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Congenital tooth-bearing tumor of the eyelid leading to lacrimal system obstruction

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ABSTRACT

Purpose: We present an unusual case of a congenital lesion presenting with concomitant chronic dacryocystitis. The clinical presentation, examination, management, and histopathology are reviewed.

Observations: A healthy male infant born at 37 weeks gestation presented with an isolated painless 5mm congenital mass of the left medial lower eyelid. Parents also reported episodic epiphora and discharge from the left eye. A surgical excision of the mass revealed an underlying dacryocystitis and the presence of a formed tooth. A dacryocystorhinostomy was performed together with a repair of the soft tissue defect. Histopathology revealed components of disorganized epithelial and mesenchymal tissues including a tooth, skeletal muscle, fat, fibrous tissue, nonkeratinized epithelium, and myelinated nerves. A diagnosis of an odontogenic choristoma of the eyelid was made. Furthermore, a lacrimal sac culture was positive for oxacillin-susceptible *Staphylococcus aureus* with pathological evidence of chronic dacryocystitis.

Conclusions and Importance: Odontogenic choristoma is a very rare finding in the periocular region with only a few cases reported in the literature. Awareness of clinical findings from this case may allow for a more accurate clinical diagnosis and understanding of the embryologic mechanisms underpinning eyelid and nasolacrimal development. Timely management of this condition is critical to ensure normal oculofacial development and prevent future complications.

1. Introduction

Although eyelid masses are clinically common in the pediatric population, congenital masses of the palpebral region are rare. When located medially, these tumors may lead to obstruction of the lacrimal drainage system. Excluding hordeolums and chalazia, epidermal cysts and dermoid cysts have been found to be the most common pediatric eyelid tumors.[1] However, the differential diagnosis may include meningoencephaloceles,[2] capillary hemangiomas,[3] sudoriferous cysts,[4] nasal gliomas,[5] dacryocystoceles,[6] dacryocystitis,[7] rhabdomyosarcomas,[8] nodular fasciitis,[9] and phakomatous choristomas.[10]

Three cases of palpebral tumors[11–13] and eight cases of orbital tumors[14–21] containing teeth have been previously described, with only five in the English literature.[15,17,19–21] Although these lesions were initially suggested to be teratomas, the pathologic classification of these tumors is not defined with certainty. We report a congenital, infra-nasal, palpebral mass containing a tooth with obstruction of the nasolacrimal system and dacryocystitis.

While tooth-bearing masses have been well-documented in the orbit and elsewhere, silent palpebral odontogenic choristomas concomitant with chronic dacryocystitis have not been previously described. This very unusual case can not only help clinicians recognize the clinical features of this condition but also allow for a better understanding of the embryological processes involved in orofacial development.

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2. Case report

2.1. Patient history

The patient, a healthy male infant presented with a congenital mass of the left lower eyelid, noted upon delivery via cesarean section at 37 weeks of gestation. On initial physical exam, the lesion appeared as a 5 mm subcutaneous pedunculated mass inferior to the punctum of the left lower eyelid (Fig. 1). Occasional mild epiphora and discharge were noted by the parents. The lesion extended 3 mm superiorly beyond the lid margin raising a concern of amblyopia by the referring pediatric ophthalmologist. A partially mucosalized demarcation line could be noted on the superior surface. The remainder of the eyelid structure appeared intact including the inferior punctum and medial canthus. Pupils were equal, round and reactive to light, and no afferent pupillary defect was noted. Fundus examination was unremarkable. The cornea was clear, sclera, and conjunctiva were intact. Extraocular movements were full and the orbits were symmetric. No abnormalities were noted in the contralateral eye. The intraoral exam was normal and the rest of the physical exam was not significant. Six weeks after the initial evaluation, the mass remained grossly unchanged although the parents reported episodic epiphora and discharge. To address the lacrimal system, improve aesthetics, and decrease the risk of amblyopia, the decision was made to excise the mass and explore the nasolacrimal passages.

