Iris Stromal Cyst in a 6-Month-Old With Rapid Progression to Angle Closure

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Case Report: A 6-month-old female presented with an iris cyst in the left eye, first identified at age 4 months. The patient was seen by an ophthalmologist at age 6 months and referred for further management. Our examination 2 weeks later demonstrated a 6 mm diameter iris cyst originating from the anterior surface of the inferior iris, occupying the inferior two thirds of the anterior chamber and obscuring the pupil. Intraocular pressure was normal and the remainder of the anterior chamber was formed. Because of concern for the development of amblyopia, the patient was scheduled for an examination under anesthesia and iris cyst removal 2 days later.

In the intervening 2 days, the patient’s mother noted worsening photophobia and tearing. At the time of surgery, the intraocular pressure was 51 mm Hg in the left eye. Anterior examination demonstrated interval development of shallowing of the anterior chamber with irido-corneal and cyst-corneal touch. The iris cyst had increased to 8 mm in size and filled the entire pupillary aperture. The iris cyst was excised, and histopathology confirmed the diagnosis of an iris stromal cyst.

Conclusions: Congenital stromal cysts of the iris can enlarge, threatening amblyopia and secondary glaucoma in children. Although angle closure is a known theoretical complication of iris stromal cysts, actual cases are rare in the literature. This case demonstrates the importance of serial examinations to monitor progression of iris stromal cysts, particularly in young children.

Key Words: iris cyst, pupillary block, glaucoma, amblyopia

Primary iris cysts may be divided into those of the iris pigment epithelium and the iris stroma, with the latter being far rarer.1 Iris stromal cysts are more frequently unilateral and predominately occur in children.2 They are characterized by their anterior position, translucency, and visible vessels overlying the cyst wall. Histopathologically, these cysts are characterized by multilayered stratified squamous epithelium with or without goblet cells.2 Although most cysts do not require treatment, congenital stromal cysts can enlarge to threaten the visual axis and risk amblyopia in children.3,4

Intraocular pressure elevation and glaucoma are possible complications of iris cysts. Mechanisms for intraocular pressure elevation in these cases include cyst rupture with subsequent blockage of the trabecular meshwork by their contents (referred to as “mucogenic” glaucoma), pigment dispersion, or secondary angle closure.5,6 Pupillary block is reported as a theoretical complication of iris cyst, but documented cases of this occurring are rare.7,8 Here, we report a case of a rapidly growing iris stromal cyst in a 6-month-old child resulting in occlusion of the pupillary aperture resulting in elevated intraocular pressure and acute angle closure.

CASE REPORT

A 6-month-old female was referred for evaluation and management of an iris cyst of the left eye. The cyst was first noticed on the inferior iris at age 4 months by the patient’s mother, who had noted increasing photophobia and squinting of the left eye. There was no history of prior amniocentesis, trauma, or any other medical comorbidities. The patient was seen by an ophthalmologist at the age of 6 months, at which time the cyst was noted to be ~4 mm in diameter with no obscuration of the visual axis. The patient was then referred to us for management.

At the time of our evaluation 14 days later, the child was able to fix and follow in both eyes. Intraocular pressure was 8 mm Hg in the right eye and 5 mm Hg in the left eye by rebound tonometry. The right pupil was 4 mm in the dark, 2 mm in the light, and round and briskly reactive. The left pupil was obscured by a cystic lesion.

Anterior examination of the right eye was unremarkable. In the left eye, the cornea was clear, and the anterior chamber was deep and formed with a 6 mm diameter cystic structure filling the inferior two thirds of the anterior chamber. The cyst made contact with the corneal endothelium inferi orly. The cyst was pigmented but translucent with visible vessels on its surface (Fig. 1A). The base of the lesion could not be directly visualized, though it appeared to originate from the anterior surface of the inferior iris. Funduscopic examination of the right eye was normal and was not visualized in the left eye. B-scan ultrasonography of the left eye revealed a normal globe contour without any appreciable mass lesions. Given the total pupillary obstruction in the left eye and concern for the development of amblyopia, the patient was scheduled for examination under anesthesia and iris cyst removal 2 days after examination.

On the day of surgery, the patient’s mother reported an increase in the patient’s epiphora and photophobia. Examination under anesthesia at that time was notable for intraocular pressures of 17 mm Hg in the right eye and 51 mm Hg in the left. In the left eye there was diffuse conjunctival injection and mild corneal miosis/cycloplegia. The anterior chamber was flattened inferiorly with irido-corneal touch peripherally and cyst-corneal touch centrally, with a small amount of aqueous present surrounding the iris cyst. Superiorly, the anterior chamber was shallowed to approximately one quarter of one central corneal thickness. The iris cyst had increased to 8 mm in diameter and filled the pupillary aperture, abutting the pupillary margin (Fig. 1B). Anterior segment optical coherence tomography demonstrated a fluid filled cyst in contact with the corneal endothelium and irido-corneal touch peripheral to the cyst (Fig. 1C).

