

Myopic Degeneration and it's Low Vision Management- A Review

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Abstract

Myopic degeneration is a condition characterized by the progressive expansion of the eyeball axial length which is not stabilized during the adulthood, causing a progressive degeneration. These changes result in visual impairment, changes in contrast sensitivity, reduced dark adaptation, visual field changes and electrophysiological changes. This causes extremely high amount of nearsightedness causing a major alteration of the shape or globe of the eye, which may lead to profound visual loss. The etiology of myopic degeneration is still not clear, but the influence of genetic and environmental factors has been increasingly recognized.

This disease is a subtype of high myopia, which is characterized for refractive values lower than -6 diopters and axial length greater than 26.5 mm. The prevalence of degenerative myopia is almost about 2% worldwide. Myopic degeneration may lead to visual impairment and hence low vision management and rehabilitation should be considered in these cases.

Keywords: Pathological Myopia; Myopic Degeneration; Visual Impairment; Low Vision; Blindness; Low Vision Rehabilitation

Introduction

Myopic degeneration is a condition characterized by the progressive expansion of the eyeball axial length which is not stabilized during the adulthood, causing a progressive degeneration [1-4]. The progressive expansion of the eyeball leads to the formation of posterior staphyloma and degenerative changes of the sclera, choroid, Bruch's membrane, retinal pigment epithelium and neural retina. These changes result in visual impairment, changes in contrast sensitivity, reduced dark adaptation, visual field changes and electrophysiological changes [5]. Pathological Myopia is an extremely high amount of nearsightedness that causes a major alteration of the shape or globe of the eye, which may lead to profound visual loss attributed to the structural damage of the retina [1]. The hallmark of this condition is an expansion of the ocular posterior segment associated with increasing axial length [1].

The recent WHO publication on myopia defined it as: a vision-threatening condition occurring in people with myopia, usually high myopia that comprises diffuse, patchy macular atrophy with or without lacquer cracks, CNV, and Fuchs' spot [6].

Etiology

The etiology of myopic degeneration is still not clear, but the influence of genetic and environmental factors has been increasingly recognized. There is a strong belief that degenerative myopia may be genetically inherited, but the form of inheritance is still poorly understood [2,7]. The incidence of this condition varies so much between the various ethnic groups.

Genetic markers that may be associated with genes for myopia have been located on human chromosomes 1, 2, 12 and 18. There is some genetic information on the short arm of chromosome 2 in highly myopic people [8]. It has been reported that a gene for familial high myopia with an autosomal dominant transmission pattern could be mapped to human chromosome 18 and 12 [9].

The genetic defects responsible may be transmitted between the generations in a variety of ways, and may produce very different degrees of myopia in different family members [10].

Prevalence

This disease is a subtype of high myopia, which is characterized for refractive values lower than -6 diopters and axial length greater than 26,5 mm. The prevalence of degenerative myopia is almost about 2% worldwide however, it is progressively increasing year by year around the world [11-13]. It is ranked seventh in the United States in the cause list of legal blindness, fourth in Hong Kong and second in China and Japan [10]. Prevalence of myopia is 47.16% of total refractive error (among these 6.56% high myopia) in Nepal [14] who might also have risk in progressing to degenerative myopic changes.

High myopia is a major cause of legal blindness in many developed countries [15]. The impact of myopic retinopathy on visual impairment is important because it is often bilateral and irreversible, and it frequently affects individuals during their productive years.

Ocular changes seen on myopic degeneration [5,7,16,17] are:

Overall

- Prominent looking eyes (due to increased axial length of the eyeball)
- Large cornea and deep Anterior Chamber
- High degree of myopia (More than -6.00 Ds)
- Cataract
- Glaucoma
- Vitreous liquefaction and detachment..

