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MULTIMODAL ULTRA WIDE FIELD AND RETRO MODE IMAGING IN A BENIGN CONCENTRIC ANNULAR MACULAR DYSTROPHY

Oral

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Purpose:

We present a case of a benign concentric annular macular dystrophy (BCAMD). BCAMD is a progressive autosomal dominant macular dystrophy characterized by parafoveal hypopigmentation and a bull's eye configuration.

Methods:

A 62-year-old woman presented with blurring of vision in the left eye during the last years, with no history of hemeralopia, nyctalopia, photophobia, no significant family history, no current or recent pharmacological therapies. The patient underwent a comprehensive ocular examination, a multimodal ultra wide field and retro mode imaging, FAF and retinography. We also requested the electroretinogram (ERG), a Goldmann visual field and a genetic counseling.

Results:

BCVA in the right eye (OD) was 20/20 and left eye (OS) was 20/25. Fundus examination showed in both eyes a bull's eye maculopathy with foveal sparing followed by hyperplasia of RPE in the temporal sector in OS. Fundus autofluorescence showed foveal hyperautofluorescence followed by annular hypoautofluorescence and hyperautofluorescence. Spectral domain optical coherence tomography (SD-OCT) of both eyes showed foveal thinning, loss of outer nuclear layer, outer plexiform layer, ellipsoid zone in the parafoveal area, saving a small island of ellipsoid at the fovea (flying saucer sign). Multicolor imaging showing concentric annular areas of hypopigmentation and hyperpigmentation around fovea. ERG was normal. Genetic test was negative.

Conclusions:

BCAMD is a rare macular dystrophy, caused by mutation in the interphotoreceptor matrix. In our case, for the first time, retromodal and ultra wide field imaging allows us to describe BCAMD. Their role was of a fundamental importance to demonstrate the central over functioning of the underlying RPE as the parafoveal RPE was atrophic.