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Optical coherence tomography angiography of iris microhemangiomatosis



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ABSTRACT

Purpose: To report optical coherence tomography angiography (OCTA) of iris microhemangiomatosis. *Observations:* A 75-year-old asymptomatic Caucasian man was found to have bilateral pupillary vascular lesions during cataract evaluation. Visual acuity was counting fingers in the right eye (OD) and 20/40 in the left eye (OS) with normal intraocular pressures in both eyes (OU). In each eye there were multifocal, round, dark red, pinpoint vascular tufts at the pupillary margin, randomly distributed and numbering 1 in OD and 7 in OS, each measuring 0.2–0.3 mm in diameter and without active bleeding or hyphema. Fundus examination OU was normal. By fluorescein angiography, the multifocal pupillary vascular tufts demonstrated mild staining without leakage. By OCTA, the tufts were clearly delineated and were fed by normal appearing radial iris vessels. OCT b-scan documented the optically dense vascular tufts at 0.1 mm in thickness and angio-overlay confirmed blood flow emanating from the deep iris stroma. Observation was recommended with the option of cataract surgery to improve vision.

Conclusions and importance: Non-invasive imaging of iris microhemangiomatosis with OCTA delineates the vascular lesion with flow arising from the posterior iris stroma.

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1. Introduction

Iris microhemangiomatosis is a benign vascular lesion that can lead to intermittent blurred vision, hyphema, and elevated intraocular pressure.^{1–4} Currently, the diagnosis is made by recognition at slit-lamp biomicroscopy and confirmed with anterior segment intravenous fluorescein angiography (FA).⁵ This tiny lesion, however, is often overlooked and in some cases, can be confused with iris neovascularization.

Recently, optical coherence tomography angiography (OCTA) has emerged as a non-invasive vascular imaging modality, allowing segmental analysis of vascular flow. OCTA is used primarily for retinal imaging, but adjustments in technique can provide non-invasive vascular imaging of the anterior segment.^{6–9} Herein, we report a case of iris microhemangiomatosis adequately imaged with OCTA, providing information in a non-invasive fashion.

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2. Case report

A 75-year-old asymptomatic Caucasian man was noted to have pupillary margin iris abnormalities in both eyes (OU) during cataract evaluation. He denied prior episodes of blurred vision or hyphema. Past medical history revealed controlled hypertension and there was no previous ocular or family history.

On examination, visual acuity was counting fingers in the right eye (OD) and 20/40 in the left eye (OS) with normal intraocular pressures OU. Slit lamp biomicroscopy disclosed nuclear sclerosis OU and posterior subcapsular cataract OD. In addition, there were multifocal pinpoint, dark red, vascular tufts at the pupillary margin OU (Fig. 1). The right eye demonstrated a single lesion at 12:00 o'clock measuring 0.3 mm in width while OS had seven lesions measuring 0.2 mm–0.3 mm in width located at 1:00, 3:00, 4:00, 5:00, 6:00, 9:00, and 10:00. There was no hyphema, neovascularization, or inflammation. Fundus examination OU was normal.

By anterior segment fluorescein angiography (FA), the vascular tufts were barely visible as pinpoint areas of pupillary margin staining and no leakage. By optical coherence tomography angiography (OCTA) (Optovue RTVue Avanti XR, Optovue Inc, Fremont

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Case report

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Fig. 1. A 75-year old Caucasian man with bilateral iris microhemangiomatosis. Slit-lamp photograph (A) of the left eye revealed multifocal iris microhemangiomatosis, each manifesting as a pinpoint red tuft at the pupillary margin (arrows). On optical coherence tomography angiography (OCTA) (B), six tightly coiled vascular tufts (arrows) are noted, fed by non-dilated iris vessels. OCT b-scan (C) through one of the lesions demonstrated an optically dense tuft resting on the lens with normal iris stroma and iris pigmented epithelium. OCT b-scan with flow overlay (red) (D) confirmed the vascular flow within the mass, originating from the deep iris stroma. The flow, seen at the iris pigmented epithelium level, could be projection artifact and could represent flow in the stroma more superficially. (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

CA, USA), using the anterior segment module, the en-face image showed tightly coiled pupillary margin vascular loops fed by nondilated, normal-appearing iris vessels (Fig. 1). By OCT b-scan, each tuft appeared optically dense and was located at the level of the posterior iris, resting on the lens (Fig. 1). The OCT flow overlay confirmed circulation within the tufts with flow emanating from vessels of the deep iris stroma (Fig. 1). These findings were consistent with iris microhemangiomatosis and, given the absence of hyphema, observation was advised with the option of cataract surgery to improve vision.

