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Case Report

Occult Uveitis-Glaucoma-Hyphema Syndrome Caused by PCIOL with Peripheral Capsular Tear

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Keywords
Cataract surgery · Uveitis-glaucoma-hyphema syndrome · Uveitis/inflammation · Glaucoma

Abstract
Introduction: Uveitis-glaucoma-hyphema (UGH) syndrome is an infrequent but severe complication following intraocular lens implantation, characterized by anterior chamber inflammation and elevated intraocular pressure (IOP). This report presents a rare case of late-onset UGH syndrome induced by a well-positioned 1-piece posterior capsular intraocular lens (PCIOL) with a bulb of the haptics extruding through a peripheral capsular tear in a 90-year-old female, 17 years post-cataract surgery. ***Case Presentation:*** The patient presented with persistent blurred vision, recurrent anterior uveitis, and uncontrolled IOP despite medical therapy. Extensive evaluation, including ultrasound biomicroscopy, failed to identify the underlying cause, necessitating surgical intervention to control IOP. During concurrent goniotomy and canaloplasty, a PCIOL haptics was discovered protruding through a peripheral capsular tear, establishing the diagnosis. Following PCIOL-haptic amputation and goniotomy and canaloplasty, the patient experienced significant improvement in symptoms and IOP control, with complete resolution of UGH syndrome. ***Conclusion:*** This case highlights the necessity of considering atypical causes in persistent postoperative uveitis and IOP elevation and emphasizes the role of surgical intervention in managing complex cases.

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Introduction

Uveitis-glaucoma-hyphema (UGH) syndrome is a rare, but well-recognized complication of intraocular lens (IOL) implantation. The condition typically results from mechanical irritation of adjacent anterior chamber (AC) angle structures, commonly by a malpositioned AC IOLs, sulcus-lying posterior chamber IOLs, soemmering's ring, or cysts [1]. This chafing can precipitate breakdown of the blood-aqueous barrier, thereby releasing pigment, red blood cells, white blood cells, and protein into the AC [2]. What often ensues is the classic clinical phenotype of UGH syndrome: accumulation of mixed cells in the AC, formation of microhyphema or hyphema, and obstruction of outflow structures leading to increased IOP [2]. With advancements in IOL design and composition and surgical techniques, the incidence of UGH syndrome has declined from a mean of 2.2–3% to 0.4–1.2%, according to recent studies [2]. The incidence of UGH syndrome is even more rare in the setting of a well-positioned posterior chamber intraocular lens (PCIOL) within the capsular bag.

In this report, we present a unique case of recurrent, unilateral, steroid-unresponsive anterior uveitis and elevated IOPs presenting 17 years after uncomplicated cataract extraction. Despite appearing in the bag on UBM, a haptics of the in-bag PCIOL pierced through the capsular bag was ultimately responsible for the occult cause of UGH syndrome in this patient. The CARE Checklist has been completed by the authors for this case report, attached as supplementary material (for all online suppl. material, see https://doi.org/10.1159/000538063).

Case Presentation

A 90-year-old woman presented to an ophthalmology clinic with a 1-year history of blurry vision in the left eye. She had a recent history of elevated intraocular pressure (IOP) in her left eye, along with recurrent episodes of AC inflammation. Despite starting her on steroid, maximum-tolerated ocular hypotensive meds, and valacyclovir for a presumed ocular herpetic disease, her IOP remained uncontrolled at 68 mm Hg. She was referred to our glaucoma service for further evaluation. Her ocular history includes uncomplicated bilateral cataract extraction with PCIOL implant 17 years prior to the initial presentation. There was no history of eye or head trauma.

