An Atypical Presentation and Management of Late-Onset Uveitis-Glaucoma-Hyphema (UGH) Syndrome: A Case Report of Single-Piece Intraocular Lens

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ABSTRACT

We present the diagnostic and management challenges of a rare case of late-onset Uveitis-Glaucoma-Hyphema (UGH) syndrome following uneventful cataract surgery. A 70-year-old female patient presented to our clinic with complaints of blurred vision. Intraocular pressure (IOP) was 48 mm Hg in the left eye. To control the refractory IOP, Ahmed glaucoma valve tube implantation was performed. Postoperatively, IOP decreased to 18 mm Hg. Two months later, the patient continued to report blurred vision. Slit-lamp biomicroscopy revealed a clear cornea, a +++ cell reaction in the anterior chamber with pigments, and a well-centered posterior chamber intraocular lens (PCIOL). Dilated fundus examination showed recurrent vitreous haze/hemorrhage. A pars plana vitrectomy (PPV) was planned to address the vitreous hemorrhage and evaluate the IOL position, which was incompletely assessed due to the mid-dilated pupil. During the procedure, a spatula was inserted under the middilated pupil to reveal adhesions between the iris and the IOL haptic and it was found that one haptic of the IOL was in the sulcus. The IOL and capsular bag complex was explanted. The new IOL was implanted using the flattened haptic end technique. The surgery concluded with PPV to remove the vitreous hemorrhage. Six months later, corrected visual acuity was 20/25 and IOP was 18 mm Hg. This case demonstrates that even a well-centered IOL can, in rare instances, lead to UGH and that a definitive diagnosis may only be established during surgical exploration. Therefore, adopting a multidisciplinary approach is crucial in addressing late-onset UGH syndrome.

Keywords: Uveitis-Glaucoma-Hyphema Syndrome (UGH), Intraocular Lens (IOL), PPV, Vitreous Hemorrhage, AGV

INTRODUCTION

Uveitis-Glaucoma-Hyphema (UGH) syndrome, resulting from the contact between the intraocular lens (IOL) and uveal tissue, was first described by Ellingson in 1978 [1]. This syndrome presents with a broad range of symptoms, including iris transillumination defects, pigment dispersion, intraocular inflammation, hyphema, and elevated intraocular pressure (IOP), or glaucoma. Despite advancements in surgical techniques and modern IOL designs, UGH syndrome remains clinically relevant due to late-onset IOL dislocations, complex surgeries, and secondary interventions [2, 3]. Initially, UGH syndrome was primarily associated with older model anterior chamber IOLs. However, recent reports indicate that UGH can also occur with posterior chamber IOLs placed in the sulcus with insufficient capsular support, or IOLs fixated to the iris or sclera [4, 5]. Notably, single-piece acrylic IOL haptics outside the capsular bag pose a risk for UGH by contacting and rubbing against the iris.

In UGH's uveitis component, findings such as cystoid macular edema, which are easily identified with advanced imaging techniques, stand out [6]. However, in our clinical

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observations, the most challenging pathology to diagnose within UGH syndrome is often glaucoma. Since high IOP in these patients is usually attributed to long-term topical steroid use or angular changes of anterior chamber IOLs, UGH may be overlooked [3]. Additionally, sulcus-placed IOLs, scleral fixation surgery, or IOLs fixated to the iris with uveal tissue contact can contribute to this pathology.Recently, vitreous hemorrhage has been noted as a more common presentation than hyphema in UGH's hemorrhagic component. This typically results from the contact of sclera-fixated IOLs with the ciliary body or iris [7]. In pseudophakic elderly patients without known retinal vascular disease and with posterior vitreous detachment, acute onset of vitreous hemorrhage warrants differential diagnosis of UGH and investigation of IOL-uveal tissue contact. In cases where UGH symptoms are inadequately assessed, some patients may not receive an accurate diagnosis, and the underlying cause can be missed.

Topical treatments are often employed in UGH management due to the possibility of symptom recurrence and prolonged remission periods [8]. However, when IOP cannot be controlled, or progressive glaucomatous atrophy develops, surgical management may be required. Surgical options include IOL repositioning, explantation, trabeculectomy, drainage device procedures, or pars plana vitrectomy.

