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Abstract

Diffuse choroidal atrophy is observed as an ill-defined yellowish lesion in the posterior fundus. Diffuse atrophy starts from the peripapillary region and eventually extends to the macula. OCT shows a marked thinning of the choroid in the area of diffuse atrophy. In some cases, almost the entire choroid is lost leaving sporadically remaining large choroidal vessels.

Keywords

Diffuse choroidal atrophy · Peripapillary diffuse choroidal atrophy · Macular diffuse choroidal atrophy · Choroidal thinning · Choriocapillaris

Diffuse choroidal atrophy is observed as an ill-defined yellowish lesion in the posterior fundus (Fig. 9.1). Diffuse atrophy primarily occurs in the peripapillary region as "peripapillary diffuse choroidal atrophy (PDCA)" and eventually extends to the macula as "macular diffuse choroidal atrophy (MDCA)" [1–3]. MDCA is uncommon below the age 40 or axial length shorter than 27 mm [4]. Diffuse atrophy is not uniformly yellow but shows granular yellowish appearance. However, the fundus color may look different according to the degree of fundus pigmentation among races (Fig. 9.2). Thus, the diagnosis of diffuse atrophy solely depending on fundus photos tends to be tricky especially in less pigmented eyes, and OCT-based diagnosis is recommended.

OCT shows a marked thinning of the choroid in the area of diffuse atrophy (Figs. 9.3 and 9.4). The subfoveal choroidal thickness in eyes with MDCA is usually below 100 μ m and the mean choroidal thickness is 50 μ m based on a clinic-based study [4]. In most cases, the choroid is almost absent except for sporadically remaining large choroidal vessels. Large choroidal blood vessel can be observed to protrude to ward the retina [5]. However, even in the area where most of the choroidal layer is absent, can be as thin as 12 micron or so, the RPE layer and outer retina are present by OCT (Fig. 9.4). It might explain a relatively preserved vision in eyes with diffuse atrophy. Such disproportionate thinning of the choroid compared to the surrounding tissue (retina and sclera) might be a key phenomenon in diffuse atrophy.

OCT angiography (OCTA) can detect choriocapillaris flow impairment, even though the visualization of the choroidal circulation remains a challenge in eyes with pathologic myopia. The OCTA in eyes with diffuse atrophy shows the low-density choriocapillaris, while medium and large sized choroidal vessels remained (Fig. 9.3) [6, 7].

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Macular Diffuse Choroidal Atrophy

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Fig. 9.1 Macular diffuse choroidal atrophy (MDCA) in an Asian patient. (Left) Left fundus of a 47-year-old woman with an axial length of 31.3 mm and with the best-corrected visual acuity (BCVA) of 1.0 shows granular yellowish, ill-defined MDCA covering almost the entire

macular area. (Right) OCT images in a horizontal section (upper image) as well as in a vertical section (lower image) show that most of the choroid is absent and only large choroidal vessels are sporadically present. Subfoveal choroidal thickness is $40 \ \mu m$



Fig. 9.2 Macular diffuse choroidal atrophy (MDCA) in a Caucasian patient. (Left) Right fundus of a 65-year-old Caucasian man with an axial length of 30.0 mm and with the best-corrected visual acuity

(BCVA) of 1.0 shows MDCA. Lacquer cracks are also seen. (Right) A horizontal OCT section shows that the choroid is very thin and the sub-foveal choroidal thickness is $25 \ \mu m$



Fig. 9.3 Extremely thin choroid in a case with macular diffuse choroidal atrophy (MDCA). (Left) Right fundus of a 70-year-old woman with an axial length of 30.2 mm shows MDCA. (Right) Vertical OCT section shows that most of the choroid is absent and subfoveal choroidal thick-

ness is as thin as 12 μ m. However, the best-corrected visual acuity (BCVA) still maintains 1.5. OCT also shows myopic retinoschisis with an epiretinal membrane superior to fovea. The inferior sclera is also very thin and the orbital fat is clearly seen



Fig. 9.4 Choroidal thinning and focal areas of choriocapillaris signal voids in a case with macular diffuse choroidal atrophy (MDCA). (Left) Right fundus of a 65-year-old man with an axial length of 30.3 mm and with the best-corrected visual acuity (BCVA) of 1.5 shows typical yellowish, ill-defined MDCA covering the entire macular area. (Right Top and Right Middle) OCT images show that the choroid is extremely thin

in the area of MDCA and the subfoveal choroidal thickness is $30 \mu m$. The entire thickness of the sclera is clearly visible. Vertical OCT image (Right, middle) also shows the inward bulge of the macular area, diagnosed as "dome-shaped macula." (Right bottom) OCT angiography shows the focal area of choriocapillaris signal voids with unmasking of the medium and large choroidal vessels seen as bright flow signals

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