Report of a Conjunctival Myxoma Case and Review of the Literature

Myxoma is a benign soft tissue tumor that presumably derives from primitive mesenchyme. It is characterized histopathologically by abundant mucoid material, a loose meshwork of reticulin fibers, and relatively small numbers of spindle- and stellate-shaped cells. The stromal matrix is rich in glucosaminoglycans and hyaluronic acid, and it is sparse in collagen and vascular structures. Myxoma can appear as localized disease or as a component of Carney complex, which includes cutaneous and cardiac myxomas, multiple pigmented lesions, and endocrine overactivity, leading to significant morbidity and mortality. Myxoma is the most common primary cardiac tumor. This tumor can arise in the skeletal muscle, bone, skin, genitourinary system, gastrointestinal tract, and nasal sinuses. It can rarely involve ocular structures and has been recognized in the eyelid, orbit, and conjunctiva.

In a review of 2455 conjunctival lesions submitted to an ophthalmic pathology laboratory, only 4 patients (0.002%) were found to have conjunctival myxoma. In a clinical review of 1643 patients with conjunctival lesions, myxoma was found in 1 case (<0.001%). Because of its rarity, conjunctival myxoma can simulate other conjunctival tumors, such as amelanotic nevus, amelanotic melanoma, squamous cell carcinoma, lipoma, and cyst. We describe a 31-year-old African American man who had a yellow, translucent subconjunctival mass that proved to be a conjunctival myxoma on histopathological examination.

Report of a Case. A 31-year-old African American man had a 2-year history of conjunctival swelling in his left eye. The lesion was diagnosed as a conjunctival cyst, and drainage was attempted. Eighteen months later, the swelling worsened and a solid conjunctival mass was noted, prompting referral to the Oncology Service, Wills Eye Hospital, Philadelphia, Pa. The patient had no history of ocular trauma and was otherwise healthy.

The visual acuity was 20/20 OU. The right eye was normal. External examination of the left eye disclosed a well-circumscribed, yellow-pink, translucent mass on the nasal bulbar conjunctiva (Figure 1). The mass had 2 parts, including a solid basal part measuring 12 × 11 × 6 mm and a cystic apical part measuring 5 × 5 × 2 mm. The clinical differential diagnosis included amelanotic nevus, amelanotic melanoma, myxoma, fibrous histiocytoma, and lipoma. The lesion was excised using the previously described no-touch technique.

Histopathological examination revealed a lesion composed of loose, myxoid areas that alternated with areas of spindle and stellate cells, which was consistent with benign conjunctival myxoma (Figure 2). Both cellular groups had moderately large hyperchromatic and slightly pleomorphic nuclei and were embedded in a loose mucoid stroma. The stroma contained reticulin fibers, sparse small blood vessels, and collagen fibers (Figure 2). Some of the tumor cells had clear, circular, intranuclear inclusions, and some contained clear cytoplasmic vacuoles. Numerous mast cells were scattered throughout the myxoid stroma (Figure 3).

Systemic evaluations of the patient for cardiac, endocrine, and
cutaneous abnormalities including echocardiography and levels of thyroid-stimulating hormone, growth hormone, and adrenocorticotropic hormone were negative. After 11 years of follow-up, the patient remained healthy without recurrence of the conjunctival mass or evidence of a systemic abnormality.

Comment. Myxoma rarely occurs in the conjunctiva. To our knowledge, only 22 well-documented cases of conjunctival myxoma have been published in the English literature (Table). The tumor usually occurs in adults and is uncommon in children. The mean age of the 22 published cases was 48 years (median age, 50 years; age range, 18-80 years), and 2 patients (9%) were younger than 20 years. Both sexes were affected equally. Similarly, a review of 58 patients who all had soft tissue myxomas revealed a mean age of 55 years, with only 1 patient (2%) younger than 20 years.

