ECRONICON OPEN ACCESS

EC OPHTHALMOLOGY
Case Report

Approach to Adult Combined Hamartoma of the Retina and Retina Pigment Epithelium Patient

Gamze Maden[1]*, Bora Deniz Argon[1], Şeyma Gülcenur Özturan[1], Akın Çakir[1], Mehmet Egemen Karataş[2] and Mustafa Nuri Elçioğlu[1]

[1]Department of Ophthalmology, Okmeydani Training and Research Hospital, Istanbul, Turkey

[2]Department of Ophthalmology, Sisli Hamidiye Etfal Training and Research Hospital, Istanbul, Turkey

***Corresponding Author:** Gamze Maden, Department of Ophthalmology, Okmeydani Training and Research Hospital, Istanbul, Turkey.

Received: June 28, 2018; **Published:** September 28, 2018

Abstract

A Combined Hamartoma of the Retina and Retina Pigment Epithelium (CHR-RPE) was detected in the right eye of the 43-years-old female patient who had complaints of low visual acuity and blurriness in her right eye since her childhood after the ophthalmologic examination.

Since CHR-RPE could be confused with many malignant tumors, we did want to draw attention to the true diagnosis.

Keywords: *Hamartoma; Retina Pigment Epithelium; Tumor*

Introduction

CHR-RPE is a rare benign fundus lesions that can be confused with Choroidal Malignant Melanoma, Choroidal Nevus, Retinoblastoma, Morning Glory Anomaly, Epiretinal Membrane, Adenoma or Adenocarcinoma of RPE, Melanocytoma or Congenital Hypertrophy of Retinal Pigment Epithelium [1]. Therefore, true diagnosis is important for prevention of radical treatment interventions.

Purpose in the Study

Our purpose in this study is to present a case with CHR-RPE, which admit to our clinic, with differential diagnosis.

Case Presentation

A 43-year-old female patient was admitted to our polyclinic with a complaint of low vision and blurred vision in her right eye since childhood. On examination, her aided visual acuity was hand motion in right eye, 8/10 in left eye. And in the right eye she had 45 Δ D exotropia without alternation. Her anterior segment biomicroscopic examination was natural except for a mildly posterior subcapsular cataract in the right eye. Intraocular pressure was normal in both eyes (14 mmHg/14 mmHg). Fundus examination of the right eye showed a large amount of gliotic bands on the papilla and in the posterior pole of retina, and a thick and elevated scar in the vicinity. In addition to that atrophic macula and a thickened and elevated scar region in the periphery of temporal zone and gliotic bands on it were observed in the same eye. There were no lesions on the fundus examination of the left eye.

There were numerous small vessels in the lesion with increased tortuosity. Existence of Epiretinal Membrane and the vessels with increased tortuosity caused the traction on the surface of the retina (Figure 1a and 1b). FFA images; Early phase FA displays; optic nerve head looks blurry due to epiretinal membrane which is extending from macular area to optic nerve head (Figure 2), OCT image is shown figure 3.

