International Journal of Science and Research Archive
eISSN: 2582-8185
Cross Ref DOI: 10.30574/ijsra
Journal homepage: https://ijsra.net/

(CASE REPORT)

Check for updates

A rare incidence of pigmented paravenous retino-choroidal atrophy in rheumatoid arthritis

Aishwaraya K *, Suresh K, Saravanan R and Kannan M

Department of Ophthalmology, Southern Railways Headquarters Hospital, Perambur, Chennai, India.

International Journal of Science and Research Archive, 2025, 14(01), 153-156

Publication history: Received on 30 October 2024; revised on 18 December 2024; accepted on 20 December 2024

Article DOI: https://doi.org/10.30574/ijsra.2025.14.1.2434

Abstract

Pigmented paravenous retinochoroidal atrophy is a rare disease characterized by perivenous aggregations of pigment clumps associated with peripapillary and radial zones of retinochoroidal atrophy that are distributed along retinal veins. A 55-year-old female presented to our hospital with complaints of floaters in both eyes of 2 years duration. She is a known case of Dry eye and Rheumatoid arthritis on treatment. Fundoscopic examination found bony spicule pigmentation and retinochoroidal atrophy along the retinal veins, bilaterally without macular involvement or signs of inflammationSpectral domain optical coherence tomography imaging revealed lamellar macular hole and thinning of outer retina at the region of pigment deposition along the veins in both eyes. No intervention was done and was advised to follow up. Pigmented paravenous retinochoroidal atrophy is a rare disease characterized by perivenous aggregations of pigment clumps associated with peripapillary and radial zones of retinochoroidal atrophy that are distributed along retinal veins. It has been described in the literature in association with diseases like Behcet's disease, Vogt Koyanagi Harada syndrome, measles, rubeola, tubercular spondylitis, syphilis, and sarcoidosis. PPRCA in a case of Rheumatoid arthritis, to our knowledge has not been reported.

Keywords: Rheumatoid arthritis; Paravenous retinochoroidal atrophy; Retinal pigmented lesions; Syphilis

1. Introduction

Pigmented paravenous retinochoroidal atrophy is a rare disease characterized by perivenous aggregations of pigment clumps associated with peripapillary and radial zones of retinochoroidal atrophy that are distributed along retinal veins (1). Various etiology like degeneration, heredity, inflammation, and idiopathy have been hypothesized (2). Being a rare disease that is not well understood, it is usually asymptomatic, non-progressive, or slowly progressive. PPRCA involves the RPE primarily, with secondary atrophy of the underlying choroidal vasculature (3,4).

2. Case description

A 55-year-old female nurse by occupation hailing from Chennai presented to our hospital with complaints of diminution of vision right eye and floaters in both eyes of 2 years duration. She is a known case of Dry eye and Rheumatoid arthritis on treatment for the past 9 years. She was on methotrexate 7.5mg weekly and hydroxychloroquine for 6 years which was stopped 3 years back. She is on sulfasalazine for the past 3 years. There was no family history of inherited ocular diseases, no history of infection or inflammation in the eye in the past. No history of any other comorbidities. Her best corrected visual acuity (BCVA) was 6/12 in the right eye and 6/9 in the left eye. The anterior segment was unremarkable in both eyes. No relative afferent pupillary defect was found. The intraocular pressure was normal. Fundus examination showed bone-spicule pigmentation and retinochoroidal atrophy along the retinal veins, bilaterally, without macular involvement or signs of inflammation (Figure 1 and 2).

