Epibulbar osseous choristoma

a clinicopathological case series and review of the literature

Epibulbäre Knöcherne Choristome

Eine klinisch-pathologische Fallserie und Überblick der Literatur

Maud Vachette

Alexandre Moulin, MD

Leonidas Zografos, MD

Ann Schalenbourg, MD

Jules-Gonin Eye Hospital
University of Lausanne
Switzerland

No conflict of proprietary interest for any author.
Running head: Epibulbar osseous choristoma

Corresponding Author: Dr Ann Schalenbourg, MER
Jules-Gonin Eye Hospital
15, Avenue de France
CH-1004 Lausanne
Switzerland
Tel: +41.21.626.8111
Fax: +41.21.626.8889
Mail: ann.schalenbourg@fa2.ch

PRECIS

Dieses Artikel beschreibt eine seltene Fallserie von epibulbären knöchernen Choristomen in 3 männliche Patienten zwischen 3 Monaten und 11 Jahren, von denen eines einen komplexen Choristom dargestellt hat, weil es mit einem Dermolipom assoziiert war. Wir haben ebenfalls einen Überblick der Literatur durchgeführt.

ABSTRACT

Background: Choristomas are benign, congenital tumours composed of normal tissue in an abnormal location. Osseous choristomas represent the rarest form of epibulbar choristomas, with now 65 cases reported in the literature. We did a retrospective clinicopathological study of all patients with epibulbar osseous choristoma observed at our institution since 1982 and updated the last review of the literature.

History and Signs: Three Caucasian male patients, aged between 3 months and 11 years, were identified. All osseous choristomas were located under the superotemporal bulbar conjunctiva of the right eye.

Therapy and Outcome: All lesions were managed with surgical excision. Histopathology revealed the presence of lamellar bone in all cases, one of which was associated with a dermolipoma.

Conclusions: We report a small rare case series of 3 epibulbar osseous choristomas and did a review of the literature. In one patient, the osteoma was associated with a dermolipoma, corresponding to the fourth reported complex choristoma of this type, in an otherwise normal eye, in the literature.

KEYWORDS: Osteoma; choristoma; dermolipoma; epibulbar; bone and bones

ABSTRACT

Hintergrund: Choristome sind gutartige, angeborene Tumore von normalem Gewebe an abnormaler Stelle. Knöcherne Choristome sind mit nun 65 Fällen in der Literatur, die seltenste Form der epibulbären Choristome. Wir haben eine retrospektive klinisch-pathologische Studie durchgeführt, die alle Patienten der Jules-Gonin Augenklinik mit epibulbären knöchernen Choristomen seit 1982 einschliesst, und mit der Literatur verglichen.

Anamnese und Befund: Drei kaukasische männliche Patienten im Alter zwischen 3 Monaten und 11 Jahren wurden identifiziert. Alle knöchernen Choristome befanden sich unter der superotemporalen bulbären Konjunktiva des rechten Auges.

Therapie und Verlauf: Alle Läsionen wurden chirurgisch exzidiert. Histopathologisch wurde in allen Fällen Lamellenknochen nachgewiesen. Einer der Fälle war zudem mit einem Dermolipom assoziiert.

Schlussfolgerung: Wir berichten über eine seltene Fallserie von 3 epibulbären knöchernen Choristomen. Ein Fall war mit einem Dermolipom assoziiert, der vierte beschriebene Fall dieser Art von komplexen Choristomen in einem ansonsten normalen Auge.

SCHÜSSELWÖRTER: Osteom; Choristom; Dermolipom; epibulbär, Knochen

BACKGROUND

Choristomas are benign, congenital tumours composed of normal tissue in an abnormal location [1]. Simple choristomas contain one tissue type, whereas complex choristomas combine tissue derived from 2 germ layers, ectoderm and mesoderm [2].

Simple ocular choristomas can be dermoids, dermolipomas, lacrimal gland or respiratory cyst choristomas, as well as epibulbar osseous choristomas. The latter consist of pure bone on the surface of the globe. They are the rarest form of epibulbar choristomas [3]. Usually, they are diagnosed during the two first decades of life though the eldest patient reported was 79 years old [4].

We performed a retrospective chart study of all patients with epibulbar osseous choristoma at the Jules-Gonin Eye Hospital (Lausanne, CH), since 1982. We reviewed the literature, identifying articles through a computerized search in MEDLINE. Keyword terms were (“Osteoma”[Mesh]) AND Epibulbar and (“choristoma”[Mesh]) AND “Bone and Bones”[Mesh] AND Epibulbar.

