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Congenital conjunctival cyst detected by prenatal ultrasound

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ABSTRACT

Purpose: The purpose of this manuscript is to report a rare case of an orbital cyst detected intrauterine with sonography.

Observation: A 23-year-old female presented for routine prenatal monitoring when an orbital cyst was detected with a transabdominal ultrasound. Uncomplicated cesarean section was performed at 38 weeks gestation with proptosis of the left globe being noted on ophthalmic examination of the newborn. Magnetic resonance imaging was conducted which demonstrated a cystic mass suspicious for lymphatic malformation. Surgical intervention was performed shortly after birth with pathological results demonstrating non-keratinizing squamous epithelium consistent with conjunctivoid variant of dermoid cyst. Post-operative examination demonstrated significant improvement of the exophthalmos with minimal lagophthalmos.

Conclusions: Congenital orbital cysts are a rare phenomenon with only a handful of cases being detected during the fetal period. We recommend physicians include this rare variant of dermoid cysts in their differential diagnosis for orbital lesions that can cause proptosis in the fetal period.

Prenatal ultrasound is a useful tool for early detection of many structural ophthalmic anomalies such as hypertelorism, dacryocystocele, septo-optic dysplasia, microphthalmia, anophthalmia, and cataracts.[1] Congenital orbital cysts are rarely diagnosed in the intrauterine period with only a handful of cases reported in the literature.[2–4] Prompt diagnosis is key as orbital cysts have the potential to cause irreversible vision loss and cosmetic disfigurement. The differential diagnosis of congenital orbital cysts include teratomas, colobomatous cyst, and lymphatic malformation.[5] We present a case of a congenital conjunctival dermoid cyst that was detected with prenatal ultrasound during the 30th week of gestation. To our knowledge, this is the fourth case of conjunctival dermoid cysts detected in utero.[2–4] This manuscript adhered to the ethical principles outlined in the Declaration of Helsinki as amended in 2013 and is Health Insurance Portability and Accountability Act (HIPAA) compliant. The patient's guardian gave consent for identifiable photograph.

1. Case presentation

A 23-year-old healthy female in her 30th week of gestation presented to the obstetrics and gynecology clinic for a routine transabdominal ultrasound which demonstrated a left orbital cystic mass measuring 36 x 33 × 18 mm. A follow-up ultrasound was performed 4 weeks later which demonstrated a stable cystic mass (Fig. 1). Cesarean section was performed at 38 weeks of gestation without any complications. Magnetic resonance imaging (MRI) of the brain and orbits was performed one day after delivery for further evaluation of the cystic mass. The MRI findings demonstrated a 3.4 x 2.7 × 2.1 cm lesion with intraconal and extraconal components most suspicious for a lymphatic lesion as well as a possible decreased size of the left optic nerve (Fig. 2). Examination under anesthesia (EUA) was conducted when the newborn was 1 week old.

External exam demonstrated severe exophthalmos with inability to close the left eye (Fig. 3). Portable slit lamp exam demonstrated increased horizontal length of the bilateral upper and lower eyelids with significant conjunctival injection and diffuse punctate epithelial erosions in the left eye. The anterior chamber appeared deep and quiet in the right eye but slightly shallow in the left eye with dilated peripupillary vessels on the iris. The lens appeared clear in both eyes. Dilated fundus examination demonstrated 0.1 c/d ratio bilaterally with no evidence of optic nerve head edema or pallor. An inferolateral orbitotomy

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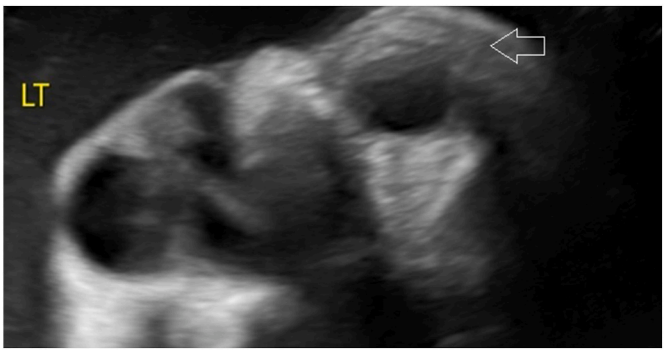

