Retinal diseases: Are we missing Glaucoma?

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Retinal disease can be complicated by a variety of glaucomas, usually of the secondary variety. The glaucoma might be easily overlooked while attention is focused on the acute visually threatening retinal affliction. Glaucoma may appear in several retinal and vitreo retinal disorders, and retinal manifestations of specific glaucoma syndromes. Understanding these conditions and the nature of their association with glaucoma can aid the identification and timely management of these forms of glaucoma.

Retinal disorders associated with glaucoma :

Several vitreo retinal diseases may be complicated by glaucoma .Here an attempt has been made to highlight the most frequent associations of retinal diseases with glaucoma seen in practice.

Age related macular degeneration :

The late forms of age related macular degeneration like geographic atrophy and exudations affect about 7% of individuals over 70 yrs. This exudation may result inchronic retinal detachment which may lead to neovascularglaucoma .In some cases exudation may cause massive vitreal, subretinal, or suprachoroidalhaemorrhage leading to anterior displacement and rotation of the lens-iris diaphragm with angle-closure glaucoma. Ghost cell glaucoma may occur as a complication of vitreous haemorrhage from age-related macular degeneration.

It has been seen that prescription of anticoagulants to elderly, hypertensive ARMD patients may also infrequently cause massive posterior segment haemorrhage and subsequently angle closure glaucoma…

Retinitis pigmentosa :

The reported overall glaucoma incidence varies between 2.98% to 10% of all cases of retinitis pigmentosa. But the prevalence of primary open-angle glaucoma ranges from 2 to 12%, whereas that of primary angle-closure glaucomain RP patients over the age of 40 is 1%.These category of patients prove to be both a diagnostic and therapeutic challenge.visual field changes In RP like midperipheral scotoma that eventually evolve into a constricted central island of vision may mimic or obscure classic glaucomatous field loss. Similarly waxy pallor of the optic disc in RP can obscure disc cupping and erosion.Because of these features that mimic glaucoma it difficult to detect progression of glaucoma in RP.

Retinitis pigmentosa-like syndromes may mimic glaucomatous field loss, and the pattern of advancement of visual field loss in true retinitis pigmentosa may parallel that of typical open-angle glaucoma.

Vitreoretinal syndromes:

A number of syndromes involving retinal or vitreoretinal degeneration and glaucoma have been reported.Of utmost importance isStickler's Syndrome {hereditary arthroophthalmopathy}, an autosomal dominant disorder that demonstrates multiple systemic abnormalities like flattened facies, high-arched palate, hearing loss, scapular abnormalities, and knee pain due to spondyloepiphysealdysplasias and ophthalmic manifestations like prominent peripheral vitreoretinal membranes associated with retinal detachment, high myopia, nuclear sclerotic cataracts, and glaucoma .The glaucoma has been described as an open-angle type with both congenital and later-onset varieties.

Other vitreoretinal syndromes resulting in glaucoma include Norrie disease, retinal dysplasia ,Wagner disease and Walker-Warburg syndrome.

Retinal Detachments:

A study by Becker et.al has reported that 5.8% of patients with retinal detachments (31 of 530 patients) had unrecognized primary open-angle glaucoma in the contralateral eye, with the diagnosis based on disc analysis, visual fields, tonometry, and/or tonography.In another study,the glaucoma preceded the retinal detachment historically or by clinical evidence in 7.3% (60 of 817 patients) and followed the retinal detachment in 2.2% (18 of 817 patients).Some reports further suggested a possible association between the use of miotics and acute retinal detachments.

Schwartz Syndrome-patients with a unilateral elevation of intraocular pressure, ranging from 29 to 55 mm Hg, with a rhegmatogenous retinal detachment.The elevated pressures respond poorly to glaucoma medications, and successful retinal repair result in a normal IOP and outflow facility.Thus it is important to exclude an occult retinal detachment in any medically unresponsive unilateral glaucoma.

Retinopathy of Prematurity :

Thirty percent of eyes with severe ROP may develop angle-closure glaucoma.

In those eyes with the cicatricial phase of the disease, crowding of the anterior ocular structures predisposes to a pupillary block glaucoma.angle-closure glaucoma can occur in a seeing eye despite high myopia,in later life. The glaucoma mechanisms in these severely afflicted eyes may be multifactorial and include iris neovascularization.Late-onset angle-closure glaucoma has been reported in patients ranging from 12 to 45 years of age.

Patients with retinopathy of prematurity have a lifelong risk of developing angle-closure glaucoma.

Intraocular tumors :

Both benign and malignant tumors can produce a secondary glaucoma.Although uveal melanomas are often initially associated with relative hypotony,but subsequent growth lead to glaucoma through either open- or closed-angle mechanisms.Most common of these mechanisms are pigment dispersion and direct tumor invasion of the angle.Others include obstruction of the trabecular meshwork with the tumor and inflammatory cells, and angle closure from neovascularization, peripheral anterior synechiae, suprachoroidalhemorrhage, and mechanical effects of the tumor mass.

In a study, it was found that glaucoma rate was only 2% in eyes with choroidal melanomas but rose to 17% in eyes with melanomas of the ciliary body.

Myopia:

Relationship between refractive error and primary open-angle glaucoma is currently unclear.Some studies suggest that glaucoma and myopia are linked through the factors related to optic nerve susceptibility.

