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A RARE CASE OF PIGMENTED PARAVENOUS RETINOCHOROIDAL ATROPHY: A CASE REPORT

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ABSTRACT Pigmented Paravenous Retinochoroidal Atrophy (PPRCA) is a rare retinal disorder characterized by bilateral pigment clumping and retinochoroidal atrophy distributed along retinal veins. The disease is often asymptomatic and discovered incidentally during routine ophthalmic examination. **Case Presentation:** A 14-year-old female presented to the ophthalmology outpatient department with complaints of diminution of vision. Fundus examination revealed characteristic bilateral pigment clumping and chorioretinal atrophy distributed along the retinal veins, suggestive of Pigmented Paravenous Retinochoroidal Atrophy. Optical coherence tomography demonstrated retinal pigment epithelium and outer retinal alterations. The diagnosis of PPRCA was established based on clinical findings. **Conclusion:** PPRCA is an uncommon retinal condition with distinctive fundus features. Recognition of its characteristic appearance helps differentiate it from inherited retinal dystrophies and inflammatory retinochoroidal disorders.

KEYWORDS : Pigmented Paravenous Retinochoroidal Atrophy, PPRCA, Retinal Degeneration, Retinal Pigment Epithelium, Fundus Imaging.

INTRODUCTION

Pigmented Paravenous Retinochoroidal Atrophy (PPRCA) is a rare retinal disorder characterized by pigment clumping and retinochoroidal atrophy distributed predominantly along the retinal veins. The condition is usually bilateral and symmetrical, although unilateral and asymmetric presentations have been reported. First described by Brown in 1937, PPRCA is considered an uncommon form of retinal degeneration with an uncertain etiology. Various hypotheses, including hereditary, developmental, and post-inflammatory mechanisms, have been proposed; however, its exact pathogenesis remains poorly understood.

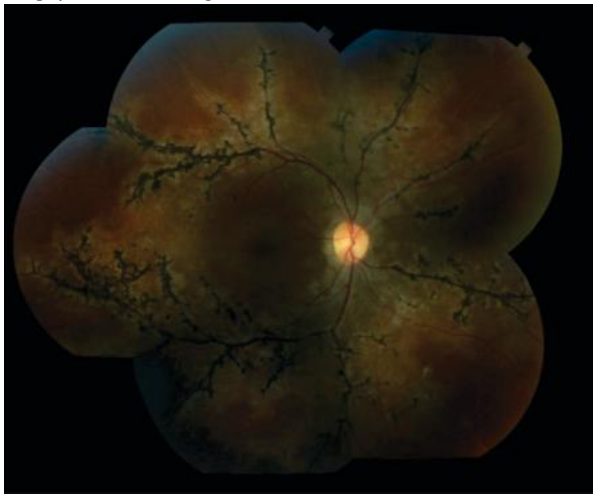
Clinically, PPRCA is often asymptomatic and is frequently detected incidentally during routine fundus examination. Some patients may present with mild visual disturbances, including blurred vision, night blindness, or visual field defects. Visual acuity is generally preserved unless there is involvement of the macular region. Characteristic fundus findings include bone-spicule-like pigment clumping along retinal veins, retinal pigment epithelium (RPE) degeneration, and areas of chorioretinal atrophy adjacent to the affected vessels. The macula is usually spared in the early stages, contributing to the relatively favorable visual prognosis observed in most cases.

Advances in multimodal retinal imaging have improved the diagnosis and understanding of PPRCA. Fundus autofluorescence typically demonstrates hypoautofluorescent areas corresponding to RPE atrophy with hyperautofluorescent borders, while optical coherence tomography (OCT) reveals thinning of the outer retinal layers with loss of photoreceptors and RPE disruption. Fluorescein angiography may show window defects secondary to RPE atrophy, and electroretinography findings range from normal to mildly reduced responses.

Because of its rarity and resemblance to other pigmentary retinopathies, particularly retinitis pigmentosa and post-inflammatory chorioretinal degenerations, PPRCA remains an important diagnostic challenge. We report a rare case of Pigmented Paravenous Retinochoroidal Atrophy highlighting its characteristic clinical appearance and multimodal imaging findings, thereby contributing to the limited literature available on this uncommon retinal disorder.

Case Study

A 14-year-old female presented to the Ophthalmology Outpatient Department with outward deviation of the right eye noted for several years. Ocular examination revealed a right exotropia measuring approximately 45 prism diopters. Binocular vision assessment demonstrated deep suppression. Visual acuity assessment suggested associated refractive error. Cycloplegic refraction was advised for further evaluation. Optical coherence tomography (OCT) could not be performed/was not available at the time of evaluation. However, the diagnosis was established based on characteristic fundus findings demonstrating paravenous pigment clumping and retinochoroidal atrophy distributed along the retinal veins.



CONCLUSIONS

Pigmented Paravenous Retinochoroidal Atrophy is a rare retinal disorder with characteristic paravenous pigment clumping and retinochoroidal atrophy. Awareness of its clinical presentation facilitates accurate diagnosis and avoids unnecessary interventions.

Previous studies have reported OCT findings in PPRCA, including thinning of the outer retinal layers, disruption of the ellipsoid zone, retinal pigment epithelium atrophy, and loss of photoreceptors corresponding to areas of paravenous degeneration. In the present case, OCT imaging was not available; therefore, correlation with these structural changes could not be performed.

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