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CASE REPORT

Orbitoconjunctival Myxoma: A Case Report and a Brief Literature Review

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Abstract: A 50-year-old male presented with painless swelling in the superonasal sector of his left eye that had appeared a year before and was unresponsive to topical steroids. He had a history of trauma to the affected eye two years prior to presentation. Examination revealed a 1 cm cystic neoformation in the episcleral subconjunctival area near the superonasal orbital margin, with normal ocular motility and no diplopia. Computed tomography (CT) showed a cystic formation between the anterior superomedial orbit and superolateral surface of the medial rectus muscle, without intraconal space involvement. Magnetic resonance imaging (MRI) indicated that the formation was hyperintense on T2 and hypointense on T1, with avid post-contrast enhancement and no extension into the surrounding structures. The neoformation was surgically removed by opening the conjunctiva, isolating the medial rectus muscle, and detaching the mass from uninfiltrated tissues. Histological examination, initially suspected to be a post-traumatic inclusion cyst, revealed an orbitoconjunctival atypical myxoma without signs of malignancy. Complete excision was deemed definitive therapy. Postoperative systemic and cardiac evaluation excluded additional myxomatous lesions. Six months after surgery, the patient remained recurrence-free.

Keywords: orbitoconjunctival myxoma, orbit, conjunctiva, Carney complex, case report

Introduction

Myxomas are benign, locally infiltrative tumours that originate from the mesenchymal cells. These can affect any age group and usually develop throughout the body affecting primary skeletal muscles, skin, heart and genitourinary system.[1] Soft tissue myxomas are categorized into five types: intramuscular, cutaneous, juxta-articular, nerve sheath, and aggressive angiomyxoma.[1] Rarely myxomas involve the head-neck district.[2] Several reports[3–14] have described ocular tissues involvement, usually occurring in orbit, conjunctiva, cornea and eyelid. They most commonly develop in middle-aged adults with no clear gender predilection.[3–14]

Ocular myxomas can arise either as a single disease or as a part of a systemic life-threatening condition called the Carney complex, which is characterized by cutaneous and cardiac myxomas, pigmented lesions, and endocrine overactivity.[15] Given the risk of fatal cardiac complications, patients with ocular myxomas should undergo systematic screening including dermatological examination for characteristic pigmented lentigines, echocardiography to detect potentially lethal cardiac myxomas and endocrine evaluation for Cushing syndrome and acromegaly. Early cardiac detection is paramount as myxomas represent the leading cause of mortality in Carney complex.[15]

In ocular region myxomas consist of stellate and spindle-shaped cells that merge in a myxoid matrix rich in glycosaminoglycans with few vascular structures.[3,5,10,11] Immunohistochemically, tumour cells can stain positively for

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vimentin and CD34.[3,6] Herein we describe a singular case of a 50-year-old Caucasian man presenting with a progressive painless conjunctival swelling in the left eye that was diagnosed as a myxoma involving either orbit and conjunctiva.

Case Presentation

A 50-year-old male patient presented to our center complaining of the appearance of a painless swelling in the superonasal sector of his left eye, which had slowly grown in the previous year. The lesion did not regress with the previously prescribed topical corticosteroid therapy. The patient reported a history of blunt trauma to the affected eye two years earlier. His medical history was otherwise unremarkable. Visual acuity was 20/20 in both eyes, and intraocular pressure was normal. Physical examination revealed the presence of a soft pink episcleral-subconjunctival cystic neoformation of approximately 1 cm that was introflected within the superonasal orbital margin, reaching near the corneal limbus (Figure 1A and B).

No alterations in ocular motility were observed and the patient did not complain of diplopia. The fundus exam was unremarkable. The first diagnostic hypothesis was a post-traumatic epidermoid inclusion cyst. In order to confirm or reject this hypothesis, we performed further investigation. Computed tomography (CT) showed a cystic formation located between the anterior superomedial portion of the bony orbit and the superolateral surface of the medial rectus muscle, which did not affect the intraconal space, and its posterior limit did not cross the border between the first and second Benedict spaces (Figure 2).[16]

