Leiomyosarcoma of the Conjunctiva: Case Report

Konjonktivanın Leiomiyosarkomu

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Geliş Tarihi/*Received:* 06.05.2009
Kabul Tarihi/*Accepted:* 17.07.2009

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ABSTRACT We present a case with leimoyosarcoma of the conjunctiva and orbit. A 70-year-old male patient who admitted with an exophytic mass covering interpalbepral fissure is presented in this case report. The past medical history was positive for a recurrent mass after two simple excisions with a preliminary diagnosis of pterygium. The orbital tomography demonstrated a 2 x 1.5 x 1.6 cm mass involving cornea. The mass had scleral and medial rectus muscle involvement, as seen intraoperatively. The mass was excised including medial rectus muscle (modified enucleation) and the conjunctiva was also excised with a 5 mm tumor-free area. The histopathologic examination confirmed the diagnosis of leimyoscarcoma. Postoperative radiotherapy was administered. Conjunctival involvement is a rare clinical form of orbital leiomyoscarcoma. Atypical conjunctival lesions and recurrent pterygia should be carefully approached and pathological examination should be performed to rule out malignant lesions like leiomyosarcoma.

Key Words: Leiomyosarcoma; pterygium; conjunctiva

ÖZET Konjonktiva ve göz küresinin leiomiyosarkomunu sunuyoruz. Yetmiş yaşında erkek hasta interpalpebral fissürü kaplayan eksofitik kitle ile başvurdu. Tıbbi özgeçmişinde tekrarlayan kitle nedeniyle iki kez basit eksizyon operasyonları mevcuttu ve bunlarda ön tanı pterigium idi. Orbital tomografide 2 x 1.5 x 1.6 cm boyutlarında, korneayı içine alan kitle saptandı. Operasyon sırasında kitlenin sklera ve medial rektus kasını da tuttuğu gözlendi. Kitle medial rektus kası da dahil olmak üzere eksize edildi (modifiye enükleasyon). Konjonktiva 5 mm tümörsüz alan bırakılarak çıkarıldı. Patolojik inceleme leiomiyosarkom tanısını kesinleştirdi. Postoperatif radyoterapi uygulandı. Konjonktiva tutulumu, orbital leiomyosarkomun nadir bir klinik formudur. Atipik konjonktiva lezyonlarına ve tekrarlayan pterigiuma dikkatle yaklaşılmalı ve leiomiyosarkom gibi malign lezyonları dışlamak için patolojik inceleme yapılmalıdır.

Anahtar Kelimeler: Leiomiyosarkom; pterijiyum; konjonktiva

Turkiye Klinikleri J Med Sci 2011;31(6):1544-6

Leiomyosarcoma is a malignant tumor of the smooth muscle cells. Although it is typically located in the uterus and in the gastrointestinal tract, it is seldomly reported in the ocular structures.[1-3]

In this study, a case with leiomyosarcoma of the conjunctiva is reported, which is the third one reported in this location.

CASE REPORT

A 70-year-old male patient was admitted with a recurrent exophytic mass in the interpalpebral fissure of the right eye (OD). The patient's past medi-

doi:10.5336/medsci.2009-13341

cal history consisted of two surgeries of the same eye within the last two years, with a preliminary diagnosis of pterygium. Unfortunately, the excised material was not examined histopathologically, and the clinical diagnosis was not confirmed.

On examination, the visual acuity was light perception in OD and 20/20 in the left eye (OS). On biomicroscopy, there was an exophytic mass originating from the medial limbus covering the entire cornea OD (Figure 1). Anterior and posterior segment and pupillary examination could not be performed OD, whereas it was normal OS. Eye motility examination revealed restricted adduction and abduction of OD, but OS had full range of motion.

Orbital computerized tomography revealed a 2 x 1.5 x 1.6 cm mass involving cornea OD. The mass seemed to extend intraocularly to the lens border, medially.

Patient was prepared for a modified enucleation surgery. During the surgery, tumor mass was seen invading the medial rectus muscle. Therefore, medial rectus muscle was also excised to 10 mm from the visible tumor margin. Conjunctiva was excised 5 mm from the visible tumor margin and cryotherapy was applied to the remaining conjunctival borders. Gross microscopy of the enucleation material revealed extension of the tumor to the sclera and intraocular structures (Figure 2). Trichrome staining of the tumor slides demonstrated extensive red staining of the smooth muscle cells (Figure 3).

Further pathological examination demonstrated atypical mesenchymal cells with large hyperchromatic round-oval nuclei and cytoplasm with bipolar extension showing irregular bundle formation in various directions (Figure 4). There were extensive atypical mitoses in every field. Immunehistochemical staining showed positivity for smooth muscle actin (SMA), H-Caldesmon, desmin and vimentin, which confirmed the diagnosis of leiomyosarcoma (Figure 5). Further staining with CD-68, S-100, keratin and HMB-45 were negative. CD-117 was also negative. These results were used to rule out malignant fibrous histiocytoma, schwannoma, spindle cell squamous carcinoma and malignant melanoma, respectively.

Patient was referred to the Oncology Department and a full body tomography was performed to rule out a possible distant metastasis. Patient was given a total dose of 6600 cGy in 33 sessions of radiotherapy (in fractions of 200 cGy) to the orbital region. Two years after surgery, the patient was symptom-free with no recurrences or distant metastasis, as investigated by the referred Oncologist.

DISCUSSION

Leiomyosarcoma of the conjunctiva has been reported previously in two case studies.[1,2] The first case was a 20-year-old woman with xeroderma pigmentosum, who also had numerous cutaneous squ-

FIGURE 1: Exophytic mass is seen originating from the medial limbus and covering the entire cornea of the left eye.

