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Case Report

Bilateral Aqueous Misdirection Syndrome After Years of Uneventful Phacoemulsification in Hyperopic Patient

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Abstract

Aqueous misdirection syndrome (AMS) is a rare but serious condition that can present after routine phacoemulsification surgery. This paper highlights a case of bilateral aqueous misdirection which developed after years of uneventful cataract surgeries in both eyes. It developed after 2.5 years in the left eye (LE) and after 5 years in the right eye (RE). The patient has hyperopic eyes with patent peripheral iridotomies performed prior to the cataract surgeries and managed later successfully with Nd: YAG hyaloidotomy and atropine eye drops in one eye while the other eye required glaucoma filtration surgery with vitrectomy to break the aqueous misdirection cycle. Patients with potential risk factors carry a lifelong chance of developing aqueous misdirection syndrome, therefore counseling the patients is crucial to keep them aware about the possible complications after any intraocular surgery and the treatment options to avoid the devastating outcomes.

Introduction

The aqueous misdirection syndrome (AMS), also referred to as malignant glaucoma, was first described in 1869 by Von Graefe as an elevated intraocular pressure (IOP) accompanied by shallowing of the central and peripheral anterior chambers (AC) in the absence of choroidal effusions or hemorrhage with a patent peripheral iridotomy (PI). It can occur immediately after intraocular surgery, including phacoemulsification, or it may take months or years. In hyperopic patients with a history of narrow or closed angle glaucoma, the risk is high. [1]

When the anterior chamber is shallow after uneventful phacoemulsification with intraocular lens (IOL) implantation, the differential diagnosis includes incision leakage, pupil block, capsular block syndrome, plateau, or pseudo plateau (ciliary body cysts), anteriorly subluxated IOL bags, supraciliary or choroidal effusions/hemorrhages, and malignant glaucoma.[2]

AMS's exact mechanism is still unclear, but it could be explained by ciliolenticular block, which could be caused by anterior movement of the lens-iris diaphragm, poor vitreous conductivity, and choroidal expansion. A subsequent closure of the angle resulted in elevated IOP without response to medical intervention or peripheral iridotomy (PI). [3]

Stepwise management of aqueous misdirection is advised and to begin medically with administration of cycloplegics and IOP suppressants. When eyes were refractory to pharmacotherapy, Neodymium-doped yttrium aluminum garnet (Nd: YAG) laser assisted PI followed by hyaloidotomy to create a channel for free flow of aqueous into the anterior chamber. Surgical options for AMS are mainly iridozonulohyaloidectomy (IZH) with vitrectomy. [4]

Case Report

A sixty-four-year-old woman, not known to have any comorbidities, was seen for the first time in our ophthalmology clinic in Al-Nahdha Tertiary Hospital in 2016. The patient was referred to our center with a complaint of gradual decreasing in vision in both eye (BE).

On examination, the patient's Best Corrected Visual Acuity (BCVA) was 6/12P in the Right eye (RE) and 6/12 in the left eye (LE). The refraction was identifying hyperopic eyes as follow: RE (+ 6.25 / -0.75 @175), LE (+6.5 / -0.5@45). The IOP was 24 mmHg and 17 mmHg in RE and LE respectively.

The anterior segment examination was normal in both eyes except of clinical evidence of shallow AC with patent PI (the PI was done long time back in different ophthalmic center). Left eye showed evidence of Pseudoexfoliation syndrome (PEX). BE showed cataract changes more in LE.

Dilated fundus examination was normal in both eyes except of early disc glaucomatous changes with cup-to-disc ratio (CDR) of 0.7, 0.5 in RE and LE respectively. Gonioscopy examination in BE showed Grade 2 angles that opens with indentation to Grade 3 according to Shaffer's system.

Base line investigations usually done in our center as routine assessment for new and suspected glaucoma patients, including Optical Coherence Tomography (OCT) of Retinal Nerve Fiber Layer (RNFL) and Macula and Visual Field (VF) test.

Our patient RNFL OCT showed, thinning in Superior quadrant with average thickness of 75 μm in RE and overall good RNFL in all quadrants with average thickness of 107 μm in LE.

OCT macula was normal with normal morphology and Central Macular Thickness (CMT) of 207 μm in RE and 180 μm in LE. While (VF) 24-2 revealed few but nonsignificant changes. In the right eye, Glaucoma Hemifield Test (GHT) was outside normal limits; Mean Deviation (MD): -5.53; pattern standard deviation (PSD): 2.16. comparing that to the left eye, it showed (GHT) of Borderline; (MD): -6.64; (PSD): 2.33.

