

CASE REPORT

Uveitis-glaucoma-hyphema syndrome associated with an in-the-bag square-edge intraocular lens

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Abstract

A 54-year-old woman presented with recurrent redness and blurred vision of the left eye with elevated intraocular pressure (IOP) for one year. She was treated as “iridocyclitis” and “Posner-Schlossman syndrome” at the local hospitals. However, the patient developed intermittent ocular inflammation and hyphema. Patient had a cataract surgery and intraocular lens (IOL) implantation in the left eye one year before at the local hospital. A diagnostic procedure was performed and the possible pathogenesis was discussed.

Key words: Uveitis-glaucoma-hyphaema syndrome; chronic postoperative endophthalmitis; intraocular lens design

Presentation of case

A 54-year-old woman presented with recurrent redness and blurred vision of the left eye with elevated intraocular pressure (IOP) for one year. This patient had uneventful cataract phacoemulsification and posterior chamber intraocular lens (IOL) (Acrysof Multipiece MA60MA, Alcon) implantation for both eyes at the local hospital in September 2016. Her personal history included 6 dpt myopia. The patient was discharged with a best-corrected visual acuity (BCVA) of 20/20.

Eleven months after surgery, the patient presented with sudden acute blurred vision, redness and pain in her left eye and was diagnosed with “iridocyclitis” at a local hospital. The BCVA was 20/30, and the intraocular pressure (IOP) was 11 mmHg. The patient was treated with topical tobramycin, dexamethasone, and pranoprofen. The inflammation is steroid responsive initially, however, it was not completely controlled after long-term treatments. Moreover, after 4 months of topical administration, the IOP was higher than 50 mmHg. Prednisolone acetate eye drops were used to replace

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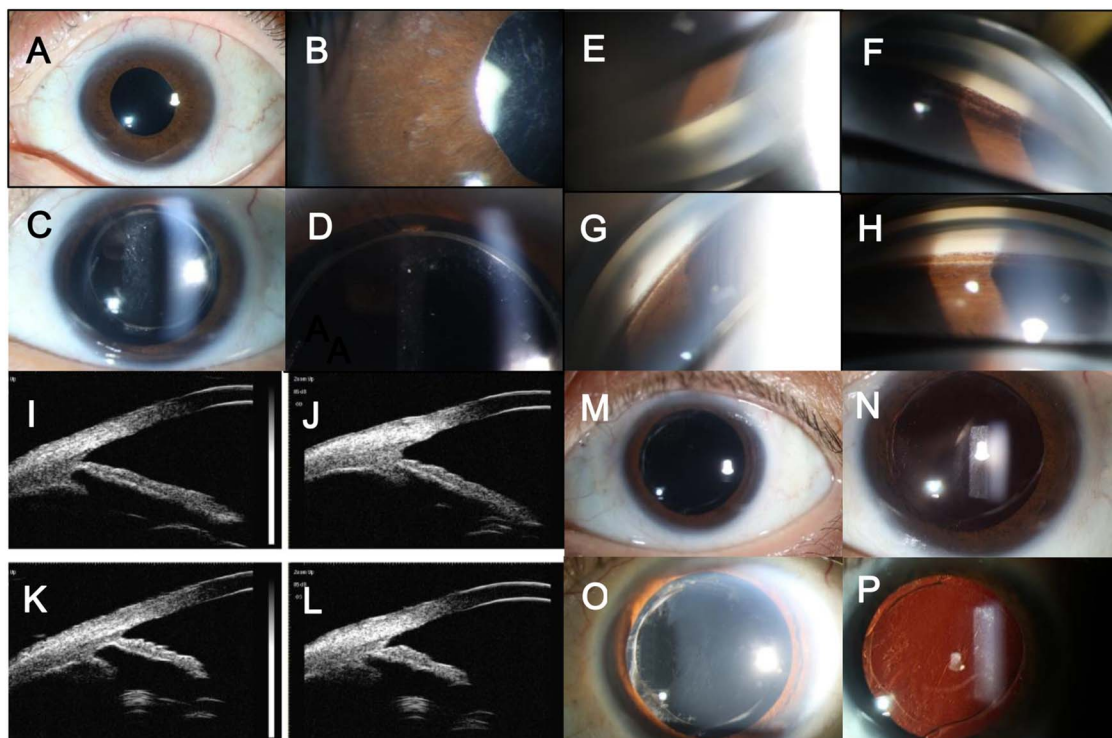