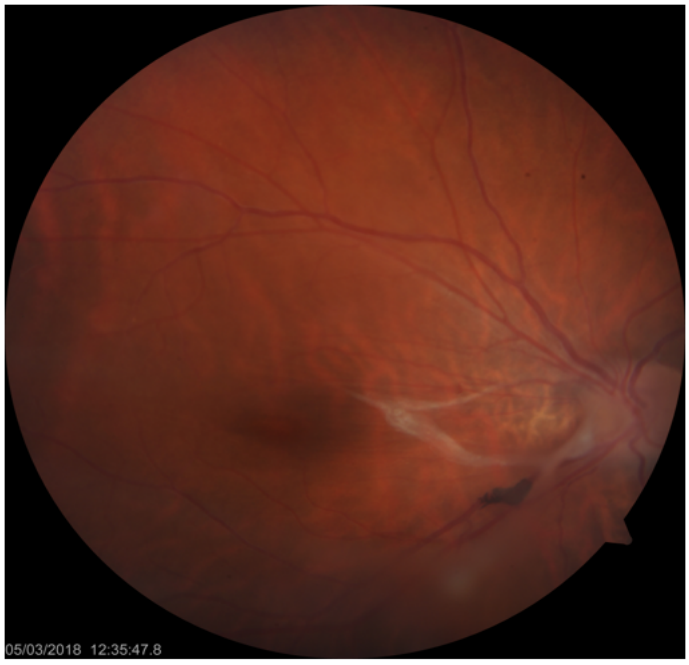

Figure 1a: *Fundus photo shows CHR-RPE.*

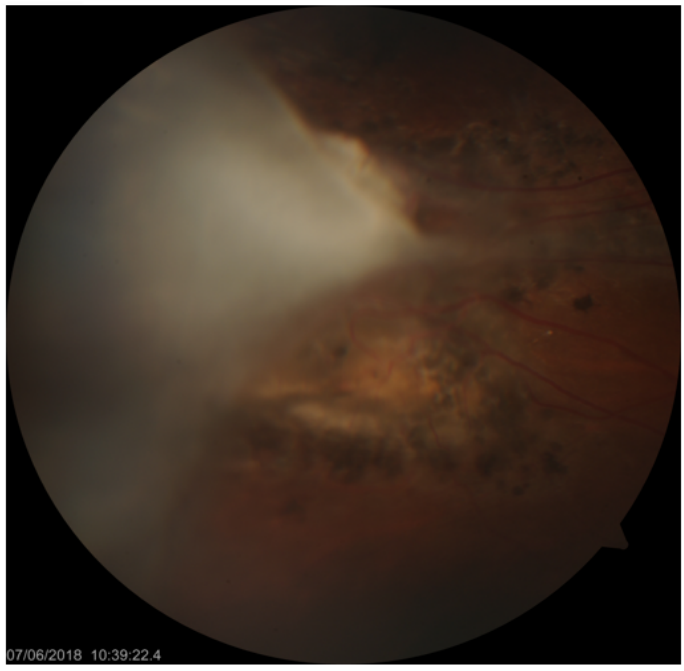

Figure 1b: *Fundus photo shows CHR-RPE.*

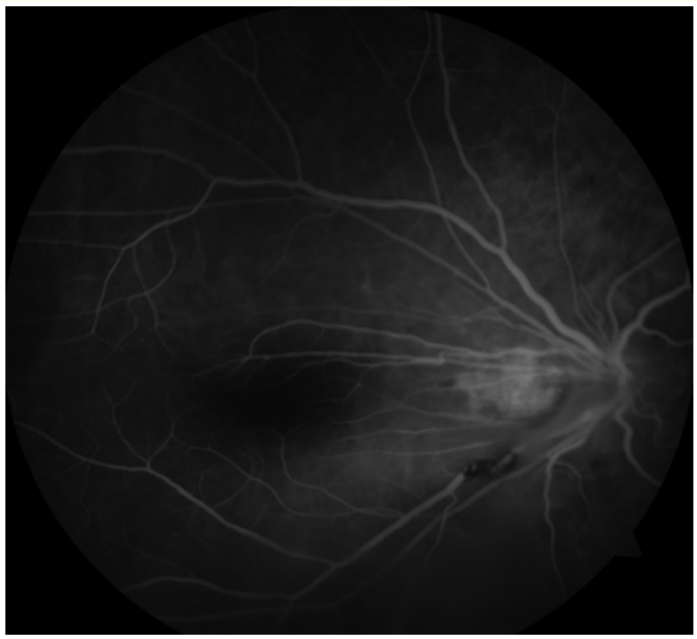

Figure 2a: *FFA image shows early phase image.*

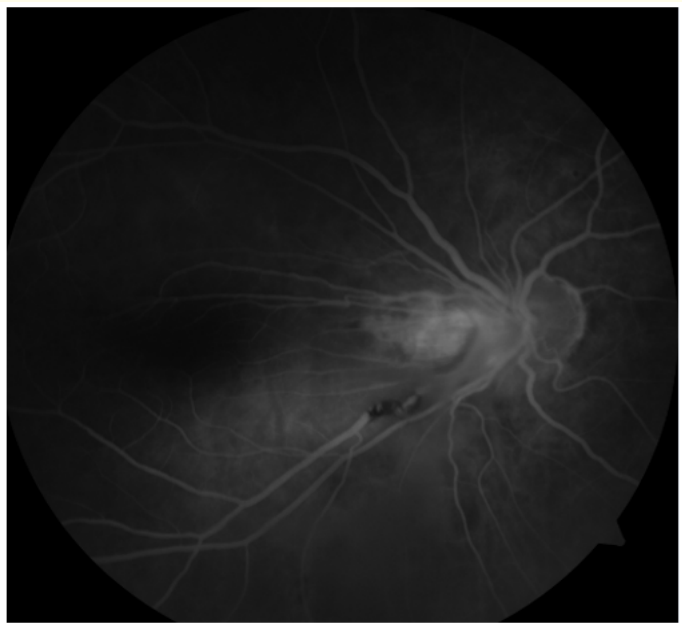

Figure 2b: *FFA image shows mid-phase image.*

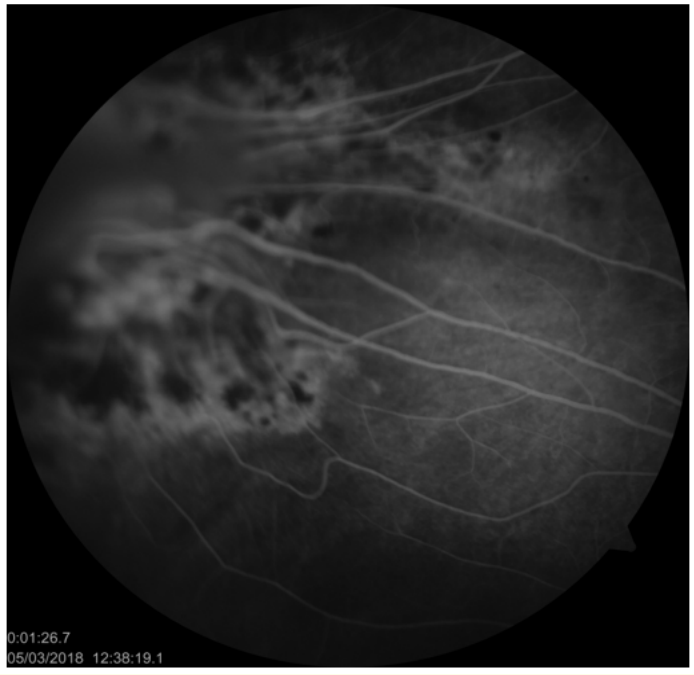

Figure 2c: *FFA image shows mid-phase image (temporal quadran).*

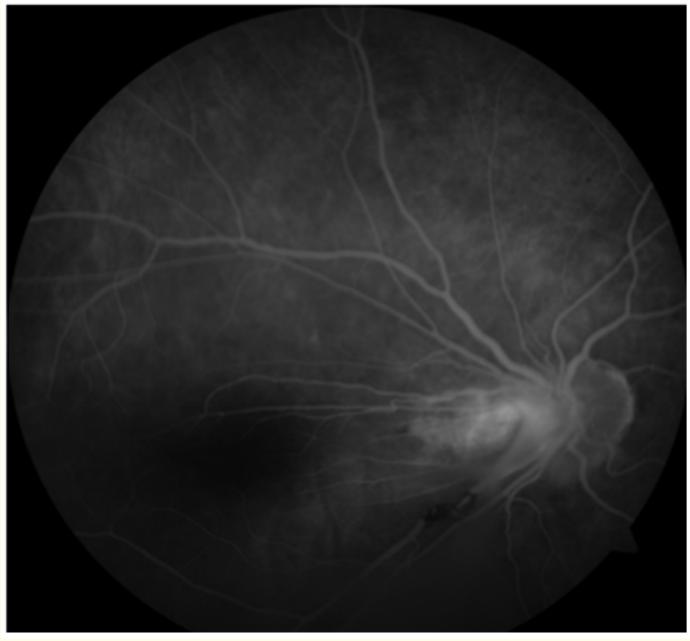

Figure 2d: *FFA image shows late phase image.*

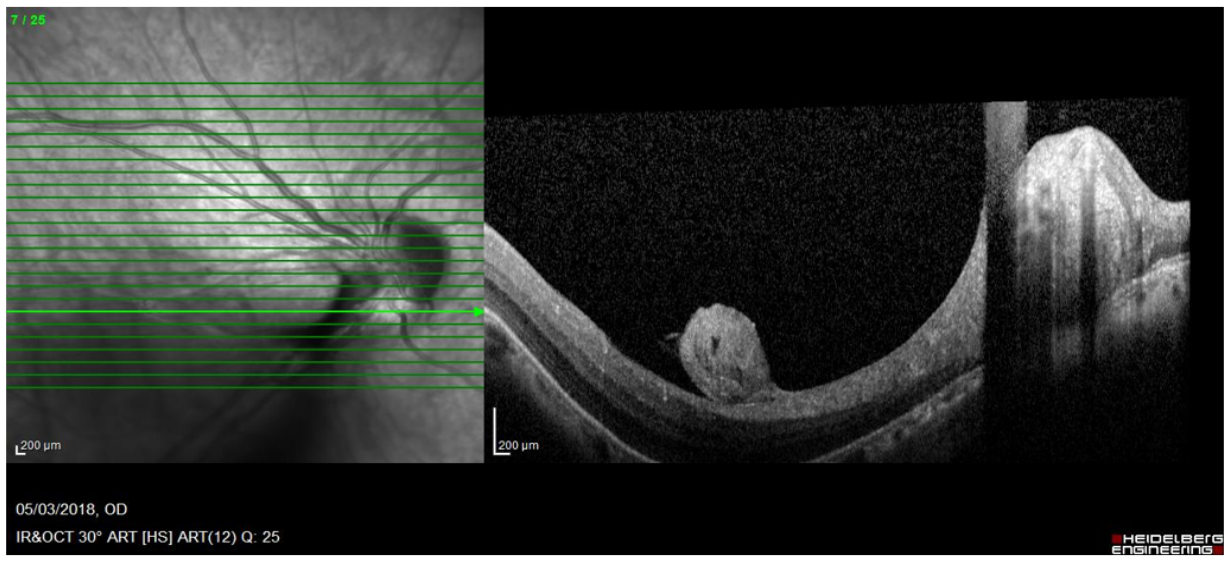

Figure 3: *OCT image shows that the CHR-RPE. It is observed that the integrity of the retinal pigment epithelium layer is impaired in this image.*

Discussion

70% of CHR-RPE patients are female [1]. The patients commonly present with painless decrease in VA (60%), strabismus (18%), floaters (5%), ocular pain (3%) and leukocoria (< 1%). The diagnosis is usually made by routine eye examination. CHR-RPE is placed in the macular zone, optic nerve, juxta-papillary zone and mid-periphery [2]. The presenting VA depends on the location of the lesion [3]. CHR-RPE lesion contains elevation from retina at a certain level, pigmentation, increase the tortuosity of vessels and glial proliferation [4].

Development of Choroidal neovascularization, Vitreous hemorrhage and macular hole are also secondary cause of vision loss. Our patient's lesion was extending from optic nerve head to periphery of temporal zone and due to macular traction her VA was at the level of hand motion.

