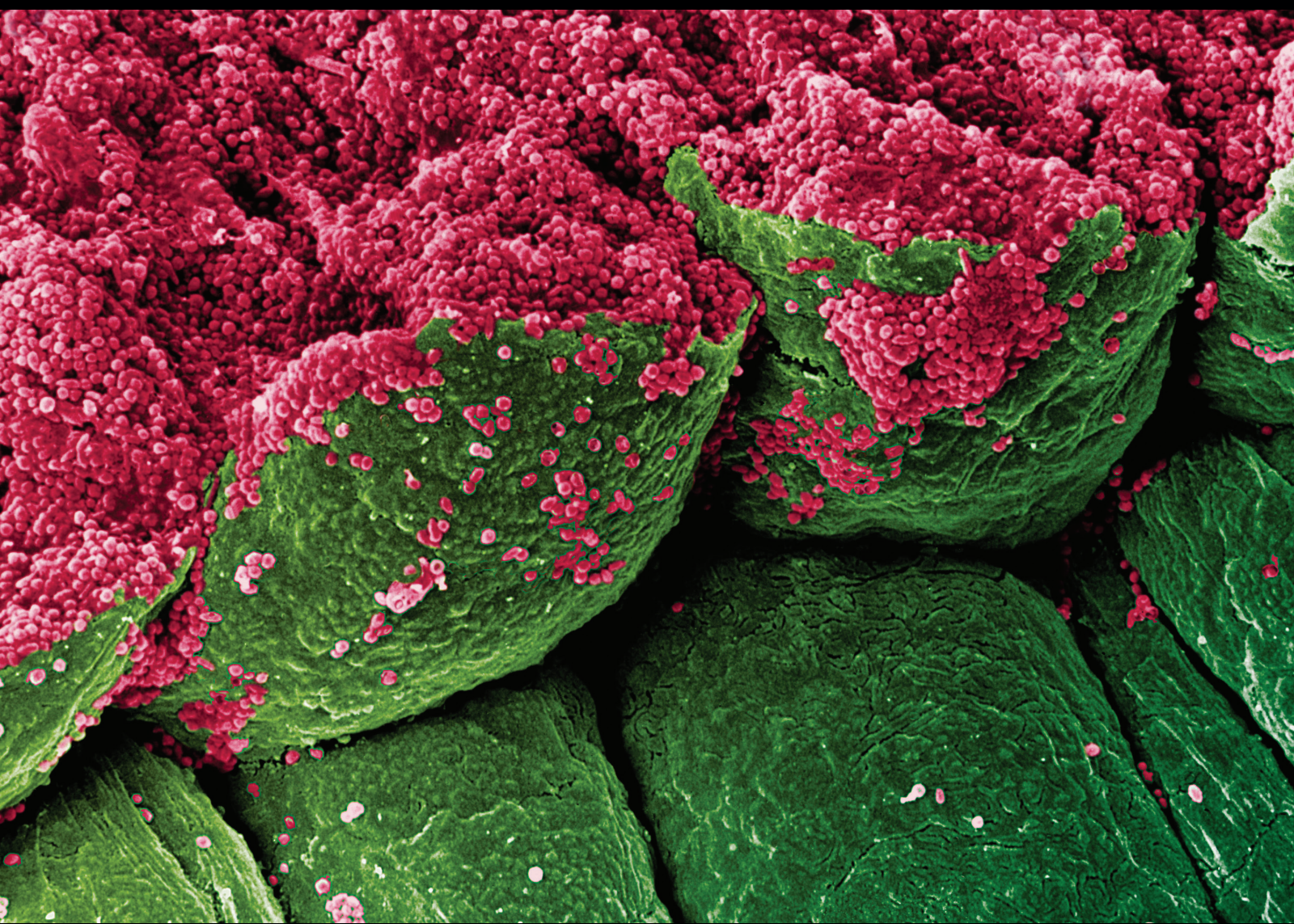
Case Reports in Ophthalmological Medicine

Optical Coherence Tomography Angiography

Special Issue Editor in Chief: Stephen G. Schwartz
Guest Editors: Harry W. Flynn, Andrzej Grzybowski, Avinash Pathengay, and Ingrid U. Scott



Optical Coherence Tomography Angiography

Case Reports in Ophthalmological Medicine

Optical Coherence Tomography Angiography

Special Issue Editor in Chief: Stephen G. Schwartz
Guest Editors: Harry W. Flynn, Andrzej Grzybowski, Avinash Pathengay, and Ingrid U. Scott

Editorial Board

Contents

Hindawi
Case Reports in Ophthalmological Medicine
Volume 2018, Article ID 7140164, 2 pages
https://doi.org/10.1155/2018/7140164

Editorial

Optical Coherence Tomography Angiography

Stephen G. Schwartz,[1] **Harry W. Flynn Jr.**,[1] **Andrzej Grzybowski**,[2,3] **Avinash Pathengay**,[4] **and Ingrid U. Scott**[5,6]

[1]*Department of Ophthalmology, Bascom Palmer Eye Institute, University of Miami Miller School of Medicine, Miami, FL, USA*
[2]*Institute for Research in Ophthalmology, Poznan, Poland*
[3]*Department of Ophthalmology, University of Warmia and Mazury, Olsztyn, Poland*
[4]*Retina and Uveitis Department, LV Prasad Eye Institute, GMR Varalakshmi Campus, Visakhapatnam, Andhra Pradesh, India*
[5]*Department of Ophthalmology, Penn State College of Medicine, Hershey, PA, USA*
[6]*Department of Public Health Sciences, Penn State College of Medicine, Hershey, PA, USA*

Correspondence should be addressed to Stephen G. Schwartz; sschwartz2@med.miami.edu

Received 25 March 2018; Accepted 26 March 2018; Published 6 June 2018

Optical coherence tomography angiography (OCT-A) is a relatively new, noninvasive outpatient imaging test that provides both structural and functional information about the macula and midperipheral retina. OCT-A is complementary to traditional imaging modalities, such as fundus photography, fluorescein angiography (FA), and spectral domain optical coherence tomography (SD-OCT).

Advantages of OCT-A include visualization of vascular flow signal, *en face* imaging, no need for dye injection, and segmentation of the posterior segment structures from the vitreomacular interface, through the retinal layers, and to the choroid. Disadvantages of OCT-A include increased costs and longer acquisition times than SD-OCT. Because of the longer acquisition times, it may be difficult to obtain high-quality OCT-A images in eyes with poor visual acuity that cannot fixate well.

The use of OCT-A is currently best established in the care of patients with retinal vascular diseases, choroidal vascular diseases, and ophthalmic oncology. Its role appears to be emerging in patients with other macular and retinal diseases (including uveitis) as well as optic nerve diseases and glaucoma.

