Acute lens-induced glaucomas: A review

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1. Introduction

An acute elevation of intraocular pressure (IOP) can be related to lens-induced factors. This phenomenon can occur spontaneously or be related to trauma to the ocular and adnexal structures. A study of these conditions is imperative, since improper management leads to frequent complications and poor visual outcomes.

Lens-induced glaucomas (LIGs) can be divided into non-traumatic and traumatic types (Figure 1). The non-traumatic types of LIGs are commonly seen in the form of phacomorphic glaucoma (PG), phacolytic glaucoma, phacoantigenic uveitis and lens-particle glaucoma[1]. This group of patients usually have good vision in the other eye, leading to a delay in seeking treatment for the cataract in the affected eye[2]. LIGs can also be associated with certain syndromes such as Marfan’s and homocystinuria, causing ectopia lentis[3]. The dislocated lens in such situations can elevate the IOP through various mechanisms. Traumatic LIGs are associated with lens subluxation/dislocation, lens rupture or rarely a hypersecretory glaucoma[4].

This review was done with the aim of studying the various types of LIGs, their current management and future possibilities. Apart from the textual literature, online search was conducted using search engines such as PubMed, Scopus, Google Scholar, ClinicalKey and the Virtual Library of the Ministry of Health, Malaysia.
2. Glaucoma associated with cataract formation

Acute non-traumatic LIGs can be associated with cataract formation. The clinical presentation of all these is nearly similar with complaints of sudden pain and redness in the involved eye. Usually, a preceding history of gradually progressive decreased vision is also present.

2.1. PG

2.1.1. Definition

This condition is caused by a swollen, cataractous lens causing pupillary or angle block and elevating IOP[5]. PG is a misnomer, since these patients may not show typical glaucomatous optic nerve degeneration and it is preferable to term it as acute phacomorphic angle-closure[6].

2.1.2. Mechanism

PG may occur during the mature stage of cataract formation due to increased contents of the lens (cortex). This makes the zonules loose, causing the lens to move forward. In a mid-dilated position of the pupils, the posterior vector of the dilator muscle of the iris is maximum. This pushes the iris against the lens causing a pupillary block. The collection of aqueous humor in the posterior chamber leads to an iris bombe configuration. Subsequently, if the condition is untreated, the iris will continue to be pushed forward, blocking the angle. In some cases, there may not be a pupillary block and the peripheral iris/ciliary body is pushed forward, blocking the angle. In such a scenario, if the condition is not managed acutely, permanent synechial closure may occur, causing chronic elevation of IOP, despite removal of the lens. An intumescent lens may also occur in the early stage of cataract formation due to physiological changes in the capsule and lens fibers. Lens hydration causes it to swell up leading to pupillary block[3,7,8].

2.1.3. Clinical features

Symptoms of PG include acute redness with pain in one eye and headache. The signs include raised IOP with congested conjunctival/ episcleral vessels, shallow anterior chamber and an intumescent, usually cataractous lens. Corneal edema and flare/cells in the anterior chamber may be present[3,5].

2.1.4. Differential diagnosis

PG needs to be differentiated from acute angle closure (AAC) glaucoma and traumatic anterior sub luxation of the lens. In AAC, the gonioscopic findings in the other eye can provide significant clues. In the latter situation, there is history of trauma and the depth of anterior chamber is often irregular, the part corresponding to the anterior subluxation being shallower. The edge of the subluxated lens may be visible in the pupillary area and an irido or phacodonesis may also be present[8,9].

2.1.5. Management

PG is managed by medical reduction of IOP and a peripheral iridotomy whenever possible. In cases of angle block, an argon laser peripheral iridoplasty may help to open the angle[10]. Topical steroids help to control the inflammation while hypertonic saline reduces corneal edema. Cycloplegia can shift the lens/iris diaphragm posteriorly and deepen the anterior chamber. Pilocarpine causes forward shift of lens-iris diaphragm and should be avoided[8]. The definitive treatment of PG is cataract extraction. A recent study assessing the role of phacoemulsification in PG patients showed good results. The IOPs came down from (49 ± 10.4) mmHg preoperatively to (13.2 ± 2.8) mmHg following phacoemulsification[11]. Femtosecond laser surgery has also been successfully demonstrated in a patient with PG[12]. A study comparing the efficacy of cataract surgery alone versus combined cataract-trabeculectomy did not find any difference in IOP control between the two modalities[13].