Magnetic resonance imaging (MRI) showed that this mass was hypointense on T1 (Figure 3A) and hyperintense on T2 (Figure 3B), and contrast-enhanced imaging (Figure 3C) showed avid enhancement, without extension to surrounding structures. The contrast enhancement pattern was not consistent with the initial diagnostic hypothesis (post-traumatic inclusion cyst); therefore, given its location in the anterior extraconal compartment, a transconjunctival surgery was chosen as the most direct and minimally invasive option. The neoformation was removed after opening the conjunctiva, isolating the medial rectus muscle, and detaching it from the surrounding tissues. This approach ensured preservation of adjacent structures and allowed for complete excision with clear margins to minimize recurrence risk. The piece was sent for histological examination with the clinical question of a post-traumatic inclusion cyst. Histological examination showed a nodular myxomatous neoformation with maximum dimensions of 15×11×6 mm, well delimited, non-encapsulated, pauci-cellular, including spindle cells, globular (CD34+ and Vimentin+), with voluminous, hyperchromic, and sometimes binucleate nuclei (Figure 4A and B). Immunohistochemical tests showed that these cells were positive for

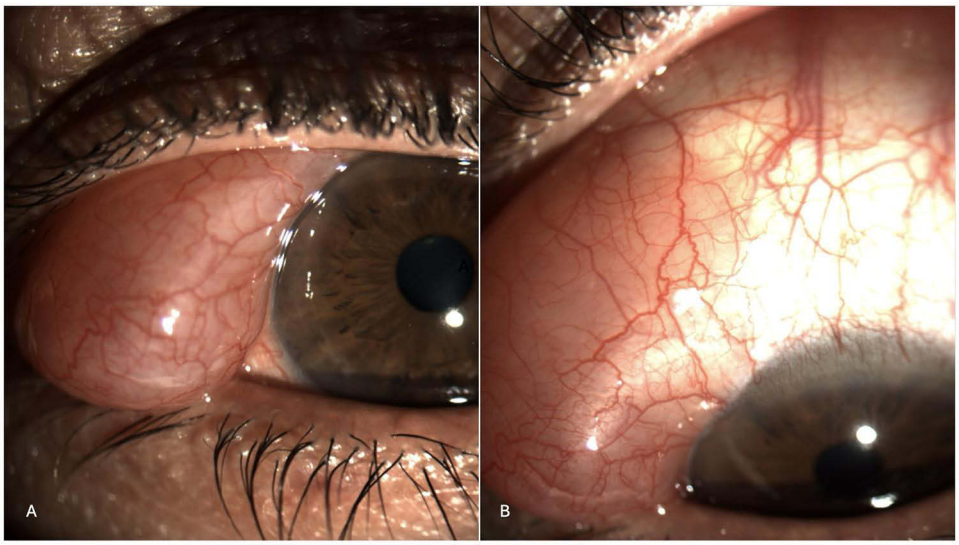

Figure 1 Slit lamp examination of the lesion showing the presence of a soft pink episcleral subconjunctival cystic neoformation of about 1 cm reaching the superonasal quadrant of the corneal limbus in primary gaze position (**A**). The mass was introflected within the superonasal orbital margin, as shown in down gaze position (**B**).

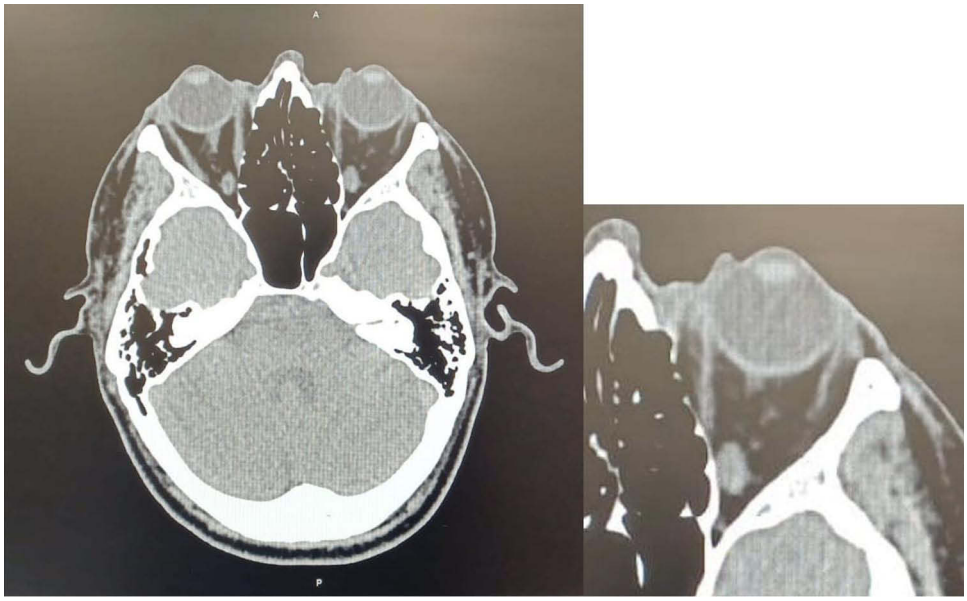

Figure 2 The CT showed the presence of a cystic formation located between the anterior superomedial portion of the bony orbit and the superolateral surface of the medial rectus muscle.