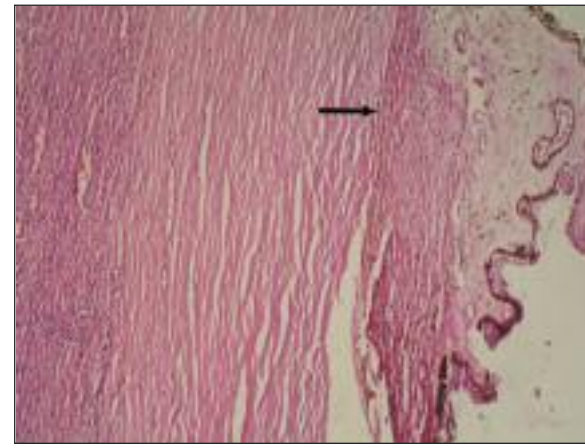

FIGURE 2: Extension of the tumor to the sclera and intraocular structures on pathological examination (Hematoxylin-eosin, x100).

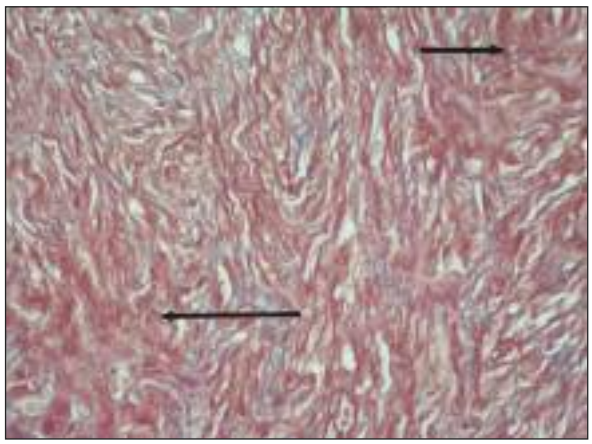

FIGURE 3: Red staining of the smooth muscle cells is seen on trichrome stain (x400).

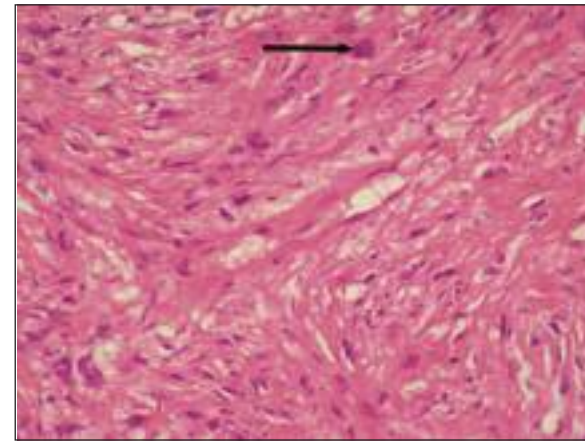

FIGURE 4: Atypical mesenchymal cells are seen forming irregular bundles in various directions (Hematoxylin-eosin, x400).

amous cell and basal cell melanomas as well as melanomas.[1] The preliminary diagnosis was leiomyoma on the biopsy, however, after rapid recurrence, exenteration was performed and a definitive diagnosis was reached. The second case was a 66-year-old man who presented with pseudopterygia after penetrating keratoplasty.[2] Actually, this patient was diagnosed with squamous cell carcinoma of the conjunctiva 26 years before the final diagnosis, but the definitive diagnosis was confirmed after repeated excision and pathological examination.

The rarity of this lesion has been confirmed in a review of 2455 conjunctival specimens by Grossniklaus et al, where no case had been demonstrated.[4] Pterygium is a much more frequent lesion and its rank is much higher in the list of differential diagnoses of similar conjunctival lesions. Although it is hard to know whether some of these patients remain undetermined, the lack of pathological workup in the prior surgeries of this case brings out the subject of misdiagnosis. We believe that, recurrent pterygial tissues and atypical pterygia should be studied by pathological examination and a definitive diagnosis should be reached, especially in developing countries in our geographical area.

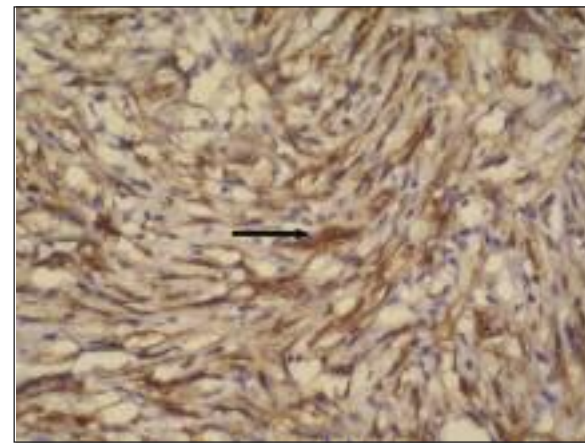

FIGURE 5: Immunohistochemical staining demonstrates high positivity for smooth muscle fibers (H-caldesmon, x400).

Nevertheless, as experienced in the earlier two cases, leiomyosarcoma may be confused histopathologically, and further workup, like immunoperoxidase study or electron microscopy may deem necessary, as in our case.

REFERENCES

1. Meekins BB, Dutton JJ, Proia AD. Primary orbital leiomyosarcoma. A case report and review of the literature. Arch Ophthalmol 1988;106(1):82-6.
2. White VA, Damji KF, Richards JS, Rootman J. Leiomyosarcoma of the conjunctiva. Ophthalmology 1991;98(10):1560-4.
3. Shields CL, Shields JA. Tumors of the conjunctiva and cornea. Surv Ophthalmol 2004; 49(1):3-24.
4. Grossniklaus HE, Green WR, Luckenbach M, Chan CC. Conjunctival lesions in adults: a clinical and histopathological review. Cornea 1987;6(2):78-116.