According to Bhartiya et al., in cases with 270° or more of synechial closure, combined cataract-glaucoma surgery should be done[14].

2.2. Phacolytic glaucoma

2.2.1. Definition

Phacolytic glaucoma is a condition characterized by leakage of lens contents through an intact capsule[9,15,16].

2.2.2. Mechanism

Previously, the pathogenesis of phacolytic glaucoma was attributed to the leakage of lens material which gets engulfed by macrophages. These distended macrophages subsequently block the trabecular meshwork (TM), affecting aqueous outflow, thus elevating IOP. Electron microscopic studies have shown macrophages with phagocytosed lens material being present in the aqueous humor and TM. However, it was seen that when needling for congenital cataracts was done, large amounts of lens material were released, yet features of phacolytic glaucoma did not develop. Later, Epstein et al. reported leakage of high molecular weight proteins (HMWPs) through intact capsules in eyes identified with phacolytic glaucoma[17]. These soluble proteins have molecular weights of more than 150 × 10^6 daltons and are capable of clogging the TM.

The cortex of hypermature cataracts has 14-fold greater amounts of HMWP, compared to immature lenses. It is assumed that HMWPs are deficient in pediatric cataracts and so phacolytic glaucoma does not develop in that age group. Currently, both macrophages and HMWPs are regarded to play an important role in the pathogenesis of phacolytic glaucoma[9,17,18].

2.2.3. Clinical features

The presentation is with an acute onset of pain and redness. The signs include congestion of the eye, corneal edema, deep AC and usually a hypermature cataract. The AC shows a significant amount of flare, often associated with characteristic iridescent or hyperreflecting particles composed of calcium oxalate or cholesterol.
Clumps of lens material may also be seen in the AC, lining the corneal endothelium or the anterior lens capsule and rarely, in the vitreous[9,19].

2.2.4. Differential diagnosis
AAC, open-angle glaucoma with uveitis, neovascular glaucoma, trauma, occult posterior segment tumor[9,19].

2.2.5. Management
Like other LIGs, the initial management involves control of IOP medically and inflammation with steroids. Manual small incision cataract surgery has proven successful in the management of phacolytic glaucoma[15,20].

2.3. Phacoantigenic uveitis

2.3.1. Definition
This rare condition was termed endophthalmitis phacoanaphylactica by Verhoeff and Lemoine[21]. It is a granulomatous, immune-mediated inflammation to lens proteins. However, the term phacoanaphylaxis is misleading since anaphylaxis requires mast cells, basophils and immunoglobulin E, none of which are present during this process[22].

2.3.2. Mechanism
Capsule rupture leads to exposure of the eye to lens antigens which are antigenically privileged. This is due to their protection by the capsules, the absence of blood vessels and nerves in the lens and the early encapsulation of them during embryogenesis. When the eye is exposed to these antigens, a severe antibody response involving polymorphonuclear leukocytes, lymphoid, epithelioid and giant cells occurs. These inflammatory cells may block the TM and elevate IOP. However, while this antigenic concept was found to be true for rabbits, it was not confirmed in humans[9].

2.3.3. Clinical features
The condition manifests days or weeks after the initial event e.g. trauma, characterized by a latent period during which sensitization occurs. However, it may occur within 24 h if the eye was previously sensitized to the lens protein. This can happen when surgery was done previously in the other eye, with retained lens matter. Occasionally, fine needle aspiration biopsy of aqueous humor needs to be performed in order to confirm the diagnosis[9,23].

On examination, there are lid edema, chemosis, conjunctival congestion, corneal edema, mutton-fat keratic precipitates, severe AC reaction and posterior synechiae[3,23].

2.3.4. Differential diagnosis
The differential diagnosis of phacoantigenic uveitis includes conditions such as sympathetic ophthalmia, other forms of uveitis, PG and lens particle glaucoma[9].

2.3.5. Management
Steroids to control inflammation and medical control of IOP should be done. In certain cases, the lens matter needs to be removed surgically by AC washout or vitrectomy[9].