2.2. Operative findings

After endotracheal intubation, dilation of the upper and lower puncta was performed. Irrigation of the nasolacrimal passages was performed revealing complete obstruction as well as purulent reflux. After injection of local anesthetic, the eyelid mass was excised at its base. Deep to the wound bed, a formed tooth was discovered and easily freed from the surrounding tissue (Fig. 2). A purulent cavity was noted medial to the tooth and the surrounding tissue appeared to be fibrotic and thickened. Any visibly abnormal tissue was excised from the wound and sent for histopathology. At this point, a standard dacryocystorhinostomy was performed with mucosal anastomosis over bicanalicular silicone Crawford stents. No other anomalies were noted within the visible nasal passage. The eyelid defect was closed in layers without requiring any tissue advancement. The patient was discharged home the same day on oral antibiotics.

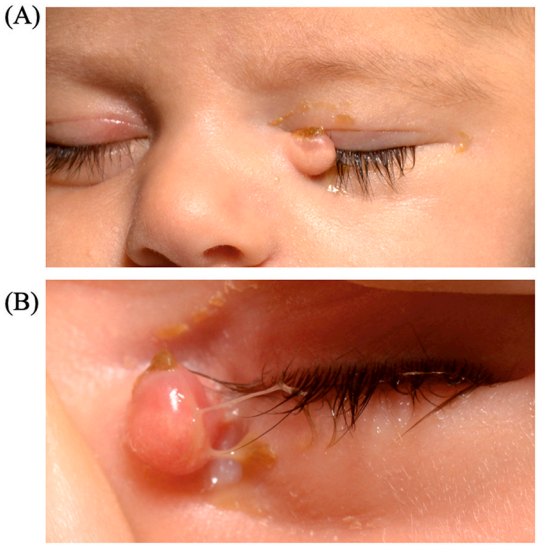

Fig. 1. (A) A 5 mm subcutaneous pedunculated mass inferior to the punctum of the left lower eyelid. (B) Close-up of the lesion reveals a pedunculated attachment to the eyelid as well as mild epiphora and discharge.

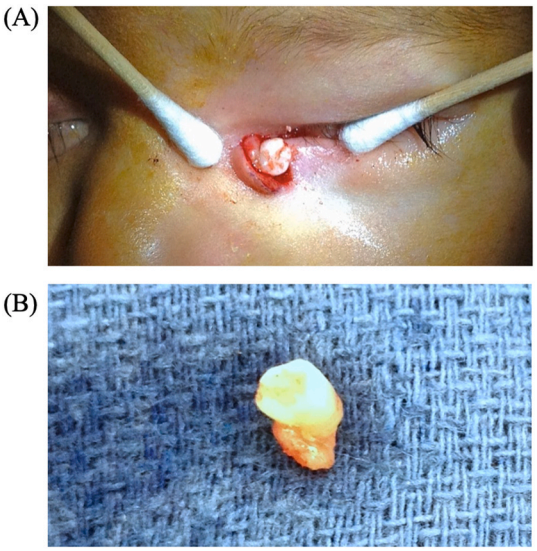

Fig. 2. (A) Intra-operative findings reveal the presence of a tooth, distortion of normal eyelid structures, and chronic dacryocystitis. (B) Gross pathology of the tooth recovered from the mass.

2.3. Post-operative course

At one week following surgery, the patient was healing well with no evidence of dacryocystitis or epiphora. The lacrimal sac culture was positive for oxacillin-susceptible *Staphylococcus aureus* with pathological evidence of chronic dacryocystitis. Pathologic examination of the mass revealed components of disorganized epithelial and mesenchymal tissues including a tooth, skeletal muscle, fat, fibrous tissue, nonkeratinized epithelium, and myelinated nerves (Fig. 3). Based on the histological analysis of the tumor, a diagnosis of odontogenic choristoma was made. There was a 2-week and 2-month postoperative follow-up with the patient with no signs of recurrence, an unremarkable physical exam, and no complications (Fig. 4).

