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Case Report

Presumed Malignant Glaucoma in an Eye with Idiopathic Anterior Uveitis, with no Prior Laser or Surgical Intervention

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A B S T R A C T

Background: Malignant glaucoma presents a clinical challenge to diagnose and manage, usually develops after laser or surgical intervention, very few cases reported of developing malignant glaucoma in a virgin eye, but diagnosis and management initiated after laser iridotomy or cataract extraction.

Case presentation: A 36 years old healthy female, presented with a red eye. The patient was diagnosed with non-granulomatous anterior uveitis in right eye (OD) and managed accordingly. Microcornea and shallow anterior chamber (AC) OD was noted, and the left eye (OS) was unremarkable. One week later on follow up her intraocular pressure (IOP) was 28 mmHg OD and was suspected to be steroid-induced. She was managed with IOP lowering medications. She was referred to glaucoma team; diagnosed as uveitic glaucoma with a narrow angle, OD IOP stabilized to normal range with topical glaucoma medications. Few months later, she had IOP of 38 mmHg on maximal tolerable topical glaucoma medication and was off steroid. AC was shallower and her angle was closed, Ultrasound Biomicroscope (UBM) indicated anterior rotation of the ciliary body (CB) OD. She was diagnosed to have malignant glaucoma. Topical phenylephrine and atropine were added to the existing IOP lowering regimen, resulting in IOP stabilization. As the AC deepened, IOP lowering medication were tapered, during the course of management she underwent laser peripheral iridotomy (LPI). Five years later, her IOP remains normal and the patient is on low dose of topical steroid and phenylephrine only.

Conclusions: ciliary body rotation should be suspected in any case of shallowing of AC, with or without increase of IOP, even without a history of surgical or laser intervention. UBM should be performed as early as possible and topical mydriatics initiated immediately, to insure successful outcome with minimal intervention.

Keywords: Ciliary body rotation; Malignant glaucoma; Ultrasound Biomicroscopy; UBM; Uveitis; Shallow anterior chamber; Intraocular pressure

List of abbreviations: OD: Right eye; AC: Anterior chamber; OS: Left eye; IOP: Intraocular pressure; UBM: Ultrasound Biomicroscopy; CB: Ciliary body; LPI: Laser peripheral iridotomy; KKESH: King Khaled eye specialist hospital

1. Introduction

Malignant glaucoma, first reported be by Albrecht von Graefe in 1869, presents a clinical challenge to diagnose and manage. The etiology of malignant glaucoma is not well understood, it is clinically characterized by normal or elevated IOP and shallowing of the central and peripheral anterior chamber due to anterior displacement of the lens-iris diaphragm. Ultrasound Biomicroscopy demonstrates anterior rotation of the ciliary body, causing iridocorneal touch and appositional angle closure in these patients. Malignant glaucoma also known as ciliary block glaucoma or aqueous misdirection glaucoma[1]. There are numerous hypotheses about the mechanism that underlies the pathology. Hoskins and Shaffer postulated that misdirection of aqueous humor flow causes accumulation of aqueous fluid behind a posterior vitreous detachment, combined with an anterior shift of the iris-lens diaphragm. They suggested a valve-like mechanism that "misdirected" the aqueous humor posteriorly[2]. Another explanation is an anterior rotation of the ciliary body processes, creating touch between the ciliary bodies and the lens, which may cause ciliary block[7]. Forward displacement of the lens blocks the communication between the posterior and anterior chambers, as well as the outlets from the eye, is the essential anatomical feature of malignant glaucoma[8]. An additional mechanism that has been suggested is the congestion of the uveal tract, which pushes the lens into an anterior position and holds it there[8], abnormal choroidal circulation may also lead to the accumulation of blood and swelling of the ciliary processes, which will obscure the flow of the aqueous fluid. Additionally, in certain cases, the lens capsule and zonules may constitute a region of resistance to flow of aqueous humor forward[9,10].

Malignant glaucoma occurs in 2% to 4% of patients with a history of either acute or chronic angle closure glaucoma who have undergone surgery[3]. In a study of 1689 patients who underwent varying ophthalmic surgeries, including glaucoma surgery, cataract surgery, or combined surgery, malignant glaucoma occurred in 1.3% of eyes postoperatively. After penetrating surgeries this complication occurred in 2.3% of the eyes. Malignant glaucoma has also been reported after LPI[4], posterior capsulotomy with Nd-YAG laser (Neodymium-yttrium-aluminum-garnet laser)[6], cyclophotocoagulation[7], and in eyes that did not undergo any previous surgical procedures[8].

Our case was managed in compliance to practices and protocols approved by the hospital medical and scientific committee and the study was reported after KKESH ethics and review committee approval.

