Ophthalmoscopic Findings in Patients of Pathological Myopia

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Abstract-

Purpose - To study the ophthalmoscopic findings in patients of pathological myopia.

Methods- This was a prospective observational study that involved 50 eyes of 25 patients with pathological myopia complaining of diminution of distant vision. Complete ophthalmic examination was done in diffuse light followed by direct and indirect opthalmoscopic examination.

Results-*There were 11 males and 14 females .The most common myopia-related macular finding in patients with high myopia was staphyloma (22%), followed by chorioretinal atrophy (18%),tigroid fundus (16%), lacquer crack (12%), retinal hemorrhage (4%), active myopic choroidal neovascularization (2%), and Fuchs spot (2%). The most common disc finding associated with high myopia was peripapillary atrophy (81%), followed by disc tilt (56%). Staphyloma and chorioretinal atrophy increased in prevalence with increasing age, increasing myopic refractive error, and increasing axial length.*

Conclusion- Typical pathological features of pathological myopia include diffuse or patchy chorioretinal atrophy, posterior staphyloma, lacquer cracks, Fuchs spots, choroidal neovascular membrane (CNV), and sometimes even foveoschisis. These pathologic changes often lead to progressive loss of vision due to a number of degenerative changes occurring at the macula. Pathological changes in high myopes start in childhood and become prominent in adulthood.

Keywords: pathological myopia, peripapillary atrophy, fuchs spots, choroidal neovascular membrane

Date of Submission: 04-07-2020

Date of Acceptance: 19-07-2020

I. Introduction

High myopia (HM) is an important cause of visual loss, especially in the younger population. It has been defined as a refractive error with spherical equivalent exceeding -6 diopters (D) and/or the axial length longer than 26.5 mm.[1,2] Pathological myopia or degenerative myopia refers to high axial myopia with characteristic pathological changes at the posterior pole.[3,4,5] Typical pathological features include diffuse or patchy chorioretinal atrophy, posterior staphyloma, lacquer cracks, Fuchs spots, choroidal neovascular membrane (CNV), and sometimes even foveoschisis.[6,7]. The inheritance of pathological myopia can vary from being autosomal dominant, autosomal recessive, X-linked recessive to a monogenic pattern. It may also be associated with certain genetic syndromes: Stickler syndromes Type 1 and 2, Type 4 Ehlers–Danlos syndrome, Knobloch syndrome, Marfan syndrome, Noonan and Down's syndrome.

Pathophysiology

Pathological changes in high myopes start in childhood and become prominent in adulthood. To begin with, there occurs excessive axial elongation. Axial elongation results in chorioretinal stretching and subsequent thinning. The mechanism behind pathological axial elongation includes an emmetropization process and involves a structural alteration of the collagen proteins. Abnormal collagen proteins may lead to degenerative changes in the retina, choroid, and sclera.

Visual acuity may be subnormal even before advanced myopic maculopathy sets in. One of the reasons behind this may be the alteration in the arrangement of photoreceptors. The arrangement of the photoreceptors in high myopes is affected due to excessive stretch in the posterior pole. This may lead to subnormal visual function (Stiles–Crawford effect). In high myopes, the cones in nasal hemiretina are aligned toward the optic nerve, whereas they are aligned toward the center of the exit pupil in temporal hemiretina. This discrepancy in receptor alignment is directly associated with the axial length.

Myopia-related complications such as posterior staphyloma and chorioretinal atrophy increase proportionally with increase in axial length.[4] Thinned out chorioretinal tissue is associated with poor blood circulation and may lead to CNV development by inducing vascular endothelial growth factor (VEGF)

expression.Furthermore, scleral thinning may cause deformation of the posterior pole leading to staphyloma formation with a shorter radius of curvature.

