



Case report

Mucogenic glaucoma in a child

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ABSTRACT

Purpose: We describe a case of secondary open-angle glaucoma due to mucin-producing congenital iris stromal cyst in a 4 year old patient.

Observations: A 4-year old female patient with a history of unilateral congenital iris stromal cyst presented with sudden-onset eye pain and redness, with markedly elevated intraocular pressure and evidence of early optic nerve damage. During the examination under anesthesia, the anterior chamber angle was open and there was no evidence of pupillary block. Ultrasound biomicroscopy revealed mildly echogenic substance filling the anterior chamber suspicious of mucoid material, which was verified by the inability to aspirate the material through a 25 gauge needle. The iris cyst was excised, and the intraocular pressure normalized spontaneously. Pathologic examination confirmed a mucin-secreting iris cyst lined with goblet cells and confirmed the mucogenic mechanism.

Conclusions and importance: This is the first reported case of mucogenic glaucoma in a pediatric patient. This rare entity should remain on the differential diagnoses of childhood glaucoma associated with nonacquired ocular anomalies. Surgical excision of the iris cyst may be curative.

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1. Introduction

Glaucoma associated with iris cysts is rare, and the mechanisms of intraocular pressure (IOP) elevation include secondary angle closure, pigment dispersion, inflammation and mucin displacing aqueous in the anterior chamber. Here, we describe the first case of glaucoma associated with mucin-producing iris stromal cyst, or mucogenic glaucoma, in a child.

2. Case report

A 4-year old female first presented to Bascom Palmer Eye Institute at the age of 22 months when her mother noticed an enlarging pigmented lesion on the right iris (Fig. 1). The patient is the product of full-term gestation, delivered by Cesarean section without complications, and is otherwise in good health with normal development. The patient's mother is a carrier of Fragile X mutation, and during gestational week 20, an ultrasound-guided amniocentesis was performed which ruled out Fragile X mutation

in the patient. There was no history of ocular trauma. The patient was diagnosed with congenital iris stromal cyst. Between the age of 22 months and 4 years, the patient underwent serial examinations under anesthesia (EUA) by an ocular oncologist every 6 months with IOP in both eyes ranged between 15 and 19 mmHg (Tonopen, Reichert, Depew, New York, USA).

At the age of 4 years and 9 months, the patient presented emergently with a one-day history of sudden onset pain and redness in the right eye. In the clinic, best-corrected visual acuity was 20/25 in the affected eye, with IOP elevated at 67 mmHg by rebound tonometer (Icare Finland, Oy, Helsinki). There was no afferent pupillary defect. The patient could not cooperate with slit lamp examination, although the anterior chamber is formed and deep on examination using an oblique slit beam from a retinoscope.¹ Topical dorzolamide/timolol fixed-combination therapy was initiated in the right eye twice daily. EUA two days later revealed an IOP of 39 mmHg (Tonopen, Reichert, Depew, New York, USA) in the right eye and 16 mmHg in the left eye under light sedation, and a focal corneal endothelial opacity in the quadrant of the cyst with mild pupillary peaking (Fig. 2). Ultrasound biomicroscopy showed a deep anterior chamber without iris bombé, and a faintly echogenic material filling the anterior chamber (Fig. 3). Gonioscopy revealed wide-open angles (Fig. 4). Examination of the fundus revealed increased cupping of the right optic disc

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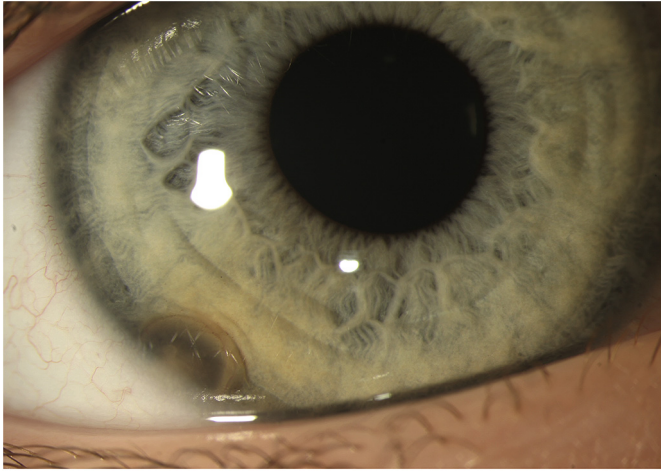


Fig. 1. Clinical photograph of a congenital iris stromal cyst on at age of 22 months.

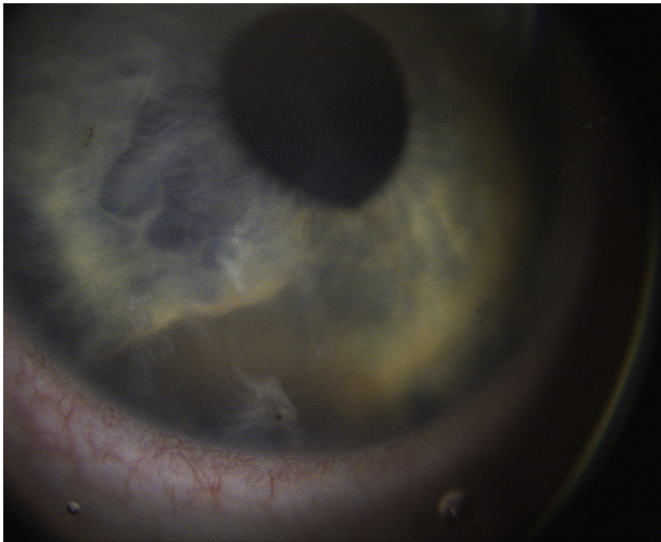


Fig. 2. Clinical photograph of the iris cyst at age 4 years 9 months. The cyst is less discreet in appearance, and there is focal corneal opacity at the quadrant of the cyst. The intraocular pressure is markedly elevated, while the cornea remained clear.

