Long-term Pattern of Progression of Myopic Maculopathy

A Natural History Study

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Objective: To investigate the long-term progression pattern of myopic maculopathy and to determine the visual prognosis of each progression stage.

Design: Retrospective, observational case series.

Participants: The medical records of 806 eyes of 429 consecutive patients with high myopia (refractive error more than −8.00 diopters [D] or axial length ≥26.5 mm) who were followed for 5–32 years were reviewed.

Methods: Participants had complete ophthalmological examinations including best-corrected visual acuity, axial length measurements, fluorescein angiography, and color fundus photography, at least once a year. The presence and type of posterior staphyloma was determined by binocular stereoscopic ophthalmoscopy. The types of myopic maculopathy included tessellated fundus, lacquer cracks, diffuse chorioretinal atrophy, patchy chorioretinal atrophy, choroidal neovascularization (CNV), and macular atrophy. None of the patients had received any type of treatment for the maculopathy.

Main Outcome Measures: The longitudinal long-term progression pattern and the visual prognosis of each type of fundus lesion.

Results: During the mean follow-up of 12.7 years, 327 of the 806 highly myopic eyes (40.6%) showed a progression of the myopic maculopathy. The most commonly observed patterns were from tessellated fundus to the development of diffuse atrophy and lacquer cracks, an increase in the width and progression to patchy atrophy in eyes with lacquer cracks, an enlargement of the diffuse atrophy, and the development of patchy atrophy in eyes with diffuse atrophy, and an enlargement and fusion of patches of atrophic areas in eyes with patchy atrophy. Eyes with tessellated fundus, lacquer cracks, diffuse atrophy and patchy atrophy at the initial examination progressed to the development of CNV. Eyes with CNV developed macular atrophy. The fusion of patchy atrophy, the development of CNV, and macular atrophy all led to significant visual decreases. A posterior staphyloma was observed more frequently in eyes that showed progression from tessellated fundus, diffuse atrophy, and patchy atrophy than those without a progression.

Conclusions: These findings indicate that myopic maculopathy tends to progress in approximately 40% of highly myopic eyes, and the pattern of progression affects the visual prognosis. Preventive therapy targeting posterior staphyloma should be considered to prevent the visual impairment caused by the progression of myopic maculopathy.

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Pathologic myopia is a leading cause of visual impairment, and is the fourth to ninth most frequent cause of blindness worldwide.[1–5] In Asian countries, pathologic myopia is the most frequent cause of visual impairment; the Tajimi Study in Japan showed that myopic macular degeneration is the leading cause of unilateral or bilateral blindness,[6] whereas in China,[7] it is the second most common cause of low vision/blindness in participants who were >40 years of age according to the World Health Organization.

Visual impairment in eyes with pathologic myopia is mainly due to the development of different types of myopic maculopathies. In highly myopic eyes, the axial elongation of the eye and the development of a posterior staphyloma result in a thinning of the retina and choroid, which then lead to the development of different types of myopic maculopathy. The impact of myopic maculopathy on visual impairment is important because the maculopathy is often bilateral, irreversible, and frequently affects individuals during their most productive years.[8]

Despite the clinical importance of myopic maculopathy, the long-term visual prognosis of each type of lesion associated with myopic maculopathy has not been determined.

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The High Myopia Clinic of the Tokyo Medical and Dental University has been in existence for 38 years, and during this time, >7000 patients with high myopia have been examined and followed. This High Myopia Clinic has allowed us to follow individual patients for long periods, and thus permits us to determine the natural course of pathologic myopia. Because we have noted that the progression of myopic maculopathy has a pattern, we believe that it is important to determine which factors influence the progression of a specific lesion and to identify which type of lesion is associated with poor vision.

A PubMed search identified only 2 articles describing the long-term progression of myopic retinopathy.[9,10] Vongphanit et al[9] reported that signs of myopic retinopathy were found in 67 eyes of 44 individuals (1.2%) among the 3583 participants of the Blue Mountains Eye Study, and a significant progression of myopic retinopathy was observed in 17.4% of these 67 eyes after a mean interval of 61 months. However, the results were limited because only a small number of patients with pathologic myopia (n = 44) were studied, and only 18 of the 44 patients had myopic retinopathy.

In the other study, Shih et al[10] reviewed the medical records of 552 highly myopic patients who were ≥40 years of age. They reported that a significant decrease of vision was found more frequently in the patients with maculopathy than those without maculopathy after 10 years using the

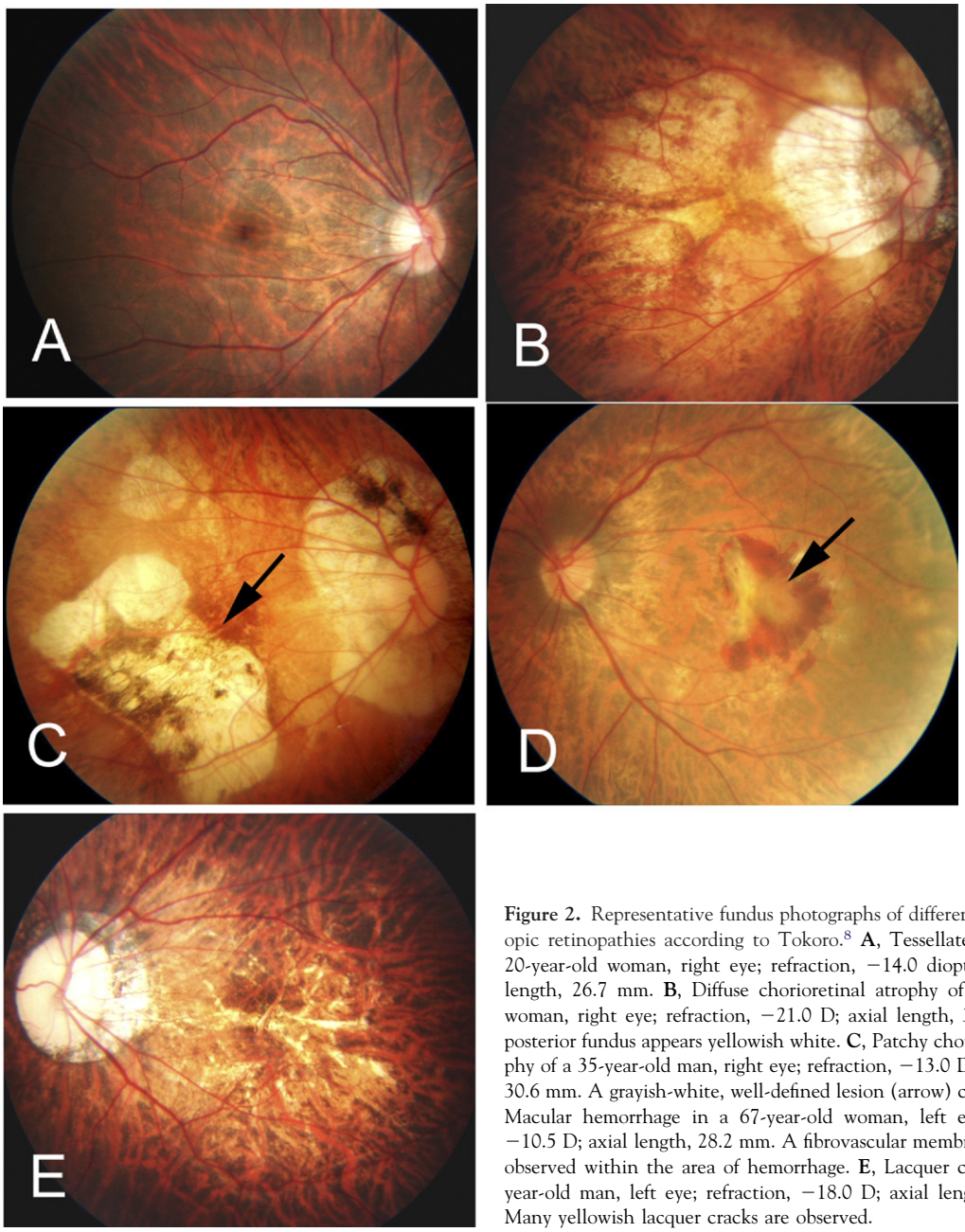

Figure 2. Representative fundus photographs of different types of myopic retinopathies according to Tokoro.[8] **A**, Tessellated fundus of a 20-year-old woman, right eye; refraction, −14.0 diopters (D); axial length, 26.7 mm. **B**, Diffuse chorioretinal atrophy of a 51-year-old woman, right eye; refraction, −21.0 D; axial length, 31.6 mm. The posterior fundus appears yellowish white. **C**, Patchy chorioretinal atrophy of a 35-year-old man, right eye; refraction, −13.0 D; axial length, 30.6 mm. A grayish-white, well-defined lesion (arrow) can be seen. **D**, Macular hemorrhage in a 67-year-old woman, left eye; refraction, −10.5 D; axial length, 28.2 mm. A fibrovascular membrane (arrow) is observed within the area of hemorrhage. **E**, Lacquer cracks in a 28-year-old man, left eye; refraction, −18.0 D; axial length, 31.0 mm. Many yellowish lacquer cracks are observed.

classification of Avila et al.[11] However, Shih et al did not focus on the progression of each type of lesion associated with the myopic maculopathy.

There are other studies on the progression of myopic maculopathy, but the studies focused on only 2 of the major lesions of myopic maculopathy, namely, myopic choroidal neovascularization (CNV)[11–16] and lacquer cracks.[17,18]

The classification of myopic maculopathy by Avila et al[11] made 25 years ago has been used most frequently in earlier studies. They graded myopic retinopathy on a scale of increasing severity from 0 to 5 as follows: M_0, normal-appearing posterior pole; M_1, choroidal pallor and tessellation (defined as the condition in which the choroidal vessels can be seen through the retina owing to reduced pigmentation or hypoplasia of the retinal pigmented epithelium[8]); M_2, choroidal pallor and tessellation with posterior staphyloma; M_3, choroidal pallor and tessellation with posterior staphyloma and lacquer cracks; M_4, choroidal pallor and tessellation with lacquer cracks, posterior staphyloma, and focal areas of deep choroidal atrophy; and M_5, posterior pole with large geographic areas of deep chorioretinal atrophy and "bare" sclera.

There are some problems with this classification because it was not based on the actual progression pattern, and posterior staphyloma was included in the classification. From our longitudinal observations of a large number of highly myopic eyes, we have evidence that a posterior staphyloma is the cause of the development of myopic maculopathy and not the reverse. In addition, eyes with lacquer cracks were placed into a relatively advanced group (M_3). However, lacquer cracks often develop at the early stage of myopic maculopathy, and they are often observed in young individuals without an obvious staphyloma or early atrophic changes of the retina.[8] We believe that these problems were from a lack of evaluating the long-term longitudinal progression of each type of fundus lesions associated with myopic maculopathy.

Thus, the purpose of this study was to determine the long-term progression pattern of each type of fundus lesion found in eyes with myopic maculopathy and to clarify the impact of each progression pattern on the visual prognosis. We shall show that the different maculopathies progressed during a follow-up period of ≥5 years, and some of the specific progression patterns led to a significant visual decrease. A posterior staphyloma was found to be an important factor that led to the progression of myopic maculopathy.

