

CHALLENGING MANAGEMENT OF TRAUMATIC POSTERIOR LENS LUXATION WITH ANIRIDIA: A CASE REPORT

Birgitta Henny Perwitasari^{1,2}, Muhammad Firmansjah^{1,2}

1. Dept. of Ophthalmology, Dr Soetomo General Academic Hospital, Surabaya

2. Dept. of Ophthalmology, Faculty of Medicine, UNIVERSITAS AIRLANGGA, Surabaya

Email: firmanjah.spm@gmail.com

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ABSTRACT:

This case report demonstrates the challenging condition and management of a patient with aphakia due to posterior lens luxation and aniridia, following a blunt trauma. The patient is a 34-year-old male with blurred vision on right eye. He had a history of blunt trauma 1.5 years prior, that resulted in corneoscleral laceration and hyphema, then underwent surgery at the time of injury. In present time, 1.5 years after the right eye trauma and surgery, the visual acuity was 6/40 cc S+7.00 C-4.00 X120 → 6/7.5 with normal intraocular pressure. Anterior segment of present right eye showed 2 remaining corneal sutures and slight neovascularization outside of visual axis. There was an absence of iris and lens on right eye, but there were visible remains of anterior capsule peripherally and vitreous body in the center. Posterior segment of right eye showed posterior lens luxation on the inferior, with retina, macula and optic nerve head within normal condition, confirmed by OCT findings. Left eye was unremarkable. The patient was diagnosed with right eye posterior lens luxation, aniridia, and anisometropia, treated with colored soft contact lens.

Posterior lens luxation without sight threatening complication can be managed conservatively. However, vitrectomy and lensectomy surgery on posterior lens luxation needs to be considered if there are complications or if the patient requires other ocular surgery. Aniridia can be acquired through ocular trauma, with treatment options ranging from colored contact lens, corneal tattoo, iris sutures, and implantable iris prosthesis. In this case, the management of aphakic condition caused by posterior lens luxation combined with aniridia includes the consideration of conservative through aphakic contact lens, or surgery of vitrectomy, lensectomy, IOL and iris prosthesis implantation.

Combined condition of post-traumatic aphakia and aniridia is a challenging situation which required many considerations.

1. Introduction

Eye trauma is a serious global health concern, which contributes as the cause of bilateral blindness in 1.6 million cases and unilateral blindness or visual impairment in 19 million cases globally. Eye trauma predominantly happen to male population.(1)

Eye injury can cause various lesion. Blunt trauma, for instance, can cause compression of the eye's globe from front to back and lead to expansion at the equator, stretching the chamber angle and iris. This can cause rupture of the blood vessels at the iris root, resulting in hyphema. Additionally, damage such as tears in the iris sphincter, iridodialysis, partial or complete aniridia, and injury to the trabecular meshwork may also occur.(1) Eye injury can also cause lens dislocation. Blunt trauma that compresses the eye from front to back can lead to lateral, superior, and inferior expansion of the eye, potentially stretching the zonules that support the lens.(2) Symptoms of lens dislocation include blurred vision, difficulty seeing clearly up close and/or far away, and double vision in one eye.(3)

Combined condition of post-traumatic aphakia and aniridia can occur and is a challenging situation which demand reconstructive surgery that is extensive and complex. There are several options to choose from when managing this condition, which needs case by case considerations. This case report demonstrates the challenging condition and management of a patient with aphakia due to posterior lens luxation and aniridia, following a blunt trauma.

