Conjunctival leiomyosarcoma: A case report and review of literature

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Leiomyosarcoma is a malignant mesenchymal tumor that is very uncommon in the conjunctiva. Nevertheless, we describe here the clinical manifestations, management, and prognosis of a rare case of leiomyosarcoma in this location. An 81-year-old male presented at a tertiary hospital with a rapidly growing mass. After performing biopsy, histopathological examination revealed the existence of a conjunctival leiomyosarcoma. On this diagnosis, a thorough metastatic screening was performed showing no enlarged lymph nodes or metastatic deposits anywhere in the body. To treat the condition, we performed an evisceration with clear margins and subsequent radiotherapy.

Key words: Conjunctiva, eye, leiomyosarcoma, sarcoma, tumor

Leiomyosarcoma is a malignant tumor that originates from the smooth muscle lineage and is considered one of the most frequent soft tissue sarcomas. It has an incidence that increases with age, with a peak at the seventh decade. The sex prevalence is highly variable and depends on its location.[1] Traditionally, soft tissue sarcomas have a high mortality rate related to their ability to recur elsewhere.[2] We describe the presentation and management of a conjunctival leiomyosarcoma.

Case Report

An 81-year-old male presented at a tertiary hospital, in June 2012, due to a rapidly growing painful growth of a 2-year-old mass in his right eye. Symptoms had developed over the previous 2 weeks. He had a prior history of retinal detachment, which had been treated with pars plana vitrectomy, encirclage, and cryopexy 8 years ago. Following treatment, the eye did not recover any vision.

On ophthalmological examination, a highly vascularized, lobulated, bright red, tender mass in his right eye was observed developing from the conjunctiva [Figs. 1 and 2]. It hindered eye closure, but did not affect ocular motility.

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A computed tomography (CT) scan of the orbit showed a soft tissue density measuring 1.7 cm × 1.1 cm × 1.5 cm located between the anterior pole of the right eye and its tarsal conjunctiva, suggestive of an exophytic lesion. However, there was no extension into the orbit and no bone erosion [Fig. 3].

We decided to perform an evisceration due to the anterior location of the lesion and placed an implant in the socket. Small tissue biopsies were taken from the sclera, conjunctiva (extending for six clock hours); the medial, superior, and lateral recti muscles. The histopathological examination revealed clear surgical margins of at least 5 mm. Macroscopically, tumor fragments were fleshy, elastic, soft, and of red-purple color with translucent whitish areas.

The histological examination revealed a spindle cell tumor, with eosinophilic fibrillary cytoplasm, blunt nuclei, fascicles of different sizes, and an occasionally storiform aspect. We also observed variable pleomorphism with a high mitotic index. In some areas, we could see myxoid stroma. Masson's trichrome stained red for cytoplasm; consistent with its muscular nature. Smooth muscle actin and vimentin expressions were positive, while cytokeratin, melan-A, HMB-45, and S-100 protein were negative [Fig. 4].

Cytologically, the diagnosis from tumor smears was of malignant spindle cell tumor concordant with leiomyosarcoma. Based on these findings, we sent the patient to the orbital department of the reference hospital for further management. More biopsies were taken near the excision area. These biopsies showed no involvement of the structures by tumor cells, so no more surgical management was performed and periodical controls were scheduled. Nevertheless, a postoperative radiation dose of 60 Gy was applied in 2 Gy per fraction with the two-isocenter technique with energy of 6 Mv each, involving the entire orbit. A whole body CT scan excluded lymphadenopathies and metastatic or primary lesions elsewhere. As per the American Joint Committee on Cancer (AJCC) staging system, the disease was classified as T1aN0M0.[3] A follow-up of 1 year revealed no tumor recurrence.