2.4. Lens-particle glaucoma

2.4.1. Definition
This is a condition resembling phacoantigenic uveitis where lens matter is released from the lens. The lens matter and engulfing macrophages clog the TM, leading to a rise in IOP[24].

2.4.2. Clinical features
The patient reports with sudden pain and redness in the affected eye following trauma or surgery. Occasionnally, these changes happen spontaneously or many years after an inciting event[25]. On examination, the cornea is often edematous, there are fluffy lens particles floating in the AC and a cataractous lens with ruptured capsules is often present[26]. In early cases, the condition is easy to diagnose on the basis of lens particles in the AC and associated history. However, in later cases, the condition can be confused with phacoantigenic uveitis, phacolytic glaucoma or other uveitic conditions associated with an open angle. In comparison to phacolytic glaucoma, the inflammation is more severe, with anterior/posterior synechia and pupillary membranes[9]. If required, aqueous aspiration studies can be done, which would demonstrate leukocytes, macrophages and cortical matter[24].

2.4.3. Management
The high IOP is controlled medically and the inflammation with steroids/cycloplegics. If the amount of lens matter is minimal, a “wait and watch” policy may be adopted. The lens matter may get absorbed spontaneously over time. However, aggressive steroid use may hinder absorption and should be treated with caution. In cases where a significant amount of lens matter is present and IOP is uncontrollable, AC washout should be done[24,25].

3. Glaucomas associated with dislocation of the lens

The characteristic feature of these conditions is the spontaneous dislocation or subluxation of crystalline lenses. The lens can move forward and cause pupillary block glaucoma. In other situations, it may push the iris forward, blocking the angle or get dislocated into the AC entirely and interrupt the aqueous outflow through the TM. In posterior dislocations, the lens may irritate the ciliary body or cause a phacolytic type of glaucoma.

3.1. Simple ectopia lentis
Simple ectopia lentis is a condition seen either congenitally in the absence of other ocular/systemic disorders or spontaneously
later in life. Both the forms show autosomal dominant (AD) inheritance[27]. However, some autosomal recessive (AR) families have also been described[28]. The condition is characterized with bilaterally symmetrical lens dislocations which can occur supero-temporally or into the AC. Associated glaucoma and retinal detachment are other characteristic features[9,29].

3.2. Ectopia lentis et pupillae

Ectopia lentis et pupillae is a rare AR disorder. It is characterized by small, subluxated lenses and oval or slit shaped pupils displaced in the opposite direction from that of lens subluxation. Associated ocular abnormalities include: severe axial myopia, enlarged corneal diameters, iris transillumination defects, poor pupillary dilatation, persistent pupillary membranes, iridohyaloid adhesions, prominent iris processes, retinal detachment and glaucoma[30,31].

3.3. Marfan syndrome

Marfan syndrome is a pleiotropic AD disorder with an incidence of 1 per 5–10 000 births. In almost 80% of the patients, the abnormality involves mutation in the protein fibrillin 1 gene[32]. The condition is diagnosed on the basis of revised Ghent nosology and other diagnostic techniques[33]. Patients have characteristic bilateral ectopia lentis superotemporally. The dislocation is usually stable throughout life. Symptoms of fluctuating blurred vision, monocular diplopia and occasional pain are present. Glaucoma is associated with lens dislocation, aphakia or a congenital anomaly of the angle[9].

3.4. Homocystinuria

Homocystinuria is an AR disorder of methionine metabolism, due to the deficiency of cystathionine-β-synthetase. Systemic features of this condition include mental retardation, skeletal disorders, fine fair hair, shuffling gait and thromboembolic episodes, which often cause death by the age of 20 years[34,35].

Ocular features include ectopia lentis, secondary glaucoma and optic atrophy. The most characteristic ocular finding in homocystinuria is ectopia lentis. The lens dislocation is usually bilateral and inferior. The lens may cause pupillary/angle block glaucoma with subsequent glaucomatous optic nerve degeneration, if undetected[36]. In some cases, the lens dislocation may be preceded by a progressive myopia[35].