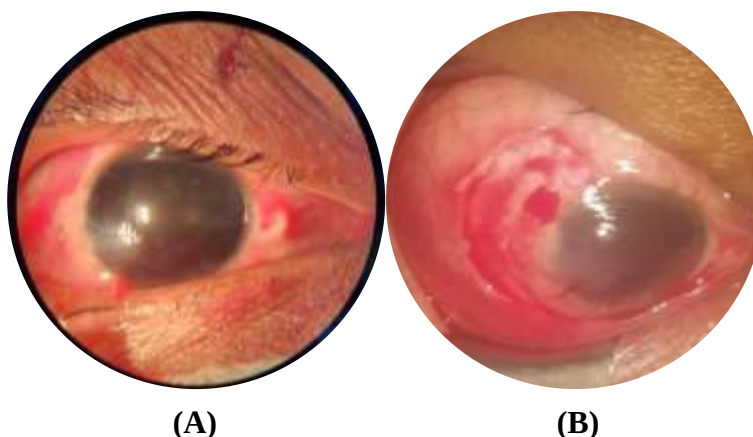
2. Case Presentation

The patient in this case is a 34-year-old male who came to outpatient clinic as an old patient with blurred vision on his right eye. The patient had a history of eye trauma on his right eye 1.5 years prior, due to work accident when a piece of pipe was broken and its fragment bounced off and hit the patient's right eye. The pipe was about 10cm long. After the incident, the patient complained of pain, blurred vision and bleeding on his right eye. The patient claimed to never had any complaints on both eyes before the accident, apart from the patient's history of needing to wear minus lensed spectacles.



Figure 1. Condition of both eyes 1.5 years prior after trauma showed skin laceration on maxilla region and right eye eyelid spasm (taken with patient's consent, courtesy of Dr. Soetomo General Academic Hospital)

The patient was brought to our emergency department after the incident. At that time, there was a skin laceration on maxillary region and spasm of right eyelid (Figure 1). The diagnosis at that time was right eye globe rupture. Based on Birmingham Eye Trauma Terminology (BETT)(4), the patient's eye injury was classified as open globe, type A (rupture), grade 4 (visual acuity < 1 meter finger counting to light perception), pupil difficult to be evaluated, zone III (posterior to 5mm from limbus) with complication of full thickness corneoscleral rupture, and hyphema (Figure 2). The iris and lens at the time of trauma was difficult to be evaluated because of hyphema, but no iris prolapse through the globe laceration was seen.



(A)

(B)

Figure 2. Condition of right eye 1.5 years ago at the time of eye trauma before (A) and after (B) conjunctival peritomy during surgery to clearly identify corneoscleral laceration (taken with patient's consent, courtesy of Dr. Soetomo General Academic Hospital)

The patient then underwent globe exploration and corneoscleral laceration suture under general anesthesia approximately 24 hours after the incident in our emergency surgery room. After the surgery, the right eye showed sutured corneoscleral laceration with formed anterior chamber (Figure 3). The skin laceration on maxilla region was also sutured.



Figure 3. Condition of right eye 1.5 years prior after globe saving surgery, note the sutured corneoscleral laceration with formed anterior chamber using air bubble (taken with patient's consent, courtesy of Dr. Soetomo General Academic Hospital)

The sutures on cornea were removed during the patient's outpatient control visit approximately 3 months after the surgery, but 2 sutures on 7 o'clock position could not be removed because they were imbedded deep inside the cornea. The sutures of the skin laceration on maxilla region were also completely removed.

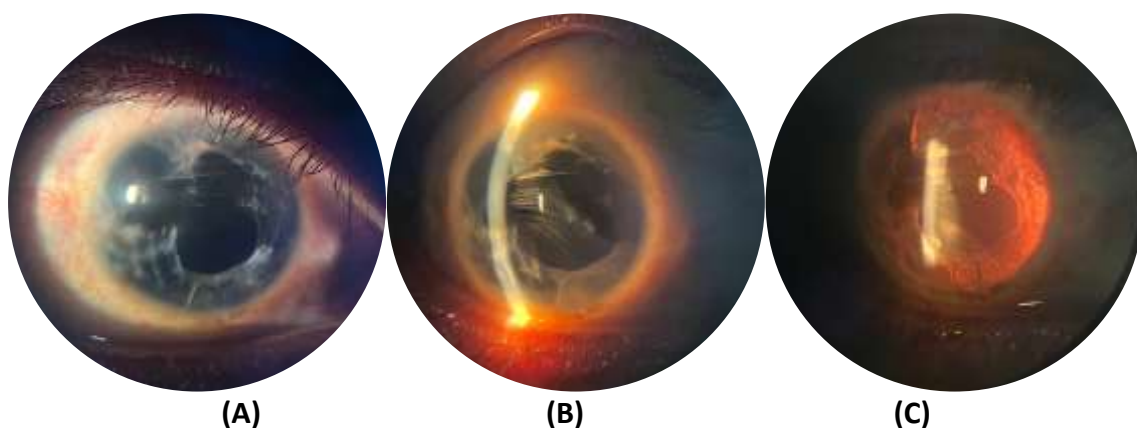


Figure 4 Current condition of right eye through slit lamp examination under diffuse illumination (A), slit illumination (B) and retro illumination (C) showed the absence of iris and lens, with anterior capsule remains and visible vitreous body (taken with patient's consent, courtesy of Dr. Soetomo General Academic Hospital)

On present time, 1.5 years after the incident and surgery, the patient showed significant recovery with no complaints other than right eye blurred vision and occasional glare. No diplopia, redness or eye discharge was experienced by the patient. However, the patient claimed to have worn minus lensed spectacles since the last 10 years but does not remember the exact lens power of his spectacles. The patient's current occupation is a car driver and he would often feel slight disturbance while driving due to his current right eye's condition.



