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Case Report

Complexity in Managing Primary Open Angle Glaucoma Eye with Lens Dislocation

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Abstract

Background: Lens dislocation due to weakness of zonular dehiscence may cause lens detachment from the patellar fossa. Surgeons providing treatment for this case should be prepared for unexpected complications following surgery.

Objective: To report a case of lens dislocation with primary open angle glaucoma and the complexity in its management.

Case Report: A 60-year-old man presented at the clinic with pain and blurred vision in his right eye. A dislocated lens and vitreous body at anterior lens was detected with 6/30 of visual acuity. Optic discs examination revealed glaucomatous cupping along with intraocular pressure (IOP) of 40 mmHg. The therapy plan entailed intra capsular cataract extraction with implantation of posterior iris fixated intraocular lens, combined with trabeculectomy with releasable suture. However, on the first day after surgery, it was noted that the patient suffered hypotonia and choroidal effusion, leading to disrupted vision and prolonged complications.

Conclusion: Lens dislocation with the presence of vitreous in the anterior chamber and high IOP is a difficult case to treat, therefore it is imperative that early detection and appropriate treatment is carried out to ensure optimal final result.

Keywords: *Dislocated Lens; Iris Fixated IOL; Cataract Surgery; Choroidal Effusion; Glaucoma*

Background

Lens subluxation, or dislocated lens, is due to weakness of zonular dehiscence that may lead to complete lens detachment from the patellar fossa. The condition often causes complications during lens extraction surgery [1]. The etiologies of weak zinni' zonular are associated with pseudoexfoliation syndrome [1,2], ocular trauma [3], laser peripheral iridotomy [4], uveitis [5], retinitis pigmentosa [6], age, diabetes mellitus [7], Marfan syndrome [8], Weill-Marchesani syndrome [9] and in many cases, the etiology is idiopathic [7].

Symptoms of zonular dehiscence may be chronic or acute, and include various forms of reduced vision. Clinical signs detected are tremulous iris, phacodonesis with or without increased intraocular pressure (IOP), or the presence of vitreous in pupillary aperture. Those surgeons providing treatment for such cases should be prepared for unexpected situations during and after surgical intervention. Modified surgical procedures for lens dislocation have been reported in many cases depending on the area of lens zonular dehiscence, the etiologies, and selection of IOL (intraocular lens) that are to be implanted [10,11].

Moreover, postoperative hypotonia, due to aqueous leakage, may occur leading to structural complications, such as choroidal effusion, cystoids macular edema, or even permanently reduced vision [12,13]. This study reports the dynamic complexity management of patient with dislocation lens and high IOP. Detailed management of this patient is documented and the outcome revealed that the patient's vision was restored successfully.

Case Report

A 60-year-old man was admitted to hospital one day prior to admission with complaints of redness in the right eye, blurred vision, pain around the eye with no nausea nor vomiting. Patient had history of diabetes mellitus, hypertension, shortened blood coagulation and well-controlled recurrent asthma. There was no family history of glaucoma. Reports obtained from other hospitals showed that examinations two months ago revealed vision of right eye to be 6/7.5, IOP 32 mmHg, also a senile cataract was detected. On his left eye, the visual acuity was 6/6 and IOP was 24 mmHg. In both eyes, no abnormalities were found in anterior segments and retinal examination showed no signs of diabetic retinopathy. Patient did not receive any form of treatment for high IOP.

On eye examination it was found that visual acuity of his right eye was 6/30 and not correctable. Visual acuity measured in left eye was 6/6. Examination using Goldmann applanation tonometry showed IOP of the right eye to be 42 mmHg, while the left eye was 30 mmHg. Slit-lamp examination of the right eye revealed hyperemic conjunctiva, edematous corneal epithelium, deep anterior chamber with no cells or flare, and vitreous body appearing from the posterior of the lens into the anterior chamber. There was no tremulous iris around the vitreous body. Grade 2 senile cataract was detected, according to Burrato classification. On the fundus evaluated using super field lens (Volk, USA), a cup-to-disc ratio (C/D ratio) was 0.8 on his right eye and 0.6 on his left eye. There were also deep glaucomatous cupping and nasalization of the vascular mechanism. The other retinal conditions were difficult to determine. On the contrary, examination of the left eye revealed normal conjunctiva, cornea and lens. The diagnosis for the patient was an open-angle primary glaucoma in both eyes and lens subluxation, senile cataract of the right eye.

