Definition of Pathologic Myopia (PM)

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Abstract

The definition of pathologic myopia had not been standardized for a long time, and pathologic myopia was often confused with high myopia. These two are distinctly different; "high myopia" is defined as an eye with a high degree of myopic refractive error, and "pathologic myopia" is defined as myopic eyes with the presence of pathologic lesions in the posterior fundus. The changes are the presence of myopic maculopathy equal to or more serious than diffuse chorioretinal atrophy and/or the presence of a posterior staphyloma.

Keywords

Pathologic myopia · High myopia · Posterior staphyloma
Diffuse atrophy · Axial length

1.1 Importance of Myopia and Pathologic Myopia

Myopia is defined as a refractive condition of the eye in which parallel rays of light entering the eye are brought to a focus in front of the retina when the ocular accommodation is relaxed [1]. This refractive status is dependent on the axial length, and a disproportionate increase of the axial length of the eye can lead to myopia, called axial myopia, or a disproportionate increase in the refractive power of the eye can also lead to myopia, called refractive myopia. The WHO Report defines myopia as "a condition in which the refractive error (spherical equivalent) is ≤ -0.50 diopter (D) in either eye" [2].

Myopia is a significant public health concern worldwide [2–4], and the rapid increase in the prevalence of myopia is of considerable concern to health care personnel and governments [5, 6]. It is estimated that by 2050, there will be 4.8 billion people with myopia which is approximately one-half (49.8%) of the world population. Of these, 938 million individuals will have high myopia which is 9.8% of the world population [5]. This is important because even mild myopia can be a risk factor for other ocular disorders [3, 7, 8]. However, it is uncertain whether the prevalence of pathologic myopia increases in parallel with an increase of myopia or high myopia.

Eyes with pathologic myopia have different types of lesions in the posterior fundus, called myopic maculopathy, which can lead to a significant reduction of central vision [9, 10]. In fact, myopic maculopathy in eyes with pathologic myopia is a major cause of blindness worldwide and especially in east Asian countries [11–15].

The definitions of myopia and pathologic myopia had not been standardized, and the term "pathologic myopia" was often confused with "high myopia." These two are distinctly different pathologies; "high myopia" is defined as an eye with a high degree of myopic refractive error, and "pathologic myopia" is defined as myopic eyes with the presence of pathologic lesions in the posterior fundus. Duke-Elder defined "pathologic myopia" although he used the term "degenerative myopia" as "the type of myopia which is accompanied by degenerative changes occurring especially in the posterior pole of the globe" [16].

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Curtin [17, 18] showed that the refractive error and axial length in eyes with the same type of staphylomas varied considerably and suggested that these measurements were unreliable indicators of pathologic myopia. He suggested that the morphology of the posterior staphyloma would be a much more reliable measure for diagnosing pathologic myopia.

1.2 Classification of Myopia According to Refractive Error (Spherical Equivalent) (Table 1.1)

Myopia is classified into low myopia, moderate myopia, and high myopia. The cut-off values for the different degrees have not been consistent among studies. The WHO Report defined "high myopia" as "a condition in which the objective refractive error (spherical equivalent) is ≤ −5.00 D in either eye" [3]. Very recently, Flitcroft on behalf of the International Myopia Institute (IMI) proposed a set of standards to define and classify myopia [1]. Low myopia is defined as a refractive error of ≤ −0.50 and > −6.00, and high myopia is defined as refractive error of ≤ −6.00 D [1]. The Japan Myopia Society proposed a category of "moderate myopia" between "low myopia" and "high myopia" (http://www.myopiasociety.jp/member/guideline/index.html). According to this society, low myopia is defined as a refractive error of ≤ −0.50 and > −3.00 D, moderate myopia is ≤ −3.00 and > −6.00 D, and high myopia is ≤ −6.00 D. A summary of the modified definition is presented in Table 1.1.

1.3 Classification of Pathologic Myopia (Table 1.1)

Pathologic myopia is classified as being present when myopic eyes have characteristic lesions in the posterior fundus. The changes are the presence of myopic maculopathy equal to or more serious than diffuse choroidal atrophy (equal to Category 2 in the META-PM classification [9]) and/or the presence of a posterior staphyloma [19]. The cut-off values of the myopic refractive error and axial length are not set for the definition of pathologic myopia because a posterior staphyloma has been reported to occur even in eyes with normal axial length (Fig. 1.1) [20] or in eyes with axial lengths <26.5 mm [21]. This suggested that pathologic myopia is considered an independent pathology of the axial elongation of the eye. Although an axial elongation mainly occurs in the equatorial region of the eye, pathologic myopia is characterized by a formation of posterior staphyloma as suggested by Spaide [22]. Thus, the mainly affected area is different between high myopia and pathologic myopia.

Optical coherence tomographic evaluations showed that the progressive choroidal thinning and a formation of Bruch's membrane defects in the macular region were key phenomena associated with myopic maculopathy, the lesions of myopic maculopathy are better classified by their appearance in the OCT images (see Chap. 4 for OCT-based classification) [23].

