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Successfully Treated Inflammatory Myofibroblastic Tumor Case of Conjunctiva and Cornea

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ABSTRACT

Inflammatory myofibroblastic tumor (IMT) is a tumor composed of myofibroblasts and a mixed inflammatory infiltrate. IMT is seen extremely rare in the anterior orbit. It might be confused with malignancy frequently. We present the case of a 60 year-old male with IMT of the conjunctiva and cornea, successfully treated by tumor resection and sistemic corticosteroid treatment.

KEYWORDS Cornea, Conjunctiva, Corticosteroid, Inflammatory myofibroblastic tumor

Introduction

Inflammatory myofibroblastic tumors (IMTs) are primarily seen in visceral and soft tissue of children and young adults. However, IMTs can be seen throughout adulthood. The etiology of IMTs is unclear and they rarely show metastasis (1). They consist of myofibroblastic spindles accompanied by an inflammatory infiltrate of plasma cells, lymphocytes, and eosinophils (1, 2).

IMTs are involved in several anatomic sites including the soft tissues, lungs, bladder, spleen, breasts, and colon. They can also occur in the orbital region, but are rarely seen in the anterior orbital region. To the best of our knowledge, only two case of IMTs of the anterior orbit have been reported (2, 3). Here, we report on the case of a 60-year-old male with an IMT of the conjunctiva and cornea that was successfully treated by a tumor resection and systemic corticosteroid treatment.

Case Report

A 60-year-old man presented with a complaint of pain and visual loss in the right eye. The patient had previously been diagnosed with leprosy and had a history of decreased visual acuity for the past two years and a visual loss over the last two months. The mass, which filled the palpebral fissure and completely covered the cornea, had an irregular surface with purulent secretions on its surface that were identified in an ophthalmic examination (Figure 1A-B). Visual acuity was light perception without projection (p+p-) in the right eye and counting fingers at 2 meters in the left eye. Both the right and left eyelid skins were loose due to lagophthalmos. A vascular leukoma in the inferior half of the cornea and conjunctival hyperemia were observed in the left eye. Magnetic resonance imaging demonstrated normal bulbus oculi integrity with a contrast-enhanced mass, 28x10 mm in diameter, anterior to the right bulbus oculi (Figure 2). The mass in the right eye was excised along with healthy conjunctival tissue under general anesthesia. Because of the infiltration of the cornea, it was partially scraped with a crescent knife (Figure 3). An amniotic membrane transplantation was performed on the cornea and open conjunctival area because the conjunctival opening was large and there was a wide eyelid opening with conjunctival defects caused by leprosy. A temporary tarsorrhaphy was performed for eyelid laxity. Postoperatively, the patient was treated with moxifloxacin eye drops six times daily and per oral amoxicillin/clavulanate capsules twice daily. Upon pathological examination of the removed mass with hematoxylin and eosin staining, we determined the mass was composed of a proliferation of short spindle-like fibroblasts and myofibroblasts accompanied by an inflammatory infiltration of plasma cells, lymphocytes, and eosinophils (Figure 4). The spindle cells were seen arranged in a herringbone, fascicle or storiform, irregular, and poorstained. Immunohistochemistry showed that part of the tumor cells stained positive for muscle-specific actin (MSA) (Figure 5). Other markers, including desmin, s-100 protein, and CD34, were negative. A pathological examination reported the mass as an inflammatory myofibroblastic tumor. After pathological diagnosis, systemic prednisolone therapy was given at 1 mg/kg/day for one month. Visual acuity was hand motion level at the 1-month postoperative visit. The superior half of cornea was clear with a vascularized corneal leukoma in the inferior half of cornea (probably due to lagophthalmos), and a mature cataract was observed (Figure 6A-B). Permanent tarsorrhaphy was performed after two months. An 18-month follow-up examination found no evidence of recurrence.

Discussion

Although IMTs are seen most frequently in the first two decades of life, the age range extends throughout adulthood. The

etiology of IMTs are unclear (1). Some authors have suggested their etiology is an unusual response to precipitating factors such as trauma and localized infection, as a post-inflammatory reparative process, or as a reaction to chronic inflammation (4, 5). The finding of human herpesvirus-8 DNA sequences and overexpression of human interleukin 6 and cyclin D1 has been reported in some cases (1). In our case, loosening of the eyelids, because of the leprosy, may have resulted in chronic irritation.

