A Case of Traumatic Phacocele in a Patient with Aniridia

Mandeep Tomar[1], Gaurav Sharma[1], Tarun Sood[2], Vandna Sharma[1], Richa Dhiman[1], Bhup Singh[1]

[1]*Department of Ophthalmology, Dr. Rajendra Prasad Government Medical College, Tanda Kangra, Himachal Pradesh, India*

[2]*Civil Hospital Sarkaghat, District Mandi, Himachal Pradesh, India*

Summary

An 18 year old boy with bilateral aniridia presented to us with a history of trauma to his left eye with a ball. Visual acuity was finger counting at 1 metre OD and perception of light OS. Past records revealed history of cataract surgery and intraocular lens implantation for congenital cataract in his right eye 8 years back and superiorly subluxated crystalline lens in left eye. Slit lamp examination showed presence of a bluish white subconjunctival mass superonasally in his left eye. The right eye had aniridia associated keratopathy, epithelial downgrowth and posterior capsular opacification. Exploration revealed a scleral perforation and phacocele. During suturing and repositioning of the prolapsed uveal tissue, non expulsive hemorrhage occurred which was managed and closure of the wound was done. Postoperatively, the eye was hypotonous with visual acuity of perception of light. We concluded that traumatic phacocele is a rare event and in patients with Aniridia, it is a surgical challenge because of tissue weakness that may lead to excessive bleeding, difficulty in suturing, and slow wound healing. Our main aim should be to extract the phacocele and maintain the globe integrity in such cases.

Delhi J Ophthalmol 2019;29;117-119; Doi http://dx.doi.org/10.7869/djo.464

Keywords: *Aniridia, Phacocele*

Introduction

Aniridia is an ocular disorder affecting the cornea, iris, intraocular pressure, lens, fovea, and optic nerve. Individuals with aniridia characteristically show nystagmus, impaired visual acuity (usually 20/100 - 20/200), and foveal hypoplasia.[1] The term phacocele is derived from the Greek word "phaco" meaning lens and "kele" meaning hernia. Phacocele (synonym =lenticele) is defined as the dislocation/ hernia of crystalline lens into the subconjunctival space through scleral rupture. It is a rare event caused by severe indirect blunt trauma and it has been reported that phacocele occurs in less than 13% of all lens dislocation.[2] Several predisposing factors which favour scleral rupture with lens dislocation are previous large surgical scar of cataract extraction/trebeculectomy/old trauma, diseases of eye coat like scleritis and connective tissue disorders and long term topical medications.[3]

Case Report

An 18 year old boy presented to us with a history of pain and redness in his left eye following blunt trauma with a tennis ball 1 week back. He admitted that vision in both eyes had been poor since childhood. The patient was a known case of bilateral aniridia with juvenile open angle glaucoma for which he was instilling timolol 0.5% eye drops off and on.

On ocular examination, visual acuity was counting finger at 1 metre OD and perception of light OS. Intraocular pressure (IOP) was 14 mmHg OD and 8 mmHg OS on NCT. Slit lamp examination showed bilateral complete aniridia with jerk nystagmus and presence of a bluish white subconjunctival mass of 8×6.5 mm superonasally in his left eye (Figure 1 and 2). Posterior segment and fundus examination could not be done owing to aniridia associated keratopathy (AAK), epithelial downgrowth, thick posterior capsular opacification in the right eye (Figure 3 and 4) and AAK, corneal edema and Descemet's folds in the left eye. Past records revealed history of cataract surgery and intraocular

Figure 1: *Showing bluish white subconjunctival mass with surrounding purple bluish scleral thinning*

Figure 2: *Showing bluish white subconjunctival mass with surrounding purple bluish scleral thinning*

Figure 3: *Showing aniridia associated keratopathy (AAK) in right eye and severe photophobia*

Figure 4: *Showing aniridia associated keratopathy (AAK) in right eye*

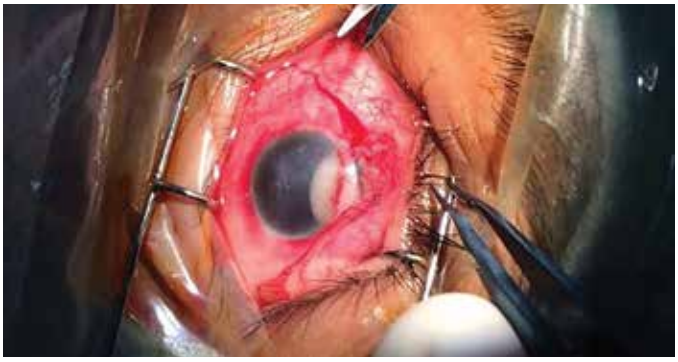

Figure 5: *Intraoperative picture showing phacocele with scleral defect at superonasal quadrant after peritomy*

Figure 6: *Postoperative day 1 picture with overlying intact scleral sutures after removal of phacocele*