This special issue, which had opened for 6 months in the second half of 2017, focuses on various clinical applications of OCT-A.

V. M. Villegas et al. present two patients with choroidal nevi, including one halo nevus, and report decreased vascular flow signal in most or all layers on OCT-A. This decreased vascular flow signal is proposed to be due to blockage from the choroidal nevus, true diminished blood flow (ischemia), or unknown causes. Because choroidal melanoma frequently demonstrates increased vascularity on FA, the authors propose that OCT-A might represent a noninvasive test to screen suspicious nevi for evidence of early malignant transformation.

V. Shah et al. report one patient with a unilateral congenital retinal macrovessel in the macula. OCT-A demonstrates replacement of the normal foveal avascular zone (FAZ) by abnormal vascular bifurcations, yet this disturbance of the FAZ is associated with relatively normal foveal anatomy, as imaged by swept-source OCT, and a best-corrected visual acuity of 20/20.

B. M. Hampton et al. present one patient with bilateral choroidal neovascularization (CNV) due to punctate inner choroidopathy (PIC). In this patient, OCT-A demonstrates bilateral submacular abnormal vessels consistent with CNV, and FA demonstrates late vascular leakage, confirming the diagnosis.

V. M. Villegas and J. L. Kovach report one patient with bilateral macular telangiectasia type 2 (MacTel2) and unilateral subretinal neovascularization (SNV). In the eye with SNV, OCT-A demonstrates abnormal submacular vessels consistent with SNV. In the fellow eye, OCT-A demonstrates

abnormal vessels temporal to the center of the macula consistent with nonproliferative MacTel2.

P. Shah et al. present two patients with acute central retinal artery occlusion (CRAO), including one with cilioretinal artery sparing, imaged with both FA and OCT-A. In both patients, the images obtained by FA and OCT-A are very similar. Because of this similarity, the authors propose that patients with acute CRAO in whom OCT-A can be obtained might not require additional imaging with FA.

M. Kaya et al. report one patient with chronic combined cilioretinal artery occlusion and central retinal vein occlusion (CRVO). In this patient, FA performed 10 months after the combined occlusion is relatively normal but OCT-A demonstrates a wedge-shaped area of decreased vascular flow signal consistent with the cilioretinal artery occlusion. The authors propose that different imaging studies may be relatively more useful in the acute and chronic phases of this disease.

S. Wu et al. present a somewhat similar patient with combined CRAO and CRVO. In this patient, symptoms began immediately following cataract surgery with retrobulbar anesthesia. OCT-A demonstrates profound decreased vascular flow signal in the superficial and deep retinal plexuses but relatively normal vascular flow signal in the choriocapillaris and choroid.

T. Y. A. Liu et al. report one patient with CNV due to presumed ocular histoplasmosis syndrome (POHS). Three monthly injections of intravitreal bevacizumab were given. On follow-up examinations up to 6 months, CNV activity is not detectable by FA or SD-OCT but is detectable by OCT-A. The authors propose that OCT-A might be more sensitive than FA for this indication.

H. Hamoudi et al. present one patient with unilateral Purtscher retinopathy following thoracic trauma sustained in a motor vehicle accident. OCT-A demonstrates decreased vascular flow signal in the superficial and deep retinal capillary plexuses of the affected eye and is normal in the fellow eye.

V. S. Chang et al. report one patient with unilateral retinal arterial macroaneurysm (RAM) treated with two injections of intravitreal bevacizumab. At presentation, OCT-A demonstrated increased vascular flow signal in the walls of the RAM. After treatment with bevacizumab was initiated, follow-up OCT-A studies demonstrate progressive reduction in the vascular flow signal within the walls of the RAM, suggesting progressive sclerosis of the lesion.

A. Fukutomi et al. present one patient with nonischemic CRVO that progressed to ischemic CRVO. At presentation, OCT-A demonstrated some reduction in vascular flow signal in the small capillaries but relatively preserved vascular flow signal surrounding the FAZ. Subsequent OCT-A studies demonstrate progressive expansion of the area of decreased vascular signal flow, consistent with progressive expansion of capillary nonperfusion.

In summary, this special issue contains many interesting case reports, which collectively illustrate many uses of OCT-A in patients with retinal vascular disease, choroidal vascular disease, and ocular oncology.

Acknowledgments

The Guest Editors express great appreciation to all authors for their excellent contributions and to all peer reviewers for their critical assistance. In addition, the Guest Editors thank the Editorial Board of this journal for their approval of this important topic and their continuous support in successful publication of this special issue. Finally, the Lead Guest Editor wishes to thank his four colleagues for their valuable expertise and dedication. We are very confident that this special issue increases our understanding of OCT-A as an emerging technology that can help clinicians better manage their patients.

Stephen G. Schwartz
Harry W. Flynn Jr.
Andrzej Grzybowski
Avinash Pathengay
Ingrid U. Scott

Hindawi
Case Reports in Ophthalmological Medicine
Volume 2018, Article ID 1354217, 4 pages
https://doi.org/10.1155/2018/1354217

Case Report

Sequential Observations of Conversion from Nonischemic to Ischemic Central Retinal Vein Occlusion Using Optical Coherence Tomography Angiography

Akira Fukutomi, Kotaro Tsuboi, Hikari Ono, Yuichiro Ishida, and Motohiro Kamei

Department of Ophthalmology, Aichi Medical University, Nagakute, Japan

Correspondence should be addressed to Kotaro Tsuboi; tsuboi.koutarou.230@mail.aichi-med-u.ac.jp

Received 28 December 2017; Revised 1 March 2018; Accepted 20 March 2018; Published 23 April 2018

Academic Editor: Stephen G. Schwartz

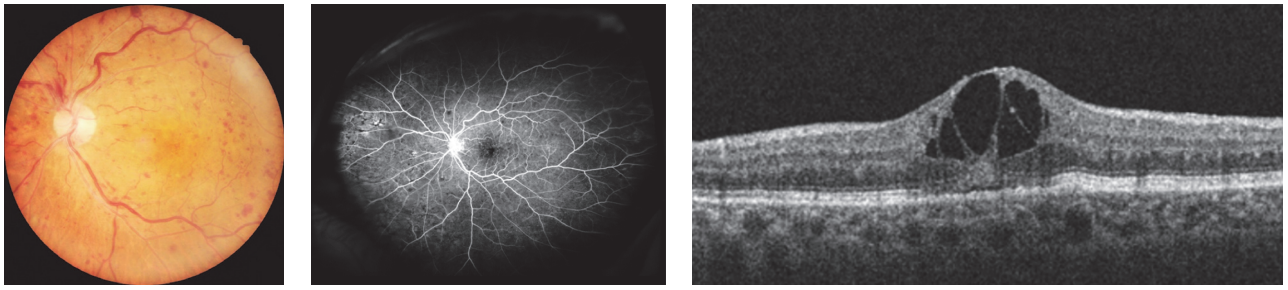
We report the sequential changes of retinal vessels observed by optical coherence tomography angiography (OCTA) in a case of nonischemic central retinal vein occlusion (CRVO) that converted to ischemic CRVO. An 81-year-old woman visited our Retina Clinic because of visual acuity loss in the left eye. Funduscopic examination showed venous tortuosity and intraretinal hemorrhage in all four quadrants of the fundus. OCT showed macular edema. Fluorescein angiography (FA) and OCTA showed loss of small capillaries. Nonischemic CRVO was diagnosed. Antivascular endothelial growth factor (VEGF) treatment resolved the edema and improved visual acuity. However, during follow-up, capillary dropout was observed on OCTA, which gradually enlarged. Eventually, FA confirmed the conversion to ischemic CRVO. In this case, sequential observations using OCTA showed that nonischemic CRVO did not convert to ischemic CRVO abruptly but occurred stepwise. Additionally, vascular changes began around the veins and blood flow changes were observed more clearly in deep capillary plexus than in superficial capillary plexus.

