Limbal Complex Choristoma – A Rare Unexpected Lesion Diagnosed by Histopathology

Abstract

Complex choristoma contains a variety of tissues derived from more than one germ layer. Complex ocular choristoma is a very rare lesion. Very few cases have been reported so far. It may or may not cause visual abnormality depending on its location. Sometimes, it is associated with other congenital abnormalities. Here, we report a case of complex limbal choristoma which was diagnosed at adult age in a 58-year-old male patient and confirmed by histopathological examination.

Keywords: *Complex choristoma, histopathology, limbus*

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Introduction

The term "choristoma" depicts the presence of normal tissue at abnormal places.[1,2] A variety of choristomatous lesions have been reported previously such as dermoid, dermolipoma, osseous choristoma, and lacrimal choristoma in different components of eye.[1] However, complex choristoma is a very rare entity which contains a variety of tissues derived from more than one germ cell layer.[1] Limbal/epibulbar choristomas have been reported as congenital nonneoplastic lesions often associated with rare genetic syndromes such as organoid nevus syndrome, nevus sebaceous of Jadassohn, and Goldenhar syndrome.[3,4] However, complex choristoma with cartilage component has rarely reported in previous literature. We report a rare case of epibulbar complex choristoma diagnosed on histopathology examination of a resected ocular mass at the right eye in a 58-year-old male patient.

Case Report

A 58-year-old male patient presented with fleshy whitish mass at medial aspect of the right eyeball associated with foreign body sensation and watering. On ocular examination, it was a whitish fleshy mass at the medial aspect of the right limbal region measuring 0.8 cm × 0.5 cm [Figure 1]. The mass was fixed with sclera with a whitish smooth surface without any involvement of angle of the eye and palpebral conjunctiva. Visual acuity was 1/6 for the right eye and 3/6 for the left eye. The right eye had mature cataract and the left eye had immature cataract. On the right eye, cornea appeared normal and anterior chamber did not reveal any abnormality. Contralateral eye did not reveal any other significant abnormality except immature cataract. Intraocular pressure was normal in both eyes.

The mass was excised under local anesthesia and repaired by primary sutures. Specimen was sent for histopathology examination. Histopathology revealed a mass composed of mature cartilage and fibrocollagenous tissue below the conjunctival epithelium with small foci of well-differentiated acinar structures and ducts of lacrimal gland [Figures 2 and 3]. Histopathological diagnosis of complex choristoma of limbus was done. He had no postoperative complications and no recurrence was noted after 1-year follow-up.

Discussion

Complex choristoma is a very rare abnormality of eye which comprises more than two types of ectopic tissues.[1] It may be an isolated pathology or associated with various congenital or developmental abnormalities such as facial nevus of Jadassohn, Goldenhar syndrome, and bony cranial defects.[5] Presentation of complex limbal choristoma may be at birth or shortly after birth or in adult life. Most of the ocular choristomas involve epibulbar area, ocular adnexa, and choroid, but classical

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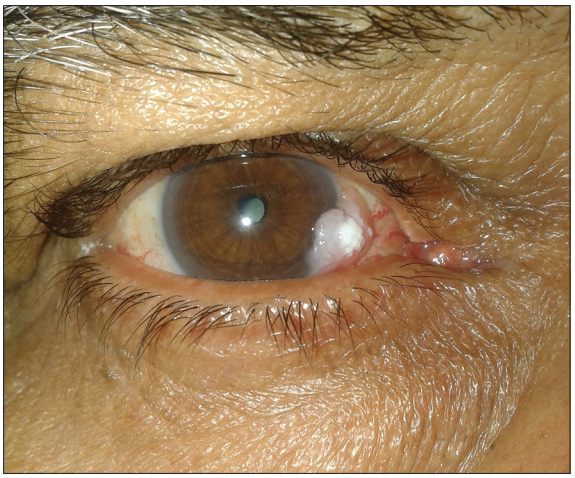

