Malignant Glaucoma: Report of a Presumed Bilateral Spontaneous Case.

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Background: ***Malignant glaucoma is an ocular emergency and may lead to blindness if not promptly recognized and treated effectively. It is usually reported after trabeculectomy, and spontaneous or bilateral cases are not common in the literature.***
Case report: ***This article reports the case of a Nigerian woman who developed spontaneous bilateral malignant glaucoma with resultant visual impairment. A literature review on the subject is also undertaken. Possible etiology and management challenges of the case are also discussed.***
Conclusion: ***This study is aimed at drawing attention to this potentially blinding eye disease, and ultimately improves knowledge on its diagnosis and management, to reduce the visual morbidity.***

Key words: Malignant glaucoma, closed angle glaucoma, blindness, visual impairment, aqueous misdirection, vitrectomy

Introduction

The term malignant glaucoma was first used by Von Graefe[1] in 1869, to describe an ocular condition characterized by elevated intraocular pressure (IOP), a shallow or flat AC, in the presence of a patent iridectomy. This was in post trabeculectomy patients. It is also been called aqueous misdirection, ciliary block, cilio-vitreal block, and cilio-lenticular block glaucoma, in recognition of the possible aetiopathogenesis of this disease entity. However, this definition has evolved over the years, and malignant glaucoma now includes a range of secondary angle closure glaucomas, unresponsive to conventional anti-glaucoma therapies[2]. The term "malignant" however, does not connote a neoplastic process. The most common scenario for malignant glaucoma is post trabeculectomy[3-5], however, association with other clinical conditions have been reported. These include; cataract surgery with or without insertion of intraocular lens (IOL)[6], laser iridotomy[7], miotic therapy[8], glaucoma drainage tube implantation, insertion of a large IOL[9], laser capsulotomy, intravitreal tramcinolone injection[10], ocular keratomycosis, needling of a trabeculectomy bleb[11] and acute hydrops in Down syndrome patients with keratoconus.

In as much as the precipitating factors can be recognized as highlighted above, there are instances where is it difficult to pinpoint the triggering factor for malignant glaucoma. Reports of spontaneous malignant glaucoma are not common[12,13] but reports from an African or Nigerian setting are very sparse. This article was aimed at reporting a case of presumed bilateral spontaneous malignant glaucoma in a Nigerian woman with resultant blindness, discuss the possible aetiology, and management challenges, and ultimately draw attention to this potentially blinding eye disease.

Case Report

The patient was a 35 year old fashion designer who presented to our clinic with a three day history of blurring of vision and a two day history of complete loss of vision. There was associated ocular pain, redness and photophobia, with headaches. There was no history of nausea or vomiting. There was no history of ocular trauma or application of herbal or traditional eye medications. She was not a spectacle user and there was no other significant ocular history. She was not a known hypertensive or diabetic patient.

Five days prior to presentation, she delivered her second child via vacuum extraction at a private clinic. There was a history of post partum haemorrhage, necessitating transfusion of five pints of blood and admission in the private centre. Two days following delivery the patient noticed sudden

blurring of vision which deteriorated to complete loss of vision two days after. An eye doctor was said to have been called to review her by the private hospital after onset of ocular symptoms, who was said to have made a diagnosis of acute angle closure glaucoma and commenced her on some topical ocular medications. She was subsequently referred to our unit for further management, because there was no improvement in her ocular status.

At presentation, she was pale, but not in any obvious distress, not febrile, and not jaundiced. Other systemic examinations were essentially normal. Ocular examination in both eyes revealed a visual acuity (VA) of no light perception (NPL), mild puffiness of the lids, conjuctival injection and chemosis, cornea edema and opacity with a peripheral circular and yellowish hue (Figure 1). The Patient's consent for the scientific use of the photograph was obtained.