Discussion

White *et al*. reported a case of a 66-year-old male with a leiomyosarcoma involving the conjunctiva and also the cornea.[4] He had a previous history of a symptomatic conjunctival lesion 26 years ago. Once the diagnosis of the tumor was reached, the patient was treated with subtotal exenteration with no other concomitant treatment. No evidence of metastatic disease was seen at the time of the

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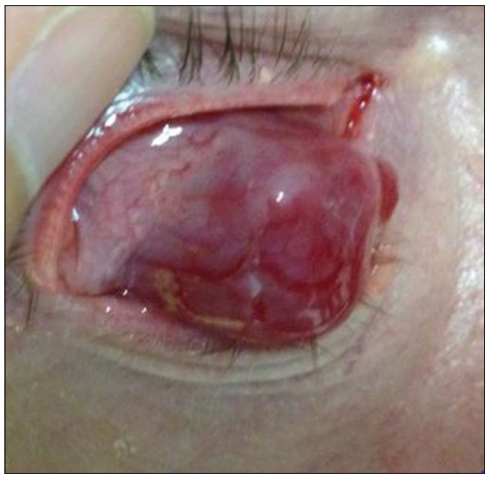

Figure 1: Color photograph of the anterior segment of the right eye showing a pink, fleshy, vascularized conjunctival mass

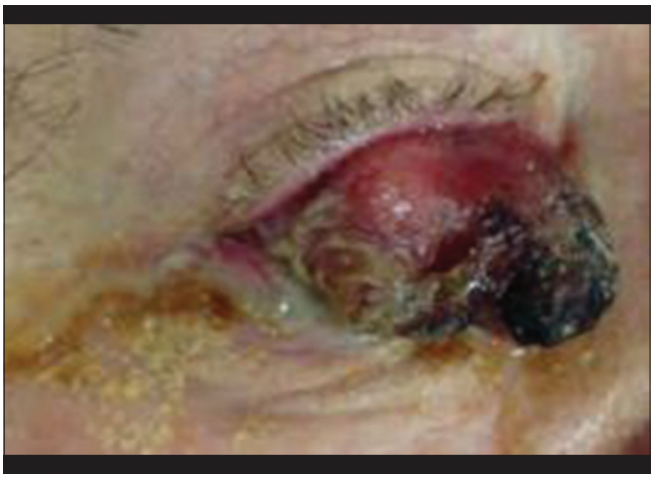

Figure 2: Color photograph of the anterior segment of the right eye showing a heterogeneous conjunctival mass, with necrotic tissue, scabs, and purulent discharge 2 weeks later

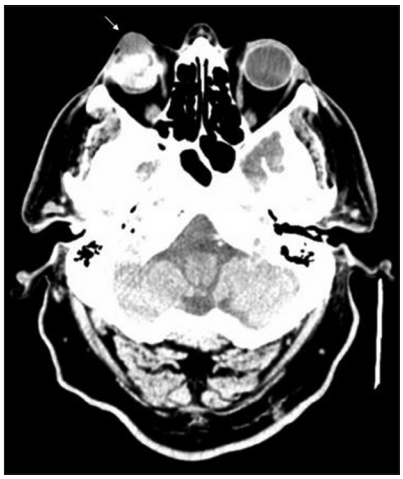

Figure 3: Computed tomography scan of the orbit showing a soft tissue mass between the anterior pole of the right eye and its tarsal conjunctiva (arrow)

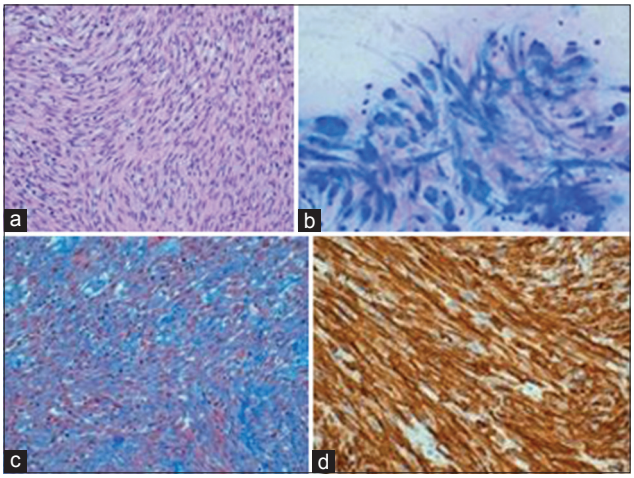

Figure 4: (a and b) Tumor with pleomorphic and myxoid areas with inflammatory infiltrate (H and E, ×20). (c) Spindled areas with mitoses and moderate pleomorphism (H and E, ×40). (d) Neoplastic cells, positive for smooth muscle actin (smooth muscle actin, ×20)

surgery, and a CT scan 6 months later demonstrated that there was no residual neoplasm. A 2-year survival period was reported.