Table 1.1 Summary of definitions of various types of myopia (modified from Flitcroft et al. [1])

Term	Definition
Myopia	A condition in which the spherical equivalent refractive error of an eye is ≤ −0.50 D when ocular accommodation is relaxed.
Low myopia	A condition in which the spherical equivalent refractive error of an eye is ≤ −0.50 D and > −3.00 D when ocular accommodation is relaxed.
Moderate myopia	A condition in which the spherical equivalent refractive error of an eye is ≤ −3.000 D and > −6.00 D when ocular accommodation is relaxed.
High myopia	A condition in which the spherical equivalent refractive error of an eye is ≤ −6.000 D when ocular accommodation is relaxed.
Pathologic myopia	Myopia that accompanies characteristic myopic fundus changes (the presence of myopic maculopathy equal to or more serious than diffuse choroidal atrophy or the presence of posterior staphyloma)

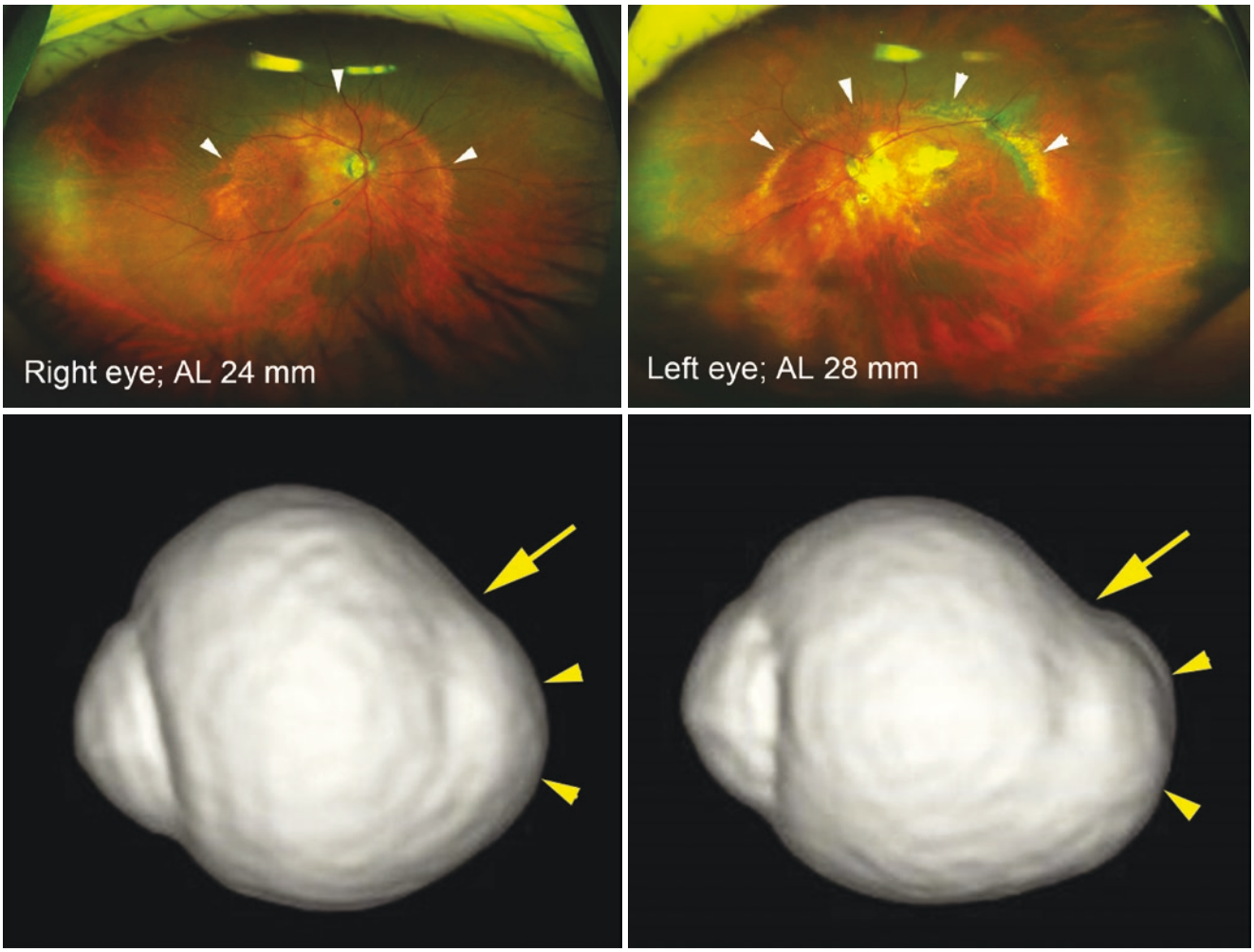

Fig. 1.1 Posterior staphyloma seen in an emmetropic fellow eye of the patient with unilateral high myopia (modified with permission from [20]). Top Row: Wide-field fundus imaging shows upper margin of wide macular staphyloma (arrowheads) both in the highly myopic left eye (axial length; 28 mm) as well as non-myopic right eye (axial length; 24 mm). Please note that the posterior fundus of the right eye is almost normal and the staphyloma edge is outside the conventional 50 degree fundus photo. Bottom Row: Three-dimensional magnetic resonance imaging (3D MRI) of both eyes. Left eye (right image) shows a clear posterior staphyloma as posterior outpouching (arrowheads). The upper margin of staphyloma is seen (arrow). The right eye also shows a similar staphyloma with the upper edge of staphyloma, although the degree of staphyloma is milder

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