3. Discussion

Vascular lesions of the iris are rare, representing 2% of all iris tumors.¹ Iris microhemangiomatosis is a benign vascular lesion consisting of minute clusters of tightly coiled vascular loops at the pupillary margin that can lead to spontaneous hyphema with occasional increased intraocular pressure.^{2–4} While the etiology remains unknown, association has been made with myotonic dystrophy and idiopathic juxtafoveolar retinal telangiectasia.^{10,1} No correlation exists for gender or race; however, a retrospective study of 3680 iris tumors found that iris microhemangiomatosis was observed mostly in patients above the age of 60 years.¹ Management of the lesion includes observation or argon laser photocoagulation for recurrent hyphema.^{4,12,13}

The diagnosis of iris microhemangiomatosis is often overlooked. The condition is typically established at slit-lamp biomicroscopy with occasional visualization of active bleeding from a lesion.¹ Anterior segment fluorescein angiography (FA) can be helpful in characterizing the lesion and ruling out iris neovascularization. On FA, the lesion demonstrates early hyperfluorescence with late staining and often involves more of the pupillary margin than was visualized clinically.^{5,13} While FA is helpful in supporting the diagnosis, its invasive nature limits its use in clinical practice.

A newer technology, optical coherence tomography angiography (OCTA), allows non-invasive visualization of ocular blood vessels. OCTA has advantages over FA including no need for intravenous dye injection, rapid speed of image acquisition, and threedimensional visualization of ocular tissue permitting segmental analysis of microvascular anatomy. Although there has been limited work regarding OCTA of the anterior segment^{6–9} (Li Y, et al. IOVS

2015; 56: ARVO E-Abstract 4512), Skalet et al. have reported OCTA in iris melanocytic lesions and Allegrini et al. have reported OCTA in iris nevus.^{8,9} This is the first report of OCTA in iris microhemangiomatosis.

The en-face OCTA image in this case demonstrates non-dilated, normal-appearing iris vessels giving rise to tightly coiled vascular tufts at the pupillary margin. Previous histopathologic evaluation demonstrated iris microhemangiomatosis as a hamartomatous vascular mass with thick-walled stromal blood vessels surrounded by loose connective tissue.¹⁴ Analysis of the cross-sectional OCT bscans with angio-overlay in this case localized the microhemangiomatosis to the posterior iris stroma. This 3-dimensional segmental analysis is of value compared to 2-dimensional FA and distinguishes iris microhemangiomatosis from iris neovascularization which localizes more anteriorly.¹⁵ However, as with OCTA of the posterior segment, projection artifact is present with OCTA of the iris.^{16,17} Much of the flow seen at the level of the iris pigmented epithelium on cross-sectional OCT b-scan with angiooverlay could be artifactual, similar to projection artifact seen with posterior segment imaging. In both OCTA and FA, skilled ophthalmic photography is necessary as motion artifact can significantly hinder adequate image acquisition. Future work may include comparison of FA with OCTA for different disease processes of the anterior segment, as has been done with imaging of the retina.18

In summary, this is the first report of OCTA of iris microhemangiomatosis. Although FA has traditionally been effective in highlighting iris vascular lesions, the noninvasive nature and depth-localizing strengths of OCTA are appealing. We anticipate that OCTA will prove to be a valuable tool for anterior segment vascular imaging.

Patient consent

Consent to publish the report has been obtained from the patient in writing.

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Authorship

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

Conflict of interest

No conflicting relationship exists for any author.

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