Patients and Methods

Approval was obtained from the Ethics Committee of the Tokyo Medical and Dental University to perform this retrospective study. The procedures used during the examinations conformed to the tenets of the Declaration of Helsinki, and patients had signed an informed consent form for fluorescein angiography. The medical records of consecutive patients with pathologic myopia (myopic refraction > 8 diopters [D] or axial length ≥26.5 mm), and a minimum follow-up of 5 years and were examined in the High Myopia Clinic at Tokyo Medical and Dental University were

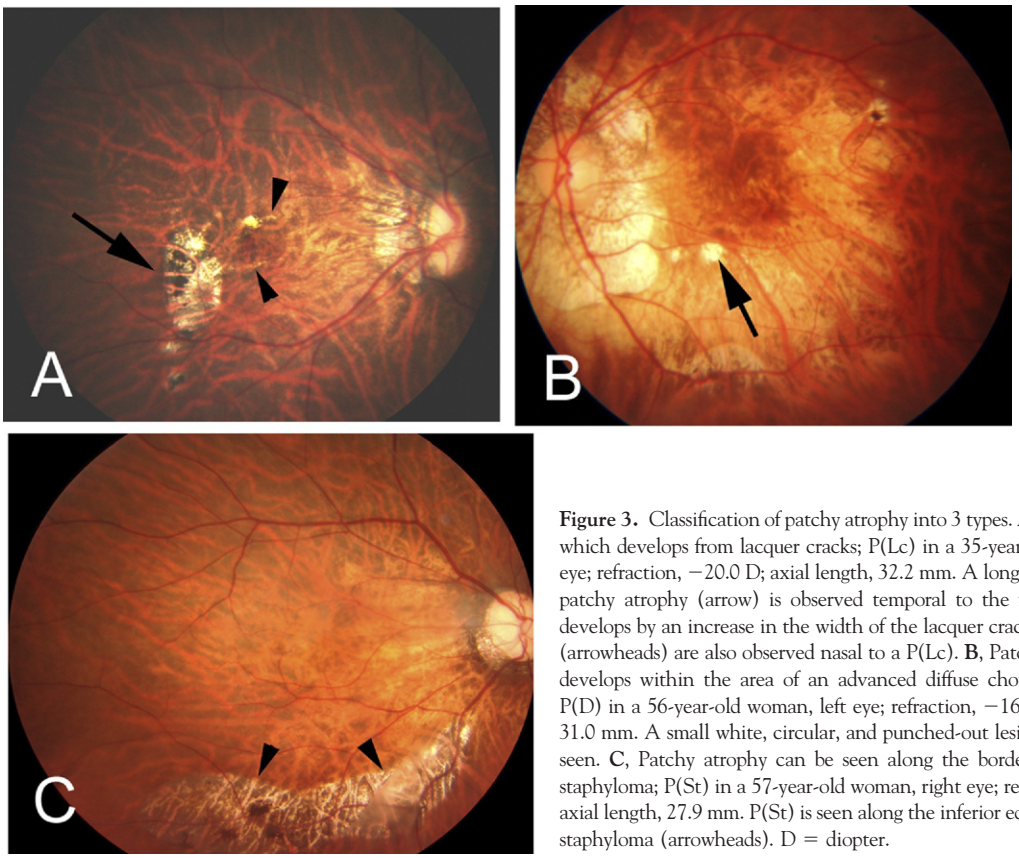

Figure 3. Classification of patchy atrophy into 3 types. **A**, Patchy atrophy, which develops from lacquer cracks; P(Lc) in a 35-year-old woman, right eye; refraction, −20.0 D; axial length, 32.2 mm. A longitudinally oriented patchy atrophy (arrow) is observed temporal to the fovea. This lesion develops by an increase in the width of the lacquer cracks. Lacquer cracks (arrowheads) are also observed nasal to a P(Lc). **B**, Patchy atrophy which develops within the area of an advanced diffuse chorioretinal atrophy; P(D) in a 56-year-old woman, left eye; refraction, −16.5 D; axial length, 31.0 mm. A small white, circular, and punched-out lesion (arrow) can be seen. **C**, Patchy atrophy can be seen along the border of the posterior staphyloma; P(St) in a 57-year-old woman, right eye; refraction, −12.0 D; axial length, 27.9 mm. P(St) is seen along the inferior edge of the posterior staphyloma (arrowheads). D = diopter.

retrospectively analyzed. When the patients were <5 years old, pathologic myopia was defined as a refractive error of more than −4.0 D (spherical equivalent), and when the patients were between the ages of 6 and 8 years, pathologic myopia was defined as a refractive error above −6.0 D according to the definitions by Tokoro.[19] All patients had 1 of the myopic maculopathy lesion at the initial examination.

The exclusion criteria included a history of other ocular disorders, such as dense cataract, glaucoma, diabetic retinopathy or other retinal vascular diseases, age-related macular degeneration, or a history of vitreoretinal surgery, which might affect the visual acuity. Because the purpose of the study was to determine the natural course of myopic retinopathy, patients who received any treatments for the myopic fundus lesions, such as laser photocoagulation, surgical treatment, photodynamic therapy, or intravitreous injection of bevacizumab for myopic CNV, were excluded. We did not perform laser photocoagulation or surgical treatment on any of the patients with myopic CNV because earlier studies had reported that these treatments were not effective.[16,20–23] Also, because photodynamic therapy was approved by the Ethics Committee of our University only in September 2004, none of the patients with >5 years of follow-up underwent photodynamic therapy or more recent treatments, including intravitreal injection of bevacizumab. For the analysis of the change of visual acuity during the follow-up period, patients who underwent cataract surgery during the follow-up period were also excluded.

All of the patients received a complete ophthalmological examination including best-corrected visual acuity (BCVA) using a Landolt C chart, measurement of axial length, dilated fundus examination by indirect ophthalmoscopy, slit-lamp examination of the posterior fundus using a +78-D lens, fluorescein angiography, and color fundus photography at least once a year. To quantify the differences in the BCVA, all of the BCVAs were converted to the logarithm of the minimum angle of resolution units. The axial length was measured by A-scan ultrasonography (Ultrascan, Alcon, Fort Worth, TX) ≥5 times for each eye, and the average value was used for the analysis. The presence and type of the posterior staphyloma were determined by binocular stereoscopic ophthalmoscopy. The staphylomas were classified into 10 types: types I–V were primary staphylomas, and types VI through X were compound staphylomas (Fig 1; available online at http://aaojournal.org).

The myopic maculopathy was initially classified according to the definition by Tokoro[8]: tessellated fundus (Fig 2A), diffuse chorioretinal atrophy (Fig 2B), patchy chorioretinal atrophy (Fig 2C), and macular hemorrhage (Fig 2D). However, we modified the classification based on our clinical impressions and the results of our earlier studies.[18,24] Although lacquer cracks were originally included in the category of diffuse atrophy in Tokoro's classification, we have classified lacquer cracks as an independent lesion (Fig 2E) because we have found that lacquer cracks progressed to different types of lesions and not only to diffuse atrophy.[18] Although macular hemorrhage was subclassified into 2 types of lesions—myopic CNV and simple macular hemorrhage according to the definition by Tokoro[8]—we focused on CNV alone in this study, because a simple hemorrhage can be caused by the formation of new lacquer cracks and be spontaneously resolved.[24] Thus, we classified eyes with a simple hemorrhage as having lacquer cracks lesion.

We also subdivided patchy atrophy into 3 subtypes. One type progressed by an increase in the width of the lacquer cracks.[18] This type of patchy atrophy had a horizontal or vertical orientation and usually developed near the fovea (Fig 3A). We named this type of lesion patchy atrophy P(Lc). The second type of patchy atrophy developed as a circular lesion in eyes with advanced diffuse chorioretinal atrophy (Fig 3B). We named this type of lesion patchy atrophy P(D). The last type of patchy atrophy developed along the edge of posterior staphyloma (Fig 3C), and we named this type of lesion patchy atrophy P(St).

The classification of the maculopathies and the determination of whether they had progressed were made by 3 of the authors (KH, KOM, NS) independently. For the patients who had several different progression patterns, we examined the subsequent progression patterns in addition to the first progression pattern to the initial fundus lesion. In cases of a disagreement, the fundus photographs were examined jointly, and an agreement was reached in the classification among the 3 authors in all of the patients.

Statistical Analyses

The comparison of patient's age, refractive error, and axial length for the different types of myopic maculopathy or for different progression patterns was made using Mann-Whitney *U* test. The comparison of the frequency and the type of posterior staphyloma was made by chi-square or Fisher's exact probability tests. The changes in the initial and final BCVA in each progression pattern were analyzed using paired *t* tests or Wilcoxon signed-ranks tests. $P<0.05$ was considered significant.

Results

Characteristics of Patients and Eye at Initial Examination

The medical records of 2710 eyes of 1355 patients who had been examined in the High Myopia Clinic at Tokyo Medical and Dental University were reviewed. From these 2710 eyes, 1904 eyes were excluded: 1568 eyes because their follow-up period was <5.0 years; 198 eyes because they had less than −8.0 D of myopia or axial length <26.5 mm, or retinal pathology; 86 eyes because they had less than −8.0 unilateral myopia; and 52 eyes for other retinal pathologies. The retinal pathologies included retinitis pigmentosa, retinal detachments, amblyopia, and phthisis bulbi. In the end, we

Table 3. Patient and Study Eye Characteristics at Initial Examination

No. patients (eyes)	429 (806)
Gender	
Male	147 (278)
Female	282 (528)
Age (yrs), mean (SD) (range)	41.1±16.7 (4 to 74)
Refractive error (D), mean (SD) (range)	−13.4±4.9 (−5.0 to −36.0)
Axial length (mm), mean (SD) (range)	28.7±1.9 (25.0 to 35.4)
Baseline logMAR, mean (SD) (range)	0.26±0.46 (−0.18 to 2.00)
Posterior staphyloma	530 (65.8%)
Type of posterior staphyloma	
I	141 (26.6%)
II	274 (51.7%)
IX	93 (17.5%)
Other	22 (4.2%)
Fundus lesion at the initial examination	
Tesselated fundus only	276 (34.2%)
Lacquer cracks	117 (14.5%)
Diffuse chorioretinal atrophy	491 (60.9%)
Patchy chorioretinal atrophy	163 (20.2%)
CNV	91 (11.3%)
Macular atrophy	45 (5.6%)
Follow-up period (years), mean (SD) (range)	12.7±6.2 (5.0 to 32.1)

CNV = choroidal neovascularization; D = diopters; logMAR = logarithm of minimal angle of resolution; SD = standard deviation.

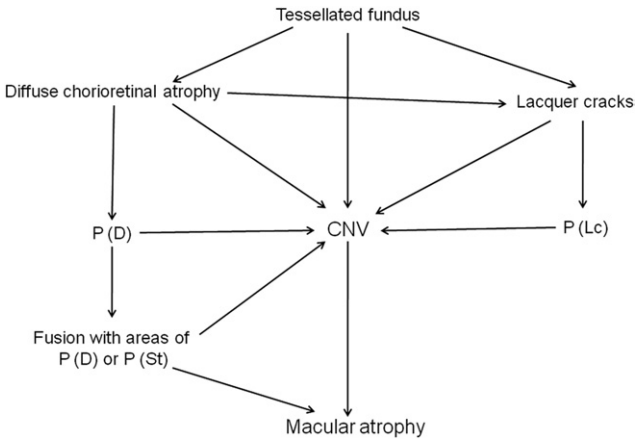

Figure 4. A scheme showing a progressive pattern of various lesions of myopic maculopathy. CNV = choroidal neovascular membrane; P(D) = patchy atrophy which develops in the posterior fundus accompanying diffuse atrophy; P(Lc) = patchy atrophy which develops from lacquer cracks; P(St) = patchy atrophy which develops along the edge of posterior staphyloma.

investigated 806 eyes of 429 patients. It should be noted that in some of the patients, only 1 eye met the inclusion criteria.

The characteristics of the 1568 eyes that were excluded because their follow-up period was <5.0 years are shown in Table 1 (available online at http://aaojournal.org). A statistical comparison of the clinical characteristics of the 827 patients who were excluded to that of the 429 patients included in this study showed that the differences in the baseline characteristics were not significant (Table 2, available online at http://aaojournal.org).

The clinical characteristics of the 806 highly myopic eyes of the 429 patients are summarized in Table 3. In the end, there were 147 men and 282 women with a mean age of 41.1±16.7 years (range, 4–74) at the time of the initial examination. The mean refractive error was −13.4±4.9 D, and the mean axial length was 28.7±1.9 mm. It is important to note that there were 61 eyes whose myopic refractive error met the criterion of −8.0 D or greater but whose axial lengths were <26.5 mm, that is, they had refractive myopia. There were also 67 eyes whose axial length met the criterion of >26.5 mm but whose refractive error was less than −8.0 D, that is, they had axial myopia. The mean follow-up period was 12.7±6.2 years.

