Pigmented Paravenous Retinochoroidal Atrophy: Case Report

Pigmente Paravenöz Retinokoroidal Atrofi

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ABSTRACT A 60 year-old female admitted to our clinic after experiencing difficulty in near vision. Examination revealed a best corrected visual acuity of 10/10 in both eyes. The intraocular pressures were in normal limits. Slitlamp evaluation revealed normal anterior segments. Fundus examination revealed retinal pigment epithelium (RPE) atrophy symmetrically in both eyes along the courses of the retinal veins. Fluorescein angiography (FA) showed hyperfluorescence in the areas with RPE atrophy and hypofluorescence in the pigmented areas due to the masking effect of RPE. Optical coherence tomography revealed normal anatomy of the macula in both eyes. There were various absolute and relative scotomas in the visual field (Humprey) in the corresponding paravenous areas. The electroretinogram showed subnormal responses in both eyes. The serology of toxoplasma, syphilis and cytomegalovirus CMV were negative. Upon these clinical findings patient was diagnosed as pigmented paravenous retinochoroidal atrophy. During the 3-year follow up period, patient`s clinical findings remained stable.

Key Words: Retinal pigment epithelium; visual fields

ÖZET Altmış yaşında kadın hasta kliniğimize yakın okuma gözlüğü için başvurmuştu. Yapılan oftalmoskopik muayenede her iki göz görmeleri tashihle 10/10 düzeyinde idi. Ön segment muayeneleri her iki gözde doğal olup, göz içi basınçları normal sınırlar içerisinde bulundu. Fundus muayenesinde, paravenöz alanda her iki gözde de simetrik olarak retina pigment epiteli (RPE) atrofisi ve pigmentasyon izlendi. Yapılan fundus flöresein anjiyografi incelemesinde RPE atrofisi olan alanlarda hiperflöresans ve pigmentasyon alanlarında ise blokaja bağlı hipoflöresans izlendi. Optik koherens tomografi incelemesinde makular anatomi doğal idi. Humphrey görme alanı tetkikinde atrofik paravenöz alanlara uyan rölatif ve absolu skotomlar saptandı. Yapılan elektrofizyolojik testlerde ise her iki gözde subnormal cevaplar alındı. Elektroretinogram cevabında üçte birlik bir azalma saptanırken rod cevabında %50'lik azalma mevcut idi. Hastanın serolojik testlerinde sifiliz, tokoplazma ve sitomegalovirüs ait sonuçlar negatif bulundu. Hastaya mevcut bulgularla pigmente paravenöz retina koroidal atrofi tanısı kondu. Hasta bu tanı ile kliniğimizce 3 yıldır periyodik aralıklarla takip edilmekte olup kliniğinde herhangi bir progresyon izlenmedi.

Anahtar Kelimeler: Epitelyumun retinal pigmenti; görme alanları

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Pigmented paravenous retinochoroidal atrophy (PPRCA) was firstly described by Brown in 1937. The characteristic findings in PPRCA are bilateral, symmetrical retinal pigment epithelium (RPE) atrophy along the course of the veins and variable pigment migration adjacent to these areas. The etiology is unknown and the diagnosis is made based on the fundus findings. Since the patients are asymptomatic, they are diagnosed during routine ophthalmolo- gical examination.[1] The diagnosis of this rare disease is challenging most of the time because many ophthalmological conditions may cause similar fundus findings. In addition, sporadic character and male predominance make the diagnosis more difficult in female cases.[2,3] We present here a PPRCA case diagnosed in a lady below.

CASE REPORT

A 60 year-old female admitted to our clinic after experiencing difficulty in near vision. The patient did not have any systemic disease or any drug to be used for a long time. There was not any trauma in the history of the patient. Examination revealed a best corrected visual acuity of 10/10 in both eyes. The intraocular pressures were in normal limits. Slitlamp evaluation revealed normal anterior segments. Fundus examination revealed RPE atrophy symmetrically in both eyes along the courses of the retinal veins. The optic nerve, macula and retinal vessels were normal in both eyes (Figure 1). There was no any inflammation finding in the vitreous. The patient did not complain about any night vision problem (nyctalopia). The color vision test performed by Ishihara coloured plates was normal in both eyes.