* Corresponding author: Aishwaraya K

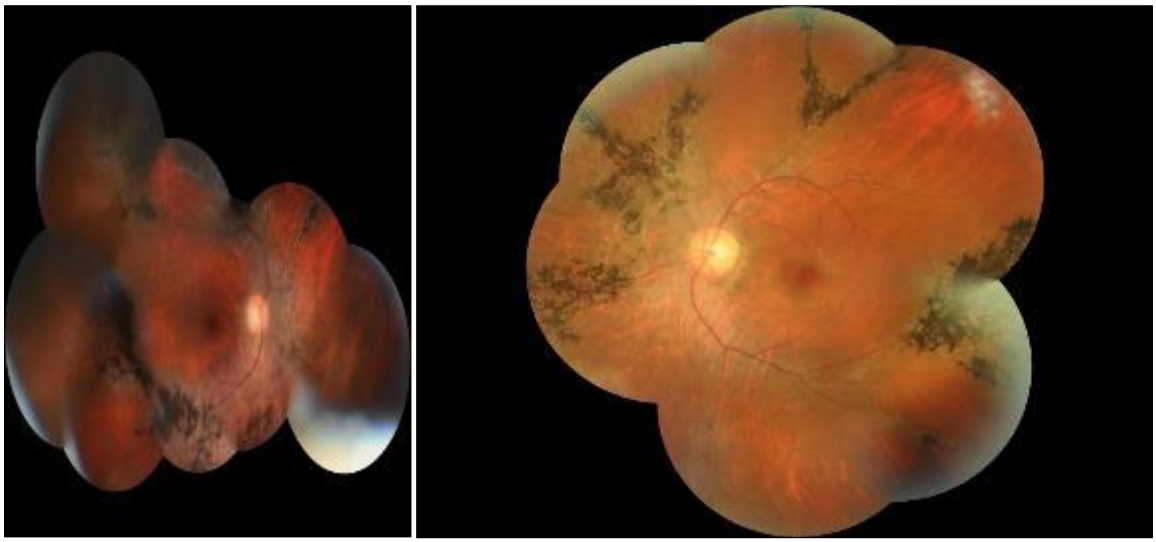

Figure 1 Composite colour fundus photograph of right and left eye showing pigment clumps along the retinal veins and chorioretinal atrophy

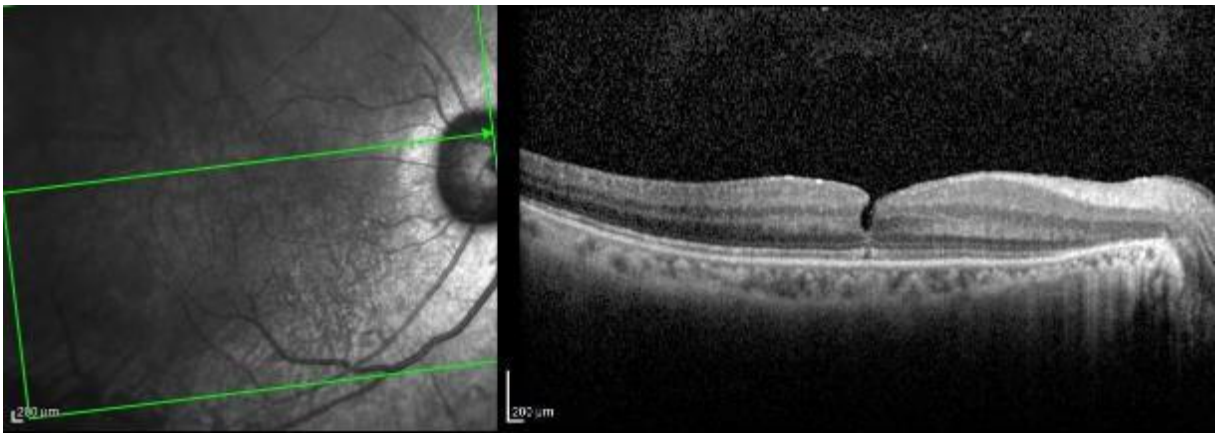

Figure 2 SD-OCT of the right eye showing Lamellar macular hole involving inner retinal layers

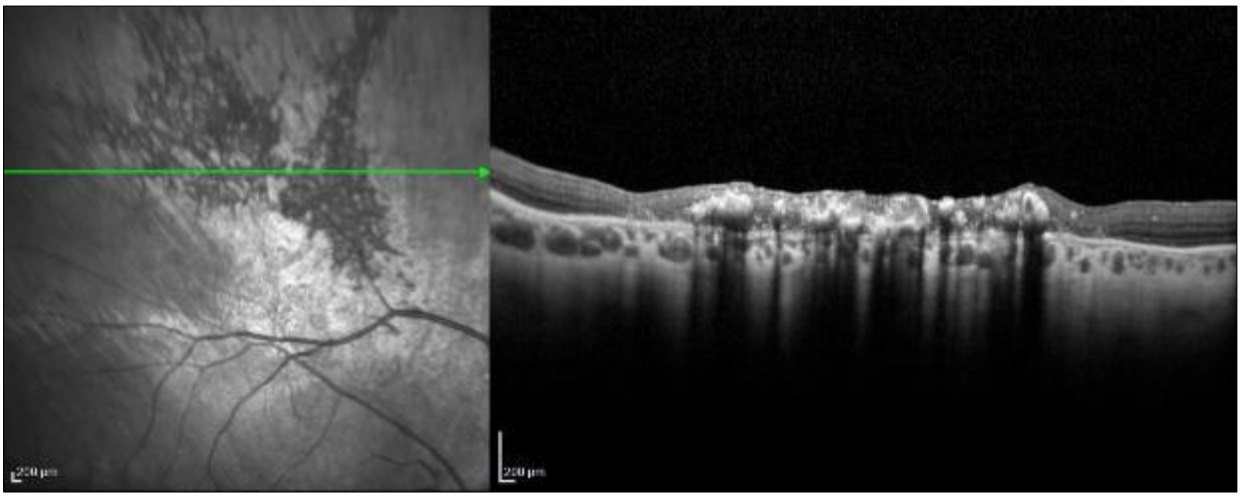

Figure 3 SD-OCT of the right eye shows thinning of the outer retina, loss of outer segments, outer nuclear layer, and inner nuclear layer at the region of pigment deposition.

