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D. Ranganathan, Pediatrix Medical Group of Georgia
Phoebe Lenhart, Emory University
George Hubbard, Emory University
Hans Grossniklaus, Emory University

Final published version: http://dx.doi.org/10.1038/jp.2010.116

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Accessed October 18, 2019 8:11 PM EDT
Lacrimal gland choristoma in a preterm infant, presenting with spontaneous hyphema and increased intraocular pressure

D Ranganathan\textsuperscript{1}, P Lenhart\textsuperscript{2}, GB Hubbard\textsuperscript{2}, and H Grossniklaus\textsuperscript{2}

\textsuperscript{1}Neonatal Intensive Care Unit, Pediatrix Medical Group of Georgia at Piedmont Hospital, Atlanta, GA, USA
\textsuperscript{2}The Emory Eye Center, Emory University School of Medicine, Atlanta, GA, USA

Abstract

We report a case of intraocular lacrimal gland choristoma presenting very early in a preterm infant with hyphema, a mass lesion and raised intraocular pressure. Enucleation of the involved eye, which is the treatment in most cases, was performed and prosthesis was fitted successfully. An interesting additional finding in our patient was a choroidal defect, not reported to date with other cases in the literature.

Keywords

preterm; intraocular pressure; lacrimal gland choristoma; enucleation; choroid rupture

Introduction

Choristoma is a congenital tumor of tissues not normally present in the involved area. Lacrimal gland choristoma or ectopic lacrimal gland tissue is an uncommon lesion of the eye that can be seen in the orbit, outer eye, conjunctiva, cornea or sclera. Intraocular lacrimal gland choristomas (IOLGCs) are even more uncommon, and can present in early childhood as a cystic mass and with secondary complications, such as iris atrophy, glaucoma, cataract and loss of vision. We report a very early presentation of IOLGC in a preterm twin, at 6 weeks of age (corrected gestational age 35 + 3/7 weeks), with hyphema, a mass lesion and raised intraocular pressure (IOP).

Case

MM, twin A, was born at 29 + 3/7 weeks gestation, the product of \textit{in vitro} fertilization, to a 30-year-old Caucasian mother, who received 2 doses of betamethasone 2 days before delivery. The prenatal laboratories were unremarkable, and family history and past history were non-contributory. She was delivered by cesarean section for maternal pregnancy-
induced hypertension, with elevated blood pressure and proteinuria. Birth weight was 1360 g and Apgars were 2 at 1 min and 7 at 5 min. The infant had respiratory distress and was intubated in the delivery room and placed on mechanical ventilation. She had a typical neonatal intensive care unit course for a baby born preterm at <30 weeks gestation.

**Eye examinations**

At 6 weeks of age, a routine screening eye examination for retinopathy of prematurity showed spontaneous hyphema in the right eye with a yellowish mass in the inferonasal quadrant. Increased IOP was also noted, although the infant was upset during the examination. She was started on steroids for possible juvenile xanthogranuloma versus persistent fetal vasculature. Both eyes showed stage 0, zone 2 to 3 retinal vascularity. Follow-up eye examination in 1 week showed some clearing of the hyphema, a more vascularized mass and much higher IOP. At this time corneal haze was also noted, so brinzolamide drops, twice a day to the right eye, were added to lower the IOP. The working diagnosis was still juvenile xanthogranuloma. Follow-up examination showed decreased IOP, clearer cornea, a small clot and iridodialysis around the mass. The fundus examination was normal. Retinal vasculature was still immature, with no retinopathy of prematurity. At 2 months and 10 days of age (corrected gestational age 38 + 3/7 weeks), eye examination showed an irregular, small pupil on the right with iris atrophy and forward displacement of peripheral iris with a cystic mass. The differential diagnosis included iridocorneal endothelial dystrophy, juvenile xanthogranuloma, benign iris cyst and malignant tumor. She was referred to the corneal and retinal specialist for evaluation under anesthesia, and for undergoing a high-magnification anterior segment examination.

