Acta Scientific Ophthalmology (ISSN: 2582-3191)

Volume 7 Issue 3 March 2024

Case Report

A Case Report of Bilateral Pigmented Paravenous Retinochoroidal Atrophy

Sunanda Sarkhel* and Aditi Singal

Department of Optometry and Vision Science, Amity University, Haryana, India

***Corresponding Author:** Sunanda Sarkhel, Department of Optometry and Vision Science, Amity University, Haryana, India.

Received: January 17, 2024

Published: February 11, 2024

Abstract

Pigmented paravenous retino-choroidal atrophy (PPRCA) is an uncommon disease characterized by peripapillary and radial zones of retinochoroidal atrophy distributed along the retinal veins with perivenous aggregations of pigment clumps in a bilateral manner. The disease aetiology is still unclear. It was earlier considered to be post-inflammatory, but recent reports of familial cases suggest hereditary nature of this condition [1]. It is also considered a variant of retinitis pigmentosa (RP) and can develop RP in further course of time. We emphasize regular follow up of PPRCA patients and screening of the family member owing to its hereditary nature.

Keywords: Retinitis Pigmentosa (RP); Eye; Retinal

Introduction

A 52 year old female came with complains of blurring of vision in the left eye since past 4 months. She is a known case of hypertension on treatment since last 6 months. The best corrected visual acuity recorded in the right eye was 6/9, N6 and the left eye was finger counting at 3 meters, <N36. Anterior segment examination of both the eyes were within normal. Fundus examination showed normal optic nerve head with peripapillary retinochoroidal atrophy that extended along the retinal veins with bone spicule pigment accumulation distributed mainly at distal parts of the atrophic areas in both the eyes with involvement of the macula in the left eye (Figure 1). There were no signs any past inflammation in either eye. Investigations were ordered to confirm these findings. Fundus auto-fluorescence showed areas of hypo auto-fluorescence along the vessels in both the eyes and at the macula in the left eye (Figure 2). The full visual field analysis showed depressed fields in both the eyes (Figure 3). The scotopic and photopic responses on electro-retinogram were reduced for both the eyes (Figure 4). A clinical diagnosis of bilateral pigmented paravenous retinochoroidal atrophy (PPRCA) was made based on the above investigations.

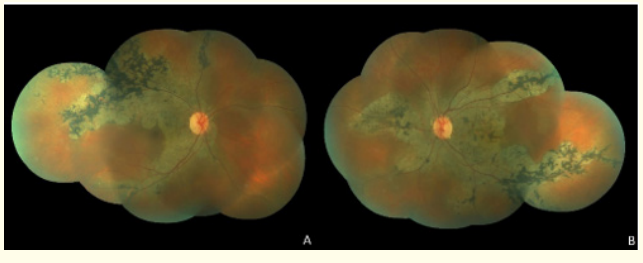

Figure 1: Colour fundus photo of the right eye (A) and the left eye (B) showing peripapillary retinochoroidal atrophy extending along the retinal veins with bone spicule pigments distributed at distal parts of the atrophic areas in both the eyes with involvement of the macula in the left eye.

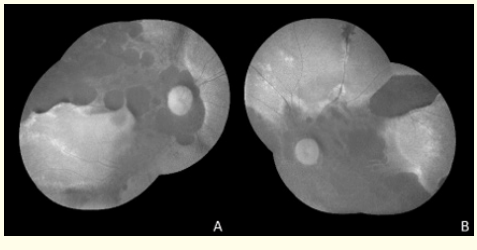

Figure 2: Fundus auto-fluorescence of the right eye (A) and the left eye (B) showing areas of hypo autofluorescence along the vessels in both the eyes and at the macula in the left eye.