Summing the previous mentioned findings, the patient was labeled as case of cataract changes in both eyes with clinical evidence of Primary Angle Closure Glaucoma (PACG) with previous patent PI in BE.

The patient was counseled regarding the ocular status and was given topical timolol to be used twice a day and was booked for cataract surgery to be done one month after the first visit. Right eye unevenful phacoemulsification with (IOL) implantation surgery was done as planned followed by the same surgery to the left eye a month later. The patient was kept on routine follow ups postoperatively and was happy with no obvious complaints.

Suddenly after around two and half years of left eye cataract surgery the patient presented to our emergency department with acute LE blurring of vision associated with severe headache. On examination, the RE was quiet but LE BCVA was 6/18P and IOP of 51 mmHg. Slit lamp examination showed shallow AC (figure 1-a, b) with Patent PI (figure 1-c). The diagnosis of pupillary block was excluded with the presence of patent PI and B-Scan showed no evidence of choroidal pathology. The diagnosis of aqueous misdirection was confirmed and the patient managed in the office immediately with intravenous acetazolamide 500 mg along with full topical IOP suppressants.

After 1 hour, IOP dropped to 31 mmHg and patient was discharged on topical glaucoma medications, topical atropine eye drops three times a day and acetazolamide tablets 250 mg three times a day.

On subsequent clinic visits, the IOP was around 21 mmHg, so augmentation of the presented PI was decided to be performed followed by (Nd: YAG) laser assisted anterior hyaloidotomy (figure 1-d). The outcome was significant with consistent IOP measures of around 16 mmHg; the patient was kept on topical IOP suppressants along with atropine eye drops and was maintaining good and controlled IOP on regular clinic visits.

After almost 5 years of the right eye uneventful cataract surgery, the patient presented to our department with the same complaint (figure 2-a). The BCVA in right eye was 6/18, the IOP was 30 mmHg and anterior segment examination showed shallow AC with patent PI. Medical treatment was initiated with IOP suppressants and atropine eyedrops along with Nd:YAG laser PI augmentation and anterior hyaloidotomy. Patient did not improve immediately, so surgical options were offered to avoid any potential long term devastating outcomes but the patient decided to continue on medical treatment only. Luckily, after close observations, the IOP in RE was controlled for a while and measured around 12mmHg on multiple visits.

The patient was not bothered by the atropine frequent use and her mid-dilated pupil did not cause any subjective visual disturbance.

Unfortunately, that did not last for so long. After one year of keeping RE on medical treatment only, the IOP started to be more in the higher side reaching almost 40 mmHg. Surgical intervention was again advised to the patient and finally agreed. Combined surgery of glaucoma filtering procedure by Ahmed Glaucoma Valve along with vitrectomy as anterior approach through the previous PI (figure 2-b, c).

On the latest clinic visit, both eyes BCVA was 6/18 and IOP was 15 mmHg and 14 mmHg in RE and LE respectively. The patient was seen multiple times in the clinic and was keeping good and controlled measures.

