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Epibulbar complex choristoma including lacrimal gland, cartilage, and adipose tissue

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Abstract

Choristomas are congenital lesions which appear in abnormal regions, preserving their size and shape over years. Choristomas are named according to their tissue content: those that include more than one tissue are called complex choristomas and constitute only a small proportion of all limbal dermoids. In this case, a 9-month-old child presented with a limbal dermoid on the superotemporal aspect of his right eye. The dermoid was surgically excised because he was at risk of anisometric ambylopia due to the induced high astigmatism. Pathological examination demonstrated a complex choristoma consisting of cartilage, lacrimal gland, and mature adipocyte tissue. Complex choristomas are important in differential diagnosis of limbal dermoid.

Introduction

Choristomas are congenital lesions characterized by ordinary tissue present at aberrant locations with nearly no developmental potential.[1] Choristomas are classified based on their tissue content. Choristomas are defined as complex when numerous tissues, such as lacrimal gland, bone, cartilage, nerve, muscles or brain, are present.[1] On the ocular surface, complex choristoma may penetrate the cornea and can appear cystic.[2] The clinical appearance of epibulbar complex choristomas may vary in location and extent, ranging from localized forms to lesions that cover whole surface of the eye.[2] In this case report, a 9-month-old child presented with epibulbar complex choristoma with ectopic lacrimal gland, cartilage and adipose tissue.

Case Report

A 9-month-old boy was presented by his parents to the local hospital, complaining of a white lesion present at birth in his right eye which has grown substantially since birth. There was no family history of ocular abnormality or disease. Neither were there any complications with his mother's pregnancy nor birth.

Upon examination, a limbal dermoid was observed involving the superotemporal aspect of the right bulbar ocular surface from the 8 o'clock to 11 o'clock position and extending into the peripheral cornea. No abnormality was found during dilated fundoscopy.

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Conflict of Interest: The authors report no conflicts of interest.
Contributions: All authors contributed equally.
Accepted for Publication: April, 2019

Excision of the limbal dermoid was performed under general anesthesia out to preclude the possibility of anisometropic amblyopia due to the induced high astigmatism in his right eye. The main site of the incision was the superotemporal conjunctiva on the patient's right eye. Removal of corneal portion of the dermoid was performed with a crescent blade while protecting underlying cornea. The conjunctival surface was closed by slippage of the nearby healthy conjunctiva and closure with 8-0 interrupted vicryl sutures.

Post-operatively, a therapeutic bandage contact lens was applied to promote epithelization of the corneal surface, which occurred during the first post-operative week. No peri-operative or post-operative complication were observed.

The excision lesion was sent for pathological examination. The excised yellowish specimen, measuring 0.7 x 0.6 x 0.3 cm was fully sampled. Histopathologically examination disclosed lacrimal gland tissue with acini filled with zymogen granules associated with adipocytes in the connective tissue below the conjunctival epithelium. Well-developed chondroidal tissue enclosed by a perichondrium was present. No sebaceous glands or hair follicles were observed in cross-sections. The contents of excision material were confirmed to be a conjunctival / limbal complex choristoma including of ectopic lacrimal gland, mature cartilage, and mature adipose tissue (figure 1).

Discussion

A significant majority of complex choristomas with ectopic lacrimal tissue are unilateral and affect the peripheral cornea, bulbar conjunctiva, and limbus.[3] The majority of complex choristomas are congenital lesions with no immature component, and these lesions tend to retain their size and shape. The temporal portion of the globe is the primary region of complex choristomas.[4] In a reported large series of 261 cases of limbal dermoid (154 females and 107 males with a mean age of 4 ± 3 years), 124 cases were dermoid choristomas, 76 were lipodermoid choristomas, 10 were epibulbar osseous choristomas, and only 5 of them were complex choristomas.[5]

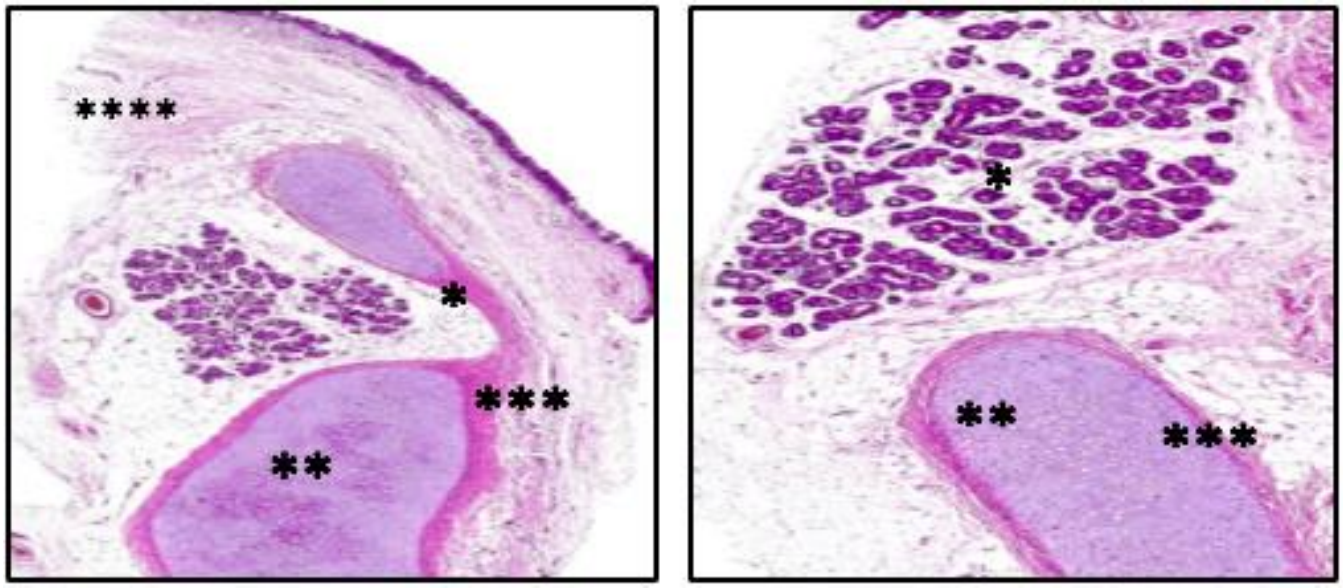

Figure 1

Hematoxylin and eosin stained sections of the limbal dermoid demonstrated a complex choristomas with well-developed glandular(*), chondroidal(**), & adipose(***) tissues within local connective tissue below conjunctival epithelium(****).

Complex choristomas are easily identified on pathologic evaluation. This specific type of complex choristomas, epibulbar complex choristoma including lacrimal gland, cartilage, and adipose, is seen rarely. A similar case of epibulbar complex choristoma has been previously reported though amblyopia developed as the surgical management required a lamellar keratopasty.[6] Though it is rare, complex choristoma should not be left out of the differential diagnosis of limbal dermoid.

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