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Long-Term Progression of Fundus Changes in Adults (2)

Reina Saito and Yuxin Fang

Abstract

Three cases with multiple progression patterns of myopic maculopathy in a long-term follow-up are presented.

Keywords

 $\label{eq:long-term} \begin{array}{l} \mbox{Long-term follow-up} \cdot \mbox{Progression} \cdot \mbox{Myopic maculopathy} \cdot \\ \mbox{Diffuse atrophy} \cdot \mbox{Patchy atrophy} \cdot \mbox{MNV} \end{array}$

R. Saito \cdot Y. Fang (\boxtimes)

Department of Ophthalmology and Visual Science, Tokyo Medical and Dental University, Tokyo, Japan

27.1 Case 1-Maintaining Good Vision Despite a Marked Enlargement of Diffuse Atrophy and Development of Myopic MNV (Figs. 27.1, 27.2, 27.3, 27.4, 27.5 and 27.6)



Fig. 27.1 Right and left fundus at the initial visit (1986) of a 31-yearold man show a small area of yellowish diffuse atrophy around the optic disc. The best-corrected acuity is 1.0 in the right eye and 0.7 in the left.

Refractive error is -15.0D in the right eye and -17.5D in the left. Axial length is 29.4 mm in the right eye and 29.9 mm in the left



Fig. 27.2 Fifteen years later (September 2001), the area of diffuse atrophy and peripapillary atrophy have enlarged. Peripapillary intrachoroidal cavitation is seen lower to the optic disc in the right eye. Two

lesions with pigmentation are seen lower to the macula in the left eye. The best-corrected visual acuity is 1.0 in both eyes. Axial length is 30.1 mm in the right eye and 31.3 mm in the left



Fig. 27.3 Thirty years later (February 2016), diffuse atrophy has extended to cover the entire posterior pole in both eyes. Right fundus shows mild pigmentation in the macula and a small patchy atrophy has

appeared lower to the macula. The peripapillary atrophy has further enlarged and extended around the optic disc in both eyes



Fig. 27.4 Thirty-one years later (May 2017), he complained the sudden blind spot in his right eye. Oblique OCT scan across the fovea shows subretinal hyper-reflective tissue (arrows), which represents a myopic macular neovascularization (MNV). The MNV shows hyper-

fluorescence (arrow) in the fluorescein angiogram. After 1 month following intravitreal injection of ranibizumab, OCT shows a complete disappearance of the myopic MNV



Fig. 27.5 Ultra wide-field OCT images of the right eye (Left) and the left eye (Right) (July 2018) show the dome-shaped macula in vertical scans bilaterally. No evident OCT features suggesting staphyloma edges (e.g., gradual choroidal thinning toward the staphyloma edge as well as

scleral inward protrusion) are seen, which means that there is no obvious staphyloma despite the dome-shaped macula in those axially elongated eyes. A defect of the retinal pigment epithelium (RPE) and Bruch's membrane are seen in the area of patchy atrophy in the right eye



Fig. 27.6 Wide-field fundus images at the last visit (February 2019). The best-corrected visual acuity is 1.0 in both eyes. Axial length is 32.3 mm in the right eye and 32.3 mm in the left

27.2 Case 2-Development of Patchy Atrophy, Followed by MNV Development Along The Foveal Edge of Patchy Atrophy, and Finally a Formation of Large Macular Atrophy by Fusion of MNV-Related Macular Atrophy and Patchy Atrophies (Figs. 27.7, 27.8, 27.9, 27.10, 27.11, 27.12,

27.13, 27.14, 27.15, 27.16 and 27.17)



Fig. 27.7 Right fundus at the initial visit (August 1996) of a 41-yearold woman shows diffuse choroidal atrophy around the optic disc. An ill-defined pigmented lesion is seen lower temporal to the macula. A temporal peripapillary crescent is also seen. The best-corrected visual acuity is 1.0. Refractive error is -15.8 D and an axial length is 29.5 mm



Fig. 27.8 Seven years later (November 2003), two lesions of patchy atrophy have appeared lower to the macula. These two lesions appear separated by a large choroidal vein



Fig. 27.9 Eleven years later (November 2007), myopic macular neovascularization (MNV) has developed along the foveal edge of a fused patchy atrophy (arrow). Fluorescein angiogram shows dye leakage from the MNV (arrow). The best-corrected visual acuity has decreased to 0.6. She received a total of three injections of intravitreal bevacizumab



Fig. 27.10 Seventeen years later (October 2013), the right fundus shows a pigmentation of scarred MNV, an enlargement of original patchy atrophy and a development of multiple new lesions of patchy atrophy. Vertical OCT shows the well-defined subfoveal hyper-

reflectivity compatible with a scarred MNV. In the area of patchy atrophy, an increased penetration of the light into deep tissues is seen due to the RPE defect. Dome-shaped macula is also seen. The best-corrected visual acuity is 0.2