Spectral-domain optical coherence tomography (SD-OCT) imaging found an obvious thinning of the outer retina, loss of outer segments, outer nuclear layer, and inner nuclear layer at the region of pigment deposition along the veins in both eyes but the microstructure of the macula remained intact (Figure 3). SD-OCT of the right eye showed a Lamellar macular hole involving inner retinal layers (Figure 4).

3. Discussion

This disease was first described by Hewitson-Brown in 1937 and he termed it retinochoroiditis radiate. The cause of the condition is unknown. It has been described in literature in association with diseases like Behcet's disease, Vogt Koyanagi Harada syndrome, measles, rubeola, tubercular spondylitis, syphilis, and sarcoidosis.[2] Mutation within the crumbs homolog 1 (CRB1) gene has been reported(5).

A non-inflammatory cause is referred to as primary, while inflammation-associated PPRCA is referred to as secondary or pseudo-PPRCA(1). PPRCA is commonly bilateral and symmetric. The major complaints of patients with PPRCA are asymptomatic or mild blurred vision(6). The disease is diagnosed by the typical funduscopic features of bilaterally symmetrical accumulation of pigment and retinochoroidal atrophy along the retinal veins. There is no gender predilection(7). Fluorescein studies in young patients with this entity revealed RPE atrophy with preservation of the choroidal vasculature underlying the atrophic RPE(3). In patients with a more advanced stage of the disease, choroidal atrophy has been observed in the involved areas of peripapillary posterior pole pigmentary sheen(4).

In the mild form of the disorder, there are only a few scattered areas with minimal evidence of retinochoroidal atrophy and paravenous pigmentation. In intermediate cases, bone spicule pigment accumulates around the majority of the veins and a minimal amount of retinochoroidal atrophy or regional retinochoroidal atrophy is observed. By contrast, marked cases are associated with peripapillary and posterior pole pigmentary sheen and diffuse areas of retinochoroidal atrophy adjacent to extensive and heavy paravenous pigment accumulation(8).

Fluorescein angiography, indocyanine green angiography, and electrophysiological tests may be used to confirm the diagnosis. Currently, no specific treatment modality is available for PPRCA. Very few cases of this disease have been reported in India to the best of our knowledge. PPRCA in a case of Rheumatoid arthritis, to our knowledge has not been reported.

4. Conclusion

PPRCA is a clinical diagnosis and its association with various clinical features like lamellar macular hole and its association with rheumatoid arthritis which has not been reported before helps us in screening patients, deciding upon the follow-up frequency, and explaining the natural course of the disease. This report adds to our knowledge of the various clinical manifestations and associations of PPRCA.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

References

[1] HUANG HB, ZHANG YX. Pigmented paravenous retinochoroidal atrophy (Review). Exp Ther Med. 2014 Jun;7(6):1439–45.

[2] Kükner AŞ, Yilmaz T, Çelebi S, Aydemir O, Ulaş F. Pigmented Paravenous Retinochoroidal Atrophy. Ophthalmologica. 2003;217(6):436–40.

[3] Chisholm IA, Dudgeon J. Pigmented paravenous retino-choroidal atrophy. Helicoid retino-choriodal atrophy. Br J Ophthalmol. 1973 Aug;57(8):584–7.

[4] Pearlman JT, Kamin DF, Kopelow SM, Saxton J. Pigmented paravenous retinochorodial atrophy. Am J Ophthalmol. 1975 Oct;80(4):630–5.

[5] McKay GJ, Clarke S, Davis JA, Simpson DAC, Silvestri G. Pigmented paravenous chorioretinal atrophy is associated with a mutation within the crumbs homolog 1 (CRB1) gene. Invest Ophthalmol Vis Sci. 2005 Jan;46(1):322–8.

[6] Brooks DN, Potter JW, Bartlett JD, Nowakowski R. Pigmented paravenous retinochoroidal atrophy. J Am Optom Assoc. 1980 Dec;51(12):1097–101.

[7] Hayasaka S, Shibasaki H, Noda S, Fujii M, Setogawa T. Pigmented paravenous retinochoroidal atrophy in a 68-year-old man. Ann Ophthalmol. 1991 May;23(5):177–80.

[8] Noble KG, Carr RE. Pigmented paravenous chorioretinal atrophy. Am J Ophthalmol. 1983 Sep;96(3):338–44.