# Management du glaucome malin : revue de la littérature

# Malignant glaucoma management: literature review

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#### RÉSUMÉ

Le glaucome malin reste une complication redoutable de la chirurgie oculaire.il peut se produire spontanément ou après une chirurgie oculaire; le plus souvent une chirurgie de glaucome. Le diagnostic clinique se fait dans le cadre d'une iridotomie périphérique patente et d'un aplatissement axial de la chambre antérieure. La pression intraoculaire est habituellement élève mais peut être normale dans certains cas. L'étiologie exacte de cette pathologie n'est pas reconnue et plusieurs mécanismes sont proposés. Cette revue traite la physiopathologie, le diagnostic différentiel, les modalités d'imageries et les stratégies thérapeutiques actuelles pour contrôler cette forme rare de glaucome secondaire.

#### Mots-clés

Malin, glaucome, management

#### SUMMARY

Malignant glaucoma remains a challenging complications of ocular surgery. It has been reported to occur spontaneously or after any ophthalmic procedure, it is most commonly encountered after glaucoma surgery. The clinical diagnosis is made in the setting of a patent peripheral iridotomy and axial flattening of the anterior chamber. Intraocular pressure is usually elevated, but it may be normal in some cases. The exact etiology of this condition is not fully understood, several mechanisms have been proposed. This review discusses pathophysiology, differential diagnosis, imaging modalities, and current treatment strategies for this rare form of secondary glaucoma.

# **Key-words**

Malignant, glaucoma, management

#### INTRODUCTION

Malignant glaucoma (MG) was described for the first time by Van Grafe in 1869, This condition is defined as following: Shallow or flat of both central and peripheral anterior chamber with increased or normal intra ocular pressure in absence of pupillary block and posterior segment pathology (1).

It is defined by the European Glaucoma Society as secondary angle closure glaucoma with posterior pushing mechanism, caused by ciliary body and iris rotating forward (2). It was also defined as aqueous misdirection, lens block angle closure and ciliary block glaucoma nevertheless exact mechanism of MG remains controversial (3).

Malignant glaucoma is a rare but serious complication that represents a challenging problem.

This review will go overdiagnosis, physiopathology and treatment of the MG.

## **EPIDEMIOLOGY**

Malignant glaucoma is a rare condition with an incidence of 0.4 to 6% (4).

The average age of patient presenting malignant glaucoma is 70-year-old with female predominance (5).

MG is generally found following glaucoma surgery for primary angle closure glaucoma inphakic, aphakic and pseudophakic eye (6,7) and may occur at any time after surgery (8). It can also occur following peripheral laser iridotomy (9), sclera flap suture lysis (10), trabeculectomy bleb needling (11) and following other surgeries including cataract surgery (10), scleral buckling (12), parsplanavitrectomy keratoplasty (13) and even spontaneously (14) or by way of infection (15) or retinal vein occlusion (16).

# Risks factors

Patient with a history of malignant glaucoma in the fellow eye is at higher risk for this complication. Other risk factors are: Axial hyperopia, nanophtalmos (17), chronic angle closure, iris plateau configuration, zonular laxity (pseudo exfoliative syndrome) and Leakage of filtering bleb (18).

# Clinical features, imaging and differential diagnosis

The first management step of this pathology is making an accurate diagnosis.

Patients with malignant glaucoma present painful red eye with rapid decrease of visual acuity. Clinical examination

found flattering of both central and peripheral anterior chamber with often an increased intraocular pressure (IOP). Due to IOP rise, patient examination can be difficult and imaging (UBM and Anterior segment OCT (AS OCT)) is helpful in one hand to confirm the diagnosis and in the other hand to rule out differential diagnosis (19,20). Both technologies show anterior chamber shallowing, irido-corneal touch appositional angle closure, and iris apposition. AS OCT offers fast scanning speed and noncontact imaging. However, compared to AS OCT, UBM can better explore structures surrounding posterior chamber mainly ciliary body by showing its anterior rotation during malignant glaucoma which is helpful to eliminate differential diagnosis.

In fact, three entities should be excluded: suprachoroidal hemorrhage, pupillary block and choroid effusion (21).

#### **PATHOPHYSIOLOGY**

It is important to understand malignant glaucoma mechanism which is also called: ciliary block glaucoma, aqueous misdirection syndrome, cilio-vitreal block glaucoma and positive vitreous pressure glaucoma. It has not to be considered as one disease but a multifactorial condition where the exact etiology of this pathology is not yet fully understood. Three pathogenic mechanism have been proposed.

Shaffer and Hoskins suggested that posterior accumulation of the aqueous humour, behind detached vitreous, induced the forward movement of the iris-lens diaphragm. They postulated the existence of a valve-like mechanism which is "misdirecting" the aqueous humour posteriorly (22).

The second theory is proposed by Chandler. He suggested that lens zonules laxity combined with vitreous pressure result in lens movement onward (23).

Quigly and al proposed that a choroidal expansion is responsible of the vitreous pressure increase. The compensatory aqueous outflow causes the anterior chamber shallowing (24).

Never minded is the mechanism, a same result: a vicious cycle. The increasing transvitreal pressure causes a poor conductivity of the vitreous. This create a cilio-vitreal blockage with an anterior displacement of the lens iris diaphragm, ananterior chamber closing and an elevation of the intraocular pressure which will aggravate the vicious cycle (Fig 1).

