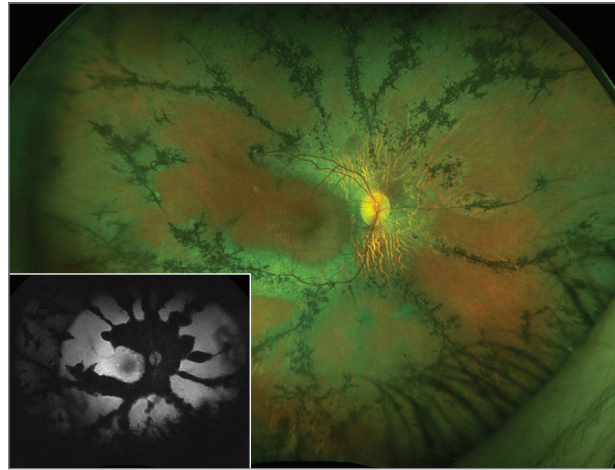


Ophthalmic Images

Simultaneous Pigmented Paravenous Retinochoroidal Atrophy and Retinitis Pigmentosa in the Contralateral Eye

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A Fundus photography, paravenous retinochoroidal atrophy



B Fundus photography, retinitis pigmentosa

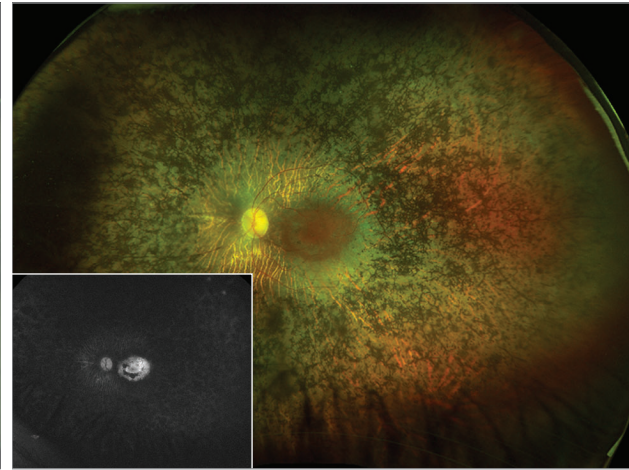


Figure. Fundus photographs and autofluorescence images (insets) of both eyes were evaluated. A, Right eye shows paravenous retinochoroidal atrophy. B, Left eye shows retinitis pigmentosa.

A 59-year-old woman was referred to our clinic. Her parents were consanguineous (first cousins). In 2013, her best-corrected visual acuity (BCVA) was 20/25 OD and 20/40 OS. Fundus examination showed well-demarcated areas of pigmented retinochoroidal atrophy along the vascular arcades with normal optic disc in the right eye, consistent with pigmented paravenous retinochoroidal atrophy (PPRCA) (Figure, A). In the left eye, fundus examination showed diffuse bony spicules, disc pallor, and

attenuated arterioles suggestive of retinitis pigmentosa (RP) (Figure, B). After 10 years, BCVA was 20/30 in OD and 20/400 OS, which correspond with the expected evolution of each separated disease.¹

Both PPRCA and RP are conditions that classically present bilaterally and symmetrically.¹ There are few reports where both diseases occur in the same patient.^{2,3} Different variants in the *CRB1* gene have been previously reported for both separated conditions.⁴ No pathogenic variant was identified in whole-exome sequencing, including *CRB1*.

ARTICLE INFORMATION

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REFERENCES

1. Huang HB, Zhang YX. Pigmented paravenous retinochoroidal atrophy (review). *Exp Ther Med*. 2014;7(6):1439-1445. doi:10.3892/etm.2014.1648
2. Aoki S, Inoue T, Kusakabe M, et al. Unilateral pigmented paravenous retinochoroidal atrophy with retinitis pigmentosa in the contralateral eye: a case report. *Am J Ophthalmol Case Rep*. 2017;8:14-17. doi:10.1016/j.ajoc.2017.08.003
3. Ratra D, Chandrasekharan DP, Aruldas P, Ratra V. Concurrent retinitis pigmentosa and pigmented

paravenous retinochoroidal atrophy phenotypes in the same patient. *Indian J Ophthalmol*. 2016;64(10):775-777. doi:10.4103/0301-4738.195009

4. 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;46(1):322-328. doi:10.1167/iov.04-0734