Case Report

Acute hydrops as an atypical presentation of primary congenital glaucoma

Abeer A. AlHazzani[1], Eman M. Al-Sharif[1,2], Saleh A. Al-Obeidan[1,3]

Access this article online

Quick Response Code:

Website: www.saudijophthalmol.org

DOI: 10.4103/1319-4534.347309

Abstract:

Primary congenital glaucoma (PCG) is a disease of childhood characterized by elevated intraocular pressure (IOP) that causes stretching of the eye's outer coats, namely sclera and cornea. This results in the elongation of the eyeball and expansion of the horizontal corneal diameter giving the appearance of a buphthalmos eye. Aggressive disease with high IOP readings causes excessive mechanical stretching that may be poorly tolerated by the corneal Descemet's membrane, leading to large breaks in it with subsequent corneal edema due to sudden influx of the aqueous humor into the exposed stroma, resulting in acute corneal hydrops. While acute hydrops is a potential sequel of PCG, it is considered one of its rare presentations. We present two cases who presented to our hospital with acute hydrops secondary to PCG. Both patients were managed surgically where the first patient underwent combined trabeculotomy-trabeculectomy with mitomycin C, while the second patient underwent deep sclerectomy with mitomycin C. The surgical procedures effectively controlled the IOP and aided in clearing corneal edema in both patients. Early diagnosis and timely surgical intervention are of paramount importance to improve visual outcomes, enhance ocular maturation, and prevent potential irreversible vision loss, especially in this young-age group of patients that are prone to amblyopia.

Keywords:

Buphthalmia, Descemet's membrane, edema, hydrops, intraocular pressure, primary congenital glaucomav

Introduction

Primary congenital glaucoma (PCG) is a congenital structural anomaly of the trabecular meshwork and anterior chamber angle that leads to obstruction of the aqueous outflow, increased intraocular pressure (IOP), and subsequent optic nerve damage. The exact etiology of PCG remains unknown. Several theories have been put forward, with some proposing the presence of a membrane that covers the anterior chamber angle, called Barkan's membrane, which is thought to decrease outflow facility.[1] Others proposed abnormal thick trabecular membrane beams, the absence of intertrabecular spaces in deeper layers of the trabecular meshwork, and the presence of an amorphous material in the subendothelial region of Schlemm's canal.[2] Moreover, a study that examined the histopathological characteristics of the angle in PCG in newborns demonstrated the following features: partial absence with posterior displacement of Schlemm's canal, hypoplastic trabecular meshwork, and broad ciliary muscle attachment to the trabecular meshwork along with anterior insertion of the hypoplastic iris forming a pseudomembrane.[3] PCG most commonly affects children from birth up to 3 years of age, with a preponderance in males. Its estimated prevalence in the general population is approximately 1 in 10,000 live births. While it carries no racial predilection, a higher prevalence had been observed among communities with consanguineous marriages.[4]

The diagnosis of PCG poses some challenges as it is limited by the patients' young age. Nonetheless, early recognition and treatment are of supreme importance since it is a major cause of childhood blindness. The common presenting signs and symptoms include corneal clouding with an enlarged horizontal diameter, high IOP, abnormally deep anterior chamber due

[1]Department of Ophthalmology, College of Medicine, King Saud University, [2]Department of Ophthalmology, College of Medicine, Princess Nourah bint Abdulrahman University, [3]Department of Ophthalmology King Saud University, Riyadh, Saudi Arabia

Address for correspondence: Dr. Eman M. Al-Sharif, Department of Ophthalmology, College of Medicine, Princess Nourah bint Abdulrahman University, P.O. Box: 84428, Riyadh 11671, Saudi Arabia. E-mail: eman.sharif@hotmail.com

Submitted: 20-Sep-2019
Revised: 15-Dec-2020
Accepted: 30-Dec-2020
Published: 13-Jun-2022

How to cite this article: AlHazzani AA, Al-Sharif EM, Al-Obeidan SA. Acute hydrops as an atypical presentation of primary congenital glaucoma. Saudi J Ophthalmol 2021;35:356-9.

