Yemeni Journal of Medical and Health Research

Bilateral Simultaneous Pseudophakic Glaucoma in a Patient without Known Causative Risk Factors - A Case Report

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DOI: https://doi.org/10.47372/yjmhr.2025(14).2.1

Abstract

Malignant glaucoma is one of the most challenging ophthalmic conditions. The aim of this article is to report a case of bilateral simultaneous malignant glaucoma in a pseudophakic female patient with no history of any major risk factor responded well to medical treatment. A sixty-six years old female patient presented to our outpatient clinic having posterior subcapsular cataract in both eyes. Phacoemulsification was done for both eyes one week apart. Both surgeries were done uneventfully. On the first postoperative day of the second eye, the patient presented to our clinic with bilateral visual loss, high intraocular pressure and bilateral axial shallowing of anterior chambers. Slit-lamp fundus examination and ocular ultrasound were normal for both eyes. A diagnosis of bilateral spontaneous pseudophakic glaucoma was made and vigorous medical treatment was initiated. On the next day there was dramatic response to medical treatment.

Keywords: Malignant Glaucoma, Pseudophakic Glaucoma, shallow anterior chamber, aqueous misdirection and lens–iris diaphragm.

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*الزرق الخبيث ثنائي الجانب المتزامن في مريض بدون عوامل خطر مسببة معروفة ـ تقرير حالة

محمد علي العنسي

ملخص الدراسة

المقدمة: يعد الزرق الخبيث (الجلوكوما الخبيثة) أحد أكثر حالات العيون تحديًا، والهدف من هذه المقالة هو تسجيل حالة زرق خبيث ثنائي الجانب (بالعينين) ومتزامن لمريضة لديها عدسة بلورية كاذبة (بعد عملية استخراج المياه البيضاء وزرع عدسة اصطناعية في العينين).

المريضة ليس لديها قصة مرضية لأي عامل خطورة مهم للزرق.

مريضة تبلغ من العمر 66 عامًا حضرت إلى العيادة الخارجية لدينا وهي تعاني من إعتام تحت محفظي خلفي لعدسة العين اليمنى واليسرى (مياه بيضاء ثنائية الجانب).

تم إجراء عملية استخراج المياه البيضاء بالفاكو وزرع عدسة اصطناعية (عدسة بلورية كاذبة) لكلا العينين بفاصل أسبوع واحد بين العمليتين. وقد تم إجراء العمليتين بدون أي مضاعفات.

في اليوم الأول بعد إجراء العملية الجراحية في العين الثانية، وأثناء إجراء الفحص الروتيني في العيادة للمتابعة بعد العملية، تبين أن المريضة تعاني من فقدان النظر، ارتفاع ضغط العينين مع ضحالة الحجرة الأمامية المحوري للعينين.

فحص قاع العين بالمصباح الشقي ضمن الحدود الطبيعية في العينين، تصوير الشبكية والسائل الزجاجي بالموجات فوق الصوتية للعينين ضمن الحدود الطبيعية.

تم تشخيص الحالة على انها زرق بلورة كاذبة تلقائي ثنائي الجانب (الزرق الخبيث) نتيجة التوجه الخاطئ للخلط المائي.

تم البدء بالعلاج الدوائي فوراً وبشكل مكثف وفي اليوم التالي لوحظ استجابة فعالة للعلاج الطبي.

الاستنتاج: يمكن أن يحدث الزرق الخبيث بعد إجراء جراحة استخراج المياه البيضاء في غياب عوامل الخطر الرئيسية

يعد التعرف المبكر على الزرق الخبيث وعلاجه أمراً ضرورياً لتقليل خطر فقدان شديد ودائم للبصر.

الكلمات المفتاحية: الزرق الخبيث – زرق البلورة الكاذبة (زرق العدسة الكاذبة) – حجرة أمامية ضحلة – التوجه الخاطئ للخلط المائي.

قسم طب العيون – كلية الطب والعلوم الصحية – جامعة ذمار - اليمن

Introduction

Malignant glaucoma (MG) was described first by Von Graefe in 1869 [1] as a rare post operative complication following trabeculectomy surgery in glaucoma patients. The condition was characterized by acute elevated intraocular pressure (IOP), a shallow or flat anterior chamber (AC), in the presence of a patent iridectomy. He used the term "malignant" glaucoma because it did not respond to conventional therapy and its tendency to progress causing severe and permanent vision loss. Other terms have been used to describe this condition as ciliary block glaucoma [2] and aqueous misdirection [3].

