preventative treatment, our patient may have benefited from antimigraine prophylaxis given that his vision loss was preceded by hundreds of transient episodes.

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Unilateral retinitis pigmentosa: clinical and electrophysiological diagnosis

Retinitis pigmentosa (RP) is a group of hereditary disorders characterized by gradual atrophy and cell death of the photoreceptors and adjacent cell layers of the retina, the common feature of which is progressive deterioration in vision. Initial presenting symptoms of the disease include night blindness, with gradual deterioration of the light-sensitive cells of the retina, causing diminution of vision in daylight in the later stages. Presentation is bilateral and symmetrical.

Unilateral retinitis pigmentosa (URP) is an uncommon entity involving RP-like changes in one eye with the fellow eye completely unaffected.[1] Proper diagnosis of URP requires that all other pigmentary retinopathies simulating RP be excluded and that the clinical and electroretinography (ERG) findings of RP be well defined in one eye and completely absent in the contralateral eye. The François and Verriest criteria are necessary for the diagnosis of URP: (*i*) occurrence of characteristic findings of RP in one eye; (*ii*) fundus findings and full-field ERG within normal limits in the healthy eye; (*iii*) exclusion of infectious, inflammatory, and vascular reasons for RP-like fundus changes; and (*iv*) a sufficiently long period of observation (>5 years) to rule out delayed onset in the unaffected eye.[2] Herein, we report 3 cases of URP confirmed by characteristic findings seen on ophthalmoscopy, visual field testing, and ERG.

Case Report

Case 1

A 44-year-old female presented for an annual eye examination with the chief complaint of blurring of distance and near vision. She was diagnosed with RP in her right eye (RE) at age 35 years, in 2008 at our institute. Distance acuities with her glasses in place were 20/20 OU. Slit-lamp evaluation of the anterior segment was unremarkable. Posterior segment evaluation of the RE revealed waxy pallor of the optic disc, arteriolar narrowing, and variable amounts of bone-spicule–shaped pigment deposits in the midperipheral retina with sparing of posterior pole (Fig. 1A). The fundus findings of the left eye (LE) were within normal limits (Fig. 1B). Full-field 120-point screening test with a Humphrey perimeter showed a residual small central island of vision in the RE with normal findings in the LE (Fig. 1D, E). The scotopic and photopic flash ERG showed nearly extinguished responses in the RE, with normal amplitudes in the LE (Fig. 1C).

Case 2

A 43-year-old male who was diagnosed with URP 6 years previously visited our institute for an annual eye examination. He had stable distance and near visual acuities in both eyes for 6 years. Best-corrected visual acuity (BCVA) at the time of presentation to us was 20/20 OU. Slit-lamp evaluation findings of the anterior segment were unremarkable. Fundus examination did not reveal any abnormalities in the RE (Fig. 2A). In contrast, fundus evaluation of the LE revealed narrowing of the retinal arterioles, optic disc pallor, and midperipheral retinal pigment clumping (Fig. 2B). The full-field 120-point screening test with a Humphrey perimeter showed normal findings in the RE with a residual small central island of vision in the LE (Fig. 2D, E). The full-field ERG showed a barely detectable rod, cone, and 30-Hz response in the LE with normal response in the RE (Fig. 2C).