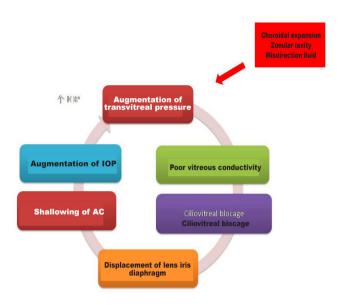


Figure 1: Differentes mecanism of malignant glaucoma

#### **MANAGEMENT**

The purpose of the treatment is to stop the vicious cycle and restore normal aqueous flow. Treatments strategies are based on medical, laser and surgical therapy.

# **Medical management:**

Medical management is the first step to control malignant glaucoma. Simmons reported that 50% of patients with MG respond positively to medical treatment alone (4).

Cycloplegics agents induce ciliary muscle relaxation that participates to tighten the lens zonules and the iriod-cristallian diaphragm is pushed posteriorly (21).

Hyperosmotics are mandatory to ensure medical management success (25).In fact, they induce vitreous drying, contributing to reduce its volume and deepen the anterior chamber. They, also, possibly increase vitreous permeability.

Aqueous suppressants decrease aqueous humourposterior pooling by reducing its production (26). They contribute to decrease the IOP: topical beta blockers and oral acetazolamide are prescribedfor this purpose (27). Topical steroid can be used to reduce inflammation (28).

Therapeutic plan include atropine 1% two times daily, phenylephrine 10% associated to intravenous

hyperosmotic agent (mannitol), and local and oral aqueous suppressants (acetazolamide and betablockers). Miotics are contraindicated. Life long treatment with atropine could be required to ovoid recurrence.

Medical management is usually tried for 3 to 5 days before deciding to switch to other therapeutic alternative, namely laser and surgery, depending on clinical finding (26).

# Laser therapy:

Laser treatment should be used as the second line approach. It's target is to establish a direct communication between restorevitreous cavity and anterior chamber using Nd laser YAG capsulotomy and hyaloidotomy (27) or by eliminating ciliovitreal blockage by lasering ciliary processes.

#### Anterior hyaloids rupture:

Nd laser YAG capsulotomy and hyaloidotomy is considered in aphakic and pseudophakic eyes. It releases the trapped aqueous from the vitreous and allow a fluent communication between the anterior and posterior chamber of the eyethanks to the rupture of posterior capsule and anterior hyaloid membrane. The protocol of using is: Power: 1 to 2 mJ, Laser Shots: 2 to 5.Efficiency of the Nd laser YAG, in cases of refractory MG to medical treatment, was reported by several series (29-31).

### Laser of the ciliary processes:

The second alternative of laser therapy is lasering the ciliary processes.

Trans-scleral cyclodiode laser in pseudophakic eyes helps to eliminate ciliovitrealblockage by shrinking the ciliary process. It can also help reducing aqueous humour production.

A single session is usually sufficient. Setting: Power: 1.5 - 2 w, Time: 2 - 3 S, 20 - 30 pulses, 1 - 2 quadrants (32). Another alternative is to perform direct argon laser through a peripheral iridotomy to eliminate an abnormal contact between the ciliary process and the vitreous body (33).

## Surgical management:

For refractory MG, surgical treatment is indicated. The main goal is to remove the vitreous body and facilitate the aqueous flow between the anterior and posterior chamber (34). This approach was firstly adopted and defended by Chandler and al, who proceeded to a simple aspiration of the anterior vitreous and trapped aqueous via a

large 18G sclerotomy (35). After that, many techniques were discussed. In fact, the key factor in the surgical management is the lens's state. On pseudophakic eyes with MG, core vitrectomy results to a complete resolution of the MG in 65% to 90% of cases (36,37); Partial vitrectomy with irido-zonulo-hyoloidetomy in aphakic and pseudophakic patients is recommended.

In case of phakic patient, vitrectomy without lens remove leads to the resolution of the MG in only 25% of cases (38); therefore, the attitude adopted actually is a core vitrectomy with phacoemulsification associated to a zonulo-hayoidectomy-iridectomy. In fact, with this complete surgical technique, the resolution of MG in phakic eyes can reach 82% of cases (39). In the most severe cases, the recommended procedure is parsplanavitrectomy with iridozonulectomy and phacoemulsification.

## **PREVENTION**

As Malignant glaucoma is a severe complication, prevention is primordial by screening predisposing eyes in order to apply prophylactic measures: longtime treatment with atropine, avoid myotics and transscleral diode laser before surgery. Anterior chamber shallowing is a very important risk factor: it can be avoided by using viscoelastic, tight scleral suturing and adjustable sutures. In eye with non-medically controlled primary angle closure glaucoma, lens extraction should be undertaken rather than trabeculectomy since there is a lower risk of malignant glaucoma during cataract surgery¹and if it occurs the management is easier in pseudophakiceyes; prophylactic vitrectomy combined with phacoemulsification is recommended in case of malignant glaucoma history in the fellow eye (40).

# CONCLUSION

Malignant glaucoma remains a therapeutic challenge. Pathophysiology of this condition remains not completely understood and advances in imaging will probably help to better understand key factors and involved mechanisms. Prognosis improvement is based on appropriate and timely interventions and prophylactic measures.

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