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EPIBULBAR OSSEOUS CHORISTOMA: A RARE ENTITY

Shanthala P. R¹, Shreesha Khandige².

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ABSTRACT: Choristoma of the eye are common epibulbar and ocular tumours in children, but osseous choristoma of the eye are very rare with few case reports being published in literature. We report one such case which presented in a 14 year female child who had small tiny nodule in the left eye present since birth with gradual increase in size. Excision biopsy was done. Histologically, nodule was composed of mature lamellated bone with a surrounding connective tissue.

KEY WORDS: Osseous choristoma, lamellated bone

INTRODUCTION: Choristomas are congenital lesions representing normal tissue in an abnormal location. Ocular choristomas are the most common epibulbar and orbital tumours in children. Epibulbar choristomas affect the cornea, limbus or subconjunctival space and may cause astigmatism. Occasionally these may be familial and may occur in association with Goldenhar syndrome or epidermal nevus syndromes.^[1] Four categories of ocular choristomas described are dermoid, dermolipoma, complex choristoma and single tissue choristoma. The osseous choristomas belong to the last category where in these contain only mature lamellated bone.^[2] Epibulbar osseous choristomas are rare, only about 65 cases of osseous choristomas have been reported in literature.^[3]

CASE REPORT: A 14 year old female child presented with tiny nodule at the temporal aspect of the left eye globe which was present since birth and had gradual increase in size. On ophthalmic examination of left eye, a lobulated firm epibulbar swelling with fleshy mass was seen extending to outer canthus. Rest of the ophthalmic examination findings like vision, refraction, and fundoscopy were normal in both eyes. The routine blood and urine investigations were within normal limits.

Provisional clinical diagnosis was dermoid or ectopic lacrimal gland. Per operatively, the nodule was found to be at superotemporal aspect of eye away from the limbus. Nodule was firm to hard in consistency with loose attachments to sclera. Excision biopsy was done. The excised nodule received macroscopically was, a single 5x3x2 mm white tiny nodule bony hard in consistency. The nodule was decalcified for study. Microscopically, the H&E stained sections of the decalcified nodule was composed of mature lamellated bone and fibrous connective tissue capsule on outer aspect. Thus, the diagnosis of osseous choristoma was made.

DISCUSSION: The epibulbar osseous choristomas are choristomatous lesion of conjunctiva containing only bone. Originally named as epibulbar osteoma, was first described by Von Grafe in 1863. Later Beckman and Sugar gave the nomenclature osseous choristoma in 1964 as they are presently known.^[4] The fascinating part of these lesions are their origin, they are said to arise from congenital nests of pluripotent mesoderm.

The lesions are typically noted at birth and may have potential for slow growth.^[5] These commonly are unilateral but rarely may be bilateral.^[6] The usual site of occurrence is the temporal

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quadrant or the superotemporal region of the eye, but may occur at other sites. Though commonly seen in children occurrence in older people have also been reported.^[7] The lesion in our case was unilateral with the typical site of occurrence, but had presented at a later age of 14 yrs. The osseous choristomas though are congenital lesions, a history of trauma may be given in some which drew the attention. Surgical excision may be indicated to improve vision, for cosmetic purpose or to impede growth. Some of these lesions may be adherent to the underlying sclera or rarely muscle and may complicate the excision procedure. In our case also the lesion was loosely attached to sclera which was released. A preoperative orbital imaging with CT scan or USG may be of help to rule out the adhesion.^[1,4]

Macroscopically most osseous choristomas are of small size < 8mm and are firm to hard in consistency. Histologically these contain mature lamellated bone with haversian systems and may be surrounded by a thin connective tissue capsule. Rarely some may have bony trabeculae with marrow elements.^[8] Clinically, differential diagnosis of the osseous choristomas is broad and includes dermoid, prolapsed orbital fat, papillomas, dermolipomas, and complex choristomas. Thus, in most cases the histopathological examination reveals the nature of the lesion.

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Fig 1. Microphotograph of the ocular lesion showing mature lamellated bone surrounded by fibrous connective tissue. (H&E 10 X)

AUTHORS:

1. Shanthala P. R,
2. Shreesha Khandige.

PARTICULARS OF CONTRIBUTORS:

1. Assistant Professor, Department of Pathology, Yenepoya Medical College.
2. Professor, Department of Pathology, Yenepoya Medical College

NAME ADDRESS EMAIL ID OF THE CORRESPONDING AUTHOR:

Shanthala P. R.,
Yenepoya Medical College,
Deralakatte, Mangalore – 575018.
Karnataka, India.
E-mail: bhatshanthala@yahoo.co.in

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