Pigmented paravenous retinochoroidal atrophy

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Pigmented paravenous retinochoroidal atrophy (PPRCA) is a rare variant of retinochoroidal atrophy (RCA) characterized by bilateral pigment accumulation along the retinal veins and RCA.[1] The disease etiology remains unknown. Majority of the cases are sporadic but some are hereditary in nature.[2]

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Case

A 24-year-old female presented with complaint of diminution of vision in both eyes (OU) for 1 year, with no past history of inflammation, trauma, or nyctalopia and no family history of retinal disorders. Best-corrected visual acuity was 6/6p OU. Biomicroscopic examination of anterior segment was unremarkable OU. Fundus examination showed pigment clumps along the retinal vascular arcade with chorioretinal atrophy extending from the disc up to the equator with no involvement of the macula OU [Fig. 1a and b].

Complete blood count (CBC), erythrocyte sedimentation rate (ESR), C-reactive protein, and random blood sugar levels were within normal limits. Antinuclear antibody (ANA) test was negative and venereal disease research laboratory (VDRL) test was nonreactive. OU fundus autofluorescence showed crescentic area of hyperfluorescence adjacent to area of chorioretinal atrophy [Fig. 1c and d]. Full-field analysis showed peripheral field constriction OU [Fig. 2a and b]. OCT revealed

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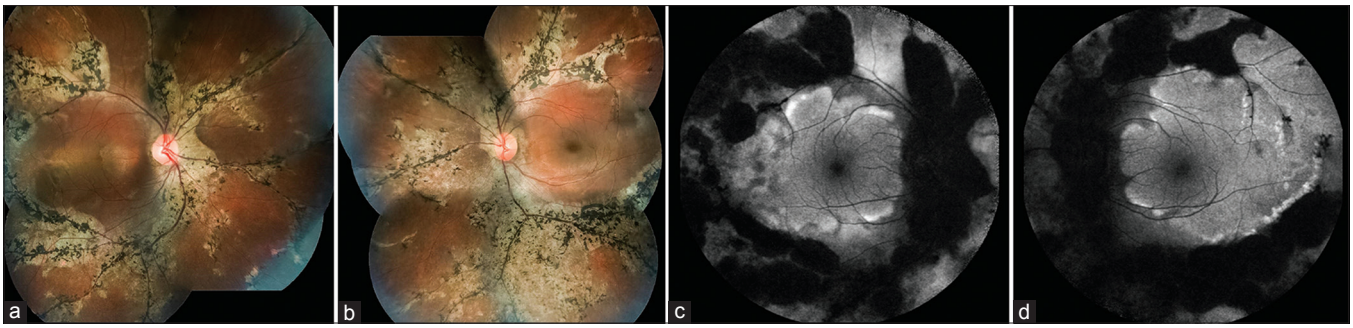

Figure 1: (a and b) Composite color fundus photographs OD and OS, respectively, showing pigment clumps along the retinal veins and chorioretinal atrophy extending from the disc to the equator. (c and d) Fundus autofluorescence photographs OD and OS, respectively, showing hypofluorescence in the areas of chorioretinal atrophy and a crescent-shaped area of hyperfluorescence adjacent to it

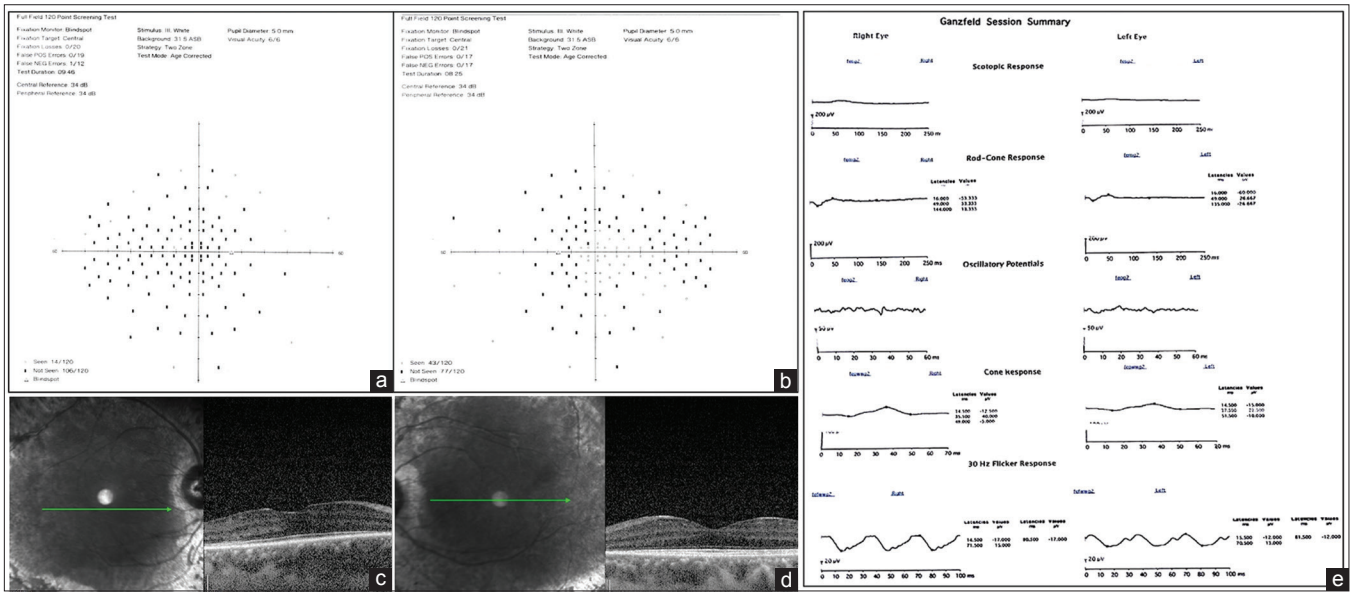

Figure 2: (a and b) Full-field analysis OD and OS, respectively, showing constriction of the peripheral field. (c and d) OCT OD and OS, respectively, showing retinal thinning. (e) ERG OU showing reduced photopic and scotopic responses

retinal thinning OU [Fig. 2c and d]. ERG showed reduced photopic and scotopic responses OU [Fig. 2e].

Discussion

Brown first reported a case of PPRCA in 1937 and termed it as retinochoroiditis radiata.[3] In PPRCA, the retinal pigment epithelium (RPE) damage leads to atrophy of the underlying choroid.[4] It is usually bilaterally symmetric but unilateral cases have also been reported.[5] The etiology remains unknown but mutations of CRB1 (crumbs homolog-1) gene have been implicated in its pathogenesis.[6] Patients usually are asymptomatic. In most cases, visual field may be normal, but some may have scotomas.[1] Fundus shows typical and characteristic pigment accumulation with RCA. Unaffected retina appears normal. Macula may be involved in some cases.[1] The differential diagnoses include chorioretinal degeneration and inflammatory diseases that cause chorioretinal atrophy. Yanagi *et al.* showed that the disease remains stationary in younger patients and slowly progress in older subjects.[7] Currently, no specific treatment modality is available for PPRCA. This photo essay highlights the salient diagnostic features in macular-sparing bilateral PPRCA in a young female.

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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.

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