Conjunctival myxoma: A case report with unique high frequency ultrasound (UBM) findings

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A 39-year-old female presented with a painless yellow-pink tumor on her right eye. High-frequency ultrasound imaging revealed an epibulbar lesion with homogenous low internal reflectivity and no evidence of intraocular invasion. The patient underwent excisional biopsy leading to a pathology diagnosis of myxoma. Additional surgical margins as well as adjuvant cryotherapy margins were followed by extensive conjunctival repair. Herein, we report on a conjunctival myxoma with unique ultrasonographic findings.

Key words: Conjunctiva, high frequency, myxoma, tumor, ultrasound, ultrasound biomicroscopy

Myxoma is a rare, benign stromal tumor of mesenchymal origin often confused with other conjunctival stromal tumors.[1-3] Conjunctival myxomas are thought to originate in Tenon's capsule and can masquerade as conjunctival lymphoma, lymphangioma, ocular surface squamous neoplasia (OSSN), or amelanotic melanoma.[2] Syndromic associations such as Carney complex and Zollinger–Ellison syndrome warrant systemic evaluation as ocular signs may precede systemic findings.[4] Herein, we describe unique ultrasonographic findings of a conjunctival myxoma.

Case Report

A female was referred to The New York Eye Cancer Center with a painless mass in the right eye for 5 years duration. Ophthalmic oncology examination revealed a visual acuity of 20/20 in both eyes. A yellow-pink subconjunctival mass lesion was noted on temporal bulbar conjunctiva on the right eye. Though there were no posterior feeder vessels, fine intrinsic tumor vascularity was present [Fig. 1]. Her intraocular pressure measurement was 15 mmHg in the right eye. Anterior segment examination (including photographic documentation of all conjunctival surfaces), gonioscopy, and dilated funduscopic examination were within normal limits. There was no palpable preauricular or cervical lymphadenopathy.

High-frequency ultrasound imaging

Ultrasound biomicroscopy showed a dome-shaped epibulbar mass, measuring 7.9 by 6.7 mm in basal dimension with 1.5 mm of tumor thickness. Internal reflectivity was uniformly low. Multiple hypoechogenic vascular foci were seen scattered throughout the tumor [Fig. 2]. There was no scleral or intraocular invasion.

Informed consent and patient care complied with the tenets of the Declaration of Helsinki. Patient information was handled according to the United States Health Insurance Privacy and Portability Act. That said, close observation for growth or primary excisional biopsy was recommended. Mutually preferring the latter, the patient underwent primary excision (2 mm margins) with intraoperative frozen section evaluation. In that, the tumor had been poorly demarcated on slit-lamp examination; an additional 5 mm excisional margin plus 2–3 mm double freeze–thaw cryotherapy-treated margins were added. Extensive mobilization of the conjunctiva and Tenon's allowed closure. All excised tissues were sent for pathology for permanent section evaluation.

Histopathology revealed an irregularly delineated, paucicellular, limbal mass composed of evenly dispersed proliferating spindle cells in a myxoid matrix without melanin pigmentation; overlying epithelium was unremarkable. Numerous small blood vessels were seen in the stroma suggestive of intrinsic tumor vascularity. Immunohistochemistry showed lesional cells label for vimentin and smooth muscle actin (SMA), while negative for S100. The stroma stained positive for Alcian blue stain. A Ki-67 stain did not show an increase in proliferation index. Overall, features were consistent with a myxoma [Fig. 3].

In consideration of her biopsy findings, systemic evaluation including an echocardiography was requested.

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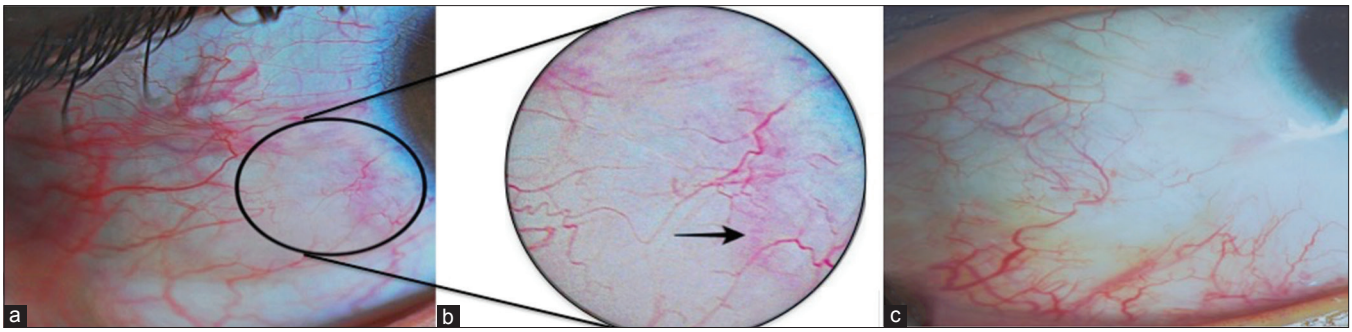

