Multimodal imaging of benign concentric annular macular dystrophy

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Key words: Benign concentric annular macular dystrophy, multimodal imaging, spectral domain optical coherence tomography

A 55-year-old woman presented with blurring of vision in the left eye since six months, with no history of hemeralopia, nyctalopia, photophobia, and no significant family history. Best corrected visual acuity in the right eye (OD) and left eye (OS) was 6/12 and 6/36, respectively. Anterior segment examination revealed early cataractous changes in both eyes. Fundus biomicroscopy and color fundus photograph revealed concentric ring of hypopigmentation followed by hyperpigmentation in the parafoveal area prominent in OS than OD [Fig. 1]. Multicolor imaging was superior in delineating the findings compared to color fundus photograph especially in early stages in OD. Fluorescein angiography showed window defect in the parafoveal area in the form of annular hyperfluorescence [Fig. 2]. Fundus autofluorescence showed foveal hyperautofluorescence followed by annular hypoautofluorescence and hyperautofluorescence [Fig. 2]. Spectral domain optical coherence tomography of OD showed foveal thinning, loss of outer nuclear layer, outer plexiform layer, external limiting membrane, and ellipsoid zone in the parafoveal area with collapse of inner retinal layers. A small island of ellipsoid was present at the fovea responsible for good visual acuity. In addition, OS showed retinal pigment epithelium (RPE) atrophy in the parafoveal area [Fig. 3].The result of electrophysiological testing was normal OU [Fig. 4].

Benign concentric annular macular dystrophy (BCAMD) is a rare macular dystrophy, first described by Duetman in 1974.[1] This is caused by mutation in the interphotoreceptor matrix proteoglycan 1 gene on chromosome 6.[2] Good visual acuity is retained till late, which explains the term "benign."[3] In our case, fundus autofluorescence showed central hyperautofluorescence that could be because of the overfunctioning of the underlying RPE as the parafoveal RPE was atrophic.

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With this case, we have described the multimodal imaging of BCAMD, which will contribute in better understanding of the underlying pathology and in differentiating it from other macular dystrophies.

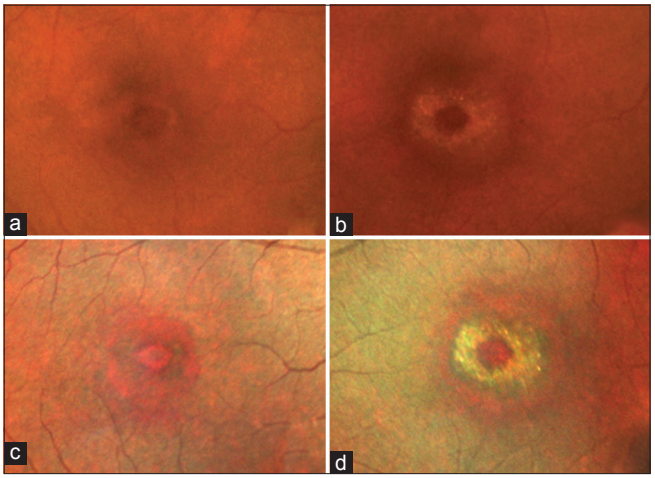

Figure 1: (a and b) Color fundus photograph OD and OS, respectively, showing concentric rings of hypopigmentation followed by hyperpigmentation around hyperpigmented fovea. (c and d) Multicolor imaging OD and OS, respectively, showing concentric annular areas of hypopigmentation and hyperpigmentation around fovea

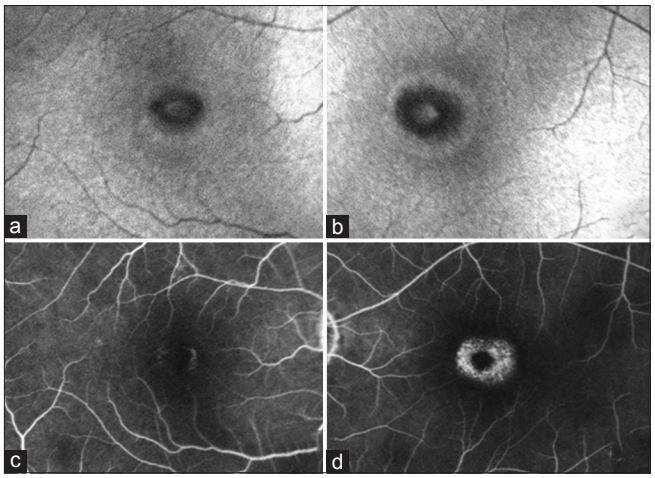

Figure 2: (a and b) Fundus autofluorescence showing central hyperautofluorescence surrounded by concentric rings of hypoautofluorescence and hyperautofluorescence. (c and d) Fluorescein angiography OD and OS, respectively, showing parafoveal annular hyperfluorescence

