

## CASE REPORT

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# Macular Neovascularization in a Patient with Pigmented Paravenous Chorioretinal Atrophy

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**ABSTRACT** A 60-year-old male patient admitted to our clinic with complaints of decreased vision, nyctalopia, and metamorphopsia in both eyes, mainly in the left eye. The best-corrected visual acuity was determined as 20/25 on the right and 20/200 on the left eye with the Snellen chart. Anterior segment examination was normal. Fundus examination revealed chorioretinal atrophic areas and bone spicules around all vascular arcades in both eyes. Optical coherence tomography displayed normal foveal architecture in the right eye; however, retinal pigment epithelial detachment, intraretinal cysts, subretinal fluid, and subretinal hyperreflective material were detected in the left eye. Examination findings and fluorescein angiography were supported by visual field and electrophysiological tests, and the patient was diagnosed as pigmented paravenous chorioretinal atrophy (PPCRA) with Type 1 macular neovascularization. We aimed to present the co-occurrence of PPCRA and macular neovascularization to the best of our knowledge, which has not been reported previously.

**Keywords:** Pigmented paravenous chorioretinal atrophy; macular neovascularization; retinitis pigmentosa; bone spicules

Pigmented paravenous chorioretinal atrophy (PPCRA) is a rare form of chorioretinal atrophy with bilateral paravenous retinal pigment epithelial atrophy and pigment aggregation.<sup>1</sup> PPCRA usually progresses slowly.<sup>2</sup> Diagnosis is based on the characteristic appearance on fundus examination, and detailed retinal imaging and retinal electrophysiology support the diagnosis.<sup>3</sup> PPCRA patients may have minimal or no electroretinogram (ERG) response under standard recording procedures.<sup>4</sup> The visual field may be normal, or the affected areas may be variable depending on the pigmentation and atrophic topography.<sup>3</sup> The underlying basis of PPCRA is not known exactly, and studies suggest that there may be inflammatory, infectious, or genetic causes.<sup>5</sup> The association of PPCRA with cystoid macular edema, epiretinal membrane, macular depigmentation, lamellar macular hole, macular retinal pigment epithelium (RPE) atrophy has been shown in the literature; how-

ever, this is the first case report that is showing its association with the macular neovascularization.<sup>3</sup>

## CASE REPORT

A 60-year-old male patient admitted to our clinic with complaints of decreased vision, nyctalopia, and metamorphopsia in both eyes, mainly on the left. In ophthalmological examination, the best-corrected visual acuity was found to be 20/25 in the right eye and 20/200 in the left eye using the Snellen chart. Anterior segment examinations of both eyes were normal, and intraocular pressure values were within normal limits. Fundus examination revealed narrowing and sclerosis in arterial structures in both eyes, decreased vein diameters, chorioretinal atrophic areas around all vascular arcades, bone spicules in the peripheral retina, and both optic discs found to be slightly pale (Figure 1). On optical coherence tomography (OCT),

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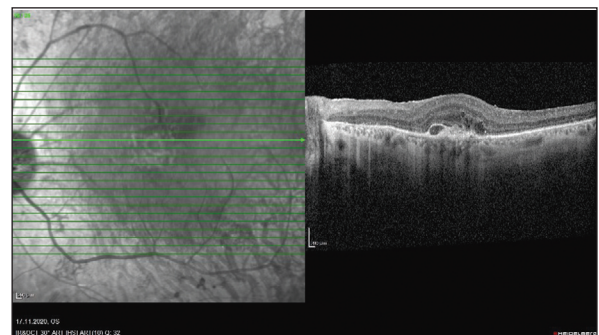
retinal layers were observed as normal except for the presence of an epiretinal membrane in the right eye. However, retinal pigment epithelial detachment (PED), intraretinal cyst, subretinal fluid, and subretinal hyperreflective material were detected in the left eye with OCT (Figure 2). Color fundus photograph, fundus fluorescein angiography (FFA) and fundus autofluorescence imaging, visual field, and ERG were performed to support the diagnosis. Fundus autofluorescence imaging showed hypoautofluorescence lesion along the temporal and nasal vascular arcades in areas compatible with the color fundus photograph in both eyes. FFA showed choroidal vessels around the vascular arcades due to window defect secondary to atrophy in the RPE in both eyes and a hypofluorescent appearance secondary to the blockage due to bone spicules was present in the retinal periphery. In addition, a hyperfluorescent area was observed in the left eye, consistent with the Type 1 macular neovascularization, with leaking of the borders in the late phase (Figure 3). Our patient had visual field loss in the form of progressive arcuate scotoma from the temporal side. There were no waves in the flash ERG and the test was considered pathological. When the patient's complaints and examination findings were evaluated together, the patient was diagnosed with the Type 1 macular neovascularization of the left eye secondary to PPCRA. For Type 1 macular neovascularization (MNV) detected in the left eye, the patient was initially injected with 3 doses of bevacizumab at monthly intervals. Macular neovascularization of the patient did not regress although receiving the 7 doses of ranibizumab injections at monthly intervals after bevacizumab enjections. The patient's anti-vascular endothelial growth factor treatment is continued at regular intervals. Consent was obtained from the patient for the publication of these findings and images.