Fluorescein angiography (FA) showed hyperfluorescence in the areas with RPE atrophy and hopofluorescence in the pigmented areas due to the masking effect of RPE. FA did not show any macular pathology (Figure 2). Optical coherence tomography (OCT) revealed normal anatomy of

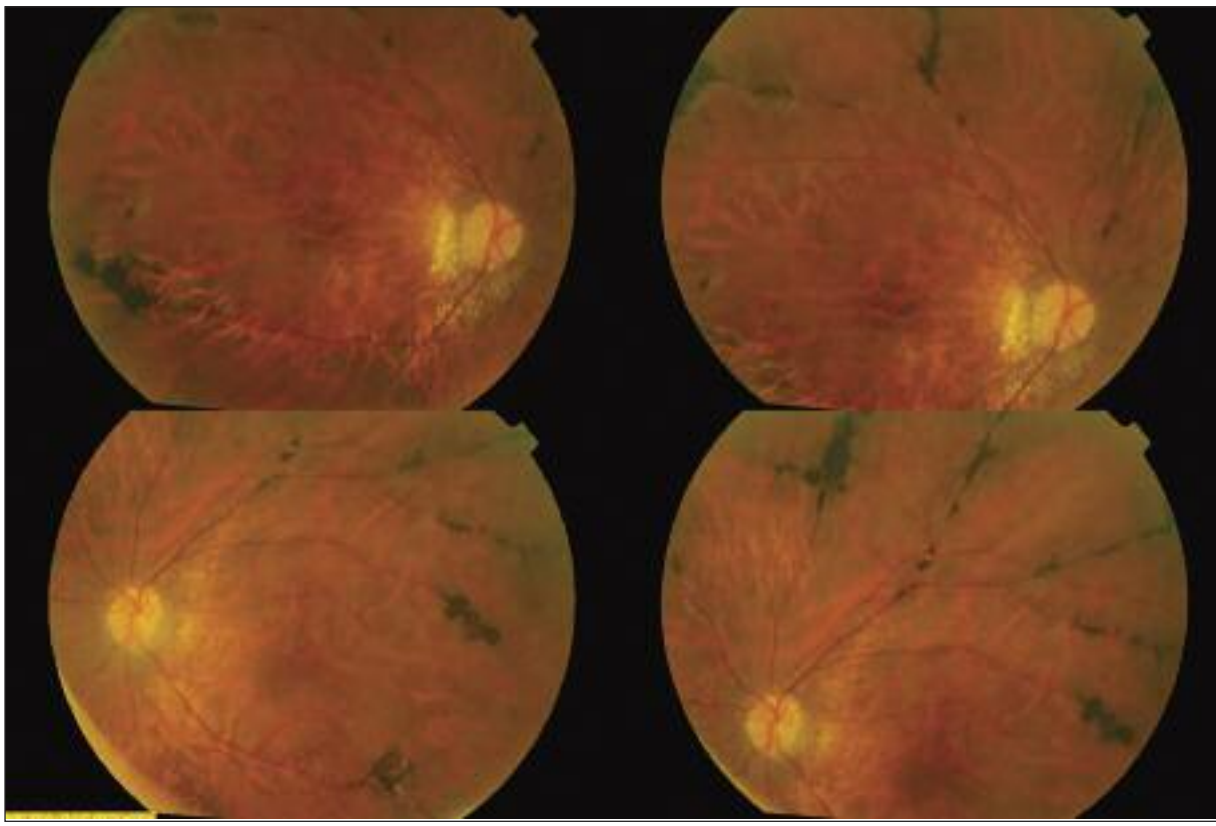

FIGURE 1: Colour fundus photos of the right and left fundi. Note typical areas of paravenous atrophy and pigment accumulation along the major vessels.