A posterior staphyloma was identified by binocular stereoscopic fundus examinations in 530 of the 806 eyes (65.8%). A type II staphyloma was the most common type (51.7%) of staphyloma followed by the type I (26.6%) and by type IX (17.5%). At the initial examination, a diffuse chorioretinal atrophy was the most frequently observed maculopathy lesion (60.9%).

Tentative Progression Pattern

From our extensive longitudinal examinations of the 806 eyes that met our inclusion criteria and from the evidence we shall present, we have arrived at a tentative progression pattern of myopic maculopathy (Fig 4). We present this diagram at this point to make it easier for the reader to follow the changes we describe and to allow the reader to determine whether this tentative progression pattern conforms to the findings.

We suggest that the first sign that a highly myopic eye had progressed to the myopic maculopathy stage was the appearance of a tessellated fundus. With increasing time, eyes with a tessellated fundus can progress to diffuse atrophy or lacquer cracks, or more rarely, directly to the formation of a CNV. Eyes with diffuse atrophy can progress to the P(D) type of patchy atrophy or to lacquer cracks or to the formation of a CNV. Eyes with lacquer cracks can progress to the P(Lc) type of patch atrophy or to the formation of a CNV. The atrophic patches in eyes with P(D) can fuse to form larger areas of atrophy, which can extend into the macular area, that is, macular atrophy. Eyes with a CNV can also progress to macular atrophy.

Our observations also showed that the timing of the progression is not uniform. The majority of the eyes with pathologic myopia pass to the tessellated fundus stage, where they remain for a relatively long time. At around the age of 40 years, a posterior staphyloma develops and the tessellated fundus progresses to the next stages, namely, diffuse atrophy and lacquer cracks. The progression of the maculopathy then proceeds relatively quickly because the staphyloma facilitates the progression.

Characteristics of Eyes at Initial Examination

The clinical characteristics of eyes with the different types of myopic maculopathy lesions at the initial examination are shown in Table 4 (available online at http://aaojournal.org). At the initial examination, 533 eyes of 237 patients had 1 type of lesion and 273 eyes of 192 patients had ≥2 different types of lesion in the same eye. The patients with a tessellated fundus were significantly younger than the patients in the other groups ($P<0.0001$). Patients

Table 5. First Progression Pattern and Clinical Characteristics in Eyes with Tesselated Fundus

Progression Type	Frequency, No. of Eyes (%)	Age (yrs)	Axial Length (mm)	Posterior Staphyloma, No. of Eyes (%)	Type of Staphyloma	Follow-up Period (yrs)
With progression (37 eyes)						
Diffuse atrophy	28 (10.1)	37.6±17.7	27.5±1.4	22 (78.6)	I: 4 eyes (18.2%) II: 14 eyes (59.1%) IX: 1 eyes (4.5%)	17.0±5.9
Lacquer cracks	8 (2.9)	39.6±14.8	28.5±1.4	7 (87.5)	I: 1 eye (14.3%) II: 5 eyes (71.4%) IX: 1 eye (14.3%)	13.7±4.7
CNV	1 (0.4)	29.0±0.0	27.1±0.0	1 (100.0)	II: 1 eyes (100.0%)	16.6±0.0
Without progression (239 eyes)						
No progression	239 (86.6)	30.9±15.6	27.4±1.2	47 (19.7)	I: 8 eyes (17.0%) II: 37 eyes (78.7%) IX: 2 eyes (4.3%)	12.0±6.3

CNV = choroidal neovascularization.

Table 6. Clinical Characteristics of Patients Who Showed Further Progression

Case No.	Age at Initial Visit (yrs)	Type of 1st Progression	At 1st Progression: Age (yrs)	At 1st Progression: Axial Length (mm)	At 1st Progression: Presence of Posterior Staphyloma	At 1st Progression: Type of Posterior Staphyloma	Type of 2nd Progression	At 2nd Progression: Age (yrs)	At 2nd Progression: Axial Length (mm)	At 2nd Progression: Presence of Posterior Staphyloma	At 2nd Progression: Type of Posterior Staphyloma
1	29	Lacquer cracks	41	27.3	+	II	P(Lc)	50	28.6	+	II
2	58	Lacquer cracks	67	25.7	+	II	P(Lc)	72	28.6	+	II
3	29	Lacquer cracks	37	29.0	+	II	P(Lc)	41	30.8	+	II
4	31	Lacquer cracks	42	28.5	+	II	P(Lc)	45	30.5	+	II
5	37	Lacquer cracks	43	29.9	+	II	P(Lc)	44	29.8	+	II
6	54	Lacquer cracks	57	28.8	+	I	P(Lc)	58	28.8	+	I
7	38	Lacquer cracks	41	26.5	+	IX	CNV	49	26.5	+	IX
8	44	Diffuse atrophy	57	27.9	+	II	P(D)	64	29.8	+	II
9	29	CNV	36	27.1	+	II	Macular atrophy	42	28.3	+	II

CNV = choroidal neovascularization; N/D = not determined because the enlargement of macular atrophy gradually occurs; P(D) = patchy atrophy

with lacquer cracks were significantly younger than those with a CNV ($P<0.00013$), and patients with diffuse atrophy were significantly younger than those with patchy atrophy ($P = 0.00016$). Patients with macular atrophy were significantly older than those with diffuse atrophy and those with lacquer cracks ($P = 0.00026$ and $P<0.0001$, respectively). The average age was least in the patients with tessellated fundus and was greatest in the patients with macular atrophy.

The eyes with a tessellated fundus were significantly less myopic than the eyes in any of the other groups ($P<0.0001$). There were no significant differences in the myopic refraction among any of the other groups. The eyes with a tessellated fundus had significantly shorter axial lengths than any of the other groups ($P<0.0001$). The eyes with patchy atrophy had significantly longer axial lengths than those with diffuse atrophy ($P = 0.0011$), those with lacquer cracks ($P = 0.0041$), and those with a CNV ($P<0.0001$). The eyes with diffuse atrophy had significantly longer axial lengths than those with a CNV ($P = 0.014$). The eyes with macular atrophy had significantly longer axial length than those with CNV ($P = 0.0004$), those with lacquer cracks ($P = 0.0036$), and those with diffuse atrophy ($P = 0.0129$). The average axial length was longest in the eyes with macular atrophy.

A posterior staphyloma was present in 27.2% of the eyes with a tessellated fundus, and in >80% of the eyes in the other groups. This difference was significant ($P<0.0001$).

Best-corrected Visual Acuity in Eyes with Different Myopic Maculopathies

To compare the BCVA among the eyes with the different types of myopic maculopathies, the eyes with a CNV or macular atrophy were excluded from the groups with diffuse chorioretinal atrophy, lacquer cracks, and patchy atrophy because the accompanying CNV or macular atrophy affected the vision noticeably, which made it difficult to evaluate the influence of the lesions on the BCVA. As a result, 372 eyes with diffuse atrophy, 75 eyes with lacquer cracks, and 74 eyes with patchy atrophy were analyzed (shown by asterisks in Table 2).

Statistical analyses showed that the eyes with a tessellated fundus had significantly better BCVA than that of any other groups ($P<0.0001$). On the other hand, eyes with macular atrophy had significantly worse BCVA than any of the other groups ($P<0.0001$). The eyes with a CNV had significantly worse BCVA than those with a tessellated fundus, diffuse atrophy, lacquer cracks, and patchy atrophy ($P<0.0001$ for all). Also, the eyes with patchy atrophy had significantly worse BCVA than those with diffuse atrophy ($P = 0.021$) and lacquer cracks ($P = 0.022$).

Progression Pattern of Myopic Maculopathy

During the mean follow-up period of 12.7 years (range, 5–32.1), 327 of the 806 eyes (40.6%) showed a progression of the myopic maculopathy. Because an accompanying CNV or macular atrophy could prevent the evaluation of the progression of the other types of lesion, the eyes with a CNV or macular atrophy were excluded in the determination of the progression pattern of the other maculopathies. As a result, the progression pattern was examined in 770 eyes: 276 eyes that had a tessellated fundus, 75 eyes that had lacquer cracks, 372 eyes that had diffuse atrophy, 74 eyes that had patchy atrophy, and 91 eyes that had a CNV. In this population, progression was observed in 37 of 276 eyes (13.4%) with a tessellated fundus, in 52 of 75 eyes (69.3%) with lacquer cracks, in 183 of 372 eyes (49.2%) with diffuse chorioretinal atrophy, in 52 of 74 eyes (70.3%) with patchy chorioretinal atrophy, and in 82 of 91 eyes (90.1%) with a CNV at the initial examination.

The progression pattern and the clinical characteristics of the fundus lesions at the initial examination are described in details below and also shown in Tables 5–12.

Eyes with a tessellated fundus at the initial examination. In the 276 eyes with a tessellated fundus at the initial examination, 28 eyes (10.1%) developed diffuse chorioretinal atrophy, 8 eyes (2.9%) developed lacquer cracks, and 1 eye (0.4%) developed a CNV (Table 5). The other 239 did not progress. The average age of the patients that had a progression was 38.3 ± 16.2 years, and that for eyes that did not show a progression was 30.9 ± 15.6 years ($P = 0.0059$).

Because there was only 1 eye that developed a CNV, this progression pattern was not included in subsequent statistical analyses. The patients who developed diffuse chorioretinal atrophy were significantly older than those without progression ($P = 0.021$). The age of the patients who developed lacquer cracks were not different from the ages of those who did not develop lacquer cracks. The axial lengths of eyes that had a progression were not different from the eyes without a progression. A posterior staphyloma was detected significantly more frequently in eyes that showed a progression than those without a progression ($P<0.0001$ for all).

A type II staphyloma was the predominant type of staphyloma in eyes with a tessellated fundus regardless of the progression pattern, and the incidence of the type of posterior staphyloma was not different between eyes that showed a progression and eyes without a progression. The mean follow-up period was signifi-

Pattern after the First Progression from Tessellated Fundus

Period from 1st to 2nd Progression (yrs)	Type of 3rd Progression	At 3rd Progression				Period from 2nd to 3rd Progression (yrs)	Age at Final Visit (yrs)	Total Follow-up (yrs)
		Age (yrs)	*Axial Length (mm)*	*Presence of Posterior Staphyloma*	*Type of Posterior Staphyloma*			
9	—						46	16.6
5	—						76	17.8
4.5	—						43	14.2
3	—						49	17.7
1	CNV	45	29.8	+	II	1.5	48	10.5
1	CNV	61	28.9	+	I	3	64	9.8
8	Macular atrophy	52	26.6	+	IX	3	54	15.8
7.5	—						69	25.2
6	—						46	16.6

originated from diffuse atrophy; P(Lc) = patchy atrophy originated from lacquer cracks.

cantly longer for patients who progressed to diffuse chorioretinal atrophy than those without progression ($P = 0.00013$). Otherwise, there was no difference in the follow-up period until the first detection of progression among the groups.

There were 9 eyes of 9 patients whose maculopathy progressed to a second pattern and to a third pattern after the initial progression during our follow-up period (Tables 5 and 6). These 9 eyes already had a posterior staphyloma at the initial examination. The incidence of the second and third progression was higher in eyes that developed lacquer cracks as the first progression from the tessellated fundus. In fact, 7 of 8 eyes that developed lacquer cracks as the first progression showed further progression. Six of these 7 eyes showed a progression to P(Lc) and 2 of these 6 eyes later developed a CNV at the edge of the fovea of P(Lc) (cases 5 and 6 in Table 6). The other eye that developed lacquer cracks as the first progression developed a CNV directly from lacquer cracks and finally progressed to macular atrophy (case 7; Fig 5, available online at http://aaojournal.org). One patient (case 8 in Table 6) developed diffuse atrophy as the first progression and later developed the P(D) type of patchy atrophy. The remaining patient (case 9) developed a CNV as the first progression, then progressed to macular atrophy, which then enlarged.