## DISCUSSION

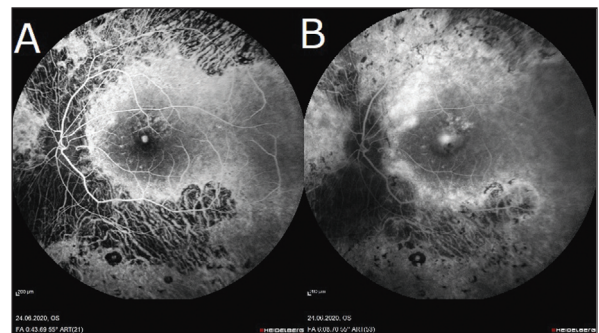
PPCRA is a rare retinal dystrophy with atrophic areas around the vascular arcades.<sup>1</sup> Although the etiology is unknown, inflammatory, infective, and genetic causes are blamed in pathophysiology of PPCRA.<sup>5</sup> Recent genetic studies have revealed that crumbs homolog 1 gene mutations are associated with PPCRA



**FIGURE 1:** Arterial narrowing and sclerosis, decrease in vein diameters, chorioretinal atrophic areas around all vascular arcades, and bone spicules in peripheral retina in both eyes.



**FIGURE 2:** Image displayed pigment epithelial detachment, intraretinal cyst, subretinal fluid, and subretinal hyperreflective material in the left eye.



**FIGURE 3:** A hyperfluorescent lesion with a clear borders corresponding to Type 1 macular neovascularization was detected in early phase of fundus fluorescein angiography. B) In the late phase, borders of the macular neovascularization blurred secondary to leakage.

and retinitis pigmentosa (RP), and these diseases have similar genetic spectra.<sup>5</sup> Ratra et al. reported a patient with PPCRA in one eye and RP in the other, trying to prove that these 2 diseases are in the same genetic spectrum.<sup>6</sup> The macular neovascularization is a very rare complication of RP, and there are limited

studies that showing this association.<sup>7-10</sup> To the best of our knowledge, there is no study showing the relationship between PPCRA, which is associated with many different macular findings, and macular neovascularization.<sup>3</sup> Retinal pigment epithelial changes, photoreceptor cell involvement, and choriocapillaris damage are constantly present in RP, and the macular neovascularization in the presence of these pathologies can always occur.<sup>8</sup> We think that the development mechanism of the macular neovascularization in PPCRA is similar to that in RP in our case of these 2 diseases belonging to a similar genetic spectrum. In 2 different cases of MNV due to RP, patients were given intravitreal bevacizumab injection, and regression was observed in MNV in the follow-ups.<sup>7,8</sup>

In our case report, the macular neovascularization was detected in the left eye of the patient due to the presence of retinal PED, drusen, and subretinal hyperreflective material in the left eye OCT in addition to chorioretinal atrophic areas around the entire vascular arch in both eyes and bone spicules in the peripheral retina. Due to the absence of drusen and PED in the right eye OCT imaging, it was concluded that the development of choroidal neovasculariza-

tion secondary to PPCRA was more likely than the association of PPCRA and exudative age-related macular degeneration in our patient. The patient whose MNV did not regress received 7 doses of ranibizumab injections at monthly intervals. However, the patient whose lesion did not regress after the injections is still receiving long-term treatment. We aimed to present our case, whose long-term treatment continues, because of this association, which is rarely found in the literature.

### Source of Finance

*During this study, no financial or spiritual support was received neither from any pharmaceutical company that has a direct connection with the research subject, nor from a company that provides or produces medical instruments and materials which may negatively affect the evaluation process of this study.*

### Conflict of Interest

*No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.*

### Authorship Contributions

*All authors contributed equally while this study preparing.*

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