

Conjunctival Leiomyosarcoma

Loo Wan-Wei^{1,3}, Ahmad-Alwi Azma-Azalina¹, Zakariah Sakinah¹, Awang Saleena², Ismail Shatriah^{3,4*} and Embong Zunaina^{3,4}

¹Department of Ophthalmology, Hospital Raja Perempuan Zainab II, Kota Bharu, Kelantan, Malaysia

²Department of Pathology, Hospital Raja Perempuan Zainab II, Kota Bharu, Kelantan, Malaysia

³Department of Ophthalmology, School of Medical Sciences, Universiti Sains Malaysia, Kubang Kerian, Kelantan, Malaysia

⁴Hospital Universiti Sains Malaysia, Kubang Kerian, Kelantan, Malaysia

Abstract

A 60-year-old lady presented with a fleshy mass at the nasal limbus of the right eye of two months' duration which was progressively increase in size. Right eye examinations showed a fleshy exophytic mass arising from inferonasal limbus with the base involving cornea and conjunctiva. There was presence of few big feeder vessels at medial part of the mass. The mass had keratinized surface and bled easily on palpation. Excision of the mass was performed in order to rule out conjunctival squamous cell carcinoma. Histopathological examination of the mass revealed malignant spindle cells arranged in interlacing bundles with prominent nucleoli, abundant eosinophilic cytoplasm and atypical mitotic activity. There were positive immunohistochemical staining for actin, vimentin and S-100. The histopathological features were consistent with leiomyosarcoma of the conjunctiva. There was no sign of recurrence at 5 month post excision.

***Corresponding Author:** Shatriah Ismail, Department of Ophthalmology, School of Medical Sciences, Universiti Sains Malaysia, 16150 Kubang Kerian, Kelantan, Malaysia; E-mail: shatriah@usm.my

Keywords: Conjunctival Mass; Squamous Cell Carcinoma; Pterygium; Exophytic Mass

Introduction

Leiomyosarcoma is a malignant tumour arising from smooth muscle. It has been reported arising primarily from the orbit and also from distant metastases. Leiomyosarcoma arising primarily from the conjunctiva is very rare, as there were only five cases being published previously. We present a rare case of conjunctival leiomyosarcoma.

Case Report

A 60-year-old lady presented with a fleshy mass at the nasal limbus of the right eye of two month's duration. The mass was progressively increasing in size and was associated with spontaneous bleeding occasionally. However, there was no eye pain, eye redness or eye discharge. She also has history of painless progressive blurring of vision in the right eye for the past two years. There was no significant history of ocular trauma or surgical procedure done before. She was not known to have any medical illness or malignancy.

The vision in the right eye was counting finger and was 6/9 in the left eye. Right eye examination showed a fleshy exophytic mass at the nasal limbus (Figure 1). The mass arise from inferonasal limbus with the base involving cornea and

conjunctiva. There were few big feeder vessels at the medial part of the mass. The surface of the mass showed some area of keratinisation and bled easily on palpation. The visible part of the cornea was clear. Anterior segment examination was normal with presence of brown mature cataract. There was no view of the right fundus due to the presence of dense mature cataract. B-scan ultrasonography of the right eye was normal with no sign of intraocular mass or retinal detachment. The left eye examination of the anterior segment was normal. The left eye fundus showed pink optic disc with cup-disc ratio of 0.3 and the retina was normal. Intra-ocular pressure of the right eye was 13 mmHg and 10 mmHg in the left eye.



Figure 1: Right eye showed a fleshy exophytic mass arising from inferonasal limbus involving cornea and conjunctiva

Excision of the mass was performed in order to rule out conjunctival squamous cell carcinoma. Histopathological examination of the mass revealed an ulcerated polypoidal tissue composed of malignant spindle cells arranged in interlacing bundles. The malignant cells have spindle to oval nuclei with irregular nuclear membrane, prominent nucleoli and abundant eosinophilic cytoplasm (Figure 2). There was also presence of occasional large pleomorphic cells with bizarre nuclei. Inflammatory cell infiltrates and pyknotic cells are seen within the tumour cells. Atypical mitotic activities are also frequently encountered. There were positive immunohistochemical

staining for actin, vimentin and S-100, and negative staining for cytokeratin, HMB-45, Desmin, CD 31 and CD 34. The histopathological features were consistent with leiomyosarcoma of the conjunctiva. Follow-up at 5 month post excision of the mass, there was no sign of recurrence. The right eye showed presence of pterygium at the nasal part of the sclera (Figure 3).

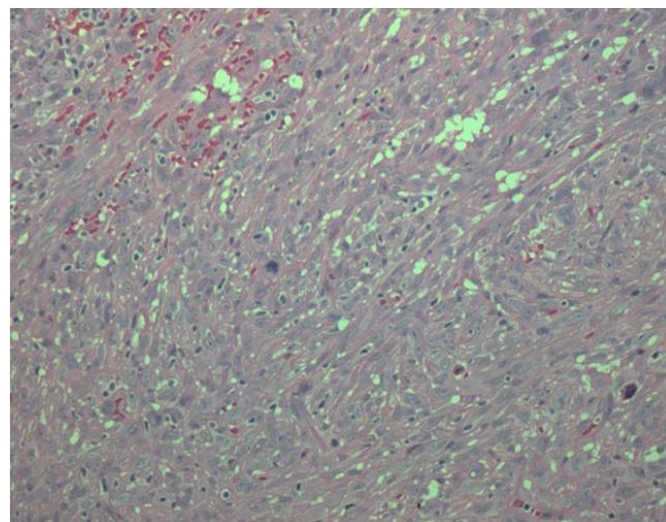


Figure 2: Malignant spindle cells arranged in interlacing bundles showing prominent nucleoli and abundant eosinophilic cytoplasm (Hematoxylin-eosin stain)

