

Uveitic Glaucoma with Complicated Cataract and Occlusion Pupillae with Neovascularization

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Abstract

Chronic uveitis encompasses a heterogeneous group of diseases, many of which being idiopathic in origin and associated with a high incidence of vision-threatening complications. This case report portrays management challenges associated with uveitic glaucoma and complicated cataract with occlusion pupillae associated with neovascularization.

A 38-year-old male presented with gradual visual loss and recurrent pain in the right eye, diagnosed as chronic anterior uveitis of idiopathic origin, with uveitic glaucoma, complicated cataract, and occlusion pupillae with iris bombe. Medical management and laser peripheral iridectomy (PI) were performed before the patient reported to us, to lower IOP, led to temporary control of IOP. We found neovascularization of the iris and pupillary membrane with obliteration of iridotomies. Nd: YAG synechiolysis was performed repeatedly. Finally under treatment, after 3 months of quiescence of the eye, Phacoemulsification with synechiolysis and IOL implantation was done. Postoperative inflammation and cystoid macular edema were controlled with oral steroids.

This case highlights all the complications of the disease which were successfully managed. Use of steroids pre and post-operatively, meticulous planning and implementation can improve visual outcome in these types of cases.

Keywords: Uveitic Glaucoma; Occlusion Pupillae; Neovascularisation; Complicated Cataract; Steroid Prophylaxis; Synechiolysis

Abbreviations: PI: Peripheral Iridectomy; LPI: Laser Peripheral Iridotomy; UG: Uveitic Glaucoma; DS: Deep Sclerectomy; MIGS: Minimally Invasive Glaucoma Surgery; IOLs: Intraocular Lenses.

Introduction

Uveitis affects over 2 million people globally, with about 10% experiencing irreversible visual loss [1,2]. It is associated with a high incidence of vision-threatening complications such as cataracts, macular edema, and

glaucoma. Uveitic glaucoma develops due to the disease itself and due to corticosteroids. Managing uveitic glaucoma with complicated cataract presents a significant treatment challenge. Neovascularization, although rare in response to inflammation, can significantly complicate this situation. We report a case of uveitic glaucoma with complicated cataract, and occlusio pupillae associated with neovascularization as the patient regained useful vision after months of treating complications of the disease and to show management of neovascularisation seen near the pupillary margin and on the cataractous lens.

Case Report

A 38-year-old male presented with gradual loss of vision and recurrent pain in the right eye of a few months duration with a history of being treated for anterior segment uveitis elsewhere. A history of three episodes of redness with pain in right eye was elicited, which had responded to topical medication. The patient had no significant systemic complaint. All investigations were done to determine the etiology of the disease but none were positive. These included complete blood count, ESR, Mantoux test, VDRL, serum ACE, Antinuclear antibodies, HLA-B27, Chest X-Ray and sacroiliac joint X-rays, and high-resolution CT scan.

At presentation, the right eye had a visual acuity of hand movement not improving with pinhole and accurate PR, and best corrected visual acuity was 6/6 in the left eye. Intraocular pressure (GAT) was 42 mm Hg in the right eye and 14 mm of Hg in the left eye. Central corneal thickness in the right eye was 531 and 540 microns in the left eye respectively.

Slit lamp examination of the right eye revealed a clear cornea and peripheral obliteration of anterior chamber with central shallowing. The pupil was small, irregular, with a thick fibrous membrane bridging the pupillary area completely. There were no signs of active uveitis of fresh keratic precipitates, aqueous flare or cells in either eye. He had a complicated cataract obscuring the fundal view. Gonioscopy, revealed anterior chamber angle to be closed in all quadrants with peripheral anterior synechiae in more than 270° area in the right eye and the left eye grade 3 (Shaffers' grading) open angles.

The left eye was pseudophakic with a well centered posterior chamber IOL implantation. The reason for early cataract surgery was not available from the patient or documents.

Fundus examination of the left eye revealed CD ratio of 0.8:1. B-scan ultrasonography of the right eye showed normal lens echoes, anechoic vitreous and normal retina-choroid complex.

