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Sir,

Optical coherence tomography findings in benign concentric annular dystrophy

Benign concentric annular dystrophy (BCAD) is a rare autosomal dominant condition first described by Deutman[1] in 1974. We present the first report of the ocular coherence tomogram (OCT) findings in this condition suggestive of new pathological abnormalities. We also describe the clinical, fluorescein, and electrophysiological findings in what is only the second case ever reported in a British journal.[2]

Case report

A 50-year-old Caucasian woman complained of blurred vision of gradual onset and was noted to have an abnormal macula appearance by her optometrist. She had no symptoms of nyctalopia and gave no history of chloroquine ingestion or the long-term use of other drugs. Her mother and sister were unaffected, but her father had had undiagnosed visual difficulties as an adult and is no longer alive. Remarkably, her best-corrected visual acuity was 6/9 OD and 6/6-3 OS with a low myopic correction. Her colour vision was 11/13 Ishihara plates OD and 9/13 OS. Over a 1-year period, there has been no change in symptoms or signs.

Anterior segment examination and intraocular pressures were normal. Fundoscopy showed bilateral annular hypopigmented areas around each fovea with central sparing. In the right macula, there was a flat well-demarcated pigmented area (Figure 1, top). Less well-demarcated pigment abnormality was seen in the left

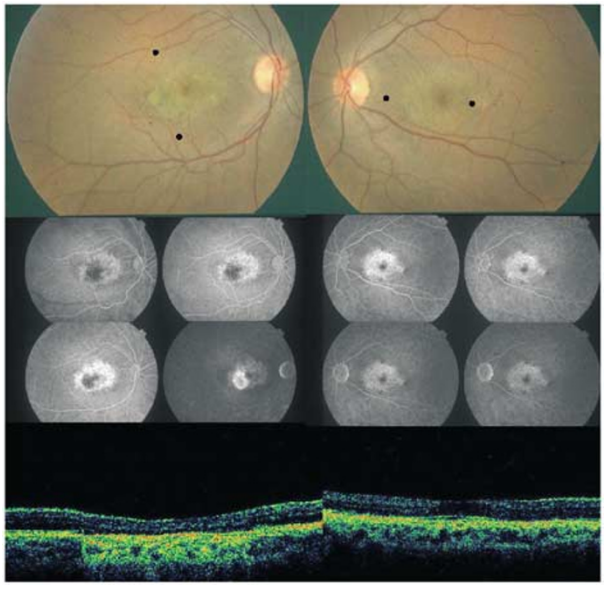

Figure 1 (Top left) Colour picture of right fundus showing pigmented macular lesion and Bull's eye macular dystrophy. (Dots refer to OCT images.) (Top right) Colour fundus photograph of left fundus showing Bull's eye macular dystrophy. (Dots refer to OCT images.) (Middle left) Fluorescein picture of (left) right fundus. and (right) left fundus.(Bottom left) OCT image of right macula on a meridian passing through the pigmented lesion between the two black dots shown on the colour photograph. (Bottom right) OCT image of left macula on a meridian passing through the fovea between the two black dots shown on the colour photograph.

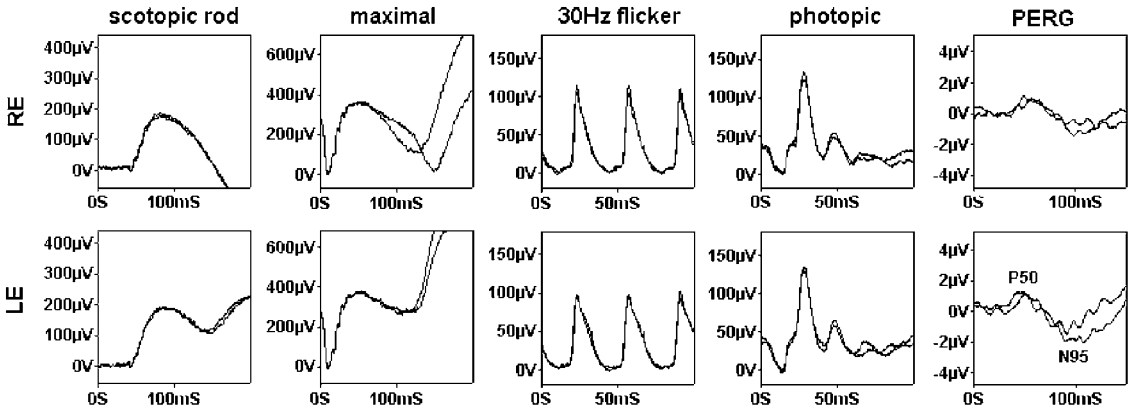

Figure 2 Electrophysiological findings demonstrating a mildly subnormal pattern ERG on the right eye, slightly more abnormal on the left with normal full-field ERGs.

macula again with no clinically detectable retinal thickening (Figure 1, top). Fluorescein angiography confirmed the clinical findings of an annular ring of RPE loss (Figure 1, middle). The peripheral retina in each eye appeared normal.