MG has been reported following several ocular surgeries such as glaucoma and cataract surgeries with anterior or posterior chamber intraocular lens (IOL) implantation [4,5]. MG has also been reported to occur following laser procedures such Neodymium-dopedyttrium aluminum garnet (Nd:YAG) posterior capsulotomy [6]. It has been also reported following use of some drugs such as topical miotics [7]. Pseudophakic MG refers to the development of glaucoma following cataract surgery with Implantation of intraocular lens (IOL). It may present immediately after cataract surgery within few hours or present few weeks to years later. Early recognition and treatment of MG is essential to reduce the risk of severe and permanent vision loss. The exact pathogenesis and different mechanisms concerned with the development of MG is not fully understood and remain uncertain up to date and are most likely to be multifactorial. An alteration in the anatomic relationship of the lens, ciliary body, and anterior hyaloid face resulting in a final pathway of aqueous posterior misdirection and forward movement of the iris–lens diaphragm was suggested in MG pathogenesis. Shaffer proposed posterior aqueous flow and accumulation either into or behind the vitreous cause forward displacement of the iris–lens diaphragm and anterior rotation of the ciliary body [3]. Chandler and Grant suggested that loose zonules along with pressure from the vitreous allow partial subluxation of the lens–iris diaphragm [8]. Positive pressure phenomenon secondary to choroidal expansion along with poor vitreous fluid conductivity is another mechanism proposed by Quigley and colleagues [9]. Medical management is effective in 50% of cases of MG within five days, after which laser intervention should strongly be considered [10]. If medical management fails within 5 days, or if lens-cornea touch occurs, Nd:YAG laser therapy may be attempted to disrupt the posterior capsule and anterior hyaloid face creating a communication between the posterior and the anterior segment of the eye. This procedure should be used when treating aphakic and pseudophakic MG, but not for phakic MG [11].

The aim of this article is to report a case of bilateral simultaneous MG in a pseudophakic female patient with

no history of any major risk factor responded well to medical treatment.

Case presentation

A 66 years old female patient presented to the hospital having posterior subcapsular cataract in both eyes. A written informed consent to publish this case report has been obtained from the patient. This report does not contain any personal identifying information.

The patient is diabetic for 13 years and fundus examination was normal for both eyes. The anterior chamber depth (ACD) and axial length (AXL) for the right eye (RE) were 3.32 mm and 23.06 mm respectively. The ACD and AXL for the left eye (LE)were 3.42 mm and 23.13 mm respectively. The intraocular pressure (IOP) was normal for both eyes, 18 mmHg for RE and 17mmHg for LE.
Phacoemulsification was done first for her LE. The surgery was done uneventfully. No complications had been detected during the early post operative follow up period. A week later phacoemulsification was done for the RE uneventfully as well.

Six hours postoperative to the second eye, the right eye, ophthalmic examination showed improved visual acuity in both eyes (0.66 for RE and 1.00 for LE), normal AC depth and normal IOP for both eyes. On the first postoperative day of the second eye (RE), the patient came to our hospital complaining of ocular discomfort and bilateral vision loss. Ophthalmic examination revealed bilateral conjunctival congestion, bilateral corneal haziness and bilateral diffuse shallowing of anterior chambers (axially and peripherally). Uncorrected visual acuity (UCVA) was 0.25 for the RE and 0.16 for the LE). There was bilateral myopic shift (i.e., from − 0.25 Diopters after the cataract surgery, to − 3.00 Diopters in the LE). IOP was high in both eyes (28 mmHg for the RE and 39 mmHg for the LE). No posterior segment pathology was detected by biomicroscopic fundus examination and ocular ultrasound in both eyes. NY: YAG laser peripheral iridotomy (PI) was done for the left eye to treat or to exclude any pupillary block component. No significant reduction in the IOP or deepening of the AC has occurred, so no further PI was done for the other eye. A diagnosis of bilateral simultaneous pseudophakic MG was made and vigorous medical treatment was initiated as; frequent topical steroid (prednisolone acetate 1%), atropine 1% three times a day, topical dorzolamide- timolol combination b.i.d, topical brimonidine 0.15% b.i.d, intravenous mannitol 20% 1g/kg and oral acetazolamide 250 mg t.i.d. 24 hours after medical therapy, there was dramatic improvement in both eyes. UCVA was 0.33 in both eyes, AC started to deepen and corneal haze resolved in both eyes. Systemic acetazolamide and mannitol were stopped two days later.