1. Introduction

Central retinal vein occlusion (CRVO) is a significant cause of acquired vision loss [1]. CRVO was traditionally classified into ischemic and nonischemic subtypes on the basis of the degree of retinal capillary nonperfusion. These two different types have very different outcomes. Ischemic CRVO, defined angiographically as showing at least 10-disc area of retinal capillary nonperfusion, has worse vision at initial presentation and at follow-up than nonischemic CRVO [2]. In ischemic CRVO, neovascular glaucoma (NVG) develops in at least 23% of the eyes after 15 months [3]. However, in nonischemic CRVO, development of NVG is rare. It is well known that eyes with nonischemic CRVO may convert to ischemic CRVO during follow-up. The Central Vein Occlusion Study (CVOS) reported a conversion rate from nonischemic CRVO to ischemic CRVO of 3.3% by 4 months after study entry and an incidence rate 10 times higher by 3 years [2]. In nonischemic CRVO, due to potential serious complications, frequent follow-up is needed. Recently, optical coherence tomography angiography (OCTA) has allowed visualization of microvascular abnormalities without dye injection and considered it useful for frequent follow-ups. Furthermore, OCTA has been reported that it is useful to identify eyes with low vascular densities which are at high risk of neovascular complications [4]. We report sequential changes of retinal perivascular capillaries observed using OCTA in a case of nonischemic CRVO that converted to ischemic CRVO. OCTA images were obtained using the RTVue XR Avanti (Optovue, Inc., Fremont, CA, USA) and Cirrus HD-5000 AngioPlex (Ziess, Inc., Oberkochen, Germany).

2. Case Report

An 81-year-old woman was referred to our Retina Clinic because of loss of visual acuity (VA) in the left eye 3 months ago. She had a history of uncontrolled hypertension, hyperlipidemia, and diabetes mellitus. Her best-corrected visual acuity (BCVA) was 20/20 in the right eye and 20/30 in the left eye. Funduscopic examination of the left eye showed typical features of nonischemic CRVO, including venous tortuosity and intraretinal hemorrhage in all four



(a) (b) (c)

Figure 1: Image findings in the left eye at initial visit. (a) Fundus photograph demonstrating venous tortuosity and intraretinal hemorrhage in all four quadrants of the funds. (b) Fluorescein angiography demonstrating an intact parafoveal capillary network. (c) Optical coherence tomography demonstrating inner retinal cysts.

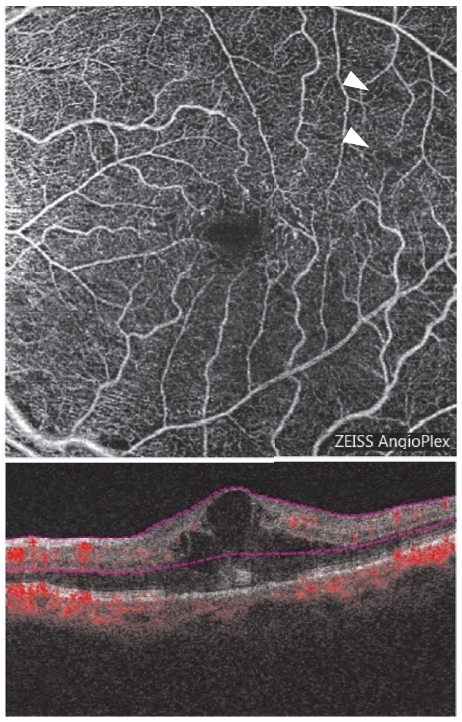
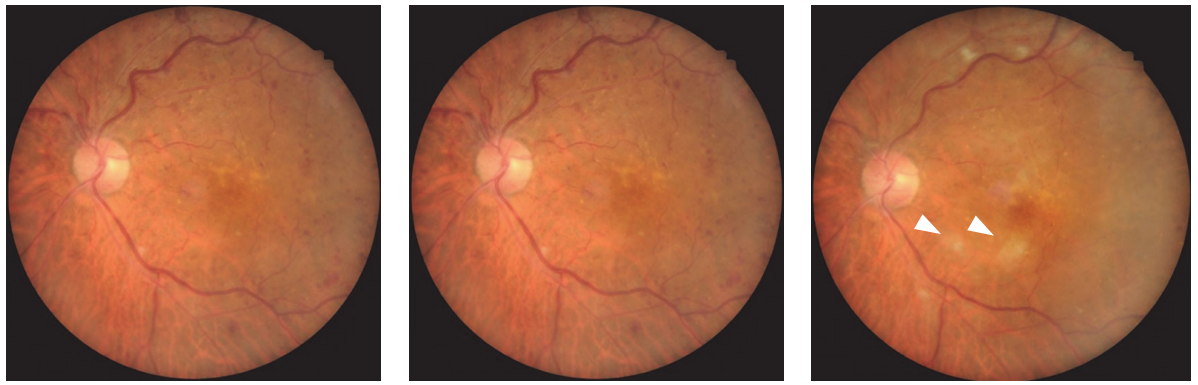


Figure 2: An OCTA image of the superficial capillary plexus in the left eye shows loss of small capillaries (arrowheads) in the upper right region.