Conjunctival myxoma is typically seen as a well-circumscribed, yellow-pink, translucent, cystic and/or solid mass. Of the 22 reported cases, the conjunctival mass was described as well-circumscribed in 20 cases (91%) and diffuse in 2 cases (9%). The color of the conjunctival mass, when mentioned, was yellow in 9 cases (50%), pink in 8 cases (44%), and white in 1 case (6%). The conjunctival lesion was translucent in 14 cases (64%) and solid in 8 cases (36%).

Conjunctival myxoma has characteristic histopathological features. It consists of scattered, relatively small numbers of stellate- and spindle-shaped cells disposed in a mucinous matrix with delicate reticulin fibers, sparse vascular structures, and mature collagen fibers. The mucinous matrix is composed predominantly of hyaluronic acid, with a lesser amount of chondroitin sulfate. Ultrastructurally, myxoma cells have intracytoplasmic vacuoles and cytoplasmic filaments arranged in loose, wavy bundles. These vacuoles are consistent with dilated rough endoplasmic reticulum. It seems that the mucoid matrix and extracellular fibrils are produced by myxoma cells. There are also intranuclear vacuoles that are consistent with invaginations of markedly folded nuclear membrane. Immunohistochemical studies showed myxoma cells were immunoreactive for vimentin and a smooth muscle actin but negative for S-100 protein, desmin, and myoglobin.

Conjunctival myxoma differs clinically and histopathologically from myxomas of extraocular soft tissues in some aspects. Unlike extraocular soft tissue myxomas, conjunctival myxoma often contains cyst-like cavities. The latter were noted in 9 of the 22 published cases. In our case, the patient was diagnosed as having a conjunctival cyst, and drainage was attempted before referral to our service. Additionally, on histopathological examination, mast cells can be observed in conjunctival myxoma.

The differential diagnosis of conjunctival myxoma includes nevus, amelanotic melanoma, fibrous histiocytoma, cyst, lymphangioma, myxoid neurofibroma, spindle cell lipoma, rhabdomyosarcoma, and myxoid liposarcoma. Unlike conjunctival amelanotic nevus and melanoma, myxoma does not have prominent intrinsic vascularity or pigmentation. Myxoid neurofibroma can have a similar clinical picture. It contains cells with spindle-shaped nuclei with thick, wavy bundles of collagen and can be associated with systemic neurofibromatosis. Both fibrous histiocytoma and myxoma stain positively with Alcian blue, which is abolished by pretreatment with hyaluronidase. Unlike fibrous histiocytoma, myxoma does not show vascularity or significant nuclear pleomorphism.

The preferred management of conjunctival myxoma is complete excisional biopsy. Of the 22 published cases with conjunctival myxoma, no recurrence or malignant transformation after excisional biopsy of the lesion was observed. In a review of 58 patients with myxomas of the soft tissue, only 2 (3%) had recurrence after 8 to 10 months following excision. These 2 patients underwent reexcision, and both were free of recurrent tumor after a mean of 5 years. Recurrence of myxoma can be related to inadequate excision, multicentricity of the tumor, the presence of genetic predisposition, or malignant transformation. In a review of 107 cardiac myxomas submitted to a pathology laboratory, atypical cells simulating malignancy were observed in only 3%.