The patient was started on systemic and topical antiglaucoma medication (Tab acetazolamide 250 mg twice daily and Brimonidine 0.2% + Brinzolamide 1% combination TDS + Timolol 0.5% BD) and cycloplegics (Eye drops Atropine 1%) followed by Nd: YAG laser Iridotomy to lower intraocular pressure and relieve associated pain. Nd: YAG laser Iridotomy had been performed twice at two different sites by other specialists to relieve the pupillary block. IOP decreased to 22 mm of Hg on maximum glaucoma medication and topical Prednisolone acetate 1% 4times/day.

Two weeks later, IOP increased to 30mm of Hg and peripheral iridotomies were found to be obliterated, multiple iridocorneal adhesions were seen with neovascularization on the iris and pupillary membrane (Figure 1A) and multiple iris bombe.

To enable free aqueous flow, Nd: YAG laser synechiolysis was performed in three sittings to perforate the tough fibrous tissue causing occlusio pupillae using low energy (0.6-0.8mJ) in the anterior offset setting, avoiding any laser shots over neovessels. Despite precautions, bleeding occurred from the new vessels around the pupillary margin but resulted in a 2-3 mm nasal opening of the occlusio pupillae. The imperforate iridotomies were also lasered. A small hyphema formed due to bleeding from the pupillary margin which cleared in a week (Figure 1B). The patient was started on 60mg/day oral prednisolone in tapering dose, after excluding diabetes mellitus. IOP decreased with this treatment.

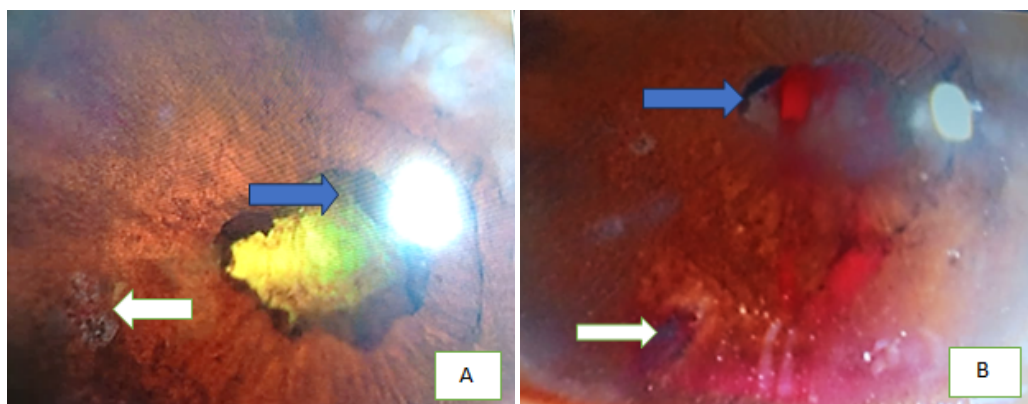


Figure 1: Slit lamp image of the anterior segment.

Figure 1A: shows complicated cataract with occlusio pupillae, neovessel over the membrane (blue arrow), and multiple peripheral iridotomies (white arrow).

Figure 1B: shows slight nasal opening of pupillary membrane and hyphema after Nd: YAG synechiolysis, opening of inferior PI.

Six weeks later, IOP was 16 mmHg (on three AGMs), nasal opening of occlusio pupillae with mild regression of iris neovascularization. The patient was kept on regular follow-up to monitor intraocular pressure and inflammation while gradually reducing the doses and frequencies of steroids, cycloplegics, and AGM. After a quiescence of the eye for

a period of three months with no further recurrences, visual rehabilitation by phacoemulsification and posterior chamber intraocular lens was carried out with synechiolysis, separation of iridocorneal adhesions and use of iris retractors (Figure 2).