In this study, we present the diagnostic and management challenges of a rare case of late-onset UGH syndrome following uneventful cataract surgery. The related treatment approaches are discussed.

CASE PRESENTATION

A 70-year-old female patient presented to our clinic with complaints of blurred vision and eye pain. History revealed that she had undergone cataract surgery six months earlier, but about 45 days post-surgery, she developed photophobia, eye pain, and reduced visual acuity. During this period, she was diagnosed with glaucoma and started on topical anti-glaucomatous therapy; however, due to persistently high IOP resistant to medical treatment, a trabeculectomy was performed. Since her symptoms did not improve, she visited our clinic with no history of trauma.

Corrected distance visual acuity was 20/40 in the right eye and 20/200 in the left eye. Tonometry measured the IOP at 14 mm Hg in the right eye and 48 mm Hg in the left eye. Corneal endothelial cell density (ECD) was 1750 mm^2/cells in the right eye and 2304 mm^2/cells in the left eye. Slit-lamp biomicroscopy revealed clear cornea with microcystic edema, superotemporal iridectomy, mid-dilated pupil (approximately 4 mm), and no visible trabeculectomy bleb. There was a ++ cell reaction in the anterior chamber with pigment, and a well-centered single-piece acrylic posterior chamber intraocular lens (PCIOL) within the capsular bag (lens type verified through patient records). Dilated fundus examination revealed vitreous haze, and the retina was attached. Optical coherence tomography showed normal retinal nerve fibre layer (RNFL) in the right eye (88 µm) and thinned in certain quadrants (60 µm) in the left eye. To control the refractory IOP, Ahmed glaucoma valve (AGV) tube implantation was performed under sedation and sub-Tenon anesthesia (**Figure 1**). Postoperatively, IOP

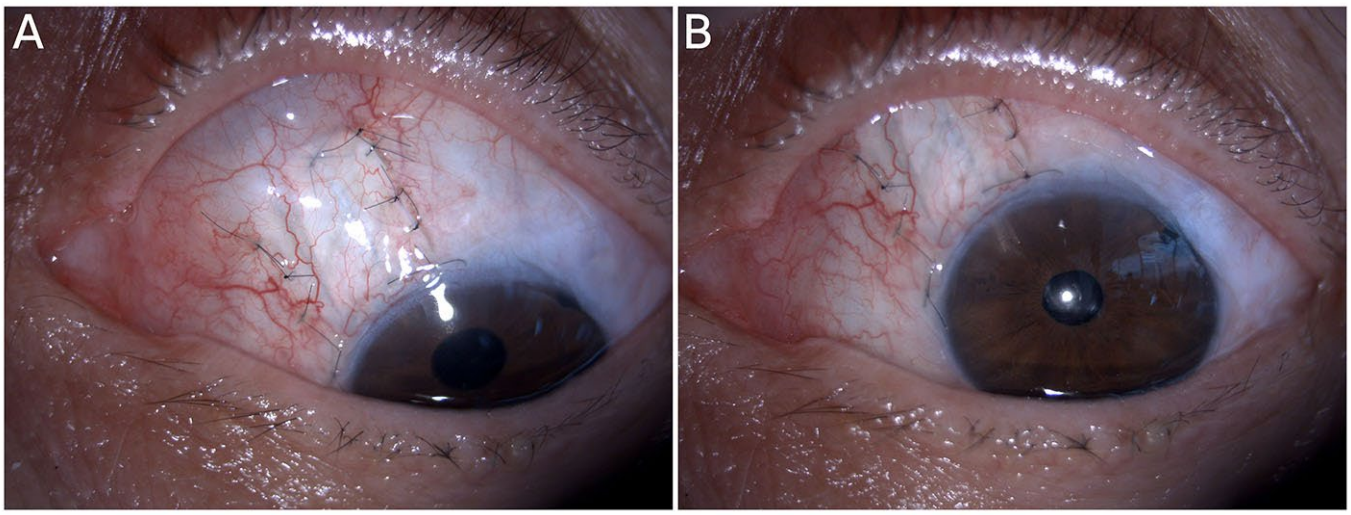

Figure 1. *View of the anterior segment and conjunctiva after Ahmed glaucoma valve (AGV) tube implantation, AGV tube not touching the iris or endothelium.*

decreased to 18 mm Hg and remained stable without the need for additional anti-glaucomatous medication over two months.