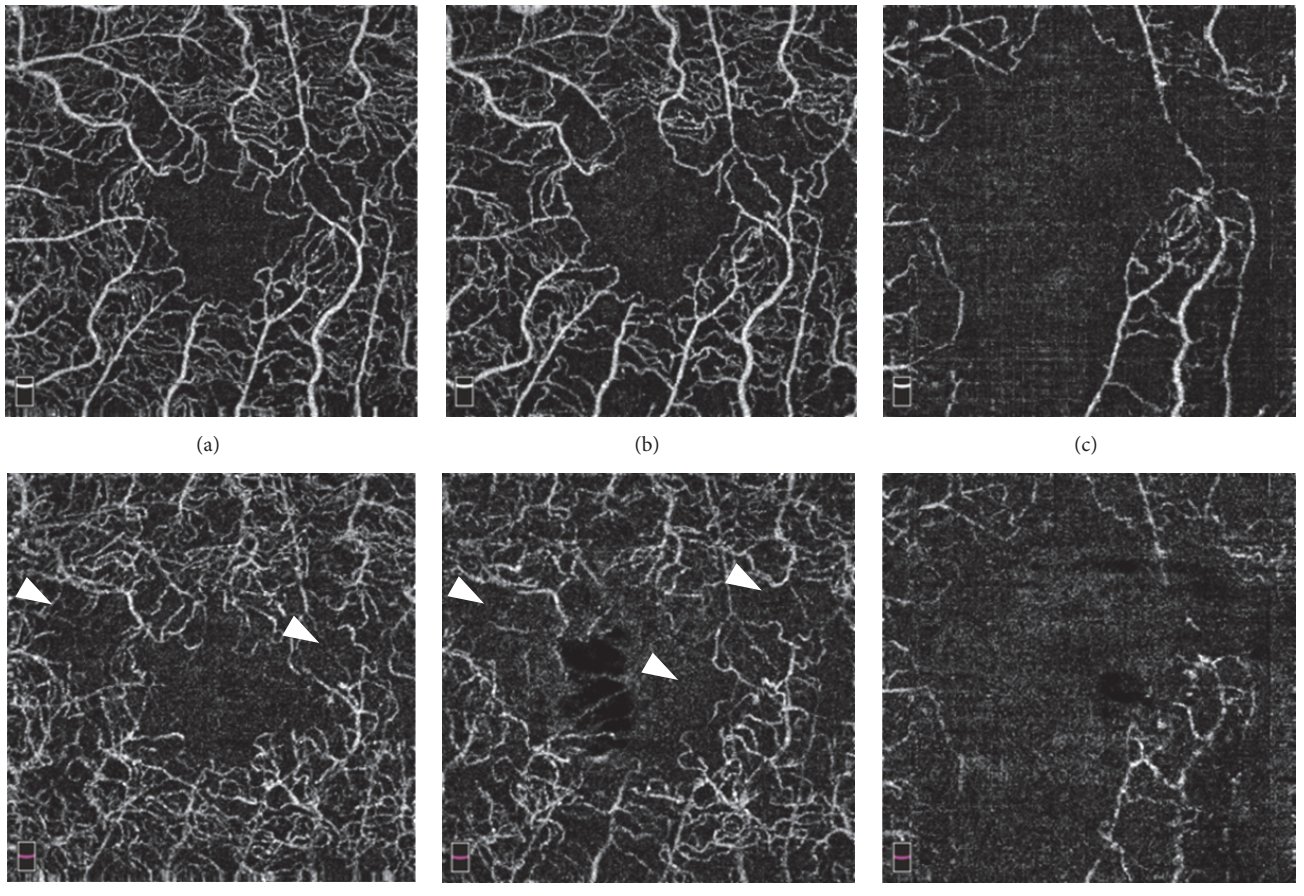
quadrants of the fundus. Fluorescein angiography (FA) and OCTA showed loss of small capillaries but nonperfusion areas were not observed, confirming a diagnosis of nonischemic CRVO. Optical coherence tomography (OCT) showed retinal thickening in the fovea with associated inner retinal cysts. (Figures 1 and 2)

Intravitreal injection of the antivascular endothelial growth factor (VEGF) drug aflibercept was initiated. One month later, visual acuity was improved to 20/25 along with gradual normalization of fundus findings associated with CRVO and macular cysts. After one more month OCTA revealed enlargement of the foveal avascular zone and broken foveal capillary ring in the superficial capillary plexus (SCP) and deep capillary plexus (DCP). Color fundus photograph demonstrated cotton wool spots located near the macula (Figure 3); OCTA revealed gradual enlargement of the avascular zone in SCP and DCP; and visual acuity was impaired. Extensive avascular areas were observed in the DCP earlier than in the SCP (Figure 4). Nine months after onset of symptoms, FA showed more than 10-disc area of retinal capillary nonperfusion (Figure 5). Conversion from nonischemic CRVO to ischemic CRVO was diagnosed. In addition, detailed sequential observations with OCTA revealed progressive capillary dropout from areas around the vein, forming avascular areas. At a specific timeframe during follow-up, the vein surrounded by avascular areas showed loss of patency (Figure 6). There was no neovascularization.



(a) (b) (c)

Figure 3: Fundus photograph images in the left eye (a) 1 month, (b) 2 months, and (c) 4 months after the initial visit. (c) Fundus photograph demonstrating cotton wool spots (arrowheads) located near the macula.



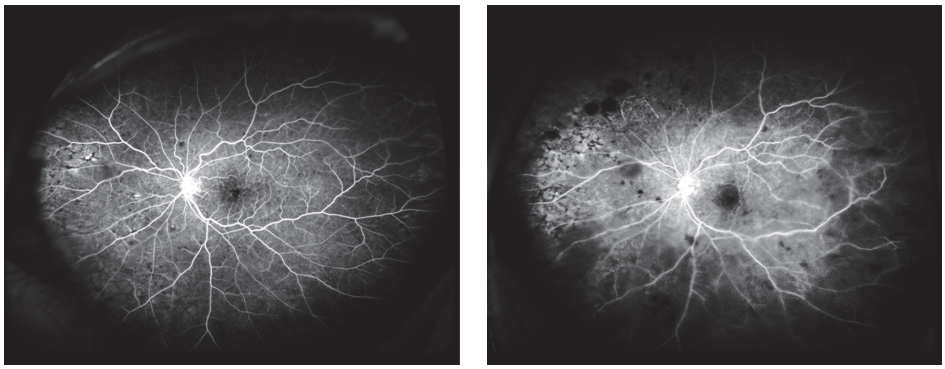
(a) (b) (c)

(d) (e) (f)

Figure 4: OCTA images of the SCP in the left eye (a) 1 month, (b) 2 months, and (c) 4 months after the initial visit. OCTA images of the DCP in the left eye (d) 1 month, (e) 2 months, and (f) 4 months after the initial visit. OCTA images demonstrating expanding nonperfusion area in both SCP and DCP. In DCP, extensive avascular areas are recognized earlier than in SCP (arrowheads).

