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Case report

Epibulbar osseous choristoma: A case report

Keegan A. Harkins [a, *], Deborah Perry [b], Donny W. Suh [a, b]

[a] Stanley M Truhlsen Eye Institute, Department of Ophthalmology and Visual Science, University of Nebraska Medical Center, Omaha, NE, USA
[b] Children's Hospital and Medical Center, Omaha, NE, USA

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ABSTRACT

Purpose: To present the case of a 12-year-old female with an epibulbar osseous choristoma.
Observations: The patient presented with right-sided conjunctival mass, which caused her discomfort. Slit lamp examination revealed a 5×5-mm, firm nodule in the superotemporal quadrant of the bulbar conjunctiva. The nodule had feeder vessels, adhered firmly to the sclera, and lacked signs of malignancy. The patient underwent excisional biopsy under general anesthesia. During this procedure, great care was taken to avoid perforation of the globe. The pathologic sections were significant for well-circumscribed osseous tissue without atypia.
Conclusions and importance: We describe diagnosis and successful surgical management of osseous choristoma the rarest subtype of ocular choristoma. With only 65 cases reported since mid-19th century, the condition remains poorly described. This report provides additional information on diagnosis and treatment of this rare condition.
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1. Introduction

A choristoma is a benign mass composed of tissue normally found in another location. Ocular choristomas can be classified as dermoid, dermatolipoma, complex choristoma (choristomas with more than one tissue type), and single-tissue choristoma. Examples of the latter are lacrimal gland, respiratory, and osseous choristomas.[1] Epibulbar osseous choristoma is the rarest type of choristoma of the eye.[2] Composed of firm deposits of bone, osseous choristomas are most commonly found in the superotemporal conjunctiva; occasionally, the mass is firmly attached to the sclera. As with all choristomas, osseous choristomas are believed to be congenital. Due to its location underneath the eyelid, the mass is usually detected once the child becomes old enough to palpate it. Here we present the case of a 12-year-old female with an epibulbar osseous choristoma.

2. Case report

A 12-year-old female without a remarkable medical, ocular, or traumatic history was referred to a tertiary care center for evaluation of right-sided conjunctival mass first noticed 3 months prior. She complained of moderate foreign body sensation underneath the eyelid in the superotemporal quadrant that was progressively becoming more uncomfortable. On examination, visual acuity was found to be 20/20 bilaterally. Intraocular pressures were 19 mmHg in both eyes. Her pupils were equal and reactive, extraocular movements were full, and alignment was orthotropic. Slit lamp examination revealed a 5×5–m, firm nodule in the superotemporal quadrant of the bulbar conjunctiva (Fig. 1). The nodule had feeder vessels and adhered firmly to the sclera, as manipulation with a cotton tip applicator revealed. Evidence of malignancy such as, multiple pigmentations, ulceration, neovascularization, or necrosis, was lacking.

The next week, the patient underwent excisional biopsy under general anesthesia. Extraocular muscles were identified prior to surgery and were displaced to avoid any damage. After conjunctival and Tenon's capsule incision of approximately 5 mm overlying the lesion, the whitish mass identified which was firmly adherent to the sclera away from the extraocular muscles. The conjunctiva and Tenon's capsule were carefully dissected to expose the mass of 5 × 5 mm. The mass was carefully separated from the sclera with a size 11 blade with a microscope on high magnification. Careful dissection was made to avoid any potential perforation of the globe. After the lesion was removed, mild thinning of the sclera was noted from chronic pressure and mass effect. Feeder vessels were carefully cauterized to control the hemorrhage. Then conjunctiva and

* Corresponding author. University of Nebraska Medical Center, 3902 Leavenworth St, Omaha, NE 68105, USA.
E-mail address: kharkins@unmc.edu (K.A. Harkins).

