



# Multimodal Imaging Characteristics of Circumscribed Choroidal Hemangioma Accompanying with Central Serous Chorioretinopathy: A Case Report

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## Abstract

A 42-year-old man presented with a temporal visual field defect in his right eye. His history revealed systemic steroid use before onset of his complaints. Multimodal imaging techniques including ultrasonography, fluorescein angiography, indocyanine green angiography, and optical coherence tomography angiography (OCTA) suggested the presence of circumscribed choroidal hemangioma (CCH) accompanying with central serous chorioretinopathy (CSCR) in the right eye, which might have worsened with systemic steroid treatment. CCH may rarely present with accompanying CSCR. Besides, OCTA is a non-invasive reliable method for the diagnosis of CCH in terms of visualizing vascular features of tumor.

**Keywords:** Central serous chorioretinopathy, choroidal thickness, circumscribed choroidal hemangioma, optical coherence tomography angiography, optical coherence tomography

## Introduction

Circumscribed choroidal hemangioma (CCH) is a benign vascular hamartoma of the choroid that generally manifests as an isolated unilateral tumor without systemic associations, and appears clinically as a round/oval orange-red mass usually posterior to the equator (1). CCHs are usually diagnosed if the patient becomes symptomatic secondary to visual loss associated with exudative foveal detachment and/or macular edema, or incidentally found during routine examination. Despite its characteristic clinical appearance, the accurate diagnosis of CCH is crucial which can be particularly challenging in chronic lesions (2). Recently, CCH has also been associated with elevated choroidal thickness and possible central serous chorioretinopathy (CSCR) (3,4).

Herein, we present multimodal imaging characteris-

tics, including optical coherence tomography angiography (OCTA), of a patient with CCH and accompanying CSCR.

## Case Report

A 42-year-old man presented to another eye clinic a year ago with a temporal visual field defect (VF) in his right eye. His history was unremarkable except systemic steroid treatment 2 weeks before onset of his complaints. Best-corrected visual acuities (BCVAs) were 0.8/1.0 (Snellen), and intraocular pressures (IOPs) were 12/13 mmHg in his right and left eye, respectively. He was initiated on systemic antimicrobials, systemic steroid therapy, and sub-Tenon steroid injections, with presumed diagnosis of neuroretinitis in the right eye. Systemic work-up was negative for HLA-B51, HLA-B27, and infectious causes.

**How to cite this article:** Karaca I, Guven Yilmaz S, Mentis J, Markov G, Tuncer S. Multimodal Imaging Characteristics of Circumscribed Choroidal Hemangioma Accompanying with Central Serous Chorioretinopathy: A Case Report. *Beyoglu Eye J* 2022; 7(4): 329-333.

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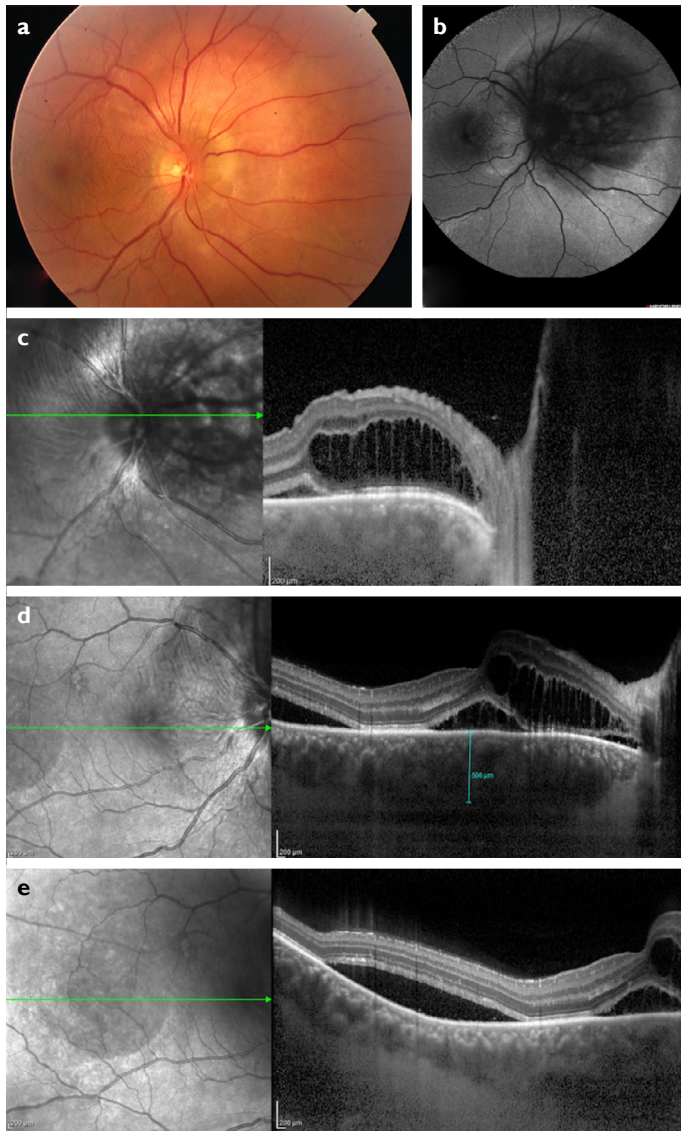
**Submitted Date:** March 14, 2022 **Revised Date:** July 26, 2022 **Accepted Date:** September 01, 2022 **Available Online Date:** November 15, 2022