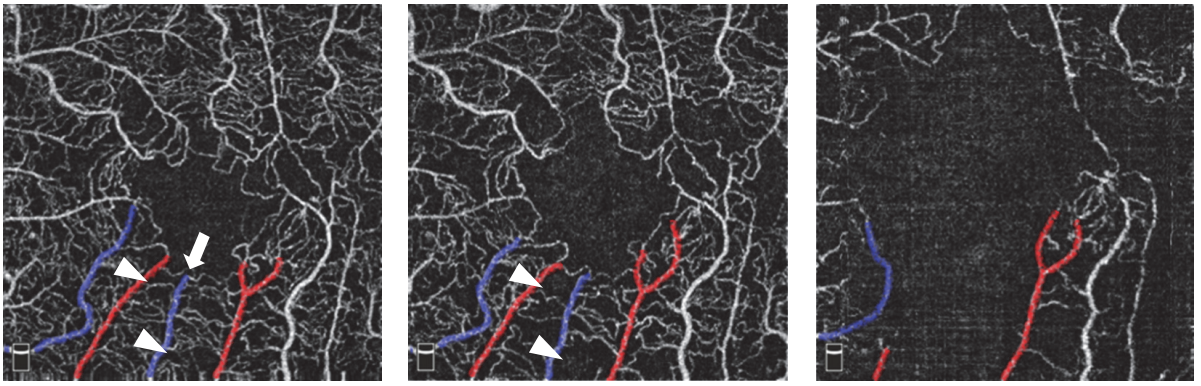
3. Discussion

We performed sequential OCTA observations on a case initially presented with nonischemic CRVO which subsequently converted to ischemic CRVO. OCTA demonstrated capillary dropout occurring preferentially around the vein and these areas gradually enlarged forming avascular lesions. Moreover, OCTA revealed more extensive avascular zones in the DCP than in the SCP. Previous study has also reported that retinal vascular abnormalities in diseases of retinal veins develop more frequently in the DCP than in the SCP [5]. Gradually enlargement of retinal capillary dropout eventually led to the



(a) (b)

Figure 5: Fluorescein angiography image in the left eye at (a) initial visit demonstrating loss of small capillaries, but nonperfusion areas werenot observed. (b) 6 months after the initial visit demonstrating more than 10 discs areas of nonperfusion.



(a) (b) (c)

Figure 6: OCTA images in the left eye (a) 1 month, (b) 2 months, and (c) 4 months after the initial visit. The artery is colored red and the vein is colored blue. Detailed sequential OCTA observations reveal progressive capillary dropout from the areas around the vein (arrow), followed by disappearance of the vein surrounded by avascular areas (arrowheads).

conversion from nonischemic CRVO to ischemic CRVO. The conversion to ischemic CRVO occurred stepwise rather than abruptly.

In conclusion, the present case suggests that conversion to ischemic CRVO starts in the areas around the vein and progresses stepwise rather than abruptly. Moreover, in OCTA study, avascular zones are more readily recognized in the DCP than in the SCP. This is the first report of sequential observations of conversion from nonischemic CRVO to ischemic CRVO using OCTA. Further study of more cases may contribute to elucidation of the mechanism of conversion to ischemic CRVO.

Conflicts of Interest

The authors declare that there are no conflicts of interest regarding the publication of this article.

References

[1] R. L. McIntosh, S. L. Rogers, L. Lim et al., "Natural history of central retinal vein occlusion: an evidence-based systematic review," *Ophthalmology*, vol. 117, no. 6, pp. 1113.e15–1123.e15, 2010.

[2] CVOS group, "Baseline and early natural history report: The Central Vein Occlusion Study," *Archives of Ophthalmology*, vol. 111, pp. 1087–1095, 1993.

[3] H. Zegarra, F. A. Gutman, and J. Conforto, "The Natural Course of Central Retinal Vein Occlusion," *Ophthalmology*, vol. 86, no. 11, pp. 1931–1939, 1979.

[4] D. Senkazi, F. Coscas, A. Sellam et al., "Optical coherence tomography angiography in retinal vein occlusion," *Retina*, pp. 1–9, 2017.

[5] F. Coscas, A. Glacet-Bernard, A. Miere et al., "Optical coherence tomography angiography in retinal vein occlusion: evaluation of superficial and deep capillary plexa," *American Journal of Ophthalmology*, vol. 161, pp. 160–171, 2016.

Hindawi
Case Reports in Ophthalmological Medicine
Volume 2018, Article ID 5474903, 4 pages
https://doi.org/10.1155/2018/5474903

Case Report

Optical Coherence Tomography Angiography of Retinal Arterial Macroaneurysm before and after Treatment

Victoria S. Chang, Stephen G. Schwartz, and Harry W. Flynn Jr.

Department of Ophthalmology, Bascom Palmer Eye Institute, University of Miami Miller School of Medicine, Miami, FL, USA

Correspondence should be addressed to Stephen G. Schwartz; sschwartz2@med.miami.edu

Received 1 November 2017; Accepted 1 February 2018; Published 6 March 2018

Academic Editor: Claudio Campa

A case of retinal arterial macroaneurysm (RAM) is presented with multimodal imaging, including commercially available optical coherence tomography angiography (OCT-A). Following treatment with intravitreal bevacizumab, reduction of flow signal through the RAM is documented. OCT-A provides useful information for the diagnosis and management of at least some patients with RAM, without the need for traditional fluorescein angiography.

1. Introduction

Retinal arterial macroaneurysm (RAM) is an acquired dilatation of a retinal arteriole typically within the first three bifurcations that may occur with varying degrees of hemorrhage, edema, and exudation. Older women are predominantly affected, and there are strong associations with systemic hypertension and arteriosclerotic disease [1]. Although RAMs may involute spontaneously, treatment can be beneficial in the setting of associated macular edema, exudate, or neurosensory retinal detachment [2].

Spectral domain optical coherence tomography (SD-OCT) and fluorescein angiography are widely used in the diagnosis and management of retinal vascular diseases. More recently, OCT angiography (OCT-A), a noninvasive imaging modality that provides structural and functional (blood flow) information from different layers of the retina and choroid, has become available [3].

Using the commercially available Cirrus 5000 with AngioPlex™ (Zeiss, Jena, Germany), the OCT-A findings of RAM in a patient treated with off-label intravitreal bevacizumab (Avastin, Genentech, South San Francisco, CA) are described. A 6 × 6 mm slab was used for all images, and no subsequent image processing was performed.

2. Report of a Case

A 70-year-old Haitian female with a history of chronic hypertension complained of decreased vision in the right eye. Visual acuity (VA) was 20/50. Fundus examination revealed a lesion along the infratemporal vascular arcade, consistent with RAM, and surrounding subretinal hemorrhage and fluid tracking into the macula (Figure 1(a)). SD-OCT showed submacular fluid (Figures 1(b) and 1(c), bottom). The RAM was well delineated on the OCT-A retina slab (Figure 1(b), top) and superficial slab (Figure 1(c), top). The patient was treated with intravitreal bevacizumab #1.