Figure 1. Slit-lamp images and clinical manifestations of eyes before and after treatment. A-D: Slit-lamp images before surgery: iris depigmentation, 3 pieces IOL with square edges uncovered by the capsulorhexis edge. E-H: gonioscopy revealed open angle with unilateral densely pigment accumulation without iris vessel abnormalities or adherence in left eye. I-L: ultrasound biomicroscopy demonstrated IOL-iris contact, slight forward displacement of the 3-pieces IOL with square edges, and the upper optic part of the IOL in close apposition to the iris. M-P: Slit-lamp images after surgery.

tobramycin dexamethasone eye drops, and brinzolamide and timolol maleate eye drops were used to control the IOP. The patient developed intermittent ocular inflammation and hypertension, then she received topical corticosteroids and anti-glaucoma treatment. Over 16 months, she reported six similar episodes, but the remission interval became progressively shorter. She was diagnosed as “Posner-Schlossman syndrome (PSS)” at another local hospital, but there was still no effective improvement.

The patient finally sought help at our hospital in October 2018. The visual acuity (VA) was 20/200, and IOP was 31.5 mmHg after oral administration of acetazolamide. Pigmented keratic precipitates (KP), tyndall (+), red blood cell floating and iris depigmentation was also found in the left eye. The IOL comprised three pieces with square edges of optical parts, which were located in the capsule bag. After the pupil dilated, we found that the IOL was not completely covered by the capsule edge, and a small amount of pearls can be seen on the posterior capsule (Fig. 1, A-D). There was a significant accumulation of brown pigment at the upper edge of the IOL. Gonioscopy revealed an open angle with unilateral dense pigment accumulation without iris vessel abnormalities or adherence in the left eye (Fig. 1, E-H). Ultrasonic biological microscopy

(UBM) (Meda md-300l; Meda Co., LTD., Tianjin, China) showed that the IOL was in contact with the iris (FIG. 1, I-L). The three-piece IOL with square edges was located in the capsule with slight forward displacement. The upper optical part of the IOL was close to the iris. No significant vitreous haemorrhage was found in the fundus. Additionally, no sign of hyphaema, pseudoexfoliation syndrome or pigment dispersion was found in the right eye with a well-centred in-the-bag IOL. The patient told that the blurred vision would get worse after running or exercise.

We diagnosed the case as uveitis-glaucoma-hyphaema (UGH) syndrome. The anterior square edge of IOL resulted in iris chafing which further led to persistent elevated IOP, bleeding and inflammation. To solve the iris friction, the IOL was removed from the capsule to control IOP and inflammation, and was replaced by a hydrophilic acrylic acid posterior chamber IOL (CT Aspina 409m; Zeiss). The one-piece plate-haptic design and overall length of 11 mm of the Zeiss IOL ensured full coverage of the implanted IOL by the capsulorhexis edge. No intravitreal antibiotic was given. The extracted IOL was immersed in 2.5% glutaraldehyde in 0.1 M phosphate buffer solution and was treated by scanning electron microscopy (SEM). The IOP and inflammation were immediately controlled after surgery. During follow-up

of one year after IOL replacement, the patient remained asymptomatic, the BCVA was 20/30 (Fig. 1, M-P), the IOP was less than 20 mmHg without topical hypotensors, and visual acuity was stable. No additional bleeding or pigment dispersion episodes occurred after surgery.

Differential diagnosis

Chronic postoperative endophthalmitis

Chronic postoperative endophthalmitis is considered as an uncommon but sight-threatening complication.¹ It is often misdiagnosed as non-infectious iritis because of topical corticosteroid therapy responsive initially. However, relapses occurred with medication tapering or stopping.² This patient in our case had acute redness in the left eye 11 months after uneventful cataract phacoemulsification and posterior chamber IOL implantation. The successful inflammation control by only topical tobramycin and dexamethasone, in combination with the clinical manifestation of persistent elevated IOP, pigmented KP, anterior chamber reaction and red blood cell floating, all supported the diagnosis of bleeding and inflammation rather than endophthalmitis. The findings of examination including the slit lamp, gonioscopy, and UBM, revealed the iris chafing was caused by the anterior square edge of the IOL.

Posner-Schlossman syndrome

PSS also known as glaucomatocyclitic crisis, is manifested as acute, unilateral, recurrent attacks of elevated IOP accompanied by mild anterior chamber inflammation.³ This patient got elevated IOP after 4 months of topical dexamethasone administration, and such clinical history made the diagnosis of PSS very unlikely.

