

BRIEF REPORTS

Malignant Melanoma of the Conjunctiva

Grace R. Dizon, MD
Leonardo R. Mangubat, MD
Alexander D. Tan, MD

*Department of Ophthalmology and Visual Sciences
University of the Philippines-Philippine General Hospital
Manila, Philippines*

ABSTRACT

Objective

To report a case of malignant melanoma of the conjunctiva.

Methods

This is a case report of a malignant melanoma of the conjunctiva seen at the University of the Philippines-Philippine General Hospital (UP-PGH).

Results

A 42-year-old male presented with an enlarging, bleeding mass on the left upper lid. There was dark, diffused pigmentation of the caruncle and the bulbar conjunctivae. A section biopsy revealed findings consistent with malignant melanoma. An orbital exenteration was performed.

Conclusion

The management of malignant melanoma of the conjunctiva should involve removal of the tumor, prevention of local recurrence, and prevention of metastasis and death.

Malignant melanoma of the conjunctiva is a rare unilateral disease occurring most frequently in patients 50 years or older.¹ Although its natural history has not been clearly established, it is potentially life-threatening.² This paper presents the history and management of a case of malignant melanoma of the conjunctiva seen at the UP-PGH.

A 42-year-old male sought medical attention for an enlarged mass on the inner portion of the left upper lid. Two darkly pigmented lesions, one measuring 1 mm x 1 mm over the bulbar conjunctiva at the medial canthus, the other measuring 3 mm x 1 mm on the inner portion of the left upper eyelid, had been present since age 10.

Visual acuity was 6/6 (20/20) in the right eye and 6/15 (20/50) pinhole to 6/6 (20/20) in the left eye. Gross examination showed a reddish-black, nodular, vascularized, and bleeding mass measuring 2 cm x 3 cm x 3 cm on the left upper palpebral conjunctiva. There was diffuse pigmentation of the caruncle and bulbar conjunctiva. The rest of the eye findings were normal.

Systemic work-up were negative. Cranial computed tomography (CT) showed a homogenous, slightly hyperdense soft-tissue mass on the left eyelid, measuring 1.5 cm x 2.2 cm x 0.8 cm and without osseous involvement. The left globe was intact.

Differential diagnosis for the pigmented lesions include a large nevus, ciliary body melanoma with extraocular extension, a pigmented conjunctival carcinoma, and melanocytoma. Grossly, these disease entities are similar; only a histological examination can differentiate them from true malignant melanoma.

A section biopsy of the mass was done showing epithelial configuration of spindle cells also seen in samples taken from the medial and lateral canthal areas.

Two principal cell types are found in malignant melanoma: the fascicular type where spindle cells are arranged in rows and the epithelioid configuration consisting of larger polygonal cells with distinct boundaries.

Malignant melanoma starts as an extraepithelial proliferation of atypical melanocytes that can spread radially (superficial spread or pagetoid growth) within the epithelium for prolonged periods and produce flat, golden brown pigmentation that may have a waxing and waning course. A vertical, invasive phase occurs later, which extends into the substantia propria. If the radial phase is short, a localized invasive nodule may become present.³ Malignant melanomas have no known association with solar exposure, but host factors play an important role in its development.²

Almost all conjunctival melanomas develop in a preexisting pigmented conjunctival lesion. Eighteen percent arise in a conjunctival nevus and can occur as a solitary circumscribed lesion, which never recurs after local excision. About 57% present as a diffuse lesion or multiple lesions associated with or arising from primary acquired melanosis (PAM) of the conjunctiva.^{2, 3} PAM usually presents in middle age with flat, granular, intraepithelial reddish-brown pigmentary change, initially noted in the bulbar conjunctiva. Clues to malignant transformation of PAM include increased thickness, a change in pigmentation, the appearance of prominent blood vessels feeding a tumor, and tethering of the

conjunctiva to the underlying sclera, where it is usually mobile.³ In this patient, the lesion behaved like it arose from a PAM by spreading to contiguous areas, developing blood vessels that fed the tumor, and increasing in thickness and size over the years.

When the melanocytes remain in the epithelium, there is no possibility for metastasis. However, once intraepithelial pagetoid tumors involve 50% to 100% of the bulbar and palpebral conjunctiva, the chance for metastasis is larger.^{1,2} Advanced melanomas ultimately invade lymphatics and spread to preauricular, submandibular, and cervical nodes before disseminating to the parotid gland, liver, subcutaneous tissues, and brain.