The patients with CHR-RPE, there is no systemic disease's symptom. However in many studies have been shown that CHR-RPE is associated with Neurofibromatosis type 1 and 2, Tuberous Sclerosis, Gorlin Syndrome [5]. It has been reported that CHR-RPE is associated with x-linked juvenile retinoschisis and optic pits, and that these tumors can be seen with optic nerve head coloboma and optic nerve head drusen [1].

There was no indication for treatment in our patient because her visual acuity was at the level of hand motion and there was 45 Δ D exotropia in the same eye. Because we thought that the level of visual acuity depends on the deprivation amblyopia.

Surgical treatment is done by peeling the epiretinal membranes with pars plana vitrectomy, which is believed to be associated with the lesion and causes decrease in visual acuity in time.

Choroidal Malignant Melanoma, Choroidal Nevus, Retinoblastoma, Morning Glory Anomaly, Congenital Hypertrophy of Retinal Pigment Epithelium, Choroidal Neovascularization, Retinoschisis should be considered in the differential diagnosis [6]. Oct sections often

show normal retinal structure in choroidal melanoma. Combined hamartomas show high reflectivity in inner retina layers with complete optical shadowing of the underlying retina. As a result, we think that OCT is an important, non, invasive and effective diagnostic tool for the differential diagnosis of CHR-RPE with choroidal malignant melanoma and avoidance of unwanted surgical interventions.

