Case Series

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Conjunctival Myxoma: Series of Three Cases of a Rare Ocular Tumour

Pathology Section

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ABSTRACT

Conjunctival myxomas are very rare benign tumours of mesenchymal origin which pose a diagnostic challenge clinically and radiologically. They usually present as asymptomatic slowly growing cystic lesions involving the bulbar conjunctiva. Two cases were with usual presentation while one was with unusual presentation. Two cases of conjunctival myxoma (59-year-old female patient and 65-year-old male patient) involving the bulbar conjunctiva and one (56-year-old female patient) in the palpebral conjunctiva are reported. Anterior segment examination and fundoscopy were within normal limits. A clinical diagnosis of conjunctival cyst, pinguecula and pyogenic granuloma was made in the three cases, respectively. Excision was done in all the cases and a diagnosis of myxoma was made based on the characteristic histological and immunohistochemical features. No systemic involvement was seen. The authors hereby report the present cases because of their rarity and to emphasise that, histopathology is required to distinguish it from other conjunctival lesions.

Keywords: Bulbar, Cyst, Palpebral, Pinguecula, Pyogenic granuloma

INTRODUCTION

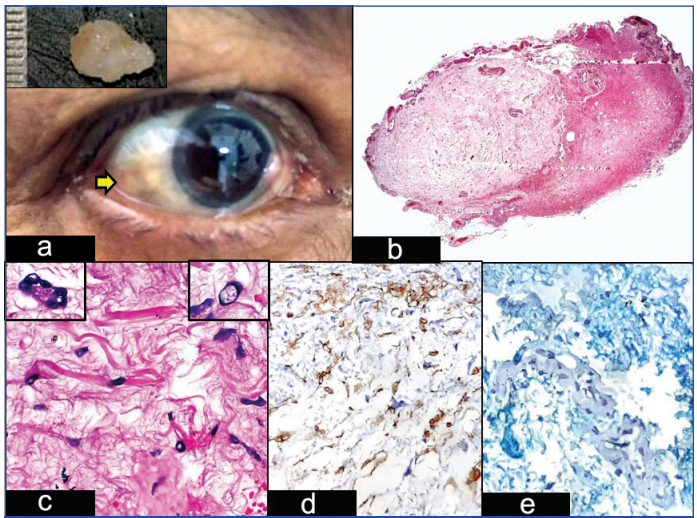
Myxomas are benign connective tissue neoplasms most commonly seen in the heart. They can rarely involve the ocular structures like conjunctiva, eyelid, cornea and orbit [1]. Conjunctival myxomas classically present as slow growing, painless, well circumscribed yellowish to pink cystic lesions [2,3]. It is important to differentiate them from other clinical mimics like conjunctival cyst, lymphangioma, myxoid neurofibroma amelanotic naevus, amelanotic melanoma and myxoid liposarcoma. Histopathology is needed to confirm their diagnosis. Microscopically, the hypocellular tumour contains spindle and stellate cells in a mucoid stroma. They are vimentin positive and S100 negative on Immunohistochemistry (IHC). They can have a localised presentation or be a part of Carneys syndrome which comprises of cardiac and extra-cardiac myxomas, spotty mucocutaneous pigmentation and endocrine overactivity [3]. Hence cardiac involvement has to be ruled out in all cases of myxoma for prompt treatment. We present three cases of conjuncival myxomas involving the bulbar and palpebral conjunctiva.

CASE SERIES

Case 1

A 59-year-old female patient presented to the Ophthalmology Department with complains of a gradually increasing swelling in the right eye that was present for six months. On examination, a well-defined yellowish-white cystic mass was seen in the temporal aspect of the right bulbar conjunctiva [Table/Fig-1a]. Routine investigations, visual acuity, intraocular pressure, anterior segment examination and fundoscopy were normal. Clinically, a diagnosis of conjunctival cyst was made and it was excised. On gross examination, a yellowish-white, soft, partly translucent nodule was seen measuring 5×4 mm {[Table/Fig-1a]-inset}. Cut section of the nodule was partly cystic and gelatinous in appearance. On microscopy, a fairly circumscribed partly cystic paucicellular tumour was seen [Table/Fig-1b], having abundant myxoid matrix with ropy collagen bundles, scattered spindle and stellate cells, presence of mast cells, and very few blood vessels [Table/Fig-1c]. Some cells showed mild pleomorphism with hyperchromatic nuclei {[Table/Fig-1c]-inset}. However, no mitotic activity was seen. Presence of intracytoplasmic and intranuclear vacuoles was seen {[Table/Fig-1c]-inset}.