Eyes with lacquer cracks at initial examination. In the 75 eyes that had lacquer cracks at the initial visit, 32 eyes (42.7%) showed an increase in the width of the cracks and these eyes progressed to the P(Lc) type of patchy atrophy, 10 eyes (13.3%) developed a CNV, 10 eyes (13.3%) had an increase in number of lacquer cracks, and 23 eyes (30.7%) did not progress (Table 7). In the eyes that had lacquer cracks at the initial examination, the ages and axial lengths of the patients that progressed were not significantly different from those without a progression. All of the eyes that progressed to the P(Lc) type of patchy atrophy or developed a CNV had a posterior staphyloma. A posterior staphyloma was observed significantly more frequently in eyes that had progressed to P(Lc) than the eyes without a progression ($P = 0.003$). The incidence of posterior staphyloma in eyes that showed an increase in number of lacquer cracks was not different from the eyes without a progression. Although the incidence of type IX staphyloma in the eyes that progressed tended to be higher than those without a progression, the difference was not significant, probably because only a relatively small number of patients had this type of staphyloma. The mean follow-up period was significantly longer in the patients who progressed to P(Lc) than those without progres-

Table 7. First Progression Pattern and Clinical Characteristics in Eyes with Lacquer Cracks

Progression Type	Frequency, No. of Eyes (%)	Age (yrs)	Axial Length (mm)	Posterior Staphyloma, No. of Eyes (%)	Type of Staphyloma	Follow-up (yrs)
With progression (52 eyes)						
P(Lc)	32 (42.7)	40.4±13.1	29.8±1.5	32 (100)	I: 7 eyes (21.9%) II: 16 eyes (50.0%) IX: 7 eyes (21.9%) Others: 2 eyes (6.3%)	13.9±6.6
CNV	10 (13.3)	50.1±12.2	28.9±1.2	10 (100)	I: 4 eyes (40.0%) II: 3 eyes (30.0%) IX: 3 eyes (30.0%)	12.3±6.3
Increase in number	10 (13.3)	36.0±14.4	30.2±1.0	9 (90.0)	I: 2 eyes (22.2%) II: 6 eyes (66.6%) IX: 1 eye (11.1%)	8.9±3.1
Without progression (23 eyes)	23 (30.7)	44.9±14.6	30.1±1.6	17 (73.9)	I: 4 eyes (23.5%) II: 12 eyes (70.6%) IX: 1 eye (5.9%)	9.0±4.0

CNV = choroidal neovascularization; P(Lc) = patchy atrophy originated from lacquer cracks.

Table 8. Second, Third, and Fourth Progression Pattern from

Case No.	Age at Initial Visit (yrs)	Type of 1st Progression	At 1st Progression				Type of 2nd Progression	At 2nd Progression			
			Age (yrs)	*Axial Length (mm)*	*Presence of Posterior Staphyloma*	*Type of Posterior Staphyloma*		*Age (yrs)*	*Axial Length (mm)*	*Presence of Posterior Staphyloma*	*Type of Posterior Staphyloma*
1	57	P(Lc)	57	28.5	+	II	CNV	57	28.5	+	II
2	42	P(Lc)	43	30.1	+	I	CNV	44	30.1	+	I
3	51	P(Lc)	52	30.1	+	IX	CNV	53	30.1	+	IX
4	42	P(Lc)	43	29.2	+	II	CNV	45	29.2	+	II
5	52	P(Lc)	57	29.7	+	IX	CNV	59	29.7	+	IX
6	48	P(Lc)	50	29.6	+	II	CNV	54	29.6	+	II
7	55	CNV	59	27.8	+	II	Macular atrophy	62	27.8	+	II
8	33	CNV	52	30.7	+	IX	Macular atrophy	55	30.7	+	IX
9	54	CNV	55	29.4	+	IX	Macular atrophy	58	29.4	+	IX
10	67	CNV	72	27.1	+	I	Macular atrophy	74	27.1	+	I
11	52	CNV	62	28.9	+	IX	Macular atrophy	64	28.9	+	IX
12	37	CNV	50	30.4	+	I	Macular atrophy	52	30.4	+	I
13	60	CNV	61	28.0	+	I	Macular atrophy	64	28.0	+	I
14	28	CNV	34	28.5	+	II	Macular atrophy	36	28.5	+	II
15	34	CNV	35	31.4	+	II	Macular atrophy	37	31.4	+	II

CNV = choroidal neovascularization; P(Lc) = patchy atrophy originated from lacquer cracks.

sion ($P = 0.005$). There was no difference in the follow-up period between the other progression patterns and those without progression.

There were 15 eyes of 15 patients who had a second and third progression pattern after the first progression from lacquer cracks at the initial examination (Table 8). All of these 15 eyes already had a posterior staphyloma at the initial examination. The progression pattern was divided into 2 clear paths. Six of the eyes progressed to the P(Lc) type of patchy atrophy and later developed a CNV at the foveal edge. All of these eyes developed macular atrophy. A representative case is shown in Figure 6. The remaining 9 eyes that developed a CNV as the first progression later developed macular atrophy.

Diffuse chorioretinal atrophy. In 372 eyes that had diffuse chorioretinal atrophy at the initial visit, 100 eyes (26.9%) had an enlargement of the diffuse atrophy, 72 eyes (19.4%) progressed to the P(D) type of patchy atrophy, 8 eyes (2.2%) to lacquer cracks, 6 eyes (1.6%) to a CNV, and 189 (50.8%) did not progress (Table 9). Three eyes progressed to 2 different types of lesions in the same eye. The patients who developed a CNV or developed P(D) were significantly older than those without a progression ($P = 0.043$ and $P = 0.0065$, respectively). There were no differences in age between the patients who progressed to an enlargement of the diffuse atrophy and those without progression. The axial lengths of the eyes that showed an enlargement of the diffuse atrophy or developed P(D) were significantly longer than those without a progression ($P = 0.02$ and $P<0.0001$, respectively). A posterior staphyloma was detected significantly more frequently in the eyes that showed an enlargement of the diffuse atrophy and those that developed a P(D) than those without a progression ($P<0.0001$ and $P<0.001$, respectively).

Table 9. First Progression Pattern and Clinical Characteristics in Eyes with Diffuse Atrophy

Progression Type	Frequency, No. of Eyes (%)	Age (yrs)	Axial Length (mm)	Posterior Staphyloma, No. of Eyes (%)	Type of Staphyloma	Follow-up (yrs)
With progression (183 eyes)						
Enlargement of area	100 (26.9)	44.0±13.7	29.7±1.5	95 (95.0)	I: 23 eyes (24.2%) II: 52 eyes (54.7%) IX: 13 eyes (13.7%) Others: 12 eyes (12.0%)	14.1±5.6
P(D)	72 (19.4)	48.6±14.3	30.6±1.6	72 (100)	I: 15 eyes (20.8%) II: 31 eyes (43.1%) IX: 21 eyes (29.2%) Others: 5 eyes (6.9%)	14.8±6.6
Lacquer cracks	8 (2.2)	30.3±19.7	30.0±0.7	8 (100)	I: 2 eye (25.0%) II: 6 eyes (75.0%)	17.7±3.3
CNV	6 (1.6)	58.3±6.2	28.3±1.3	6 (100)	I: 2 eyes (33.3%) II: 3eyes (50.0%) IX: 1 eyes (16.6%)	11.0±3.0
Without progression (189 eyes)	189 (50.8)	42.1±16.1	29.1±1.7	128 (67.8)	I: 46 eyes (35.9%) II: 69 eyes (53.9%) IX: 7 eyes (5.5%)	12.1±6.2

CNV = choroidal neovascularization; P(D) = patchy atrophy which develops in the posterior fundus accompanying diffuse atrophy.

Lacquer Cracks and Its Clinical Characteristics

Period from 1st to 2nd Progression (yrs)	Type of 3rd Progression	At 3rd Progression				Period from 2nd to 3rd Progression (yrs)	Age at the Final Visit (yrs)	Total Follow-up (yrs)
		Age (yrs)	*Axial Length (mm)*	*Presence of Posterior Staphyloma*	*Type of Posterior Staphyloma*			
0.5	Macular atrophy	60	28.6	+	II	2.0	74	16.7
1.5	Macular atrophy	45	30.1	+	I	1.5	47	5.0
1.5	Macular atrophy	55	28.6	+	IX	3.0	59	7.8
2.0	Macular atrophy	49	30.0	+	II	4.0	54	11.8
2.0	Macular atrophy	62	29.2	+	IX	3.0	73	20.7
4.0	Macular atrophy	56	29.6	+	II	2.0	56	7.2
3.0	—						66	11.3
3.5	—						56	22.6
3.5	—						61	6.5
2.0	—						74	6.5
2.5	—						73	20.7
2.5	—						52	15.0
3.0	—						82	22.0
2.5	—						37	8.8
2.5	—						43	9.2

The eyes that had progressed to a P(D) had the type IX staphyloma more frequently than the eyes without a progression ($P<0.0001$). In fact, a type IX staphyloma was observed in only 5.5% of the eyes without a progression, but in 29.2% of the eyes that developed a P(D). The patients who showed an enlargement of the diffuse atrophy, the development of P(D), or the development of lacquer cracks had significantly longer follow-up periods than those without progression ($P = 0.0032$, $P = 0.0011$, and $P = 0.0058$, respectively).

There were 9 eyes of 8 patients with diffuse atrophy that progressed to a second and third type of maculopathy (Table 10). Seven of the 9 eyes had a posterior staphyloma at the initial examination, and the remaining 2 eyes (both eyes of case 1) did not. Both eyes of case 1 developed lacquer cracks despite the absence of a posterior staphyloma, and then progressed to P(Lc), and then showed an enlargement of the P(Lc) with the development of a posterior staphyloma (Fig 7, available online at http://aaojournal.org). The 6 eyes (cases 2–7) that developed a CNV from a diffuse atrophy later progressed to macular atrophy. The remaining eye that developed P(D) from diffuse atrophy later developed a CNV at the foveal edge, and finally progressed to macular atrophy (Fig 8).

Patchy chorioretinal atrophy. In the 74 eyes that had patchy atrophy at the initial visit, 50 eyes (67.6%) showed an enlargement of the patchy areas, 10 eyes (13.5%) showed a fusion with P(D) or P(St), 2 eyes (2.7%) developed a CNV, and 22 eyes (29.7%) did not progress (Table 11). The 10 eyes that showed a fusion of P(D) or P(St) had an enlargement of the area of patchy atrophy.

There were no significant differences in the ages of patients with each progression pattern and those without a progression. The axial length was significantly longer in the eyes that had an enlargement of the patchy areas or a fusion with other P(D) or P(St) than eyes without progression ($P = 0.012$ and $P = 0.001$, respectively). A posterior staphyloma was observed in all of the eyes that had an enlargement of the area, those that developed a CNV, and those that had a fusion with other P(D) or P(St), whereas a posterior staphyloma was observed in 81.8% of the eyes without a progression. The eyes that showed a fusion of the patchy atrophy with other P(D) or P(St) had type IX staphyloma significantly more frequently (90.0%) than eyes without a progression (11.1%; $P<0.0001$). Also, the eyes that showed an enlargement of the area of atrophy had type IX staphyloma significantly more frequently than those without a progression ($P = 0.041$). The mean follow-up period was significantly longer in the eyes that showed an enlargement of the area and had a fusion with other P(D) or P(St) than the eyes without progression ($P = 0.0035$ and $P = 0.022$, respectively).

Among the 74 eyes with patchy chorioretinal atrophy, 25 eyes had the P(Lc) type, 40 eyes had the P(D) type, and 20 eyes had the P(St) type of patchy atrophy at the initial examination. In the 40 eyes with P(D), 22 eyes (55.0%) had an increase in the area, and 10 eyes (25.0%) had a fusion with other atrophic areas. In 20 eyes with P(St), 18 eyes (90.0%) showed an increase in the area and 10 eyes (50.0%) showed a fusion with other patchy atrophy. The development of a CNV was found only in the 2 eyes (8.0%) with the P(Lc) type. In the 25 eyes with P(Lc), 16 eyes (64.0%) had an increase in the area of atrophy and none of the patients showed a fusion with other patchy atrophy.