Fundus changes

- Diffuse tessellation of the fundus (thinning of the RPE)
- Peripapillary scleral crescent (failure of the choroid and RPE to extend to disc margin)
- Posterior staphyloma (thinning and bulging of sclera posteriorly due to overstretching)
- Lacquer Cracks in the Bruch's membrane (Yellowish appearing linear breaks in Bruch's membrane)
- Fuchs' Spot (A rounded black lesion as a result of RPE hyperplasia usually overlying a previous choroidal neovascular membrane)
- Retinal Holes and Detachment (Due to peripheral retinal degeneration and vitreous liquefaction)
- Choroidal neovascularization
- Subretinal hemorrhage

- Tilting of the optic disc
- Geographic areas of atrophy of the retinal pigment epithelium and choroids.
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Based on long-term clinical observations that showed the progression patterns and associated factors of the development of myopic choroidal neovascularization (CNV), the META-PM Study [18] has classified myopic maculopathy lesions into five categories (Table 1). Additional features were added to these categories and were included as “plus signs”: (1) lacquer cracks and (2) myopic CNV. Fuchs' spots were categorized under the term of myopic CNV as it represented a scarred form of myopic CNV. “Plus signs” have been separately listed owing to its strong association with central vision loss; however, they did not fit into any particular category and might develop from, or coexist, in eyes with any categories of lesions. Based on this classification, pathologic myopia has now been defined as myopic maculopathy Category 2 or above, or the presence of “plus” sign or posterior staphyloma [18,19].

Category	Features	Plus lesion
Category 0	Normal Fundus	Myopic CNV, Lacquer cracks
Category 1	Tessellated Fundus	
Category 2	Diffuse Chorioretinal Atrophy	
Category 3	Patchy Chorioretinal Atrophy	
Category 4	Macular Atrophy	

Table 1

Visual functions

Visual Acuity

Visual acuity may be correctable to 20/20 initially. But in advanced cases central acuity can be severely affected as a result of structural changes in the central retina (Posterior staphyloma; myopic macular degeneration).

Visual field

High magnitude of myopia can result in a variety of field defects. Central ring-shaped scotoma, hemianopic and quadrantic defects are commonly encountered in the presence of posterior staphyloma.

Contrast sensitivity

There is a loss of higher spatial frequency in high degrees of myopia.

Associations with other conditions

Pathological myopia has been associated with other ocular and systemic diseases. These conditions include Down's syndrome, Ocular Albinism, Infantile Glaucoma, Marfan's Syndrome, Retinopathy of Prematurity, Ehler's-Danlos Syndrome, low birth weight, and maternal alcoholism. Patients with these diseases or conditions should be considered "at risk" for pathological myopia and carefully monitored [7].

Signs/symptoms

These include the sudden appearance of flashes of lights (photopsia), like lightning flashes. Also, floaters, little shadowy dots, and/or cobwebs, shadowy strands could be the first sign of an impending retinal detachment or a tear of the retina. If the patient experiences the sudden onset of both flashes and floaters at the same time, the risk of retinal detachment is extremely high. In a retinal detachment, we may notice impairment in one area of your vision described like a curtain coming down. This can occur from any direction including the right or left side or from the top or bottom of the vision. Visual acuity may be reduced markedly or mildly depending on the site of the detachment.

Investigations

- Retinoscopy/Refraction
- Tonometry
- Ultrasound (A and B Scan)
- Optical Coherence Tomography (OCT)
- Fundus Fluorescein Angiography (FFA)
- Fundus photography.

Treatment and control

Degenerative myopia is a challenge for eye care practitioner since this condition requires long term medical care and comprehensive exploration during the progressive course of the pathology. It is often suggested that high myopic patients should adopt certain measures to prevent the progression of the condition like: avoid staying in rooms with artificial lighting for long time and the importance of establishing regular medical follow-up due to their pathological condition [20]. Degenerative myopia may be preventable in the future. Today, however, the usual treatments are limited to optical correction, intra-ocular pressure control, and attention to complications that may occur. The impact of myopic retinopathy on visual impairment is important because it is often bilateral and irreversible, and it frequently affects individuals during their pro-

ductive years. Refractive status, ageing, axial elongation and retinal degeneration appear to be the main determinants for a successful visual outcome.

The different treatment options available are:

- **Optical:** PAL's, MyoVision™ spectacles and Defocus Incorporated soft Contact (DISC) Lens [21,22], Defocus Incorporated Multiple Segments (DIMS) spectacle lenses [23], SightGlass Vision [The CYPRESS study].
- **Medical:** 7-methylxanthine [21], Atropine [24,25].
- **Surgical:** Scleral Buckling, Donor sclera for posterior staphyloma.
- **LASER:** LASER photocoagulation for central and peripheral retinal degenerations.