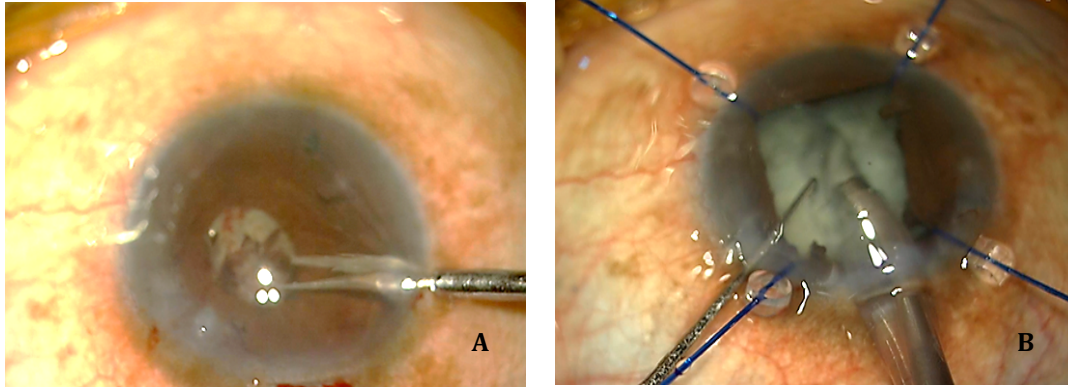


Figure 2: Intraoperative images.

Figure 2A: Viscoelastic was injected beneath the iris through the small nasal opening; pupillary membrane cut with a micro scissor and peeled off using micro forceps.

Figure 2B: Iris retractors applied to expand the pupil; soft mature cataract was successfully removed using low energy and low bottle height.

Post operatively, there was intense inflammation with the development of hypopyon and elevated IOP of 38 mmHg. This was controlled with tapering doses of oral prednisolone

acetate, acetazolamide tablets, topical anti-glaucoma medication, and cycloplegics (Figure 3).

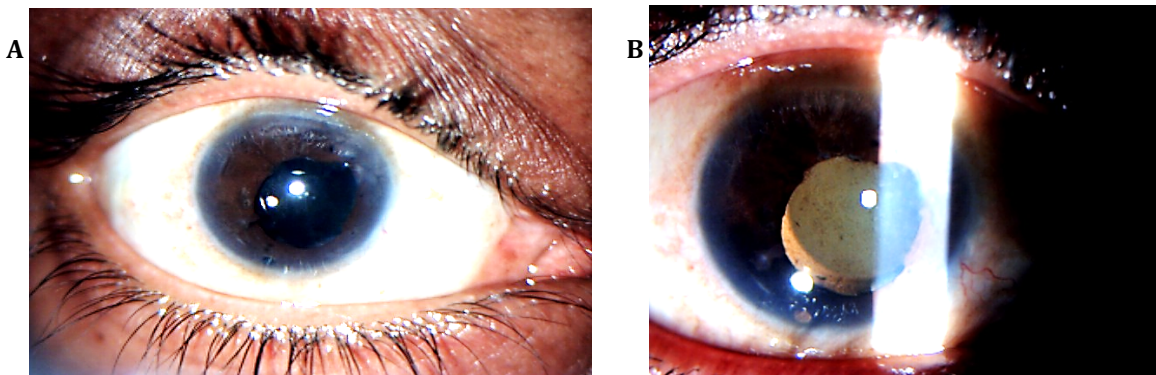


Figure 3A & 3B: Postoperative 6 week slit lamp image of dilated right eye showing patent PI, IOL in place and posterior capsular plaque.

The patient regained finger count vision at 4 metres, and fundus examination showed glaucomatous optic neuropathy with a CD ratio of 0.7:1 (Figure 4). OCT macula revealed cystoid macular edema for which he was treated with topical Difluprednate 0.05% and posterior sub-Tenon's injection of Triamcinolone acetonide (40mg/ml). Three months post-

surgery, the patient's right eye had a BCVA of 6/24 and an IOP of 12 mm Hg, while using a combination of 2% Dorzolamide and 0.5% Timolol drops, with resolving macular edema (Figure 5). The patient has been kept on three monthly follow up.