3. Discussion

The case presented here is of a very rare eyelid mass featuring a tooth and chronic dacryocystitis in an infant. The histologic diagnosis of this lesion is more consistent with an odontogenic choristoma than a teratoma. Teratomas are defined histologically as congenital tumors consisting of disorganized tissue from all three germ layers: ectoderm, mesoderm, and endoderm. If only two layers are present, some have described these lesions as "teratoid".[13] Teratomas may be found in various locations. Extragonadal teratomas derive from pluripotent germ cells that fail to migrate to the gonadal ridge during the 4th and 5th week of gestation. The germ cells undergo abnormal maturation cues due to their dislocation, and their pluripotent status gives them the potential to mature into any adult or fetal tissue. Hence, they are often characterized as having all three embryonic germ layers.[22] Sacrococcygeal teratomas are the most common, representing 59–65% of incidences, and head and neck teratomas occur in 5% of cases.[23,24]

Choristomas, by comparison, are congenital tumors of normal tissue in abnormal (ectopic) locations. Jakobiec et al. proposed that the few described periorbital teeth-containing tumors are actually odontogenic choristomatous displacements.[13] Two possible embryonic explanations are offered: (1) a displaced tooth bud or anlage may become lodged in the first branchial arch without any embryonic oral epithelium, or (2) multipotent oral epithelium may become displaced within the mesenchyme leading to tooth development. The latter is the favored theory. This theory can explain several differences between the presentation of these lesions as compared to true teratomas, including more mature

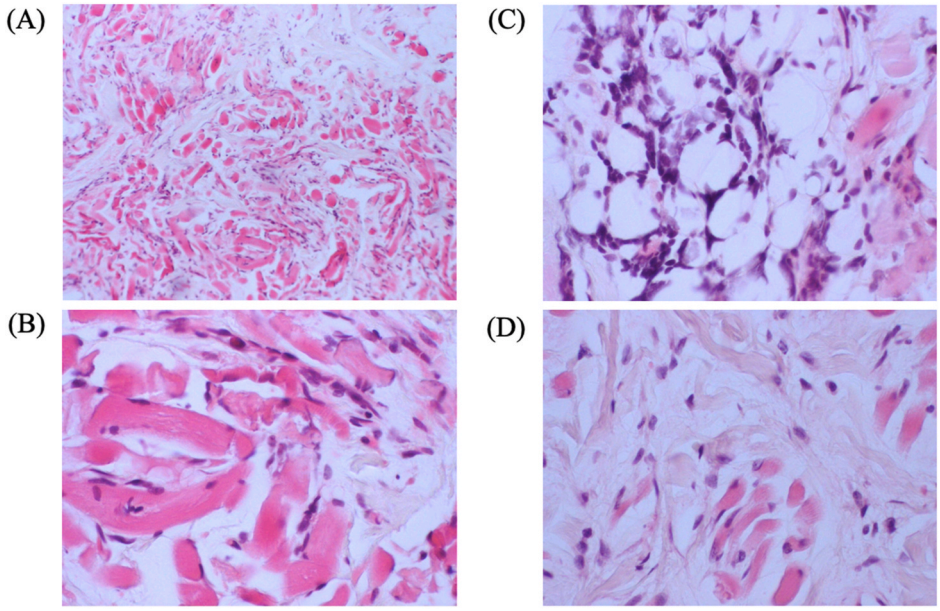

Fig. 3. (A, B) Histopathology findings of left medial canthal lesion at low (A) and high (B) magnification (hematoxylin-eosin stain) reveal skeletal muscle, fat, fibrous tissue, nonkeratinized epithelium, and myelinated nerves. (C, D) Histopathology findings of the posterior lacrimal sac at low (C) and high (D) magnification (hematoxylin-eosin stain) reveal a small fragment of nonkeratinized epithelium overlying a fibrovascular response with many lymphocytes.

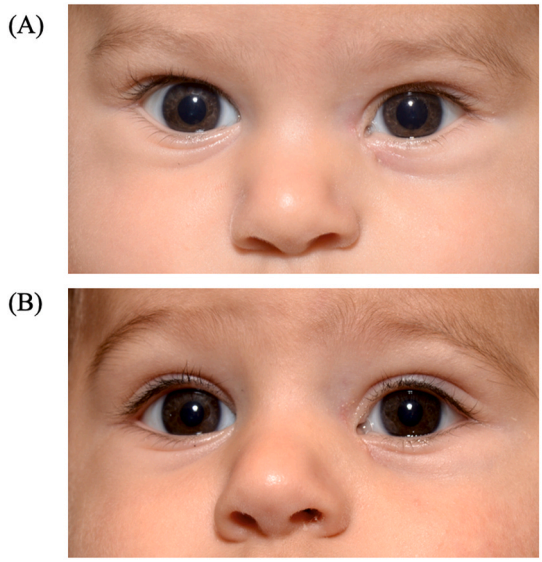

Fig. 4. Postoperative follow-up at 2 weeks (A) and 2 months (B).

