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A Rare Case of Conjunctival Myxoma Initially Misdiagnosed as a Conjunctival Inclusion Cyst

Dear Editor,

Myxoma, derived from the primitive mesenchyme, is a benign and slow-growing soft tissue tumor that is considered to be one of the most common tumors of the heart. Myxomas can also involve the skeletal muscle, genitourinary or gastrointestinal system, and nasal sinuses. Ocular myxomas, however, are extremely rare and can masquerade as lymphoma, ocular surface squamous neoplasia, or amelanotic melanoma. Myxomas can occur as components of syndromic associations such as the Carney complex, in which case warrants systemic evaluation as ocular signs may precede systemic findings. Here we report a rare case of conjunctival myxoma that was initially misdiagnosed at a local clinic as a conjunctival cyst due to its lack of typical features previously described in literature.

A 45-year-old female first presented to clinic complaining of a painless, yellow, cyst-like lesion of the left eye of 1-year duration. She was scheduled for needle drainage at a local clinic under the clinical diagnosis of a conjunctival cyst. She presented to the outpatient department at Severance Eye Hospital for a second opinion. Her past medical and ocular history was unremarkable. Upon ophthalmologic evaluation, her visual acuity was 20 / 20 in the both eyes. Her intraocular pressure was normal. Slit-lamp examination revealed a well-circumscribed, nontender, yellow and translucent mass measuring 3.0 mm × 1.8 mm in the temporal bulbar conjunctiva of the left eye (Fig. 1A). The overlying conjunctiva was not congested. The lesion was completely excised under local anesthesia (Fig. 1B). Histopathological examination revealed spindle and mast cells in hypocellular myxoid stroma. The cells showed intranuclear vacuoles and multinucleation. Immunohistochemical staining tested positive for CD34 but negative for S-100 (Fig. 1C, 1D). Ki67 was less than 1%. Accordingly, conjunctival myxoma was diagnosed. Postoperatively, the patient was treated with 1.5% levofloxacin and fluorometholone eye drops four times a day for one month. She was referred to the department of internal medicine for extensive cardiac and endocrine workup. After 6 months, the patient showed no recurrence, metastasis, or any evidence of systemic abnormalities.

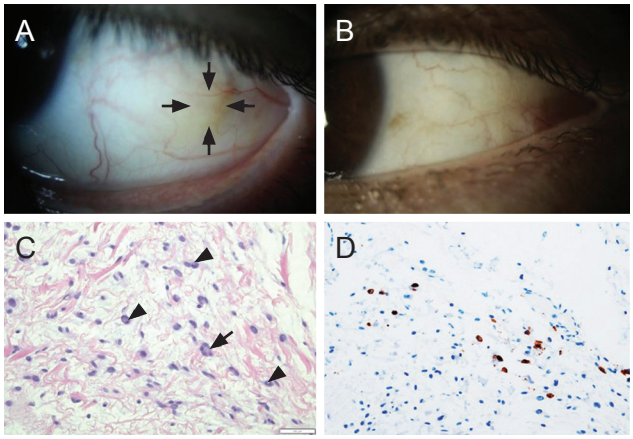

Fig. 1. Clinical and microscopic presentation of conjunctival myxoma. (A) Slit-lamp examination shows a translucent, yellow, cyst-like lesion (margins indicated with arrows) of the left temporal conjunctiva. (B) Slit-lamp photography at postoperative 6-month follow-up. (C) Conjunctival myxoma showing spindle and fusiform cells distributed throughout a mucinous matrix with cells having intranuclear vacuoles (arrowheads) and multinucleation (arrow) (H&E, ×200). (D) C-kit (CD117) immunohistochemical staining reveals scattered mast cells in stroma (×200). Written informed consent was obtained from the patient.

Conjunctival myxoma is an extremely rare benign tumor with an incidence of 0.001% to 0.002% among conjunctival lesions [1]. To our knowledge, 30 cases have been reported since the original published report in 1913 [2]. Conjunctival myxomas usually present as a slow-growing, nontender and well-circumscribed pink lesion. In most reported cases, the overlying conjunctiva was injected with prominent engorged vessels and sometimes exhibited subconjunctival hemorrhage [3]. The differential diagnosis includes amelanotic melanoma, lymphoma, lymphangioma, myxoid neurofibroma, or rhabdomyosarcoma. Under biopsy, the characteristic histopathological features of conjunctival myxoma are spindle and fusiform cells distributed through-

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out a hypocellular myxoid stroma. Conjunctival myxomas are treated by complete excision [4]. Although most reported cases of conjunctival myxoma were localized occurrences, some cases presented as a component of Carney complex, an autosomal dominant syndrome that can present with multiple endocrine tumors and cardiac myxomas. A thorough systemic evaluation is important, including cardiac echocardiography to rule out cardiac myxoma, and hormone tests to rule out endocrine abnormalities. Ophthalmic manifestations may precede vascular embolic events which account for over 10% of sudden death in patients with Carney complex. Conjunctival myxoma may also be associated with the Zollinger-Ellison syndrome [5]. Whether conjunctival myxoma later induces systemic involvement or is a sequelae of systemic disease, is an issue yet to be elucidated. As such, long-term follow-up of systemic evaluation is imperative, as systemic involvement could arise later on. However, recurrence or malignant transformations are known to be extremely rare.

Most cases of conjunctival myxoma described in literature show an erythematous mass with prominent congested vessels. The conjunctival myxoma in this case had an atypical clinical appearance from cases previously described in literature. In this case, a yellow, cyst-like lesion lacking congestion made it almost impossible to differentiate from a conjunctival cyst. Had the patient underwent drainage at a local clinic, prompt systemic evaluation and investigation may have been overlooked.

For indeterminate conjunctival lesions, lesions with engorged vessels, and lesions with history of refractory topical medication or failed aspiration and drainage, most ophthalmologists consider further intervention for accurate diagnosis. As with our case, excisional biopsy should be considered even in some patients who present with conjunctival cysts to assess for myxoma which may warrant a prompt and thorough systemic evaluation. It is our role as ophthalmologists to be aware of the various clinical presentations of conjunctival myxomas for accurate diagnosis.

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Conflict of Interest

No potential conflict of interest relevant to this article was reported.

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