

Infantile glaucoma presenting as a case of “red eye”

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RÉSUMÉ : Le glaucome infantile est une forme de glaucome congénital qui se manifeste au cours de la première année de vie. Les enfants, comme les adultes, peuvent développer une forme de glaucome aigu pouvant conduire à la cécité chez l'enfant. L'identification et le traitement rapides sont essentiels afin d'éviter ce dénouement tragique. L'enfant dont le cas est discuté dans le présent article présentait une rougeur à l'œil qui fut diagnostiquée par la suite comme étant un glaucome. Cet article passe en revue la physiopathologie du glaucome infantile, les techniques d'examen et les stratégies de traitement.

Key words: pediatric, glaucoma

Introduction

Infantile glaucoma is a form of developmental glaucoma that presents in the first year of life. Children, like adults, may develop acute glaucoma, and childhood glaucoma may lead to blindness. Prompt recognition and treatment is essential if we are to avoid this tragic outcome. The child discussed in this case presented with a red eye, which was subsequently diagnosed as glaucoma. This paper reviews the pathophysiology of infantile glaucoma, examination techniques and treatment strategies.

Case report

A 6-month-old boy was presented to the Deaconess Immediate Treatment Center (Buffalo, NY) with a red, apparently painful, right eye. Prenatal, birth and neonatal history were unremarkable. He was breast-fed, fully immunized, otherwise healthy, and had reached age-appropriate developmental milestones. His mother indicated that, on the morning of presentation, he had awakened very fussy with a red and tearful right eye. When it failed to improve after several hours, she sought medical attention. She stated that there had been no similar episodes in the past, no recent vomiting, no apparent respiratory infection and no history of ocular trauma.

Examination revealed an afebrile male infant who was photophobic, resisted eye opening, and was crying incon-

solably. Inspection of the right eye demonstrated marked conjunctivitis and tearing. Ophthalmoscopy revealed that the right cornea was cloudy and that the pupil was 4 mm and poorly reactive to light. Corneal size was estimated at 15 mm and ocular movements were full. The left eye appeared normal, with a clear cornea and no conjunctivitis. The remainder of the physical exam was unremarkable.

Corneal clouding, conjunctivitis and tearing suggested glaucoma; therefore, the child was transferred to the Children's Hospital of Buffalo, where he was examined under anesthesia by a pediatric ophthalmologist. His intraocular pressure was elevated at 40 mm Hg, and his cup-to-disc ratio was increased at 0.8, but the retina was otherwise normal. A diagnosis of infantile glaucoma was made and urgent goniotomy performed.

The child required a subsequent goniotomy several months later but is doing well at 2 years of age. He has been followed by a pediatric ophthalmologist and remains on levobunolol hydrochloride 0.25% (Betagan®) eye drops twice daily. Vision in the right eye was well preserved and recently measured at 6/12. Acuity in the unaffected eye is age appropriate at 6/8.

Discussion

This child was presented with a red eye and in obvious distress. Differential diagnoses would include conjunctivitis,

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iritis, corneal abrasion, trauma, megalocornea, allergic reaction and glaucoma. The critical physical finding in this case was corneal clouding, a classic feature of glaucoma.

Glaucomas are classified as primary, congenital (infantile) or secondary. Primary glaucomas include open- and closed-angle varieties, but over 90% of all glaucomas are open-angle.¹

Open-angle glaucoma has an insidious onset and often causes no symptoms until irreversible visual impairment has occurred. The cause of increased intraocular pressure is overproduction of aqueous humor and obstruction of aqueous drainage from the anterior chamber through the trabecular meshwork (Canal of Schlemm). The treatment is medical and directed toward these 2 pathophysiological mechanisms. Beta-adrenergic blocking agents (e.g., timolol, betaxolol and levobunolol) reduce aqueous humor production,² and miotic drops (e.g., pilocarpine or carbachol) cause pupillary constriction, which draws the root of the iris away from the angle, reducing obstruction to aqueous outflow. Table 1 summarizes medical treatment modalities.

“Acute” (closed-angle) glaucoma preferentially affects people with narrow anterior chamber drainage angles. In such individuals, intraocular pressure rises abruptly when the root of the iris obstructs the angle and prevents aqueous efflux. Acute attacks may be precipitated by mydriatic agents or chronic congestive changes affecting the trabecular meshwork. The treatment of closed-angle glaucoma is urgent laser iridectomy,¹ but pre-operative administration of miotic drops and osmotic agents (e.g., glycerol or acetazolamide) is warranted.

Secondary glaucomas are due to acquired changes of the lens and uveal tract, or to trauma.³

Congenital glaucoma, as described in this case, results from trabeculodysgenesis (dysgenesis of the drainage

framework) during fetal development, which may be inherited as an autosomal recessive trait.⁴ In affected children, the iris is hypoplastic and does not insert properly into the trabecular surface. Because the drainage system is dysfunctional, aqueous pressure builds in the anterior chamber and is transmitted to the optic disc.

Continued pressure causes pathologic “cupping” of the disc and ultimately leads to permanent optic nerve damage and blindness.⁵ This form of glaucoma requires surgical goniotomy or trabeculotomy.⁶

Congenital glaucoma presents with 4 common signs. *Tearing* (epiphora) results when corneal stretch irritates corneal nerve endings. *Corneal enlargement* may occur in children under 3 and is a marker of increased intraocular pressure. Normal corneal diameter should be 12 mm or less in infants under 1 year of age. *Corneal clouding* results when tears in Dehrcemet’s membrane render it permeable, allowing aqueous humor to infiltrate the stroma.¹ *Photophobia* is typical, and children with glaucoma will usually hide their eyes from light to reduce pain.

In children over 3 years, glaucoma often presents with increasing myopia and little or no pain. In this age group, it may be asymptomatic and present late, after visual loss has already occurred. Fundoscopic findings include blurring of the disc margin and an increased cup-to-disc ratio (the normal being 0.6) in one eye.^{3,6}

ED management of pediatric glaucoma requires prompt referral to a pediatric ophthalmologist for definitive examination (under anesthesia, if necessary), surgical treatment and follow-up. If an appropriate consultant is not rapidly available, emergency physicians may initiate the medical therapy described above; however, drug treatment is merely a stopgap measure and does not take the place of prompt surgical consultation.^{3,6,7}

Table 1. Medical management of acute glaucoma

Medication	Strength / concentration	Dosage
Miotics*		
Carbachol	0.75%, 1.5%, 2.25%, 3.0%	1–2 drops stat, then bid or tid
Pilocarpine	0.5%, 1%, 2%, 3%, 4%, 6%	2 drops stat, then bid to qid
Beta-adrenergic blocker†		
Timolol	0.25%, 0.5%	1 drop stat, then bid
Levobunolol	0.25%, 0.5%	1 drop stat, then 1–2 drops bid
Betaxolol	0.25%	1 drop stat, then 1–2 drops bid
Hyperosmolar agents†		
Glycerol	1 mL/kg of 50% solution po	1 dose stat
Mannitol	1–2 g/kg of 20% solution IV	1 dose stat
Acetazolamide	250-mg tablets or IV solution	500 mg po or IV stat, then 250 mg po or IV q6h

*To constrict pupils and enhance aqueous drainage.
†To reduce aqueous production.

Conclusion

Red eyes in children are not always infectious or allergic. A brief examination of the cornea and optic disc should lead to correct diagnosis and expedient treatment of this serious condition.

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