Case Report

A 36 years old healthy female presented with best corrected visual acuity of 20/40 in the right eye (OD) with -14 Diopter (D) sphere, -3 (D) cylinder, axis 30°, and 20/100 in the left eye (OS) with +2.5 (D) sphere, -2 (D) cylinder, axis 5°.

Patient had asymmetrical measurement between both eyes, her right eye had a Microcornea, shallow AC and normal fundus. The cornea was normal size OS, AC was normal and she had an old macular scar, which was the reason behind her OS low vision, with OS amblyopia and anisometropia

Axial length was 25.89 mm OD and 21.78 mm OS, AC depth was 2.05 mm OD and 3.18 mm OS. Lens thickness was 5.7 mm OD and 3.77 mm OS.

The patient presented to King Khalid Eye Specialist Hospital, Emergency department in April 2012 complaining of ocular irritation. On examination, intraocular pressure (IOP) OU was within normal limits. The right eye had +1 reaction in AC. She was managed accordingly and started on prednisolone acetate ophthalmic suspension 1% (Pred Forte, Allergan) OD. A week later during follow up in the clinic, her IOP was 28 mmHg OD and 18 mmHg OS, measured by Goldmann applanation tonometry. The patient was diagnosed with steroid-induced IOP elevation. Her steroid was tapered, and the patient was prescribed a fixed combination of topical 2% dorzolamide/0.5% timolol (Xolamol, Jamjoom Pharma) and her IOP decreased to 18 mmHg OD. The patient was then referred to a glaucoma specialist, seen on May 20, 2012. The specialist noted a shallow AC, IOP of 23 mmHg OD and 18 mmHg OS, she was diagnosed to with uveitic glaucoma, with occludable angles with IOP above target, brimonidine tartrate 0.15% ophthalmic solution (*A*lphagan P, Allergan) was added.

On June 2012 uveitis was quiescent and the IOP was 18 mmHg on topical Xolamol, Alphagan and 1% Rimexolone ophthalmic suspension (Vexol) OD. Her uveitis kept on flaring and was managed with Pred Forte, and IOP was not elevated, she was still on Xolamol and Alphagan, with IOP at 15 mmHg OD and 18 mmHg OS.

On September 2012 the patient presented complaining of blurring of vision OD, IOP was 38 mmHg, she was on Xolamol and Alphagan, mild flare was noted in the AC. The patient was prescribed oral Acetazolamide (Diamox) and glycerol to reduce the IOP and she was referred back to glaucoma team. On examination the patient had severe AC shallowing with anterior displacement of the lens - iris diaphragm, AC depth was 0.83 mm (prior to this incidence it was 2.05 mm) and UBM showed ciliary body rotation (**Figures 1,2,3 & 4**). Patient was put on maximum topical glaucoma medication, atropine 1% ophthalmic suspension and phenylephrine hydrochloride 2.5% minims. Diamox was halted as the patient did not tolerate it well. A few days later her IOP was 24 mmHg OD, AC started to deepen, and AC angle was open OD. Over the next follow up visits, her IOP normalized to mid to lower teens. Laser peripheral iridotomy was performed under topical anesthesia using the Ellex Tango laser (Ellex Medical Laser Pt., Adelaide, SA, Australia) YAG mode, pilocarpine was not instilled prior to the procedure as a precaution (our protocol of pre-laser preparation for LPI does recommend pilocarpine, however it is a well-known factor in causing ciliary body rotation).

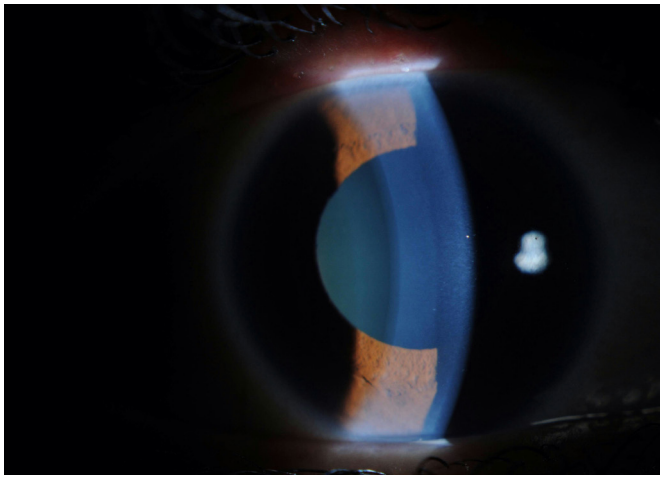

Figure 1: Shallow AC OD.