Discussion and Conclusion

PPRCA is a rare disease characterized by perivenous aggregations of pigment clumps associated with peripapillary retinochoroidal atrophy that extend along the retinal veins. It is commonly

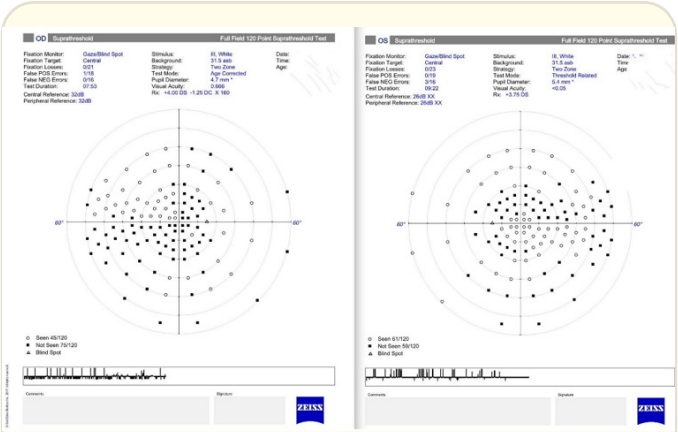

Figure 3: Full visual field analysis of both the eyes showing depressed fields.

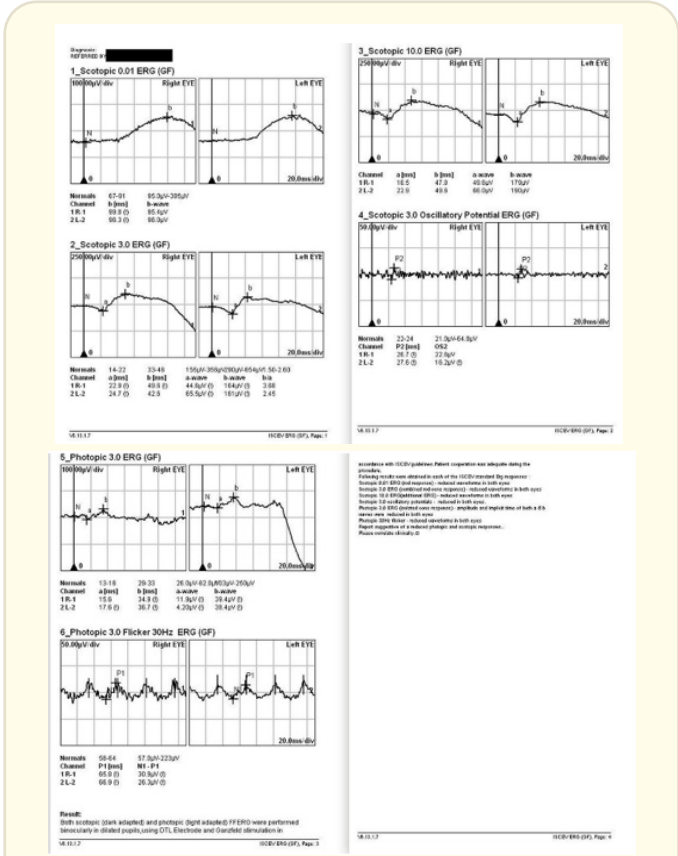

Figure 4: Electroretinogram of both the eyes showing reduced scotopic and photopic responses.

bilateral and symmetric. Patients are usually asymptomatic unless the macula is involved. The disease process is non-progressive or subtly progressive. The cause of the condition may be idiopathic, although a dysgenetic, degenerative, hereditary aetiology or even an inflammatory cause has been hypothesized.[1]It is also considered as a variant of retinitis pigmentosa (RP) [2]. These patients should be on regular follow up as some PPRCA patients develop RP in further course of time or their family member develops retinal degenerative disease, as previously reported [3].

Bibliography

1. Huang HB and Zhang YX. "Pigmented paravenous retinochoroidal atrophy (Review)". *Experimental and Therapeutic Medicine* 7.6 (2014): 1439-1445.

2. Choi JY., *et al*. "Natural course of ocular function in pigmented paravenousretinochoroidal atrophy". *American Journal of Ophthalmology* 141 (2006): 763-765.

3. Aoki S., *et al*. "Unilateral pigmented paravenous retinochoroidal atrophy with retinitis pigmentosa in the contralateral eye: A case report". *American Journal of Ophthalmology Case Reports* 8 (2017): 14-17.