CASE REPORT

Late Onset Uveitis-glaucoma-hyphema Syndrome with Out-the-bag Placement of Intraocular Lens

Benjamin Zhou[1], Vladislav P Bekerman[2], David S Chu[3], Albert S Khouri[4]

Received on: 22 May 2021; *Accepted on:* 29 June 2021; *Published on:* 23 January 2023

Abstract

Aim: To report a case of uveitis-glaucoma-hyphema (UGH) syndrome secondary to a tilted toric intraocular lens (IOL).

Background: Over the past few decades, upgrades in lens design, surgical techniques, and posterior chamber IOLs have drastically decreased the incidence of UGH syndrome. We present a rare case of UGH syndrome developing 2 years after a seemingly uneventful cataract surgery and its subsequent management.

Case description: A 69-year-old female presented with episodes of sudden visual disturbance in her right eye 2 years after a seemingly uneventful cataract surgery with placement of a toric IOL. Workup included ultrasound biomicroscopy (UBM), which revealed a tilted IOL and confirmed haptic-induced iris transillumination defects consistent with the diagnosis of UGH syndrome. The patient underwent surgical repositioning of the IOL, which led to the resolution of UGH.

Conclusion: Uveitis-glaucoma-hyphema developed from a tilted toric IOL inducing posterior iris chaffing. Careful examination and UBM revealed the IOL and haptic out of the bag position, which was critical in determining the underlying UGH mechanism. The surgical intervention led to the resolution of UGH syndrome.

Clinical significance: In patients with a history of uneventful cataract surgery who develop UGH-like symptoms, continued examination of implant orientation and haptic position is critical in preventing the need for future procedures.

Keywords: Cataract surgery, Out-the-bag intraocular lens delayed dislocation, Uveitis-glaucoma-hyphema syndrome.

Journal of Current Glaucoma Practice (2022): 10.5005/jp-journals-10078-1381

Background

Uveitis-glaucoma-hyphema syndrome was initially described in 1978 by Ellison as a condition of iris chafing caused by anterior chamber IOLs, classically presenting as a triad of uveitis, glaucoma, and hyphema. Iris chafing breaks down the blood-aqueous barrier, triggering an inflammatory cascade and causing hyphema. The inflammatory cascade can lead to obstruction of the trabecular meshwork, causing an increase in intraocular pressure (IOP). First-line medical therapy involves the use of anti-inflammatory agents like topical steroids and pressure-lowering medications for temporary relief; often, however, the IOL must be removed or repositioned to treat the culprit. Although first described in anterior chamber IOLs, UGH syndrome has been associated with a variety of other causes, including posterior in-the-bag and out-the-bag IOLs, and iris-supported IOLs. Here, we describe a case of a 69-year-old female who developed UGH syndrome 2 years after an uneventful cataract surgery due to a tilted toric posterior chamber IOL.

Case Description

A 69-year-old female was initially referred for neuro-ophthalmic care at our institution following two episodes of sudden painless visual disturbance in her right eye. Her past medical history was significant for mitral valve prolapse, scoliosis, hypertension (controlled with medications), and osteopenia. Past ocular history was significant for uneventful bilateral cataract extraction with implantation of posterior chamber toric IOL 2 years prior to presentation. She awoke to both episodes with transient visual loss, reporting "everything (was) blurred," that lasted 12 hours before resolving. She sought medical care after experiencing her second episode. At this evaluation, her right IOP was 42 mm Hg. Topical steroids and three topical IOP-lowering agents were used to control this episode. Due to the unclear etiology of the patient's visual symptoms, a neurologic workup was also conducted, including magnetic resonance imaging, echocardiogram, and carotid ultrasound. All imaging was interpreted as normal.

On our initial examination, there was no relative afferent pupillary defect, and uncorrected visual acuity was 20/25 + 2 right eye [oculus dexter (OD)] and 20/25 – 1 left eye [oculus sinister (OS)]. IOP was 18 mm Hg OD and 14 mm Hg OS. Humphrey visual fields were full in both eyes [oculus uterque (OU)], and optical coherence

[1,2,4]Institute of Ophthalmology & Visual Science, Rutgers New Jersey Medical School, Newark, New Jersey, United States

[3]Institute of Ophthalmology & Visual Science, Rutgers New Jersey Medical School, Newark New Jersey, United States; Metropolitan Eye Research and Surgery Institute, Palisades Park, New Jersey, United States

Corresponding Author: Albert S Khouri, Institute of Ophthalmology & Visual Science, Rutgers New Jersey Medical School, Newark, New Jersey, United States, Phone: +19739722045, e-mail: albert.khouri@rutgers.edu

How to cite this article: Zhou B, Bekerman VP, Chu DS, *et al.* Late Onset Uveitis-glaucoma-hyphema Syndrome with Out-the-bag Placement of Intraocular Lens. J Curr Glaucoma Pract 2022;16(3):205–207.