Steroid and antibiotic eye drops were provided for the right eye qid, and dorzolamide and brimonidine-purite 0.1% eye drops were given bd daily for both eyes. Oral 250 mg acetazolamide tablet was also given 3x daily and Aspar-K® tablet 1x daily. In addition, other medications were also given to control diabetes and hypertension, while treatment of anticoagulation was temporarily discontinued. The patient was then prepared for lens extraction surgery under general anesthesia with implantation of capsular tension ring and anterior vitrectomy with intraocular lens-in-the-bag, followed by trabeculectomy added with mitomycin C(MMC) on the right eye.

Surgery was started with a-biplane corneo-scleral 6-mm incision as the main port, and afterward Ophthalmic viscosurgical device (OVD) was injected into anterior chamber to maintain the depth of the chamber and protect the corneal endothelium. During the procedure, it was found that the lens was actually displaced into the vitreous body, hanging by only one third of a zonule of Zinn. Immediately, the lens was hooking from two sites by two hookers, first from the main port and second from the new port, made 90^{o} apart, aimed to support the lens from the back. OVD was reinjected behind the lens. Subsequently, the whole lens could be retracted towards the anterior chamber. ICCE was then performed and remnant vitreous body was removed from anterior chamber using anterior vitrectomy instrument. Acetylcholine hydrochloride (Miochol®; Bausch and Lomb, Irvine, CA, USA) was injected until pupil size was smaller than 6 mm. Iris claw aphakic IOL was implanted (VerisyeTM- Abbott Medical Optics, Inc). The IOL body was inserted into posterior chamber through the pupil and both claws were enclaved into posterior iris surface using the appropriate instrument. After evaluating the IOL position to ensure it had been placed at the expected site, the corneo-scleral incision was closed using interrupted 10-0 nylon sutures. Thereafter, trabeculectomy with releasable suture supplemented with MMC was performed, and OVD aspirated, removed and replaced with balanced salt solution.

Following surgery, Cravit® and Pred Forte® eye drops were given 6x daily. On the first day following surgery we examined the right eye and found that visual acuity was 1/300 with IOP of 3 mmHg. Hyperemic conjunctiva, edematous cornea with folded-in Descemet's membrane, shallow anterior chamber, loosened corneo-scleral sutures and bleb were well-formed with positive Seidel Test from the cornea limbal area suture. We observed hyphema at 1/3 corneal diameter; however, the position of retro pupillary IOL was satisfactory and the haptic IOL behind iris on central pupil with both clips attached optimally to the iris body. However, the anterior chamber was narrow, the angle remained closed (Figure 1) and the fundus only vaguely evaluated. On USG examination, there was kissing choroidal effusion. On examination of left eye, we found visual acuity of 6/6 with IOP of 14 mmHg. Eye drops were continued and oral steroid with 16 mg methylprednisolone 1 x 2 tablets daily were given. The following day, corneo-scleral resuturing was performed using topical anesthesia.

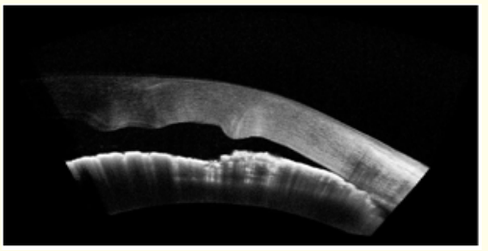

Figure 1: *The anterior chamber was narrow and the angle remained closed.*

A week later, the right eye revealed a visual acuity of 6/60 with IOP of 4 mmHg, marked corneal descemet's membrane folds and the eye chamber well-maintained. However, choroidal detachment still remained. On examination, funduscopy revealed cup disc ratio of 0.8 and glaucomatous cupping with nasalization was also detected. Although no dot retinal hemorrhages, hard and soft exudates, nor neovascularization were found, photo of fundus showed massive choroidal effusion, especially in temporal site, as seen in figure 2. Eye drop treatment for right eye was continued, likewise oral methylprednisolone until the 14th day. However, the retina remained detached and folded. Methylprednisolone oral was continued at the same dose, along with Pred Forte® and Cravit® eye drops 4x daily. The symptoms persisted over two weeks, with IOP 4 mmHg, thus required trans-scleral conjunctival flap covered up trabeculectomy flap. Methylprednisolone was gradually discontinued after 1.5 months along with with Pred Forte® and Cravit® eye drops.

