ISSN: 0975-833X

Available online at http://www.journalcra.com

International Journal of Current Research
Vol. 17, Issue, 08, pp.34369-34371, August, 2025
DOI: https://doi.org/10.24941/ijcr.48727.08.2025

INTERNATIONAL JOURNAL OF CURRENT RESEARCH

RESEARCH ARTICLE

PIGMENTED PARAVENOUS CHORIORETINAL ATROPHY MISDIAGNOSED AS SERPIGINOUS CHORIORETINITIS

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ARTICLE INFO

Article History:
Received 11th May, 2025
Received in revised form
24st June, 2025
Accepted 19th July, 2025
Published online 30th August, 2025

Keywords:
Pigmented Paravenous Chorioretinal Atrophy, Serpiginous Choroiditis, Chorioretinal Atrophy, Ophthalmology, Retina, Uveitis, Ocular disease.

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ABSTRACT

Pigmented paravenous chorioretinochoroidal atrophy (PPCRA) is a rare iatrogenic disorder, characterized by pigment accumulation along the distribution of retinal veins. The findings are usually incidental with minimal effect on vision. This report presenting a patient of PPCRA associated with rod dystrophy misdiagnosed as Serpiginous Chorioretinitis (SC)." In page 2, Figure 1: replace (left sided) with (upper) and (right sided) with (lower).

Citation: Masood Bagheri. 2025. "Pigmented Paravenous Chorioretinal Atrophy Misdiagnosed as Serpiginous Chorioretinitis". *International Journal of Current Research, 17, (08), 34369-34371.*

INTRODUCTION

Pigmented paravenous chorioretinal atrophy (PPCRA) is a relatively uncommon, bilaterally symmetrical stationary disease of unknown origin, characterized by chorioretinal atrophy (CRA) affecting retinal pigment epithelium (RPE) and choriocapillaris with retinal pigmentation, presenting bone spicule pigment clumping in paravenous distributions [1]. Usually, it is described as sporadic and non-progressive or slowly progressive, generally without macular involvement. However, there are also very few hereditary cases or macular involved in the literature [2]. PPCRA has appeared in the literature under the appellations of Pigmented paravenous retinochoroidal atrophy (PPRCA). As hypothesized and proven in this study, choroidal atrophy in the paravenous region occurs before RPE and outer retinal involvement. Thus, PPCRA is a more accurate terminology, with choroidal circulation being the primarily affected component, and retinopathy developing as the secondary changes. We aimed to report a case of PPCRA associated with rod dystrophy. To the best of the authors' knowledge, based on the extensive literature review, it is the first report presenting a case of PPCRA associated with rod dystrophy. The patient was misdiagnosed with serpiginous choroiditis (SC), revealed by precise examination of the multimodal imaging. The case was a 46-year-old Iranian man presenting for follow-up with a diagnosis of SC who was treated with corticosteroid and immunomodulator therapy. He mentioned that his visual disturbance started 12 years ago with painless, progressive diminution of vision of insidious onset in both eyes. He was diagnosed with SC 5 years ago and treated with prednisolone and cyclosporine. At a previous follow-up visit a month ago, he was treated with periocular corticosteroid injection (trans-septal triamcinolone acetonide) after being diagnosed with active disease and cystoids macular edema (CME) on spectral domain optical coherence tomography (SD-OCT). The patient had an unremarkable hereditary family history of eye diseases, systemic condition or trauma. No immunological or hematological evidence of infectious or inflammatory disease was found in the patient in examination tests. The patient's chest radiograph was normal, and his tuberculin skin test, syphilis, toxoplasmosis, and cytomegalovirus serology were negative. The last ophthalmic examination revealed a normal anterior segment examination with pink and sharp optic discs in the posterior segment with a widespread region of CRA along the retinal vessels in both eyes; but, the macula was spared. Multiple areas of bony spicule pigmentation were noted along the veins with mildly vascular attenuation (Figure 1). This feature was mistaken with pseudopodal CRA and misdiagnosed with SC in previous examinations. Multimodal imaging helped us make the accurate diagnosis. Best corrected visual acuity was 10/40 and 15/40 and intraocular pressure (IOP) was 12 and 11 in the right and left eyes, respectively. Also, no relative afferent papillary defect (RAPD) was found. Visual field testing showed a large peripheral scotoma in both eyes corresponding to the atrophic retinal area. SD-OCT cross