CASE REPORTS

Patient 1

An 11-year-old boy was referred to the Jules-Gonin Eye Hospital because of a lump on his right eye, with a foreign body sensation since 3 months. Medical history was unremarkable. Visual acuity was 10/10 in both eyes. Slit lamp examination revealed a superotemporal, non-pigmented, multinodular mass (Fig. 1A-B), which was adherent to the sclera and hard on palpation. On computerised tomography (CT) scan of the orbit, the lesion was calcified, leading to the diagnosis of an epibulbar osseous choristoma (Fig. 1C). Surgical excision was performed, without complications.

Histopathology of the whitish firm nodule identified central well-organized lamellar bone, surrounded by periosteum and embedded in fibro-adipose tissue (Fig. 1D).

Patient 2

A 3-month-old boy was referred because of a congenital yellow-orange tumour on the right eye, extending under the superotemporal conjunctiva anteriorly towards the limbus (Fig. 2A).

Cycloplegic refraction and motility examination were within normal limits. With a presumed diagnosis of dermolipoma, the lesion was observed; as it had increased 3 months later, surgical excision was performed. During surgery, the mass was shown to invade the lateral rectus muscle. Close to this insertion, a separate, hard mass was exposed, the aspect of which evoked an osseous choristoma. Through lamellar dissection, this lesion was separated from the superotemporal sclera and also removed.

Histology of the first tumour revealed a dermolipoma composed of mature fat cells, covered by dermis in which hair follicles and sebaceous glands were observed. At the surface, the squamous epithelium was partially keratinized in some areas and a granular layer could be identified (Fig.2B). The second, hard mass was surrounded by dense fibrous tissue and consisted of a small fragment of bone with Haversian canals (Fig. 2C) and larger, haemorrhagic, partially fibrosed cavities filled with mature blood cells (Fig. 2D), leading to the diagnosis of an epibulbar osseous choristoma.

Patient 3*

A 7-year-old boy, complaining of an irritated right eye, was brought to the Emergency room. The parents had noticed, close to the upper temporal fornix, a cyst-like nodule that was punctured. When the lesion remained unchanged, surgical excision was proposed. During surgery, the mass was noted to be hard and adherent to the sclera.

Histologically, the beige nodule consisted of a well-defined lamellar bone, accompanied by elastic fibrous tissue.

*The pictures of this case, without a description of the case history, were published in 1984 [5].

DISCUSSION

We report a rare consecutive case series of 3 male patients with epibulbar osseous choristoma, including one associated with a dermolipoma.

In 1863 [6], Von Graefe reported the first case of osseous choristoma, with the tumour interestingly located under the tarsal conjunctiva. Spencer-Watson described the first epibulbar osseous choristoma in 1871 [7].

Pathogenesis is not well known. A history of trauma or inflammation is sometimes associated, but the former might only have drawn attention to the lesion, while the latter could be secondary. Boniuk and Zimmerman suggested an atavistic origin [8]. In a comparative study of the scleral ossifications in different classes of vertebrates, Bonafonte questioned this theory [9]. Most authors believe that osseous choristomas represent an abnormal development of the embryonic pluripotential mesenchyme [1].

Being the rarest form of epibulbar choristoma, 63 cases of epibulbar osteoma in 62 patients have now been published [1,4,10-13]. We updated Gayre's meta-analysis of 2002 on 51 cases [1], which included already our third patient [5], and added the more recently published cases [4,10-13] as well as our other two, totalling 65 cases in 64 patients. We found that of the 55 tumours with a reported location, 42 were localised in the superotemporal quadrant (76%), 11 temporally (20%), one inferotemporally (2%) and one superiorly (2%). Of the 41 lesions where laterality is mentioned, 28 occurred in the right eye (68%). However –in contrast to our cases-, 24 of the 41 lesions with a given gender were reported in females (59%).

Although most epibulbar osteomas occur as isolated lesions, Gayre identified 7 cases of osseous choristomas associated with other ectopic tissue [1], to which we added Verity's second [13], Tsai's first [4], and our second patient, resulting in 10 cases of 65 (15%). The other choristomatous tissues included cartilage, lacrimal gland, nerve, fibrous connective or adipose tissue. However, we could only identify 3 cases where a well defined dermolipoma [14-15] or dermoid [16] was associated with an epibulbar osteoma in an otherwise normal eye, our second case becoming the fourth ever reported. These combined lesions are complex choristomas; they are rare and have also been reported in deformed eyes [15] or in association with the organoid naevus syndrome [2-3].