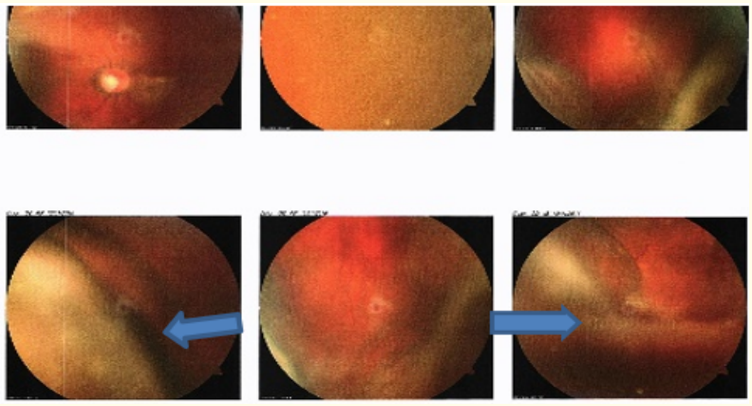

Figure 2: *Cup disc ratio of 0.8 and glaucomatous cupping with nasalization was detected (above) and massive choroidal effusion was seen on day 1 (arrow, below).*

Two months after surgery, visual acuity of right eye was improved to 6/7.5 with IOP of 12 mmHg. Patient presented with clear cornea and well-maintained eye chamber. During follow-up, we carried out visual-filed "Humphrey" analysis, as well as retinal nerve fiber layer by using ocular coherence topography (CIRRUS™ HD-OCT Carl Zeiss, Meditec AG, Germany). Patient was closely monitored, particularly his visual field, the structure of retinal nerve fiber layer and optic disc, according to the procedures of glaucoma management. His visual field revealed double arcuate defect with mean deviation of -23 dB corresponding to flattened superior and inferior area of retina nerve fiber layer (RNFL) on OCT on his right eye. Meanwhile, depression of both nasal areas on visual field with MD-8 dB and of superior RNFL on OCT were detected on left eye. Six months following surgery, treatment by fix combination dorzolamide and brinzolamide eye drops bd was continued in order to manage the IOP and to maintain the intra ocular pressure within safe range (10 - 12 mmHg on his right eye and 12 - 14 mmHg on his left eye).

Discussion

Spontaneous lens dislocation accompanied with prolonged untreated primary open angle glaucoma presents a difficult case to treat. We performed lens extraction combined with MMC trabeculectomy and releasable suture on the eye. During surgery the lens dropped unexpectedly into vitreous; however, the surgeon was able to treat the affected area by holding the crystalline lens [11,14]. The situation occurred when the surgeon opened the cornea and, as a result, IOP dropped with no pressure from the back behind the eye, thus resulting in the luxated crystalline lens slipping back into the vitreous.

Our patient did not show any signs of pseudoexfoliation and there was no history of eye injury. Immature cataract that occurs over the age of 50 is a common degenerative process [1,2,4,7]. Moreover, the patient also had high intraocular pressure (> 40 mmHg) without treatment, which put him at a higher risk for degenerated and disintegrated zonule of Zinn. The progressive zonule dehiscence was worsened by underlying comorbidities, such as diabetes mellitus [7], heart disease and asthma with frequent use of local steroid medication.

In addition, careful observation when performing slit-lamp examination, including findings such as tremulans iris, phacodonesis and vitreous appearance in the pupil or in the anterior chamber may establish earlier diagnosis of lens dislocation [10]. Our patient showed signs of phacodonesis, which was confirmed by the presence of vitreous in front of the lens. High IOP was detected due to vitreous' pupillary blockage of the aqueous humor flow, that confirmed as POAG. It has been reported that ultrasonic biomicroscopy (UBM) can evaluate accurately zonular fragility, laxity or even detachment of zonular fibers [15].

On discovery of such a condition, the surgeon has to predict that unexpected situations might occur during the surgery. Therefore, the appropriate surgical procedures and instruments should be organised carefully. There are various surgical procedures that can be selected depending on the extent of zonular dehiscence, the availability of facilities and surgical technique of cataract surgery [2,10,11,14,16-18].

According to classification by Hoffman, severe lens subluxation is a condition that occurs if lens edge reveals more than 50% of the pupil [10]. In most cases, position of lens displacement is only vaguely evaluated despite dilation of the pupil; however, we may assume that if the position of the lens is still in line with the iris, then this would indicate a mild lens subluxation only, since the remaining zonular fibers are still healthy and strong. If there is an obvious tremulous iris, then it may be assumed that lens dislocation and dehiscence is quite severe [10].