It is a benign lesion, it is valuable in terms of prevention of misdiagnosis and unnecessary treatment just because it can be confused with intraocular malignant tumors.

Conclusion

The patient was considered CHR-RPE with this data. It is important to keep in mind that it is not often encountered but can interfere with the diseases we often see.

Bibliography

1. Augsburger JJ., *et al*. "Combined hamartoma retina and retinal pigment epithelium". *Yanoff Ophthalmology* 9 (1093): 156.
2. Shields CL., *et al*. "Combine hamarto- ma of the retina and retinal pigment epithelium in 77 consecutive patients visual outcome based on macular versus extramacular tumor location". *Ophthalmology* 115.12 (2008): 2246-2252.
3. İnoue M., *et al*. "Successful treatment of sub-foveal choroidal neovascularizastion associated with combined hamartoma of the retina and retinal pigment epithelium". *American Journal of Ophthalmology* 138.1 (2004): 155-156.
4. Shields CL., *et al*. "Optical coherence tomographic findings of combined hamartoma of the retina and retinal pigment epithelium in 11 Patients". *Archives of Ophthalmology* 123.12 (2005): 1746-1750.
5. Tsai P and O'Brien JM. "Combined hamartoma of the retina and retinal pigment epithelium as the presenting sign of neurofibromatosis-1". *Ophthalmic Surgery, Lasers and Imaging* 31.2 (2000): 145-147.
6. İskovicch J and Acherman C. "An epidemiological study of posterior uveal melanoma in Israel. 1961-1989". *International Journal of Cancer* 61.3 (1995): 219-225.

Volume 9 Issue 10 October 2018