Source of support: Nil

Conflict of interest: David S Chu: Grant Support: Mallinckrodt; Consultant: Aldeyra, Bausch and Lomb, Dompé, Kriya, Santen. Speaker Bureau: Bausch and Lomb

tomography showed superior thinning OD. Fundus exam was within normal limits with vertical cup-to-disk ratios of 0.4 in both eyes. Initial assessment ruled out a neurologic etiology, and a uveitic workup was recommended due to the presence of inflammation.

After continuing to experience recurrent episodes of blurred vision, the patient was referred to our glaucoma clinic 5 months after initial presentation under a presumptive diagnosis of uveitic glaucoma. She remained on topical steroids and three IOP-lowering agents at this time. Visual acuity was 20/20 OD and 20/25 OS; IOP was 20 mm Hg OD and 16 mm Hg OS. Examination at this time was notable for a superior crescent-shaped transillumination defect superiorly in the right iris (Fig. 1A). Due to the recurrent spontaneous episodes of blurred vision, UBM scan was done to evaluate the position of the IOL. UBM of the right eye showed a tilted IOL with its superior haptic in the sulcus (Fig. 1B). These findings confirmed a diagnosis of UGH syndrome.

The patient underwent surgery to reposition the right haptic. Three months following lens repositioning, the patient was tapered off all topical medications. Visual acuity was 20/25 + 1 OD and 20/20 – 1 OS with IOP of 15 mm Hg OU.

Discussion

Uuveitis-glaucoma-hyphema syndrome can progress insidiously, presenting years after uncomplicated cataract surgery and IOL placement. Although initially described as anterior chamber IOLs causing iris chafing, UGH syndrome can commonly be caused by the malposition of a single piece IOL in the sulcus.[1,2] Sometimes, the haptic is placed in the sulcus during initial positioning due to poor pupillary dilation or IOL dislocation from the bag into the sulcus.[1] In an autopsy study in 1986, 47% of 75 eyes examined were found to have one haptic in-the-bag and one in the sulcus placement; only 32% of eyes had both haptics in-the-bag.[3]

While in-the-bag haptic placement greatly reduces the risk of developing UGH syndrome, cases where UGH syndrome developed from in-the-bag haptic IOL placement have been reported. Various causes include displacement of an IOL optic by an iridociliary cyst,[4] anterior bowing of a haptic due to IOL deformation during initial placement,[5] a tear in the capsule causing rubbing against uveal tissue by a protruding haptic,[2] chafing of the iris due to pseudoexfoliation syndrome and pseudophacodonesis,[6] and from extensive capsule fibrosis causing formation of a Soemmerring ring.[1] In addition, posterior chamber IOL dislocation as a late complication of cataract surgery with both in-the-bag and out-the-bag IOL placement has been reported with increasing frequency recently.[7]

In the case described, we suspect that the mechanism of UGH syndrome is due to out-the-bag misplacement of a toric IOL haptic during the time of initial lens insertion. Alternatively, this may have occurred during intraoperative manipulation or rotation as is needed with toric IOL axis alignment. A larger capsulorhexis diameter may also contribute to the haptic "slipping" out of the bag during rotation. Over time, the tilted IOL resulted in the superior haptic chafing against the posterior iris causing anterior chamber inflammation and hyphema. Interestingly, the patient's visual acuity in this eye remained at 20/20 for 2 years, and outside of the transient blurring episodes, the patient did not complain of progressive blurring, distortions in vision, or dysphotopsias. The initial inflammation and IOPs were controlled by corticosteroids and IOP-lowering drops while surgery was considered. Importantly, UBM was critical in determining the underlying cause of UGH syndrome and determining future management. After surgical repositioning of the haptic, no further recurrences of visual loss were noted on follow-up.