There were 12 eyes of 12 patients who progressed to a second and third stage after the first progression from patchy atrophy (Table 12). All of the 12 eyes had a posterior staphyloma at the initial examination. The further progression pattern was divided into 2 clear paths. The eyes of cases 1 and 2 that developed a CNV from patchy atrophy at the edge of the foveal P(Lc) as the first progression progressed to macular atrophy. The other 10 eyes of 10 patients that had a fusion with other P(D) or P(St) had an enlargement of the patchy atrophy and progressed to macular atrophy. A representative case is shown in Figure 9.

Choroidal neovascularization. In the 91 eyes with a CNV at the initial visit, 82 eyes (90.1%) developed a macular atrophy around the regressed CNV and 9 eyes did not progress (Table 11). Because of the small number of the eyes without progression, statistical analyses showed no significant difference in the patients' age, axial length, or prevalence as well as the type of staphyloma between the eyes with and without progression.

Influence of Each Progression Pattern on Visual Prognosis

A summary of the BCVA at the initial examination and at the time of the progression for each progression pattern is presented in

Table 10. Second and Third Progression Pattern from

Case No.	Side	Age at Initial Visit (yrs)	Type of 1st Progression	At 1st Progression				Type of 2nd Progression	At 2nd Progression			
				Age (yrs)	*Axial Length (mm)*	*Presence of Posterior Staphyloma*	*Type of Posterior Staphyloma*		*Age (yrs)*	*Axial Length (mm)*	*Presence of Posterior Staphyloma*	*Type of Posterior Staphyloma*
1	R	14	Lacquer cracks	21	30.6	—		P(Lc)	23	30.9	—	
	L	14	Lacquer cracks	22	30.7	—		P(Lc)	23	29.4	—	
2	R	54	CNV	58	27.4	+	II	Macular atrophy	61	28.0	+	II
3	L	52	CNV	54	28.8	+	II	Macular atrophy	56	30.2	+	II
4	R	60	CNV	68	28.9	+	IX	Macular atrophy	73	28.8	+	IX
5	R	70	CNV	71	26.9	+	I	Macular atrophy	73	26.9	+	I
6	L	59	CNV	61	27.4	+	I	Macular atrophy	63	29.1	+	I
7	R	51	CNV	58	30.6	+	II	Macular atrophy	60	30.6	+	II
8	L	43	P(D)	47	28.7	+	II	fuse with other P(D)	53	28.8	+	II

CNV = choroidal neovascularization; L = left; N/D = not determined because an enlargement of macular atrophy occurs gradually; P(D) = patchy

Table 13. To avoid confusion, the analysis was performed only on the first progression pattern from each myopic maculopathy. Statistical analyses showed that the development of a CNV and macular atrophy significantly worsened the vision ($P<0.0001$). The fusion of patches of atrophy with other areas in eyes classified with P(D) or P(St) was the only other progression pattern that caused a significant decrease in vision (Fig 8).

Discussion

Our results showed that 40% of the highly myopic eyes had progression of myopic maculopathy during a mean follow-up of 12.7 years. Vongphanit et al[9] reported that after a mean period of 61 months, a significant progression of myopic retinopathy was observed in only 8 eyes (17.4%). This incidence was much lower than our results. One reason for the difference may be because our population was a hospital-based group and their population[9] was a community-based group. Also, the patients who stopped visiting the High Myopia Clinic might have been those who did not show a visual decrease or progression of myopic maculopathy. Another reason might be that they examined their patients after only 5 years, and our results showed that the progression required a considerable time, especially in eyes

Table 11. Progression Pattern and Clinical Characteristics in Eyes with Patchy Atrophy or Choroidal Neovascularization

Retinopathy at Initial Visit	Progression Type	Frequency, No. of Eyes (%)	Age (yrs)	Axial Length (mm)	Posterior Staphyloma, No. of Eyes (%)	Type of Staphyloma	Follow-up (yrs)
Patchy atrophy (74 eyes)	Enlargement of area	50 (67.6)	47.9±12.7	30.7±1.6	50 (100)	I: 13 eyes (26.0%), II: 15 eyes (30.0%) IX: 19 eyes (38.0%) Others: 3 eyes (6.0%)	14.7±6.5
	Fuse with other P(D) or P(St)	10 (13.5)	52.1±10.0	32.0±1.1	10 (100.0)	II: 1 eye (10.0%) IX: 9 eyes (90.0%)	16.4±6.3
	CNV	2 (2.7)	57.5±9.5	28.0±0.8	2 (100)	II: 1 eye (50.0%) IX: 1 eyes (50.0%)	14.0±0.6
	No progression	22 (29.7)	47.0±12.2	29.7±1.7	18 (81.8)	I: 6 eyes (33.3%) II: 9 eyes (50.0%) IX: 2 eyes (11.1%) Others: 1 eye (5.6%)	9.8±5.4
CNV (91 eyes)	Macular atrophy	82 (90.1)	50.0±13.5	28.8±1.7	78 (95.1)	I: 15 eyes (19.5%) II: 39 eyes (50.6%) IX: 20 eyes (26.0%) Others: 4 eyes (5.1%)	11.8±5.9
	No progression	9 (9.9)	42.6±13.9	29.4±1.4	7 (77.8)	I: 1 eye (14.3%) II: 3 eyes (42.9%) IX: 2 eyes (28.6%) Others: 1 eye (1.4%)	9.9±5.1

CNV = choroidal neovascularization; P(D) = patchy atrophy which develops in the posterior fundus accompanying diffuse atrophy; P(Lc) = patchy atrophy which develops from lacquer cracks; P(St) = patchy atrophy which develops along the edge of posterior staphyloma.

Diffuse Atrophy and Its Clinical Characteristics

Period from 1st to 2nd Progression (yrs)	Type of 3rd Progression	At 3rd Progression: Age (yrs)	At 3rd Progression: Axial Length (mm)	At 3rd Progression: Presence of Posterior Staphyloma	At 3rd Progression: Type of Posterior Staphyloma	Period from 2nd to 3rd Progression (yrs)	Type of 4th Progression	Age (yrs)	At 4th Progression: Axial Length (mm)	At 4th Progression: Period from 3rd to 4th Progression (yrs)	Age at Final Visit (yrs)	Total Follow-up (yrs)
1.5	Enlargement of P(Lc)	32	32.2	+	II	9.0					36	21.5
1.0	Enlargement of P(Lc)	32	32.2	+	II	9.0					36	21.5
2.5	—										72	13.5
1.5	—										63	11.0
4.0	—										73	13.3
1.5	—										76	6.2
2.0	—										73	13.5
1.5	—										63	11.9
6.0	CNV	57	28.7	+	II	10.0	Macular atrophy	59	30.5	3.0	60	16.8

atrophy which developed from diffuse atrophy; P(Lc) = patchy atrophy which develops from lacquer cracks; R = right.

with a tessellated fundus. In addition, their cohorts probably had less severe myopic retinopathies than those in our study. Because the number of patients with myopic retinopathy was small in their study, it would be difficult to determine the long-term progression of myopic retinopathy from their observations.

One of the difficulties in interpreting the earlier studies was that the definition and the classification of myopic maculopathy were different. Vongphanit et al[9] defined myopic retinopathy as a posterior staphyloma, myopic conus, lacquer cracks, Fuchs' spot, and myopic chorioretinal atrophy. In the classification of Avila et al,[11] myopic retinopathy included tessellated fundus, posterior staphyloma, lacquer cracks, and patchy atrophy. In our study, we defined myopic maculopathy as a tessellated fundus, diffuse chorioretinal atrophy, lacquer cracks, patchy chorioretinal atrophy, CNV, and macular atrophy.

Based on our classification, we examined the progression pattern of the myopic maculopathy for each type of fundus lesion. The results of our longitudinal study indicated that the incidence and type of progression were different for the different types of fundus lesion. Only 13.4% of eyes with a tessellated fundus showed a progression of myopic maculopathy during the follow-up period, whereas 69.3% of eyes with lacquer cracks, 49.2% of eyes with diffuse atrophy, 70.3% of eyes with patchy atrophy, and 90.1% of eyes with CNV showed a progression of the myopic maculopathy. These findings suggest that myopic maculopathy tends to progress more quickly after the myopic maculopathy has advanced past the tessellated fundus stage.

Statistical comparisons of the clinical characteristics at the initial examination showed that the patients with tessellated fundus were significantly younger than any of the other groups, and were significantly less myopic than any of the other groups. Also, a posterior staphyloma was present in significantly fewer eyes with a tessellated fundus than any of the other groups. These findings suggest that patient age, degree of myopia, axial length, and the presence of posterior staphyloma might be important factors for the progression of myopic maculopathy. These findings also support our hypothesis that the tessellated fundus may be the first fundus lesion observed in eyes with pathologic myopia.

Because the progression of each stage of myopic maculopathy had different paths, we first examined the first progression pattern of each fundus lesion. The development of a CNV was found to develop from a tessellated fundus, lacquer cracks, diffuse atrophy, and patchy atrophy (Tables 5, 7, 9, and 11). However, the incidence of progression to a CNV was higher in the eyes with lacquer cracks or patchy atrophy than from a tessellated fundus which confirmed our earlier results.[17] Once developed, most of the eyes with a CNV showed a progression to macular atrophy. Thus, 1 longitudinal progression pattern of myopic maculopathy was the progression from a tessellated fundus, lacquer cracks, diffuse atrophy, and patchy atrophy to a CNV and then to macular atrophy (Fig 4).

Other than the development of a CNV, the progression pattern was different for the different types of maculopathies. In the eyes with a tessellated fundus, the most frequent pattern was the progression to diffuse chorioretinal atrophy (10%), and the second most frequent pattern was the development of lacquer cracks (3%). Lacquer cracks also developed in eyes with diffuse atrophy at a comparable incidence of 2.2% (8 of 272 eyes); however, the degree of diffuse atrophy in these 8 eyes was mild. The fact that lacquer cracks did not develop in eyes with more severe myopic maculopathy, such as patchy atrophy, CNV, and macular atrophy, indicates that lacquer cracks develop at an earlier stage than these more severe maculopathies. This suggests that a long, linear rupture of Bruch's membrane that is clearly observed as yellowish lacquer cracks ophthalmoscopically might develop when mechanical tension is placed on the eyes with less-atrophied and relatively thick retinas such as those with a tessellated fundus or mild diffuse atrophy. Although many minute ruptures of Bruch's membrane might occur in highly atrophied retinas, these

Table 12. Second and Third Progression Pattern from

Case No.	Age at Initial Visit (yrs)	Type of 1st Progression	At 1st Progression				Type of 2nd Progression
			Age (yrs)	*Axial Length (mm)*	*Presence of Posterior Staphyloma*	*Type of Posterior Staphyloma*	
1	60	CNV	67	28.7	+	IX	Macular atrophy
2	47	CNV	48	27.2	+	II	Macular atrophy
3	53	Fuse with other P(D) or P(St)	62	30.3	+	IX	Macular atrophy
4	30	Fuse with other P(D) or P(St)	51	32.7	+	IX	Macular atrophy
5	53	Fuse with other P(D) or P(St)	63	32.9	+	IX	Macular atrophy
6	55	Fuse with other P(D) or P(St)	60	32.3	+	IX	Macular atrophy
7	55	Fuse with other P(D) or P(St)	61	30.9	+	IX	Macular atrophy
8	69	Fuse with other P(D) or P(St)	72	30.5	+	IX	Macular atrophy
9	54	Fuse with other P(D) or P(St)	63	33.9	+	IX	Macular atrophy
10	45	Fuse with other P(D) or P(St)	62	31.9	+	IX	Macular atrophy
11	45	Fuse with other P(D) or P(St)	60	32.5	+	II	Macular atrophy
12	62	Fuse with other P(D) or P(St)	70	32.1	+	IX	Macular atrophy