Immunohistochemically, tumour cells showed diffuse expression of vimentin [Table/Fig-1d], focal expression of smooth muscle actin and were negative for S100. The myxoid matrix stained positively with alcian blue stain at pH 2.5. Ki 67 proliferation index was less than 1% [Table/Fig-1e]. A diagnosis of conjunctival myxoma was rendered based on the microscopic and IHC findings. Further systemic evaluation was advised to rule out presence of associated tumour syndromes. On follow-up, patient was asymptomatic and underwent two dimensional (2D) echo and Electrocardiography (ECG) to rule out cardiac involvement which were normal. There was no skin pigmentation or endocrine abnormality.

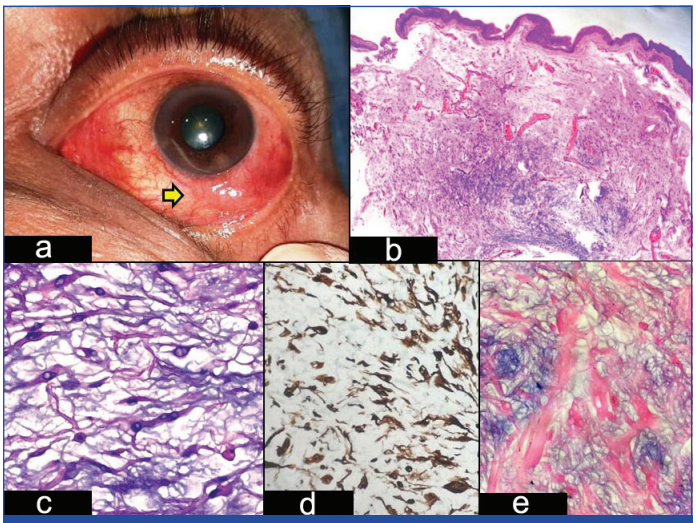


[Table/Fig-1]: a) Clinical picture with arrow showing a yellowish pink nodule on the temporal bulbar conjunctiva of the right eye. Inset shows specimen as a glistening yellowish-white nodule measuring 5×4 mm; b) Well circumscribed tumour nodule with paucicellular solid area and cystic area with haemorrhage (20x); c) Spindle and stellate cells with collagen fibres. Inset shows intracytoplasmic and intranuclear vacuoles (400x); d) Tumour cells exhibiting vimentin positivity (100x); e) Myxoid matrix highlighted by alcian blue stain (100x).

Case 2

A 65-year-old male patient presented to the Ophthalmology Department with complains of swelling in the left eye that was present for one year associated with mild redness since one week. On examination, a cystic mass was seen in the infero-temporal aspect of the left bulbar conjunctiva which was diagnosed as

pinguecula and it was excised [Table/Fig-2a]. Grossly, a partly cystic translucent nodule was seen measuring 5×4 mm with gelatinous cut surface. On microscopy, a subconjunctival paucicellular tumour was seen having myxoid matrix with scattered spindle cells, stellate cells, blood vessels and ropy collagen bundles. Some cells showed mild pleomorphism with intracytoplasmic and intranuclear vacuoles [Table/Fig-2b,c]. Immunohistochemically, tumour cells showed strong and diffuse positivity for vimentin [Table/Fig-2d] with focal expression for smooth muscle actin. S 100 was negative. The myxoid matrix was positive for alcian blue stain at pH 2.5 [Table/Fig-2e]. Ki 67 proliferation index was less than 1%. On follow-up, the patient was asymptomatic without skin pigmentation or endocrine dysfunction. Cardiac involvement was ruled out by 2D echocardiography which was normal.