A week later, ophthalmic examination showed deep AC in both eyes with normal IOP and improvement in UCVA (0.5 for both eyes). Unfortunately, posterior synechiae started to form in both eyes inferiorly. Atropine and topical brimonidine were stopped and topical steroid tapering started. The patient kept on

topical dorzolamide- timolol combination twice a day.

A month later, the IOP and ACD were normal in both eyes. The ACD was 4.38 mm in RE and 3.5 mm in LE as shown by the Pentacam imaging, fig (1,2)). UCVA was 0.66 for the RE and 0.5 for the LE corrected to 0.66 by -1.25D lens. Posterior synechiae persisted in both eyes with IOL capture in the RE, Fig (3,4). The topical dorzolamide-timolol combination was stopped and no further medications were prescribed.

Three months later the ACD and IOP remained within normal ranges. The UCVA and the best corrected visual acuity (BCVA) remained unchanged and the patient was very satisfied. Regular follow up visits were instructed and alarm signs of recurrency were explained to the patient.

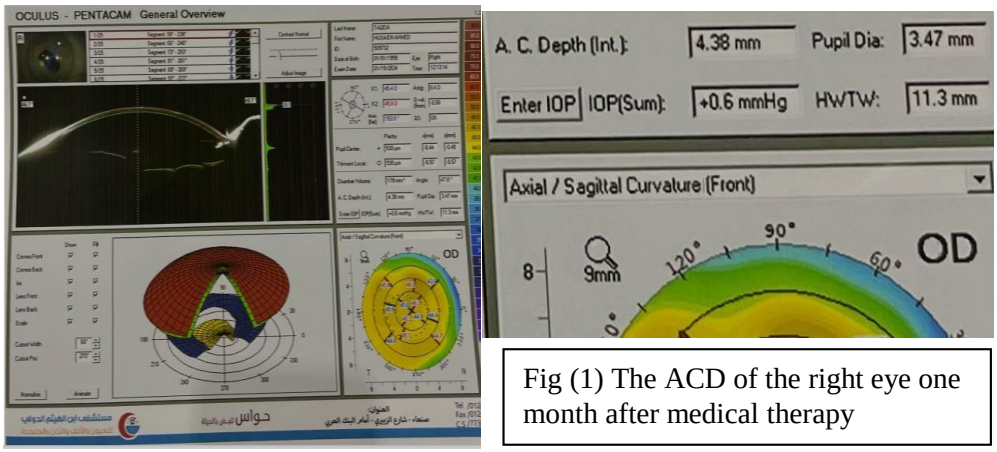

Fig (1) The ACD of the right eye one month after medical therapy

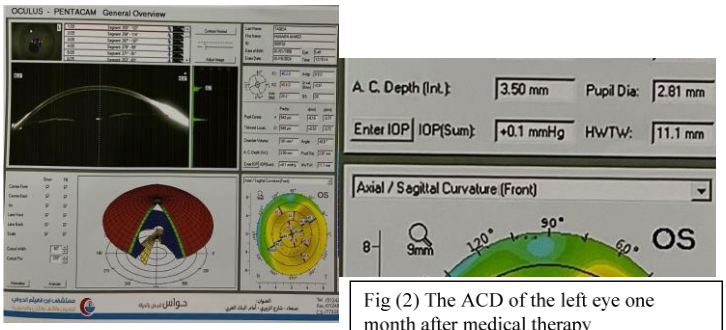

Fig (2) The ACD of the left eye one month after medical therapy

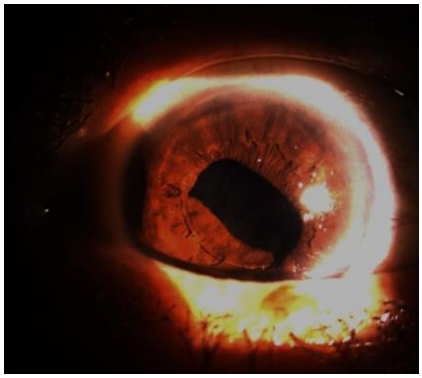

Fig (3) posterior synechia and IOL capture in the right eye one month after medical therapy

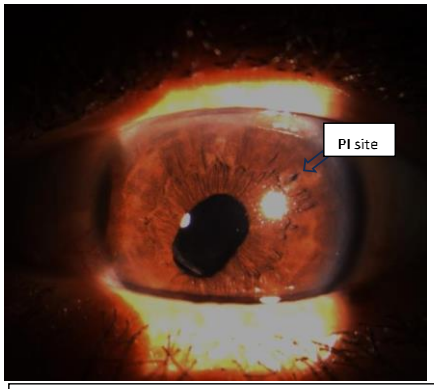

Fig (4) posterior synechia left eye one month after medical therapy. The pupil is semidilated

Discussion

MG is one of the most complex types of secondary glaucoma to manage and it can progress to permanent blindness without proper management. It is a rare post operative complication of many ocular surgeries and can occurs spontaneously as mentioned above. Early diagnosis and treatment are essential to prevent permanent vision loss. The diagnosis of MG is a clinical diagnosis of exclusion, and many symptoms and signs are nonspecific [12].