At postoperative month two, the patient continued to report blurred vision. Corrected distance visual acuity was 1 meter per second, and IOP was 22 mm Hg. Slit-lamp biomicroscopy revealed a clear cornea, an AGV tube not in contact with the iris or cornea, a +++ cell reaction in the anterior chamber with pigment, and a well-centered PCIOL. However, no transillumination defect or iris chafing was observed on preoperative examination. Dilated fundus examination showed recurrent vitreous haze/hemorrhage. Surgical options were discussed with the patient, and a pars plana vitrectomy was planned to address the vitreous hemorrhage and evaluate the IOL position, which was incompletely assessed due to the mid-dilated pupil. During the procedure, a spatula was inserted under the mid-dilated pupil, revealing adhesions between the iris and IOL haptic. Despite being within the capsular bag, one of the IOL haptics had exited the capsule and adhered to the iris, leading to a UGH diagnosis. Adhesions were lysed to release the haptic. Due to the presence of haptic-iris adhesion and zonular weakness, it was recognised that the IOL could not be placed in the capsular bag and it was decided that the IOL should be explanted.

The single-piece IOL and capsular bag were then grasped with forceps, brought into the anterior chamber, and explanted using the "grasp, pull, and refold technique" [9]. Briefly, in this technique, the leading haptic of the IOL is extracted through the main incision, then the IOL is grasped at the haptic-optic junction and rotated counterclockwise approximately 180 degrees to refold within the corneal main incision for explantation. This prevents contact with the corneal endothelium and the angle structure. A new intraocular lens was implanted using the flattened haptic end intrascleral IOL fixation technique described previously [10]. The surgery concluded with pars plana vitrectomy to remove the vitreous hemorrhage (**Video 1**).

Video 1. Release of the IOL adherent to the iris causing UGH, followed by explantation of the IOL, intrascleral fixation of the new IOL and removal of vitreous haemorrhage by pars plana vitrectomy. Access link: https://www.dropbox.com/scl/fi/nbshdip3zm0lcek18rzze/Video-1.mp4?rlkey=bobg7r2o8n64yg2udyywj17zd&st=yxmz17mi& dl=0

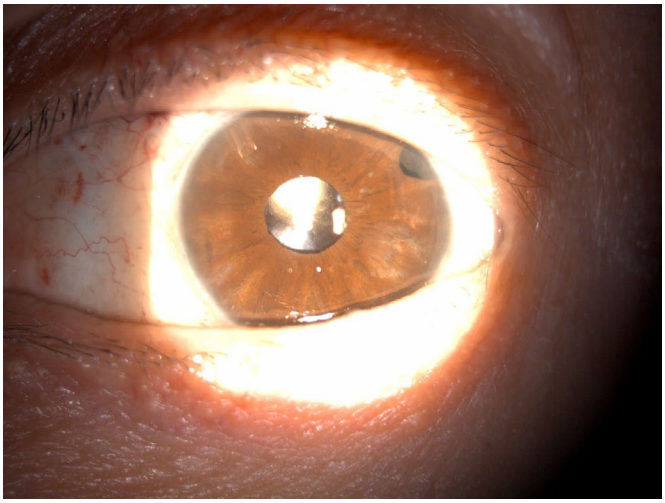

Figure 2. *Slit lamp biomicroscopy at the final visit revealed a clear cornea, an AGV tube not in contact with the iris or cornea, a calm anterior chamber and a well-centred PCIOL fixed to the sclera.*

In the first week postoperatively, corrected visual acuity improved to 20/25, and IOP decreased to 19 mm Hg, remaining stable without anti-glaucomatous therapy. Slit-lamp examination showed a well-centered PCIOL with no anterior chamber reaction or lens tilt. Fundus examination revealed no pathological findings. At follow-up 6 months later, corrected visual acuity was 20/25 and IOP was 18 mm Hg, with no signs of UGH upon slit-lamp and dilated fundus examination (**Figure 2**). At the final visit, postoperative corneal ECD was 1617 mm2/cells in the affected eye.