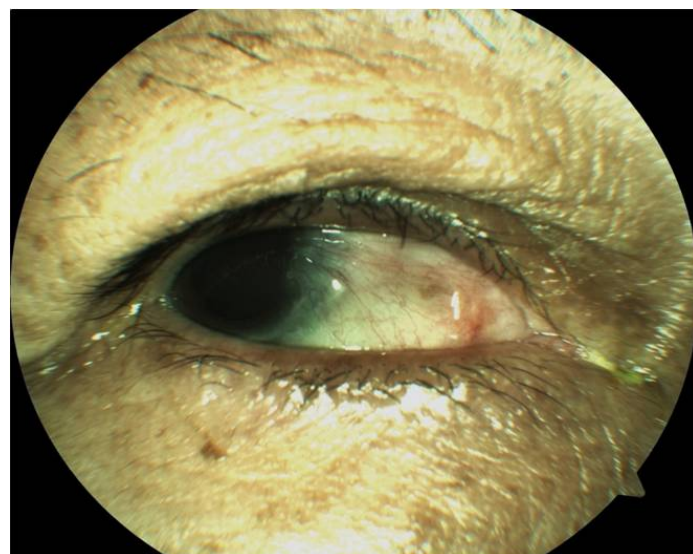


Figure 3: Right eye showed presence of pterygium nasally with no sign of recurrence at 5-months post excision

Discussion

Leiomyosarcoma is a smooth muscle malignancy in which it frequently arises from uterus, stomach, intestines, blood vessels and skin. Primary orbital leiomyosarcoma occurs rarely [1, 2], so does secondary leiomyosarcoma which arises after irradiation or from metastatic disease.

Conjunctival leiomyosarcoma is very rarely seen, as to date, there are only five cases being reported [3-7]. Table 1 summarizes the five cases of reported conjunctival leiomyosarcoma in the literature from 1976-2012. Histopathological and immunohistochemical staining confirmed the diagnosis of leiomyosarcoma. First case was reported by de Wolff-Rouendaal [3] in 1976 where a 20-year-old lady with history of xeroderma pigment sum presented with limbal tumour which was initially diagnosed to be leiomyoma. The tumour recurred rapidly after excision and subsequently required exenteration. Leiomyosarcoma was suspected but the final histopathological diagnosis was never being confirmed as the original slides were not available anymore. White et al. [4], Yoon et al. [5] and Katircioglu et al. [6] reported case of leiomyosarcoma of the conjunctiva. Those three cases presented differently but there was one similarity found in all the three cases- they were initially diagnosed or suspected to be a

different pathology but final immunohistochemical examination confirmed the diagnosis of conjunctival leiomyosarcoma. White et al. [4] reported a 66-year-old male who presented with pseudopterygia after penetrating keratoplasty, whom was previously diagnosed to have squamous cell carcinoma of the conjunctiva 26 years before that. Definitive diagnosis of leiomyosarcoma was made after repeated excision and histopathological examination. It remains unknown whether the second tumour that occurred at the same site was the same as the initial lesion. Yoon et al. [5] reported a case in which the patient had been diagnosed to have pterygium at the site of tumour and excision of pterygium was done a year before. The mass was then excised to rule out conjunctival intraepithelial neoplasm but eventually the definitive diagnosis of leiomyosarcoma of conjunctiva was made after analysis by immunohistochemical staining. In the case reported by Katircioglu et al. [6], a 70-year-old male presented to them with recurrent exophytic mass. There were two surgeries done for the same eye within the last two years with a preliminary diagnosis of pterygium without any histopathological examination. Modified enucleation was done for the patient and definitive diagnosis of leiomyosarcoma with involvement of medial recti muscle was made.

Table 1: Reported cases of conjunctival leiomyosarcoma

Author/Year	Age/Gender	Presentation	Initial impression	Treatment
De Wolff-Rouendaal, 1976	20/Female	Limbal lesion	Leiomyoma	Recurred after excision. Subsequently required exenteration
White et al, 1991	66/Male	Pseudopterygia	Squamous cell carcinoma	Extensive excision
Yoon et al, 2006	59/Male	Limbal lesion	Conjunctival intraepithelial neoplasm	Excision
Katircioglu et al, 2009	70/Male	Recurrent limbal lesion	Not stated	Modified enucleation
Kenawy et al, 2012	37/Female	Conjunctival mass	Not stated	Wide excision

A recent case of primary leiomyosarcoma of conjunctiva published by Kenawy et al. [7] reported a 37-year-old lady with history of cutaneous melanoma of the abdominal skin which was successfully treated with excision with no

evidence of recurrence or distant metastases. She presented with a smooth conjunctival vascularized tumour with computed tomography of the orbit showed soft tissue mass arising from the medial rectus insertion without bone or deep involvement.

Wide surgical excision was done included some medial and superior recti muscle fibers.

In this particular case that we have encountered, excision of the conjunctival mass was carried out initially to rule out conjunctival squamous cell carcinoma. Review at 5 months post excision showed no local recurrence but long term follow-up and further surveillance are required.

Conjunctival squamous cell carcinoma is far more commonly seen than conjunctival leiomyosarcoma, evidenced by no case demonstrated in the major review of tumours of the conjunctiva and cornea done by Shields et al. [8]. The rarity of this smooth muscle cells malignancy requires analysis by immunohistochemical staining for definitive diagnosis. Histological method alone is not sufficient to diagnose this rare disease.

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