Figure 1: (a) Slit-lamp photograph of the right eye shows a yellow-pink temporal epibulbar mass, (b) magnified view showing intrinsic tumor vascularity (arrow), and (c) slit-lamp photograph at 4-month follow-up

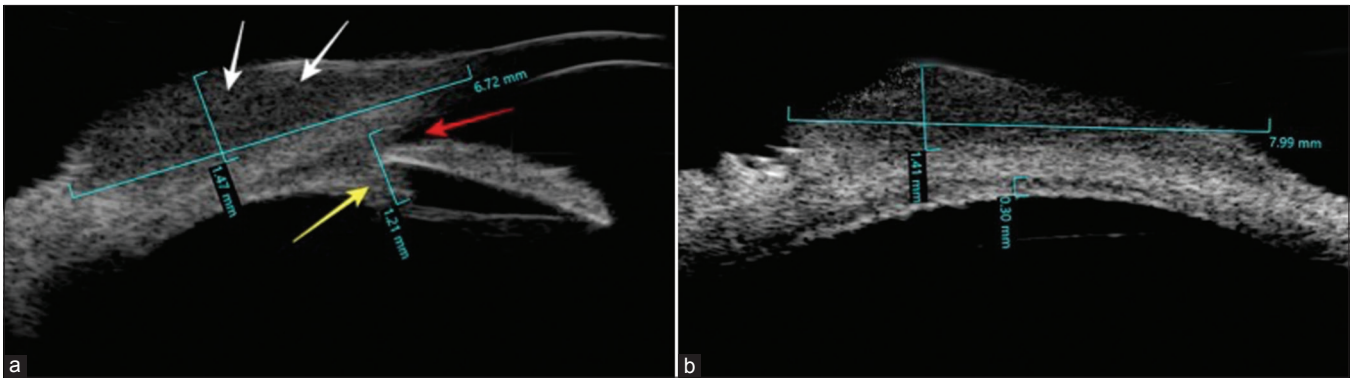

Figure 2: High-frequency ultrasound image of the myxoma (a) longitudinal section showing a dome-shaped epibulbar mass 6.7 mm in basal dimension with 1.5 mm of thickness. It reveals uniform low internal reflectivity with multiple vascular hypoechogenic foci (white arrows). The sclera is intact. The anterior chamber angle is sharp (red arrow) and ciliary body thickness is normal (1.2 mm; yellow arrow). (b) Transverse section showing basal dimension of 7.9 mm, intact sclera, and no uveal thickening

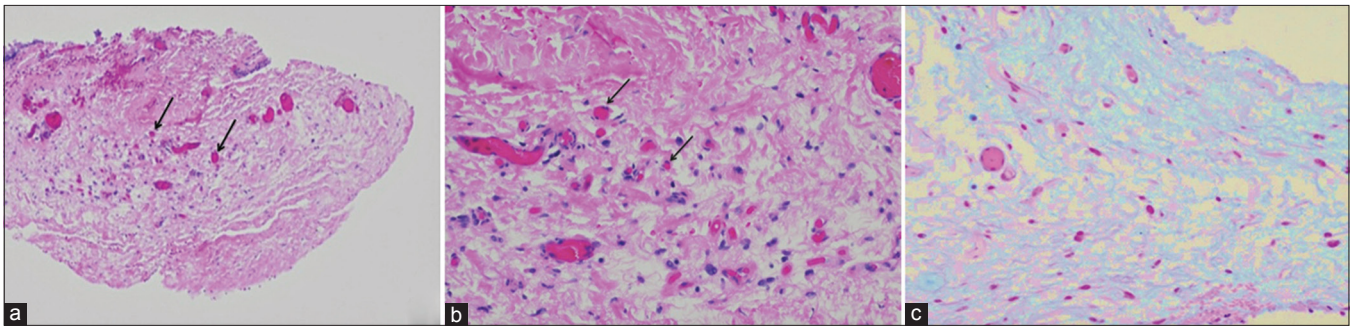

Figure 3: (a) Intrinsic vasculature (arrows) (H and E, ×4), (b) paucicellular myxoid matrix with fine blood vessels (arrow) (H and E, ×20), and (c) alcian blue stain positive (×20)

Discussion

Conjunctival myxoma was first reported in 1913.[5] Using the key words myxoma, conjunctiva, ocular, epibulbar, stromal, tumor on PubMed Central, we could find only 48 cases published in literature.[1] Mean age at presentation is 47.6 years with no gender predilection.[1] As in our case, epibulbar myxomas typically present as a solitary smooth, yellow-pink, temporal bulbar conjunctiva mass, with sizes varying from 4 to 20 mm.[2] Intrinsic vascularity and intraocular extension have not been reported. We could find no reports on the high-frequency ultrasound characteristics of conjunctival myxoma.