In the case that the detached zonule of Zinn is less than 1800, the modern surgical procedure of phacoemulsification may still be performed by placing a segmental ring to hold and fixate the posterior capsule, such as the capsular tension ring (CTR) [10,19]. However, if the zonular dehiscence is more than 1800, a direct whole lens extraction (intra capsular cataract extraction approach) or a similar highly complex technique should be performed. The latter procedure requires the implantation to be sutured into sclera to maintain lens stability in the capsular bag, such as Cionni CTR device implantation in subluxated capsular bag [17].

In a situation when there is no supporting lens capsule, selecting a proper IOL is an essential step. There are three options of implantation procedures; namely, an anterior chamber (AC) lens, an iris fixation (claw) IOL or a scleral fixation posterior chamber (PC) IOL [18-23]. AC IOL implantation was the standard procedure in the 80's, followed by PC sclera fixation [10,20,22]. On years later, it was known that AC-IOL could lead to secondary glaucoma, as well as the increasing rate of corneal endothelial loss.24 Various techniques have been demonstrated to date, such as fixating IOL haptic under the sclera flap with 10-0 or 9-0 sutures, or tissue glue [10,19,20,24].

Iris fixation IOL with claw haptics imbricated on the iris surface gained popularity as it simplified the surgical technique by enclaving it into iris surface [21-26]. On the other hand, iris fixation (claw) IOL is also prone to exacerbate corneal endothelial damage if placed anteriorly. Recently, there appears to be an innovation trend to enclave this lens in the posterior surface of the iris. This method demonstrates a long-term effect as no damage is caused to corneal endothelium [28,29]. However, this approach certainly needs higher surgical skills since direct or indirect complications may arise in the case that the claw haptic is not well-enclavated into the iris, resulting in the IOL falling into the vitreous or even into the retina [28-30].

Hypotonia is another complication that may occur as a result of aqueous leakage due to loosened cornea limbal suture, loosened releasable suture, or as a complication after trabeculectomy. The very low IOP may lead to visual disability, choroidal effusion, maculopathy, flat anterior chamber, optic neuropathy and cataract. These complications are often resolved by suturing the leakage [12,13,31]. Ocular ultrasound is useful in distinguishing the differences among other choroidal lesions, such as haemorrhage, tumor mass or inflammation. Choroidal effusions are usually painless, presenting as a shallow anterior chamber and low IOP with typically hypotony maculopathy and blurred vision. Cycloplegia adjusts the anterior chamber and angle due to posterior rotation of the iris-lens diaphragm. Topical steroids are advisable to reduce inflammation, often a contributing factor for serous choroidal effusion. If choroidal effusion still persists, oral steroid treatment, as anti-inflammatory dose, serves as treatment of choice to suppress further inflammation [31-33]. In the case that massive choroidal effusion occurs, the integrity of anterior chamber should be restored immediately; however, immediate correction of the loosened corneo-scleral sutures may be insufficient to resolve hypotonia.

There might be a possibility of another overfiltration aqueous leakage from flap sclera, requiring trans-scleral conjunctival flap resuturing [34]. In some cases, a number of hypotonia eyes could be left untreated for longer than 6 months; however, early intervention will certainly result in a more optimal end result. Prolonged hypotonia may cause irreversible fibrosis within the retina and sclera. Up until now, many techniques have been introduced to overcome hypotonia, such as bandage lens, intra bleb autologous blood injections and laser therapy (argon, Nd:YAG) or simple flap suture [35,36]. After long-term conjunctival compression suture, IOP increase has been shown to lead to restoration of the architecture of the retina, allowing realignment of photoreceptor and visual recovery [33,34,37].

Two months following procedure, it responded positively to treatment and hypotonia was resolved immediately, along with associated serious complications. Although the visual acuity of this patient has not been restored completely due to either corneal astigmatism or the possibility of tilted or decentered IOL apposition, the condition is nevertheless well-tolerated by using spectacle correction. Moreover, the patient is now able to perform confidently his daily working activities.

Conclusions

Lens subluxation or dislocation, with the presence of vitreous in the anterior chamber with high IOP is a difficult case to treat, and must be managed appropriately without delay. Surgical preparation for any unexpected situations must be anticipated and carried out by competent surgeons in an established eye-care facility. Post-operative hypotonia with choroidal effusion may lead to permanent disrupted vision. This situation necessitates early reintervention. We were able to demonstrate that well-timed management and attentive observation can lead to optimal restoration of vision.

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