

Synchronous conjunctival melanoma and lentigo maligna melanoma*

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Abstract: Lentigo maligna has an extensive and neoplastic character. It typically progresses slowly and may eventually develop into an invasive melanoma, which is called lentigo maligna melanoma. Ocular melanoma is the second most common type of melanoma. The uvea is the most common site of origin of ocular melanomas, while conjunctival melanoma accounts for about 1-5% of cases. In this article, we describe a rare case of synchronic conjunctival melanoma and lentigo maligna on the face.

Keywords: Melanoma; Nevi and melanomas; Hutchinson's melanotic freckle

INTRODUCTION

In 1892, Hutchinson described a form of melanoma *in situ* that affected sun-exposed areas in the elderly and named it "senile freckles". Later, Dubreuilh coined the term lentigo maligna (LM), and demonstrated the extensive and neoplastic nature of the lesion. It typically progresses slowly and may eventually develop into an invasive melanoma, which is called lentigo maligna melanoma.¹ The peak incidence is between 65-80 years. Risk factors include fair-skinned individuals with intense photodamaged skin.¹

Lentigo maligna clinically presents itself as a smooth pigmented macule of variable size and color.¹ Dermoscopy plays a key role in the diagnosis of LM, revealing characteristics such as gray dots, gray globules, dark rhomboidal structures, and asymmetric pigmented follicular openings.²

Ocular melanoma is the second most common type of melanoma. The uvea is the most common site of origin of ocular melanomas, comprising more than 80% of cases, while conjunctival melanoma accounts for about 1-5% of cases. Although most ocular melanomas are primary, cutaneous melanoma metastases may also occur in the ocular region, which represent less than 5% of all metastases to the eye and orbits.³ Conjunctival melanomas originate from melanocytes located in the basal layers of the conjunctival epithelium. Unlike other mucous membranes of the bulbar conjunctiva is directly exposed to solar radiation.

According to Jovanovic *et al.*, conjunctiva melanoma occurs almost exclusively in white people, and only less than 1% are African-American patients. The incidence of this type of melanoma increases with age and it appears more frequently in the fifth and sixth decades of life.³⁻⁵

Conjunctival melanoma typically presents as a raised pigmented lesion, although sometimes it may be amelanotic, surrounded by prominent blood vessels or areas of primary acquired melanosis (PAM). It can appear in any conjunctival region including the bulbar conjunctiva, the eyelid, the fornices, semilunar fold, and caruncles. In most cases, the lesion develops at the limbus. Complaints about the sudden appearance of a local pigmented lesion is common, while irritation and pain are rare events.³⁻⁵

The mainstay treatment of conjunctival melanoma is tumor resection with wide surgical margin. In most cases, adjunct therapy is associated with cryotherapy. Some authors recommend the use of additional brachytherapy. Orbital exenteration is reserved for cases in very advanced stages. Local tumor recurrence has been reported in 60% of patients. Metastases occur most commonly to the preauricular and submandibular lymph nodes. Mortality rate in 10 years is between 23% and 30%.^{4,6}

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CASE REPORT

We report an 85-year-old male patient complaining of a pigmented lesion in the right malar region and an ulcerated lesion of about 5 cm on the back. The lesions had been present for 2 years. Dermoscopy of the malar lesion showed multiple shades ranging from brown to black and dark rhomboidal structures with asymmetric perifollicular pigmentation (Figure 1).

Physical examination revealed a brownish and unilateral pigment in the conjunctival lesion. The patient reported to be unaware of the existence of the lesion. Ophthalmologic evaluation showed an ocular melanoma. Therefore, we scheduled a biopsy of the lesion (Figures 2 and 3).