This myxoma demonstrated a dome apical shape and low internal ultrasonographic reflectivity [Fig. 2]. The noteworthy feature was the intrinsic vascularity evidenced by the multiple hypoechogenic foci, which was confirmed on histopathology. Other conjunctival tumors with low internal reflectivity typically demonstrate monomorphic tissue patterns. They include conjunctival melanoma, lymphoma, and OSSN.[6-8] In contrast, high-frequency ultrasound of a lymphangioma can demonstrate multicystic, irregularly reflective architecture. Though rare entities, such as malignant fibrous histiocytoma, myxoid neurofibroma, myxoid liposarcoma must be considered, most will be differentiated by histopathological analysis.[1,2]

Histopathology of conjunctival myxoma shows scanty, spindle-shaped cells in a mucinous matrix rich in hyaluronic acid.[1,2] Immunohistochemical staining is positive for vimentin, alcian blue

and negative for S-100, desmin, and myoglobin.[1,2] In our case, the unusual feature was the fine intrinsic vascularity and SMA positivity.

The treatment of myxoma is complete excision.[1,2] However, in view of intrinsic tumor vascularity and clinically ill-defined margins, we supplemented excision and added double-freeze–thaw cryotherapy to increase our margin safety. Amniotic membrane graft was not utilized, as primary closure was possible. Amnion is a transplanted tissue and thus carries a possible risk of virus transmission and dislocation prior to epithelialization. Conjunctival myxoma has not been shown to recur or undergo malignant transformation.[1] Though not seen in our case, eye cancer specialists must consider its association with Carney complex and Zollinger–Ellison syndrome.[9,10]

Conclusion

High-frequency ultrasound findings of a conjunctival myxoma are similar to a conjunctival lymphoma, OSSN, and melanoma but differ from tumors with more developed intrinsic vascular or fibrous components. Preoperative high-frequency ultrasound imaging, correlated to histopathology, revealed a distinct vascular pattern along with no evidence of intraocular invasion.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

References

1. Ríos Y Valles-Valles D, Hernández-Ayuso I, Rodríguez-Martínez HA, Medina-Cruz A, Salcedo-Casillas G, Rodríguez-Reyes AA, *et al.* Primary conjunctival myxoma: Case series and review of the literature. Histopathology 2017;71:635-40.
2. Sharma N, O'Hagan S, Phillips G. Conjunctival myxoma – Atypical presentation of a rare tumour: Case report and review of literature. BMC Ophthalmol 2016;16:54.
3. Herwig MC, Wells JR, Grossniklaus HE. Conjunctival stromal tumor: Report of 4 cases. Ophthalmology 2012;119:682-7.
4. Correa R, Salpea P, Stratakis CA. Carney complex: An update. Eur J Endocrinol 2015;173:M85-97.
5. Magalif: Myxoma conjunctivae. Klin Monatsbl Augenheilkd 1913;51:844.
6. Garcia JP Jr., Spielberg L, Finger PT. High-frequency ultrasound measurements of the normal ciliary body and iris. Ophthalmic Surg Lasers Imaging 2011;42:321-7.
7. Finger PT, Tran HV, Turbin RE, Perry HD, Abramson DH, Chin K, *et al.* High-frequency ultrasonographic evaluation of conjunctival intraepithelial neoplasia and squamous cell carcinoma. Arch Ophthalmol 2003;121:168-72.
8. Finger PT, Perry HD, Kempin S, Iacob CE. High-frequency ultrasound of extranodal limbal rosai-dorfman disease: Affecting the conjunctiva, sclera, and cornea. Cornea 2007;26:888-90.
9. Kennedy RH, Flanagan JC, Eagle RC Jr., Carney JA. The carney complex with ocular signs suggestive of cardiac myxoma. Am J Ophthalmol 1991;111:699-702.
10. Ramaesh K, Wharton SB, Dhillon B. Conjunctival myxoma, Zollinger-Ellison syndrome and abnormal thickening of the inter-atrial septum: A case report and review of the literature. Eye (Lond) 2001;15:309-12.