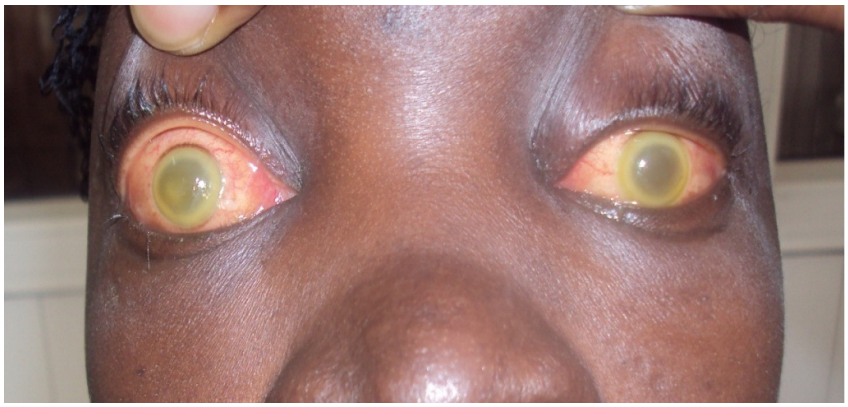

Figure 1. Photograph of Patient Showing Decompensated Cornea in Both Eyes.

There was no view of the anterior chamber of the eyes or further ocular structures. Intra ocular pressure (IOP) measurement using hand held perkins applanation tonometer revealed a pressure of over 50mmgh in both eyes. A preliminary diagnosis of acute angle closure glaucoma with cornea decompensation was made at this point.

The patient was commenced on Latanoprost and timolol eye drops and acetazolamide tablets to lower the IOP, ibuprofen eye drops to control ocular inflammation, and analgesics tablets. Intravenous mannitol 60mg over 20mins was also given. Examination of the packed cell volume (PCV) revealed 15% value; patient was also requested to carry out an ocular ultrasound, and serial monitoring of the IOP was instituted. Haematinics were also commenced, and obstetrics consult was requested. However patient's IOP did not respond significantly to antiglaucoma medication as the IOP remained above 50 mm Hg two days after admission. A diagnosis of malignant glaucoma was entertained at this point. Part of the line of management of obstetricians was a request that the patient be transfused. However, due to financial constraints, the patient opted to discharge herself two days after admission.

Discussion

Malignant glaucoma is a potentially blinding eye disease, if not promptly recognized and appropriately managed. Even though the definition by Von Graefe[1] described a particular clinical scenario post trabeculectomy, the term malignant glaucoma as it is used now probably reflects the clinical course and prognosis and may not be restricted to a single disease entity. Consequently, malignant glaucoma has been associated with many secondary angle closure glaucomas resistant to conventional management protocols.

The exact precipitating factor in this patient is not known, hence the presumed spontaneous terminology given to this case. One of the reasons for this is the late presentation to our facility. At the time of her presentation, vision in both eyes was already NPL, and the corneas were already

decompensated. This rendered further detailed ocular examination impossible. The persistently high IOP over a prolonged period had already caused end stage and irreversible damage to the optic nerve, as well as the cornea. The difficulty and resistance to conventional treatment modalities of malignant glaucoma was demonstrated in this case.

The exact patho-physiology of malignant glaucoma is poorly understood, but the conventional believe is that of Shaffer[14] who proposed that it is due to aqueous misdirection into the vitreous at the level of the ciliary body, lens and anterior vitreous face, with consequent increase in vitreous volume and pressure. He coined the term ciliary block glaucoma. According to the work of Epstein et al[15] it was postulated that there is additional thickening and decreased permeability of the anterior hyaloid surface, with limitation of anterior flow of aqueous humour into the anterior chamber. These lead to forward movement of the iris-lens diaphragm, shallowness of the anterior chamber, and secondary angle closure. However, Quigley[16] had a contrary opinion on the pathway to the development of forward movement of the iris lens diaphragm and resultant angle closure. They contradicted the conventional idea of aqueous misdirection because it contravenes physical laws. They believe that for aqueous misdirection to occur, there needs to be a unidirectional valvular mechanism in place to prevent forward movement of the misdirected aqueous from the vitreous back into the posterior chamber, but no such one way mechanism has been found to exist at the anterior hyaloids surface. Quigley[16] proposed two possible ocular conditions for the development of malignant glaucoma. They implicated choroidal expansion and abnormal vitreous fluid conductivity. Increases in orbital venous pressure and the volume of choroidal extra vascular space will produce choroidal expansion. The latter is thought to be more likely to be responsible for sustained choroidal expansion.