Figure 5 Current unremarkable condition of left eye (taken with patient's consent, courtesy of Dr. Soetomo General Academic Hospital)

Upon latest examination, the best corrected visual acuity of right eye was 6/40 cc S+7.00 C-4.00 X120 → 6/7.5 with no pinhole improvement and left eye was 6/95 cc S-3.25 → 6/6. The intraocular pressure was 12 mmHg on both eyes. Anterior segment examination on right eye showed no eyelid spasm and no conjunctival or pericorneal injection on conjunctiva. There were 2 sutures visible on cornea at 7 o'clock position, with slight corneal neovascularization on 12 o'clock position. On right eye there was an absence of iris and lens, but we can see anterior capsule remains peripherally and vitreous body in the center. The iris of right eye was completely absent without any remains (Figure 4). Left eye condition was unremarkable, with normal iris and pupil (Figure 5)

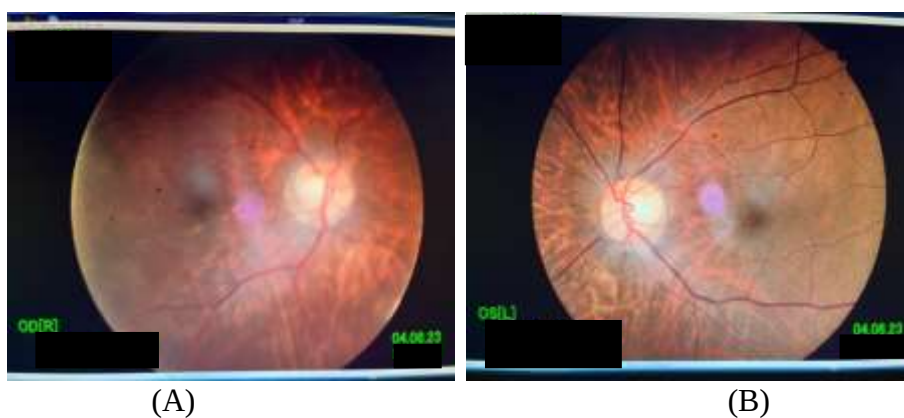


Figure 6. Fundus photograph of right eye (A) and left eye (B) (taken with patient's consent, courtesy of Dr. Soetomo General Academic Hospital)

Fundus examination on both eyes showed positive fundus reflex with normal optic nerve head rim and color. There was a myopic crescent on optic nerve head of both eyes and also tigroid retina

found on both eyes. The macular reflexes of both eyes were normal (Figure 6). Upon fundus examination of right eye using 90D lens and slit lamp, posterior lens luxation was found on the inferior, however this finding was unfortunately cannot be showed through fundus photograph.

