

Rare case of simultaneous manifestation of pigmented paravenous retinochoroidal atrophy and retinitis pigmentosa in contralateral eye

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Key words: PPRCA, Retinitis Pigmentosa, CRB1 gene

A 22-year-old female came to our hospital with chief complaints of narrowing of the visual field in her left eye (OS) since 8 years which was progressive, while she had no complaints in the right eye (OD). She gave no history of night blindness. Her best-corrected visual acuity was 20/20 in OD and 20/30 in OS.

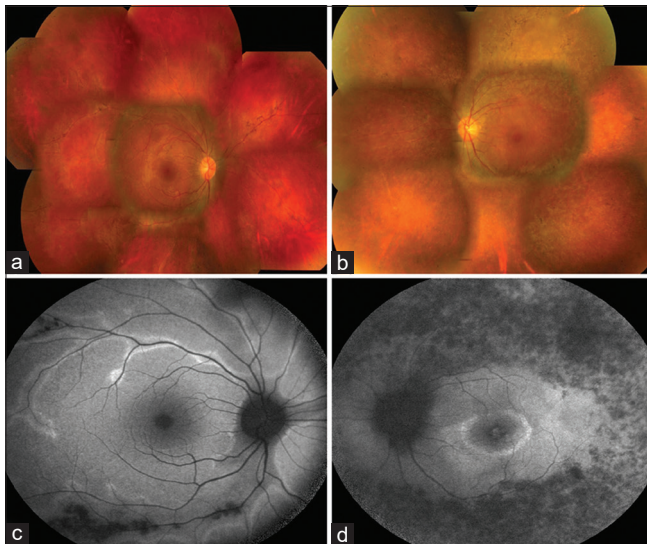


Figure 1: Fundus photography together with autofluorescence (a) showing paravenous retinochoroidal atrophy while (b) shows peripheral retinal pigmented epithelium atrophy with diffuse bone spicule pigmentation. (c and d) Arcs of increased autofluorescence surrounding the area of retinal pigment epithelium atrophy

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Funduscopy examination revealed OD having peripapillary retinochoroidal atrophy (PPRCA)^[1,2] that extends along the retinal veins with bone spicule pigment accumulation distributed mainly at distal parts of the atrophic areas, leaving the macula and the optic disc intact [Fig. 1a and c]. The OS had peripheral retinal pigmented epithelium (RPE) atrophy with diffuse bone spicule pigmentation leaving only a small island of the unaffected macula suggesting retinitis pigmentosa (RP) [Fig. 1b and d].^[3] We observed arcs of increased autofluorescence surrounding the area of RPE atrophy in OD, while parafoveal ring-shaped area of increased autofluorescence in OS [Fig. 1c and d]. Optical coherence tomography demonstrated normal foveal contour and ellipsoid zone in OD; however, OS had cystoid edema with normal ellipsoid zone in foveal region and absent in para-foveal and peripheral to it [Fig. 2]. Visual field examination revealed an enlargement of physiologic blind spot and paracentral arcuate scotoma in OD and a concentric contraction of visual field in OS [Fig. 3]. The OD showed reduced amplitudes (particularly b-wave) and prolonged latencies, while OS demonstrated nonrecordable pattern in the rod and cone electroretinograms [Fig. 4]. Recent genetic research has found CRB1 (Crumbs 1) gene, the mutations within which are associated with both PPRCA and RP.^[4]

Simultaneous manifestation of PPRCA and RP observed in this case is rare^[5] and substantiates that these are sister diseases

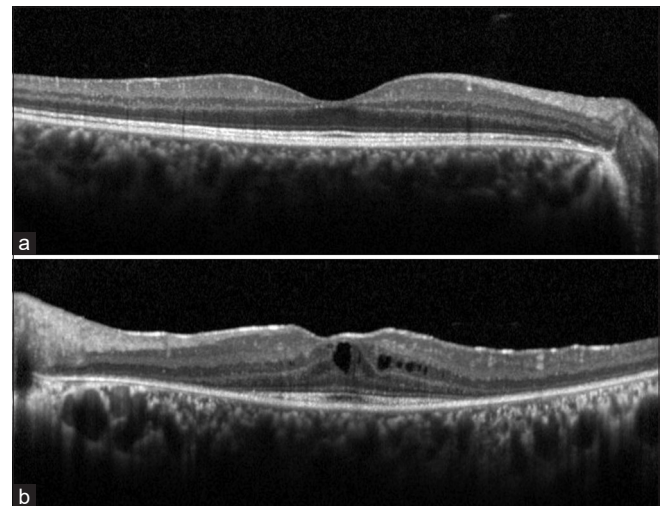


Figure 2: Optical coherence tomography demonstrated normal foveal contour and ellipsoid zone in OD; however, OS had cystoid edema with normal ellipsoid zone in foveal region and absent in para-foveal and peripheral to it

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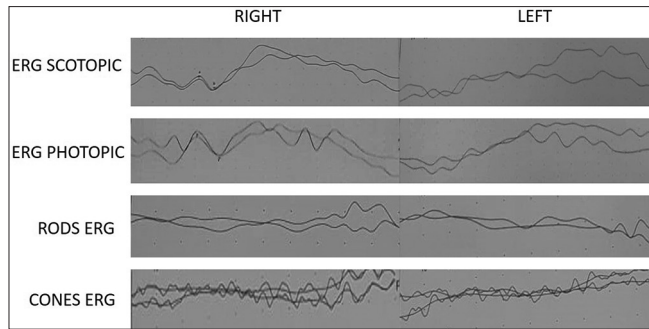


Figure 3: Electroretinogram (ERG). The right eye showed reduced amplitudes, while left eye demonstrated a nonrecordable rod and cone ERG

of the same genetic spectrum. This case also highlights the asymptomatic and nonprogressive nature of PPRCA.

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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Nil.

Conflicts of interest

There are no conflicts of interest.

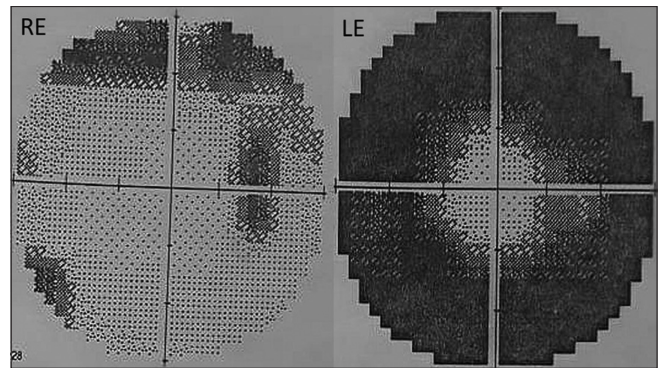


Figure 4: Humphrey visual field: grey scale revealed (RE) paracentral arcuate scotoma and (LE) showed concentric contraction of visual field

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