*Beyoglu Eye Training and Research Hospital - Available online at [www.beyoglueye.com](http://www.beyoglueye.com)*

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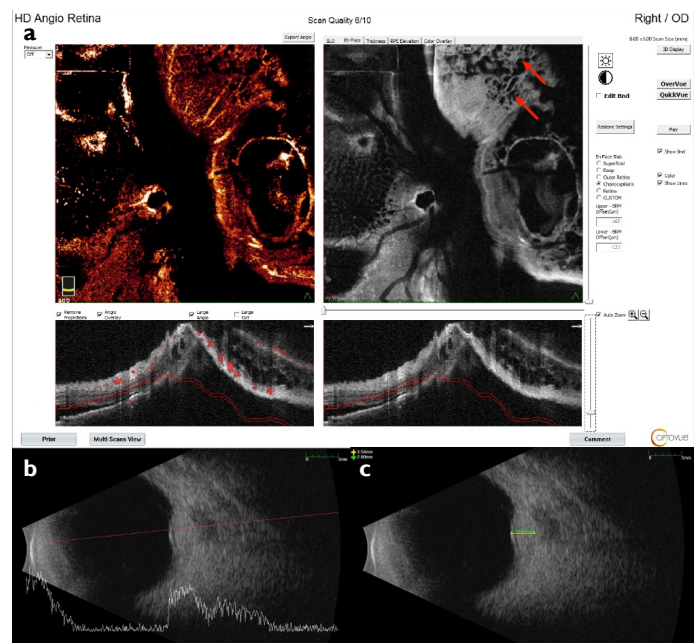


Later, the patient was admitted to our clinic due to lack of significant response to previous treatment. He was under systemic steroid treatment (60 mg/day) for 3 weeks at that time. BCVA was 0.5 (Snellen), IOP was 21 mmHg in his right eye. Slit-lamp examination revealed no anterior chamber cells or vitritis. There were centrally located focal RPE changes and orange pigmentation nasal to optic disk (OD) (Fig. 1). EDI-OCT showed peripapillary smooth, anterior