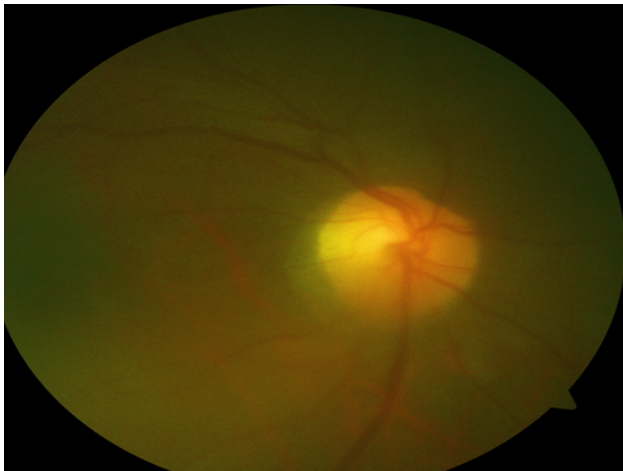


Figure 4: Fundus image of the right eye showing hazy view with CDR 0.7:1.

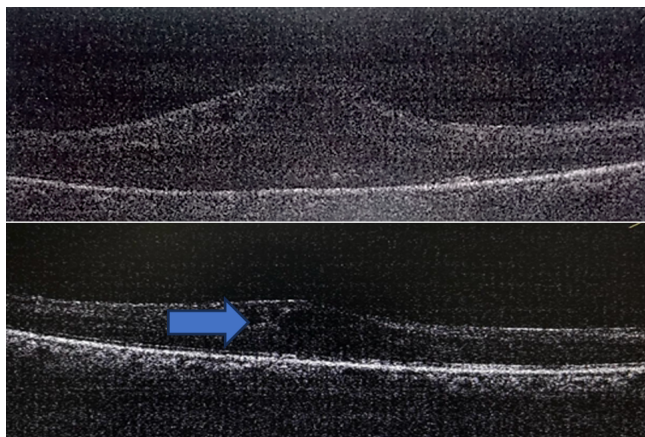


Figure 5: OCT macula showing resolving macular edema.

Discussion

Chronic uveitis encompasses a heterogeneous group of diseases, many of which are idiopathic in origin [3], with high incidence of complications, and has a variable long-term visual prognosis. This case highlights all the complications of the disease successfully managed with visual rehabilitation of reasonable vision of 6/24.

Anterior segment neovascularization is a rare but well-known complication of uveitis, may develop in response to either anterior or posterior segment inflammation or ischemia. Angiogenesis occurs when the balance between angiogenic and nonangiogenic factors favours the former [4].

The cause of neovascularisation of the iris and pupillary membrane in this case was possibly due to long-standing

subclinical inflammation due to chronic anterior uveitis, as it regressed on treatment with oral and topical steroids and post-operative fundus examination did not reveal any peripheral retinal detachment or proliferative vitreoretinopathy. Presence of neovascularization further complicated this challenging case.

Identifying new blood vessels in patients with uveitis requires a high degree of suspicion, careful examination, and sometimes can be missed. Neovascularization increases the risk of bleeding in uveitis and even spontaneous hyphema have been reported by Fong DS, et al. [5]. Neovascularization in this case was initially missed by the treating doctor, and the patient underwent laser iridotomy, which resulted in the flare-up of inflammation, obliteration of iridotomies, and formation of multiple iridocorneal adhesions.

There is always an element of inflammation, be it the primary feature or a secondary, in uveitic neovascularization. Therefore, local or systemic corticosteroids are almost always indicated for its treatment. There are reports in literature where corticosteroids have been shown to produce resolution of disk neovascularization in sarcoidosis, juvenile rheumatoid arthritis, toxocariasis, and HLA-B27-associated uveitis. They are shown to inhibit the growth of new blood vessels and cause existing ones to diminish [6]. Additionally, they may also reduce inflammation, which in turn decreases the stimulus for abnormal blood vessel growth [7,8]. The mainstay of treatment to control inflammation and its sequelae in our patient was steroids both topically and oral, taking care to prevent its systemic complications with oral Pantoprazole and calcium tablets.