One month later, the patient reported subjective improvement, although the VA remained 20/50. Fundus examination showed decreased blood and subretinal fluid, with lipid exudates in the macula. Fundus photography was not performed at this visit. SD-OCT showed reduced submacular fluid (Figures 2(a) and 2(b), bottom). Lipid exudates appeared as scattered hyperreflective signaling. The OCT-A retina slab (Figure 2(a), top) and superficial slab (Figure 2(b), top) demonstrated diminished signal due to artifact but also reduced flow signal through the RAM. The patient was treated with intravitreal bevacizumab #2.

Two months after presentation, VA improved to 20/30. Fundus examination showed apparent sclerosis of the RAM,

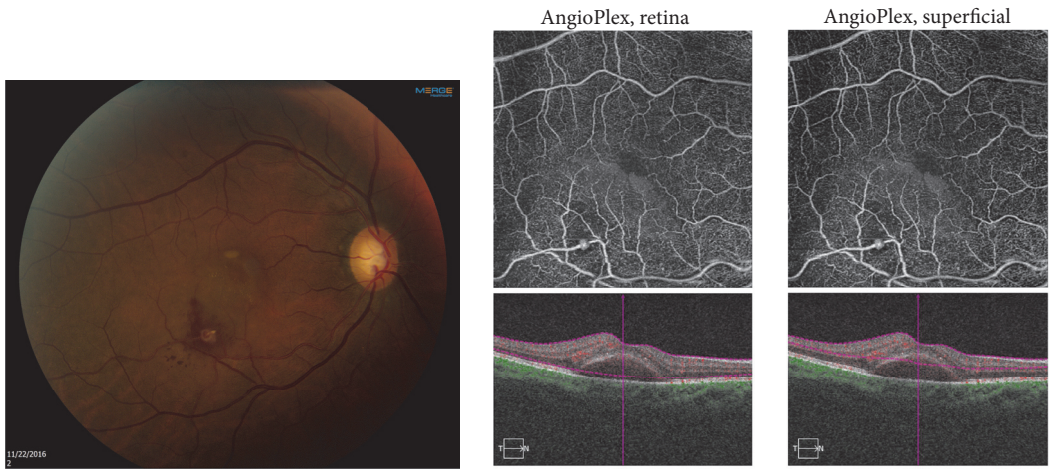


Figure 1: A 70-year-old female presented with decreased vision in the right eye. (a) Fundus photography shows the retinal arterial macroaneurysm (RAM) along the infratemporal vascular arcade, with surrounding subretinal hemorrhage and fluid. (b) TOP: optical coherence tomography angiography (OCT-A) retina slab clearly delineates the RAM. BOTTOM: spectral domain optical coherence tomography (SD-OCT) shows submacular fluid. (c) TOP: OCT-A superficial slab reveals the RAM. Bottom: SD-OCT shows submacular fluid.

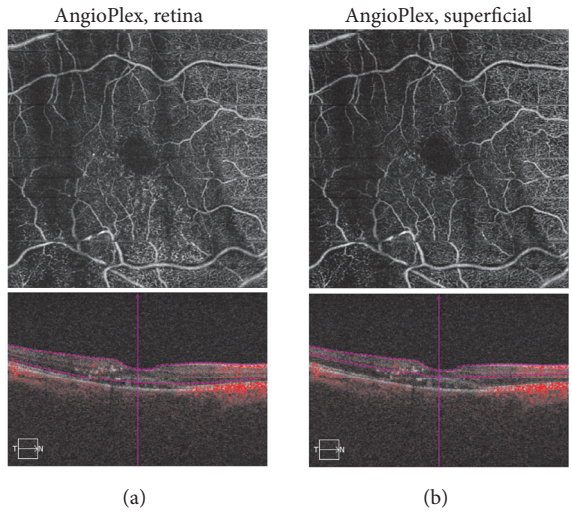


Figure 2: One month following presentation, following intravitreal bevacizumab #1. (a) TOP: optical coherence tomography angiography (OCT-A) retina slab demonstrates reduced signal due to artifact and decreased flow signal through the retinal arterial macroaneurysm (RAM). BOTTOM: spectral domain optical coherence tomography (SD-OCT) reveals reduced submacular fluid. (b) TOP: OCT-A superficial slab shows reduced signal due to artifact and decreased flow signal through the RAM. BOTTOM: SD-OCT reveals decreased submacular fluid.

with persistent lipid exudate in the macula (Figure 3(a)). SD-OCT showed further reduction of submacular fluid (Figures 3(b) and 3(c), bottom). The OCT-A retina slab (Figure 3(b), top) and superficial slab (Figure 3(c), bottom) demonstrated reduced signal flow through the RAM with persistent flow through the normal arteriole. The patient was observed.

Four months after presentation, VA improved to 20/25. Fundus examination showed persistent sclerosis of the RAM, with reduced lipid exudate in the macula (Figure 4(a)). SD-OCT showed restoration of the macular contour (Figures 4(b) and 4(c), bottom). The OCT-A retina slab (Figure 4(b), top) and superficial slab (Figure 4(c), bottom) showed further reduced signal flow through 4 the RAM with persistent flow through the normal arteriole. The patient was observed and was subsequently lost to follow-up.

3. Discussion

Many patients with RAM improve spontaneously, especially if the center of the macula is not involved. For patients with visual loss, treatment options include laser, antivascular endothelial growth factor (anti-VEGF) agents, and combination therapies [4]. Laser for RAM has been reported using

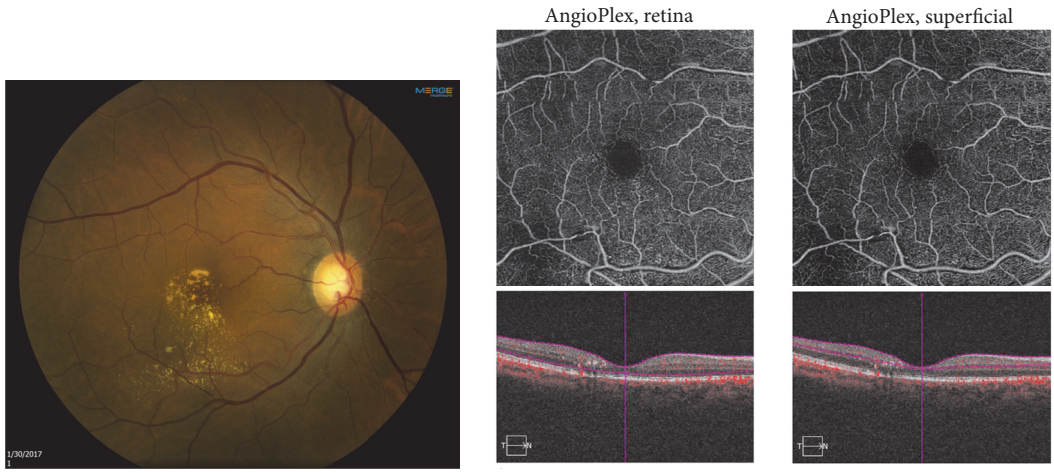


Figure 3: Two months following presentation, following intravitreal bevacizumab #2. (a) Fundus photography shows apparent sclerosis of the retinal arterial macroaneurysm (RAM) with lipid exudate in the macula. (b) TOP: optical coherence tomography angiography (OCT-A) retina slab demonstrates reduced flow signal through the retinal arterial macroaneurysm (RAM). BOTTOM: spectral domain optical coherence tomography (SD-OCT) shows restoration of the foveal contour. (c) TOP: OCT-A superficial slab reveals reduced flow signal through the RAM. BOTTOM: SD-OCT shows decreased submacular fluid.