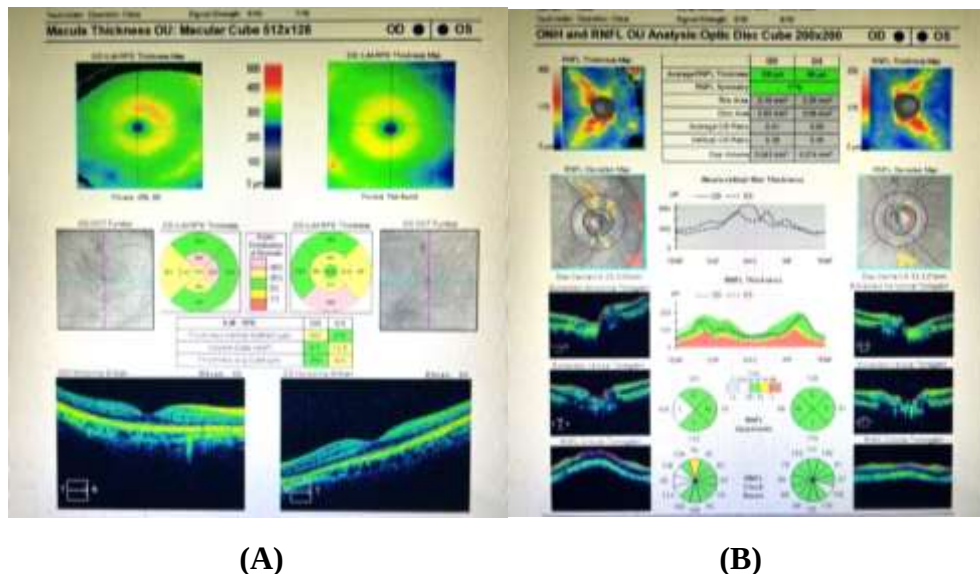


Figure 7. Optic Coherence Tomography (OCT) of both eyes showed normal condition of macula (A) and optic nerve head (B) (taken with patient's consent, courtesy of Dr. Soetomo General Academic Hospital)

Optical coherence tomography (OCT) examination of macular cube and optic nerve head of both eyes were normal (Figure 7). Macular OCT of both eyes showed normal inner limiting membrane – retinal pigment epithelium (ILM-RPE) thickness. Optic nerve head OCT showed normal retinal nerve fibre layer (RNFL) thickness and normal C/D ratio on both eyes.

Based on all clinical and ancillary examination findings, this patient was diagnosed with right eye posterior lens luxation, right eye aniridia, and anisometropia. This patient is originally planned to undergo surgery of vitrectomy, lensectomy, intraocular lens (IOL) implantation in the sulcus with possibly iris prosthesis implantation. If iris prosthesis implantation is not possible to be done, other option for this patient's surgery is vitrectomy with lensectomy and IOL implantation followed by colored contact lens, or conservative management with colored aphakic contact lens. Due to the scarce availability of iris prosthesis in our hospital, surgery was not done in this patient. Instead, the patient was planned to be given colored soft contact lens with the power S+7.00 D (Figure 8). The patient's vision of right eye was able to be corrected to 6/9.5 with no pinhole improvement using the fitted colored soft contact lens.



Figure 8. Patient's right eye after colored soft contact lens fitting (taken with patient's consent, courtesy of Dr. Soetomo General Academic Hospital)

3. Discussion

Posterior lens dislocations are often caused by trauma, where Partial or complete damage to the zonular fibers can result in the lens becoming subluxated or dislocated into the posterior chamber.(5) Blunt eye injuries often cause disruption of zonule fibers, leading to the dislocation of the natural lens or intraocular lens (IOL) implant into the vitreous body or anterior chamber in 9% of cases.(6) Posteriorly dislocated lens can often be seen on the vitreous body during fundus examination. In this case, the patient had a history of blunt trauma when a 10 cm fragment of a broken pipe bounced off and hit his right eye. The traumatic impact at the time of the incident was possibly high enough that it caused corneoscleral rupture and posterior lens luxation.

Posterior lens dislocation can cause several complications, which are secondary glaucoma, retinal detachment, cataract, and vision loss. Refractory glaucoma, persistent uveitis, or corneal damage are conditions following posterior lens dislocation that require urgent or emergency intervention.(3)

Management of lens dislocation depends on the direction of the dislocation. Anterior lens dislocation often disrupts the anterior chamber and cornea, therefore usually requires lens removal surgery. On the contrary, posterior lens dislocation may be managed conservatively by correction with aphakic contact lenses.(3) If there are no sight-threatening complications like elevated IOP or corneal decompensation, conservative management may be preferred for posterior lens dislocation, particularly in patients with good vision in the other eye or those who are not medically fit for surgery. This conservative management includes aphakic contact lens fitting for visual rehabilitation and regular follow-up to be aware of possible complications that may require surgical intervention such as increased IOP, bullous keratopathy, cystoid macular edema, retinal break, and retinal detachment.(7) For successful contact lens fit, the contact lens must not cause discomfort for the wearer, and there should be significant visual acuity improvement with the use of contact lens.(8)

Lens removal surgery for posterior dislocations should be considered in patients who cannot tolerate correction, experience symptoms despite correction, or develop other sight-threatening complications as mentioned earlier. Additionally, if the patient needs surgery for another posterior chamber condition, lens removal may be performed at the same time.(3) The method of surgery for traumatic posterior lens dislocation depends on the surgeon's expertise or preference, usually require pars plana vitrectomy and lensectomy, followed by intraocular lens implantation.(6)

The patient in this case fortunately did not experience complications other than decreased vision. The patient's vision of right eye was decreased but fortunately can still be corrected to reach visual acuity of 6/9.5. The patient's IOP was also normal with no signs of uveitis and severe corneal damage. The patient also showed normal retinal condition evaluated through fundus examination and OCT. However, managing this patient's condition of traumatic posterior lens dislocation becomes challenging due to the presence of aniridia.