CD34 and vimentin (Figure 4C and D). No evidence of densely cellular areas or a preponderance of curvilinear vessels, investigate also with CD31 antibody, were found in the sections examined.

The cell proliferation index Ki67 (30–9 clone, Ventana Roche) was low (1%). Immunoreactivities for Desmin (DE-R-11, Ventana Roche), Smooth Muscle actin (1A4, Sigma Aldrich), S100 estrogen receptor (SP1, Ventana Roche), progesterone receptor (1E2, Ventana Roche), MUC4 (8G7 clone, CellMarque), CD 68 (KPI, Roche Ventana), ALK-1 (ALK01, Roche Ventana), and β-catenin (14, CellMarque) were negative. Given the absence of pronounced cytologic atypia, high proliferative activity, tumor necrosis, or rich vascular network, the lesion was not considered at risk of malignant transformation.[5] A diagnosis of atypical orbitoconjunctival myxoma was established and complete excision was considered definitive therapy. The patient subsequently underwent systemic and cardiac evaluations, including echocardiography, to exclude additional myxomatous lesions.

At six months postoperatively, there were no signs of recurrence, and systemic screening did not reveal any other lesions.

Written informed consent was provided by the patient to have the case details and any accompanying images published.

Discussion

In the present report, an unusual presentation of an atypical myxoma simultaneously involving both the orbit and conjunctiva was described. The patient presented with a slowly progressive, painless subconjunctival swelling, and a history of prior ocular trauma, with imaging features initially suggestive of a post-traumatic inclusion cyst. However, histopathological examination ultimately revealed a diagnosis of atypical myxoma with simultaneous orbitoconjunctival presentation which represents an exceptional and clinically relevant finding.

Typically, slow-growing, painless masses such as myxomas involving orbital soft tissue or adjacent bone may enlarge considerably before diagnosis.[13] Histogenetically, myxomas are tumours of primitive mesenchymal tissue, where cells with pyknotic nuclei produce a myxoid stroma that is poor in collagen and rich in hyaluronic acid.[6,13] Imaging is not decisive for diagnosis but is useful for early evaluation. Indeed, CT usually displays a well-defined soft tissue mass, which on MRI appears homogenous with low or intermediate signal intensity on T1-weighted images and high signal intensity on T2-weighted images.[17] Definite diagnosis depends on histological examination, which confirms the benign nature of the lesion and allows differentiation of myxomas from other diseases presenting with myxoid degeneration.[3,5,13]