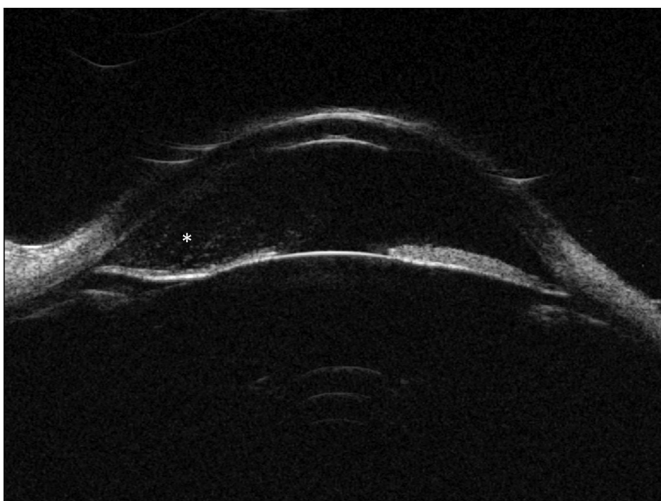


Fig. 3. Ultrasound biomicroscopy examination of the right eye. The anterior chamber is deep with open angles. There is no iris bombé. The anterior chamber is filled with a faintly echogenic material (asterisk).

compared to prior photographs (Fig. 5). Axial length by contact A-scan biometry was 21.5 mm on the right and 21.2 mm on the left.

An attempt to aspirate the anterior chamber content with a 25 gauge needle yielded no fluids, which suggests a secondary open-angle mechanism of IOP elevation due to a viscous fluid displacing the aqueous. The decision was made to excise the iris cyst with concurrent irrigation of the anterior chamber. Following the creation of a large scleral flap of approximately 90% depth (Fig. 6a), two iridotomies were created using a 23 gauge vitrector (Fig. 6b), and a sectoral iris flap was created using intraocular scissors (Fig. 6c, d). The scleral bed was incised and the anterior chamber was entered at the scleral flap hinge, and a large globule of gelatinous material spontaneously presented (Fig. 6e, f). The iris was externalized and was excised at the base (Fig. 6g). The scleral flap was repositioned and closed using interrupted 10-0 nylon sutures followed by closure of the conjunctiva (Fig. 6h).

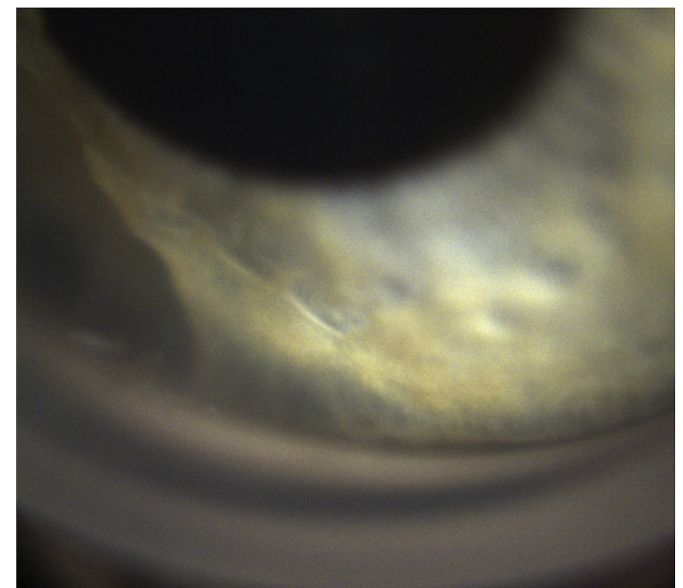
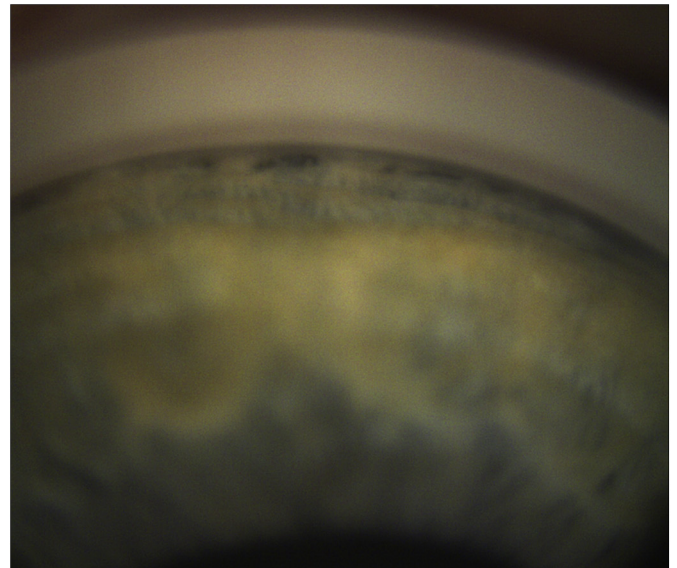


Fig. 4. Gonioscopic photograph of the superior and inferior angles. The angles in the affected eye were wide-open, with a visible ciliary body band circumferentially.

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