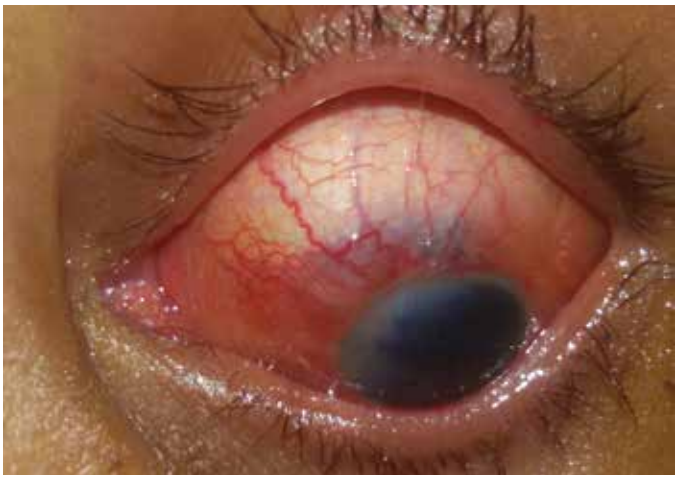

Figure 7: *At 2 months postoperatively*

lens implantation for congenital cataract in his right eye 8 years back and superiorly subluxated crystalline lens in the left eye.

Exploration under peribulbar anaesthesia was done and a scleral perforation of 4.5-5 mm size was found superonasally from 10 to 1 o'clock with phacocele and uveal prolapse. Phacocele was removed and during repositioning of the prolapsed uveal tissue, a profuse non expulsive hemorrhage occurred intraoperatively which was managed by rapid closure of the wound using interrupted vicryl 6-0 sutures, intravenous mannitol and digital ocular pressure. Postoperatively, the eye was hypotonous with full chamber hyphaema. The patient was managed conservatively on oral prednisolone 1mg/kg body weight in tapering doses, oral acetazolamide 250 mg three times a day, topical steroids and antibiotics, along with cycloplegic eye drops. At 1 week postoperatively, the hyphaema started to clear, IOP was 10 mmHg with visual acuity of perception of light. Patient was discharged and kept under follow up.

Discussion

After blunt trauma, ocular rupture tends to occur in the superior nasal sector due to the projection of energy caused by the impact in the temporal region, where impacts occur more frequently.[4] Energy is projected towards the superior, posterior and nasal sectors; the globe collides with the trochlea and orbital wall.[5,6,7] Rupture usually occurs at 2.5 mm and is concentric to the limbus, where tense and deep scleral fibers are transformed into a delicate lamella of pectineous ligament.[6] The predominant site of indirect scleral rupture is the superonasal quadrant[8], followed by the superotemporal quadrant.[9] The scleral rupture frequently occurs between the limbus and spiral of Tillaux.[9]

Due to poor compliance, the patient had elevated intraocular pressure since a very long time and sudden compression – decompression forces during trauma in a predisposed eye like aniridia with lens subluxation may be a contributing factor for subconjunctival dislocation of lens and also for intraoperative non-expulsive choroidal hemorrhage. Since visual potential was already very low in this case, our main aim was to remove the phacocele, repair the underlying scleral defect and achieve the integrity of globe, which we were able to achieve successfully.

Conclusion

Traumatic phacocele is a rare event and in patients with aniridia, it is a surgical challenge because of tissue weakness that may lead to excessive bleeding, difficulty in suturing, and slow wound healing. Our main aim should be to extract the phacocele and maintain the globe integrity in such cases. Of course, evidence based on one case cannot be conclusive but in rare cases such as phacocele in aniridia, any observation may be worthwhile.

References

1. Hingorani M, Williamson KA, Moore AT, van Heyningen V. Detailed ophthalmologic evaluation of 43 individuals with PAX6 mutations. *Invest Ophthalmol Vis Sci* 2009; 50:2581–90.

2. McDonald PR, Purnell JE. The dislocated lens. *JAMA* 1951; 145:220-6.
3. Santos-Bueso E, Sáenz-Francés F, Díaz-Valle D, Troyano J, López-Abad C, Benítez-del-Castillo JM, et al. Ocular rupture associated with lensdislocation to the subconjunctival space. *Arch Soc Esp Oftamol* 2007; 82:641-644.
4. Berlin HS, Ritch R. The treatment of glaucoma secondary to aniridia. *Mt Sinai J Med* 1981; 48:11.
5. Nelson LB, Spaeth GL, Nowinski TS, Margo CE, Jackson L. Aniridia. A review. *Surv Ophthalmol* 1984; 28:621-642.
6. Sony P, Khokhar S, Panda A. Traumatic lenticele. *Clin Experiment Ophthalmol* 2005; 33: 76-77.
7. Sathish S, Chakrabarti A, Prajna V. Traumatic subconjuntival dislocation of the crystalline lens and its surgical management. *Ophthalmic Surg Lasers* 1999; 30:684-68
8. Lister W. Some concussion changes met within military practice. *Br J Ophthalmol* 1924; 8:305-18.
9. Cherry PM. Indirect traumatic rupture of the globe. *Arch Ophthalmol* 1978; 96:252-6.

Cite This Article as: *Tomar M, Sharma G, Sood T, Sharma V, Dhiman R, Singh B. A Case of Traumatic Phacocele in a Patient with Aniridia.*

Acknowledgments: Nil

Conflict of interest: None declared

Source of Funding: None

Date of Submission: 26 August 2018
Date of Acceptance: 18 December 2018

Address for correspondence

Mandeep Tomar MS, DNB
Senior Resident
Department of Ophthalmology,
Dr. Rajendra Prasad Government Medical College, Tanda Kangra,
Himachal Pradesh - 176001, India
Email id: sunnyigmc85@gmail.com

Quick Response Code