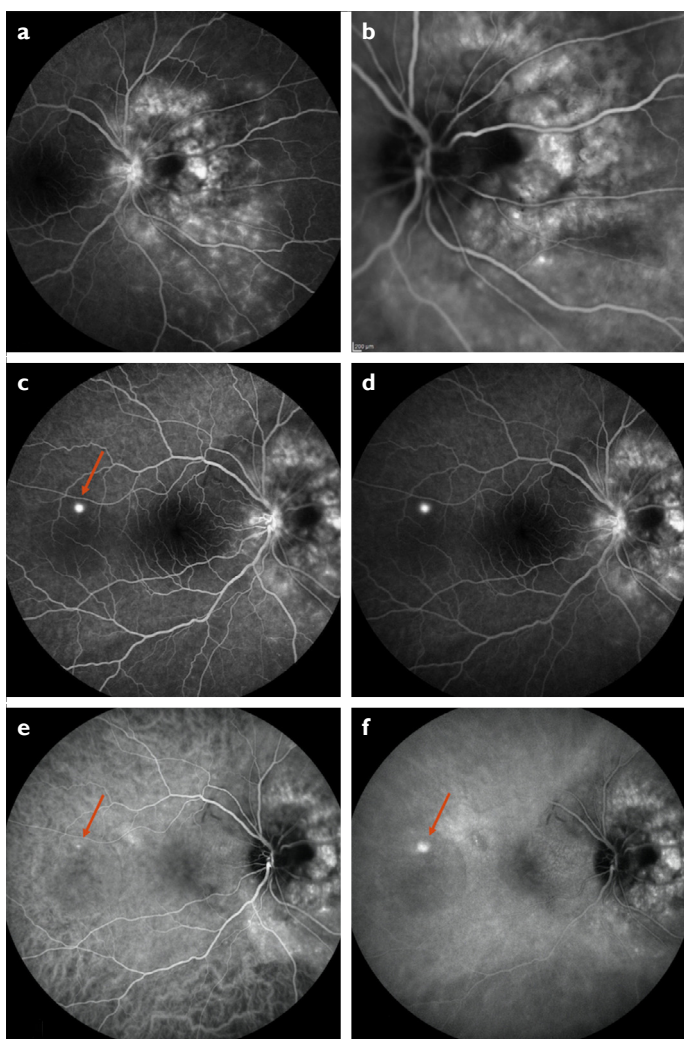
**Figure 1.** (a) Colored fundus photography and (b) fundus autofluorescence images at the time of admission to our clinic. In addition, enhanced depth imaging (EDI)-OCT findings indicating (c) low-to-medium homogeneous reflective signals at the choroidal layer of the peripapillary region, in addition to smooth, anterior sloping of RPE with subretinal and intraretinal fluid particularly nasal to the OD. (d) Subretinal fluid and intraretinal retinoschisis-like cystoid edema at the macula extending toward the OD, along with elevated subfoveal choroidal thickness. (e) Non-central, isolated serous retinal detachment temporal to the macula.

sloping of RPE, and subretinal and intraretinal fluid at the macula. In addition, isolated serous retinal detachment was presented temporal to macula. Subfoveal choroidal thickness (SFCT) was increased ( $523\mu$ ), indicating a pachychoroid spectrum (Fig. 1) (5). OCTA revealed well-defined network of interconnected vessels (Fig. 2a). B-scan ultrasonography (USG) indicated an acoustically solid lesion with high internal reflectivity (tumor thickness= $2.8\text{ mm}$ ) and associated retinal detachment (total thickness= $3.5\text{ mm}$ ) (Fig. 2b and c). A-scan USG showed typical see-saw pattern consistent with a choroidal hemangioma. FA and indocyanine green angiography (ICGA) established the presence of CCH and accompanying CSCR which might have worsened with systemic steroid treatment (Fig. 3). Orbital magnetic resonance imaging also demonstrated a tumor consistent with CCH which is hyperintense to vitreous in T1-weighted images and isointense to vitreous in T2-weighted images. The patient was then referred and managed by an ocular oncologist (ST). Systemic steroid treatment was discontinued with rapid tapering, and photodynamic therapy (PDT) was performed with multiple spots with a size of  $4400\mu\text{m}$  in three overlapping areas (overall lesion size= $7000\mu\text{m} \times 4000\mu\text{m}$ ), standard power of  $600\text{ mW/s}$ , and duration of 83 s for each area. The subretinal fluid (SRF) was totally resolved 2 months after PDT. The tumor thickness was reduced to 1.4 mm associated with small retinoschisis area over the tumor at 6 months of follow-up (Fig. 4). The images of the fellow eye are given in Figure 5.



**Figure 2.** (a) Optical coherence tomography angiography revealed well-defined network of interconnected varying sized enlarged vessels (red arrow). (b) B-scan ultrasonography indicating a solid lesion with high internal reflectivity and (c) associated retinal detachment (RD) (tumor thickness was  $2.8\text{ mm}$ , tumor + RD thickness was  $3.5\text{ mm}$ ).