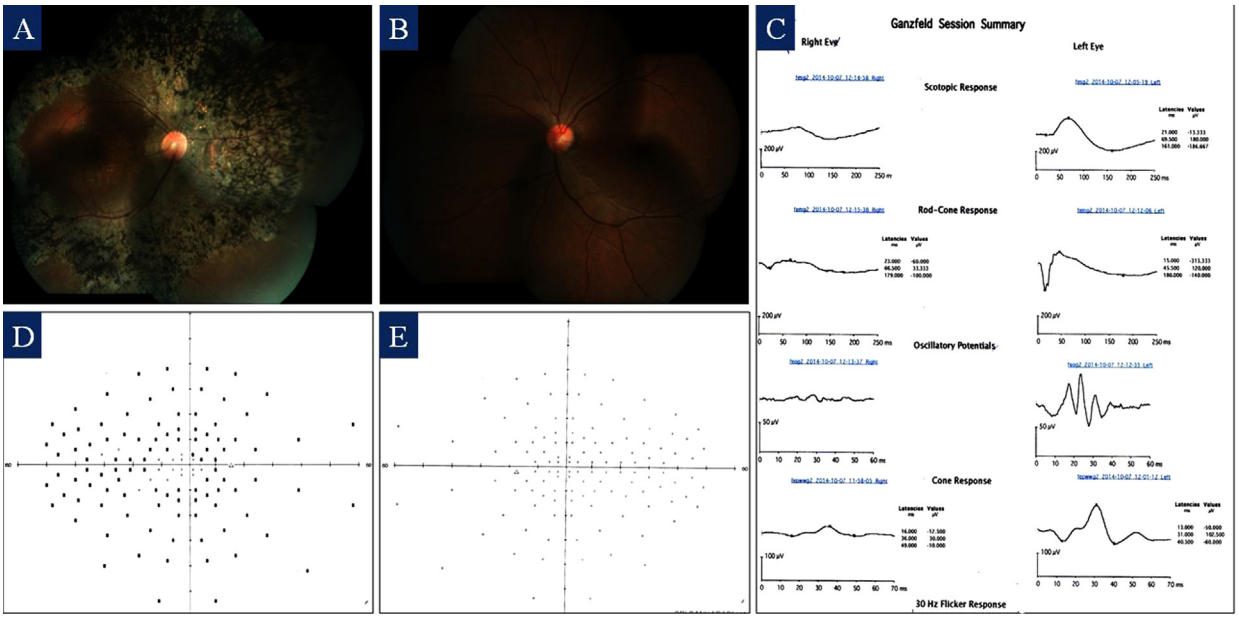

Fig. 1—Colour fundus image of the RE (A) showing RP-like changes; findings of the LE (B) are within normal limits. Result of full-field 120-point screening test with a Humphrey perimeter of the RE (D) revealed loss of peripheral field with a preserved central island, and the LE showed a normal field (E). Electroretinographic record of the RE showed extinguished response with normal amplitude in the LE (C). RE, right eye; RP, retinitis pigmentosa; LE, left eye.

Case 3

A 23-year-old male with a long history of visual disturbance who had been diagnosed with RP in his LE 5 years ago visited our institute for an annual eye examination. His chief complaint was progressive constriction of the peripheral visual field with preservation of a central island of vision in the LE. At the time of last visit, BCVA was 20/20 OU. The biomicroscopic anterior segment examination was normal in BE. Although the characteristic findings of RP were obvious in the fundus examination of the LE, the RE was completely normal (Fig. 3A, B). Despite the restricted visual field with central island in the LE on the computerized full-field 120-point screening test, the RE was totally normal

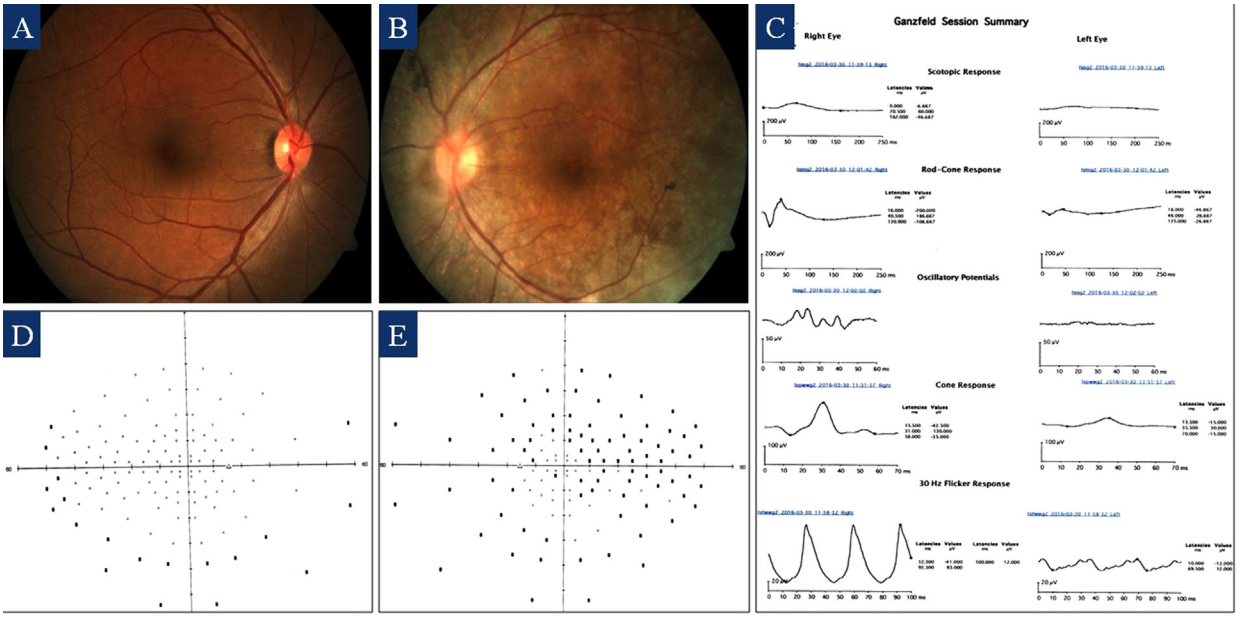

Fig. 2—Colour fundus findings of the RE (A) are within normal limits. The LE shows characteristics of RP (B). Full-field 120-point screening test revealed normal field in the RE (D) and preserved central island in the LE (E). Electroretinographic record of the RE is within normal limits, and the LE shows extinguished response (C). RE, right eye; RP, retinitis pigmentosa; LE, left eye.