Fig. 1. Intrauterine ultrasound at the 34th week of gestation demonstrating a left orbital cystic lesion (arrow).

was then performed at the same time as the EUA by incising the conjunctiva over the lesion in the inferolateral fornix. Subconjunctival dissection was performed 360° around the anterior aspect of the lesion dissecting the conjunctiva off, allowing anterior access to the lesion. Further dissection into the orbit was performed. At this point, given the lesion was cystic on imaging, incision was made into the anterior aspect of the lesion and immediate egress of green-yellow fluid was noted (Fig. 4). The cystic lesion subsequently collapsed and the external wall of the cyst was dissected from surrounding orbital tissues for the anterior 75 % of the lesion. Dissection was not performed for the most posterior aspect given its close proximity to the optic nerve. The dissected portion of the cyst wall was then excised and sent for pathology leaving just the most posterior aspect of the lesion behind. The cystic fluid was sent for culture which demonstrated no growth. Pathology of the cyst wall demonstrated a benign cyst with non-keratinizing squamous epithelium and focal cuboidal epithelium consistent with conjunctivoid variant of dermoid cyst.

On postoperative day one, ophthalmic examination was remarkable for 2 mm lagophthalmos in the left eye. Exophthalmos was significantly improved but not completely resolved. Aggressive lubrication was advised.

The patient was lost to follow-up until postoperative week ten. At this time, the patient's mother reported that the left eye varied from being completely closed to partially open. She admitted to non-compliance with the lubricating ointment. Cycloplegic retinoscopy demonstrated +2.00 sphere on the right and −3.00 sphere on the left. Anterior segment examination demonstrated pupils reactive to light with grossly full extraocular movements. Intraocular pressure was soft to palpation in both eyes. An inferior cornea scar was noted with diffuse punctate epithelial erosions in the left eye (Fig. 5).

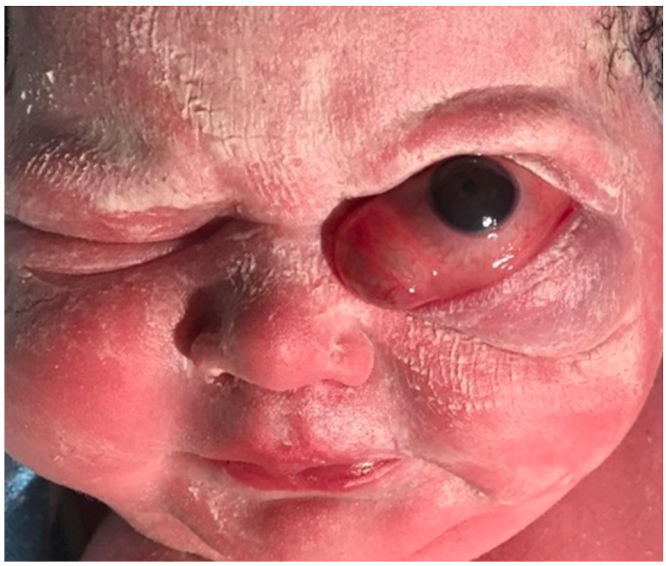

Fig. 3. External ophthalmic examination one week after birth demonstrating the cystic lesion inferiorly and nasally with significant proptosis.

2. Discussion

Dermoid cysts are the most common orbital tumor in the pediatric population.[6] Typical dermoid cysts are lined by keratinized, stratified squamous epithelium similar to that of normal skin. However, in rare instances they can be lined by nonkeratinized epithelium or cuboidal epithelium with or without goblet cells resembling conjunctiva which was first reported by Jakobiec et al.[7] Beyond histological differences, cutaneous and conjunctival dermoid cysts can also differ in age of presentation and location. Cutaneous orbital dermoid cysts typically present in the first decade of life, whereas conjunctival dermoid cysts are often found in young adults with the average age at presentation of 24 years.[8] In terms of location, cutaneous orbital dermoid cysts are predominantly found in the superotemporal region versus the nasal predisposition of conjunctival cysts. Conjunctival dermoid cysts are thought to be secondary from developmental sequestration of the epithelium during the neonatal period; however, despite their theorized congenital origin, most conjunctival cysts are acquired following trauma or surgery.[8]