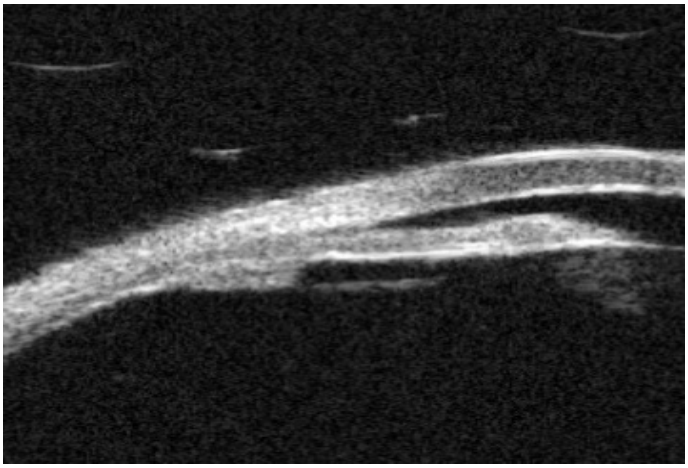

Figure 2: OD UBM shows ciliary body rotation.

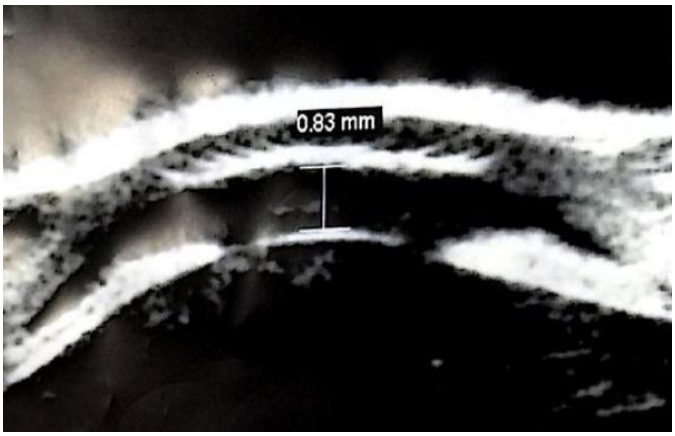

Figure 3: OD UMB shows shallow AC.

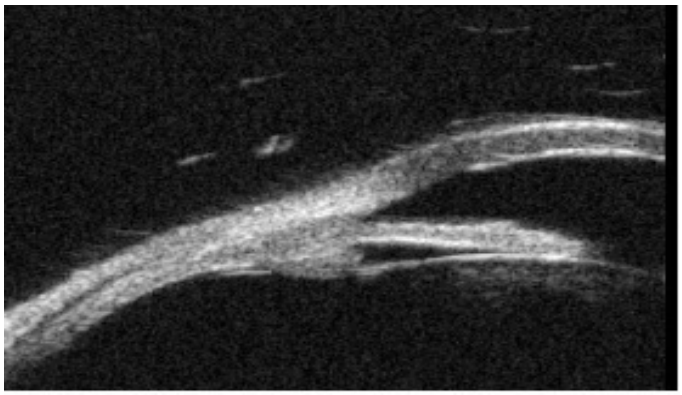

Figure 4. OS UBM.

A few months later the patient stopped atropine and phenylephrine, as it was interfering with accommodation. The patient was a school teacher and her right eye was the dominant eye, as OS had a macular scar, she had difficulty reading paperwork. On follow up her AC started to became shallow (confirmed by UBM), without an IOP elevation. She was prescribed a nightly dose of phenylephrine and all her glaucoma medications were stopped. At last, follow up on August 2018, her LPI was patent and IOP was in the lower teens.

Discussion

Malignant glaucoma is a rare form of glaucoma, which typically follows intraocular laser or surgical intervention. The etiology of the mechanism is not completely understood, which creates some difficulties in the standardization of the nomenclature. Certain authors suggest that the malignant glaucoma group should exclude cases of pupillary block or choroidal detachment[3].

Uveitis, on the other hand, is considered one of the non-surgical predisposing factors for glaucoma, which could be due to angle closure and pupillary block or may be open angle glaucoma as in our case. Our patient was diagnosed with open angle glaucoma secondary to steroid use, on her first episode of IOP elevation[26].

It is estimated that around 20% of uveitis patients will eventually develop what is known as uveitic glaucoma[12]. Less commonly, malignant glaucoma might couple with uveitis results in angle closure due to the rotation of the ciliary body[13].

It is thought that malignant glaucoma could be related to unique ocular anatomy. Lynch et al. suggested that it occurs more frequently in small eyes with an anatomically narrow iridocorneal angle[4], similar to our patient's right eye. Our patient had a microcornea and shallow anterior chamber OD compared to the normal left eye. This shallow anterior segment predisposes to angle closure glaucoma[5]. However, this is not the only mechanism in this pathology. Peripheral iridotomy, which eliminates pupillary block, does not prevent fluid accumulation in the narrow anterior segment, which may lead to further closure of the angle. If the aqueous humour is misdirected toward the vitreous cavity instead of the posterior chamber, symptoms of malignant glaucoma will manifest.