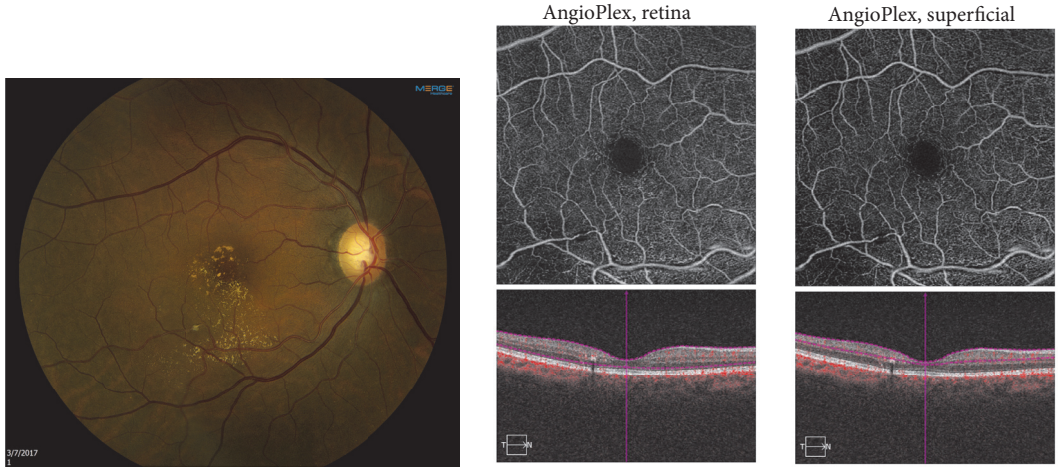


Figure 4: Four months following presentation. (a) Fundus photography demonstrates the sclerosed retinal arterial macroaneurysm (RAM). (b) TOP: optical coherence tomography angiography (OCT-A) retina slab reveals further reduced flow signal through the RAM. BOTTOM: spectral domain optical coherence tomography (SD-OCT) reveals resolution of the submacular fluid. (c) TOP: OCT-A superficial slab shows reduced flow signal through the RAM. BOTTOM: SD-OCT reveals improved macular contour.

yttrium aluminum garnet (YAG), conventional photocoagulation (argon or krypton), and subthreshold treatment using infrared diode [5]. If the RAM causes vitreous hemorrhage, then pars plana vitrectomy techniques may be considered [6].

In the present case, OCT-A demonstrated a focal outpouching of the vessel with a hyperreflective lumen consistent with an active RAM. The lesion became hyporeflective following anti-VEGF treatment, suggesting that it had involuted.

OCT-A can provide clinically useful information in the diagnosis and management of RAM. However, motion artifact and signal attenuation due to hemorrhage may cause degradation of the image quality. Nonetheless, OCT-A is able to produce high resolution images, while avoiding the long acquisition time and invasive nature of fluorescein angiography [7, 8].

In patients with RAM in whom an adequate OCT-A can be obtained, fluorescein angiography may not be necessary. Further experience with OCT-A may clarify the precise role of this technology in diagnosing and managing retinal vascular diseases.

Conflicts of Interest

Dr. Schwartz declares that he has received consulting fees within the last three years from Alimera and Welch Allyn.

All other authors declare that there are no conflicts of interest regarding the publication of this paper.

Acknowledgments

This case report was partially supported by NIH Center Core Grant P30EY014801 and by an Unrestricted Grant from Research to Prevent Blindness to the University of Miami.

References

[1] M. F. Rabb, D. A. Gagliano, and M. P. Teske, "Retinal arterial macroaneurysms," *Survey of Ophthalmology*, vol. 33, no. 2, pp. 73–96, 1988.

[2] F. Pichi, M. Morara, C. Torrazza et al., "Intravitreal bevacizumab for macular complications from retinal arterial macroaneurysms," *American Journal of Ophthalmology*, vol. 155, no. 2, pp. 287–e1, 2013.

[3] T. E. de Carlo, A. Romano, N. K. Waheed, and J. S. Duker, "A review of optical coherence tomography angiography (OCTA)," *International Journal of Retina and Vitreous*, vol. 1, article 5, 2015.

[4] A. Cahuzac, C. Scemama, M. Mauget-Faÿsse, J.-A. Sahel, and B. Wolff, "Retinal arterial macroaneurysms: Clinical, angiographic, and tomographic description and therapeutic management of a series of 14 cases," *European Journal of Ophthalmology*, vol. 26, no. 1, pp. 36–43, 2015.

[5] M. B. Parodi, P. Iacono, L. Pierro, A. Papayannis, S. Kontadakis, and F. M. Bandello, "Subthreshold laser treatment versus threshold laser treatment for symptomatic retinal arterial macroaneurysm," *Investigative Ophthalmology & Visual Science*, vol. 53, no. 4, pp. 1783–1786, 2012.

[6] S. Koinzer, J. Heckmann, J. Tode, and J. Roider, "Long-term, therapy-related visual outcome of 49 cases with retinal arterial macroaneurysm: A case series and literature review," *British Journal of Ophthalmology*, vol. 99, no. 10, pp. 1345–1353, 2015.

[7] A. C. Tan, G. S. Tan, A. K. Denniston et al., "An overview of the clinical applications of optical coherence tomography angiography," *Eye*, 2017.

[8] P. J. Rosenfeld, M. K. Durbin, L. Roisman et al., "ZEISS Angioplex™ Spectral Domain Optical Coherence Tomography Angiography: Technical Aspects," *Developments in Ophthalmology*, vol. 56, pp. 18–29, 2016.