A small superior peripheral iridectomy was performed via a superior paracentesis to relieve the pupillary block, and the anterior chamber immediately deepened (Fig. 2A). The cyst was decompressed with a 23-G needle (Fig. 2B). A portion of the cyst wall was excised and sent for pathology. A viscoelastic agent was used to dissect the cyst away from the corneal endothelium. A vitrector was used to remove the anterior wall of the cyst (Fig. 2C) and intraocular cautery was applied to the base of the cyst (Fig. 2D).
Pathology demonstrated a strip of anterior iris stroma with one side lined by ocular surface-type epithelium, including goblet cells, consistent with iris stromal cyst. In postoperative follow-up, the iris cyst recurred 6 weeks after their initial surgery, requiring reoperation for further excision and cautery.

**DISCUSSION**

Primary iris stromal cysts are uncommon and comprise a small portion of all primary iris cysts. Although most cysts typically do not cause vision-threatening complications, enlarging iris stromal cysts may result in amblyopia or elevated intraocular pressure, especially in pediatric patients in whom they are more likely to occur. Pupillary block is a frequently described theoretical complication of primary iris cysts, but there are few documented reports of this actually occurring. Thomas et al reported a case of chronic angle closure glaucoma due to multiple iris and ciliary body cysts found at the pupillary margin in a 37-year-old male. Paul et al reported elevated intraocular pressure in an 18-month-old caused by an inferior intrastromal cyst with occludable angles for 270 degrees, but occlusion of the pupil was not present in this report. Obata et al reported a congenital iris pigment epithelial cyst arising from the pupillary margin which resulted in pupillary block and iris bombe. To our knowledge, our patient is the first reported case of a primary iris

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**FIGURE 1.** Preoperative multimodal imaging of the left eye. A, External photo of the left eye 2 days before surgery demonstrates a 6 mm iris cyst arising from the inferior iris with cyst-cornea touch inferiorly. B, External photo of the left eye from examination under anesthesia shows interval enlargement of the cyst, pupillary block, and shallowing of the anterior chamber. C, Anterior segment optical coherence tomography (Leica Biosystems, Buffalo Grove, IL) shows cyst (arrowhead) and iris (arrow) both in contact with the corneal endothelium with a small portion of aqueous remaining adjacent to the cyst.

**FIGURE 2.** Intraoperative photographs of cyst excision. A, A superior peripheral iridectomy is performed to relieve pupillary block. Pupillary margin is seen pulled away from the iris cyst wall. B, The iris cyst contents are aspirated with a 23-G needle. C, A 23-G vitrector is used to remove the anterior portions of the cyst, with the base of the cyst better visualized on the anterior surface of the inferior iris. D, Intraocular cautery is applied to the base of the cyst to prevent recurrence.
stromal cyst arising from the anterior surface of the iris occluding the pupillary aperture and causing acute angle closure. The rate of progression of our patient’s iris cyst was remarkable: during a documented period of 16 days, the cyst progressed from a visually insignificant lesion to a size large enough to occlude the visual axis and the pupil.

Our differential diagnosis for the mechanism of the patient’s acute rise in intraocular pressure included angle closure due to occlusion of the pupillary aperture, and malignant glaucoma, given the significant flattening of the anterior chamber. However, a small area of aqueous remained in the anterior chamber superiorly and in the space immediately surrounding the iris cyst, and the anterior chamber deepened rapidly after the creation of a surgical peripheral iridectomy. Typically, in malignant glaucoma, the flattening of the anterior chamber persists despite presence of a patent peripheral iridectomy or iridotomy. In our patient, the occlusion of the pupillary aperture by the iris cyst and the response to a peripheral iridectomy suggest that the iris cyst prevented the passage of aqueous humor from the posterior to the anterior chamber, thus causing an elevation in intraocular pressure in a mechanism similar to classically described pupillary block, in which the crystalline lens is responsible for the resistance to flow of aqueous humor.

The vast majority of primary iris cysts do not progress or cause visual complications and therefore require no treatment. In cases where treatment is required, approaches include fine needle aspiration, laser treatment, and surgical excision. A stepwise approach beginning with minimally invasive treatment first has been recommended. In our case, however, surgical excision was performed because of the rapid cyst growth, obscuration of the visual axis, development of angle closure, and high risk of amblyopia. This report demonstrates the importance of serial examinations and thorough patient and family counseling to detect potentially vision-threatening complications of iris stromal cysts, particularly in young children.

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