Clinical Features



<u>Myopic conus/crescent</u> -Peripapillary scleral expansion leads to a sharply defined concentric area of depigmentation adjacent to the optic disc where the inner surface of sclera is visible. It occurs due to a disparity between the area of the sclera and the retinal pigment epithelium (RPE)-choriocapillaris complex and a premature termination of this complex ahead of the optic disc edge. It can either be a scleral crescent alone or choroidal crescent or both. Based on the extent, it can be a temporal conus, nasal conus, inferior conus, or annular conus. Temporal conus is reported to be the most frequent type.[4]

<u>Tessellation</u> -Generalized depigmentation due to RPE atrophy leads to a tigroid appearance of the fundus.

<u>Posterior Staphyloma (Scarpa's Staphyloma)</u> - It is an outward protrusion of all coats of the posterior pole and is considered pathognomonic . Spaide defined a posterior staphyloma as "outpouching of the wall of the eye that has a radius of curvature less than the surrounding radii of curvature. It is seen as a secondary depression with bending of vessels at the margin and a dark crescentic nasal reflex.

<u>Macular Chorioretinal Atrophy</u> - Chorioretinal atrophy occurs due to progressive thinning of the choroid, disappearance of choroidal vessels, and loss of RPE and photoreceptors. The cause behind atrophy is probably choroidal vascular occlusion and abiotrophic degeneration. Chorioretinal atrophy is of two types – diffuse atrophy and patchy atrophy.[7] Diffuse atrophy appears as yellowish-white areas of atrophy with ill-defined borders while patchy atrophy is grayish-white, well-defined area of atrophy, and produces an absolute scotoma on visual fields. Patchy atrophy predisposes to CNV development.

<u>Lacquer Cracks</u> - Lacquer cracks are breaks in the Bruch's membrane at the macula in highly myopic eyes, usually associated with a posterior staphyloma. These appear as multiple yellowish-white irregular lines, usually horizontally oriented, and coursing the posterior pole. Lacquer cracks can be linear or stellate, and sometimes show branching, crisscrossing, or both. These occur more commonly in males and decrease with aging.

Förster-Fuchs' Spot - It is a raised, pigmented, round, or elliptical lesion that is predominantly dark but can have a gray, yellow, red, or green hue. Forster-Fuchs' spots arise due to proliferation of RPE associated with choroidal hemorrhage. These are primarily small scars formed following degeneration and neovascularization

<u>Choroidal Neovascular Membrane</u> - Macular CNV is a one of the most common complications that results in reduced central vision . Myopic CNV develops in 10% of high myopes and 30% myopes eventually develop CNV in the other eye as well. It appears as a grayish subretinal membrane with hyperpigmented borders. As the retina is thin, bleeding does not usually obscure these lesions and they are easily visible on clinical examination.

<u>Macular Hole</u> - Macular hole formation tends to occur at a younger age as compared to idiopathic age-related macular holes. The degree of myopia and axial length has been shown to have an inverse correlation to the age of onset of the macular hole. Macular hole formation in myopic eyes may be related to the early onset of vitreous degeneration with development of tangential traction at the level of the premacular cortex.

II. Method And Material

This was a prospective observational study that involved 50 eyes of 25 patients with pathological myopia complaining of diminution of distant vision. Patients were recruited from the OPD of MLB MEDICAL college, Jhansi ,Uttar Pradesh and were followed from 1st november 2019 - 1st april 2020. It was performed under the Helsinki Declaration of 1975, as revised in 2000. The necessary permission from the Ethical and Research Committee was obtained for the study.

Inclusion criteria

1. All patients who presented to the OPD of MLB medical College Jhansi with the complaint of dimunition of distant vision and diagnosed case of pathological myopia were included.

Exclusion criteria

1.Patients with ocular systemic diseases(like hypertension, diabetes) that could affect the retina.

2.Patients with other retinal disorders

3.Patients with recent intraocular surgery

4.Patients with the history of trauma

5.Patients with dense cataract

5.Mentally or physically unfit patients

All patients were subjected to a detailed history taking, refraction using Topcon autorefractometer and best corrected visual acuity (VA) measurement. All patients had complete ophthalmic examination including biomicroscopic slit lamp examination, fundus examination with 90D lens and fundusphotography and optical coherence tomography.

Optical coherence tomography examination was done through dilated pupils, OCT examination was done through a dilated pupil using commercially available Cirrus HD-OCT Model 4000 - Carl Zeiss Meditec, Inc., Dublin, California, USA or Spectralis OCT Heidelberg Engineering.