The differential diagnosis in our case of uveitic glaucoma was narrow angle verses steroid responder. Once the anterior chamber became shallower centrally and periphery and other causes of acute angle closure were excluded when UBM showed typical picture of ciliary body rotation, malignant glaucoma were considered, as with the case previously reported by Huang[27]. The patient was diagnosed and managed as angle closure uveitis glaucoma, and once postoperative IOP spiked to 52 mmHg despite a patent peripheral iridotomy and progressive anterior chamber shallowing, ciliary body rotation was suspected and patient was diagnosed with malignant glaucoma.

Furthermore, swelling of the ciliary processes due to inflammation, or miotics can cause critical narrowing of an already anatomically narrow space between the lens equator and the ciliary body, and eventually block forward aqueous flow[11]. During our management course we avoided the use of pilocarpine to avoid worsening of the condition.

Other possible differential diagnosis, is idiosyncratic uveal effusion syndrome as a reaction to Diamox use, ultrasound showed no choroidal effusion.

Once diagnosed, the treatment options of malignant glaucoma are categorized into two wide categories, conservative medical therapy, and surgical management. Firstly, the aim of the conservative medical therapy is to decrease the production of the aqueous humour, shrink the vitreous, and simultaneously lower the resistance in the path of the aqueous flow to the anterior chamber[14]. Medical therapy constitutes of Mydriatics - cycloplegics: which causes paralysis to the ciliary muscle, tightening of the zonule apparatus, and moving the lens iris diaphragm backward. Osmotically active agents: causes increase in the blood osmolality, which will lead to the movement of the aqueous fluid toward the hyperosmotic plasma. This will result in dehydration and shrinkage of the vitreous body and make it possible to retract the iris-lens diaphragm and deepen the AC. Aqueous production suppressants such as beta blockers and carbonic anhydrase inhibitors, to reduce aqueous inflow, which

will eventually decrease the volume of the fluid directed toward the vitreous. Corticosteroids: to stop inflammatory process and reduce ciliary body edema, additionally, it minimizes the inflammatory adhesions of the lens and the vitreous body to the ciliary body[15].

Previous studies of malignant glaucoma have reported that approximately 50% of patients respond to conservative medical therapy[5]. A study by Debrouwere et al., reported a 100% recurrence rate of malignant glaucoma in patients who underwent only conservative management despite the good initial response to therapy[16]. Our patient had a recurrence of AC shallowing and ciliary body rotation on UBM when she stopped her mydriatics. However, the cycle was stopped before she developed high IOP and conservative treatment was successful in breaking the cycle. In our case, these results have been sustained over 6 years until the time of writing. We have elected to prescribe a daily single maintenance dose of mydriatics to ensure that ciliary body rotation does not recur.

If conservative treatment fails, surgical management can be initiated, including laser or surgical intervention. Laser is usually used as adjunctive management option along with conservative medical therapy. This usually involves a combination of laser iridotomy with anterior hyaloidotomy and posterior capsulotomy. Using this maneuver improves the outcomes by creating direct communication between the vitreous, the posterior chamber, and the AC. This procedure can restore the normal dynamics of aqueous humour flow in patients with malignant glaucoma[17]. Our patient was phakic and the right eye is the good dominant eye, hence, this mode of treatment was not recommended and only laser peripheral iridotomy was performed during the course of medical management, especially as she was responding well to conservative treatment. Based on these observations we believed there was no indication for more extensive surgical intervention. A similar case to ours was reported in 1975, where malignant glaucoma developed in eye with no previous surgeries, LPI and maximum indicated medications failed to control IOP and the patient required cataract extraction with anterior vitrectomy to control IOP[8].

Surgical intervention is indicated when both the laser and the medical conservative therapies lack effectiveness. The main goals of surgical intervention in malignant glaucoma are to reducing the IOP and restore the correct anatomical relationship between the vitreous body, lens, and ciliary body. Additionally, surgical intervention aims to correct flow of the aqueous humour from the posterior segment toward the AC[18]. Achievement of correct flow and equalization of the pressure between the posterior segment and the anterior segment is an indicator of the effectiveness of the surgery [14].

In conclusion, ciliary body rotation and malignant glaucoma should be suspected in any case of shallowing of AC[19-25], with or without increase of IOP, even without a history of surgical or laser intervention. UBM should be performed as early as possible and topical mydriatics initiated immediately even with a shallow AC and closed angle, laser and surgery are warranted in cases that do not respond.

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