Although Beckman and Sugar described, already in 1961, a 'tooth-shaped opacity' on orbital X-ray [19], it's only the arrival of CT scan in the eighties that facilitated a preoperative diagnosis. Differential diagnosis includes age-related calcifications involving the insertion of the lateral recti muscles [20], an extraocular retinoblastoma extension [11] or an intraorbital foreign body [13].

Epibulbar osteomas are benign tumours, without any report of an intraocular extension or malignant transformation. While growth of other epibulbar choristomas has been well documented [1], it is controversial in osseous choristomas. Some authors noticed a slow

increase in size [1,8], whereas the Shields state that these lesions don't grow [3]. In our second case, we speculate that it was the adipose tissue that increased in volume.

Indications for surgery include the need for diagnosis, cosmesis, ocular inflammation and tearing. As these lesions can involve the rectus muscle as well as the sclera, the surgeon needs to prepare for strabismus surgery and –rarely- a scleral graft [3]. Alternatively, in minimally symptomatic cases, a conservative attitude can be adopted [1-3].

REFERENCES

1. Gayre GS, Proia AD, Dutton JJ. Epibulbar osseous choristoma: case report and review of the literature. Ophthalmic Surg Lasers 2002; 33(5): 410-415

2. Pe'er J. Conjunctival stromal tumors. In: Singh AD, Damato BE, Pe'er J, Murphree AL, Perry JD ed. Clinical ophthalmic oncology. Philadelphia: Saunders Elsevier Inc, 2007; 150-157

3. Shields JA, Shields CL. Conjunctival and epibulbar choristomas. In: Shields JA, Shields CL ed. Eyelid, conjunctival and orbital tumors. 2nd ed. Philadelphia: Lippincott, Williams & Wilkins, 2008: 249-265

4. Tsai ASH, Lee KYC, Al Jajeh I et al. Epibulbar osseous choristoma: a report of two cases. Orbit 2008; 27(3): 231-233

5. Zografos L, Uffer S. Epibulbar osteoma, J Fr Ophtalmol 1984; 7(10): 625-626

6. Von Graefe A. Tumor in submucosen gewebe lid-bindehaut von eigenthumlicher, beschaffenheit. Klin Monatsbl Augenheilkd 1863; 1: 23

7. Spencer-Watson W. An ivory exostosis growing from the sclerotic coat of the eye. Transactions of the Pathological Society of London 1871; 22: 227-228

8. Boniuk M, Zimmerman LE. Epibulbar osteoma (episcleral osseous choristoma). Am J Ophthalmol 1962; 53: 290-296

9. Bonafonte S, Aguirre A. Is the human epibulbar osteoma an atavism? Ocul Ther Surg 1985; 4: 157-160

10. Osoba Y, Dodd CL, Bonshek R. Epibulbar osseous choristoma. Eye 2001; 15: 547-548

11. Marback EF, Sout TJ, Rao NA. Osseous choristoma of the conjunctiva simulating extraocular extension of retinoblastoma. Am J Ophthalmol 2003; 133: 825-827

12. Kim BJ, Kazim M. Bilateral symmetrical epibulbar osseous choristoma. Ophthalmology 2006; 113: 456-458

13. Verity DH, Rose GE, Uddin JM et al. Epibubar osseous choristoma : benign pathology simulating an intraorbital foreign body. Orbit 2007; 26(1): 29-32

14. Santora DC, Biglan AW, Johnson BL. Episcleral osteocartilagenous choristoma. Am J Ophthalmol 1995; 119: 654-655

15. Hered RW, Hiles DA. Epibulbar osseous choristoma and ectopic lacrimal gland underlying a dermolipoma. J Pediatr Ophthalmol Strabismus 1987; 24(5): 255-258

16. Ferry AP, Hein HF. Epibulbar osseous choristoma within an epibulbar dermoid. Am J Ophthalmol 1970; 70: 764-766

17. Wagenmann A. Über einen merkwürdigen Fall von Dermoid Geschwulst mit rudimentarer Entwickelung des Auges. Archiv für Ophthalmologie 1889; 35: 111-146

18. Schieck F. Eine bislang unbekannte Missbildung des Auges. Bet Dtsch Opthalmol Ges 1910; 36: 311.

19. Beckman H, Sugar HS. Episcleral osseous choristoma : report of two cases. Arch Ophthalmol 1964; 71: 377-378

20. Gonnering RS, Fuerste FH, Lemke BN et al. Epibulbar osseous choristomas with scleral involvement. Ophthal Plast Reconstr Surg 1988; 4(1): 63-66

Figure legends

Figure 1

(A, B): Slit lamp examination revealing a whitish multinodular mass located superotemporally in the right eye

(C): Coronal CT scan of the orbit showing a calcified density on the right superotemporal sclera

(D): Epibulbar lesion, composed of mature lamellar bone, surrounded by periosteum and embedded in fibro-adipose tissue. (Hematoxylin-eosin; original magnification 26x)

Figure 2

(A): Yellow-orange congenital tumoral formation, located superotemporally in the right eye.