This patient had a history of post partum haemorrhage, necessitating transfusion of seven pints of blood. It is not unlikely that this patient with a low socio-economic status might have had some underlining anaemia even during pregnancy. We postulate that this chronic aneamia, further complicated by acute blood loss might have precipitated extravasations of fluid into the extra vascular space, including the choroid. Another possibility in this patient is perhaps use of sulphonamide drugs. In the report by et al, the isolated malignant glaucoma was later found to have been precipitated by microspherophakia.

The pathway to blindness in this patient (NPL in both eyes) was most likely optic neuropathy from the extremely high, sustained and uncontrolled IOP. The decompensation of the cornea is also related to the high IOP. The poor visual prognosis of malignant glaucoma is well recognized. Unique features of this case were the bilateralism and absence of an apparent predisposing factor. Bilateral presentation of malignant glaucoma is not common[17], and cases without an apparent precipitating factor[18,19] are also rarer. To the best of our knowledge, this is the first report of such a case in Nigeria.

At presentation, the aim of treatment of this patient was effective reduction of IOP, even though vision had been irreversibly affected. She was treated with topical (latanoprost, timolol) and systemic (oral acetazolamide and intravenous mannitol) IOP reducing drugs, as well as cycloplegic agents. However, these had little or no effect, as the IOP remained high for the two days she spent on admission. The difficulty in managing malignant glaucoma is well known[2,20] and a significant proportion of patients might need surgical intervention[18, 21] when medical treatment fails to bring about IOP reduction, through reduction in aqueous production, vitreous shrinkage, widening of the ciliary ring, and eventual posterior movement of the iris lens diaphragm. However, medical treatment is usually the first line of management, and usually consists of:

- Strong cycloplegic agents such as atropine, to paralyze the ciliary muscle, increase zonular tension, and cause flattening and posterior movement of the iris lens diaphragm, thereby opening up the anterior chamber angle.
- Reduction of aqueous production using topical beta blockers (timolol), topical oral carbonic anhydrase inhibitors (acetazolamide), and alpha adrenergic agonist (brimonidine).
- Osmotic agents (IV mannitol, oral glycerol or isosorbide) to shrink vitreous volume and encourage posterior movement of the iris lens diaphragm.

Overall, there is a reduction in IOP using a combination of the above regimen, and this is said to be effective in about 50% of patients[22]. A maintenance dose of atropine is said to be often required to prevent future recurence[20] after gradual discontinuation of other drugs. However, some patients may not respond to medical therapy and surgical intervention may therefore be necessary. A commonly performed procedure is pars plana vitrectomy with or without lens extraction.[23] Anterior hyaloidectomy in an aphakic or pseudophakic eye, lens extraction with anterior vitrectomy in a phakic eye, cyclodialysis, and posterior sclerostomy have also been used. Laser procedures found to be beneficial include; argon laser shrinkage of the ciliary processes[24], ND-YAG laser capsulotomy and hyaloidotomy in aphakic or pseudophakic eyes[25].

The visual outcome in this patient resulting from prolonged uncontrolled raised IOP could perhaps have been prevented by an aggressive early intervention on prompt referral from the primary care givers. This highlights the need to ensure that general practitioners are able to recognize with ocular emergencies, able to give initial palliative intervention if necessary and ensure prompt referral above all.