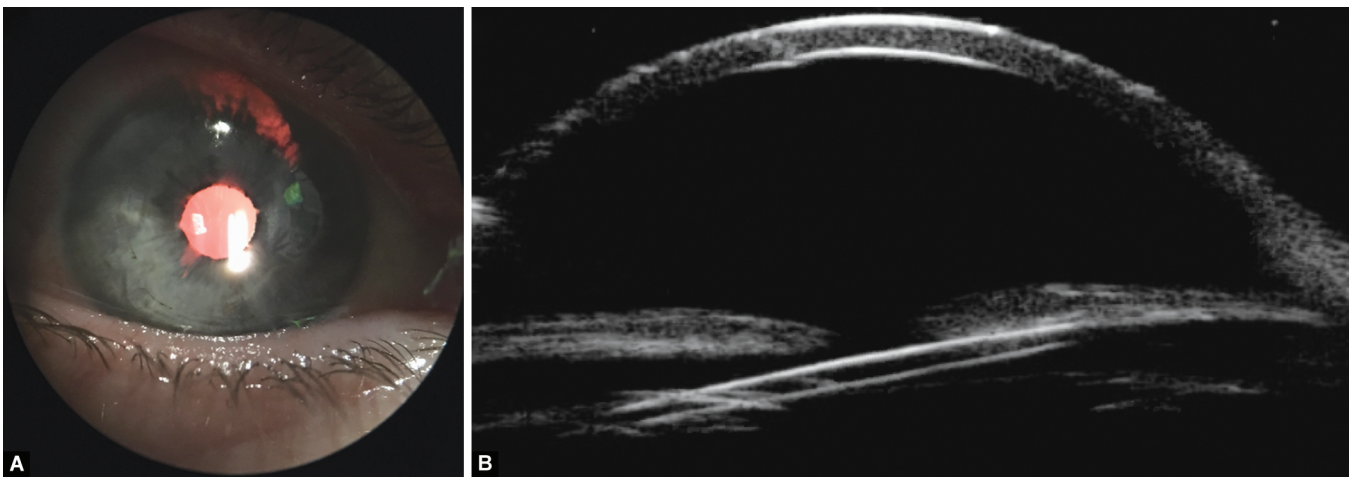
In patients with a history of uneventful cataract surgery who develop UGH-like symptoms, carefully examining the implant orientation, and haptic position is critical in determining the underlying mechanism. UBM is a helpful tool to help illustrate posterior chamber anatomy and confirm diagnosis. Prompt surgical intervention focused on repositioning the IOL in the capsular bag can eliminate and reverse the morbidities of UGH syndrome.

Conclusion

With advancements in surgical technique and IOL design, the incidence of IOL haptic malposition in the sulcus has decreased.[2] Nonetheless, its occurrence should not be overlooked, and all cataract and refractive surgeons should keep a high level of suspicion for this possibility even after favorable refractive outcomes.

Clinical Significance

To our knowledge, this is the first case of UGH syndrome being reported secondary to a tilted toric IOL with a delayed presentation. Continued examination of implant orientation and haptic position is critical in preventing the need for future procedures.



Figs 1A and B: Uveitis-glaucoma-hyphema syndrome on clinical exam. Slit lamp photography of a crescent-shaped transillumination defect of the superior right iris (A). UBM of the tilted IOL with superior haptic in the sulcus and evidence of iris-haptic touch (B)

Orcid

Benjamin Zhou https://orcid.org/0000-0002-0723-4702

References

1. Bryant TK, Feinberg EE, Peeler CE. Uveitis–glaucoma–hyphema syndrome secondary to a Soemmerring ring. J Cataract Refract Surg 2017;43(7):985–987. DOI: 10.1016/j.jcrs.2017.07.002
2. Badakere SV, Senthil S, Turaga K, et al. Uveitis-glaucoma-hyphaema syndrome with in-the-bag placement of intraocular lens. BMJ Case Rep 2016;2016:bcr2015213745. DOI: 10.1136/bcr-2015-213745
3. Cheung AY, Price JM, Hart JC Jr. Late-onset uveitis-glaucoma-hyphema syndrome caused by Soemmering ring cataract. Can J Ophthalmol 2019;54(4):445–450. DOI: 10.1016/j.jcjo.2018.09.001
4. Foroozan R, Tabas JG, Moster ML. Recurrent microhyphema despite intracapsular fixation of a posterior chamber intraocular lens. J Cataract Refract Surg 2003;29(8):1632–1635. DOI: 10.1016/s0886-3350(03)00122-6
5. Boutboul S, Letaief I, Lalloum F, et al. Pigmentary glaucoma secondary to in-the-bag intraocular lens implantation. J Cataract Refract Surg 2008;34(9):1595–1597. DOI: 10.1016/j.jcrs.2008.04.054
6. Zhang L, Hood CT, Vrabec JP, et al. Mechanisms for in-the-bag uveitis-glaucoma-hyphema syndrome. J Cataract Refract Surg 2014;40(3):490–492. DOI: 10.1016/j.jcrs.2013.12.002
7. Pueringer SL, Hodge DO, Erie JC. Risk of late intraocular lens dislocation after cataract surgery, 1980–2009: a population-based study. Am J Ophthalmol 2011;152(4):618–623. DOI: 10.1016/j.ajo.2011.03.009