All full-field flash electroretinograms (ERGs) were within normal limits, but pattern electroretinograms (PERG) were mildly subnormal in keeping with dysfunction confined to the maculae (Figure 2). Electro-oculograms were within normal limits.

Strikingly, optical coherence tomography (OCT3 Zeiss Humphrey Division, Dublin, CA, USA) demonstrated an increased thickness of the RPE in the area of abnormal pigmentation in the right eye with increased reflectivity extending into the choroid (Figure 1, bottom). There was thinning of the neurosensory retina overlying this, from 215 to 141 μm, probably due to loss of the photoreceptor layer. OCT of the left eye, through the fovea, demonstrated an additional line in the region of the RPE similar to that seen in shallow serous elevation of the fovea again with increased reflectivity in the underlying choroid.

Comment

Well-preserved acuity, even at 50 years of age, the retinal and angiographic appearance and mild colour vision and PERG abnormalities suggest that this patient has benign concentric annular (macular) dystrophy (BCAD). Other focal RPE dystrophies such as central areolar macular dystrophy, pattern dystrophy, Stargardt's-like macular dystrophy, pigment epithelial dystrophy, and North Carolina macular dystrophy were excluded either because of the mild acuity loss or retinal appearance.

The family originally described[1] and subsequent cases [2–5]are reported with similar retinal appearances and good visual acuities.[1] The most interesting report on the 10-year follow-up of the originally described cases[1] questioned the 'benign' nature of the condition.[6] This follow-up showed further loss of acuity and colour vision, a bone-spicule peripheral retinopathy, and further decline of ERG responses suggestive of a cone–rod degeneration. A Leu579Pro mutation in the interphotoreceptor matrix proteoglycan 1gene (IMPG1) on chromosome 6 has been suggested as the causative defect in a recent linkage analysis study.[7]

These OCT findings are unusual and to our knowledge have not been described previously in this condition. Abnormal areas of high and low reflectivity under the retinal pigment epithelium are reminiscent of OCT finding seen in adult vitelliform macular dystrophy,[8] where increased reflectivity under the RPE has been attributed to pigment epithelial cell clumping and low reflectivity to fluid accumulation. The relatively normal neurosensory retina at the fovea probably explains the good visual acuity and mild PERG abnormalities. There are as yet no histopathological studies of BCAD with which to compare these results. These novel findings describe pathological changes that will contribute to our understanding of the evolution of BCAD.

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Sir,

Inferior oblique myectomy *vs* recession—its clinical significance

We read with great interest and would like to congratulate Shipman and Burke for their paper comparing the results of inferior oblique myectomy and recession.[1] The homogenicity of their sample population adds strength to their findings. Their results confirm the efficacy of single muscle surgery, but we would like to question their interpretation of the results and the conclusion that 'inferior oblique muscle myectomy may be the procedure of choice giving a better and more predictable long term outcome.' While a 1-year difference between 1.75^{Δ} and 3^{Δ} may be statistically significant, we wonder how clinically significant this is likely to be, given that a difference of 1.25^{Δ} can entirely be attributable to a small change in head positioning.[2] Furthermore, we question the basis of concluding that myectomy has a more predictable outcome. They have shown in Table 2 that the range of hyperdeviation in contralateral gaze at 12 months was much more in the myectomy group (-5 to $+16$) as compared to the recession group (0 to $+9$). This should make recessions more predictable. We are also concerned that there have been some patients with overcorrection in the myectomy group that might represent a group of very unhappy patients, their new eye position going against their long-term head posture. We feel the conclusions have been overstated.

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Sir,

Reply to letter on inferior oblique paper

Patients with symptomatic unilateral right inferior oblique overaction/superior oblique underaction may describe diplopia that is initially confined to levo-elevation or levo-depression, and eventually can progress into primary position. The goal of surgery is to achieve as large a field of diplopia-free vision as is functionally possible without the need to assume a compensatory head position. Ideally, this surgical outcome should not then recede with time.