Glaucoma: The silent thief of sight

A team of Ophthalmologists discuss Glaucoma, the silent thief of sight owing to its painless nature of irreversible vision loss. They give the basics on its risk factors, aetiology, diagnosis and treatment

Introduction

Glaucoma is often referred to as the silent thief of sight owing to its painless nature of irreversible vision loss. It is an optic neuropathy with characteristic optic disc changes, typical visual field defects with or without raised Intra Ocular Pressure (IOP) of greater than 21mmHg. Primary open angle glaucoma (POAG) is the most common form of the disease: a type of glaucoma with a characteristic glaucomatous optic disc, typical visual field defect and an open angle evidenced on gonioscopy with or without IOP of 21mmHg and above.

POAG is a major cause of visual impairment and blindness in Sub-Saharan Africa (SSA). The WHO World Vision Report 2019 estimated that globally, 76million people have glaucoma and 6.9 million people have visual impairment secondary to glaucoma. More recently, the Lancet Global Health commission report on Global Eye Health reported glaucoma as the third leading cause of blindness globally (3·61 million, CI 2·81–4·42).

In Uganda, the population-based prevalence of glaucoma is unknown. A systematic review of Rapid Assessment of Avoidable Blindness (RAAB) data from Africa in 2013 reported that glaucoma contributed to 38.5% of all blindness, the second in Africa after Nigeria which had 42%.

Risk factors for Glaucoma

The risk factors of developing and progression of POAG were identified in prospective glaucoma studies as well as population-based studies. These were classified as "who is at risk of developing glaucoma" "who has glaucoma" and "who is at risk of developing blindness from glaucoma". In summary, the risk factors for risk of developing glaucoma (incident cases) were increasing age, higher IOP, lower systolic blood pressure (BP) to IOP ratio (BP/IOP), lower mean diastolic ocular perfusion pressure (diastolic BP minus IOP), thinner central corneal thickness (CCT), and a positive family history of glaucoma. The risk factors for "who has glaucoma" (prevalent cases) were increasing age (more cases in the higher age groups [70-79 years] compared to the lower groups [40-49 years]), gender (more in males than females), raised IOP, positive family history of glaucoma, race (more in blacks compared to whites), lower Body Mass Index (BMI) especially in men and history of cataract surgery. The risk factors for developing blindness from glaucoma included increasing age (more than 60 years), IOP greater than 31mmHg, race (more aggressive in blacks than whites), delayed presentation to hospital and untreated glaucoma. Cases of early-onset glaucoma (age less than 40 years) follow genetic inheritance patterns with mutations that are highly penetrant.

In summary, the most common as well as modifiable risk factor for glaucoma aetiology, progression and blindness is a high intraocular pressure.

Aetiology

Glaucoma can be primary or secondary. Primary glaucoma has no identifiable cause with a list of risk factors that have been coined following large glaucoma trials.

Secondary glaucoma has identifiable and sometimes modifiable causes some of which include; prolonged use of Ocular and systemic corticosteroids, ocular trauma, ocular surgeries, lens induced glaucoma, Pseudo exfoliation syndrome among others.

Diagnosis

Diagnosis is based on a detailed patient history, full ocular examination including tonometry (measurement of IOP), gonioscopy (evaluation of the anterior chamber angle), anterior and posterior segment imaging and perimetry (Visual field Test).

In the history; keen attention is drawn to risk factors discussed above, presence of systemic illnesses and other secondary causes of glaucoma.

The cornea may be enlarged and sclera thinned with bluish-coloration in childhood glaucoma owing to the flexibility of the collagen fibres. The entire eye may be buphthalmic in paediatric glaucomas.

The lens; may have a cataract and swollen to cause anatomical narrowing of the anterior chamber angle, may have leaky protein and consequential lens-induced glaucomas present.

Goniotomy: The anterior chamber angle assessed for patency, presence of pseudo exfoliative substance, pigment dispersion, blood, and other material that could block it or close it causing angle closure glaucoma. The anatomy is also assessed for; recession in case of trauma, as well as degree of openness.

Intraocular Pressure is normally between 9 and 21mmHg. Pressure higher than 21mmHg should be further investigated.

Parchymetry; this measure the corneal thickness. Thin corneas are a risk of development and progression of glaucoma.

Characteristic optic nerve changes in glaucoma include but not limited to cupping, nasalisation of the blood vessels, thinning of the retinal nerve fibre layer, disc bleeds, RNFL kinks, and Lamina dot sign among others.