to increased axial length, and optic nerve cupping. Bilateral disease tends to be more frequent than unilateral disease.[5]

A well-known but rare presenting manifestation of infantile glaucoma is acute corneal hydrops, which is considered a medical emergency requiring urgent ophthalmic attention to prevent irreversible vision loss in this group of patients. The development of acute corneal hydrops is explained by the sudden rupture of the Descemet's membrane, which consequently allows the aqueous humor to enter the corneal stroma, leading to acute corneal swelling and opacification. The observed disruptions of Descemet's membrane are usually large; hence, the resultant corneal edema tends to take longer time to resolve and the visual outcome is generally worse compared to smaller disruptions of Descemet's membrane (Haab's striae). In such cases, early surgical intervention is mostly needed to control the elevated IOP and to clear the corneal edema, thus preventing amblyopia and enhancing visual development. Combined trabeculotomy-trabeculectomy (CTT) is one of the popular effective interventions that has been reported to have good success rates in various ethnic groups.[6,7] We report, herein, two atypical cases of PCG that initially presented to our institute with acute corneal hydrops. These cases highlight the importance of recognizing PCG as one of the potential causes of acute hydrops in infants.

Case Reports

First patient

A 12-day-old male infant, product of an uncomplicated gestation that led to spontaneous vaginal delivery, presented to our hospital due to the sudden onset of left corneal opacification and eye protrusion that developed over days shortly after birth [Figure 1a and b]. The family reported positive family history of congenital glaucoma and consanguinity. On examination under anesthesia, the IOP was found to be 41 mmHg and 27 mmHg in the right and left eye, respectively. Pachymetry revealed a central corneal thickness of 411 mm and 980 mm in the right and left eyes, respectively. Slit lamp examination revealed bilateral corneal haze of Grade 1 in the right eye and Grade 4 in the left eye, with a large intrastromal fluid cleft.[8] Corneal diameters were 11.5 mm in the right eye and 13 mm in the left eye. The right eye exhibited a deep and quiet anterior chamber with a clear lens, and fundoscopic evaluation showed a cup–disc ratio of 0.65. Evaluation of the structures behind the cornea was not possible in the left eye due to significant edema; therefore, B-scan ultrasonography was done preoperatively which revealed large optic nerve cupping. The infant was diagnosed with bilateral congenital glaucoma with acute hydrops in the left eye. He subsequently underwent deep sclerectomy (DS) with mitomycin C in the right eye and CTT with mitomycin C in the left eye. One day postoperatively, IOP dropped to 10 mmHg in the right eye and 15 mmHg in the left eye. The corneal clarity was completely restored in the right eye and significantly improved in the left eye. One month postoperatively, the IOP was 18 mmHg in both eyes and slit lamp examination showed a clear cornea in the right eye and a centrally scarred cornea in the left eye with clear periphery. Interestingly, the significant corneal edema that was present preoperatively almost cleared with complete disappearance of the fluid cleft [Figure 1c].

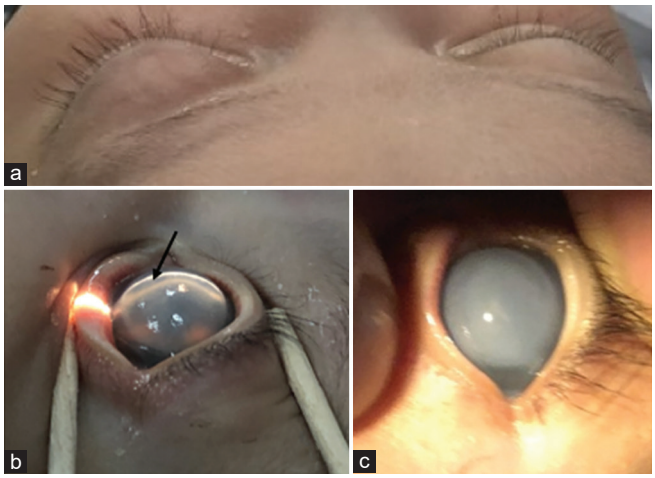

Figure 1: (a) External photograph showing the protrusion of the left eye on presentation. (b) The intracorneal fluid cleft clearly demonstrated in the left eye on presentation (black arrow). (c) The remnant corneal scarring 1 month after surgery

