

Adherent Episcleral Osseous Choristoma

Devina Gogi,¹ Rana Sherwani²

¹Institute of Ophthalmology and ²Department of Pathology, Jawaharlal Nehru Medical College, Aligarh Muslim University, Aligarh, India

This report is of a rare congenital lesion, an episcleral osseous choristoma, which was found firmly adherent to the underlying sclera in an adolescent girl. Careful lamellar scleral dissection of the lesion enabled total excision of the lesion with histopathology revealing a lamellar bone structure characteristic of this lesion.

Key Words: Choristoma, Osteoma

Asian J Ophthalmol 2006;8(1):35-36

Introduction

Episcleral osseous choristoma (epibulbar osteoma), a rare congenital benign lesion presenting as an epibulbar lesion, is typically seen in the superotemporal quadrant 5 to 10 mm from the limbus.¹⁻⁷ To date, only 52 osseous choristomas have been reported in literature.⁸ Osseous choristomas are generally seen in essentially healthy eyes and are usually loosely attached to the underlying sclera.⁸ This report is of the management of a patient with episcleral osseous choristoma that was firmly adherent to the underlying sclera, a type that is rarely described in the literature.⁹

Case Report

A 16-year-old girl presented in 2002 with a pea-sized nodule in the upper temporal quadrant of the left eye, which had been present since childhood but had slowly increased in size during the previous year (Figure 1). There was no history of injury or any episode of pain or discomfort. The patient's best-corrected visual acuities were 6/6 bilaterally. Examination of the left eye revealed an 8-mm diameter hard non-tender quadrangular firm lesion 7 mm from the limbus at the 2 o'clock position in the upper temporal quadrant. The lesion was firmly fixed to the underlying sclera, had a relatively smooth surface, and showed

no evidence of hair or any adenexal structures protruding from the surface. Lid fissure height and levator function were symmetrical. No obvious diplopia was noted and full ocular motility was present. External examination of the right eye, anterior segment examination of both eyes, and fundus examination of both eyes were normal.

Differential diagnoses of dermoid cyst, dermolipoma, prolapsed orbital fat, or conjunctival granuloma were made. Echography revealed superficial scleral

involvement of this high-density lesion. Conservative observation of the lesion was advised, but local excision of the lesion was done under general anaesthesia on request from the patient's carers and for aesthetic reasons. The tumour was found to be firmly adherent to the underlying sclera. Blunt and careful lamellar scleral dissection of the lesion was performed leaving more than half of the scleral thickness. The raw area of the sclera was covered with conjunctiva, which was closed with a running 6.0 vicryl suture. The hard nodule measured 8 mm in diameter and 3 mm in thickness (Figure 2). Histopathological examination revealed a dense lamellar bone with haversian canals surrounded by a rim of fibrous connective tissue (Figure 3).

At the 2-year follow-up visit, the patient was asymptomatic with no evidence of local recurrence of the lesion.

Discussion

The origin and nomenclature of osseous choristoma has always been a subject of controversy.¹⁻⁴ First described by Grafe in 1863 as epibulbar osteomas,¹ these lesions



Figure 1. Photograph of the right eye of the patient showing episcleral osseous choristoma in the upper temporal quadrant.

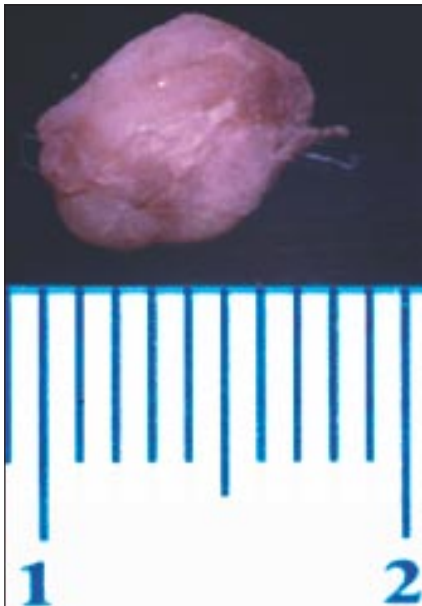


Figure 2. Excised lesion.

were thought to be teratomas arising from the conjunctiva. However, they proved to be congenital quiescent malformations composed only of mature compact bone. Henceforth, Beckman and Sugar preferred to call them 'osseous choristoma',⁴ a nomenclature that has persisted into modern literature.