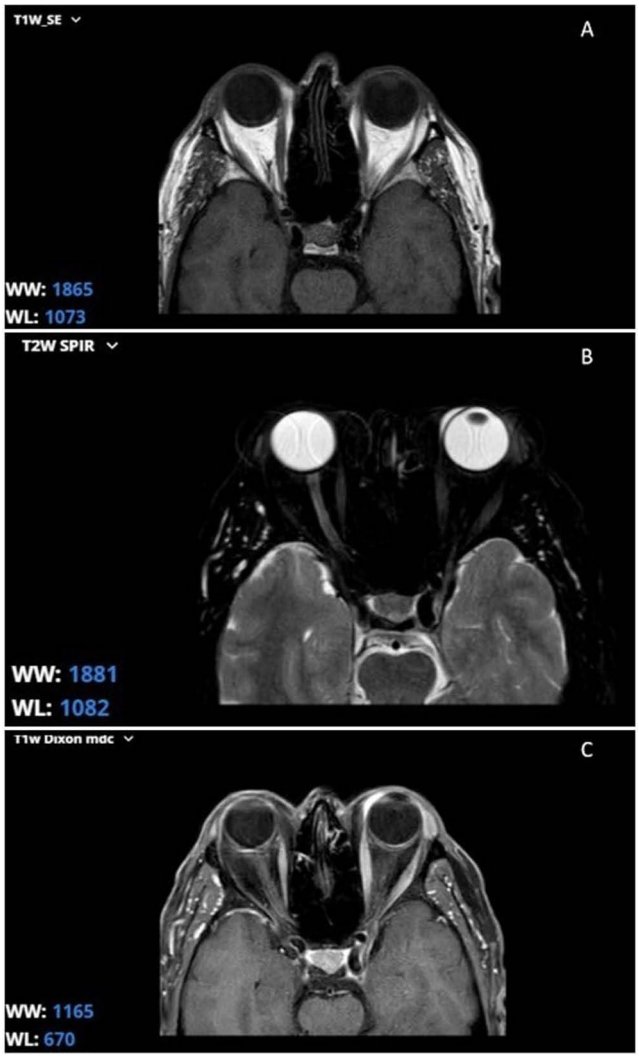

Figure 3 MRI showed a lesion that was hypointense on T1 (**A**) hyperintense on T2 (**B**) and the contrast-enhanced imaging (**C**) showed a avid enhancement, without extension to surrounding structures.

Myxomas can affect different structures of the eye, including the cornea, conjunctiva, eyelid, and orbit. To date, 31 cases of corneal myxomas have been reported. Corneal myxoma is described as a primary condition or reactive process secondary to traumatic events or previous corneal diseases. It is usually a circumscribed whitish gelatinous mass that commonly extends under the corneal epithelium.[8–10] Furthermore, 70 cases of conjunctival myxomas have been described, with only one located in the tarsal conjunctiva involving the eyelid margin.[18] The morphological appearance is defined as a yellow-pink, smooth tumour generally located in the temporal bulbar conjunctiva, which may resemble a conjunctival cyst.[3,4] Eyelid myxoma is rare and typically appears as a small, translucent, nodular mass that often manifests as painless swelling of the lid and tends to recur after surgical excision. Only 6 cases of eyelid myxoma[11,12,14,19] have been described in the literature. To date, 29 cases of orbital myxoma have been reported.[5,6,13] Orbital myxomas manifest themselves as slow-expansive masses that may remain asymptomatic until causing proptosis.[5,6,13] According to the literature, the treatment of choice is represented by excisional biopsy,[20] with a low number of recurrences usually occurring after incomplete resection and no metastasis. However, in the presence of small and asymptomatic lesions, periodic observation may be considered.[7,9,13] Owing to the low mitotic activity, chemotherapy and radiotherapy are ineffective for these tumors, although a successful medical response to cyclophosphamide has been described.[21,22] Systemic evaluation is useful to rule out the presence of a syndromic disorder; in particular, echocardiographic examination is mandatory to exclude the presence of cardiac myxomas.[15]

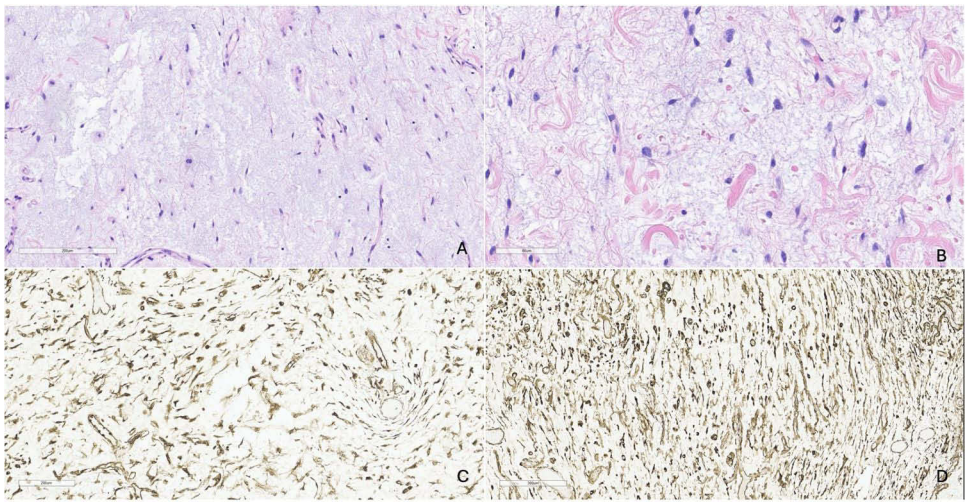

Figure 4 Histological and immunohistochemical appearance of the atypical orbito-conjunctival myxoma. Hematoxylin and eosin stains showed a non-encapsulated paucicellular myxoid lesion with fine vascular network including spindle/ stellate and rare globous cells with voluminous, hyperchromic nuclei, sometimes binucleated (**A** and **B**). Vimentin immunostain positive (Ventana, clone V9) (**C**) CD 34 immunostain positive (Ventana, clone QBEnd/10) (**D**). Scale bars: A = 200 µm; B = 60 µm; C = 200 µm; D = 300 µm.