Second patient

A 5-month-old male infant, a product of an uncomplicated gestation that led to spontaneous vaginal delivery, presented to our hospital due to clouding of the right cornea recognized by the parents 1 day before presentation. The family reported a history of excessive tearing and a larger right eye since birth, for which they never sought medical attention. The parents are third-degree relatives, but they reported no family history of glaucoma or trauma to the right eye. On examination under anesthesia, the IOP was found to be 37 mmHg and 11 mmHg in the right and left eye, respectively. Slit lamp examination revealed corneal haze in the right eye of Grade 4 with a diameter of 14 mm and a clear cornea in the left eye with a diameter of 11 mm.[8] The left eye exhibited a deep anterior chamber with a clear lens, and fundoscopic evaluation revealed a healthy optic nerve. Evaluation of the structures posterior to the cornea was not possible in the right eye due to the haze. B-scan ultrasonography was done preoperatively which revealed moderate right optic nerve cupping. The infant was diagnosed with unilateral right eye congenital glaucoma with acute hydrops, and he subsequently underwent nonpenetrating DS with mitomycin C. One day postoperatively, the IOP dropped to 10 mmHg. At follow-up 3 years later, the IOP was maintained within normal in the operated eye (10 mmHg on average) and the cornea was clear with an oblique scar [Figure 2]. However, poor fixation was noted in the right eye with left eye preference as the child developed a large angle esotropia with high astigmatism in the right eye (+5.50 diopters) causing anisometropic amblyopia.

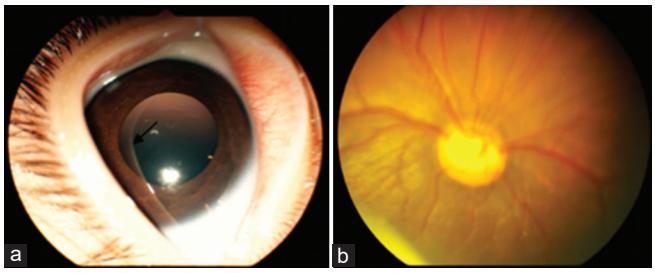

Figure 2: (a) External photograph of the right eye showing the break in Descemet's membrane after healing (black arrow). (b) Fundus photograph of the right eye showing moderate optic nerve cupping

Discussion

Acute corneal hydrops is an uncommon presentation of PCG with only a few cases reported in the literature. Acute corneal hydrops is actually rare in infants, and perhaps, PCG in addition to birth trauma is one among the most common causes in this age group.[9] These patients usually have significant corneal edema and may consequently manifest with intrastromal fluid clefting, as seen in in our first patient, and in accordance to what was previously reported in the literature.[10,11] It had been postulated that these breaks usually heal by primary intention, resulting in minimal scar formation when treated early.[7,11] Detecting these breaks in Descemet's membrane might be difficult on initial presentation as they are usually obscured by edema.[7] However, ultrasound biomicroscopy can be utilized in such patients to demonstrate these breaks in Descemet's membrane.[11]

One of the key factors to consider in these patients is early detection and timely surgical intervention to prevent potential blindness, due to scarring and imminent amblyopia. Remarkably, our patients' IOPs were well controlled after undergoing surgery, and there was rapid clearance of corneal edema. Yet, considerable central corneal scarring developed especially in our first patient in spite of undergoing urgent CTT. We hypothesize that this is most likely related to the severity of the disease and the size of the break in Descemet's membrane. In one of the largest case series that reported the surgical outcomes of 31 cases (38 eyes) of infantile glaucoma presenting with acute corneal hydrops over a 23-year period, all patients underwent CTT. IOPs dropped by an average of 53.05% and corneal edema cleared in all of the cases. Nevertheless, the same study also reported moderate vision recovery that was mainly attributed to the development of irregular corneal astigmatism because of Haab striae, which crossed the visual axis in the majority of patients and healed with scarring.[6] Similarly, our second patient was found to have poor visual recovery in the right eye upon follow-up because he developed high astigmatism which resulted in anisometropia, further contributing to his amblyopia along with the large angle esotropia. On the other hand, despite the limited follow-up period of our first patient, it is expected that the visual outcome will be poor due to the significant corneal scar that might require penetrating keratoplasty for visual rehabilitation.