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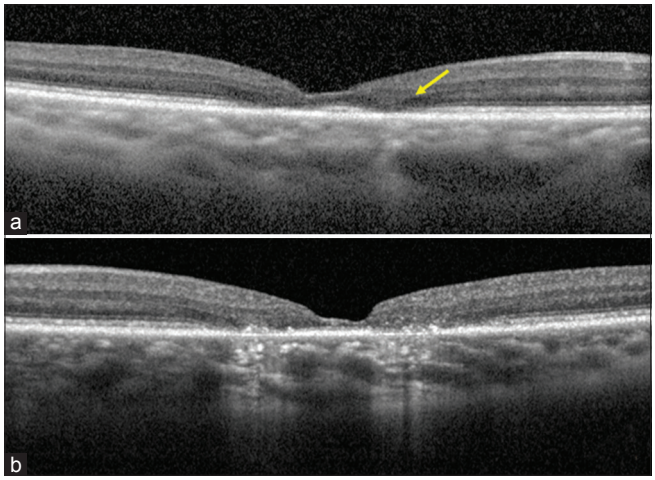

Figure 3: (a) Optical coherence tomography (OCT) of right eye showing foveal thinning, loss of outer nuclear layer, outer plexiform layer, external limiting membrane, and ellipsoid zone in the parafoveal area with collapse of inner retinal layers (arrow) in the parafoveal area. In addition, (b) OCT of left eye shows RPE atrophy in the parafoveal area

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

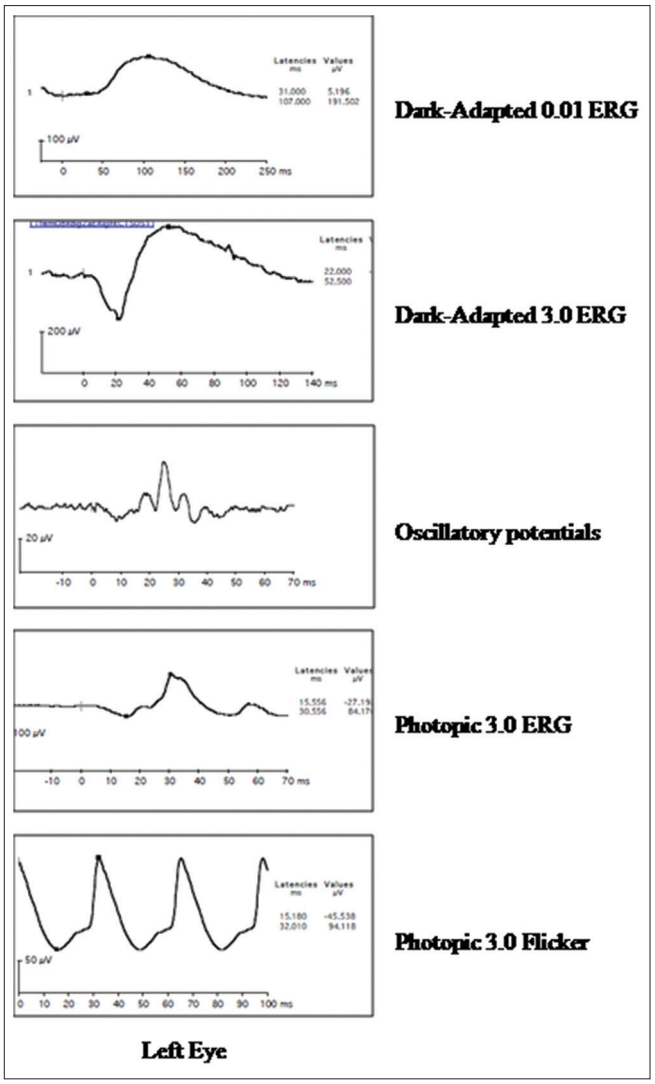

Figure 4: Electroretinogram of left eye shows normal photopic and scotopic response

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