In this case, the patient's medical history suggested the possibility of a post-traumatic epidermoid inclusion cyst. Epidermoid tumours in the orbit are extremely rare. They can be subdivided into primary and secondary types, with the latter usually being caused by traumatic injuries.[22–24] However, as these tumours usually appear as encapsulated masses with internal contents consisting of keratin and ectodermal elements in ectopic positions,[22–24] histological examination excluded this diagnosis. Furthermore, considering the histopathological and immunohistochemical differential diagnoses, the following myxomatous entities were considered: low-grade fibromyxoid sarcoma, inflammatory myofibroblastic sarcoma, atypical myxoma, and low-grade myxofibrosarcoma.

MUC4 negativity excluded low-grade fibromyxoid sarcomas. The failure to find Reed-Sternberg-like cells (similar to virocytes) and the absence of an inflammatory exudate allowed the exclusion of inflammatory myofibroblastic sarcoma.

Since positivity for CD34 and vimentin, together with poor cellularity, are characteristics of both atypical myxoma and low-grade myxomafibrosarcoma (both with risk of local recurrence), the latter was excluded following macroscopic evaluation of the lesion with its nodular appearance and specific tumor boundaries.

Conjunctival stromal tumor (COST), a rare benign tumor arising from mesenchymal cells, was also considered in the differential diagnosis. Indeed, COST, a slow-growing yellow-white tumor, can appear as a cystic or nodular lesion.[25]

Lam et al[26] reported that spindle-shaped cells' immunoreactivity for CD34 supports COST diagnosis rather than conjunctival myxoma. However, Alvarado and colleagues[27] confirmed immunoreactivity for CD34 in published cases of conjunctival myxoma. Qin and coauthors[28] proposed the term "conjunctival myxoid stromal tumor" as a new entity, suggesting the need for further studies to delineate the differences between these entities. Typically, myxomas are localized to a single ocular region, making this case noteworthy for its simultaneous involvement of the orbit and conjunctiva. The dual involvement of these distinct ocular structures presents a rare diagnostic challenge and highlights the importance of comprehensive imaging and histological examination for accurate identification and management. Histologically, the features of this myxoma are similar to those of other myxomatous lesions present in different eye tissues, suggesting that management should prioritize the lesion itself rather than its specific location. Successful surgical excision without evidence of malignancy serves as a definitive therapy, consistent with the existing literature on the primary treatment approach for myxomas.[23] Additionally, the absence of myxomatous lesions upon systemic screening reinforces the localized nature of the condition. Nonetheless, regular follow-up is recommended, with clinical examination every 6–12 months for the first years to monitor for local recurrence, along with screening for Carney complex manifestations including annual echocardiography.

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Some limitations of the present report must be acknowledged. Firstly, as a single patient observation, its generalizability is inherently limited, and broader conclusions regarding management or prognosis should be drawn with caution. Secondly, the diagnosis relied heavily on histopathological and immunohistochemical analysis to differentiate it from other myxoid lesions, a process that may not be routinely available in all clinical settings and highlights the diagnostic challenge these lesions present. Thirdly, the relatively short postoperative follow-up period of six months does not allow definitive assessment of long-term recurrence risk, particularly in atypical variants that may exhibit locally aggressive behaviour.

Conclusion

In conclusion, this case represents a rare instance of an orbitoconjunctival atypical myxoma, highlighting the diagnostic complexities of myxoid ocular lesions and the value of integrating clinical, imaging, and histopathological findings. Complete surgical excision remains the cornerstone of treatment, with systemic screening essential to exclude associated syndromic conditions. This case expands the spectrum of ocular myxomas and underscores the importance of maintaining a broad differential diagnosis when evaluating atypical conjunctival or orbital masses. Future multicenter studies collecting data on similar rare cases would be valuable to better understand the clinical behaviour, optimal management, and long-term outcomes of ocular myxomas.

Institutional Approval

Institutional review board (IRB) approval was not required for the publication of the case details.

Informed Consent Statement

Informed consent was obtained from the patient.

Patient Consent

Written informed consent was provided by the patient to have the case details and any accompanying images published.

Author Contributions

All authors made a significant contribution to the work reported, whether that is in the conception, study design, execution, acquisition of data, analysis and interpretation, or in all these areas; took part in drafting, revising or critically reviewing the article; gave final approval of the version to be published; have agreed on the journal to which the article has been submitted; and agree to be accountable for all aspects of the work.

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Disclosure

The authors declare no conflicts of interest.

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