



## Case report

## Solitary retinal hemangioblastoma findings in OCTA pre- and post-laser therapy



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## ABSTRACT

**Purpose:** To report optical coherence tomography angiography (OCTA) findings pre- and post-thermal laser therapy of a rare, asymptomatic, unilateral, isolated retinal hemangioblastoma.

**Observations:** A 40-year-old asymptomatic Caucasian man was found to have an isolated, unilateral retinal hemangioblastoma after referral for possible retinal hole. Comparison of OCTAs pre- and post-thermal laser therapy demonstrated a significant reduction of blood flow toward and within the lesion, in addition to contraction of the lesion and constriction of the accompanying feeder vessels. A 4-month follow-up OCTA showed marked reduction of lesion size, and caliber of treated vessels.

**Conclusions and importance:** Non-invasive OCTA imaging of a retinal hemangioblastoma demonstrated decreased blood flow signal and vessel caliber after treatment. OCTA shows potential as a new tool to improve our understanding of the effectiveness of therapy in these lesions.

## 1. Introduction

Retinal hemangioblastomas, also known as retinal capillary hemangiomas, are lesions composed of enlarged clusters of retinal capillary endothelial cells with dilated retinal vessels feeding and draining the tumor. While the tumor itself is benign, treatment may be required when vision is compromised by subretinal or intraretinal exudation involving the macula.<sup>1</sup>

Optical coherence tomography angiography (OCTA) has emerged as a new, non-invasive imaging modality for visualizing the retinal blood vessels.<sup>2</sup> The broad applications of this technology are still being realized, and we present a case to demonstrate the applicability of OCTA in the evaluation of retinal hemangioblastoma and the response to therapy.

## 2. Case report

A 40-year-old asymptomatic Caucasian man was found to have an isolated, unilateral retinal hemangioblastoma after referral for potential retinal hole. Patient ocular and systemic history was significant only for mild myopia (-2.00 diopters OU).

On examination, visual acuity was 20/20 OU with glasses correction. Slit lamp evaluation was unremarkable. On dilated examination of the retina, the right eye revealed a lesion consistent with a hemangioblastoma at the inferotemporal arcade (Fig. 1). Fluorescein angiography

(FA) of the right eye demonstrated a singular mass of the temporal quadrant with increasing hyperfluorescence. Two retinal arterioles appeared to enter the lesion with one large draining vein (Fig. 2). OCT of the right eye demonstrated an isoreflective vascular mass throughout most of the layers of the retina with no surrounding fluid or exudates. OCTA showed a robust network of blood vessels with flow signal in the lesion (Fig. 3). OCTA also clearly delineated the three dilated and slightly tortuous feeder vessels, which appeared to anastomose within the lesion. These classical clinical findings of dilated, tortuous vessels leading to and away from a vascular tumor were consistent with the diagnosis of retinal hemangioblastoma. The patient was sent for genetic testing of the von Hippel-Lindau tumor suppression gene and a follow up visit for treatment was scheduled.

Genetic testing results came back negative. At the follow-up visit, after discussion with the patient, thermal laser therapy was initiated. An argon green laser set at 200-micron spot size, .30 sec duration with power setting at 300-350 mW was applied first to the two arterioles and then to the one venule blanching them to a point where no flow was seen within the vessel column. The lesion itself was then lasered. Fifteen minutes post therapy, OCTA demonstrated a discontinuity of the flow through the lasered vessels surrounding the lesion and decrease flow within the lesion itself. OCTA also showed an overall constriction of blood vessels within and associated with the lesion (Fig. 3).

At 4-month follow-up, the patient was doing well, and remained asymptomatic. The repeated OCTA demonstrated further constriction

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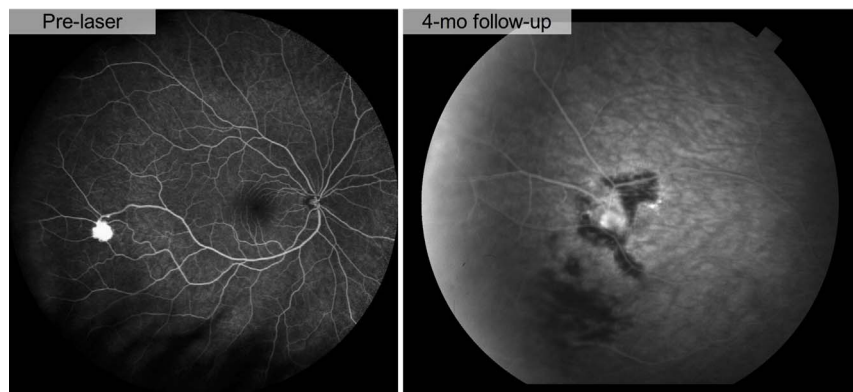


**Fig. 1. Fundus Photos Pre-laser Therapy and at 4-month Follow-up.** Top: Color fundus photo prior to laser therapy shows the retinal hemangioblastoma with two dilated feeder arterioles entering and one vein exiting the lesion. Bottom: 4-month follow-up reveals the fibrosed lesion and laser scars overlying the feeding and draining vessels. The vessels appear constricted compared to pre-laser images. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

and straightening of the lasered vessels. One lasered arteriole no longer appeared to enter the retinal hemangioblastoma. The remaining arteriole and venule had persistent flow and appeared to anastomose within the lesion, and the lesion itself was much smaller (Fig. 3).