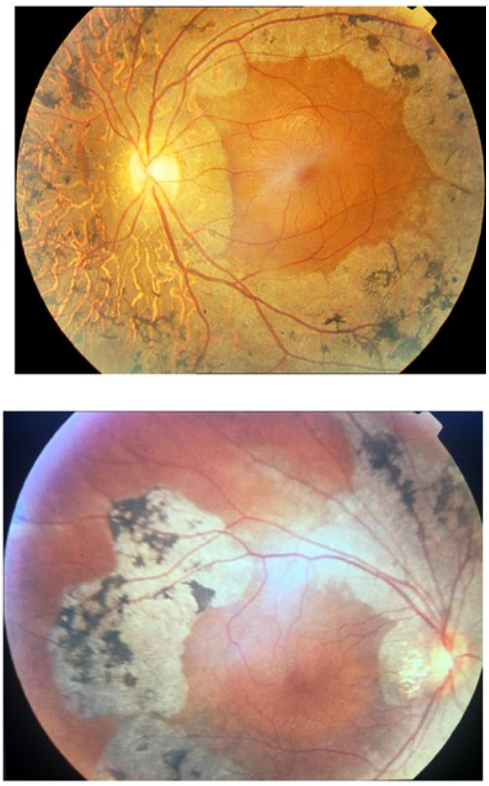

Figure 1. Fundus photograph of left (upper) and right eye (lower) showed chorioretinal atrophy and retinal pigmentation with presenting bone spicule pigment clumping in paravenous distributions

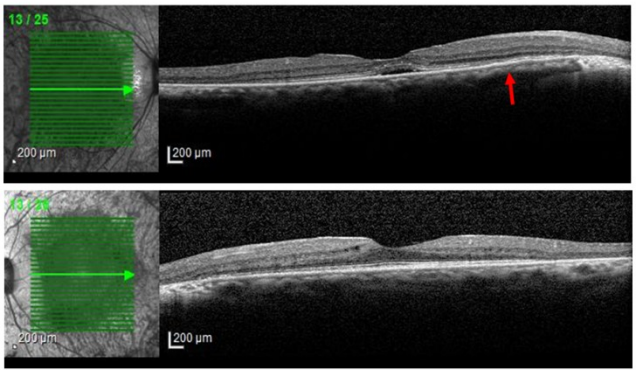

Figure 2. Spectral domain-optical coherence tomography (SD-OCT) of right (upper) and left eye (lower) showed normal outer and inner retina layers and choroid in the spared central macular region with presence of foveal sub-retinal fluid in right eye (red arrow)

scan through the retinal lesions showed significant RPE atrophy and migration with pigment clumping and loss of the entire outer retinal layers (outer nuclear layer, external limiting membrane, myoid zone, ellipsoid zone, and inter-digitation zone) and choroidal atrophy with only large Haller's layer vessels remaining. The SD-OCT scan revealed a highly reflective choroidal signal due to retinal thinning in the affected retina and choroidal atrophy with normal outer retina and RPE in the junctional zone between the damaged and intact regions. Retina and choroid in the spared central macular region appeared normal, except for the presence of foveal sub-retinal fluid in the right eye (Figure 2). Fundus autofluorescence (FAF) and infra-red reflectance (IR) imaging revealed geographic, well-demarcated regions of hypoautofluorescence and hyperreflectance, respectively, corresponding to the atrophic patches of chorioretinal lesion along the paravenous retinal area (Figure 3).

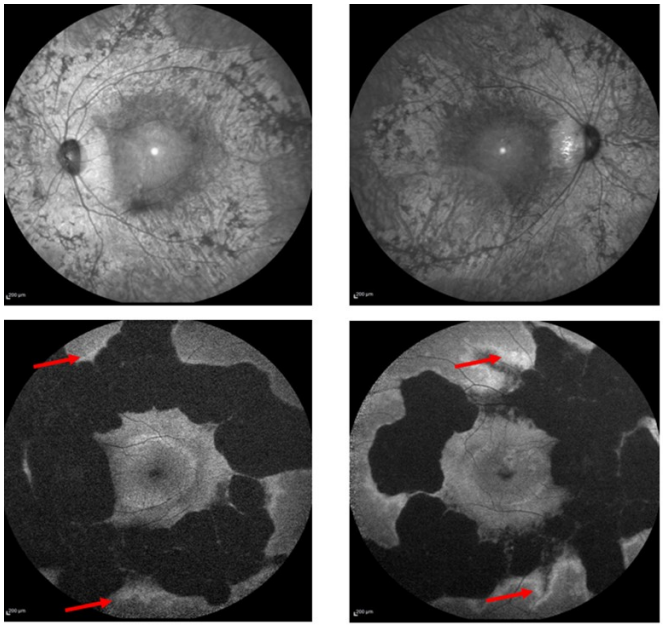

Figure 3. Infra-red reflectance (IR) (upper) and fundus autofluorescence (FAF) (lower) imaging revealed geographic, well-demarcated region of hyperreflectance and hypoautofluorescence, respectively, corresponding to the atrophic patches of chorioretinal lesion along the paravenous chorioretinal atrophy. On FAF imaging, junctional hyperautofluorescent (red arrows) appeared prior to eventual RPE atrophy. In this patient, hyperautofluoresent on the border is consistent with progressive RPE atrophy with an active retinochoroidal atrophic process.