Management of malignant melanoma of the conjunctiva is difficult and the subject of many controversies. Complete excision of the tumor and prevention of local recurrence, metastasis, and death are the goals.² Options include a wide excision of the melanoma with cryotherapy of the margins,⁵ wide excision with topical chemotherapy such as mitomycin-C,⁴ (Mitomycin C, Kyowa, Tokyo, Japan), wide excision with radiotherapy, and orbital exenteration. For small circumscribed melanomas, beta-irradiation with strontium-90 surface applicators, proton-beam therapy, gamma emissions from cobalt-60 or cobalt plaques have been found effective.⁴

Due to the extensive involvement of the conjunctivae, including the caruncle and upper lid, an orbital exenteration was done in this patient. Histopath results showed largely epithelioid configuration of spindle cells. The eyeball, optic nerve, lacrimal gland, and all surgical margins were negative for tumor.

Malignant melanoma is the most serious of the conjunctival malignancies. Nearly 50% of patients have recurrence after resection, while 26% develop metastasis after 10 years.¹ Prognosis is often poor for patients with malignant melanoma in the palpebral conjunctiva, fornices, plica, caruncle, or eyelid margin. Among these patients, mortality is 2.2 times higher.² Prognosis improves for patients with lesions found in the bulbar conjunctiva.¹

Proximity to the lymphatic system may explain why some locations are less favorable than others.² Survival rates are worse for the following: lesions arising from PAM, those with an initial thickness of more than 4 mm, lesions arising from an unfavorable site, multifocal disease in favorable sites, mixed spindle and pure epithelioid cells with lymphatic invasion, high mitotic index, and lesions that grow vertically.^{1,2}

In this patient, prognosis was considered poor because the tumor involved the palpebral conjunctiva, fornices, and caruncle, and the lesions arose from a PAM. Whether orbital exenteration can prevent metastasis is uncertain. Some studies have reported that the procedure did not lead to increased survival rate.⁴

References

1. DePotter P, Shields CL, Shields JA, Menduke H. Clinical predictive factors for development of recurrence and metastasis in conjunctival melanoma: A review of 68 cases. *Br J Ophthalmol* 1993; 77: 624-630.
2. Paridaens A, Minassian D, McCartney A, Hungerford J. Prognostic factors in primary malignant melanoma of the conjunctiva: A clinicopathological study of 256 cases. *Br J Ophthalmol* 1994; 78: 252-259.
3. Paridaens A, McCartney A, Hungerford J. Multifocal conjunctival melanoma and acquired melanosis sine pigmento. *Br J Ophthalmol* 1992; 76: 163-165.
4. Hungerford J. Management of ocular melanoma. *Br Med Bull* 1995; 51: 694-716.
5. Shields J, Shields C, DePotter P. Surgical management of conjunctival tumors. *Arch Ophthalmol* 1997; 115: 808-815.

Retinal Dysplasia*

Mary Ellen A. Sy, MD
Romeo B. Espiritu, MD

Department of Ophthalmology and Visual Sciences
University of the Philippines-Philippine General Hospital
Manila, Philippines

ABSTRACT

Objective

To report a case of retinal dysplasia.

Method

This is a case report of retinal dysplasia, the first documented case seen at the University of the Philippines-Philippine General Hospital (UP-PGH).

Results

A three-month-old female presented with bilateral leukocoria and an intraocular mass in the right eye. The right eyeball was enucleated and histopathology results revealed retinal dysplasia.

Conclusion

Ocular ultrasonographic and cranial computerized tomography are helpful in the diagnosis of suspected retinal dysplasia.

Retinal dysplasia consists of an abnormal proliferation of developing retina, producing tubular structures with a rosette-like appearance in cross section.¹ It appears to be associated with a single pathogenetic basis: the separation of the retina during a critical stage of its differentiation from its underlying pigment epithelium.²

We are presenting the first reported case of retinal dysplasia seen at the UP-PGH.

A three-month-old female, the second of twins, born preterm at 7 months (1,400 grams) via Caesarean section,

*Presented at the Annual Meeting of the Philippine Academy of Ophthalmology, November 2003