Case Report

Resolution of Combined Hamartoma of Retina And Retinal Pigment Epithelium (CHRRPE) with Following Pars Plana Vitrectomy

Monika Kapoor,[1] Siddharth Madan,[2] Priyanka Sankaran,[1] Preeti Singh,[1] Sarita Beri[1]

[1]*Department of Ophthalmology, Lady Hardinge Medical college and Associated Hospitals, University of Delhi, New Delhi, India.*
[2]*Department of Ophthalmology, University College of Medical Sciences and Associated GTB Hospital, University of Delhi, New Delhi, India.*

Abstract

Combined hamartoma of retina and retinal pigment epithelium (CHRRPE) is a benign tumor located at the posterior pole. The pathogenesis of the lesion is unknown however some authors propose that the tumor may arise from undifferentiated ectopic progenitor cells which were otherwise destined for RPE and demonstrate failure to completely differentiate. These cells then undergo hyperplasia and accumulate in the neurosensory retina. Optical coherence tomography (OCT) assists in classification of the lesions based on location and differentiates from an epiretinal membrane (ERM). Surgical intervention is warranted in cases that produce macular distortion with/without associated ERM or retinal detachment and those producing visual complaints. Similar was the case with a 26-year-old male who presented with unilateral metamorphopsia that was attributed to CHRRPE with a grade two ERM. Pars plana vitrectomy with ERM peeling resulted in resolution of disabling symptoms and anatomical success as visible on OCT.

Delhi J Ophthalmol 2021;32;74-75; Doi http://dx.doi.org/10.7869/djo 695

Keywords: *CHRRPE, Epiretinal Membrane, Combined Hamartoma Of Retina And Retinal Pigment Epithelium, Pars Plana Vitrectomy, Omega Sign*

Introduction

Combined hamartoma of retina and retinal pigment epithelium (CHRRPE) are rare, solitary, nonhereditary unilateral oval to bean shaped tumors that are benign with no malignant potential. These tumors are composed of glial, vascular, and RPE components. The lesion is usually an ill-defined gray retinal mass with the presence of tortuous or straightened retinal blood vessels. Retinal traction might also be observed. The lesions are usually stable. The CHRRPE lesions involve either the optic nerve or the macula. CHRRPE involving the macula show progressive loss of vision as in the case of a 26-year-old male due to the presence of epiretinal glial tissue and vitreous traction.[1,2]

Case report

The patient presented with diminution and distortion of vision in his right eye (OD) for last one year. Best corrected visual acuity (BCVA) was 6/12 partial OD with a refractive correction of -0.75 DS/-0.50 DC x 180 degrees and 6/6 (with plano) in the left eye (OS). Anterior segment examination and intraocular pressures were normal in both the eyes. Fundus examination OD showed grade two epiretinal membrane (ERM) based on the Gass classification, manifesting with macular distortion along-with a CHRRPE (Figure 1a). Fundus examination OS was normal. Fundus fluorescein angiography showed distortion of vessels near the lesion and ERM (Figure 1b). All secondary causes for ERM were ruled out. Optical coherence tomography (OCT) OD showed the typical omega sign (Figure 1c) and incomplete posterior vitreous detachment. In view of deteriorating visual acuity, vitreoretinal traction (Figure 1d) and surface irregularity, pars plana vitrectomy (PPV) with ERM peeling without any internal limiting membrane peeling along-with intravitreal 20 % sulfur hexafluoride (SF6) injection in the vitreous cavity was performed. A VA of 6/9 was achieved on post-operative day 40 with amelioration of metamorphopsia that was documented on Amsler's chart (Figure 1e-f).

Discussion

Dragging of the fovea is seen in all CHRRPE lesions involving the macula and in around 42 % of extra-macular lesions. Fundus photography is very helpful for documentation and follow-up. OCT findings include omega-shaped in¬filtration of the inner retina that is lined posteriorly by the outer plexiform layer. OCT classifies the CHRRPE lesions into three zones based on their location i.e. macular/peripapillary- Zone1; midperiphery-Zone 2 and located in far periphery as Zone 3. The lesions in Zone 1 need frequent follow-up however the peripheral lesions can be observed. The complications associated with CHRRPE include reduced VA due to ERM, amblyopia, retinoschisis, holes, choroidal neovascular membrane or an accompanying retinal detachment.[3]

Epiretinal membranes that are associated with eye diseases such as retinal vascular occlusion, diabetic retinopathy, trauma, retinal detachment, exudation, haemorrhage, choroidal involvement or any ocular surgical intervention are called secondary ERM's. This patient had no signs of intraocular inflammation. Spontaneous separation of ERM's is observed in cases of CHRRPE.[4] Combined hamartoma of retina and retinal pig¬ment epithelium contains retinal blood vessels which appear abnormal and have a certain degree of traction at the vitreoretinal interface and are accompanied with prominent pre-retinal gliosis and tortuosity of the retinal blood vessels. These clinical features of CHRRPE can create diagnostic dilemma with the clinical appearance of complex ERM.