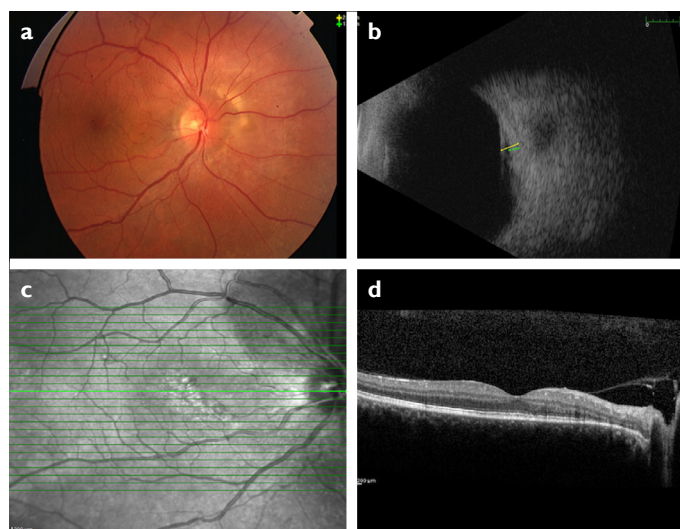




**Figure 3.** (a-d) Fluorescein angiography showed early spotty and late diffuse, intense hyperfluorescence nasal to the OD, in addition to early hyperfluorescent spot with gradual enlargement in the late phases (“inkblot” pattern) at the temporal region of the macula. (b-f) Indocyanine green angiography indicated intense hyperfluorescence of choroidal vessels in the arterial phase with late staining nasal to the OD, along with choroidal hyperpermeability temporal to the fovea.

## Discussion

Despite incidental diagnosis in asymptomatic cases, particularly young patients with CCH located nasal to OD often present with visual symptoms such as metamorphopsia, decreased BCVA, and VF defects which are related to SRF, cystoid edema, etc (1,2). The diagnosis is challenging especially in chronic lesions as they simulate CSCR or malignancies. Shields et al. (1) documented CSCR as the most likely non-tumor etiology among suspected diagnoses, since CCH might be linked to SRF. Therefore, to avoid unnecessary interventions, ancillary testing (USG, FAF, EDI-OCT, FA, ICGA, and lately OCTA) is essential for accurate diagnosis of CCH and associated changes.

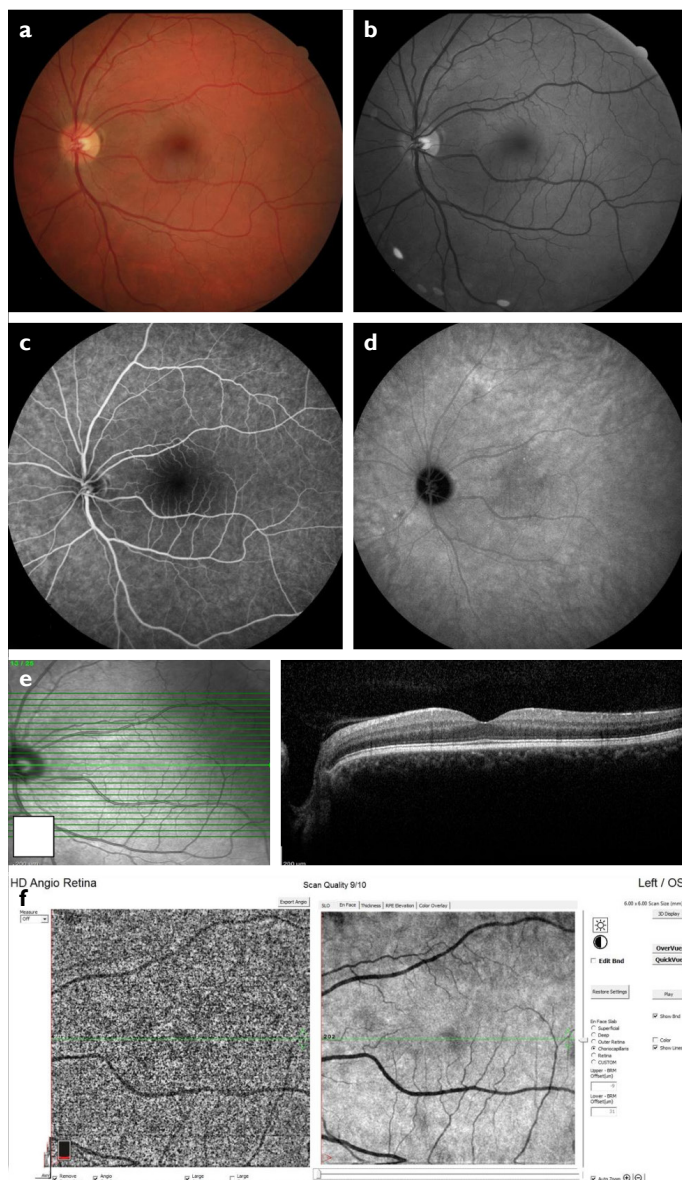


**Figure 4.** Six months after PDT (a) colored fundus photography, (b) B-scan ultrasonography showing tumor thickness reduced to 1.4 mm with resolved RD, and the presence of only retinoschisis over the OD. (c) OCT section demonstrating totally resolved subretinal and intraretinal fluid at the macula.