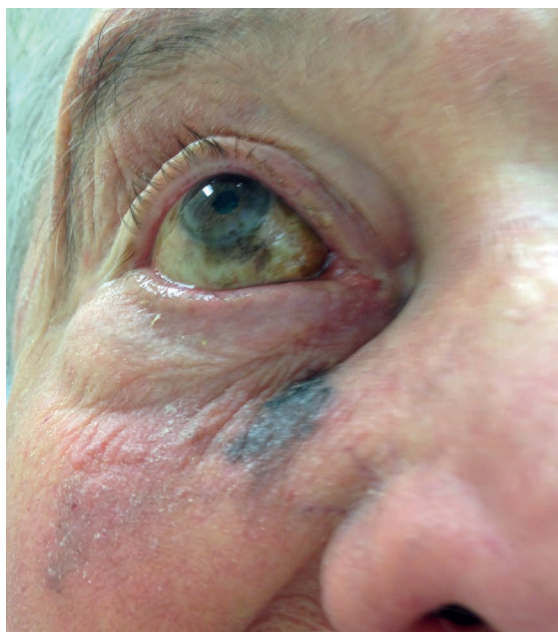


FIGURE 1: Conjunctival melanoma and lentigo maligna melanoma



FIGURE 2: Dermoscopy of the ocular lesion

The patient underwent excision of both lesions with the following histopathological diagnoses: lesion in the right malar region – lentigo maligna melanoma, Breslow 0.7mm, Clark III, mitotic index = 0/mm², absence of ulceration, perineural invasion, undetected angiolymphatic invasion, discrete perineural infiltration of lymphocytes, absence of regression, no satellitosis, and free margins; lesion on the back – nodular basal cell carcinoma, ulceration, and undetected angiolymphatic invasion (Figure 4).

After a biopsy of the lesion, we diagnosed ocular melanoma *in situ* (Figure 5).

The indicated treatment for an ocular melanoma *in situ* is the surgical resection, but the patient refused treatment and failed to show for the scheduled appointments.

DISCUSSION

Conjunctival melanoma is a rare disease that may arise de novo or from pre-existing conditions such as PAM or conjunctival nevus. About 60% of melanomas arise from PAM. Studies show



FIGURE 3: Ocular biopsy

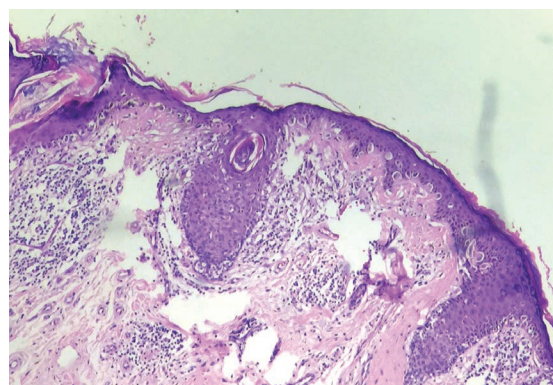


FIGURE 4: Continuous and atypical melanocytic-epidermal proliferation extending to the follicular infundibulum. The dermis presents intense basophilic degeneration of collagen (Hematoxylin & eosin X100)

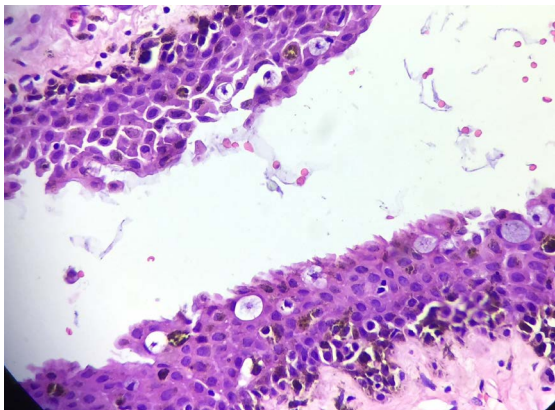


FIGURE 5: Atypical melanocytic proliferation in the conjunctival epithelium with the presence of Paget cells. (Hematoxylin & eosin X400)

similar pattern of increased incidence of cutaneous melanoma and conjunctival melanoma, suggesting a possible link to sunlight exposure and its role in the etiology of conjunctiva melanomas.^{3,4,7,8}

The present case demonstrates the importance of dermoscopy for a precise clinical diagnosis and the importance of multidisciplinary work. □

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