Short communication

Atypical melanocytic nevus of the limbus. A case report

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A B S T R A C T

Case report: A 78-year-old-woman presented with a corneal non-pigmented vascularized tumor of her left eye, of 2 months onset, but with no previous ocular disorders. Surgical excision was performed, and the histopathological study showed the lesion to be an atypical melanocytic nevus of the limbus.

Discussion: Corneal pigmented lesions tend to occur as a result of conjunctival or sclero-corneal limbus lesions spreading or arising de novo from melanocytic cells that have migrated following corneal injury. A biopsy should be carried out to type and distinguish benign lesions (nevus) from pre-malignant or malignant lesions (primary acquired melanosis or conjunctival melanoma).

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Nevus melanocítico atípico limbar. A propósito de un caso

R E S U M E N

Caso clínico: Mujer de 78 años que se presenta con una tumoração corneal no pigmentada y vascularizada de ojo izquierdo de dos meses de evolución y sin antecedentes oftalmológicos. Se realizó exéresis y estudio histopatológico con diagnóstico de nevus melanocítico atípico limbar.

Discusión: La pigmentación corneal procede, en la mayoría de los casos, de la extensión de lesiones de conjuntiva y limbo, o de la migración de sus melanocitos tras una agresión corneal previa. Debemos realizar biopsia y estudio anatomopatológico puesto que, aunque debemos descartar enfermedad precancerosa y maligna (melanosis adquirida primaria y melanoma conjuntival), también existen condiciones benignas como el caso que nos ocupa.

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Introduction

Corneal tumors tend to be the spreading of a primary palpebral or conjunctival tumor.\(^1\) Reports of melanocytic lesions of the cornea are rare; they originate in the conjunctiva and sclerocorneal limbus.\(^2\) However, other cases are true corneal-primary lesions due to melanocyte migration after previous corneal damage.\(^3\)

Case report

A 78-year-old patient seen for a tumor with 2-month progression after she noticed a “red eye” in the mirror. She had no previous ophthalmological disorders.

Left eye biomicroscopy showed a granulomatous tumor with a sessile base of about 6 mm in diameter, embossed with well-defined edges, with transparent tissue and multiple blood vessels within it, without bleaching or pigmentation (Fig. 1). The tumor was predominantly limbal with corneal involvement; we were unable to determine its depth (anterior segment OCT was not available). Visual acuity was 0.7 corrected in both eyes; the rest of the examination was normal.

Due to the nature of the lesion, surgery was performed with complete excision and anatomopathological examination. Since tumor depth was unknown, we waited for availability of a donor corneal graft in the event that a lamellar or penetrating keratoplasty would be needed. Finally, simple treatment was performed under retrobulbar anesthesia; there was no deep corneal invasion, and it was resected without needing a corneal graft.

Pathological examination showed conjunctival and corneal epithelium with melanocytic proliferation in intraepithelial/subepithelial nevi (Fig. 2) with consistent architectural atypia in thecic horizontalization and fusion (Fig. 3) and occasional mitosis in surface layers, but with good maturation to the depth without significant cytologic atypia. Melanin pigment was scarce. Immunohistochemistry confirmed the cells’ melanocytic nature; these were Melan-A and HMB-45 positive, the latter diminishing in depth, as well as proliferative activity, which was measured with Ki-67 (Fig. 4). Histopathological and immunohistochemical results led to a diagnosis of atypical melanocytic compound nevus of the cornea.

Post-operative course was uneventful; 6 months thereafter, the cornea remained clear without signs of recurrence (Fig. 5).

Discussion

Based on biological behavior, conjunctival and corneal lesions of melanocytic origin can be classified as benign (nevus and melanocytosis), precancerous (primary acquired melanosis, PAM), and malignant (racial melanosis and melanoma).

PAM is a proliferation of intraepithelial melanocytes that generate brownish conjunctival plaques, poorly demarcated, which migrate on scleral tissue. It occurs in 60-year-old Caucasian individuals unilaterally. Its precancerous condition is due to the risk of progressing to melanoma (about 30%).\(^3\)

 Conjunctival malignant melanoma is multi-recurrent, difficult to treat and of poor prognosis. It may arise de novo (33%),

Fig. 1 – Preoperative corneal lesion in left eye and without any topical treatment.

Fig. 2 – Melanocytic compound nevus of the cornea. Intraepithelial/subepithelial melanocytic proliferation with sclerocorneal limbus involvement (H-E 100×).

Fig. 3 – Melanocytic compound nevus of the cornea. Intraepidermal and subepithelial component with nevi horizontalization (H-E 200×).
from a pre-existing nevus (33%) or from a PAM. Recurrence rate is around 50% at 10 years, and the mortality is between 25 and 36% at 10 years. Progression depends on its local extension, histological type and treatment efficacy. Therapeutic options include excision with possible cryotherapy, mitomycin C and radiotherapy. Exenteration is reserved for orbital extension and local destruction cases.³

There are cases reported in the literature of early malignant melanoma of the cornea.² Although it is thought that the cornea does not have pigment cells⁴,⁵ and pigment is in melanoblasts located in the basal layer of the conjunctiva and limbus epithelium, these melanoblasts have been reported to migrate toward the corneal epithelium, usually after physical lesions such as trauma, keratitis from different origins and various types of surgery.² Striated melanokeratosis refers to this corneal opacification condition due to pigment presence after corneal damage. There are 2 cases reported in people with fair skin: one case after subconjunctival injection of 5-fluorouracil, and another one in a woman with recurrent corneal erosions and conjunctival melanosis.⁵

The origin of pigmented lesions in the cornea can be attributed to sclerocorneal limbus involvement, mobilization of pigment contained in limbus cells, conjunctiva, uvea or melanoblasts migration. The size of these lesions does not exceed more than one corneal quadrant.

Other conditions that can mimic the appearance of melanin pigment are metals, hemosiderin, lipid, lipofuscin and certain substances (epinephrine and phenothiazines).³

Although whenever a corneal pigmented lesion is observed melanoma must be considered; there are reported cases of benign conditions such as melanocytic nevi.⁷

If malignancy is suspected, a biopsy (complete exeresis plus pathological study), ophthalmological follow-up and, in the event of cellular atypia, including systemic study with multidisciplinary management (internal medicine, oncology, and other); furthermore, nasal extension must be ruled out (ENT examination) and other malignancy-susceptible skin lesions.⁷

**Conflicts of interest**

The authors declare that they have no conflicts of interest.

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