Eye (2020) 34:2345–2346
https://doi.org/10.1038/s41433-020-0792-5

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CORRESPONDENCE

Use of optical coherence tomography angiography in the diagnosis of small retina lesions in Von Hippel–Lindau disease

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Received: 27 December 2019 / Revised: 19 January 2020 / Accepted: 28 January 2020 / Published online: 10 February 2020

To the Editor:

Von Hippel–Lindau disease (VHL) is a neoplastic disorder that can lead to the multisystem formation of tumors and cysts in the central nervous system, adrenal glands, kidneys, pancreas, reproductive system, and retina [1]. The earliest ocular findings of VHL are retinal capillary hemangiomas, and loss of vision can result from tumor growth, extravasation of fluid and lipid exudates from the lesions, and retinal detachment. The evaluation of retinal manifestations of VHL includes the clinical exam and fundus fluorescein angiography (FFA), but FFA is invasive and findings of small lesions can be equivocal, making the decision for treatment challenging. Optical coherence tomography with angiography (OCTA) is a novel, noninvasive imaging technique that can provide in vivo images of the retinal vasculature with higher resolution than conventional techniques. However, the ability of OCTA to confirm both the presence and absence of potential retinal lesions seen with FFA in VHL has not yet been described. We have observed two cases of retinal hemangiomas with a systemic history of VHL.

Case 1

A 22-year-old female with VHL and a history of previously treated retinal hemangiomas in both eyes presented for annual evaluation. FFA of the right eye (Fig. 1a, upper left corner) showed an area of hyperfluorescence below the inferior arcade, which was concerning for a small posterior lesion that had not been visualized on dilated fundus exam (Fig. 1a, larger image). OCTA confirmed a very small, new subclinical angioma (Fig. 1b) and laser treatment was recommended.

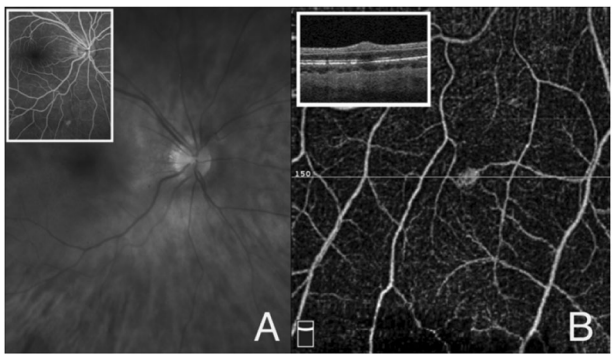

Fig. 1 Case 1 - OCTA confirms presence of a retinal angioma in a VHL patient with equivocal findings on fundus photo and FFA. Fundus photo (**a** larger image), FFA (**a** upper left corner), and en face OCTA images (**b**) of the subclinical retinal angioma in Case 1. Fluorescein angiography (**a**) shows an area of hyperfluorescence below the inferior arcade not seen in the fundus image or exam. En face OCTA imaging (**b**) of that area confirms the presence of a retinal angioma with blood flow.

Case 2

A 45-year-old male with VHL and systemic manifestations in his adrenal glands, spine, and kidneys presented for annual evaluation. Fundus autofluorescence (FAF) (Fig. 2a) showed a small area of irregularity concerning hemangioma in the inferotemporal quadrant of the right eye that was not clearly discernable on exam or fundus photo (Fig. 2b, larger image). FFA evaluation of the lesion showed an area of hyperfluorescence at the inferotemporal secondary branch in the same area concerning for a small, early angioma (Fig. 2b, upper left corner). OCTA of the area showed no

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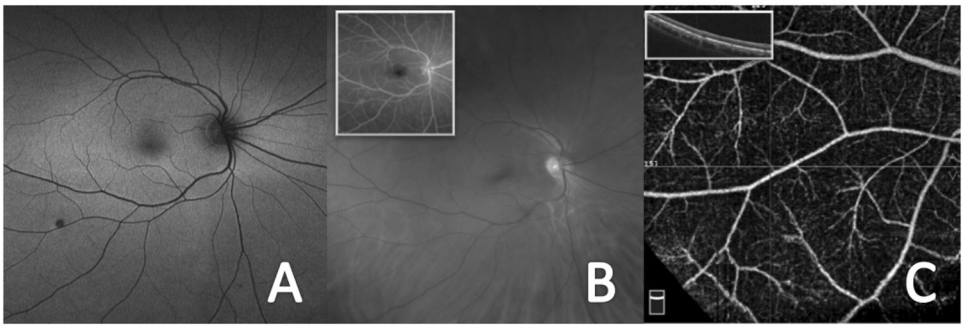

Fig. 2 Case 2 - OCTA confirms absence of angiomatous lesion in a VHL patient with irregular lesion seen on FA and FAF but not fundus photo. Fundus autofluorescence (**a**), color fundus photo (**b** larger image), FFA (**b** upper left corner), *en face* OCTA images (**c**) in Case 2. Fundus photo (**b**) of the inferior retina is unremarkable, but the fundus autofluorescence (**a**) and FFA (**b** upper left corner) highlighted an inferotemporal irregularity concerning for a subclinical small angioma. Further assessment with OCTA (**c**) did not show evidence of a vascular lesion.

evidence of a vascular lesion, and the decision was made to monitor the patient (Fig. 2c).

As demonstrated in the cases described, OCTA has promising potential as an adjunctive tool to enhance the evaluation and management of patients with VHL. In Case 1, OCTA was able to confirm the presence of a vascular lesion that had not been observed on the dilated fundus exam and an equivocal appearance on FFA. In Case 2, using OCTA in the evaluation of a small area of hyperfluorescence observed with FFA demonstrated that there was no hemangioma. In both cases, the use of OCTA aided in the clinical management of the patients. The frequency of retinal lesions ranges from 49 to 85% in individuals with VHL [2, 3] and the potential damage to the retinal tissue and subsequent vision loss underscore the importance of high-quality clinical imaging in the ophthalmic management of this patient population. The size, location, and progression of the ocular lesions in VHL guide the decision for appropriate therapy, which includes laser photocoagulation, cryotherapy, diathermy, external beam radiation, plaque brachytherapy, intravitreal anti-VEGF administration, and pars plana vitrectomy. OCTA has been used to identify feeding and draining vessels and to characterize lesions after laser treatment of retinal angiomas that were previously diagnosed by exam and/or FFA [4]. Limitations of OCTA include its inability to image the peripheral retina. Although OCTA is a relatively new technology, its increased utilization and future advances in its imaging capabilities may improve the medical management of patients with suspected vascular lesions of their retina.

Acknowledgements We thank our colleagues, Blake L. Williams MD, and Sidney A. Schechet MD from the University of Chicago, for comments that greatly improved the case report. We thank all our reviewers for their comments and reviews. We certified that all images used in this correspondence are our own images and came from the department of Ophthalmology and Vision Science at the University of Chicago, Chicago, IL, USA.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

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