Case Report

Ectopic lacrimal gland in a complex choristoma involving the lacrimal sac fossa

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Abstract

Complex choristomas involving the lacrimal drainage system are not common. Ectopic lacrimal gland within complex choristomas has been described in the eyelid and orbit. We describe a pediatric case of complex choristoma with ectopic lacrimal gland involving the lacrimal sac fossa and partly the medial eyelid and orbit along with a brief review of literature.

Keywords: Choristoma, Ectopic, Lacrimal gland, Lacrimal sac

Introduction

Choristomas are congenital lesions where histologically normal tissues with little or no growth potential are present at abnormal locations. Choristomas can be termed complex when multiple tissues such as lacrimal gland, bone, cartilage, nerve, muscles or brain are present. Choristomas with ectopic lacrimal gland have been reported in eyelids, bulbar conjunctiva, limbus, orbits and intraocular tissues. We describe a male child, 2 years of age with ectopic lacrimal gland in complex choristomas involving the lacrimal sac fossa and partly the medial eyelid along with a brief review of literature.

Case report

A male child, 2 years of age presented to us with complaints of right sided epiphora and swelling at the inner corner of the right eye since birth (Fig. 1A and B). Antenatal and perinatal history was normal and there was no past history of trauma or any systemic disease. Magnetic resonance imaging performed at the age of 6 months showed a soft tissue lesion in the lacrimal fossa with extension into the infero-medial orbit (Fig. 1C). The patient had an inferior limbal dermoid, which was excised elsewhere at the age of 1 year along with an incision biopsy of the medial mass. No further details were available. On examination there was a large, soft mass below the medial canthus involving the medial half of the lower eyelid and causing canthal dystopia (Fig. 1A). An overlying cutaneous scar was noted secondary to the past incision biopsy (Fig. 1A). A central lower lid symblepharon was noted with corneal scarring (Fig. 1B). Ultrasound B-scan showed normal intraocular contents. Examination of the lacrimal system under the operating microscope along with irrigation and probing showed the right lower lacrimal punctum and canaliculus to be absent and an upper distal canalicular block. The left eye and lacrimal system were normal. A clinical impression of right possible choristoma was made and the patient was advised for an exploration and debulking. The past scar was used for incision. Intraoperatively, the lesion was found to be predominantly composed of fat. Upon
reflecting the periosteum over the anterior lacrimal crest, the entire lacrimal sac fossa was involved with similar tumor with no anatomical delineation of the sac (Fig. 1D). The entire lesion in the lacrimal sac fossa could be removed (Fig. 1D).

Histopathological evaluation showed the lesion to be predominantly consisting of adipose tissue with intervening glandular and muscular tissues (Fig. 1E). The lacrimal fossa tissues showed clusters of adipocytes separated from large islands of acinar tissues by fibrocollagenous strands. These islands of acini were forming lobules and the acinar cells showed numerous secretory granules within their cytoplasm (Fig. 1F). Histopathological examination was suggestive of a complex choristoma with ectopic lacrimal gland. The child would be closely followed up for recurrences and a plan has been discussed with parents regarding future lacrimal rehabilitation in the form of endoscopic minimally invasive conjunctivodacryocystorhinostomy.

Discussion

Complex choristomas with ectopic lacrimal gland have been noted to have a predisposition to bulbar conjunctiva and cornea, although it has been reported from eyelids, orbits and intraocular tissues such as ciliary body and iris.1–5 Numerous theories have been proposed to explain the occurrence of choristomas and ectopic lacrimal gland and include aberrant implantation of embryonic cells destined to differentiate into lacrimal gland and aberrations during closure of fissures and sutures.5

Alyahya et al.1 studied 61 cases of ectopic lacrimal glands over a period of 50 years and found that 62.2% (38/61) of these were associated with some forms of complex choristomas. 81% of all the lesions were located at the temporal epibulbar conjunctiva. Only two of the 61 cases were correctly diagnosed preoperatively. Ferri et al.2 reported a case of an inferomedial subconjunctival dermolipoma with ectopic lacrimal gland and concluded that the location was quite atypical based on the literature review. Verb et al.5 described a phakomatous choristoma presented as a lower eyelid mass with a mechanical nasolacrimal duct obstruction. However, since this was a phakomatous choristoma, it did not have any ectopic lacrimal gland tissues. The current case however showed the choristoma with ectopic lacrimal gland to completely involve the lacrimal sac and nasolacrimal duct beyond anatomical delineation.

In summary we presented a case of a 2 year old male child with a medial canthal lesion and exploration revealed involvement of the lacrimal drainage system with a complex choristoma having an ectopic lacrimal gland tissue. It is important for the Oculoplastic surgeons and Pediatric Ophthalmologists to be aware of a possibility of ectopic lacrimal gland within the lacrimal sac fossa. Lacrimal drainage rehabilitation in these cases can be very challenging.

Conflict of interest

The authors declared that there is no conflict of interest.

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References

1. Alyahya GA, Bangsgaard R, Prause JU. Occurrence of lacrimal gland tissue outside the lacrimal fossa: comparison of clinical and histopathological findings. Acta Ophthalmol Scand 2005;83:100–3.
2. Ferri S, Shinder R. Inferomedial dermolipoma with ectopic lacrimal gland. *Ophthal Plast Reconstr Surg* 2013;29:e43–4.
3. Gordon AJ, Patrinely JR, Knupp JA, Font RL. Complex choristoma of the eyelid containing ectopic cilia and lacrimal gland. *Ophthalmology* 1991;98:1547–50.
4. Verb SP, Roarty JD, Black EH, et al. Phakomatous choristoma: a rare orbital tumor presenting as an eyelid mass with obstruction of the nasolacrimal duct. *J AAPOS* 2009;13:85–7.
5. Jung JY, Kim JH, Kim ST, et al. MR features of intraocular ectopic lacrimal tissue. *Am J Neuroradiol* 2006;27:2196–8.