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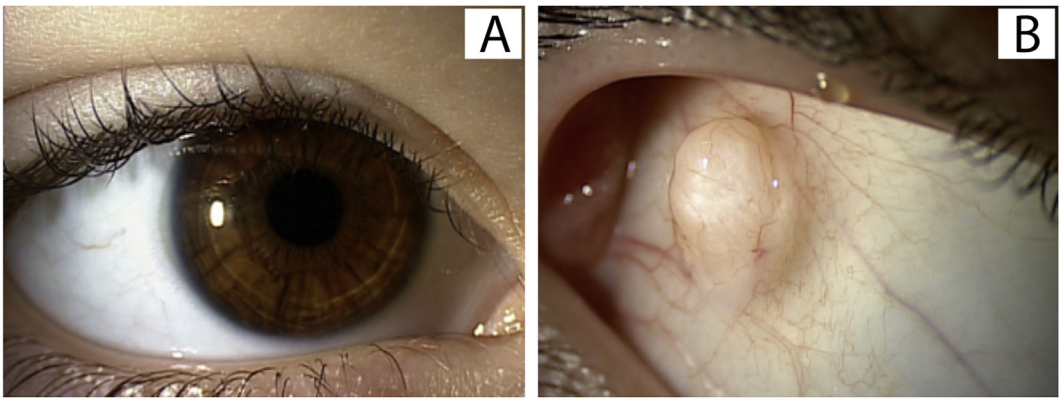

Fig. 1. Slit lamp examination photographs showing the right eye (A) and prominent conjunctival mass with some vascularization (B).

Tenon's capsule were closed with 2 interrupted 6.0 plain gut sutures. A 0.4 × 0.3 × 0.2 cm firm nodule was sent to pathology (Fig. 2). The pathologic sections were significant for well-circumscribed osseous bone without atypia. After excision, Tenon's capsule and conjunctiva were sutured to close the surgical incision site. At the patients two week follow up, her primary complaint of foreign body sensation had resolved.

3. Discussion

In this report, we describe a pediatric case of epibulbar osseous choristoma, a very rare tumor of the eye. Because only a few cases have been reported, diagnosis and management may pose a challenge, some of which are discussed below.

The differential diagnosis of a pediatric conjunctival mass includes the following entities: limbal dermoid, myxoma, scleral melanocytosis, melanoma, Kaposi's sarcoma, sebaceous carcinoma, extraocular retinoblastoma extension, and intraorbital foreign body, among others.[3–5] Shields et al. found the following etiologies in 262 children with conjunctival masses referred to an oncology service: 67% melanocytic, 10% choristomatous, 9% vascular, and 2% benign epithelial.[3] Similar pathology series by Cunha et al. and Elsas and Green found choristomas to constitute 23%–29% of conjunctival masses.[5,6]