References

1. Von Graefe A: Beitsage Zur pathologic Und Therapic des Glaucomas. Arch of Ophth 15:108. 1869.
2. Luntz MH, Rosenblatt M. Malignant glaucoma. Survey of Ophthalmology. 1987;32(2):73-93.
3. Ozeki N, Yuki K, Kimura I. Alternative approach to treating malignant glaucoma after trabeculectomy with unplanned zonulectomy. Clinical Ophthalmology. 2010;4:383 - 385.
4. Przybylska-Rybczynska I, Klosowska-Zawadka A, Pecold-Stepniewska H. Malignant glaucoma following trabeculectomy--case report. Klin Oczna. 2004;106:261-262.
5. Adegbehingbe B, Majemgbasan T. A review of trabeculectomies at a Nigerian Teaching Hospital. Ghana Med J. 2007; 41: 176-180.
6. Duy TP, Wollensak J. Ciliary block (malignant) glaucoma following posterior chamber lens implantation. Ophthalmic Surg. 1987;18(10):741-744
7. Cashwell LF, Martin TJ. Malignant glaucoma after laser iridotomy. Ophthalmology. 1992;99:651–658.
8. Rieser JC, Schwartz B. Miotic induced malignant glaucoma. Arch Ophthalmol. 1972;87:706.
9. Reed JE, Thomas JV, Lytle RA, Simmons RJ. Malignant glaucoma induced by an intraocular lens. Ophthalmic Surg 1990;21:177–180.
10. Heatley CJ, Lim KS, Siriwardena D, Barton K. Malignant glaucoma as a complication of intravitreal triamcinolone acetonide. Acta Ophthalmol Scand. 2006;84(5):712-713.
11. Mathur R, Gazzard G, Oen F. Malignant glaucoma following needling of a trabeculectomy bleb. Eye. 2002;16:667–668.
12. Manku MS. Spontaneous bilateral malignant glaucoma. Australian and New Zealand Journal of Ophthalmology. 1985;13(3):249-250.
13. Fanous S, Brouillette G. Ciliary block glaucoma: Malignant glaucoma in the absence of a history of surgery and miotic therapy. Can J Ophthalmol. 1983;18:302-303.
14. Shaffer RN, Hoskins HD. Ciliary block (Malignant) glaucoma. Ophthalmology. 1985;85:215–221.
15. Epstein DL, Hashimoto JM, Anderson PJ, Grant WM. Experimental perfusions through the anterior and vitreous chambers with possible relationships to malignant glaucoma. Am J Ophthalmol. 1979;88(6):1078-1086.
16. Quigley HA. Angle-closure glaucoma-simpler answers to complex mechanisms: LXVI Edward Jackson Memorial Lecture. Am J Ophthalmol. 2009;148(5):657-669 e651.
17. Stan C. Bilateral malignant glaucoma--case report. Oftalmologia. 2005;49(4):33-34.
18. Gonzalez F, Sanchez-Salorio M, Pacheco P. Simultaneous bilateral "malignant glaucoma" attack in a patient with no antecedent eye surgery or miotics. Eur J Ophthalmol. Apr-Jun 1992;2(2):91-93.

19. Saunders P.P., Douglas G.R., Feldman F, Stein RM. Bilateral malignant glaucoma. Can J Ophthalmol. 1992;27(1):19-21.

20. Ruben S.T., Tsai J., Hitchings R.A. Malignant glaucoma and its management. British Journal of Ophthalmology. 1997;81(2):163-167.

21. Chen SD, Salmon JF, Patel CK. Videoendoscope-guided fluorescein-assisted vitrectomy for phakic malignant glaucoma. Arch Ophthalmol. Oct 2005;123(10):1419-1421.

22. Trope GE, Pavlin CJ, Bau A, Baumal CR, Foster FS. Malignant glaucoma. Clinical and ultrasound biomicroscopic features. Ophthalmology. 1994;101(6):1030-1035.

23. Byrnes GA, Leen MM, Wong TP, Benson WE. Vitrectomy for ciliary block (malignant) glaucoma. Ophthalmology. 1995;102(9):1308-1311.

24. Herschler J. Laser shrinkage of the ciliary processes. A treatment for malignant (ciliary block) glaucoma. Ophthalmology. Nov 1980;87(11):1155-1159.

25. Halkias A, Magauran DM, Joyce M. Ciliary block (malignant) glaucoma after cataract extraction with lens implant treated with YAG laser capsulotomy and anterior hyaloidotomy. British Journal of Ophthalmology. 1992;76(9):569-570.