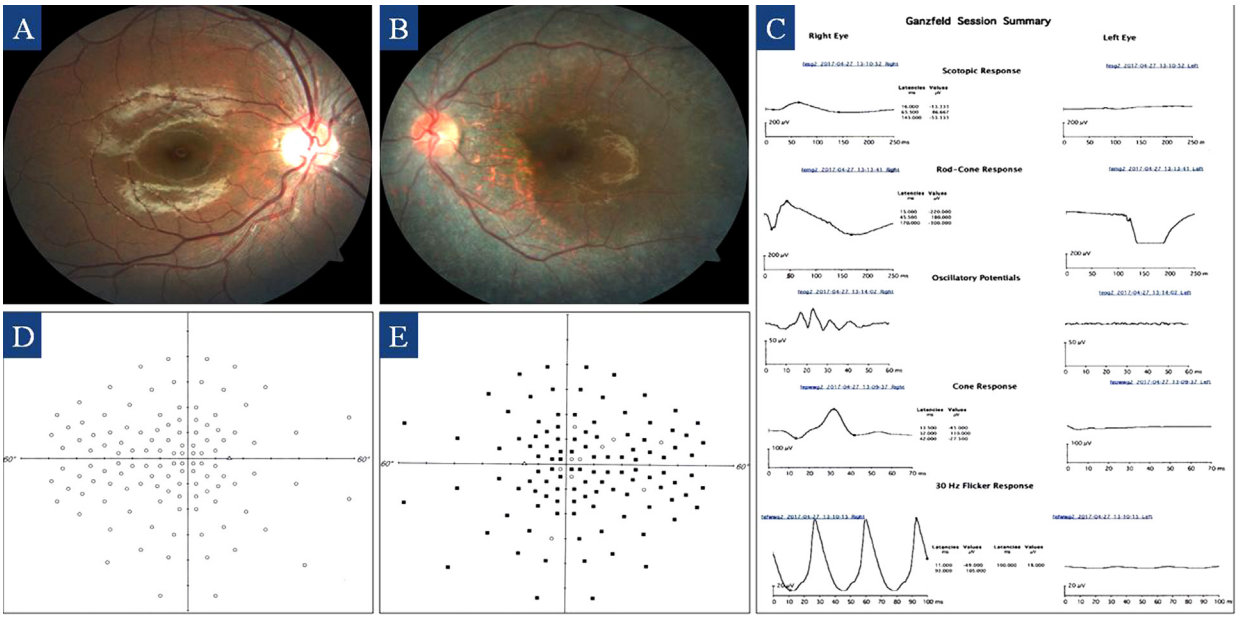

Fig. 3—Colour fundus findings of the RE (A) revealed characteristics of RP, and the LE showed normal fundus (B). Full-field 120-point screening test showed loss of peripheral visual field in the RE (D), and the LE showed normal fields (E). Electroretinographic record of the RE showed extinguished response with normal amplitude in the LE (C). RE, right eye; RP, retinitis pigmentosa; LE, left eye.

(Fig. 3D, E). The full-field ERG showed extinguished responses in the LE with normal amplitudes in the RE (Fig. 3C).

Discussion

Unilateral RP is a rare form of rod–cone dystrophy that was first described in 1948.[3] Retinal dystrophies are usually present bilaterally because of their genetic background, and unilaterality requires an explanation: one pathomechanism is attributable to the occurrence of genetic mosaics (i.e., the mutation affects only some of the cells), and the second mechanism is a somatic mutation instead of a germline mutation. Marsiglia *et al.*[4] reported the cases of 5 patients with URP and found a USH2AW4149R mutation in only one patient. Graff and Stone[5] proposed that URP is the result of a somatic mutation during embryogenesis, causing some groups of cells within the body to carry a gene mutation with the potential to cause RP. If the group of cells destined to become the retina and retinal pigment epithelium is mutant, then the clinical presentation of RP will develop in that eye alone and the fellow eye will remain normal.