UGH syndrome

UGH syndrome is a rare complication of intraocular chafing from IOL implants leading to a spectrum of iris transillumination defects, pigmentary dispersion, microhyphemas and hyphemas and elevated IOP.⁴ This syndrome is less often encountered nowadays with the advent of modern posterior chamber IOLs, particularly if the IOL is placed within the capsular bag. It happened usually in patients with sulcus fixation of posterior chamber IOL, improperly positioned anterior chamber IOLs, iris prosthesis or glaucoma drainage implants. According to previous reports, modern IOLs help increase the risk of iris touching due to thicker and sharper edges of IOL haptics.⁵ In this case, anterior chamber reaction and cells, significant accumulation of pigment at the edge of the IOL, and the malpositioned IOL supported the diagnosis of UGH syndrome. SEM showed that red blood cell and membrane-like materials attached on the surface of extracted IOL with the rough anterior square edge. The patient also reported worse symptoms

after exercises, which is also an important clue for UGH syndrome.

Clinical diagnosis

Uveitis-glaucoma-hyphaema syndrome.

IOL exchange

IOL exchanges were performed by Dr. Yi Lu under topical anaesthesia. The three-piece IOL was gently removed from the capsule, and then was dissected and extracted from the main incision. After reopening the capsule, the one-piece hydrophilic acrylic posterior chamber IOL (CT ASPHINA 409M; Zeiss) was implanted into the capsule. The pigment deposited in the anterior chamber was irrigated by cannula irrigation/aspiration (for the surgery process please refer to the Supplementary Material).

Scanning electron microscopy (SEM) of IOLs

IOLs were fixed in 2.5% glutaraldehyde solution in 0.1 M phosphate buffer for 2 h, followed by secondary fixation with 1% osmium tetroxide stationary solution. Fixed IOLs were dehydrated in ethanol-water mixtures with increasing concentrations of ethanol and ethyl acetate. Observations of the IOL surface were performed at 10 kV using SEM (JEOL JSM-6380LV; JEOL Ltd, Tokyo, Japan).

Initial SEM showed red blood cell and membrane-like materials attached on the surface of the extracted IOL (Fig. 2) and no bacterial adherence. The extracted IOL had an anterior square edge and a rough side.

Discussion of management and pathogenesis

UGH syndrome was historically associated with IOLs and had a spectrum of microhyphemas, pigmentary dispersion, inflammation and elevated IOP.⁶ With the advent of modern posterior chamber IOLs and the development of cataract surgery, the incidence of UGH syndrome had gradually decreased.⁴ Recently, however, the syndrome was found to be associated with acrylic IOLs placed in the ciliary sulcus, or even in the capsular bag (the displaced haptic went through the tear of the capsular bag).^{8,9} If the IOL is not placed properly, excessive movement of the lens may result in chafing, pigment dispersion and breakdown of the blood-aqueous barrier, leading to recurrent hyphaema, raised IOP and anterior uveitis.⁵ Because UGH is rarely reported in patients with in-the-bag acrylic IOLs, an early accurate diagnosis may be more challenging, and delayed treatment often leads to significant eye morbidity.^{7,8} Usually, eye discomfort of the patient is not commensurate with the clinical manifestations.

The pathogenesis of UGH in our patients was quite different from previous studies. We propose that the mechanism of UGH syndrome in our patient was the uncovered sharp square edges leading to iridociliary irritation and iris bleeding. The slight forward