The lungs are the most frequent site of IMTs (6). Extrapulmonary IMTs have a recurrence rate of approximately 25% related to location, resectability, and multinodularity. In some cases (< 5%), IMTs are known to metastasize (1). Recurrence is very rare following the complete excision of a solitary lesion. However, the lesion is usually unencapsulated, and this causes difficulties in estimating the extent of excision during an operation (7). Recurrences are particularly common among multinodular intra-abdominal tumors and those in delicate anatomical locations, such as vital structures; this likely reflects the difficulty of complete surgical resections (6, 8-10). Possible therapies in this case include corticosteroids, immunosuppressive therapy with cyclosporine A, chemotherapy, laser therapy, and radiation therapy (6, 11, 12).

Steroid therapy seems to be effective postoperatively. Idrees at al. (12) reported that an IMT of the larynx recovered well after laser therapy followed by grafting combined with local steroid injections. In our case, the only feasible curative surgical approach was enucleation or exenteration. However, we achieved positive results with a combination of tumor resection and systemic corticosteroid therapy.

The diagnosis of an IMT, which is based on the clinical manifestation and imaging, is always of little specificity. Therefore, histopathological and immunohistochemical examinations are essential in the diagnosis of IMT findings. Most of the IMT diagnoses are difficult to establish before surgery because of their diversified clinical and radiologic manifestations (4). In our case, did not come to the diagnosis until we received the histopathological results after surgery.

Histological features in inflammatory myofibroblastic tumors do not correlate well with clinical behavior (13). Typically, it is a circumscribed but nonencapsulated lesion. Histopathologically, it is a mixture of different inflammatory cells and mesenchymal cells, which include plasmacytes, histiocytes, lymphocytes, and spindle cells (14). As a consequence of the myofibroblastic differentiation characteristics of IMTs, immunohistochemically, almost all tumors stain positive for vimentin. Reactivity for smooth muscle actin and muscle-specific actin varies from a focal to a diffuse pattern in the spindle cell cytoplasm, and desmin is identified in many cases. Focal cytokeratin immunoreactivity is seen in about one third of all cases. Myogenin, myoglobin, and S100 protein are negative (1).

Chromosomal translocations leading to an activation of the ALK tyrosine kinase (and overexpression of the ALK protein) can be detected in approximately 50% of IMTs, but are uncommon in older patients (13).

IMTs of the head and neck are rare, but can occur in the orbit, larynx, and maxillary sinus. Coffin *et al*. (14) in their review of 84 extra-pulmonary IMTs, noted three head and neck cases (3.6%) including one orbital IMT. Sa *et al*. (3) reported a case involving a 10-year-old boy with a 2-week history of diplopia and subconjunctival mass. The patient underwent surgical debulking followed by systemic corticosteroids. Sa reported progression with invasion of the superior cornea. The patient later received radiation therapy after his parents refused a subtotal orbital exenteration. The 2-year follow-up examination showed no evidence of recurrence.

Favini at al. (2) reported a 12-year-old female with a left conjunctival nodular mass causing diplopia and a decline in visual acuity. The intimate relationship of the mass with the sclera and muscles did not allow for a complete conservative surgical resection. Therefore, they administered low-dose chemotherapy. The 2-year follow-up examination showed no radiological evidence of any tumor progression or recurrence. In the present case, as Favini *et al.* did, we were able to partially scrape with a crescent knife because of the infiltration of cornea.

In conclusion, it should be kept in mind that inflammatory myofibroblastic tumors localized in the anterior orbit might be confused with a malignancy. IMTs can be treated with partial tumor resections followed by systemic steroid therapy rather than using a radical surgical approach. The optimal management of this benign tumor should be decided individually for each patient.

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Figure Legends

Figure 1A, B: The mass fills the palpebral fissure and covers the corneal surface irregularly with purulent secretions on its surface.

Figure 2: Coronal contrast enhanced CT demonstrates contrast enhanced mass(arrows) anterior to the right bulbus covering conjunctiva.

Figure 3: The mass was excised along with healthy conjunctival tissue. The mass was partially scraped by a cresent knife because of the involvement of cornea.

Figure 4: Photomicrograph of H and E, ×200 showing features of inflammatory myofibroblastic tumor. Mixed inflammatory cells dominated by lymphocytes and plasmocytes, and the proliferation of spindle cells with a swirly growth pattern.

Figure 5 Photomicrograph of immunohistochemical staining, ×200 showing features of inflammatory myofibroblastic tumor. Spindle cells mixed with inflammatory cells staining with smooth muscle actin (SMA) (Negative staining with Desmin, S100 and CD34).

Figure 6A, B: The superior half of cornea was clear, vascularized corneal leukoma in the inferior half of cornea (probably due to lagophthalmos) and mature cataract was observed at the 1-month postoperative

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