DISCUSSION

UGH syndrome, historically described as a common complication associated with anterior or posterior chamber IOLs in the ciliary sulcus or scleral IOL fixation, persists as a clinical challenge [4, 5]. Despite significant improvements in modern IOL designs, single-piece acrylic IOL haptics that remain outside the capsular bag continue to present a substantial risk for UGH, especially upon iris contact [3].

Due to the rarity of this syndrome, clinicians may overlook the potential for UGH development in well-centered IOLs within the capsular bag. Moreover, the wide symptom range and diagnostic difficulties in cases with multiple complications make diagnosis even more challenging, leading to a delay in identifying the underlying anatomical problem [3, 11]. For example, Dossantos et al. reported a case of UGH caused by persistent lens-ciliary body friction due to a capsular tear, which went unrecognized [3].

Another significant challenge is the misconception that all three symptoms (uveitis, glaucoma, hyphema) must be present for a UGH diagnosis. However, diagnosis requires only one or more clinical findings along with an associated anatomical abnormality. This insight could aid in earlier recognition of UGH, especially in delayed presentations, and prevent misdiagnosis.

The limitations of current imaging techniques pose another diagnostic challenge. Indeed, on preoperative examination, there was no transillumination defect in the iris and no iris chafing, even in the area of the haptic in the sulcus during PPV. Anterior segment OCT may be inadequate for visualizing the posterior surface of the iris. In such cases, ultrasound biomicroscopy (UBM) can provide clear views of the IOL position and its relation to surrounding tissues, improving diagnostic accuracy. Alternatively, intraoperative endoscopic evaluation or macroscopic examination during surgery can aid in identifying haptics outside the capsule that may be in contact with uveal tissue [11].

Treatment options for UGH syndrome vary based on the underlying cause and severity of symptoms. When medical therapy is insufficient and IOP remains uncontrolled, surgical options are considered. Realignment or explantation of the IOL is often effective, and glaucoma surgery, such as trabeculectomy or the implantation of an Ahmed glaucoma valve, plays a critical role in IOP management in suitable cases. Pars plana vitrectomy is especially beneficial in cases with vitreous hemorrhage [9, 10]. In this case, despite proper IOL centration, the haptic outside the capsule formed an adhesion with the iris, illustrating that UGH's underlying causes may not always be readily visible and can only be definitively diagnosed during surgical exploration. On the other hand, more caution should be exercised in cases with middilated or small pupils which are not dilated enough for the recognition of UGH syndrome and uveal tissue-IOL contact should be investigated. In order to prevent the development of UGH in such cases with small pupils, it should be ensured that the haptics enter into the capsular sac during surgery and the edge of the capsule should be controlled by entering under the iris with Sinskey hook.

If IOL removal had been performed prior to AGV implantation, the patient's IOP—who had previously undergone trabeculectomy—might have normalized. In cases where IOP remained elevated despite IOL removal and additional medical therapy, AGV implantation could then have been considered as a next step. However, following this sequence is not always feasible in clinical practice. The patient exhibited no UGH-related findings during preoperative examinations due to the mid-dilated pupil, and the elevated IOP's association with UGH was unrecognized. This connection only became apparent intraoperatively when haptic-iris contact was observed.

In conclusion, adopting a multidisciplinary approach in managing UGH cases is essential, especially when UGH may coexist with other ocular diseases. In cases where early diagnosis is delayed or anatomical abnormalities are overlooked, UGH may lead to long-term vision loss or persistent glaucoma. Therefore, careful evaluation of symptoms in high-risk patients can facilitate early and appropriate treatment strategies.

This case demonstrates that even a well-centered IOL can, in rare instances, lead to UGH and that a definitive diagnosis may only be established during surgical exploration. Therefore, adopting a multidisciplinary approach is crucial in addressing late-onset UGH syndrome.

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