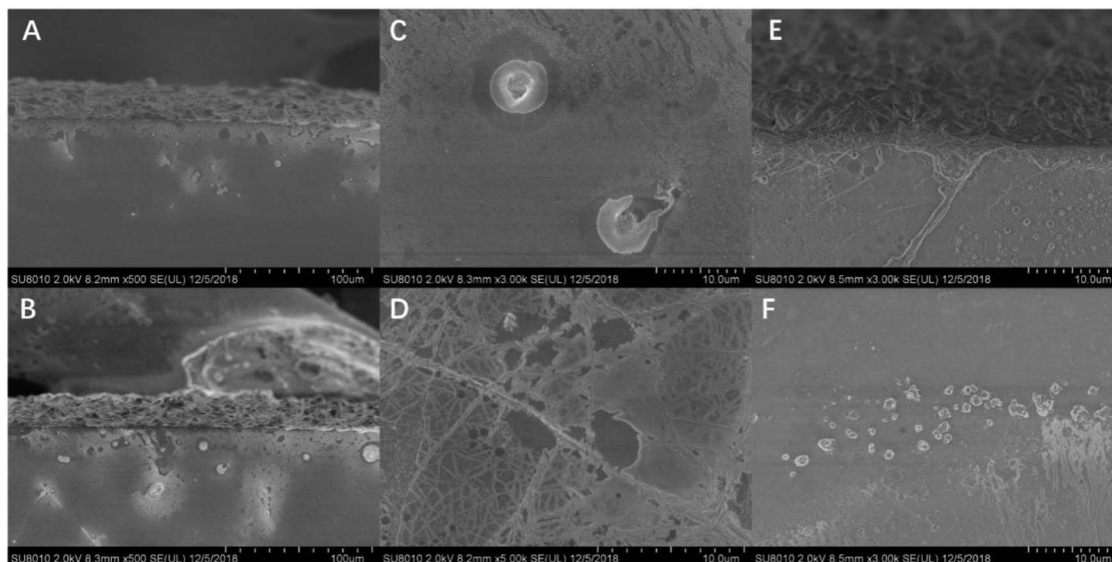


Figure 2. Scanning electron microscopy of exchanged IOL. A-B: anterior square-edge of three-piece of IOL. C: red blood attachment; D: membrane-like materials attachment. E: the rough side of three-piece of IOL. F: tiny fragments attached on the surface.

displacement of the IOL caused chafing and active retro-iridian bleeding in the contact area between the IOL and iris by the uncovered anterior square-edged IOL. However, in previous studies, chafing of the posterior iris in patients with in-the-bag IOL implantation was more often caused by the sharp square-edged haptic, zonular laxity, or IOL optic displacement.⁸⁻¹² In-the-bag sharp anterior square-edged three-piece acrylic IOL is a potential source of iris chafing in certain situations. The mechanism observed in this paper should be considered to explain concisely the pathogenesis of UGH syndrome with in-the-bag IOLs in similar situations.

We had different findings in the SEM of the extracted IOL. In this case, we found red blood cell and membrane-like materials attached to the surface of the extracted IOL. SEM also showed that the anterior square edge of the IOLs implanted in this patient was sharp and rough. In a previous study, SEM showed densely packed coccoid-like structures on the haptic surface that was also proven to be melanosomes.¹³

Given the prevalence of intracapsular acrylic IOL implantation in cataract surgery, it is important to gain new insight into uncommon IOL-related in-the-bag UGH. Because of the difficulty in early diagnosis, it can lead to severe morbidity of patients. In this case of in-the-bag UGH, the symptoms occurred 11 months after cataract surgery. The diagnosis was 16 months after the symptoms appeared, while the patient underwent long-term evaluations and treatments. We found that UBM and gonioscopy were helpful in identifying pigment dispersion, microhyphaema, IOL placement, and iridociliary chafing. In patients with UGH syndrome, topical corticosteroids and anti-glaucoma medication could reduce the intraocular inflammation and control IOL in short term. However, the recurrence of inflammation,

decreased vision and progressive glaucomatous atrophy are indicators of IOL exchange. IOL removal should be chosen rather than conservative management. Because the smooth optical edge has no potential mechanical chafing and is helpful to stabilize the capsular bag and zonule, it is better to use single-piece large-diameter acrylic IOL with a round anterior edge. In previous studies, endoscopic cyclophotocoagulation or local laser iridoplasty was also suggested to resolve extensive haptic-capsule fibrosis.^{7,10} CTR implantation was also recommended to redistribute zonular tension at the capsular equator.¹⁰

Clinicians should be aware that anterior square-edged intra-capsular IOLs may be a potential cause of iridociliary chafing, and previous studies also showed a decrease in edge glare with round edges compared to sharp ones.^{14,15} Previous studies have shown that IOL-edge microstructure and material composition may contribute to reducing posterior capsular opacity (PCO) after cataract surgery.¹⁶ Evidence of a square posterior optic edge in reducing PCO is overwhelming, and the importance of a square-edge structure is widely accepted.^{17,18} However, the uncovered anterior sharp edge of IOL may cause iris chafing and microhyphaema. All these clinical findings indicated that the design of IOL should avoid anterior square edge. Complete coverage of the optic periphery by the capsulorhexis edge and symmetrical centralization of IOL in-the-bag could decrease the prevalence of interaction with the posterior iris surface.

Supplementary material

Supplementary material is available in *Precision Clinical Medicine* online at <https://doi.org/10.1093/pccmedi/pbz026>.

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Conflict of interest

No author has any competing financial interests in relation to the work described. No conflicting relationship exists for any author.

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