This lesion is typically noted at birth or shortly thereafter,² although a few cases have been reported for which trauma drew attention to a pre-existing lesion.^{2,5} Additionally, most of the lesions reported in the literature do not change, with only a few lesions showing any change in size, as occurred in this patient.^{6,7}

The classical location of this tumour is the upper temporal quadrant.⁸ The lesion is seen as a round to oval nodule with a concave inner surface and a convex outer surface. Due to the movement of the upper eyelid pressing on the globe, it may assume the shape of a disc or a rectangle.

The presentation can be a diagnostic dilemma but the firm nature of the lesion, absence of hair and adenexal structures on the surface, and the characteristic

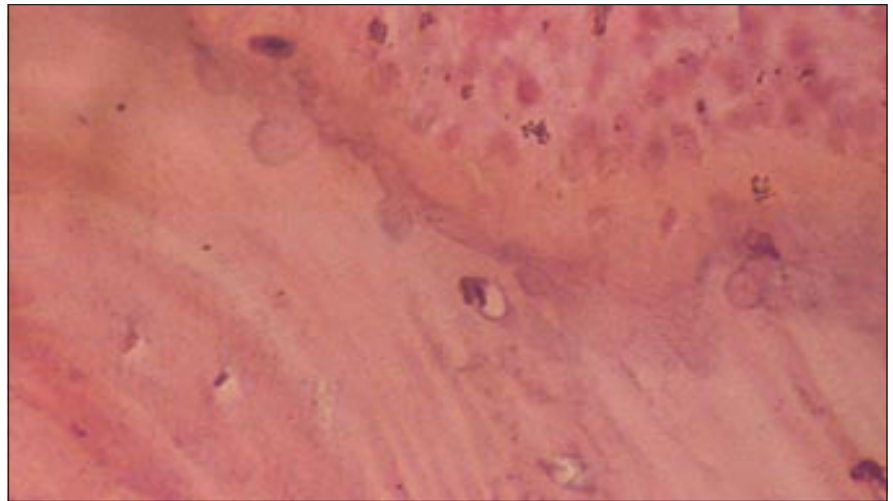


Figure 3. Histological appearance of the lesion showing bone surrounded by fibrous tissue (haematoxylin and eosin).

histopathological findings differentiate it from dermoid and dermolipoma. Furthermore, differentiation from conjunctival granuloma is relatively easy due to the presence of a predisposing factor, inflammation, and vascularity associated with conjunctival granuloma.

Usually, episcleral osseous choristomas move freely over the sclera, making local excision easy. However, they seldom adhere firmly to the muscle sheath or the underlying sclera⁹ as in this patient. In this situation, careful and blunt dissection of the lesion is mandatory before shaving off the tumour level with the sclera. For these patients, preoperative computed tomography scanning or ultrasonography is advisable to rule out dense scleral adherence. The management is controversial as there is a risk for iatrogenic globe perforation and scleral ectasia with excision. Thus, conservative management or surgery may be considered, depending on a combination of orbital imaging and clinical examination.

References

1. Grafe V. Tumour in the Submucosen Gewebe der Lid-Bindehaut von eigen-thumlicher Beschaffen-heit. *Klin*

Monatsbl F Augenheil 1863;1: 23-26.

2. Boniuk M, Zimmerman LE. Epibulbar osteoma (episcleral osseous choristoma). *Am J Ophthalmol* 1962;53:290-296.
3. Ballantyne AJ. Two cases of epibulbar osteoma. *Ophthalmologica* 1940;99: 87-95.
4. Beckman H, Sugar HS. Episcleral osseous choristoma. Report of two cases. *Arch Ophthalmol* 1964;71:377-378.
5. Ortiz JM, Yanoff M. Epipalpebral conjunctival osseous choristoma. *Br J Ophthalmol* 1979;63:173-176.
6. Sihota GS. Epibulbar osteoma. *Br J Ophthalmol* 1964;48:504-506.
7. Roch LM, Milauskas AT. Epibulbar osteomas. *Arch Ophthalmol* 1968;79: 578-580.
8. Gayre GS, Prola AD, Dutton JJ. Epibulbar osseous choristoma: case report and review of the literature. *Ophthalmic Surg Lasers* 2002;33:410-415.
9. Gonnering RS, Fuerste FH, Lemke BN, Sonneland PR. Epibulbar osseous choristoma with scleral involvement. *Ophthalmic Plast Reconstr Surg* 1988;4: 63-66.

Address for Correspondence

Dr Devina Gogi
14, Woodlea Park
Meanwood
Leeds LS6 4SH
UK
E-mail: devinagogi@hotmail.com