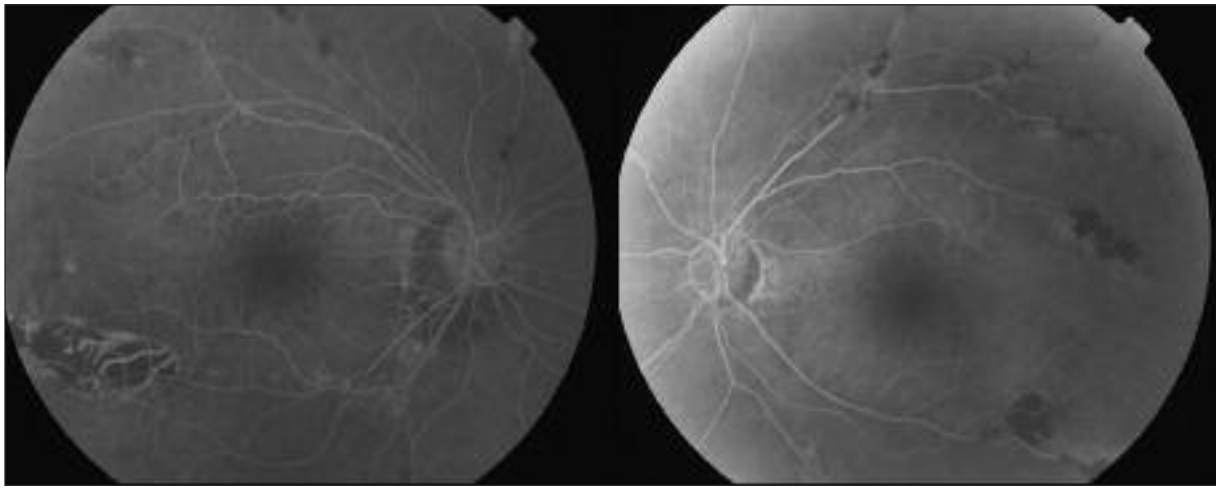

FIGURE 2: FA of the right and left fundi. Note window defects along the major vessels due to RPE atrophy and hypofluorescence due to pigment clumping.

the macula in both eyes. There were various absolute and relative scotomas in the visual field (Humphrey) in the corresponding paravenous areas (Figure 3 and 4). The electroretinogram (ERG) showed subnormal responses in both eyes. The serology of toxoplasma, syphilis and cytomegalovirus CMV were negative. Upon these clinical findings patient was diagnosed as PPRCA. During the 3-year follow up period, patient's clinical findings remained stable.

DISCUSSION

Pigmented paravenous retinochoroidal atrophy was firstly described by Brown in 1937 as retinochondroiditis radiata. Thereafter, the name of the disease was changed to PPRCA in 1962 by Franchesetti. PPRCA is an atypical form of retinitis pigmentosa. Some authors think that there is a congenital origin for this disease; some others think that this is a primary retinal degeneration.[3]

In the literature most of the reported cases are sporadic in nature.[1,3] On the other hand, it was thought that inflammatory mechanisms might play a role in the pathophysiology due to findings of some early reported cases. For instance, there was a patient with tuberculous spondylitis and another case with congenital syphilis.[4] Also, Batioglu et al showed that inflammatory causes may have a role in the etiology of PPRCA.[5] Although exact pathophysiological process remains to be elucidated, clinical specifications of this disease, both related to diagnosis and prognosis, are well described.

The presented case was a female and her ophthalmological examination showed normal anterior segments examination findings, without any findings related to ocular inflammation. In the presented case there was not any inflammatory cause and genetic predisposition for PPRCA. This case was accepted as a sporadic similar to the majority of the cases reported in the literature. Patients with PPRCA are usually asymptomatic and the levels of the visual acuities are at normal limits. The majority of cases reported in the literature are young males. Of these, only a small percentage is female.[3,6,7] For this reason we think that our female case has a clinical significance due this rarity of PPRCA in female gender.

The fundus examination revealed findings in close association with PPRCA such as symmetrically RPE atrophy along the courses of the retinal veins in both eyes. On the other hand, there were no other additional ocular pathologies involving