The first case of prenatal ultrasound detection and monitoring of a congenital orbital cyst was described in 2001 by Yen et al.[2] In this case, a left orbital cyst was detected during the 23rd week of gestation and surgically excised shortly after birth, sparing the globe. Pathological

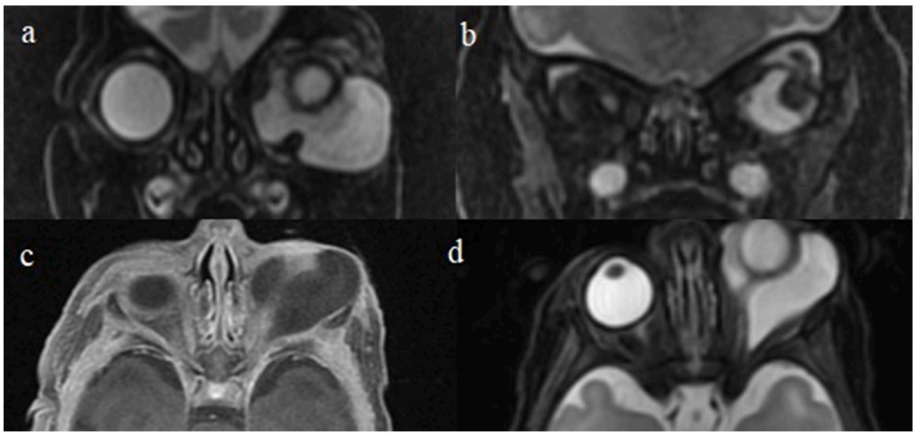

Fig. 2. Magnetic resonance imaging (MRI) orbits illustrating a left orbital cystic mass one day after delivery. A and B: coronal images; C and D: axial images.

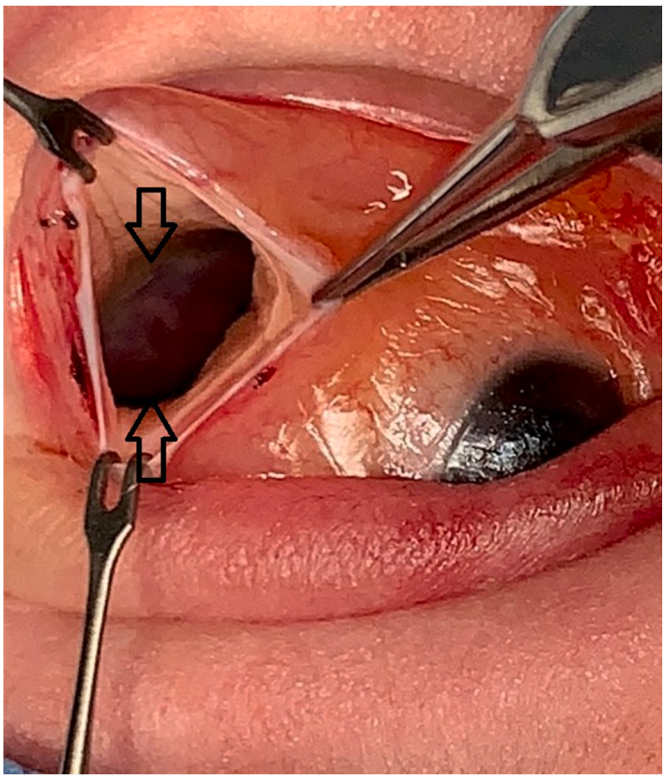

Fig. 4. Intraoperative photo demonstrating the cystic cavity at newborn age one week. Note arrows highlighting cyst borders inferiorly and superiorly.

examination demonstrated findings consistent with a conjunctival cyst. Postoperatively, the patient wore a scleral shell with good motility. Since this initial case, there have been two more reports to date of congenital conjunctival cysts being detected by prenatal ultrasound.[3,4] Singh et al. describe a case of a right orbital cyst detected during the 27th week of gestation.[4] Fetal MRI confirmed the cystic mass and surgical intervention was performed shortly after birth following a planned Cesarean section at the 38th week of gestation. Anterior orbitotomy was performed which demonstrated a benign cyst lined by squamous epithelium consistent with the diagnosis of a conjunctival cyst. At the third postoperative month, minimal proptosis was noted without evidence of exposure keratopathy. Topaloğlu et al. reported the most recent case of a left orbital cyst being detected during the 22nd week of gestation.[3] Similar to Singh et al., fetal MRI was used to further characterize the cystic lesion. Globe-sparing surgical intervention was performed after birth with pathology results demonstrating findings consistent with an conjunctival cyst. To our knowledge, our patient is the fourth reported case of congenital conjunctival cyst detected in utero.

