An Asymptomatic Circumscribed Choroidal Hemangioma: A Case Report

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Abstract
This paper reviews the clinical characteristics of a circumscribed choroidal hemangioma (CCH). This rare, benign ocular tumor often goes undiagnosed until it causes secondary complications such as macular edema. It is critical to differentiate a CCH from malignant posterior segment tumors such as choroidal melanoma. Ocular photodynamic therapy is an important treatment option for patients with symptomatic CCH.

Introduction
A circumscribed choroidal hemangioma (CCH) is a benign, vascular tumor of the choroid. Unlike diffuse choroidal hemangiomas, which are seen in younger patients with Sturge-Weber syndrome, a CCH has no systemic associations and is most often diagnosed in middle-aged patients. It is important to correctly diagnose a CCH as it can result in visual loss from secondary macular edema and can also be mistaken for a malignant choroidal tumor. Herein, we present a case of a CCH and discuss the management of these benign tumors.

Case Report
A 68-year-old Caucasian male presented to the eye clinic for a six-month follow-up visit for non-exudative age-related macular degeneration (ARMD) in both eyes. The patient’s visual acuity with correction was 20/20 in the right eye and 20/25 in the left eye, which was stable to previous visits. The examination findings were notable for an epiretinal membrane (ERM) and confluent macular drusen in both eyes. Additionally, in the right eye there was a non-pigmented, shallow dome-shaped lesion superior to the optic nerve head with indistinguishable borders (Figure 1); the lesion had no associated orange pigmentation, drusen, hemorrhage, or halos.

Diagnostic Testing:
Macular optical coherence tomography (OCT) testing revealed blunted foveal contour due to an ERM and scattered hyper-reflective disruptions of the retinal pigmented epithelium (RPE) consistent with drusen in both eyes; in the right eye, there was a focal area of subretinal fluid (SRF) associated with a hyperreflective sub-retinal lesion superior to fixation. A macular OCT scan over the lesion in the right eye displayed mild elevation of the retina.

Macular OCT-angiography (OCT-A) in the right eye showed a focal area of increased flow superior to fixation consistent with a choroidal neovascular membrane (CNVM); OCT-A of the left eye was unremarkable.

B-scan ultrasonography of the right eye revealed a minimally elevated acoustically solid choroidal lesion without subretinal fluid; the lesion was 1.3mm in thickness. A-scan ultrasonography showed moderate to high internal reflectivity of the lesion.

Figure 1. Fundus photograph of the right eye showing subtle circumscribed choroidal hemangioma.
Fluorescein angiography (FA) of the right eye exhibited focal stippled leakage in the mid and late phases superior to fixation consistent with an occult CNVM; there was an additional area of late stippled hyperfluorescence in a circumscribed area superior to the optic nerve head corresponding to the lesion seen on examination (Figure 2). The FA in the left eye showed mild staining of drusen in the later phases.

Indocyanine green angiography (ICG) of the right eye scan showed early hypercyanescence of the choroidal lesion superior to the optic nerve head with late phase “washed-out” hypocyanescence in a circumscribed area suggestive of a CCH (Figure 3A and 3B).

The patient was diagnosed with a CCH and new exudative ARMD in the right eye. Intravitreal anti-vascular endothelial growth factor (anti-VEGF) therapy was initiated for the right eye. After receiving an additional evaluation with an ocular oncologist, the CCH was managed conservatively with plans to recheck the lesion in six months.

Discussion:
Presentation:
A CCH is a congenital tumor most often diagnosed in adults in their 4th decade.\(^1,3\) These tumors are seen primarily in Caucasians and there is no sex predilection.\(^1,3,5\) Asymptomatic CCHs are usually found incidentally on fundus examination; however, over 70% of CCH patients are symptomatic at the time of diagnosis with the majority (80%) complaining of blurry vision/central scotoma.\(^1,4\) These tumors can range from 1 millimeter (mm) to 25mm with the average diameter being 6-9mm.\(^2-4\) The average tumor thickness is 2-4mm with a range of 0.5-11.5mm.\(^2-4\)

Diagnosis:
A CCH is often diagnosed based on clinical appearance and diagnostic tests. The color of these lesions varies but is often similar to the patient's background fundus appearance;\(^1,2,4\) in many cases, CCHs are only distinguishable due to the slight elevation of the lesion as was the case with our patient (Figure 1). The majority of CCHs are within the macula (67%) with the remainder being between the macula and the equator (33%).\(^1,7\)

Ancillary testing can be helpful in identifying a CCH. An OCT may show thickening of the choroid in the region of the CCH as well as any secondary SRF.\(^5,6\) An OCT-A can show the abnormal vasculature appearance associated with CCH.\(^6\) Additional testing with A- and B-scan ultrasound, FA, and ICG also can be helpful in differentiating CCH from other posterior segment tumors, including a choroidal nevus, choroidal osteoma, and choroidal metastasis/melanoma (Table 1).

Management:
Patients with asymptomatic CCHs or those with poor visual outcomes (e.g., chronic macular edema) are usually observed.\(^1,3\) Current management of a symptomatic CCH is centered upon the use of ocular photodynamic therapy (PDT).\(^1,2,12\) Ocular PDT eliminates macular SRF and can shrink the size of the tumor.\(^1,2\)
Several small retrospective cohort studies suggest that PDT is a safe and effective treatment option for symptomatic CCH patients.13,15 Newer treatment options include intravitreal anti-VEGF and steroid injections to reduce chronic macular edema and preserve visual function.1,2

Conclusion:
The CCH is a rare benign tumor that should be distinguished from malignant tumors such as choroidal melanoma. A CCH can result in visual loss from macular edema; OCT-A may be a useful non-invasive tool for the early detection of a CCH. Emerging treatment options may include intravitreal anti-VEGF and steroid injections to reduce chronic macular edema and preserve vision.1,2

References