Low vision management

Degenerative myopia represents one of the important causes of low vision. Several factors may concur to cause low vision in highly myopic eyes: choroidal neovascularization, macular atrophy, lacquer cracks, macular hole, retinal detachment, glaucoma, macular retinoschisis, complications due to cataract extraction and, most recently, complications due to refractive surgery [2,7,10,16].

The high incidence of retinal pathologies or complications associated with high myopia, inducing a total or partial vision loss, are the main causes that account for the great number of highly myopic patients with low vision. These subjects have difficulty accomplishing visual tasks, but can enhance their ability by using compensatory strategies, optical devices and by modifying the environment [26]. The different optical and low vision managements of myopic degeneration is highlighted below;

Spectacle dispensing

A small round frame, high index lenses, and antireflective coated lenses help to reduce edge thickness and peripheral distortions. For higher power, a myodisc or blended myodisc should be considered. Strong/unbreakable frames with polycarbonate or Trivex lenses in spectacles should be considered as these individuals have thick powered lenses and are more prone to accidents owing to difficulty in orientation and mobility.

Spectacle vs contact lens correction

Correction of refractive error with conventional spectacles usually improves acuity. But it produces minification of retinal image compared to contact lenses. Relative magnification produced by a

contact lens serves as an advantage over a spectacle lens of same power. Furthermore, contact lens use may be advantageous since it provides a wider macular field of view as opposed to spectacles. In addition, Contact Lens also eliminates peripheral distortion and prismatic effects experienced with high-powered spectacles secondary to its aesthetic value.

Low vision management

- For Spectacle users, use of near magnification devices may be compensated by simply removing the habitual correction to read at the appropriate working distance.
- For Contact Lens users, he may choose to use different types of magnifiers (spectacle magnifiers, hand-held magnifiers and stand magnifiers) over contact lenses.
- For spotting and scanning distance objects, telescopes (Handheld or spectacle mounted clip-on) can become handy.
- Assistive technology such as Close Circuit Television (CCTV) can provide the individual with many magnification options, polarity options and comfortable near activities.
- Direct illumination along with magnification and reading stand are helpful for performing near tasks for longer durations that may ease posture related complications as well.
- Eccentric viewing technique is beneficial, if central vision is affected.
- Peak cap and Sunglasses are best for outdoors to tackle photophobia.
- Flashlight may be helpful for poor night vision.
- Relative larger size devices like large print materials, large clocks, telephones, calendars, playing cards, etc. can be of great help for dailies and recreations.
- Talking clock, talking telephones, talking books, tape recorders, tactile cards etc. can make their dailies easier.
- Readout sphygmomanometers, large print syringes, large readout thermometers, pre-filled syringes etc. can help the visually impaired in self medical management.
- Bold line paper, Black ink with felt tipped pen for writing.
- Typoscopes, Notex, signature guides, Braille etc. are very helpful for visually impaired individuals.
- In cases of difficulty in orientation and mobility, they can be advised to use sighted guides, dog guides and canes after proper trainings from the professionals. These days, modern electronic travel aids like Pathsounder, Laser cane, Mowat sensor, Nightscope etc. can be used for easy orientation and mobility.
- Environmental modifications will be helpful for most of these individuals.

Patients with degenerative myopia have thinned retina which may easily develop tears, holes and detachments and must not involve in any physical activities that may lead to jostling and/or trauma to the eyes. Contact sports and ball sports are also dangerous for these groups. Even activities like jumping rope or volleyball can lead to a retinal detachment. Children should not participate in physical education or competitive sports without the prior review and approval of their eye care practitioners.

Uncorrected refractive errors are common cause of visual impairment and blindness worldwide. Nearly a quarter of this world population is estimated to be living with this problem [27]. To reiterate, degenerative myopia is a vision threatening condition that causes irreversible retinal changes and severe form of visual impairment. Although visual prognosis is poorer in patient with myopic degeneration, it is important to recognize the patient with myopic degeneration for successful management and rehabilitation.

Conclusion

Prevalence of myopia and pathological myopia is increasing alarmingly and so is the number of visually impaired people. Visual impairment may be an impediment for not only an individual but also a family, a society and a whole nation. Accessible Low vision management and visual rehabilitation can make a person with VI independent and hence contribute in national growth. Timely examination, regular follow up, specialty consultation and proper visual rehabilitation is a must for a person with myopic degeneration.

Financial Interest

None

Conflict of Interest

Nil

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