The majority of reported cases of URP ultimately are found to have another explanation. The differential diagnosis of URP includes the so-called phenocopies, that is, diseases that mimic the clinical appearance of RP. Therefore, before establishing the URP diagnosis, it is essential to rule out other possible etiologies such as trauma, retained metallic intraocular foreign body, infection (syphilis, toxoplasmosis, rubella, chickenpox, measles, cytomegalovirus), inflammation, retinal toxicity (i.e., chloroquine, chlorpromazine), and vascular- (obstruction of the central retinal artery) and cancer-associated retinopathy in every patient with pigmentary retinopathy that demonstrates disparity between the eyes.[6] The François and Verriest criteria are necessary for the diagnosis of URP.[6]

Unlike RP, there is no proof to suggest that URP is an inherited condition.[7] Fundus examination, visual field testing, and ERG are necessary to confirm the diagnosis. URP appears more frequently in adult patients.[7] We have described 3 cases of unilateral RP, and these cases highlight the importance of combining clinical and electrophysiological testing to elucidate the unilateral pattern of the disease. In our patients, the characteristic signs of RP could be seen: bone-spicule pigmentary changes, pallor of the optic disc, narrowed arterioles, concentric narrowing of the visual field, and nearly extinguished flash ERG in one eye. None of our cases had evidence of trauma, nor did they have a metallic intraocular foreign body. There was also no evidence of prior retinal detachment or any segmental pigmentary changes that would be expected from detached retina in any of our cases. None of the patients had ever had any serious systemic infection, and work-up also ruled out infectious diseases that can affect the retina, such as syphilis, toxoplasmosis, rubella, chickenpox, measles, and cytomegalovirus. Therefore, in this case series, the fundus findings, ERG results, and characteristic progressive loss of peripheral field with a preserved central island observed over the extended period (at least 5 years) made URP a distinct possibility.

To conclude, URP is an uncommon entity, with fewer than 100 cases reported in the literature.[7] Therefore, clinical signs and symptoms, a minimum of a 5-year follow-up period, and confirmatory ERG and visual field testing are very helpful in elucidating the unilateral pattern of the disease and in monitoring these individuals.

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Bilateral isolated choroidal melanocytosis with isoautofluorescence

Ocular melanocytosis is a congenital hyperplasia of the melanocytes in the ocular tissues: episclera, iris, ciliary body, and choroid. It is usually unilateral but can be bilateral. It can affect the surrounding eyelids, as in oculodermal melanocytosis, or be isolated to the choroid, which is referred to as isolated choroidal melanocytosis. Isolated choroidal melanocytosis is an uncommon condition that has been reported unilaterally in a number of cases,[1,2] but bilaterally in only 5 other cases in the literature.[3–6] We present a case of bilateral isolated choroidal melanocytosis in which there is isoautofluorescence in the affected areas, suggesting an overlying healthy retinal pigment epithelium (RPE).

Case Report

A 34-year-old male from Greece presented for a full eye examination. His ocular history included blunt trauma to the right eye 1 year previously. He was not assessed for that injury but did not report any sequelae. His medical history was positive for hearing loss secondary to tympanic membrane damage from otitis as a child. He denied any other health problems and was not taking any medication.

Presenting best-corrected visual acuity was −0.15 logMAR OU. There was no pigment noted on the eyelids, iris, episclera, or sclera of either eye. Intraocular pressures were 12 mm Hg at 2:32 PM in each eye. Dilated fundus examination showed healthy optic nerves with a vertical cup-to-disc ratio of 0.30 in the right eye and 0.45 in the left eye. Examination of the retina revealed a large area of flat, confluent choroidal hyperpigmentation with feathered boarders in each eye. It included the subfoveal area and extended to the ora in both eyes. In the right eye it extended 7 clock hours, and in the left eye it extended 12 clock hours (Fig. 1). Optical coherence tomography (OCT) images (Fig. 2) were captured for each eye and are shown for the left eye. The OCT (Heidelberg OCT Spectralis) showed normal retinal structure over the pigmented areas. Fundus autofluorescence imaging also done with the Spectralis revealed isoautoflourescence over the pigmented areas (Fig. 3).

Discussion

The differential diagnosis for hyperpigmentation in the choroid includes large nevi, bilateral diffuse uveal melanocytic proliferation syndrome, diffuse choroidal melanoma, and isolated choroidal melanocytosis.[3–5,7]

Choroidal nevi are neoplasms derived from uveal melanocytes, which are commonly found on routine examination in 6%–10% of the population.[8] Most are small (1.5–5 mm in diameter), flat, and located posterior to the equator. They have minimal thickness and an irregular border and often have overlying drusen.[8] Nevi usually show hypoautofluorescence but occasionally can have hyper- or isoautofluescence.[9]

Bilateral diffuse uveal melanocytic proliferation syndrome is rare paraneoplastic syndrome that is associated with an occult or previously diagnosed systemic cancer. There is a proliferation of melanocytes in the outer choroid that are unrelated to the primary nonocular tumour by histopathology. The presentation includes decreased visual acuity, small pigment patches with moderate thickening, and focal serous detachment. It shows hyper- and hypoautofluorescence over the affected areas.[10] Prognosis is poor and