Myxomas can be a component of Carney complex, Mazabraud syndrome (bone fibrous dysplasia and intramuscular myxoma), and McCune-Albright syndrome (café au lait spots and nodulocystic goiter). Carney complex is an autosomal dominant syndrome that includes 2 or more of the following conditions: multiple foci of mucocutaneous pigmentation (face, trunk, lips, conjunctiva), cardiac and extracardiac (skin, breast) myxomas, endocrine overactivity (including Cushing syndrome), pituitary adenoma, unusual testicular tumors, and psammomatous melanotic schwannoma. Patients with myxoma, mucocutaneous pigmentation, and/or a family history of Carney complex should undergo a systemic evaluation including echocardiography, computed tomography or magnetic resonance imaging of the body, testicular ultrasound, analysis of the thyroid-stimulating hormone level, adrenocorticotropic hormone level, and genetic analysis (PRKAR1A gene at chromosome 17q2). Ophthalmic manifestations of Carney complex include facial and eyelid lentigines, pigmentation on the caruncle or conjunctival semilunar fold, and eyelid myxoma. Kennedy et al described a patient with Carney complex who had a myxoma of the palpebral conjunctiva and an additional lesion on his eyelid margin. None of the 22 published cases of...
<table>
<thead>
<tr>
<th>Source</th>
<th>Age, y/ Sex</th>
<th>Systemic Disease</th>
<th>Eye</th>
<th>Symptoms</th>
<th>Duration of Symptoms, mo</th>
<th>Clinical Features</th>
<th>Color</th>
<th>Conjunctival Site</th>
<th>Conjunctival Location</th>
<th>Outcome</th>
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<tbody>
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<td>Ffooks (1962)&lt;sup&gt;8&lt;/sup&gt;</td>
<td>72/M†</td>
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</table>

Abbreviations: F, female; M, male; NA, not applicable; OD, right eye; OS, left eye.

*Cases were published in the English literature and were well documented. For all of the patients, the therapy was excisional biopsy and the histopathological examination result was myxoma.

†Patients were white.
conjunctival myxoma in the literature were associated with Mazabraud or McCune-Albright syndromes.8-23

In summary, conjunctival myxoma can appear as a well-circumscribed, translucent, yellow-pink conjunctival mass in middle-aged patients. Management is generally complete surgical resection.

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Corneal Decompensation Following Bleb Revision With Absolute Alcohol: Clinical Pathological Correlation

Topical alcohol has been commonly used to facilitate removal of corneal epithelium.1 More recently, a technique for bleb revision has been described in which absolute alcohol is used topically to remove the epithelium of a previously existing bleb.2 Unlike other techniques in which the prior bleb is excised, this technique has the advantage of leaving the prior bleb in place while resurfacing it, thus minimizing the potential for contraction that may otherwise occur during formation of an entirely new bleb.

We report a case of a patient who suffered acute corneal decompensation following surgical bleb revision in which a large amount of absolute alcohol had been used to de-epithelialize a leaking bleb. Results of an experimental rabbit model are presented to demonstrate the possibility that absolute alcohol could compromise the corneal toxicity in this case.

Report of a Case. A 79-year-old man had a 3½-month history of a Seidel-positive bleb leak and declining visual acuity. Two and one-half years earlier, he had undergone combined phacoemulsification and trabeculectomy with 0.5 mg/mL mitomycin C for 4 minutes under retrobulbar anesthesia. On examination, his visual acuity had decreased from a baseline of 20/25 to 20/80. Application intraocular pressure measured ranged between 5 and 10 mm Hg on serial examinations. Slitlamp examination revealed a large, avascular bleb with 2 epithelial defects that were briskly positive on Seidel test. The cornea showed folds in the Descemet membrane. No corneal guttata were seen. Examinations of the cornea prior to the bleb leak showed no signs of corneal guttata, early edema, or other disease. There was no sign of guttata or other corneal disease in the fellow eye. The disc showed a 0.8 cup. The macula was flat.

The patient underwent revision of trabeculectomy using a conjunctival advancement flap under topical and subconjunctival but not intraocular anesthesia. An incision was made around the bleb and the surrounding tissues were freed from the underlying sclera. The bleb was de-epithelialized using 100% ethanol (absolute alcohol) soaked on a cotton swab. We used a cotton swab rather than the cellulose spear originally described2 because it had been observed that the anhydrous etha- nol did not expand the spear, and thus little alcohol was applied with it. More alcohol could be applied with the cotton that did absorb the alcohol. Residual epithelium was removed using a sharp blade. The conjunctival advancement flap was then sutured into place at the limbus with a combination of polyglactin sutures at the ends of the flap and a mattress-style nylon suture at the midpoint of the flap. A circumferential relaxing incision through conjunctiva was cut as posteriorly as possible to relieve tension on the flap.

One week postoperatively, the patient's visual acuity measured