Figure 1: A whitish fleshy mass at medial aspect of right limbal region

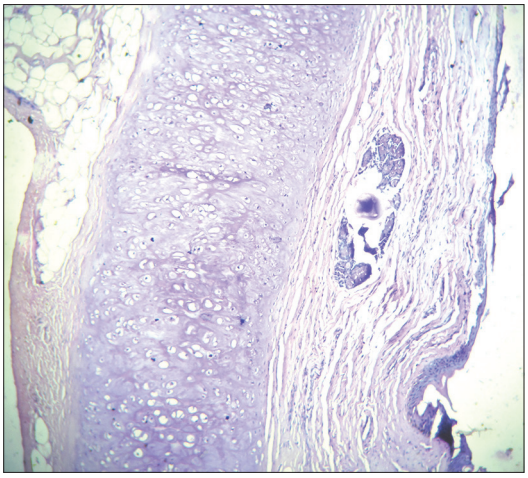

Figure 2: Photomicrograph of revealed a mass composed of mature cartilage and fibrocollagenous tissue bellow the conjunctival epithelium with small foci of well-differentiated acinar structures and ducts of lacrimal gland (low-power view, H and E)

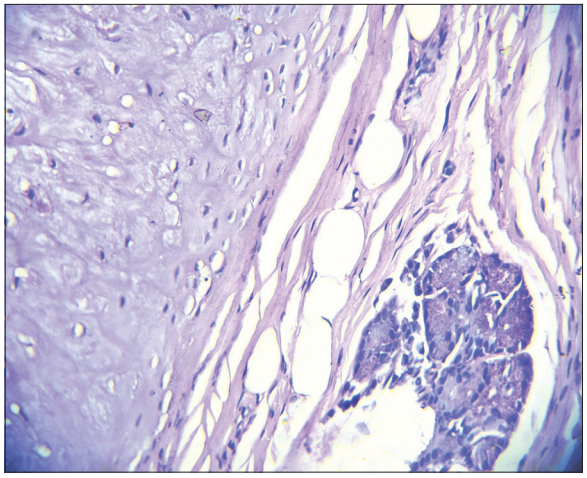

Figure 3: Mature cartilage and well-differentiated acinar structure (high-power view, H and E)

location is upper temporal region of the globe.[5] In our case, the lesion occurred at adult age without any associated neurological or developmental abnormality. In contrast, single-tissue choristoma contains ectopic neuroectodermal tissues (lacrimal gland, fat, nerve, brain tissue, bone, cartilage, and teeth) or dermis-like component.[5] Histologically, complex choristoma could be differentiated from dermoid cyst or dermolipoma by the presence of hair and adnexal structure in these lesions.[6] In the present case, the mass contains cartilaginous element, lacrimal gland tissue, fibrocollagenous tissue, and adipose tissue simultaneously. Exact pathogenesis of complex choristoma is still unknown. A hypothesis of defect in cell migration of pluripotent cells during embryonic development had been postulated and supported by subsequent authors.[3] Another hypothesis is defective differentiation due to toxic or infectious causes.[3] Clinical presentation of ocular choristoma depends on the site of involvement, size of choristomatous tissue, and growth rate of it. Mass-forming choristoma at conjunctiva does not cause significant visual abnormality. However, choristoma at corneal limbus region causes amblyopia, diplopia, irregular astigmatism, and increased intraocular pressure.

Treatment of complex choristoma depends on the size, location, and mechanical effect of the lesion. In general, wide excision of the lesion with closure is the treatment of choice.[1,7] Sometimes, keratoplasty (when cornea is involved) and repair with amniotic membrane may be necessary.[1,7]

Conclusion

Diagnosis of complex choristoma is histopathological, and it indicates the need of thorough examination of the tissue and multidisciplinary approach by neurologist, otorhinolaryngologist, dermatologist, and ophthalmologist to detect other congenital abnormalities.

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.

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