Tilted discs and enlarged optic discs and cups, often with visual field defects similar to those seen in glaucoma complicate our ability to detect progressive glaucomatous optic nerve damagein myopics.

Central Retinal Artery Occlusion-

Neovascular glaucoma occurs in approximately 15% of patients with central retinal artery occlusion, nearly always within 1-2 months of the occlusive event. Mechanism of anterior segment neovascularization remainscontroversial.These patients clearly have altered retinal perfusion and it is important to consider previously undiagnosed carotid obstructive disease.

As survival appears to be decreased in persons with a centralretinal artery occlusion, patients should undergo a systemic evaluation for collagen-vascular and cardiovascular disorders.

Central Retinal Vein Occlusion (CRVO) :

Patients with central retinal vein occlusion have anapproximately 20% risk of developing neovascular glaucoma, primarily due to the development of retinal ischemia. If major retinal capillary compromise is present, iris neovascularization may develop within 1 to 3 months and produce peripheral angle synechiae and angle closure.In rare cases, eyes with central retinal vein occlusioncan develop a reversible form of angle-closure glaucoma without iris or angle neovascularization.

In 20% to 30% of cases of central retinal vein occlusion, signs of primary open-angle glaucoma are present in the contralateral, uninvolved eye.

Diabetes Mellitus :

Epidemiological studies have addressed the association between diabetes and primary open-angle glaucoma.Many cases of iris neovascularization develop following ocular surgery, particularly following pars planavitrectomy for posterior segment complications of proliferative diabetic retinopathy, especially if lensectomy is required (risk increased by twofold) resulting in neovascular glaucoma.

In diabetic eyes requiring complicated vitrectomies, usually with lensectomy, a unique syndrome of fibrin pupillary block glaucoma occurs.Fibrin , a result of surgical

trauma, clots and occludes the pupil, resulting in an acute aphakic pupillary block glaucoma.

Coats disease :

Congenital retinal telangiectasia, or Coats' disease may lead to secondary angle-closure glaucoma by neovascular or inflammatory mechanisms.The intravitreal exudates may lead to total retinal detachment, which can limit attempts to treat the retinal ischemia with panretinal photocoagulation (PRP).

Many of these eyes ultimately requireenucleation.

Retinal Dysplasia :

The term designates a pathologic retinal lesion involving dysplastic retina in a malformed eye.This condition is seen in persistent hyperplastic primary vitreous, chromosomal trisomy syndrome 13-15, and the fetal effects of maternal ingestion of D-lysergic diethylamide (LSD). Secondary angle-closure glaucoma of the pupillary block variety may develop as a result of anterior chamber angle malformation in retinal dysplasia.

Choroidalhaemangiomas and Sturge-Weber Syndrome:

In patients with choroidalhemangiomas, the most common mechanisms of developing glaucoma include neovascular glaucoma from retinal ischemia secondary to total retinal detachment, and angle closure due to choroidal effusion.Sturge-Weber syndrome is a common cause of choroidalhemangiomas.Thirty to 50% of patients with Sturge-Weber syndrome may develop glaucoma, and case series suggest that 60% of these are congenital, with 40% having a juvenileor adult onset.Glaucoma in infancy is similar to that of other congenital forms of glaucoma.Later-onset glaucoma generally has an open angle and increased episcleral venous pressure and can be associated with supraciliary effusion.

The elevated episcleral venous pressure in this syndrome and the extent of the episcleralangioma (often more obvious at surgery when conjunctiva was reflected) is proportionate to the severity of the glaucoma.

Other Retinal Entities Associated with Glaucoma:

Unusual cases of acute angle-closure glaucoma developed in young men who are later found to have positive human immunodeficiency virus (HIV) titers.The glaucoma was of choroidalcyclocongestive type.

Examples of other retinal disorders with which cyclocongestive angle-closure glaucoma may be rarely associated areScleritis,Pars planitis,Harada's disease,Nanophthalmos.

Overview of Retinal diseases that may be complicated by glaucoma

Age related macular degeneration	Closed angle
Retinitis pigmentosa	Open and/or closed angle
Other vitreoretinal syndromes	Open and/or closed angle
Retinal detachment	Open angle
Retinopathy of prematurity	Closed angle (early and late)
Intraocular tumors	Open and/or closed angle
Myopia	Open angle
Central retinal artery occlusion	Neovascular glaucoma
Central retinal vein occlusion	Neovascular glaucoma
Diabetic retinopathy	Neovascular glaucoma,Open angle
Coats disease	Neovascular glaucoma
Retinal dysplasia	Closed angle
Choroidalhaemangiomas	Open and/or closed angle

Special points to ponder:

Since the association of retinitis pigmentosaand ARMD with glaucoma is a common occurrence, Unoprostone isopropyl a drug administered topically as eye drops, approved for the treatment of glaucoma in the United States is now being tested {is under trail} as a potential treatment for both retinitis pigmentosa (RP) and dry age-related macular degeneration (AMD).

Conclusion:

A good understanding and an organised approach to the vast array of interrelations of the retinal disorders and associated glaucomas will promote diagnostic accuracy and an effective therapeutic plan.

We are what our thoughts have made us; so take care about what you think.
Words are secondary. Thoughts live; they travel far.

- Vivekananda