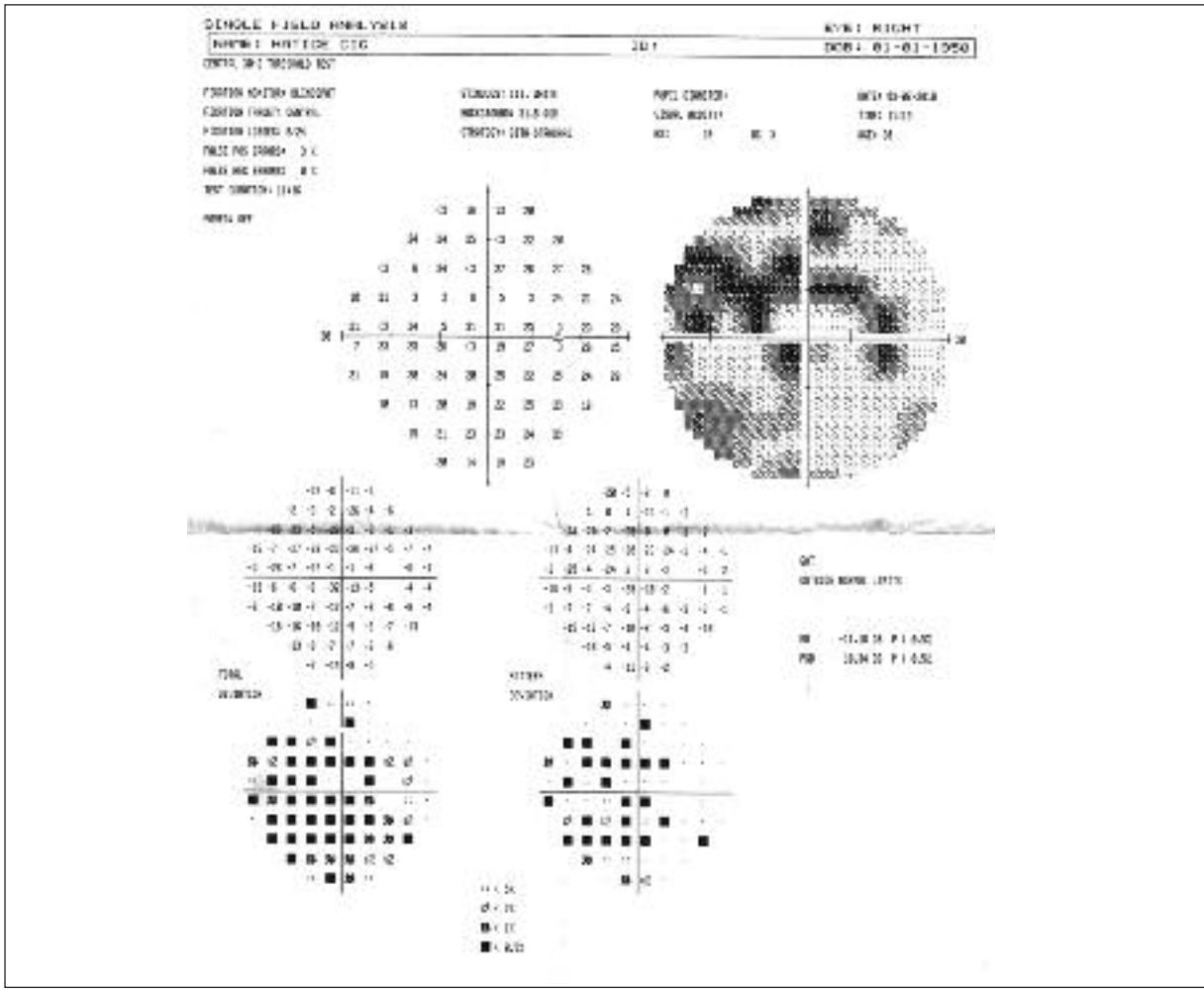

FIGURE 3: The visual field analyser (Humphrey) showed various absolute and relative scotomas in the corresponding paravenous areas (OD).

the macula or any other part of the retina. It was reported that there is atrophy of RPE along the course of large retinal veins in PPRCA. In most of the cases, the optic disc and the calibration of the retinal vessels is normal.[1,4] In some cases, in addition to RPE atrophy, there may be also atrophy of the choriocapillaris.[1,4] In a small percentage of the cases there may be involvement of the macula that is the reason of decreased visual acuity.[4,6] In severe cases, macular involvement is similar to gyrate atrophy and central areolar atrophy. Angioid streaks, helicoidal peripapillary degeneration, syphilis, sarcoidosis, tuberculosis, toxoplasma and CMV chorioretinitis, sectorial retinitis pigmentosa, pseudoretinitis pigmentosa must be differentiated from PPRCA.[3,4,8] The differential diagnosis from these diseases is made based on the typical fundus findings and the symmetrical involvement of both eyes.[4] In some reports it has been shown that in addition to classical findings of PPRCA there is also macular coloboma, drusen of the optic nerve head and retinal microangiopathy.[3]

FA showed hyperfluorescence in the areas with RPE atrophy and hopofluorescence in the areas with pigment clusters and these findings support PPRCA diagnosis. In addition, the visual field examination of the presented case showed scotomas corresponding to the RPE atrophy similar to other cases reported in the literature.[3] Although ERG of