(B): Dermolipoma composed of mature fat cells, covered by dermis containing adnexae. The overlaying squamous epithelium is partially keratinized. (Hematoxylin-eosin; original magnification 63x)

(C): Epibulbar lesion, surrounded by dense fibrous tissue and consisting of bone with Haversian canals. (Hematoxylin-eosin; original magnification 26x)

(D): Central cavity within the bone, filled with mature red blood cells, eosinophilic cells, lymphocytes and mastocytes. (Hematoxylin-eosin; original magnification 260x)

Figures

Figure 1

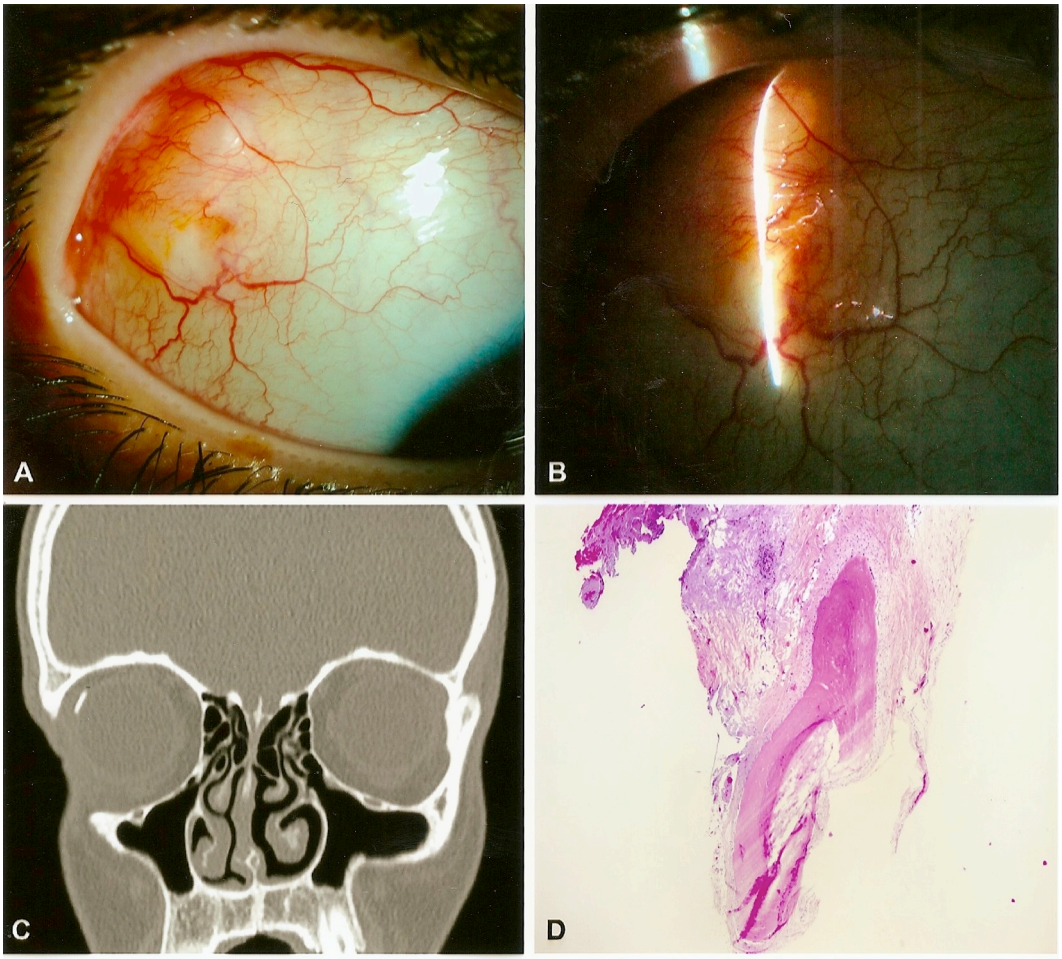

Figure 2