to our hospital. The long period of misdiagnosis resulted in a massive pigmented lesion protruding from the eye. Excision biopsy is the only way to distinguish melanoma from a benign lesion¹. In our case it was not an option due to the tumor size. Cytology of the tumor surface as well as melanoma cells staining for HMB 45⁵ and BerEp4(EA)⁶ were unhelpful in this case. Incisional biopsy is not recommended³. Massive necrosis of the tumor surface enabled us to collect a tumor fragment for histopathology that was conclusive for melanoma. Previous histologically confirmed melanoma should be excised, preferably with 3-5 mm margins¹. Cryotherapy applied to the excisional biopsy margins and tumor bed helps eliminate residual neoplastic cells¹. Adjuvant radiotherapy or topical Mitomycin C^{1-3,7} can be administered. Due to the tumor spread, exenteration was an option but our patient refused it consenting for enucleation. In the literature, exenteration has shown no statistically significant change in outcome compared with more conservative measures^{1,3}. Intraoperative 4-mm excision of healthy-looking surrounding conjunctiva after enucleation was

performed in an attempt to reduce the possibility of residual tumor cells. No cryotherapy or Mitomycin C was applied perioperatively. According to the report by Shields *et al.*³, when the cornea was involved with melanoma, the depth of invasion was into the corneal epithelium in all but one case in which stromal involvement was detected. In our patient, histopathology of the tumor documented stromal infiltration of the cornea. Conjunctival melanoma does not metastasize hematogenously, rather disseminating through regional preauricular and submandibular lymph nodes that should be palpated during examination^{1,2}. No lymph nodes of the head and neck were palpable in our patient. Staging performed when the diagnosis was made revealed multiple liver lesions that were interpreted as metastases. Our patient was referred to the oncologist for additional examination and treatment. In their case study, Shields *et al.*³ report on a 35% recurrence rate of conjunctival melanoma. In high-risk lesions, follow up should be scheduled every 3 to 4 months with monitoring for early disease recurrence. Unfortunately, our patient was lost from follow up.

Conclusion

Although being a rare entity, conjunctival melanoma should be considered on the differential diagnosis of pigmented eye surface lesions, especially when growing on pre-existing tumors such as primary acquired melanosis or nevus. Early detection of conjunctival melanoma prevents life-threatening implications that have been documented in a relatively high percentage of cases.

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Sažetak

KONJUNKTIVNI MELANOM – PRIKAZ SLUČAJA

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Konjunktivni melanom je relativno rijetka maligna bolest. Obično se javlja kao jednostrana, pigmentirana lezija koja zahvaća intrepalpebralnu bulbarnu spojnicu perilimbalno, a najčešće se razvija iz primarne stečene melanoze. Muškarac star 81 godinu, inače zdrav, došao je na pregled zbog protrudirajućeg tumora desnoga oka koji je polagano rastao unazad dvije godine. Biomikroskopski pregled otkrio je masivan, smeđe pigmentirani tumor koji se izdizao iznad temporalne 4/5 rožnice i susjednih 3 mm bjeloočnice temporalno od limbusa uzrokujući mehanički ektropij s keratiniziranom sponicom donje vjeđe. Samo limbus od 2 do 4 sata nije bio prekriven tumorom. Površina tumora širine 10 mm u bazi i visine 7 mm bila je nepravilna, nekrotična, s bogatom mrežom krvnih žila i točkastim krvarenjima. Citološkim brisom površine tumora nađene su samo upalne stanice. Bojanja na melanomske antigene HMB 45 i BerEp4(EA) bila su negativna. Zahvaljujući nekrotičnoj površini tumora bilo je moguće uzeti komadić tumora i poslati ga na histopatološku analizu kojom je postavljena dijagnoza melanoma. Kao najmanje invazivna palijativna operacija učinjena je enukleacija bez primarnog orbitalnog implanta. Također je cirkularno izrezano 4 mm susjedne vizualno nepromijenjene spojnice. Histopatološkom analizom tumora postavljena je konačna dijagnoza konjunktivnog melanoma.

Ključne riječi: *Melanom; Konjunktivne neoplazme; Melanoza; Oftalmologija*