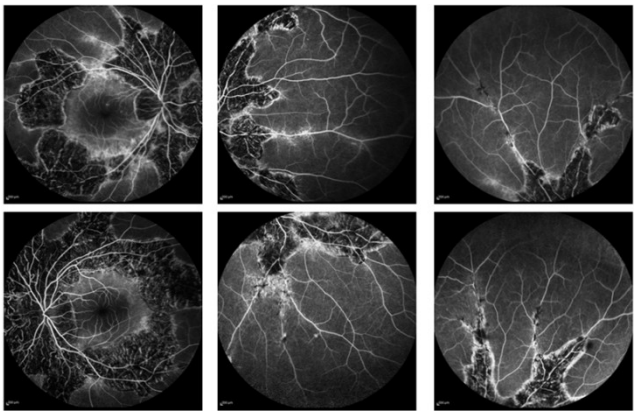

Figure 4. Fluorescein angiography (FA) imaging of right (upper) and left eye (lower) showed the paravenous abnormal areas with diffuse window defect hyperfluorescence consistent with RPE degeneration in both eyes with no active leakage on late phase. More extensive areas of choriocapillaris atrophy and blocked fluorescence in the pigment accumulation areas can be recognized.

Linear hyperfluorescence was noted on FAF, which comparatively surrounded the hypofluorescence in the peripheral paravenous distribution, appearing prior to eventual RPE atrophy, and was consistent with an active chorioretinal atrophic process in this junctional area. On fundus fluorescein angiography (FA) imaging, the paravenous abnormal areas showed diffuse window defect hyperfluorescence consistent with RPE degeneration in both eyes with no active leakage in the late phase and more extensive areas of choriocapillaris atrophy and blocked fluorescence in the pigment accumulation areas (Figure 4). Photopic electroretinogram (ERG) was intact, but scotopic ERG demonstrated flat waves in both eyes. Based on clinical exams, multimodal imaging, and electrophysiologic

studies, PPCRA associated with rod dystrophy was proposed for this patient. The amplitude of the scotopic ERG markedly disappeared, and photopic ERG remained intact, which may indicate association with rod dystrophy. However, it is unclear if the association was real or a coincidence. When atrophy involves extensive areas, the amplitude should be reduced, even in PPCRA. Flat waves are suggested for dystrophy. However, it may be challenging to differentiate PPCRA from retinal dystrophy based on only ERG findings in selected cases. All medications were discontinued, the nature of the disease was explained, and low vision aids were prescribed. The patient had stable visual acuity and ancillary testing at his follow-up for one year, although, as of this writing, he has had no long-term follow-up yet to determine if the findings have remained stable. Fundus abnormalities were more widespread in FAF and FA imaging than in fundus photography. Also, FAF more extensively revealed abnormalities than FA. It is a proper noninvasive examination method to complement a diagnosis of PPCRA. Junctional hyperautofluorescence, as seen in this patient, highlighted stressed RPE with increased lipofuscin deposition, suggesting the possibility of RPE atrophy in the future. Also, choroidal thinning and atrophy along the junctional zone on SD-OCT hypothesized choroidal circulation disorder in the paravenous region, playing a potential role in the pathogenesis of PPCRA.

Choroidal thinning and following blood flow abnormalities may cause defective metabolism in the outer retina. This may lead to an atrophic process first on the outer retina and then on RPE in the disease. As differential diagnoses, acute zonal occult outer retinopathy (AZOOR), with similar clinical appearance, distinct by photopsias within the scotoma as prevalent ophthalmic symptoms. Another differential diagnosis is retinitis pimentosa (RP), characterized by clinical features like optic disc palor and severely affected photopic and scotopic ERG on electrophysiology [3]. Limited information on this disease is published in the literature, mostly from small case series. Recently, researchers have proposed primary RPE dysfunction, retinal vascular maldevelopment, and choriocapillaris abnormalities as primary pathology in this condition. Multimodal imaging techniques have been recently used to better understand the pathogenesis of PPRCA. Various inflammatory, infectious, developmental, and even genetic conditions have been hypothesized with PPCRA. However, the exact etiopathogenesis remains unknown [4-5], While infectious and inflammatory causes have been proposed. As the macula is often spared in this pathology, patients typically carry a good visual prognosis, and visual acuity remains unaffected until advanced phases with a slowly function loss in peripheral retina [6]. Thus, in most cases, fundus abnormality was found out accidentally on a routine ophthalmic exam. PPCRA is usually discovered in distinguished fundus characteristics. However, the multimodal imaging findings help us confirm the diagnosis [6]. As masquerades have been reported in association with inflammatory conditions, a thorough search for uveitic stigmata by exam and FA should be carried out prior to diagnosis [7]. The main differential diagnosis must be with RP [8], and it has been suggested that the two diseases may stand on a spectrum. However, little or no progression without night blindness, the normal appearing regions tend to function normally, and intact or only mildly to moderately affected ERG responses distinguishes the PPCRA from RP [4,9-10].

ACKNOWLEDGEMENTS

We also extend our thanks to clinical research development center of Imam Khomeini and Mohammad Kermanshahi and Farabi Hospitals affiliated to Kermanshah University of Medical Sciences for their kind support.

Financial Support and Sponsorship: Nil.

Conflicts of interest/Competing interests: There are no conflicts of interest.

Ethics approval: The authors certify that they have obtained all appropriate patient consent forms. In the form the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understand that his name and initial will not be published and due efforts will be made to conceal his identity.

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