3.5. Weill-Marchesani syndrome

Weill-Marchesani syndrome (also known as spherophakia-brachymorpha syndrome or congenital mesodermal dysmorphodystrophy or glaucoma-lens ectopia-microspherophakia-stiffness-shortness syndrome) is inherited as an AD or AR disease with a partial heterozygotic expression[37,38]. It is characterized by short stature, brachydactyly with stiff joints, restricted movements, osteoporosis, minor facial features, mental retardation, microspherophakia, high myopia, ectopia lentis and glaucoma[39]. Glaucoma can occur due to a maldevelopment of the AC and angles, phacoanaphylactic uveitis or as a consequence of ectopia lentis[36,40,41]. The latter is assumed to be due to abnormalities in the zonules, ciliary body structure or the lens fibres. The last feature is seen in microspherophakia, a condition characterized by a smaller diameter and more spherical lens. The weak zonular ligaments allow progressive forward movement of the lens leading to ectopia lentis which can lead to pupillary/angle-closure glaucomas[42].

3.6. Other conditions

Some other rare conditions in which ectopia lentis and glaucoma can occur include hyperlysinemia, sulfite oxidase deficiency and Ehlers-Danlos syndrome[9].

3.7. Management

In mild cases, ectopia lentis can be managed by spectacle or contact lens correction[28]. In some patients, a cycloplegic agent can be tried, which tightens the zonules and reduces the dislocation. Surgical management is frequently complicated, with the need for specialized instruments such as capsular tension rings and implantation of iris- or scleral-fixated intraocular lenses (IOLs) or anterior chamber IOLs[43].

4. Traumatic lens induced glaucomas

4.1. Introduction

Traumatic glaucoma attributable to the crystalline lens can occur through multiple mechanisms. In a study by Bai et al., 21.36% of 103 eyes with traumatic secondary glaucoma had a lens subluxation or dislocation[4]. Choi et al. also found that nearly half of their cases had traumatic dislocation of the lens causing secondary glaucoma[44]. In the series reported by Stanić and Stanić, nearly one third of the patients with secondary glaucoma had dislocated lenses[45]. Netland et al. have also reported one patient with traumatic anterior lens dislocation, pupillary block and secondary angle-closure glaucoma[46].

4.2. Pathophysiology

The mechanism of traumatic lens dislocation is attributed to a sudden compressive deformation of the globe. The shock wave displaces the cornea and anterior part of the sclera posteriorly while the equatorial part of the eyeball expands in compensation.
This movement may damage the lens zonule fibers, leading to subluxation or dislocation of the lens[47]. Pupillary block glaucoma may occur due to a lens dislocating just posterior to the iris, incarcerating into the pupil or complete dislocation into the AC[48]. A lens which is completely dislocated into the AC can block the TM physically, compromising the facility of aqueous outflow. Such a lens may also cause corneal endothelial damage and traumatic uveitis[4,49]. Posterior dislocation of the lens may lead to its close approximation to the ciliary body and cause hypersecretory glaucoma. The lens may also dislocate completely into the vitreous humor. Thereafter, pupillary block glaucoma may develop due to vitreous blocking the pupil. There can be rupture of the lens capsule and release of lens particles leading to phacoantigenic uveitis and glaucoma[4,50].

4.3. Clinical features

Symptoms of lens displacement include blurring of vision, monocular diplopia and distortion of the images. Examination of such a case will show an uneven depth of AC, iridodonesis and the edge of the lens becoming visible. Pupillary block may resemble an AAC, while in phacoantigenic uveitis, severe iridocyclitis is present[47,51]. Trauma may also lead to PG. The patient develops a traumatic, intumescent cataract causing a pupillary block or direct angle compromise by mass effect[50].

4.4. Management

Patients with mild displacement may not require treatment, especially if posteriorly. However, symptomatic dislocation of the lens requires lensectomy, intracapsular cataract extraction or phacoemulsification. In mild cases, phacoemulsification with the use of capsule retractors or stabilizers, iris-sutured or scleral-fixated AC IOLs can be done depending upon the surgeon’s preference[43].

5. Conclusion

This review shows that LIGs can have various modes of mechanism and presentation. It is imperative to identify the correct type of LIG and manage it accordingly. In order to prevent such situations, regular screening of the elderly can be done by paramedical staff, volunteers and non-governmental organizations to identify cataracts early. Families with hereditary disorders should undergo regular screening to detect and monitor lens displacements, while situations leading to trauma can be avoided by personal eye protections.

Conflict of interest statement

The author reports no conflict of interest.

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