The main characteristic features of MG are; increased IOP, shallow or flat AC, normal posterior segment, no response to miotics or peripheral iridectomy, and good response to cycloplegics [13]. Along with postoperative myopic shift, all criteria of MG diagnosis were full filled in the present reported case. An angle closure secondary to pupillary block was excluded by creating a peripheral iridotomy. Many risk factors for the development of MG were reported such as the presence of MG in the fellow eye, uncontrolled angle closure glaucoma, preoperative shallow AC, hyperopia and short axial length, pseudoexfoliation and partial or total AC shallowing during surgery [4,13]. In contrast to these studies, there was no apparent risk factor detected in the present case. Similar to the reported case, Schwarts and Anderson found that MG can rarely develop in eyes with no antecedent risk factors [14]. This finding may indicate that there are still unknown mechanisms and or other risk factors involved in the etiopathogenesis of pseudophakic

MG and more studies are required to define the exact etiology, risk factors and mechanisms involved. Pseudophakic MG is an infrequent complication which can occur after cataract surgery and IOL implantation within various latencies. Anterior subluxation of the IOL due to weak zonules specially in presence of pseudoexfoliation may explains the possible cause of pseudophakic MG [15]. In addition, an intact anterior hyaloid face plays an important role in the etiopathogenesis of pseudophakic MG [16]. Contrary to these proposed mechanisms, both IOL were implanted in the bag without any subluxation, there was no zonular weakness or dehiscence and there was no pseudoexfoliation was detected in the present case. Similar to the present reported case, MG has been reported in an eye with no known causative risk factors [14].

MG may occur unilaterally or bilaterally. Similar to this article bilateral simultaneous MG have been reported in few studies [17-20] but in contrary to this article all patients in these articles developed MG spontaneously with no antecedent eye surgery.

Medical therapy is the first line of MG management achieving resolution within 5 days in 50% of cases. Medical management involves combined use of cycloplegics-mydriatics, aqueous suppressants and hyperosmotic agents. A combination of topical atropine, topical or oral carbonic anhydrase inhibitors CAIs, topical alpha-2 agonists and beta-blockers or intravenous mannitol can potentially break an attack of MG effectively. Anti-inflammatory agents are used to decrease post-operative inflammation [21].

These medications function together to reverse the clinical manifestations of MG. Cycloplegics relax the ciliary body muscle causing tightening of zonules thus resulting in backward movement of the iris-lens diaphragm. Hyperosmotic agents dehydrate and shrinks the vitreous allowing posterior movement of the iris-lens diaphragm and deepening of the AC. Aqueous suppressants decrease the flow of aqueous that can precipitate aqueous misdirection [22].

Once the condition resolves, medications can be withdrawn gradually. Osmotics and CAIs are typically withdrawn first, followed by alpha-2 agonists. Cycloplegics are usually withdrawn last but may be required long term [12]. In this case, the recommended regimen mentioned above was followed. Once the condition resolved, IOP normalized and AC formed, gradual drugs withdrawal was started including cycloplegics as the pupils didn't resume their normal size and remained mid-dilated due to posterior synechiae formation. Long term follow up is required and surgical intervention may be required latter on according to recent study [23].

Patient satisfaction and good final BCVA achieved as well as the fear of MG recurrency led to avoidance of any further surgical intervention to release posterior synechia formed or to reposit the captured IOL. The patient was advised for regular follow up visits and informed about all alarm symptoms of possible MG recurrency.

Conclusion

MG can occur after cataract surgery event in the absence of major risk factors. Early recognition and treatment of MG is essential to reduce the risk of severe and permanent vision loss.

Funding
This research received no specific grant from any funding agency in the public, commercial, or not-for-profit sectors.

Conflict of interest
The author has no any conflict of interest.

Patient consent
A written informed consent to publish this case report has been obtained from the patient. This report does not contain any personal identifying information.

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