CNV = choroidal neovascularization; P(D) = patchy atrophy originated from diffuse atrophy; P(St) = patchy atrophy along the staphyloma.

minute ruptures might not be recognized ophthalmoscopically. In addition, it might also be possible that lacquer cracks could not be detected in eyes with highly atrophied retinas such as those with patchy atrophy. Although we performed fluorescein angiography in all of the patients, fundus autofluorescence[23] or indocyanine green angiography,[24] both of which are useful for detecting lacquer cracks, were not routinely performed because these techniques were

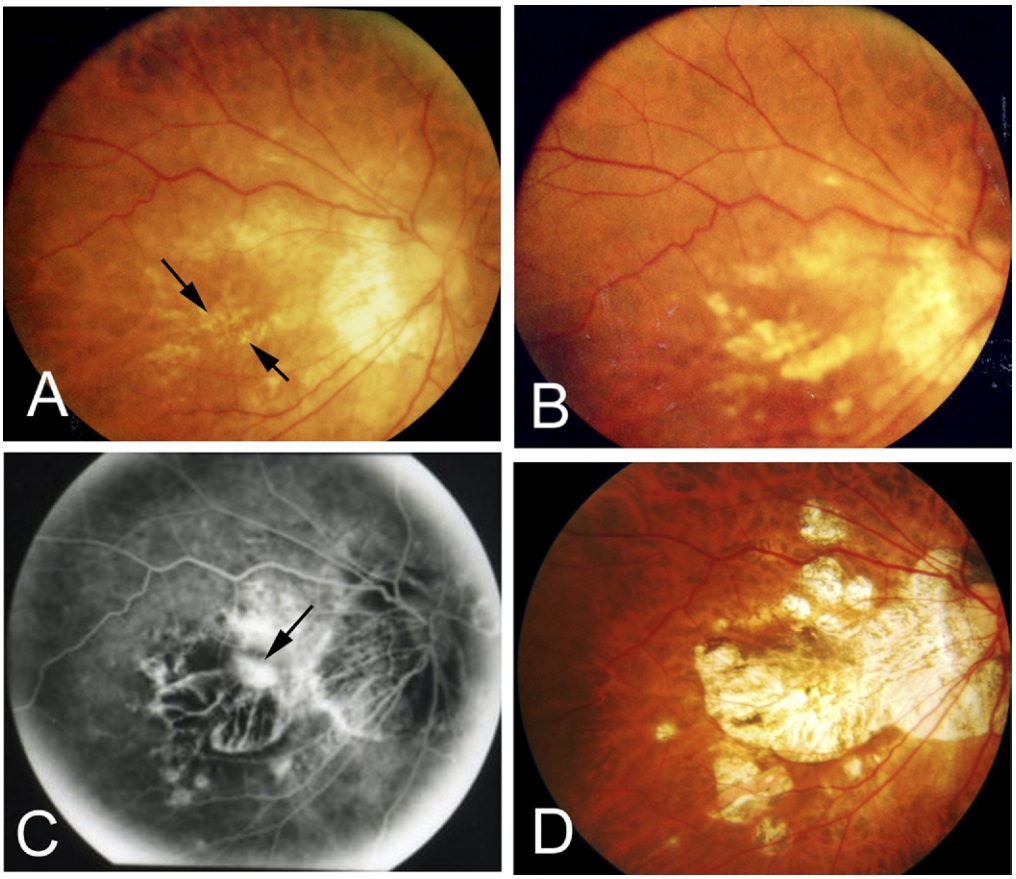

Figure 6. Complex progression from lacquer cracks in a 51-year-old woman, right eye; refraction, −13.0 D; axial length, 30.1 mm (case 3 in Table 8). **A**, At the initial examination, the right fundus shows multiple lacquer cracks in the foveal region (arrows). Best-corrected visual acuity (BCVA) was 1.0. **B**, One year later, the width of lacquer cracks has increased and progressed to patchy atrophy; BCVA was 0.8. **C**, Three years after the initial examination, fluorescein angiogram showed the hyperfluorescence due to a choroidal neovascular membrane (arrow) which developed at the foveal edge of patchy atrophy, and BCVA decreased to 0.1. **D**, Eight years after the initial examination, the posterior fundus was replaced by macular atrophy, and the BCVA decreased to 0.05. D = diopter.

Patchy Atrophy and Its Clinical Characteristics

At 2nd Progression						
Age (yrs)	*Axial Length (mm)*	*Presence of Posterior Staphyloma*	*Type of Posterior Staphyloma*	**Period from 1st to 2nd Progression (yrs)**	**Age at Final Visit (yrs)**	**Total Follow-up (yrs)**
69	29.3	+	IX	2.0	73	13.3
50	26.6	+	II	3.0	62	14.6
71	30.4	+	IX	9.0	73	19.8
53	33.2	+	IX	2.0	53	23.0
70	31.5	+	IX	7.0	73	19.8
64	32.5	+	IX	4.0	64	9.0
64	31.3	+	IX	3.0	64	9.0
73	31.5	+	IX	1.0	74	5.0
68	33.9	+	IX	5.0	69	15.4
66	31.4	+	IX	4.0	68	23.2
65	33.2	+	II	5.0	68	23.2
75	32.6	+	IX	5.0	79	17.0

not in use at the early phase of this study. This is a limitation of this study, and a prospective study incorporating fundus autofluorescence and indocyanine green angiography is necessary to make stronger conclusions.

Lacquer cracks showed characteristic progression patterns. The most frequent pattern was the progression to the P(Lc) type of patchy atrophy (42.7% of eyes). The other progression patterns included the development of CNV (13.3%), and an increase in the number of lacquer cracks (13.3%). We have reported that lacquer cracks progressed in 37 (56.1%) of 66 eyes with a mean follow-up period of 72.8 months,[18] and the most frequent pattern was also a progression to patchy atrophy in 40.5%. Fifteen eyes with lacquer cracks showed a second and third progression after the first progression. The subsequent development of macular atrophy after the CNV development was also observed in eyes with lacquer cracks as was shown in eyes with a tessellated fundus. However, the characteristic progression pattern in the eyes with lacquer cracks was the development of a CNV at the foveal edge in eyes with the P(Lc) type of patchy atrophy and a final progression to macular atrophy. Because lacquer cracks usually develop near the foveal area, P(Lc) also develops near the fovea as the width of the lacquer cracks increased. Later, a CNV developed at the foveal edge of eyes with patchy atrophy together with an enlargement of the P(Lc) as we have reported.[17] This progression pattern was observed at a relatively high incidence; 6 of 32 eyes (18.8%) which showed the progression to P(Lc) from lacquer cracks as the first progression. This suggests that we should carefully monitor these eyes and watch for the development of a CNV when lacquer cracks increase their width and progress to P(Lc).

Combining our findings on eyes with a tessellated fundus or lacquer cracks, we suggest that 1 progression pattern is from a tessellated fundus to the development of lacquer cracks to P(Lc) to a CNV at the edge of a P(Lc) to macular atrophy (Fig 4).

In eyes with diffuse atrophy, an enlargement of area of atrophy was observed in 26.9% of the eyes and a development of P(D) in 19.4% of the eyes. Nine eyes of 8 patients showed a second and third progression (Table 10). Although the 2 patterns were similar to those already shown in eyes with tessellated fundus or lacquer cracks, that is, the development of lacquer cracks to P(Lc) and the development of CNV to macular atrophy, 1 patient (case 8 in Table 10) showed a progression from diffuse atrophy to P(D) and then a fusion of the atrophic areas with other areas of P(D), then to a CNV at the foveal edge of a fused P(D), and finally macular atrophy (Fig 8). This complex pattern was the only pattern that developed a CNV from patchy atrophy other than P(Lc).

In eyes with patchy atrophy, the most frequent progression pattern was an enlargement of the area followed by fusion with other P(D) or P(St). In fact, all of the eyes that showed a fusion of patchy atrophy with other P(D) or P(St) atrophies had an enlargement of the area. Statistical comparison of the BCVA between the initial examination and at the first progression revealed that the fusion of areas with other P(D) or P(St) was the only progression pattern causing a significant decrease in vision other than the development of a CNV or macular atrophy (Table 13).

Among the subtypes of patchy atrophy, a fusion of patchy atrophy was observed only in eyes with P(D) or P(St). None of the eyes with P(Lc) showed this type of progression. Different from P(Lc), which usually develops in eyes with less atrophic retinas such as the tessellated fundus, P(D) and P(St) usually developed in eyes with more atrophic fundus such as those with severe diffuse atrophy and a deep staphyloma. This probably happens because of choroidal circulatory disturbances and the continuous high mechanical tension on the retina and choroid within the staphyloma in the highly atrophic fundus. A similar progression pattern was found in eyes with severe diffuse atrophy, where P(D) gradually enlarges and finally fuse with other P(D) or P(St). Even though the patchy atrophy usually enlarges, it usually enlarges in a centrifugal direction, and patchy atrophy that usually develops outside the fovea does not involve the central fovea after its enlargement.[25] However, when patchy atrophy fuses with other P(D) or P(St) and the fused areas further enlarge, only a small area in-

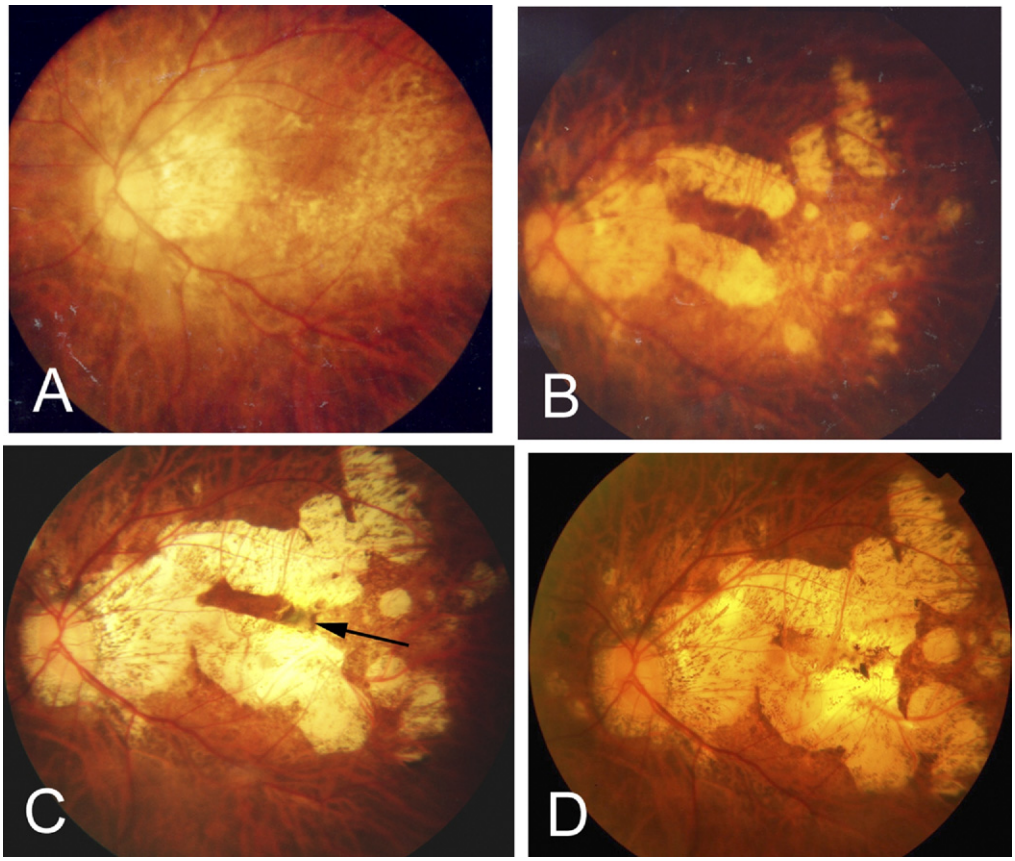

Figure 8. Complex progression from diffuse atrophy in a 43-year-old woman, left eye; refraction, −12.5 D; axial length, 28.7 mm (case 8 in Table 10). **A**, At the initial examination, the left fundus shows diffuse chorioretinal atrophy. Best-corrected visual acuity (BCVA) was 1.0. **B**, Six years later, multiple areas of patchy atrophy developed within the area of diffuse atrophy; BCVA was 0.8. **C**, Fourteen years after the initial examination, choroidal neovascular membrane (arrow) developed bridging the 2 areas of patchy atrophy situated superior and inferior to the central fovea, and BCVA decreased to 0.2. **D**, Sixteen years after the initial examination, the posterior fundus was replaced as macular atrophy, and the BCVA decreased to 0.1. D = diopter.

cluding the central fovea remains as a small island of good vision. Thus, we suggest that the significant visual decrease probably happens because the remaining central fovea might undergo nutritional and circulatory defects that would finally result in a visual decrease (Fig 9).