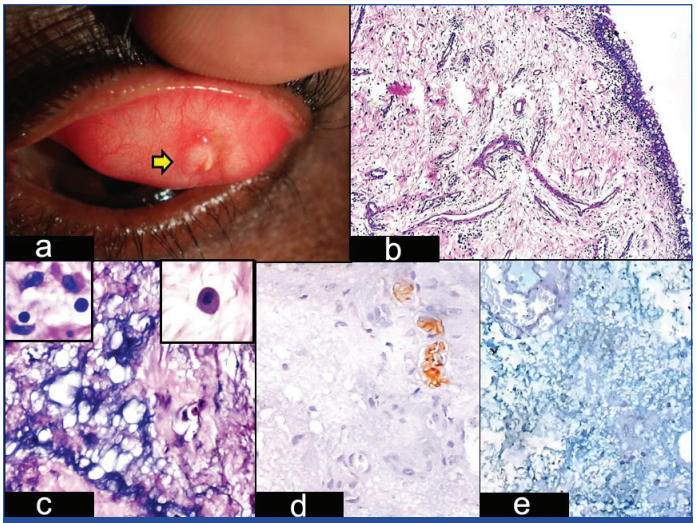
[Table/Fig-2]: a) Cystic swelling seen in the inferotemporal region of right bulbar conjunctiva; b) A subconjunctival paucicellular tumour composed of spindle cells (20x); c) Spindle and stellate cells with intracytoplasmic and intranuclear vacuoles against a myxoid background (400x); d) Tumour cells showing strong and diffuse vimentin positivity (100x); e) Alcian blue (counter stained with Haematoxylin) stains myxoid background (100x).

Case 3

A 56-year-old female patient presented to the Ophthalmology Department with itching and watering of left eye for four months and fullness in the upper eyelid since three months. On clinical examination a yellowish white, lesion measuring 5×5 mm over the upper palpebral conjunctiva was seen [Table/Fig-3a]. Clinical diagnosis was pyogenic granuloma and excision was done. Grossly, a yellowish-white, soft partly translucent nodule was seen measuring 5×5 mm. Cut section of the nodule had a gelatinous appearance. On histology a subconjunctival lesion composed of spindle and stellate cells in a myxoid background was seen [Table/Fig-3b,c]. Some cells showed pleomorphism with prominent intranuclear and intracytoplasmic vacuoles {[Table/Fig-3c]-inset}. Stroma showed ropy collagen, mast cells, lymphocytes and plasma cells and few blood vessels. On IHC, cells expressed vimentin and were negative for smooth muscle actin [Table/Fig-3d] and S100. Stroma was alcian blue positive at pH 2.5 [Table/Fig-3e]. Ki 67 proliferation index was less than 1%. No systemic involvement was present on follow-up as patient was asymptomatic and had normal 2D echocardiogram, absence of pigmentation and endocrine dysfunction.

DISCUSSION

Myxomas are benign mesenchymal tumours that commonly involve the heart, skin, bone, skeletal muscle, nasal sinuses, gastrointestinal and genitourinary system [1]. Ocular myxomas are rare and can involve the eyelid, ocular adnexa, conjunctiva or the cornea [2]. Conjunctival myxomas can occur as isolated tumours or rarely as a part of Carneys or Zollinger Ellison Syndrome with multi-systemic involvement [3-5]. The incidence of conjunctival myxomas was estimated to be 0.002% and 0.001% in two reviews of conjunctival



[Table/Fig-3]: a) Well circumscribed yellow nodule seen on the left palpebral conjunctiva; b) Paucicellular tumour composed of spindle cells along with a few blood vessels and covered by conjunctival epithelium (20x); c) Tumour showing abundant myxoid stroma with mast cells. Inset (left) shows a few atypical cells. Inset (right) shows mast cell (400x); d) Tumour cells showing vimentin positivity (100x); e) Alcian blue stains myxoid background (100x).