tooth formation, lack of all three germ layers, and differences in clinical presentation, such as mild eyelid edema. The presentation and pathology of our patient are in line with the patients discussed by Jakobeic et al. with findings of mature tooth formation, mild eye symptoms, and lack of all three germ layers in the specimen. In our case, however, the extent to which the choristoma invades the palpebral tissue seems to be more limited. The presence of a nasolacrimal sac obstruction and dacryocystitis may be due to an abnormal lacrimal sac development or compression of an otherwise normal lacrimal sac leading to tear stasis and secondary infection.

In our patient, no pre-operative imaging was performed. Due to the location of the lesion and the lack of other ocular or orbital pathology, intracranial and intraorbital extension was deemed unlikely. Other authors have suggested that computed tomographic (CT) scan or dacryocystography are beneficial for surgical planning, to decrease the risk of lacerating the canaliculi during excision of a medial canthal mass.[25] However, even with the help of preoperative imaging, the lacrimal system is still susceptible to injury that may necessitate repair.[26] In general, MRI has been suggested as the preferred modality when assessing for intracranial extensions, while CT is preferred for evaluating lesions of the nasolacrimal duct system.[27] Imaging may reveal the presence of calcifications within a heterogeneous mass suggestive of a teratoma.[28] Preoperative imaging of congenital palpebral masses may be beneficial in the setting of significant anatomical distortion or if there is a concern for intracranial or intraorbital extension. Otherwise, it is best to avoid radiation exposure in the developing child.[29]

Complete excision is the standard of care for teratomas and choristomas, with recurrences mostly noted for incompletely excised lesions.[30] Challenges with eyelid mass excisions include avoiding iatrogenic lacrimal system injury and potential dysfunction of the tear flow or eyelid function, especially in a pediatric patient. Kim et al. advocate using silastic intubation of both canaliculi during the excision to protect from inadvertent injury.[25] Indeed, silastic intubation can alert the surgeon to an iatrogenic injury that might otherwise go undiagnosed. In our patient, after completely excising the lesion, the canaliculi were probed and confirmed to have no laceration.

4. Conclusion

This case report describes the rare palpebral odontogenic choristoma with concomitant chronic dacryocystitis. Awareness of the topographic and clinical findings from this case may allow for a more accurate clinical diagnosis and understanding of the embryologic mechanisms underpinning eyelid and nasolacrimal development. Timely management of this condition is necessary to ensure normal oculofacial development and prevent future complications.

Patient consent

The patient's legal guardian consented to publication of the case in writing.

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Authorship

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Potential conflict of interest exists

We wish to draw the attention of the Editor to the following facts, which may be considered as potential conflicts of interest, and to significant financial contributions to this work:

The nature of potential conflict of interest is described below

No conflict of interest exists.

We wish to confirm that there are no known conflicts of interest associated with this publication and there has been no significant financial support for this work that could have influenced its outcome.

Intellectual property

We confirm that we have given due consideration to the protection of intellectual property associated with this work and that there are no impediments to publication, including the timing of publication, with respect to intellectual property. In so doing we confirm that we have followed the regulations of our institutions concerning intellectual property.

Research ethics

Written consent to publish potentially identifying information, such as details or the case and photographs, was obtained from the patient(s) or their legal guardian(s).

Contact with the editorial office

Someone other than the Corresponding Author declared above submitted this manuscript from his/her account in EVISE: Ebrahim Afshinnekoo <ebrahim_afshinnekoo@student.nymc.edu>

We understand that this author is the sole contact for the Editorial process (including EVISE and direct communications with the office). He/she is responsible for communicating with the other authors, including the Corresponding Author, about progress, submissions of revisions and final approval of proofs.

Declaration of competing interest

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