High-Definition Optical Coherence Tomography in a Case of Congenital Hypertrophy of the Retinal Pigment Epithelium

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ABSTRACT
The authors describe the use of high-definition optical coherence tomography (HD-OCT) in a case of congenital hypertrophy of the retinal pigment epithelium. A 40-year-old woman presented with a large flat pigmented lesion in the inferior retinal quadrant of the left eye, which was compatible with congenital hypertrophy of the retinal pigment epithelium. The lesion was studied with HD-OCT (5 line raster and macular cube 512 × 128) and the results were compared with those of the fellow eye. The volume of the cube in the normal eye measured 7.1 mm³, whereas the volume of the cube with the lesion was 6.7 mm³. The subfield thickness analysis was 153 µm in the eye with congenital hypertrophy of the retinal pigment epithelium and 188 µm in the fellow eye, showing a difference of 35 µm between the two eyes. Congenital hypertrophy of the retinal pigment epithelium studied with HD-OCT showed loss of the junction layer between the inner and outer segment of photoreceptors and of the outer nuclear layer. [Ophthalmic Surg Lasers Imaging 2010;41:S93-S95.]

INTRODUCTION
Congenital hypertrophy of the retinal pigment epithelium (CHRPE) is a flat lesion at the level of the RPE with sharp margins and is often associated with a surrounding pigmented or non-pigmented halo. Within the lesion areas of rounded hypopigmentation, so-called “lacunae” can be found. CHRPE appears as a unifocal or multifocal lesion. Rarely, a carcinoma or an adenocarcinoma develops within the lesion. The lesion examined with optical coherence tomography (OCT) was found to have an overlying photoreceptor loss with subsequent retinal thinning and minimal thickening of the RPE. It remains unclear whether the photoreceptor loss is partial or complete. In this case report, we present a unifocal CHRPE lesion examined with high-definition OCT (HD-OCT) to better define its in vivo characteristics.

CASE REPORT
In a routine dilated fundus examination of a 40-year-old woman, a large flat pigmented lesion was found on the left eye, compatible with CHRPE. Photographic documentation of the lesion was taken with a Canon fundus camera (CF-60 UD; Canon, Inc., Tokyo, Japan) and HD-OCT (Cirrus HD-OCT; Carl Zeiss Meditec, Inc., Dublin, CA). Macular cube (512 × 128) and acquisition OCT protocol were used. Retinal thickness was obtained by data analysis of the macular cube acquisition and then compared with that of the right eye, which was obtained by placing the acquisition cube in the inferior retinal quadrant at the same distance from the optic disc. The solitary CHRPE described in our case was in the left eye, located in the inferior retinal quadrant posterior to the equator (Fig. A). Its basal diameter was 5 mm.
and the distances of the margin to the optic disc and to the fovea measured 3.5 and 4.5 mm, respectively. HD-OCT (Fig. B) showed both loss of the junction layer between the inner and outer segment (IS/OS) of photoreceptors and loss of the outer nuclear layer, whereas the inner nuclear layer remained intact. It also showed RPE hyperreflectivity, which mirrors the increased thickness and causes a shadowing of the underlying choroid. Contiguous to the shadowing, the RPE layer was irregular; this feature corresponds to the large lacuna and causes a stronger light transmission into the choroid. The HD-OCT of the fellow eye (Fig. C) showed a regular and less intense RPE reflectivity, inner (thick arrow) and outer (thin arrow) nuclear layers, and defined IS/OS (arrowhead) together with a homogeneous optical transmission into the choroid.

**DISCUSSION**

CHRPE is a benign unifocal or multifocal tumor usually discovered by routine ophthalmologic examination. The first histopathologic features on CHRPE were noted by Duke et al., but the term CHRPE was adopted in 1974 when Buettner described its congenital nature. Other previous histopathologic reports showed the presence of retinal atrophy overlying the lesion. Until now, OCT studies concerning CHRPE could not demonstrate whether the photoreceptor loss was partial or complete. Although a report about CHRPE topography pointed out how typical isolated CHRPE is more frequent in the temporal retinal quadrant, we report a single lesion located in the inferior retinal quadrant. Typically, imaging those lesions with OCT is not easy due to their peripheral location in the fundus, but this has become easier due to the higher speed of the HD-OCT in capturing images. HD-OCT has a resolution of 5 µm, which allows us to distinguish retinal details close to a histological examination and
helps us to understand many retinal diseases. We are now able to identify some other CHRPE features, such as the complete loss of the photoreceptor layer, proved by the loss of both the IS/OS photoreceptors junction and the outer nuclear layer in the retina underlying the lesion, and the non-homogeneous and irregular RPE hyperreflectivity within the lesion due to lacunae. Furthermore, in our case another indirect sign of cell loss was given by volume analysis and central subfield thickness, which were less in the cube containing the lesion when compared with that in the other eye. Because this information is about a single case, its features cannot be generalized to every patient affected by CHRPE. These observations need to be studied in a large population to obtain more data about CHRPE anatomical structure.

REFERENCES