In different studies, the incidence of secondary glaucoma caused by uveitis is reported to be 10–20% [9,10]. Many mechanisms are involved alone or in combination in the pathogenesis of uveitic glaucoma (UG). Our patient had raised IOP due to multiple factors: pupillary block due to posterior synechiae and occlusion pupillae in addition to peripheral anterior synechiae. Compared to general population uveitic glaucoma patients have lower rates of overall surgical success and high rates of postoperative complications like hypotony due to ciliary body dysfunction caused by recurrent intraocular inflammation [11,12].

Nd-YAG laser peripheral iridotomy (LPI) used to relieve pupillary block due to posterior synechiae and iris bombe formation, is not always successful in UG. Failure of LPI and development of overt neovascularization after LPI in this case signifies presence of subclinical inflammation which flared up after the procedure. The other dangers of laser procedures in such eyes are due to shallow anterior chambers, increasing the risk of endothelial damage during LPI [13,14]. Laser synechiolysis is well documented in the

literature as a method to break iridolenticular adhesion in patients with seclusion pupillae to bypass the pupil block of aqueous [15], and there are reports of using laser synechiolysis to perforate fibrinous membrane in cases of occlusion pupillae in uveitic glaucomas [16]. In our patient, as the central anterior chamber was not completely obliterated, a laser synechiolysis could be done which was successful in providing immediate relief by reducing IOP and pain.

Some complications which may occur due to this procedure, such as microhemorrhaging, pigment dispersal in the anterior chamber, and elevation of intraocular pressure, are typically self-limiting [15,17]. These complications did occur after laser and were managed with steroids, glaucoma medications and atropine.

Based on multiple studies, nearly 30% of patients with UG will require surgical treatment [18,19]. It is generally accepted that either trabeculectomy [18] or valve [19] implantation are safe and, most of the time, successful procedures in the treatment of UG. The non-penetrating glaucoma procedures such as deep sclerectomy (DS) and viscocanalostomy offer the advantage of minimal post-operative anterior chamber inflammation and a reduced risk of delayed complications in UG. These procedures have shown satisfactory long-term results [20].

Recently, minimally invasive glaucoma surgery (MIGS) techniques have been introduced for treating UG with varied success. These procedures are performed as primary treatments, in combination with cataract surgery, or after failed previous methods [21].

Uveitic cataracts comprise approximately 1.2% of all cataract surgeries and are more demanding surgically [22] with far less predictable postoperative outcomes. Recurrent anterior segment inflammation in this patient resulted in a small pupil, associated with posterior synechiae, pupillary membrane and neovascularization which compromised surgical visibility, making surgery more challenging. The use of dispersive viscoelastic, iris retractors and careful removal of the pupillary membrane improved the visual outcome.

Use of corticosteroids, availability of biocompatible intraocular lenses (IOLs), and improved surgical techniques have significantly reduced complications in uveitic cataract extraction in recent years [23,24]. In our case, we used single-piece hydrophobic acrylic IOL.

Controversy about combined surgeries for cataract and glaucoma always remains. Since our patient's IOP was controlled with three antiglaucoma medications after laser treatment, we decided to first remove the large cataractous lens using conjunctiva sparing technique and consider

a second procedure later if necessary. As expected in uveitic cases, our patient developed intense inflammation postoperatively but was managed successfully with systemic and topical steroids and achieved reasonable vision in the affected eye. His intraocular pressure remains controlled with a combination of Dorzolamide – timolol twice daily, thereby not requiring any further surgery.

Conclusion

Management of uveitic glaucoma with complicated cataract is extremely demanding with high risk of complications. Anterior segment neovascularization has to be tackled aggressively before planning cataract surgery. Laser synechiolysis to remove the fibrous tissue in pupillary area can help in AC reformation and in lowering the uncontrollable intraocular pressure. Combined surgery should be avoided in these cases as it increases the risk of complications. Use of steroids pre and post-operatively, meticulous planning and implementation can improve visual outcome in these cases.

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