III. Results

A total of 50 eyes of 25 patients were studied. We included eyes with complaint of diminution of vision. There were 11 males and 14 females and 60% of the studied eyes were the right eyes.

All eyes had one or more features typical of pathological myopia (like diffuse or patchy chorioretinal atrophy, posterior staphyloma, lacquer cracks, Fuchs spots, choroidal neovascular membrane)

Table1:Ophthalmoscopic findings in patients of pathological myopia

Features		Total %	
Peripapillary atrophy	81%		
Tilted disc	56%		
Staphyloma	22%		
Chorioretinal atrophy	18%		
Tigroid fundus	16%		
Lacquer crack	12%		
Retinal hemorrhage	4%		
Choroidal neovascularization	2%		
Fuchs spot	2%		

IV. Discussion

Pathological myopia is more prevalent in the Asian population than among other racial groups. In a recent study on school-going children in North India, the prevalence of HM was found to be 1.5%.[8] Population-based studies show a higher prevalence of PM in women than in men. The Blue Mountains Eye study[9] and Hisayama study[10] reported a prevalence of 0.4% and 2.2% in women, respectively, while in men, it was 0.06% and 1.2%, respectively. Refractive status of the parents plays an important role in the development of high myopia.[11] Twin studies report high heritability values of up to 90%.[12] Currently, the most studied environmental parameter thought to be protective against development of myopia is time spent by children outdoors. Chang L, Pan CW et al [13] in their study Myopia-related fundus changes in Singapore adults with high myopia concluded that staphyloma and chorioretinal atrophy lesions were the most common fundus findings among Asian adults with high myopia. In this population, tilted discs and peripapillary atrophy

were also common, while choroidal neovascularization and Fuchs spot were rare. In contrast with Singapore teenagers, in whom tilted disc and peripapillary atrophy were common while staphyloma and chorioretinal atrophy were rare, pathologic myopia appears to be dependent on the duration of disease and, thus, age of the individual. Moriyama M et al. studied the morphology and long-term changes of choroidal vascular structure in highly myopic eyes with and without posterior staphyloma[14]. Ohno-Matsui K et al in his study concluded that patchy atrophy and lacquer cracks predispose to the development of choroidal neovascularisation in patients of pathological myopia he also described the long-term pattern of progression of myopic maculopathy [15]. Ikuno Y et al described the ocular risk factors for choroidal neovascularization in pathologic myopia, steeper posterior staphyloma, and lacquer cracks are thought to be the risk factors for development of myopic CNV.[16] Myopic CNV progresses through three main stages. In the initial phase, there is direct damage to the photoreceptors, causing central visual loss. Then, as the CNV regresses, a fibrous pigmented scar forms, referred to as Forster–Fuch's spot. Finally, chorioretinal atrophy develops around the regressed CNV, which results in a poor long-term visual outcome.

V. Conclusion

Pathological myopia can affect the macula in various ways and can lead to a dramatic fall in visual acuity. Patients present with diminision of distant vision and fundus findings include peripapillary atrophy, staphyloma ,tigroid fundus,lacunar cracks,chorioretinal atrophy, choroidal neovascular membrane and other macular changes. These all are result of stretching and thinning of retina and choroid. Under-correction of myopia leads to increase in myopia progression and therefore optimal correction is necessary. Progressive or bifocal lenses may slow the progression of myopia by limiting ocular accommodation. Increased time spent outdoors is also a protective factor for progression of myopia. Optical coherence tomography and now OCTA aid in an early diagnosis and are important imaging tools required for a serial follow-up of such eyes to prevent complications related to pathological myopia include chorioretinal atrophy, foveoschisis, choroidal neovascularization, rhegmatogenous retinal detachment, cataract, and glaucoma.

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Dr. Jitendra Kumar, et. al. "Ophthalmoscopic Findings in Patients of Pathological Myopia." *IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)*, 19(7), 2020, pp. 14-17.

DOI: 10.9790/0853-1907081417