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Imaging: Ultrasound scan of the anterior chamber may reveal anterior chamber angle closure, Optical Coherence Tomography is done to evaluate the retina nerve fibre layer, the macula, the optic disc as well as monitoring progression of the disease and timing of interventions.

Perimetry to identify visual field defects that are characteristic of glaucoma such as an early nasal step scotoma, arcuate scotomas, and constricted visual fields in advanced disease is a very useful tool in diagnosis and follow-up of glaucoma patients.

Treatment

Landmark trials in the management of glaucoma which included; The Collaborative Initial Glaucoma Treatment Study (CIGTS), Ocular Hypertension Treatment Study (OHTS), Collaborative Normal Tension Glaucoma Study (CNTGS), Early Manifest Glaucoma Trial (EMGT), United Kingdom Glaucoma Treatment Study (UKGTS) all were in agreement that primary management of glaucoma involved lowering of the intraocular pressure. These emphasized early initiation of medical therapy in those with early disease as well as patients with just an elevated IOP showing no signs of optic nerve damage; the ocular hypertension group.

Treatment initiation requires setting of a target IOP depending on the severity of disease. The various modalities of treatment are then chosen based on which one would be best to achieve the set target.

Medical treatment

Medical therapy includes different classes of IOP lowering drugs namely; Prostaglandin analogues such as Latanoprost, Bimatoprost which are first line therapy in adults, Topical beta adrenergic antagonists like Betaxolol, Timolol which are first line medical therapy in children; they should be avoided in patients with asthma and significant cardiac disease owing to their systemic absorption and mechanism of action that worsens these diseases. Carbonic anhydrase inhibitors like Acetazolamide, dorzolamide; alpha adrenergic agonists like Brimonidine; Parasympathomimetic agents such as pilocarpine and echothiophate iodide also form bulk of the medical treatment of glaucoma. Rho kinase inhibitors like Netarsudil and hyperosmotic agents which include glycerol and mannitol are also utilised in the medical treatment.

Laser treatment

Laser therapy of various forms contributes to management of glaucoma; selective laser trabeculoplasty can be done to remodel the trabecular meshwork and increase aqueous humour outflow, Laser peripheral iridotomy creates a hole in the peripheral iris providing an alternative pathway for aqueous passage into the anterior chamber and peripheral iridoplasty which causes thinning of the peripheral iris to widen the anterior chamber angle.

Surgical treatment

Trabeculectomy is the most common glaucoma surgery done in Sub-Saharan Africa. Its aim is to create a fistula between the anterior chamber and the sub-conjunctival space. This creates a stable outflow and thus reduction of IOP. This is usually indicated when maximally tolerated medical and laser therapy are insufficient or fail to prevent progressive damage. New advances however suggest earlier surgical invention to prevent progression which comes from the unstable IOP fluctuations in the medically treated groups. Tube shunt surgeries involve creating an alternative pathway for the drainage of aqueous as in Trabeculectomy however, the fistula is maintained by a drainage device which can either be valved such as the Ahmed glaucoma valve or non-valved like the Molteno and Baerveldt.

Minimally Invasive Glaucoma Surgeries (MIGS) are a group of incisional glaucoma procedures that enhance the pre-existing aqueous outflow channels and not just creating new ones as seen above. These include; stents to Schlemm canal, Trabecular Meshwork disruption, stents to the suprachoroidal space.

In children; trabecolotomy and goniotomy are the first line treatment modalities.

Cyclodestruction: This is done in management of end-stage disease without visual potential but with very high IOP. The aim is to cause destruction of the ciliary body which is responsible for aqueous production. This in turn lowers the IOP

Prognosis and Follow-up

All cases of paediatric glaucoma require life-long follow up to look out for potential complications from surgeries, progression of disease and management of complications of glaucoma. Ambylopia is by far the commonest cause of visual compromise which usually results from corneal opacification, uncorrected myopia from buphthalmic eyes; this requires urgent diagnosis and treatment. Children require early referral to vision rehabilitation centres during the course of treatment.

Adult patients whose IOPs are well controlled on whichever form of treatment may follow two paths; one of stable disease in which no progression or worsening is noted on follow-up visits or a form in which disease progression occurs despite having achieved the target IOPs. In such cases the target IOP is adjusted downward. Follow up involves routine IOP checks, Visual field tests, OCT of both anterior and posterior segment and a detailed ocular examination.

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