Fig. 27.11 Twenty-three years later (July 2019), the right fundus shows a large macular atrophy which is formed by an enlargement and fusion of MNV-related macula atrophy and patchy atrophy. OCT shows

a large area of RPE defect. However, the best-corrected visual acuity is still 0.6. The axial length is 32.1 mm





Fig. 27.12 Left fundus at the first visit (August 1996) shows diffuse atrophy around the optic disc and a temporal peripapillary crescent. The best-corrected acuity is 1.0. The refractive error is -17.3 D and the axial length is 29.8 mm

Fig. 27.13 Eleven years later (December 2007), two small lesions of patchy atrophy have appeared upper temporal to the macula. These two lesions appear separated by a large choroidal vein



Fig. 27.14 Twelve years later (July 2008), the left fundus shows myopic MNV temporal to the fovea (arrow). Multiple lacquer cracks accompanying with a development and enlargement of patchy atrophy along the course of lacquer cracks are seen in the macula area.

Fluorescein angiogram shows dye leakage from the MNV (arrow). OCT shows a subretinal hyper-reflective lesion with slight serous retinal detachment. The visual acuity has decreased to 0.5. This eye was treated with intravitreal injections of bevacizumab



Fig. 27.15 Eighteen years later (February 2014), the patient complained of a new metamorphopsia. A new myopic MNV is observed (arrow). An enlargement of patchy atrophy is seen. This eye was treated with intravitreal injections of ranibizumab



Fig. 27.16 Twenty years later (September 2016), the patient complained of a decreased vision. The visual acuity has decreased to 0.3. OCT shows that a myopic MNV has appeared in the fovea



Fig. 27.17 Left fundus at 23 years after the initial visit shows a further enlargement of patchy atrophies and a fusion with MNV-related macular atrophy. (Top Right) Vertical OCT section shows the subfoveal hyperreflectivity and MNV-related macular atrophy (arrowheads). (Bottom) Oblique OCT scan shows a large area of RPE defect and

Bruch's membrane defect corresponding to two areas of patchy atrophy. The ends of the RPE and Bruch's membrane are indicated by arrows and an arrowhead, respectively. The best-corrected visual acuity is 0.5. The axial length is 32.4 mm 27.3 Case 3-Enlargement of Diffuse Choroidal Atrophy and a Development of Patchy Atrophy Within an Area of Diffuse Atrophy (Figs. 27.18, 27.19, 27.20, 27.21, 27.22, 27.23, 27.24, 27.25, 27.26, 27.27 and 27.28)



Fig. 27.18 Right fundus at the initial visit (May 2004) of a 53-year-old man shows diffuse choroidal atrophy covering the entire posterior fundus. A temporal peripapillary atrophy is also seen. The best-corrected acuity is 1.0. The axial length is 29.0 mm and the refractive error is -13.4 D



Fig. 27.20 Four years later (October 2008), horizontal OCT image shows an extreme thinning of the choroid. Subfoveal choroidal thickness is $28 \ \mu m$



Fig. 27.19 One year later (June 2005), two small lesions of patchy atrophy have appeared upper and lower to the macula



Fig. 27.21 Six years later (October 2010), the upper lesion of patchy atrophy has enlarged. Peripapillary atrophy has also enlarged



Fig. 27.22 Twelve years later (December 2016), the upper lesion of patchy atrophy has further enlarged. New lesions of patchy atrophy have appeared in the lower fundus. Swept-source OCT image across the

two lesions of patchy atrophy shows a defect of RPE and Bruch's membrane. The subfoveal choroidal thickness is still 28 μm



Fig. 27.23 Wide-field fundus image at 15 years after the first visit (January 2019) shows slightly pigmented temporal border of posterior staphyloma (arrows). Diffuse atrophy is seen only within the staphyloma. The axial length is 31.5 mm and the refractive error is -13.3 D. The best-corrected acuity is 1.0



Fig. 27.24 Left fundus at the first visit (June 2005) shows macular diffuse choroidal atrophy and a temporal peripapillary atrophy. Please note a large macular vortex vein with ampulla (arrow). The best-corrected visual acuity is 1.2. Axial length is 29.2 mm and the refractive error is -12.3 D



Fig. 27.25 Three years later (October 2008), there is no obvious change in the left fundus. Horizontal OCT image shows the extreme thinning of choroid. Subfoveal choroidal thickness is 24 µm. Branches of macular vortex vein (between arrowheads) are seen



Fig. 27.26 Twelve years later (December 2017), the area of diffuse atrophy and peripapillary atrophy have further enlarged. A close observation of the fundus photo shows that some branches of macular vortex vein have disappeared especially in the area nasal to the fovea

Fig. 27.27 Wide-field OCT image shows branches of macular vortex vein. In the area outside macular vortex vein, the choroid is extremely thin. Subfoveal choroidal thickness is $23 \ \mu m$



Fig. 27.28 Fifteen years later (January 2019), a wide-field fundus image shows the depigmented and pigmented changes at the edge of staphyloma (arrows). Diffuse atrophy is restricted within the staphylomatous area. The axial length is 31.1 mm and the refractive error is -12.3 D. The best-corrected visual acuity is 1.2