Guerriero *et al*. in their report described a case of a 56-year-old female with a fast growing mass, originating in a previous conjunctival lesion that she had for years.[5] An examination using electron microscopy showed irregular cells and often grooved nuclei with prominent nucleoli, moderate amounts of cytoplasmic thin filaments with focal densities, immature cell junctions, and discontinuous basal lamina material along the tumoral cell surface. A CT scan showed adenopathies, which were probably of a metastatic origin. Due to the fast growth and the malignancy of the lesion, she was treated with orbital exenteration, chemotherapy, and radiotherapy.

Kenawy *et al*. reported a case of a 37-year-old female with a painful swelling area in her left conjunctiva for 8 weeks.[6] Examination showed a soft tissue mass arising from the medial rectus with no bone or deep involvement. The treatment consisted of wide surgical excision. Complete examination revealed no metastatic disease. A follow-up of 1 year revealed no signs of recurrence.

Nair *et al*. documented a first case of a 34-year-old male with the presence of a conjunctival and corneal mass of 2-year evolution that was diagnosed as conjunctival leiomyosarcoma.[7] It presented orbital extension, so the patient underwent orbital exenteration. The second documented case was of a 39-year-old male with an excised limbal mass. A later review of the histopathological slides suggested a diagnosis of conjunctival leiomyosarcoma. He was also treated with radiotherapy. A follow-up of 1 year revealed no tumor recurrence or systemic metastasis in both cases.

White *et al*. compared the characteristics of the conjunctival leiomyosarcoma to the superficial skin lesions and postulated that they both have a similar benign prognosis. However, as was reflected by Serrano and George, not only do the histologic grade, tumor size, and

tumor depth – characteristics included in the AJCC – have an influence on the prognosis, but also the anatomic site plays an important role.[1]

The heterogeneous appearance of leiomyosarcomas reflects the need to use a biopsy to reach the correct diagnosis of the specimen.[7] Due to the differential diagnosis with other types of tumors, such as spindle cell lymphoma, large cell lymphoma, melanoma, and undifferentiated sarcoma, immunohistochemical staining plays an important role in the correct identification of leiomyosarcomas.[5] Due to the hematogenous spread of leiomyosarcomas, a positron emission tomography-CT scan is also required.

The small number of leiomyosarcomas originating in this specific location hinders the existence of a standardized treatment to manage them. An option could be to follow the recommendations for other locations such as in the limbs where the main treatment in the case of localized leiomyosarcomas is surgical resection with a complete excision with wide negative margins, with or without adjuvant treatment.[1,8-10] According to the literature, three of the cases of conjunctival leiomyosarcoma underwent orbital exenteration.[4,5,7] In our case, and in the case reported by Kenawy *et al*.,[6] a more conservative approach was performed (tumor excision with clear margins). Nevertheless, due to its aggressive course and tendency to recur, an orbital exenteration or extended enucleation should be considered as the treatment of choice.

The presence of negative surgical margins, as reported in our case, has been shown to play a major role in decreased local and distant recurrences of soft tissue sarcomas and in increased overall survival.[10]

Although radiotherapy is an important option to consider in the treatment of soft tissue sarcomas of the extremities and trunk, there is no agreement on its role at other locations.[1,11] On the other hand, there is no evidence of any benefit, in terms of survival, in the use of postoperative chemotherapy in the treatment of this kind of tumor.[10] Due to this, the advanced aged of our patient, his general poor condition, and the absence of metastasis on a complete examination, we decided not to apply it.

The prognosis of this specific type and site of sarcoma is not well established by large series. In the case of the overall group of soft tissue sarcomas, the median survival time is less than a year from diagnosis of metastases.[10]

Conclusion

Histopathological examination plays an important role in the diagnosis of conjunctival leiomyosarcoma due to its rareness and low clinical suspicion. An aggressive treatment should be considered due to its high recurrence rate.

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Conflicts of interest

There are no conflicts of interest.

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