When seen at the referral center at 4 months of age, a cystic iris lesion with hyphema and elevated IOP was noted. Differential diagnoses included juvenile xanthogranuloma, atypical retinoblastoma, nevus/melanoma, medulloepithelioma, lacrimal gland choristoma and multifocal iris stromal cysts. Examination under anesthesia was carried out and showed a multicystic mass in the right eye (Figure 1). Over the next few weeks she developed increased corneal clouding, epiphora and buphthalmos, and was restarted on medications to lower IOP. Immersion ultrasound of the anterior segment under anesthesia showed a multicystic mass within the iris and ciliary body (Figure 2). Examination of the eye under anesthesia also showed corneal decompensation and choroidal ruptures. The differential diagnoses at this time included medulloepithelioma, congenital lesion and ectopic lacrimal gland. As the eye was amblyopic with secondary glaucoma, and had a poor visual prognosis, enucleation was the preferred treatment (Figure 3). Pathological examination of the right eye showed lacrimal gland choristoma of the iris and ciliary body, a choroidal defect with atrophy of the outer retinal layers and underlying retinal pigment epithelium and defect in Bruch’s membrane in this area, along with mild glaucomatous atrophy of the retina and optic nerve. Histopathology showed lobules of benign lacrimal gland tissue with dilated ducts and acini (Figure 4).
Discussion

IOLGC of the eye is extremely rare, with only 15 cases being reported in the ophthalmology literature.¹ Thirteen of them were treated by resection or enucleation. The two cases managed conservatively, one in 1997² and the other in 2007,¹ have no reported follow-up. Interestingly, in two cases treated by initial local excision, subsequent enucleation was needed due to the development of phthisis bulbi and recurrence of the tumor.³ In another case, a 12-month-old infant with a mass in the right eye from birth was diagnosed on a clinical basis as IOLGC. The mass enlarged and was removed by iridocyclectomy. However, 5 months after surgery he developed retinal detachment near the margin of the resection, which was then successfully repaired.⁴

One earlier report of IOLGC in a preterm infant was made in 1985.⁵ The infant was born at 33 weeks, presented at 11 months with blepharitis and at 17 months with photophobia, when a mass was seen. The baby was managed conservatively, but at 22 months of age enucleation was performed due to increase in the size of the mass. In another report, a 2-month-old infant was noted to have a vascular tumor in the left eye. The infant was treated with topical steroids for suspected juvenile xanthogranuloma.⁶ However, 5 weeks later the eye enlarged and IOP increased significantly. Ultrasound examination under general anesthesia showed cystic areas. As cysts are unlikely in juvenile xanthogranuloma, the diagnosis of IOLGC was carried out. Three weeks later, due to enlargement and pupillary abnormality, a resection was performed.

IOLGC was first reported in 1887,⁷ but the etiology remains unknown. Possible theories include early aberrant implantation of cells destined to become lacrimal glands, implantation with surface epithelium during lens formation, pinching off of lacrimal gland buds during closure of the fetal fissure or intraocular extension of early lacrimal gland tissue through preexisting scleral defects and aberrant differentiation, rather than aberrant implantation, of developing intraocular tissue.¹ ⁵

Although fine-needle aspiration biopsy and open biopsy have been used in some instances, the risk of spillage from a tumor, like retinoblastoma, along with amblyopia with glaucoma and poor visual prognosis, made enucleation the preferred treatment in our case, as in most other cases reported in the literature. An interesting additional finding in our patient was the choroidal defect, likely congenital in origin, but not reported to date with other cases of IOLGC. Our patient had a prosthesis placed 2 months after enucleation, when fully healed and all swelling was gone. She is doing well on follow-up.

Acknowledgments

P Lenhart, GB Hubbard and H Grossniklaus were supported in part by the Research to Prevent Blindness Inc. New York, New York and the National Institutes of Health Core Grant P30 EY06360, Bethesda, MD. The authors would like to thank the patient and family.
References


Figure 1.
Examination under anesthesia—multicystic mass.
Figure 2.
Immersion ultrasound of anterior segment shows cysts within the mass and within the iris stroma.
Figure 3.
Enucleated right eye shows a 5 × 2 mm mildly pigmented tumor in the ciliary body with central cyst. The tumor invaded the adjacent iris and protruded into the anterior chamber.
Figure 4.
Left: Histopathology shows lobules of benign lacrimal gland tissue with dilated ducts. Right: The acini are dilated in some areas (hematoxylin and eosin, left × 25, right × 100).