When we combine the findings of tessellated fundus, diffuse atrophy, and patchy atrophy, the other longitudinal progression pattern involving different categories of myopic maculopathies is hypothesized as follows: from tessellated fundus to the development of diffuse atrophy, to the development of P(D) or P(St), to the fusion with the lesions of patchy atrophy, and finally resulting in a very small morphologically unaffected area including the central fovea (Fig 9). A hypothetical scheme of the progression pattern of each fundus lesion of myopic maculopathy based on the results of this study is presented in Figure 4.

Our results showed that the development of a posterior staphyloma was critically important for the progression of myopic maculopathy. During the follow-up period, a posterior staphyloma was observed significantly more frequently in the eyes that showed any progression pattern versus those without progression. Also, the incidence of a posterior staphyloma was significantly higher in eyes that showed a progression to P(Lc) or the development of a CNV from lacquer cracks versus those without progression.

The type of staphyloma also affected the incidence of progression of myopic maculopathy. The type IX staphyloma was found to be especially important for the progression of myopic maculopathy. The eyes that progressed to patchy atrophy from diffuse atrophy were found significantly more frequently in eyes with type IX staphyloma versus the eyes without progression. The type IX staphyloma was detected in 90% of the eyes that showed a fusion with other P(D) or P(St), and was observed in only 11% of the eyes that did not show progression from patchy atrophy. We have reported that the type II staphyloma was the most prominent type in the Japanese; however, in older subjects, the incidence of type II was significantly lower and the type IX was significantly greater.[26] The progression from type II to type IX probably increases the mechanical tension on the macular area of highly myopic eyes, which then leads to the progression of the myopic maculopathy. These observations suggest that the inhibition of the development of posterior staphyloma, especially type IX staphyloma, might be a therapeutic target to prevent significant progression of myopic maculopathy. Although the treatments for independent

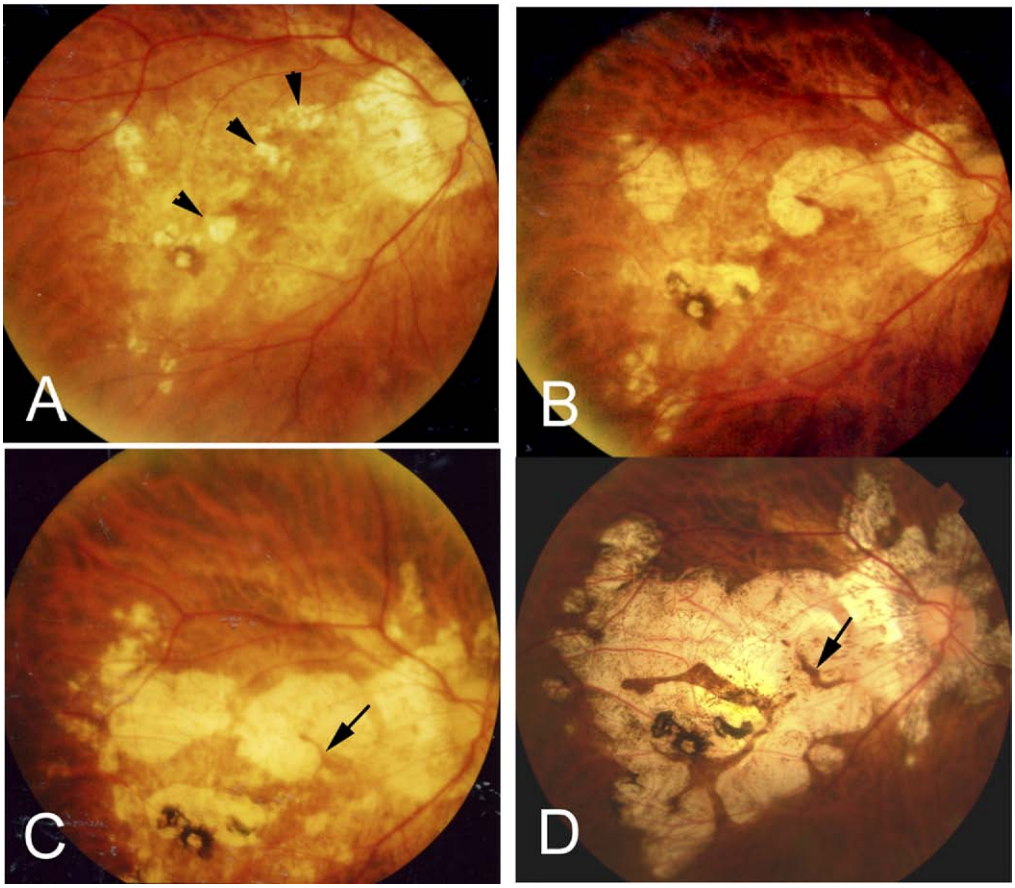

Figure 9. Fusion of areas of patchy atrophy and an enlargement of the fused area in a 45-year-old woman, right eye; refraction, −15.0 D; axial length, 32.0 mm (case 11 in Table 12). **A**, At the initial examination, the right fundus shows multiple areas of P(D) (patchy atrophy originated from diffuse atrophy; arrowheads) within the area of diffuse atrophy. Best-corrected visual acuity (BCVA) was 0.5. **B**, Fifteen years later, the areas of P(D) have enlarged and have fused with each other; BCVA was 0.5. **C**, Seventeen years after the initial examination, P(D) is further enlarged and fused with each other. The central fovea (arrow) was spared (arrow), and BCVA was maintained at 0.5. **D**, Twenty years after the initial examination, the patchy atrophy fused with each other and almost the entire posterior fundus was replaced with patchy atrophy. Although the central fovea (arrow) was still spared, the BCVA decreased to 0.1. D = diopter.

myopic fundus lesion, for example, antiangiogenic therapy against myopic CNV or vitrectomy against myopic traction maculopathy, are now being widely performed; an inhibition of staphyloma development is expected to be important to prevent the progression of myopic maculopathy.

One limitation of this study was that all of the patients did not start with the same fundus lesion. Some had tessellated fundus only at the initial examination, and others already had macular atrophy at the initial visit. However, we believe that analyses of the second and third progression patterns in the same eyes were useful to follow the different kinds of lesions of myopic maculopathy and to determine the progression patterns in the long-term. These observations led to the progression paths shown in Figure 4.

Another limitation of this study was that optical coherence tomography and visual field examinations were not routinely performed on all of the patients. Thus, it is possible that the concomitant presence of myopic traction maculopathy or myopic optic neuropathy, which might affect the vision, could have been missed. To solve this problem, optical coherence tomography and visual field examination have been routinely performed on all of the highly myopic patients since 2002, and we are planning to clarify the entire progression pattern of myopic maculopathy including myopic traction maculopathy in the future.

In conclusion, we determined the long-term progression pattern of each type of fundus lesion of myopic maculopathy, and established a scheme of the progression of myopic maculopathy. The results are clinically valuable for predicting the progression of myopic maculopathy and for predicting the vision in highly myopic patients. In addition, knowledge of the natural course of myopic maculopathies will allow us to determine the effectiveness of the treatments.

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Table 13. Comparison of Visual Acuity According to the Different Progression Patterns

Fundus Lesion at Initial Visit	First Progression Pattern	Initial BCVA	BCVA at Progression	*P*-Value
Tesselated fundus	Diffuse atrophy	0.06±0.12 (0.88)	0.06±0.16 (0.87)	NS
	Lacquer cracks	0.03±0.16 (0.93)	0.09±0.23 (0.81)	NS
	CNV	0.00±0.00 (1.0)	1.52±0.00 (0.03)	NS*
Lacquer cracks	Increase in number	0.11±0.19 (0.78)	0.14±0.18 (0.73)	NS
	P(Lc)	0.16±0.24 (0.70)	0.17±0.32 (0.68)	NS
	CNV	0.12±0.18 (0.77)	0.70±0.24 (0.20)	0.0019
Diffuse atrophy	Enlargement of area	0.17±0.31 (0.68)	0.22±0.33 (0.61)	NS
	Lacquer cracks	0.13±0.09 (0.73)	0.13±0.11 (0.73)	NS
	P(D)	0.21±0.26 (0.61)	0.31±0.38 (0.50)	NS
	CNV	0.22±0.30 (0.60)	0.74±0.32 (0.18)	<0.0001
Patchy atrophy	Enlargement of area	0.28±0.28 (0.52)	0.33±0.39 (0.46)	NS
	CNV	0.08±0.08 (0.84)	0.91±0.09 (0.12)	NS†
	Fuse with other P(D) or P(St)	0.30±0.18 (0.50)	0.62±0.35 (0.24)	0.046
CNV	Macular atrophy	0.71±0.45 (0.19)	1.07±0.41 (0.09)	<0.0001

BCVA = best-corrected visual acuity; CNV = choroidal neovascularization; NS = not significant; P(D) = patchy atrophy which develops in the posterior fundus accompanying diffuse atrophy; P(Lc) = patchy atrophy which develops from lacquer cracks; P(St) = patchy atrophy which develops along the edge of posterior staphyloma.
*Only 1 eye in this category.
†Only 2 eyes in this category.

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Footnotes and Financial Disclosures

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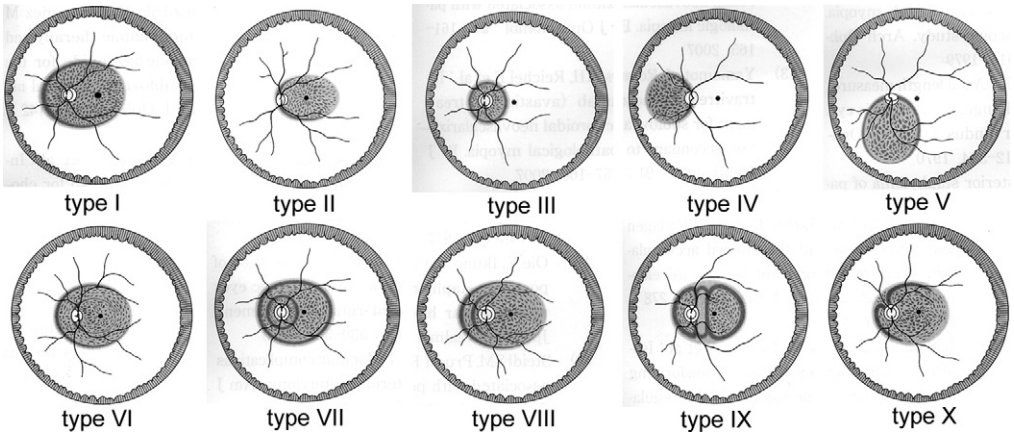

Figure 1. Drawings showing the classification of the different types of posterior staphyloma according to Curtin.[27]

Table 1. Number of Patients with Various Periods of Follow-up in Each Fundus Lesion of Myopic Maculopathy