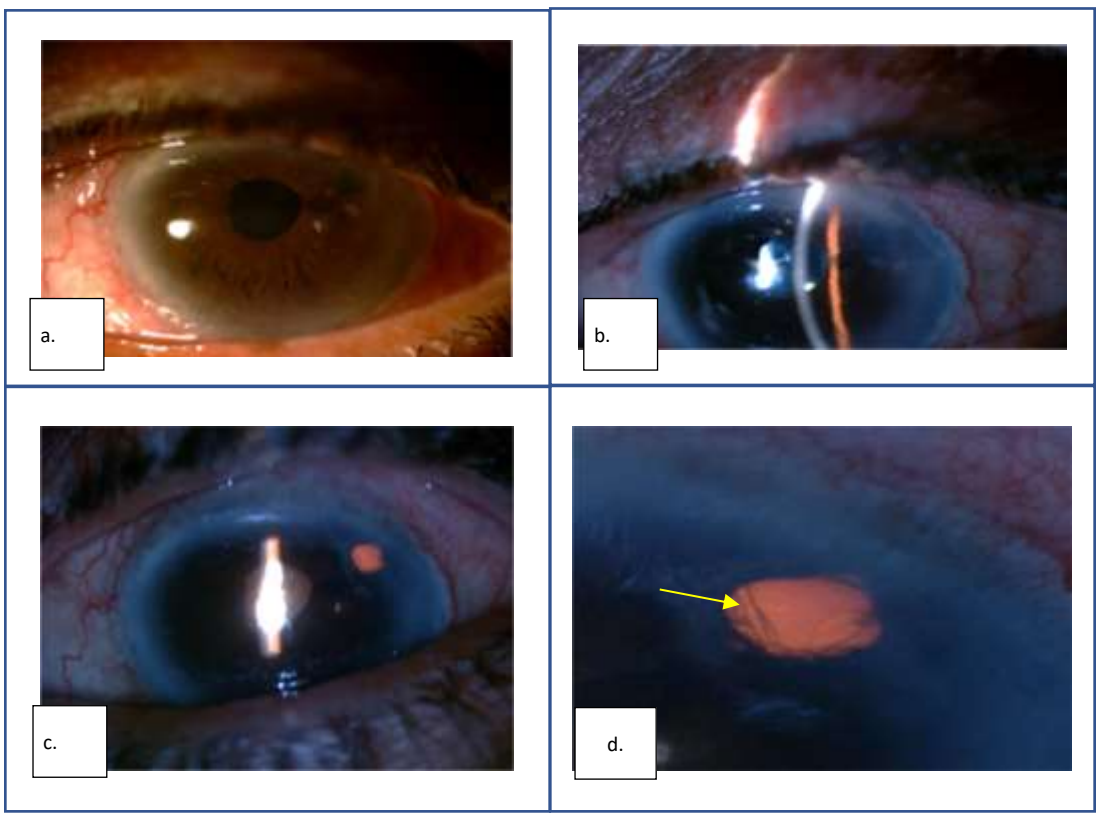

Figure 1: (a) and (b): Left eye with shallow anterior chamber. **(c)** Patent peripheral iridotomy with retro-illumination. **(d)** Left eye after Nd:YAG laser augmentation of peripheral iridotomy with anterior hyaloidotomy, yellow arrow pointing to the interface of the anterior hyaloid membrane after being disrupted by the laser.

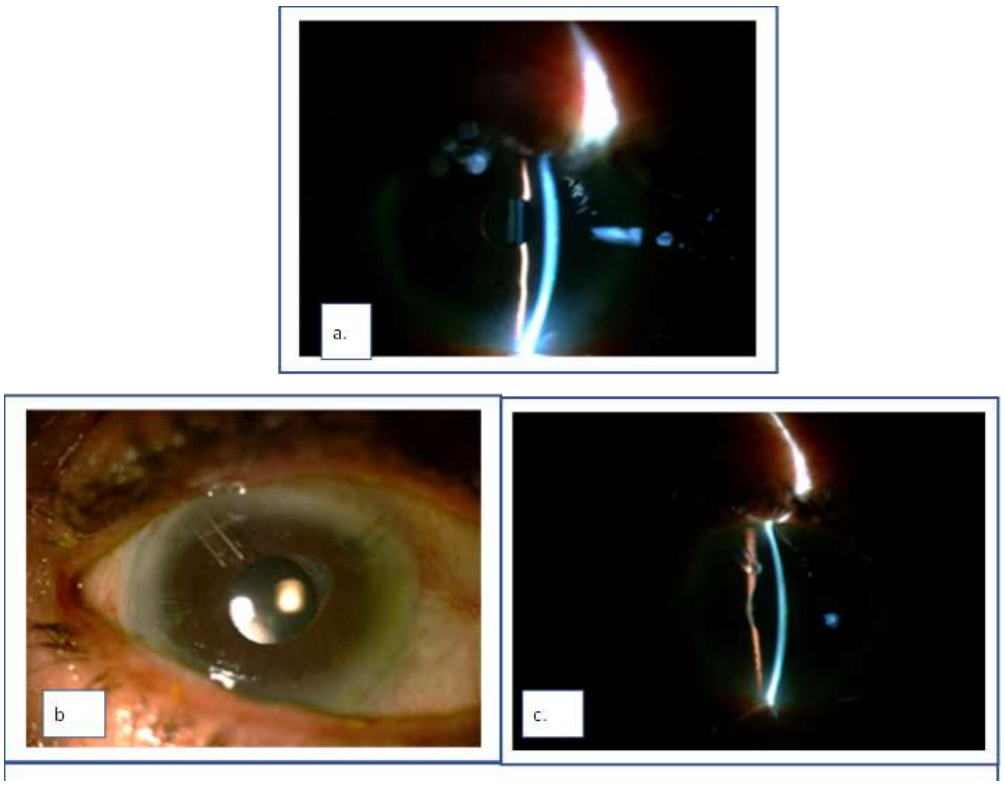

Figure 2: (a) Right eye shallow anterior chamber after having AMS. **(b) and (c):** Right eye after underwent surgical intervention, AGV**and vitrectomy.