Another study reported that the complete resolution of corneal edema in an infant presenting with acute hydrops secondary to PCG for which CTT was performed.[11] In fact, the majority of the previously published cases of PCG with acute hydrops were managed by CTT with satisfactory success evident by good IOP control and lack of any intraoperative or postoperative complications. Our second patient underwent DS, and to the best of our knowledge , this is the only patient with acute hydrops caused by PCG who underwent this procedure successfully. DS was shown to be effective and fruitful in managing childhood glaucoma.[8] All these studies collectively suggest that the choice of surgical management may vary among different physicians depending on the case, preference, and experience, whereas the constant factor governing success in such urgent cases is probably the timing of surgery where earlier intervention will likely lead to better outcomes.

In conclusion, these case reports serve as a reminder that PCG may initially present with acute hydrops, despite being less frequently so. Early recognition and timely surgical management are of paramount importance to minimize corneal scarring and improve the visual prognosis in this vulnerable age group. CTT and DS are both effective procedures that successfully control IOP and aid in effectively clearing corneal edema in these patients.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patients have given their consent for their images and other clinical information to be reported in the journal. The patients understands that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

The authors would like to thank Deanship of scientific research for funding and supporting this research through the initiative of DSR scholarship support.

Conflicts of interest

There are no conflicts of interest.

References

1. deLuise VP, Anderson DR. Primary infantile glaucoma (congenital glaucoma). Surv Ophthalmol 1983;28:1-19.
2. Anderson DR. The development of the trabecular meshwork and its abnormality in primary infantile glaucoma. Trans Am Ophthalmol Soc 1981;79:458-85.
3. Perry LP, Jakobiec FA, Zakka FR, Walton DS. Newborn primary congenital glaucoma: Histopathologic features of the anterior chamber filtration angle. J AAPOS 2012;16:565-8.
4. Mandal AK, Chakrabarti D. Update on congenital glaucoma. Indian J Ophthalmol 2011;59 Suppl: S148-57.
5. Yu Chan JY, Choy BN, Ng AL, Shum JW. Review on the management of primary congenital glaucoma. J Curr Glaucoma Pract 2015;9:92-9.
6. Mandal AK. Acute corneal hydrops in children with primary infantile glaucoma: A report of 31 cases over 23 years at the LVPEI. PLoS One 2016;11:e0156108.
7. Kaushik S, Joshi G, Pandav SS. Pathogenesis and surgical resolution

of acute hydrops in primary congenital glaucoma. Clin Experiment Ophthalmol 2018;46:569-70.
8. Al-Obeidan SA, Osman Eel-D, Dewedar AS, Kestelyn P, Mousa A. Efficacy and safety of deep sclerectomy in childhood glaucoma in Saudi Arabia. Acta Ophthalmol 2014;92:65-70.
9. McAnena L, O'Keefe M, Kirwan C, Murphy J. Forceps delivery-related ophthalmic injuries: A case series. J Pediatr Ophthalmol Strabismus 2015;52:355-9.
10. Roche O, Beby F, Dufier JL, Parsa CF. Descemet membrane rupture accompanied by stromal clefting in congenital glaucoma. Arch Ophthalmol 2008;126:1163-4.
11. Zacharia PT, Harrison DA, Wheeler DT. Penetrating keratoplasty with a valved glaucoma drainage implant for congenital glaucoma and corneal scarring secondary to hydrops. Ophthalmic Surg Lasers 1998;29:318-22.