Globe-sparing surgical intervention is the preferred management for orbital dermoid cysts. If detected in utero, we recommend close coordination with obstetrics, radiology, and ophthalmology to allow prompt diagnosis and surgical intervention within the first several weeks of birth. If not addressed in the early newborn period, it could lead to poor visual outcome from optic neuropathy or exposure keratopathy in the setting of proptosis. Surgery can be complicated if a cyst ruptures due to the inflammatory contents; however, this may sometimes be unavoidable and can be mitigated with copious irrigation. Additionally, if the dermoid is not completely excised, the remaining tissue may result in an orbitocutaneous fistula from chronic inflammation.[9]

The etiology of dermoid cysts remains mostly unknown. Of the reported cases involving prenatal detection, there have been no associated systemic manifestations. Therefore, these lesions seem to represent an isolated event. Preoperative management should be focused on correctly identifying the type of lesion and ruling out other etiologies of a neonatal orbital mass such as orbital teratoma, lymphatic malformation, meningocele, and dacryocystocele. Surgical intervention is then undertaken for prevention of secondary complications. Similar to the three previously reported cases, we opted to monitor the lesion throughout pregnancy and planned on early surgical intervention shortly after birth. Unlike those cases, however, our case is unique in that we elected not to perform complete excision given the proximity to the optic nerve. Attempting complete excision could risk optic nerve compromise, resulting in catastrophic vision loss. Our patient had a favorable surgical outcome with significant improvement of exophthalmos and minimal lagophthalmos without developing an orbitocutaneous fistula. This case demonstrates that partial excision is a viable option for more posteriorly located cysts. Despite their rare occurrence, conjunctival dermoid cysts should remain on clinician's differential for orbital lesions in the fetal and neonatal period.

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CRediT authorship contribution statement

Ryan Gabbard: Writing – review & editing, Writing – original draft. **Hunter Harrison:** Writing – review & editing, Writing – original draft. **Kenneth Chang:** Writing – review & editing, Writing – original draft. **Elizabeth Pogrebniak:** Writing – review & editing, Writing – original draft. **Katie Keck:** Writing – review & editing. **Rakesh M. Patel:** Writing – review & editing.

Patient consent:

Consent to publish the case report was obtained. Additionally, patient gave consent to publish an identifiable photograph.

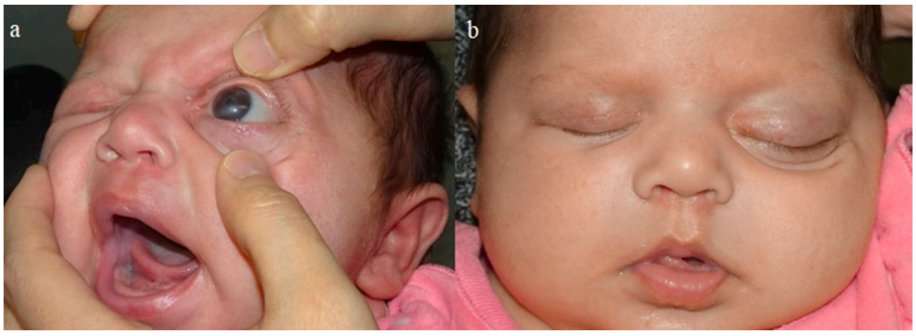

Fig. 5. Post-operative week ten – note infant is 11 weeks old. A: Anterior segment exam demonstrating the inferior cornea scar. B: External photograph illustrating significant improvement in proptosis.

Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

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None.

Declaration of competing interest

The authors declare the following financial interests/personal relationships which may be considered as potential competing interests: The authors have no conflict of interest.

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