A diagnosis of CHRRPE is more likely as it usually presents early in first or second decade of life and is not associated with underlying causes as mentioned. Moreover OCT helps in distinguishing the two entities as CHHRPE lesions have a characteristic omega-shaped disorganization of inner retinal

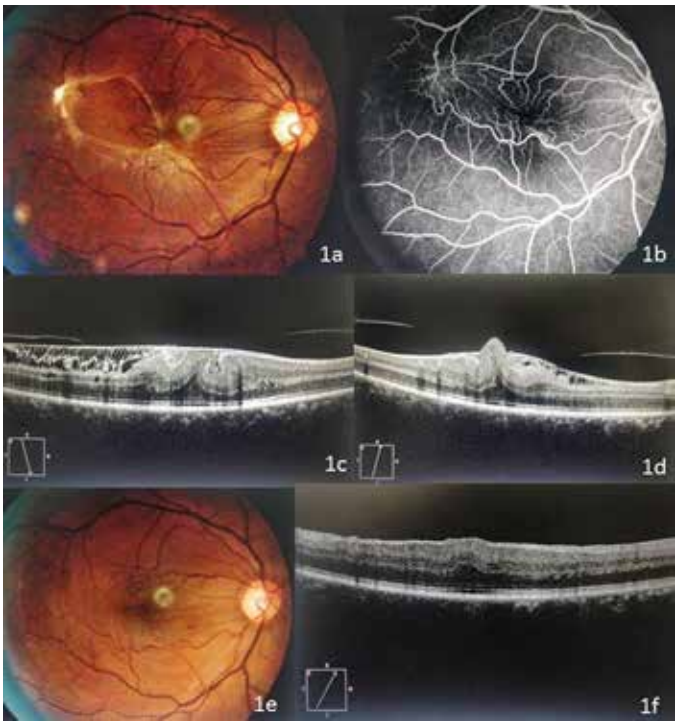

Figure 1: *Fundus photograph OD at presentation showing grade two ERM with an elevated lesion on the superotemporal aspect suggestive of CHRRPE (1a). FFA showed distortion of vessels (1b). OCT OD showed the typical omega sign characterized by localized thickening of the retina with epiretinal glial tissue accompanied-with vitreous traction and bounded posteriorly by the outer plexiform layer (1c-d). Incomplete PVD was also seen on OCT (Fig. 1c-d). Post- operative fundus (1e) and OCT (1f) at day 40 shows resolution of CHRRPE, vitreous traction and regularization of epiretinal surface.*

layers, are bounded posteriorly by the outer plexiform layer. This was also the case in this patient who no other underlying ocular pathology and therefore CHRRPE was a likely diagnosis.

Surgery aims to reduce the vitreous traction and establish surface regularity. Overlying epiretinal membrane is closely linked to the underlying hamartoma therefore forceful peeling of ERM is not advocated due to the anticipated complications.[5,6] Reports on resolution of CHRRPE with epi-retinal membrane removal performed during PPV accompanied-with disappearance of omega sign are few. Epiretinal membranes may recur after surgery however VA does improve after primary surgery even if the ERM recurs. Timely surgical interventions may yield gratifying functional and anatomical success.

References

1. Kumar V, Chawla R, Tripathy K. Omega Sign: A Distinct Optical Coherence Tomography Finding in Macular Combined Hamartoma of Retina and Retinal Pigment Epithelium. Ophthalmic Surg Lasers Imaging Retina. 2017 Feb 1;48(2):122125.
2. Shields CL, Thangappan A, Hartzell K, Valente P, Pirondini C, Shields JA. Combined hamartoma of the retina and retinal pigment epithelium in 77 consecutive patients visual outcome based on macular versus extramacular tumor location. Ophthalmology. 2008 Dec;115(12):2246-2252.e3.
3. Dedania Vs, Ozgonul C, Zacks Dn, Besirli Cg. Novel Classification System For Combined Hamartoma Of The Retina And Retinal Pigment Epithelium. Retina. 2018 Jan;38(1):12-19.
4. Kumar V. Spontaneous Separation of ERM in Combined Hamartoma of Retina and Retinal Pigment Epithelium. Ophthalmology. 2017 Sep 1;124:1402.
5. Bruè C, Saitta A, Nicolai M, Mariotti C, Giovannini A. Epiretinal membrane surgery for combined hamartoma of the retina and retinal pigment epithelium: role of multimodal analysis. Clin Ophthalmol. 2013;7:179-84.
6. Zhang X, Dong F, Dai R, Yu W. Surgical management of epiretinal membrane in combined hamartomas of the retina and retinal pigment epithelium. Retina. 2010 Feb;30(2):305-9.

Cite This Article as: Monika Kapoor, Siddharth Madan, Priyanka Sankaran, Preeti Singh, Sarita Beri. Resolution of Combined hamartoma of retina and retinal pigment epithelium (CHRRPE) with epiretinal membrane following pars plana vitrectomy. Delhi J Ophthalmol 2021; 32 (1) 74-75.

Acknowledgments: Nil

Conflict of interest: None declared

Source of Funding: None

Date of Submission: 06 Feb 2021
Date of Acceptance: 02 Apr 2021

Address for correspondence

Siddharth Madan

M.S., D.N.B, FICO, FAICO (Retina)
Assistant Professor,
Department of Ophthalmology,
University College of Medical
Sciences and Associated GTB Hospital,
University of Delhi, New Delhi
Email: drsiddharthmadan@gmail.com

Quick Response Code