Aniridia is a condition where the iris or parts of the iris is absent, which can be congenital or acquired. This condition can significantly affect visual quality because an intact iris is critical to determine the amount of light that enters the eye through pupil size and also increase the depth of focus during accommodation process.(9)

Congenital aniridia is a rare genetic eye condition characterized by the complete or partial absence of the iris from birth. In most cases, this condition is caused by a mutation in the PAX6 gene, which affects various structures of the eye. Unlike those with acquired aniridia, individuals with congenital aniridia are often diagnosed in early infancy due to the associated panocular abnormalities, including iris hypoplasia (aniridia), nystagmus, foveal hypoplasia, ptosis, and visual impairment.(10) The most common cause of acquired aniridia is ocular trauma. Blunt injuries that cause anteroposterior compression of the globe can overstretch the anterior chamber angle and iris, which can rupture the

blood vessel and root of the iris and produce iris tear and hyphema. Injury to the iris can occur as tears of the iris sphincter, iridodialysis, partial or total aniridia, and trabecular meshwork damage.(1)

In this patient's condition, it is unclear whether the aniridia condition is caused by trauma or if it is already present before the trauma as a congenital disorder. The patient did not have a history of eye complaints before the blunt trauma, and the aniridia condition is unilateral with no associated ocular anomalies such as nystagmus, or visual impairment. Therefore, the possibility of congenital aniridia in this patient is very small. However, there is a total absence of iris in this patient, with no remaining iris visible. Considering the initial condition after trauma that no prolapse iris was found; it is also a very peculiar thing if the aniridia condition of this patient is solely caused by trauma.

There are several options to restore pupil size and function for people with aniridia. Some of those options are colored contact lens, corneal tattoo, micro tying sutures, and implantable iris prosthesis.(9) Management of iris defect using sutures can be done if the iris defect is in less than 2 clock hours and for iridodialysis conditions.(1) In this case, the aniridia was total, therefore the option of iris sutures can be ruled out. The possible management option for aniridia of the patient in this case is colored contact lens and implantable iris prosthesis. However, because the patient is also aphakic due to posterior lens dislocation, the management of this patient needs to consider the combined condition of aphakia and aniridia.

Combined condition of post-traumatic aphakia and aniridia can occur and is a challenging situation which demand extensive and complex reconstructive surgery. The goal of therapy is to provide best possible visual outcome by implanting both iris prosthesis and IOL, which often needed to be fixated to the sclera to provide stable position.(11)

There are several options for reconstructive management of iris defects, mainly based on the size of iris defect and also the condition of the capsular bag.(1) For this case, the iris defect of the patient is more than 2 clock hours and the posterior capsular bag is absent, therefore the possible option management that can be selected for this patient is using CustomFlex ArtificialIris (Figure 9) with suture fixation. In patients with aniridia who experience glare and are being considered for ArtificialIris implantation, it is recommended to first conduct a trial with a cosmetic contact lens to assess whether reducing the pupil size helps alleviate the patient's symptoms.(9)



Figure 9 Two view of the CustomFlex ArtificialIris by HumanOptics (12)

However, due to the scarce availability of artificial iris in our hospital, and also the absence of serious complication of posterior lens luxation in this patient such as uveitis or glaucoma, surgery was not done in this patient. Instead, the patient was treated with colored soft contact lens to correct his condition of aphakia, aniridia and anisometropia.

4. Conclusion

Combined condition of post-traumatic aphakia and aniridia is a challenging situation which required complex reconstructive surgery with many considerations. Posterior lens luxation without sight threatening complication can be managed conservatively. However, vitrectomy and lensectomy surgery on posterior lens luxation needs to be considered if there are complications or if the patient requires other ocular surgery. Aniridia can be acquired through ocular trauma, with treatment options ranging from colored contact lens, corneal tattoo, iris sutures, and implantable iris prosthesis.

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