On examination, her visual acuity with correction was 20/60 in the right eye and 20/100 in the left. Her IOP measured by tonometry was 16 mm Hg in the right eye and 48 mm Hg in the left eye. Slit-lamp examination of the left eye demonstrated a clear cornea with microcystic edema, non-reactive pupil with a superonasal transillumination defect, 3+ mixed cells in the AC with pigments, and a well-positioned 1-piece acrylic PCIOL without donesis. Dilated fundus exam was unremarkable. Gonioscopy detected open angle with hyphema nasally and heavily pigmented trabecular meshwork inferiorly in the left eye compared to the pale trabecular meshwork in the right eye. Optical coherence tomography of the retinal nerve fiber layer (RNFL) showed normal findings in the right eye (75 μm) and superior RNFL thinning in the left (71 μm). Ultrasound biomicroscopy (UBM) of the left eye demonstrated a PCIOL in the capsular bag with no iris-haptic touch and no Soemmering's rings or foreign body in the sulcus (Fig. 1). Additional workup for uveitis, including an anterior-chamber tap for virology, chest X-ray, syphilis and Lyme disease screening, and human-leukocyte antigen-B27 serology, all yielded negative results. The patient was diagnosed with UGH syndrome of the left eye.

In order to address her poorly controlled IOP, the decision was made to perform OMNI ™ canaloplasty and goniotomy in the left eye, taking account of her age and her wish to avoid filtering surgery. When the sulcus space was filled with viscoelasticity in order to confirm the

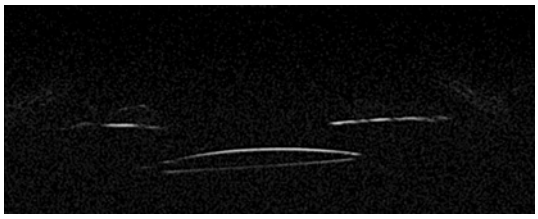

Fig. 1. UBM of the left eye demonstrating a well-positioned PCIOL with no iris-haptic touch.

IOL position, the tip of the superior haptics was found to be protruding outside the bag (Fig. 2). The anterior capsule had 360-degree coverage of the optic indicating that the haptics was breaking through a puncture in the capsular bag peripherally. Dispersive viscoelasticity and a Kuglen hook™ was used to carefully dissect the superior haptics free from the bag. Packer-Chang™ IOL cutters were used to amputate the protruded haptics close to the haptic-optic junction, and the cut haptics was removed from the eye (online suppl. Video 1). The IOL-bag complex remained stable following this maneuver. Ab-interno canaloplasty with goniotomy was performed 360 degree, and all viscoelasticity was removed from the eye, followed by intracameral antibiotics.

At 1 week postoperatively, visual acuity improved to 20/50-1 and IOP was 10 mm Hg on 4 antiglaucoma drops off oral acetazolamide. Slit-lamp exam revealed mild corneal edema and a well-seated PCIOL. At her 1-month postoperative visit, her visual acuity improved to 20/50-1 with correction and her IOP was stable at 10 mm Hg on 3 antiglaucoma drops, and slit-lamp exam findings were unchanged. At her 4-month postoperative visit, her best-corrected visual acuity was 20/30 bilaterally and her IOP was stable at 14 mm Hg without antiglaucoma drops. A slit-lamp examination and gonioscopy revealed a clear cornea with no signs of inflammation or hyphema. The patient was able to discontinue all glaucoma medications and steroid without recurrence of UGH.

Discussion

Originally described by Ellingson in 1977, the pathogenesis of UGH syndrome involves repeated mechanical iris chafing by a malpositioned IOL. Most reported cases occur in patients with ACIOLs or PCIOLs in the ciliary sulcus; however, in-the-bag PCIOLs are not entirely immune to inducing UGH syndrome. Given its rare incidence, this etiology often poses a diagnostic challenge and causes significant ocular morbidity. Zhang et al. proposed pseudophacodonesis of an unstable lens-bag complex secondary to pseudoexfoliation syndrome and focal capsular fibrosis around a square-edged haptics as two possible underlying mechanisms for in-the-bag UGH syndrome [3]. Sura et al. [4] described a similar case implicating a dislocated haptics within the capsular bag displaced by a Soemmering's ring. Interestingly, the mechanism of our patient's intracapsular UGH syndrome involved a superior haptics protruding through a peripheral tear in the bag causing secondary iris chafing and uveal tissue irritation. As a result, she experienced recurrent episodes of AC inflammation and elevated IOP.