	Type of Myopic Fundus Lesions at Initial Visit													
	Tesselated Fundus		*Lacquer Cracks*		*Diffuse Atrophy*		*Patchy Atrophy*		*CNV*		*MA*		**Total**	
Follow-up (yrs)	*No. of Eyes*	*Progression*	*No. of Eyes*	*Progression*	*No. of Eyes*	*Progression*	*No. of Eyes*	*Progression*	*No. of Eyes*	*Progression*	*No. of Eyes*	*Progression*	*No. of Eyes*	*Progression*
First visit only	133	—	15	—	130	—	19	—	22	—	26	—	301	—
<1 year	157	0	24	0	103	2	26	1	20	7	5	0	289	10 (3.5%)
1–2 years	76	0	6	0	153	6	35	2	20	12	8	1	244	21 (8.6%)
2–3 years	129	0	7	0	133	5	35	3	33	18	18	3	268	28 (10.4%)
3–4 years	101	0	8	1	167	9	28	3	28	17	5	2	263	31 (11.8%)
4–5 years	87	0	8	1	123	9	20	2	23	16	16	7	203	31 (15.3%)
≥5 years	276	37	75	52	372	183	74	52	91	82	45	36	806	327 (40.6%)

Table 2. Clinical Characteristics of Included and Excluded Patients

	Included Patients (n = 429) (follow-up ≥ 5 years)	Excluded Patients (n = 827) (follow-up < 5 years)	*P*-Value
Age (yrs), mean (SD) (range)	41.1±16.7 (4 to 74)	41.0±15.6 (4 to 81)	NS
Refractive error (D), mean (SD) (range)	−13.4±4.9 (−5.0 to −36.0)	−13.1±4.6 (−4.5 to −36.1)	NS
Axial length (mm), mean (SD) (range)	28.7±1.9 (25.0 to 35.4)	28.6±1.8 (25.0 to 36.4)	NS
Baseline logMAR, mean (SD) (range)	0.26±0.46 (−0.18 to 2.00)	0.22±0.42 (−0.18 to 2.00)	NS
posterior staphyloma	530 (65.8%)	985 (62.8%)	NS

CNV = choroidal neovascularization; D = diopters; logMAR = logarithm of minimal angle of resolution; SD = standard deviation.

Table 4. Clinical Characteristics of Eyes with Different Myopic Retinopathy at the Initial Examination

Fundus Lesion at Initial Visit	Age (yrs)	Refractive Error (D)	Axial Length (mm)	Posterior Staphyloma, No of Eyes (%)	BCVA, logMAR (decimal VA)
Tessellated fundus (276 eyes/170 patients)	31.9±15.9	−10.3±3.1	27.4±1.2	75 (27.2)	0.00±0.15 (1.00)
Lacquer cracks (117 eyes/91 patients)	44.0±13.9	−15.0±4.0	29.4±1.6	104 (88.9)	0.19±0.31 (0.65)*
Diffuse atrophy (491 eyes/292 patients)	45.7±15.0	−15.2±4.7	29.5±1.7	420 (85.6)	0.23±0.38 (0.59)*
Patchy atrophy (163 eyes/119 patients)	51.1±12.4	−16.4±5.6	30.1±2.0	156 (95.7)	0.30±0.37(0.50)*
CNV (91 eyes/83 patients)	49.4±13.6	−14.0±4.4	28.9±1.7	85 (93.4)	0.68±0.45 (0.21)
Macular atrophy (45 eyes/37 patients)	54.1±11.8	−17.6±7.1	30.4±2.3	45 (100)	1.27±0.43(0.05)

BCVA = best-corrected visual acuity; CNV = choroidal neovascularization; D = diopters; logMAR = logarithm of minimum angle of resolution.
*Eyes with accompanying CNV or macular atrophy were excluded.

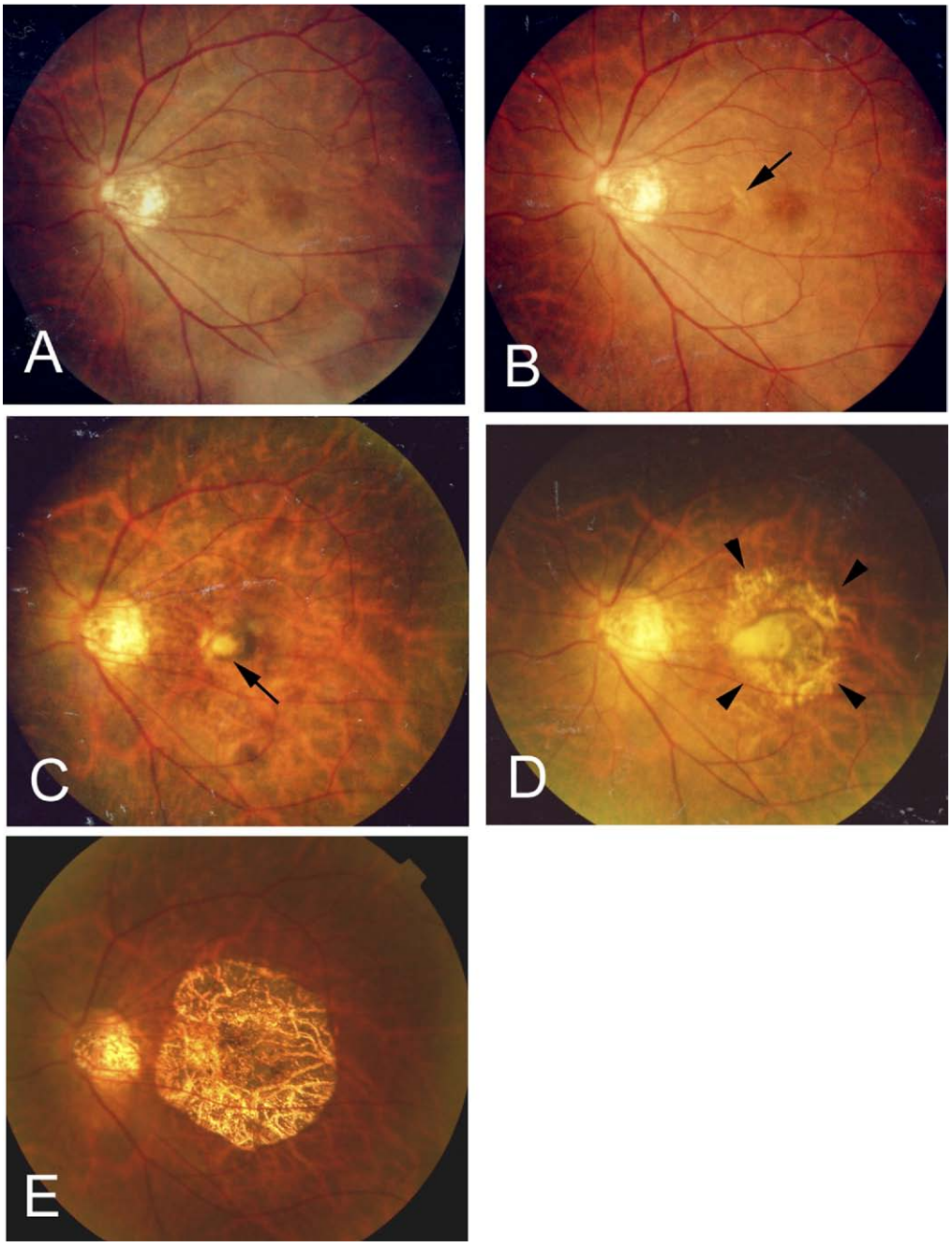

Figure 5. Complex progression from tessellated fundus in a 38-year-old man, left eye; refraction, −9.0 D; axial length, 26.5 mm (case 7 in Table 6). **A**, At the initial examination, the left fundus shows tessellated fundus. Best-corrected visual acuity (BCVA) was 1.2. **B**, Three years later, a lacquer crack (arrow) developed nasal to the central fovea. The BCVA was 1.0. **C**, Eleven years after the initial examination, a choroidal neovascular membrane (CNV; arrow) developed at the corresponding site of the lacquer crack, and BCVA decreased to 0.4. **D**, Fourteen years after the initial examination, macular atrophy (arrowheads) developed around the regressed CNV, and the BCVA decreased to 0.15. **E**, Twenty years after the initial examination, macular atrophy enlarged, and the BCVA further decreased to 0.1. D = diopter.

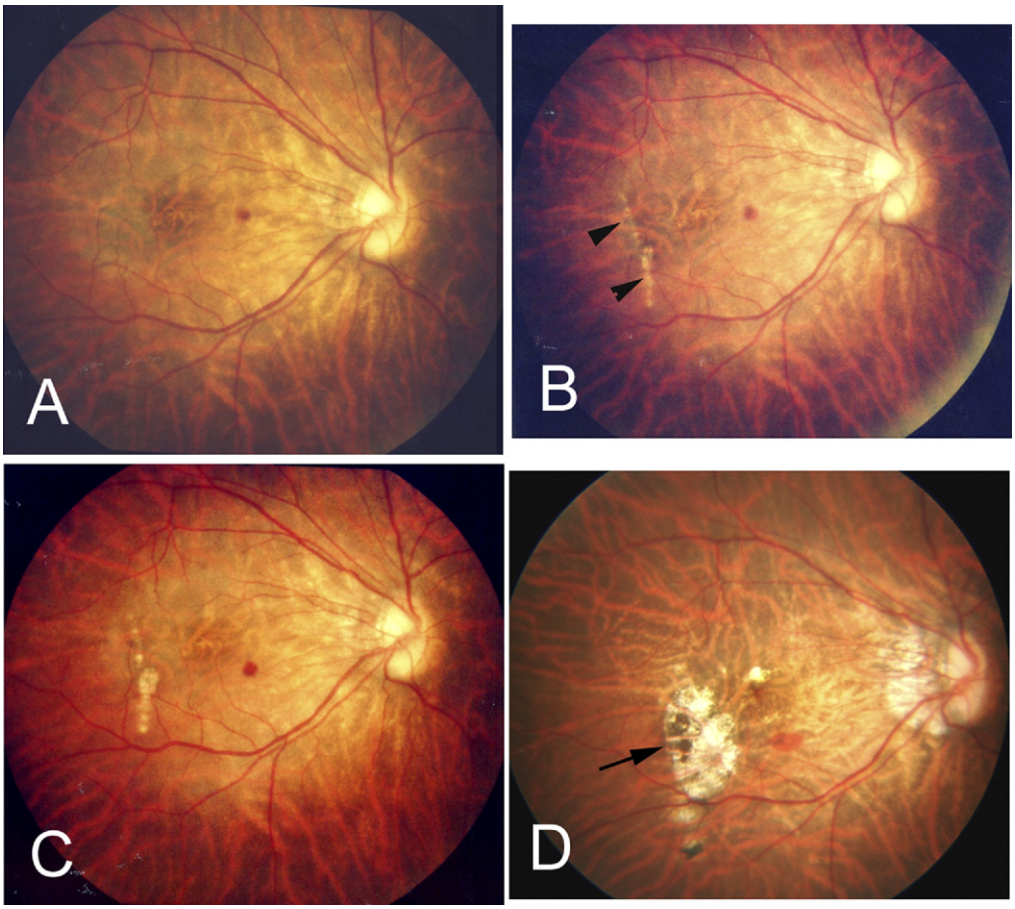

Figure 7. Complex progression from diffuse atrophy in a 14-year-old young girl, right eye; refraction, −19.0 D; axial length, 29.7 mm (case 1 in Table 10). **A**, At the initial examination, the right fundus shows mild diffuse atrophy temporal to the optic disc. A posterior staphyloma was not obvious. Best-corrected visual acuity (BCVA) was 1.0. **B**, Seven years later, the lacquer cracks developed temporal to the central fovea; BCVA was 1.0. **C**, Three months after the photograph shown in *B*, the width of lacquer cracks have slightly increased. **D**, Nine years after the initial examination, the lacquer cracks are increased in width and progressed to patchy atrophy. A myopic conus has also developed together with a development of type II staphyloma (according to Curtin[27]). The BCVA was 1.0.