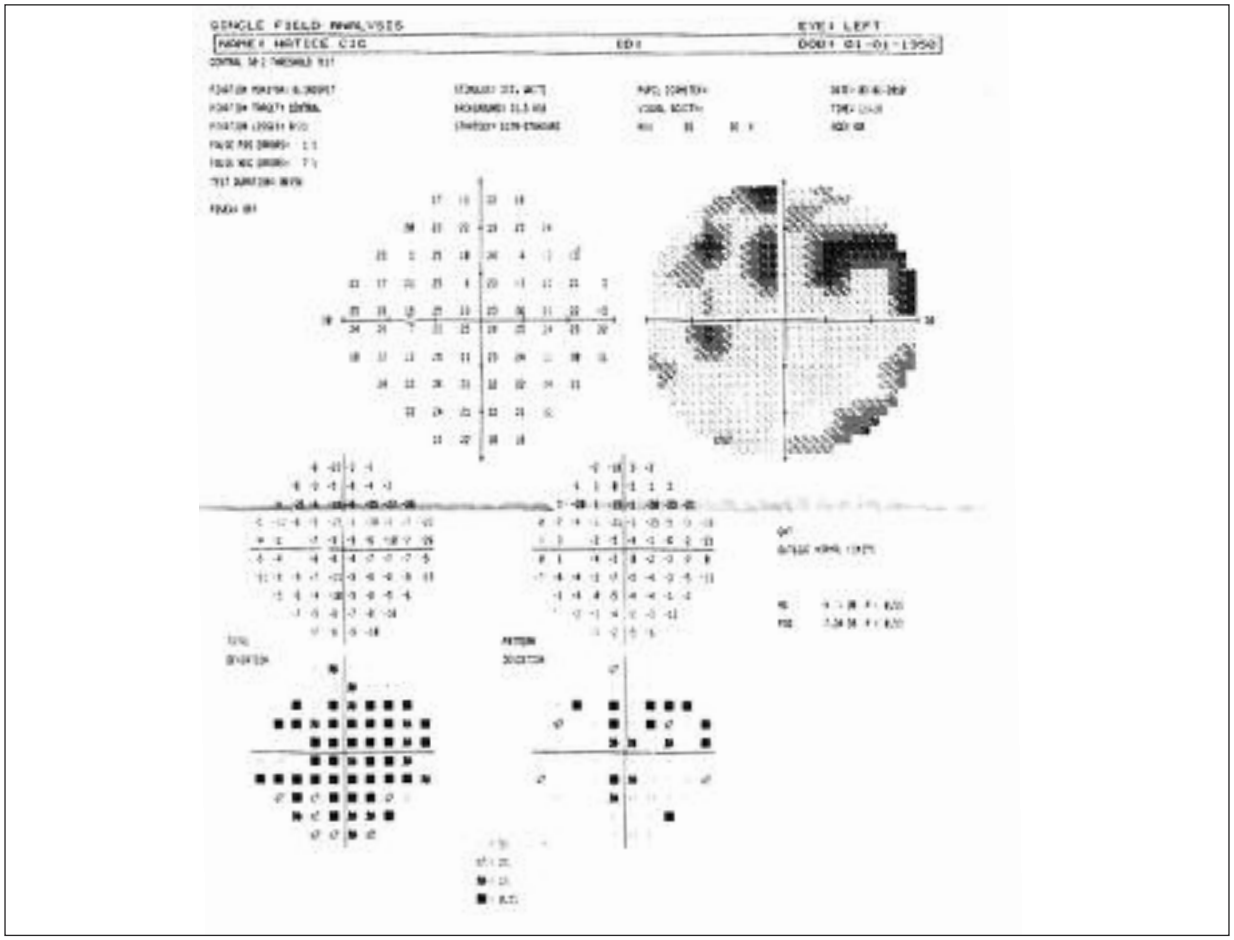

FIGURE 4: The visual field analyser (Humphrey) showed various absolute and relative scotomas in the corresponding paravenous areas (OS).

most of the cases reported in the literature was in normal range, the reported case's ERG examination showed subnormal responses in both eyes.[3] It is well known that ERG responses show deterioration if a substantial percentage of retinal photoreceptors become damaged. For this reason reported ERG response changes found in PPRCA are not even. Subnormal responses observed in this case imply such an extensive disturbance.

There was no progression in the clinical findings and no decrease in the visual acuity in a 3-year follow up time in our case. However, there may be progression in the clinical findings of some patients whose lesions progress to the posterior pole and coalesce around the optic disc.[9] In one of 3 cases that Pearlman and one of six cases that Noble and Carr reported there was progression in the clinical findings.[4,8] The progression of the RPE atrophy usually originates from the inner part of the retina to the surface of the veins. Follow-up period is three years in this case and lack of any progression sign hints that this case may remain stable. But every case must be evaluated separately, taking account its own specific conditions, in order to suppose a prognosis in PPRCA cases.

In conclusion, PPRCA is a rare clinical entity and the mechanism of the disease is not very well understood. As the presented case showed, in cases with no macular involvement, deterioration in the visual acuity is not expected to be a high probability.

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