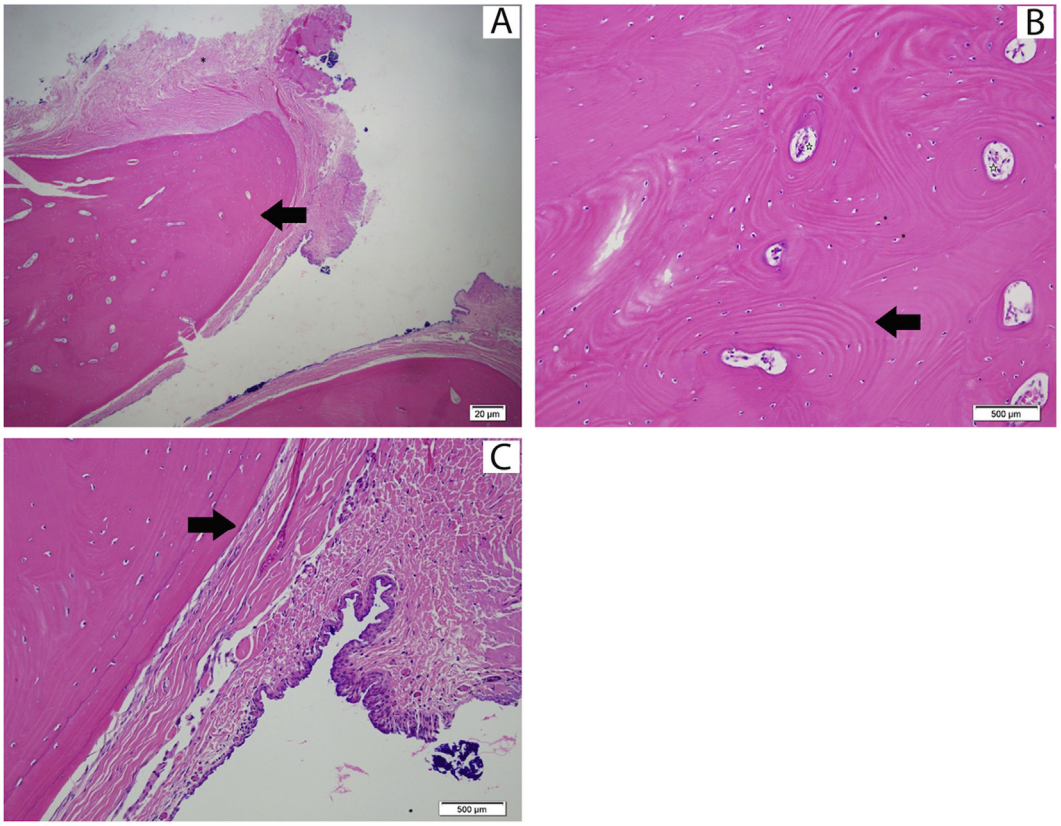

Fig. 2. Pathology section on low power magnification showing conjunctiva (*) and lamellar bone (arrow) (A). High-power magnification revealing Haversian canals (stars) surrounded by concentric light and dark rings of lamellar bone and normal appearing osteocytes (*) (B). High magnification of the well-defined junction between bone and conjunctiva (arrow) (C).

As stated in the introduction, of these orbital choristomatous masses, osseous choristomas are the rarest. The first case of epibulbar osseous choristoma was described by Von Graefe in 1863.[7] Since that time, only 65 total cases have been reported in the literature in both pediatric and adult patients. There is a slight female preponderance (59%) as well as, interestingly, a preponderance (68%) for the right eye. Our case supports these statistics. Of the cases that reported location, 76% were located in the superotemporal quadrant as our case was.[5] The mass can freely mobile or be firmly attached to the sclera or the rectus muscles.[8] Shields reports that the osseous choristomas, unlike other choristomas, have limited to no growth potential.[4] Measured growth of these masses has been reported on several occasions; however, this growth is likely attributed to the growth of tissues other than bone, eg, (lacrimal acini, dermoid, fibrous connective tissue, fibroadiopose tissue, or nerves), which can be found in 10% of these tumors.[8–9]

Frequently, osseous chorisomas get biopsied to confirm the diagnosis and rule out malignant causes. Histopathology shows compact bone surrounded by fibrous tissue.[8] Haversian canals with concentric rings of lamellar bone are common, but bone marrow is rarely found. The osteocytes appear normal.

Malignant transformation or intraocular extension of choristomas has never been reported in the literature. Therefore, it has been suggested that these lesions may be managed by observation if the lesion is typical and ultrasound or computed tomography scan has confirmed the diagnosis by finding calcium components.[2,10] When surgery is necessary, the indications include: need for diagnosis, cosmesis, tearing, foreign body sensation, irritation, or recurrent ocular inflammation. If the lesion has intimate scleral attachment requiring a partial sclerectomy, great care during surgery and consideration of preoperative imaging are needed to avoid iatrogenic globe rupture. In this case, we have taken great care to avoid globe perforation.

4. Conclusions

In conclusion, this report presents a case of a very rare conjunctival tumor. If epibulbar osseous choristoma is considered in the differential diagnosis of a pediatric conjunctival mass, it may be observed closely in asymptomatic patients with small lesions that are stable, but if there are signs of growth or new symptoms, careful surgical excision may be needed. Since the patient reported discomfort, we have elected to performed surgical excision of the mass, which successfully resolved this symptom.

Patient consent

The patient's family provided consent to publish details of this case.

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Conflict of interest

The authors of this paper listed below have no financial interests or conflicts to report.

Keegan A. Harkins, Deborah Perry, Donny W. Suh.

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Authorship

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