UGH syndrome can present anywhere from a few days to several years after cataract surgery. Smith et al. [5] found that patients with PCIOL implantation had a mean time of 4.4 years from cataract extraction to diagnosis. Our patient's postoperative course is unusual as she began to exhibit signs of anterior uveitis and IOP elevation 17 years after cataract extraction. The IOL instability seen in this case could perhaps explain this delayed onset [4]. Moreover, similarly to Badakere et al. [6], the initial tear in the capsule may have occurred

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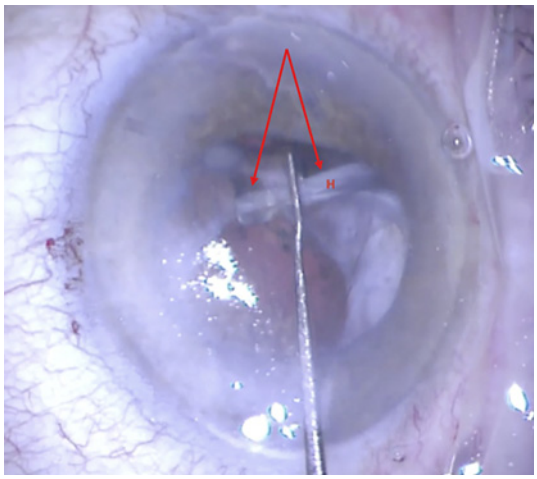

Fig. 2. Image demonstrating a Kuglen hook™ being used to manipulate the haptics (H) emerging from a puncture in the capsular bag peripherally.

when dialing the IOL into the capsular bag. We also speculate that haptic protrusion and subsequent UGH presentation may have occurred due to an unreported fall or trauma.

Prompt diagnosis and effective management of UGH syndrome is crucial to minimize the risk of chronic inflammation, recurrent hyphema, increased IOP, and optic nerve damage. UBM has proven to be indispensable in evaluating the anterior segment for malpositioned IOLs when clinical suspicion for UGH syndrome is high [7]. In our patient's case, however, UBM of the symptomatic eye demonstrated a well-positioned PCIOL with no evidence of iris-haptic touch. Due to her poorly dilated pupil and the peripheral location of the extruded haptics, surgical exploration of the IOL was required in order to ascertain the pathophysiological mechanism at play. In a case with a similar etiology where pupils were reactive, Badakere et al. [6] obtained a successful diagnosis by slit-lamp examination and dilated gonioscopy.

Due to our patient's persistently elevated IOP, goniotomy and canaloplasty was performed to acutely manage her uveitic glaucoma. This decision was predicated on both the patient's preference for minimally invasive surgery and the higher risk of filtering surgery with her age and other medical conditions, including active uveitis [8]. At 4 months postoperatively, our patient's IOP remained at 14 mm Hg off all ocular hypotensive medications. This highlights goniotomy and canaloplasty as a useful approach for the surgical management of uveitic glaucoma with pathophysiology localizing to the trabecular meshwork.

In conclusion, ophthalmologists should be aware of the various underlying mechanisms of UGH syndrome and its potential for delayed onset. Occult UGH syndrome should remain on the differential for pseudophakic patients with steroid-unresponsive, AC inflammation and persistently elevated IOP with normal UBM findings. Doing so can mitigate the risk of unnecessary testing, prolonged steroid usage, and further ocular morbidity.

Statement of Ethics

Written informed consent was obtained from the patient for publication of the details of their medical data and any accompanying images and videos. Ethical approval is not required for this study in accordance with local or national guidelines.

Conflict of Interest Statement

Dr. Jella An reports consulting fees from Alcon and Sight Sciences. All other authors have no conflicts of interest.

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Author Contributions

J.D. contributed to the chart review and literature search and wrote and edited the manuscript draft. Y.B. and O.H. contributed to the literature search and writing and editing the manuscript draft. J.A. and J.S. saw the patient in clinic, performed surgery, and edited the manuscript.

Data Availability Statement

All data generated or analyzed during this study are included in this article and its supplementary material files. Further inquiries can be directed to the corresponding author.

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