Macular congenital hypertrophy of retinal pigment epithelium (CHRPE) in a patient with best vitelliform dystrophy (BVD)

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DESCRIPTION

A 28-year-old woman presented with decreased vision in her right eye for the past 6 months. She also reported several years of low vision in both eyes. There was no history of ocular trauma or previous laser photocoagulation. Best-corrected visual acuity (BCVA) was 6/36 in the right eye and 6/18 in the left. Anterior segment was normal in both eyes. There were no signs of intraocular inflammation. Dilated fundus examination showed peripapillary atrophy and a pigmented lesion at the right macula (figure 1A). This lesion contained depigmented lacunae and had a white ring at its outer border. There was a 'scrambled egg appearance' of the left macula, suggestive of a vitelliruptive stage of best vitelliform dystrophy (BVD) (figure 1B). Spectral domain optical coherence tomography (SD-OCT) of the right macula revealed foveal atrophy, hyper-

reflectivity from the retinal pigment epithelium (RPE) hypertrophy with underlying shadowing and loss of outer retinal layers adjacent to the RPE hypertrophy (figure 1C). SD-OCT of the left macula demonstrated neurosensory detachment, elongation of the photoreceptor outer segments and a fibrotic pillar at the level of the RPE (figure 1D). A full-field electroretinogram was within normal limits in both eyes, however, an electro-oculogram showed a reduced Arden ratio in both eyes (1.1 and 1.2 in right and left eye, respectively). A diagnosis of BVD with congenital hypertrophy of retinal pigment epithelium (CHRPE) at the right macula was made.

Solitary macular CHRPE is an uncommon entity and has been reported to comprise only 1% of all cases of CHRPE.¹ The occurrence of macular CHRPE in the setting of BVD has hitherto been unreported.

Figure 1 Fundus photograph of patient with best vitelliform dystrophy. Right eye (A) showing pigmented lesion at macula, left eye (B) showing scrambled egg appearance. Spectral domain optical coherence tomography (SD-OCT) OD (C) showing retinal pigment epithelium hyper-reflectivity and foveal atrophy. SD-OCT OS (D) showing neurosensory detachment and fibronodular pillar.

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Learning points

- ▶ Congenital hypertrophy of retinal pigment epithelium (CHRPE) at the macula is rare and seen in only 1% of cases of CHRPE.
- ▶ A fibrotic pillar on spectral domain optical coherence tomography is a feature of vitelliruptive stage of best vitelliform dystrophy (BVD).
- ▶ Elongation of photoreceptor outer segments is also seen in BVD, in addition to the more common central serous chorioretinopathy.

Competing interests None declared.

Patient consent Obtained.

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REFERENCE

- 1 Shields CL, Mashayekhi A, Ho T, *et al*. Solitary congenital hypertrophy of the retinal pigment epithelium: clinical features and frequency of enlargement in 330 patients. *Ophthalmology* 2003;110:1968–76.

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