Case Report

# Combined Hamartoma of Retina and Retinal Pigment Epithelium: A Case Report

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## Abstract

**Background:** Combined hamartoma of the retina and retinal pigment epithelium (CHRRPE) is an uncommon benign tumor of the retina that often affects one eye. The typical CHRRPE consists of lesions that are somewhat elevated at the posterior pole, and the proliferative membrane frequently causes vascular distortion. Macular edema, macular holes, retinal detachments, or vitreous hemorrhages may manifest in severe cases. Patients with unusual clinical symptoms are more likely to have an incorrect diagnosis made by novice ophthalmologists.

**Case summary:** We present a bilateral involving CHRRPE lesion in a 26-year-old gentleman who came with complaints of defective vision in both eyes since childhood. His BCVA was Right eye 5/60 and left eye 6/60. His anterior segment examination showed no abnormality, with posterior segment examination showing both eyes greyish white elevated lesions involving the macula with thick fibrotic membrane causing the macular drag temporally in the right eye and supero-temporally in the left eye. OCT shows the thick ERM with the disorganized inner retinal layers suggestive of hamartoma of the Retina and Retinal Pigment Epithelium.

**Conclusion**: Although specialist retina evaluation is enough to diagnose a case of CHHRPE, associated OCT evaluation will help in differentiating the lesion from other differential diagnosis such as idiopathic Epiretinal membranes and unilateral involvement is more common but bilateral involvement can be seen as in this case.

**Keywords:** Hamartoma, Retina, CHRRPE.

## Introduction

Combined hamartoma of the retina and retinal pigment epithelium (CHR-RPE) is a benign condition that is included in pigmented lesions of the fundus. The first case of CHRRPE was documented by Gass[1] in 1973. Although it usually affects children, CHRRPE has also been documented in old or young people.[2,3] Usually, it is not linked with any systemic disease and it often affects just one eye; in a small number of cases, association with type II or type I neurofibromatosis[4] is noted.

CHRRPE Lesion can be found in the macula, juxta papillary, or periphery and consists of glial cells, vascular tissue, and sheets of pigment epithelial cells. The most typical symptoms of CHRRPE are strabismus and painless vision loss. However, depending on where the lesion is located, it may not present with any symptoms at all and can only be found on fundus examination.[5,6]

## Case Presentation

In the present case, we report a Bilateral CHRRPE lesion in a 26-year-old male who came with complaints of defective vision in both eyes since childhood. His BCVA was Right

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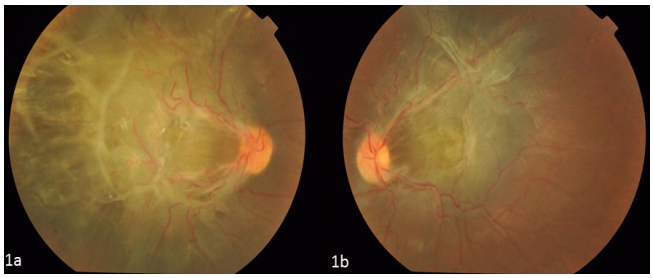
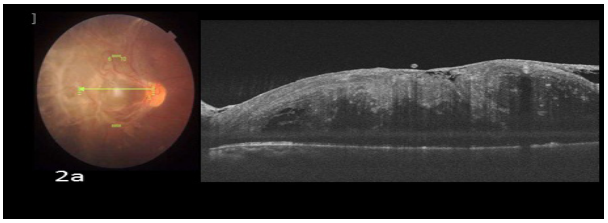



**Fig. 1:** a and b show the right and left eye with greyish-white elevated lesions involving the macula with a thick fibrotic membrane causing the macular drag temporally.



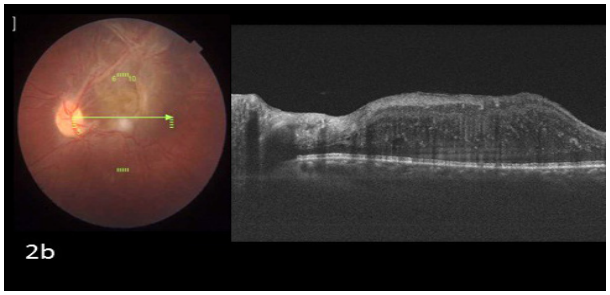




**Fig. 2:** a and b optical coherence tomography section through the lesion showing the grossly thickened and disorganized retina with a thick tractional pre-retinal membr

eye 5/60 and left eye 6/60. His anterior segment examination showed no abnormality, while posterior segment examination showed both eyes (Fig. 1a and b) greyish white elevated lesion involving the macula with a thick fibrotic membrane causing the macular drag temporally in the right eye and supero-temporally in the left eye (Fig. 2a and b). OCT shows the thick ERM with the disorganized inner retinal layers suggestive of hamartoma of the retina and retinal pigment epithelium.
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## Conclusion

Combined hamartoma of the retina and retinal pigment epithelium (RPE) is a rare, congenital, and benign tumor that may be mistaken for choroid melanoma or retinoblastoma.[7]

Although specialist retina evaluation is enough to diagnose a case of CHHRPE, associated OCT evaluation will help in differentiating the lesion from another differential diagnosis such as idiopathic Epiretinal membranes.

Since combined hamartoma of the retina and RPE presents at a young age, amblyopia prevention is paramount. Surgery for associated ERMs is still a subject of debate since visual acuity may not improve despite membrane removal. Also, it may not be possible to remove the whole ERM without damaging the retina. There are case reports of treatment of complications and non-classic presentations of combined hamartomas, such as CNV and vitreous hemorrhage.

Although unilateral presentation of the CHRRPE is the most common bilateral involvement can also be seen as presented in our case report.

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