This report demonstrated chronic CCH presented with cystoid changes in macula and co-occurring CSCR in another localization of the same eye in young male patient. We documented multimodal imaging characteristics, including OCTA of these lesions.

Recently, Kim et al. (3) reported significantly elevated SFCT of fellow eyes of CCH patients in comparison to healthy controls. They also detected RPE alterations suggesting previous asymptomatic CSCR in 20% of fellow eyes, which is higher than the expected frequency in healthy population. Thus, they propose possible association of CCH with increased SFCT and CSCR, as the hyperpermeability of choroidal vessels is common pathophysiological mechanism in both diseases (3,6). Sobol et al. (4) reported similar SFCT increment in fellow eyes of CCH patients in larger cohort. In addition, they noted elevated SFCT in the same eye of patients with extramacular CCH indicating bilateral choroidal alterations in these patients. Likewise, despite extramacular localization of CCH and SFCT was elevated in the same eye of our case which also supported the presence of diffuse, bilateral changes in choroid.

Furthermore, a dense and irregular, disorganized choroidal vascular network, characterized with dilated medium and large choroidal vessels, is already known to be present within choroidal hemangioma (7,8). These vascular changes may explain associated SRF in CCH, as it possibly arise from leakage secondary to elevated hydrostatic pressure in tumoral choroidal vessels. Sobol et al. (4) demonstrated significantly more frequent vascular disorganization (unidentification or attenuation of typical size gradient of choroidal vessels) also



**Figure 5.** Fellow eye. (a) Colored fundus photography. (b) Red-free photography. (c) Fluorescein angiography. (d) Indocyanine green angiography. (e) Optical coherence tomography. (f) Optical coherence tomography angiography.

in fellow eyes of patients with CCH. Therefore, this generalized disturbance in choroidal vascular organization may be linked to diffuse hyperpermeable state, and subsequently, result in both CSCR and SRF linked to CCH itself. In light of these possible baseline alterations in choroid, choroidal abnormalities related to CCH appear to be more widespread than previously described (2,7). Hence, as in our case, it seems reasonable to observe co-occurrence of CCH and CSCR in the same eye of a patient.

Besides, OCTA is non-invasive complementary technique providing valuable information about vascular tissue within choroidal tumors like CCH, recently (7). This report also

established that CCH composed of thin capsule surrounding large hollow which may correspond to vascular lacunae and well-defined network of interconnected vessels. Giudice et al. (9) and Cennamo et al. (10) also indicated OCTA features of CCH both before and after treatment with PDT and ruthenium-106 brachytherapy, respectively. They demonstrated regression of flow in intralacunal vessels which appeared as dark vascular channels on OCTA, secondary to vaso-occlusive effects of treatments.

## Conclusion

Patients with CCH seem to exhibit generalized, diffuse choroidal alterations, and hyperpermeable state is possibly the common underlying pathophysiology for both CCH and CSCR. Therefore, CCH may conceivably present with accompanying CSCR. Besides, OCTA should be considered as noninvasive, reliable adjunctive tool for the diagnosis of CCH, in terms of visualizing vascular features of tumor.

## Disclosures

**Informed consent:** Written informed consent was obtained from the patient for the publication of the case report and the accompanying images.

**Peer-review:** Externally peer-reviewed.

**Conflict of Interest:** None declared.

**Authorship Contributions:** Concept – I.K., S.G.Y., J.M., S.T.; Design – I.K., S.G.Y., J.M., S.T.; Supervision – S.G.Y., S.T.; Resource – I.K., S.G.Y., J.M., G.M., S.T.; Materials – I.K., S.G.Y., J.M., G.M., S.T.; Data collection and/or processing – I.K., S.G.Y., J.M., G.M., S.T.; Analysis and/or interpretation – I.K., S.G.Y., S.T.; Literature search – I.K., S.G.Y., J.M., S.T.; Writing – I.K., S.G.Y., S.T.; Critical review – I.K., S.G.Y., S.T.

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