### 3. Discussion

While retinal hemangioblastomas most often occur in the setting of von Hippel-Lindau syndrome, they have also been reported to appear as an isolated clinical entity, as our case described above.<sup>3</sup> These lesions may be present on the peripheral retina, but our case demonstrates a more posterior lesion, allowing for clearer imaging. Other vascular tumors may mimic a retinal hemangioblastoma, including a racemose hemangioma, retinal cavernous hemangioma, or retina macroaneurysm, but our case is most consistent with the clinical appearance of a retinal hemangioblastoma. Screening to rule out von Hippel-Lindau syndrome should be initiated due to the threat of systemic lesions,



**Fig. 2. Fluorescein Angiograms Pre-laser Therapy and at 4-month Follow-up.** Left: Pre-laser fluorescein with marked hyperfluorescence at 56 seconds, delineating the single isolated lesion with specific margins. Lesion feeder vessels are well-visualized. Right: A close up image at 4-month follow-up demonstrates scarring as well as reduced hyperfluorescence of the lesion and feeder vessels at 57 seconds.

including pancreatic cysts, pheochromocytoma, or renal cell carcinoma.<sup>4</sup> If available, molecular genetic analysis has emerged as a means for a definite diagnosis of VHL, as was performed in this case.

The treatment of any retinal hemangioblastoma is dependent on size and macular involvement. Smaller lesions, especially if detected early, are typically treated via photodocumentation or laser photocoagulation, often with patient preference. Peripheral lesions less than 3.00mm are most often treated with laser therapy shortly after diagnosis, before any threat of exudation. Laser applied to the lesion and the surrounding area promotes scarring and involution of feeder vessels.<sup>5</sup> Larger lesions typically require cryotherapy or plaque radiotherapy. Vitreoretinal surgery is another option, but is typically reserved for resistant cases.<sup>6</sup> The use of vascular endothelial growth factor (VEGF) inhibitors has recently risen as an additional therapy with mixed treatment outcomes for these types of lesions.<sup>7</sup>

Previously, outcome measures of these therapies consisted of patient visual outcome and/or appearance on OCT and FA.<sup>8</sup> More recently, OCTA has been used as a tool to evaluate the retinal blood vessels with depth resolved accuracy.<sup>9</sup> In our case, we show the application of OCTA in the visualization and evaluation of retinal hemangioblastomas. With OCTA, flow to the lesion can now be visualized and quantified, suggesting the potential of this modality for evaluating the outcome of laser therapy.

In summary, we report OCTA findings of pre- and post-thermal laser therapy of a retinal hemangioblastoma. OCTA is a relatively new imaging modality that should be considered for assessment of therapy effectiveness. Thermal laser therapy has been the mainstay of treatment for retinal hemangioblastomas, in both isolated cases as well as in von Hippel-Lindau Syndrome, and we anticipate OCTA will become a valuable tool for post-procedure evaluation.

### Patient consent

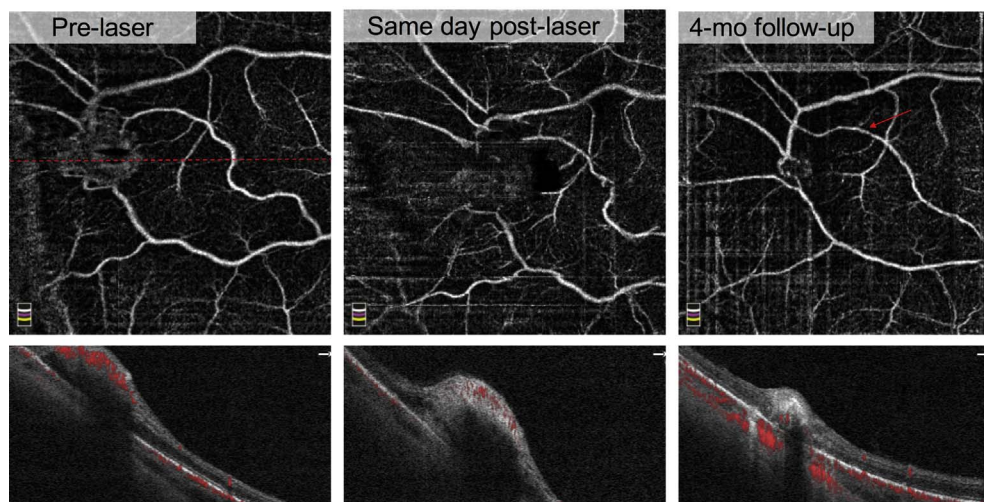
This patient was recruited in 2016 in the Department of Ophthalmology at Northwestern University in Chicago, Illinois and written informed consent was obtained. The study was approved by the Institutional Review Board of Northwestern University, followed the tenets of the Declaration of Helsinki and was performed in accordance with the Health Insurance Portability and Accountability Act regulations.

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### Conflicts of interest

The authors have no financial disclosures.



**Fig. 3.** OCT angiography (OCTA) at Pre-, Post- and 4-month Follow-up.

Left: Pre-laser *en face* OCTA shows the three major, somewhat tortuous vessels associated with the retinal hemangioblastoma. The dotted line shows the location of the B-scans, which include a red pixel overlay that represents blood flow. Pre-laser cross-sectional OCTA shows appreciable flow through the lesion. Middle: OCTA immediately post-laser reveals discontinuity of flow through the major vessels surrounding the lesion on *en face* and decreased flow within the lesion itself on *en face* and cross-section. Such discontinuity may be attributed to involution of the vessels or blockage due to swollen retinal tissue. Right: At 4 months, the vessels are less tortuous and one vessel appears to no longer enter the lesion (arrow). The lesion itself is much smaller and there is flow from the remaining arteriole to the draining venule. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

### Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

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