leisons studied by Grossniklaus HE et al., and Shields CL et al., respectively [6,7]. Though, it was first described by Magalif NI in 1913, on review of literature till date only about 50 cases of conjunctival myxomas have been reported [8]. The involvement of palpebral conjunctiva is very rare with only three cases reported uptil now [1-3]. Demicri H et al., reviewed 22 cases of conjunctival myxoma and described their clinical and histopathological features [9]. Xiong MJ and Dim DC reviewed and presented the findings of 42 cases with this lesion [3].

The tumour presented as a slow growing painless yellowish pink cystic or solid mass which commonly involved the bulbar conjunctiva. It involved the temporal region in 49% cases, nasal region in 27% and other locations in 24% cases. The mean age of presentation was 47.6 years and no sex predilection was seen. The size varied from 1×1 mm to 16×12 mm. The first two patients were asymptomatic which was similar to previously reported cases [3]. The third case presented with symptoms of watering, itching and heaviness of upper eyelid which has not been commonly reported. These symptoms may be attributed to the unusual location of this tumour. Jain P et al., reported the high frequency ultrasound findings of conjunctival myxoma which showed homogenous low internal reflectivity with no evidence of intraocular extension [10]. Grossly, conjuctival myxomas are usually well circumscribed, yellow or translucent masses which may be cystic or solid. Only three cases of diffuse involvement are described in literature [3,10].

Microscopically, the tumour is lined by conjunctival epithelium. It is paucicellular with scattered spindle and stellate cells, some showing intranuclear and intracytoplasmic inclusions. Intranuclear inclusions are due to invagination of the nuclear membrane while intracytoplasmic inclusions represent dilated endoplasmic reticulum [9]. The tumour shows collagen fibres and paucity of blood vessels. The second case in the present study showed increased vascularity with presence of lymphocytes probably because of inflammation secondary to itching which was an unusual symptom. Presence of mast cells and cystic change differentiate ocular myxomas from non ocular myxomas [11]. These tumours can show cellular pleomorphism with nuclear atypia as was seen in the present study cases. Alcian blue at pH 2.5 and mucicarmine stain mucopolysaccaride rich stroma. Tumour is vimentin positive, S100 negative and shows variable expression of SMA. All the three reported cases showed strong and diffuse positivity for vimentin. Ki-67 index is less than 5% [2,3].

Clinically, differential diagnosis of conjunctival myxomas is varied. In older patients, these tumours may be mistaken for malignant tumours such as lymphoma, ocular surface squamous neoplasia

and amelanotic melanoma. High frequency ultrasonography findings of conjunctival myxoma could not distinguish it from these tumours [10].

Microscopically, the differential diagnosis includes its close morphologic mimics with abundant myxoid stroma. Nerve sheath myxomas and myxoid neurofibromas contain dense wavy collagen fibres with spindly cells which are S100 positive. Spindle cell lipoma shows presence of adipocytes and CD 34 positive spindle cells while myxoid liposarcomas contains vacuolated lipocytes and lipoblasts with increased vascularity. Conjunctival myxomas are treated with simple excision and no recurrence has been reported. Myxomas can be a component of Carneys complex which includes cutaneous and cardiac myxomas, multiple pigmented lesions, schwannomas and endocrine overactivity and has significant morbidity and mortality [9]. Kennedy RH et al., have reported conjunctival myxoma as a part of Carneys complex in which patient had palpebral and eyelid lesions along with cardiac involvement [12]. Hence, following its diagnosis syndromic involvement of heart, endocrine system and skin has to be ruled out.

CONCLUSION(S)

Conjunctival myxomas are rare benign tumours which commonly present as cystic lesions. They can be differentiated from other benign and malignant conditions on the basis of its characteristic histological features. Hence, excision biopsy followed by histology is advisable. Moreover, its diagnosis can give a clue to underlying Carneys syndrome which needs to be ruled out by further work-up.

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