*AMS: Aqueous misdirection syndrome.

**AGV: Ahmed Glaucoma Valve.

Discussion

Management for aqueous misdirection syndrome remains challenging. Here, we highlighted a case of bilateral aqueous misdirection after few years from uneventful cataract surgeries in both eyes. Researches suggest that handling difficult cases successfully is achieved mainly after disruption of the anterior hyaloid. In our case, one eye was treated successfully with Nd: YAG hyaloidotomy and atropine eye drops while the other eye required glaucoma filtration surgery with vitrectomy to break the aqueous misdirection cycle.

There is an anatomical predisposition for aqueous misdirection. Anatomically narrow filtering angles or chronic angle-closure glaucoma characterize the majority of patients. In other studies, plateau iris configuration and hyperopia have been defined as risk factors.

Furthermore, in ultrasonic biomicroscopy, anterior rotation of the ciliary body processes may be important for blocking aqueous fluid flow.[1] Aqueous misdirection syndrome is indeed more common in women than in men. The lens is positioned more forward in women than it is in men, resulting in not only a 4% shallower anterior chamber but also a narrower space between the lens equator and ciliary body. Therefore, women are more prone to developing a misdirection of the aqueous flow.[5] Chandler et al. suggested that forward movement of the lens is caused by the flexibility of the lens's zonules combined with vitreous tension.[6]

The occurrence of aqueous misdirection in one eye significantly increases the risk to the fellow eye.[7] This what happened with our patient with a duration of 2.5 years between one eye and the other of developing the aqueous misdirection attacks.

The pathophysiology that is primarily accountable is the establishment of a pressure gradient between the posterior vitreous cavity and cerebral spinal fluid at the retrobulbar space. Fluid moves in the optic nerve's subarachnoid space due to a pressure gradient and histologic alterations in the lamina cribrosa, contributing to elevated cerebral spinal fluid pressures.[8] Further fluid migration to the subarachnoid space of the contralateral eye is facilitated by the bidirectional flow of cerebral spinal fluid, causing displacement of the vitreous body anteriorly.[9,10] Although it is difficult to hypothesize the cause of the aqueous misdirection-like spontaneous presentation within two to five years postoperatively, it is suspected that, previous surgeries, anatomic predisposition, increased resistance at the trabecular meshwork, aging, and smaller eyes can be contributory.[11,12]

In term of management, patients who are refractory to medical therapy may require laser treatment with neodymium: yttrium–aluminum–garnet, laser capsulotomy, laser hyaloidotomy, or cyclodiode laser photocoagulation.[13,14] It is believed that large optic lens or synechiae formed between the intraocular lens (IOL) and capsule may also act as a barrier to anterior flow of aqueous. In these cases, laser capsulotomy creates a patent passageway for fluid flow from posterior to the AC.[14] Invasive surgical techniques can improve treatment if individuals are still unresponsive by permanently disrupting the anterior hyaloid face, which establishes up a passage for the aqueous flow. These procedures may include vitrectomy with iridozonulectomy, hyalozonulectomy, or a complete vitrectomy–iridectomy–zonulectomy.[15]

In our patient, right eye responds to vitrectomy + surgical drainage device while left eye responds to Nd: YAG laser hyaloidotomy. Numerous studies have indicated that effective management is ultimately achieved with a combination of numerous techniques; but there is still a lack of consensus about the most optimal therapeutic strategy.

In conclusion, the goal of our treatment is to stop the cycle of aqueous misdirection by establishing a passage between the posterior and ACs in eyes refractive to medical treatment. It is also important to note that the risk to the fellow eye is significantly elevated in aqueous misdirection, and thus careful monitoring with prophylactic treatment should be considered. Careful follow-up and continued monitoring of patients who are at high risk of aqueous misdirection ensures optimal care and better prognosis. Also, counseling the high risk patients to the risk of malignant glaucoma and its symptoms even years after any ocular surgeries.

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Declaration of patient consent:

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understand that their name and initials will not be published, and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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