Hindawi
Case Reports in Ophthalmological Medicine
Volume 2018, Article ID 4318354, 4 pages
https://doi.org/10.1155/2018/4318354

Case Report

Optical Coherence Tomography Angiography of Purtscher Retinopathy after Severe Traffic Accident in 16-Year-Old Boy

Hassan Hamoudi,[1,2] **Marie Krogh Nielsen**,[1,2] **and Torben Lykke Sørensen**[1,2]

[1]*Clinical Eye Research Unit, Department of Ophthalmology, Zealand University Hospital, Roskilde, Denmark*
[2]*Faculty of Health and Medical Sciences, University of Copenhagen, Copenhagen, Denmark*

Correspondence should be addressed to Hassan Hamoudi; hassanhaidar10@hotmail.com

Received 4 December 2017; Revised 15 January 2018; Accepted 5 February 2018; Published 1 March 2018

Academic Editor: Stephen G. Schwartz

Purpose. To describe optical coherence tomography (OCT) angiography (OCTA) in a case of Purtscher retinopathy. *Methods*. A 16-year-old male underwent ophthalmological examination including color fundus photography, spectral domain OCT, OCTA, and microperimetry. Examination was performed 10 days, 1 month, and 6 months after the trauma. Diagnosis was based on the characteristic clinical presentation. *Patients*. A single patient case. *Results*. Only the right eye was affected, and all examinations of the left eye were normal. The visual acuity of the right eye was 0.03 (Snellen equivalent) at 10 days and at one month, improving to 0.16 at 6 months. The imaging confirmed the findings of Purtscher retinopathy with ischemic whitening of the retina and retinal hemorrhages and thickened inner retina on OCT. Microperimetry showed reduced sensitivity in the central macula of the right eye. OCTA revealed nonperfusion in both the superficial and the deep retinal capillary plexus of the right eye. *Conclusion*. The OCTA in traumatic Purtscher retinopathy following traffic accident showed nonperfusion in both the superficial and the deep capillary plexus of the retina. OCTA is a valuable noninvasive diagnostic examination in Purtscher retinopathy, and fluorescein angiography became redundant in this case.

1. Introduction

Purtscher retinopathy is an extremely rare condition with an estimated incidence of 0.24 per million per year. It is a condition associated with numerous forms of trauma, including cranial trauma and thoracic compression. It is an occlusive microvasculopathy, and the clinical presentation includes loss of vision of varying severity, occurring hours to days after the trauma. The funduscopic findings include whitening of the retina, multiple cotton wool spots, and bleeding of different sizes. The retinal changes are explained by acute ischemia [1, 2].

In this case report we describe a patient with Purtscher retinopathy, examined with fundus photography, spectral domain optical coherence tomography (OCT) using Heidelberg Spectralis (Heidelberg Engineering, Germany), OCT-angiography (OCTA) (Topcon, Japan), and microperimetry using Microperimeter MP-3 (Nidek Co., Ltd., Japan). The novelty in this paper is the description of Purtscher retinopathy on OCTA.

2. Case Presentation

A 16-year-old boy was involved in a car accident as a front seat passenger. The patient had no previous or current medical history. In the initial phase he was hospitalized in the intensive care unit because of multiple injuries. He suffered from bleeding in the abdominal cavity, pneumothorax, and lesions of the spleen but experienced no direct head trauma or loss of consciousness during the accident. There were no signs of pancreatitis. He underwent abdominal surgery with laparotomy and tube thoracostomy. After a couple of days his general condition was stabilized and he was transferred to the Pediatric Department at our hospital. Ten days after the trauma he complained about blurred vision on the right eye, and was therefore referred to the Department of Ophthalmology.

His presenting corrected visual acuity was 0.03 (Snellen equivalent) on the right eye and 1.0 on the left eye and was unchanged at one-month examination. Six months later, the visual acuity improved to 0.16 on the right eye and

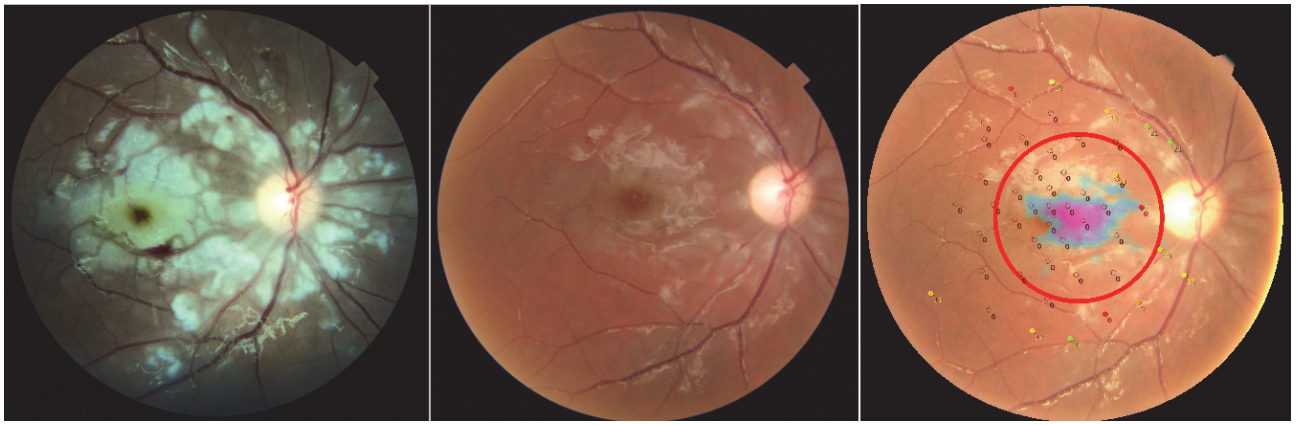


Figure 1

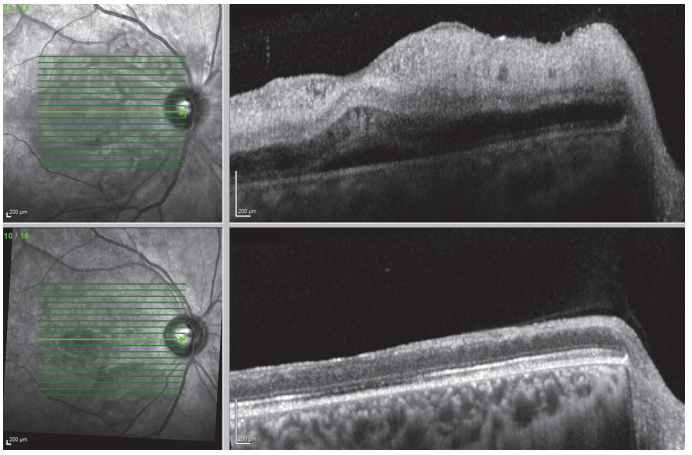


Figure 2

was still normal on the left eye. Anterior segment findings and intraocular pressure were normal. Ophthalmoscopy and fundus photo revealed an ischemic white posterior pole with cotton wool spots and intraretinal hemorrhages mainly in the macula and nasally to the optic disc. The white lesions and bleeding decreased already at the one-month visit (Figure 1), and after 6 months, the white lesions were almost resolved.

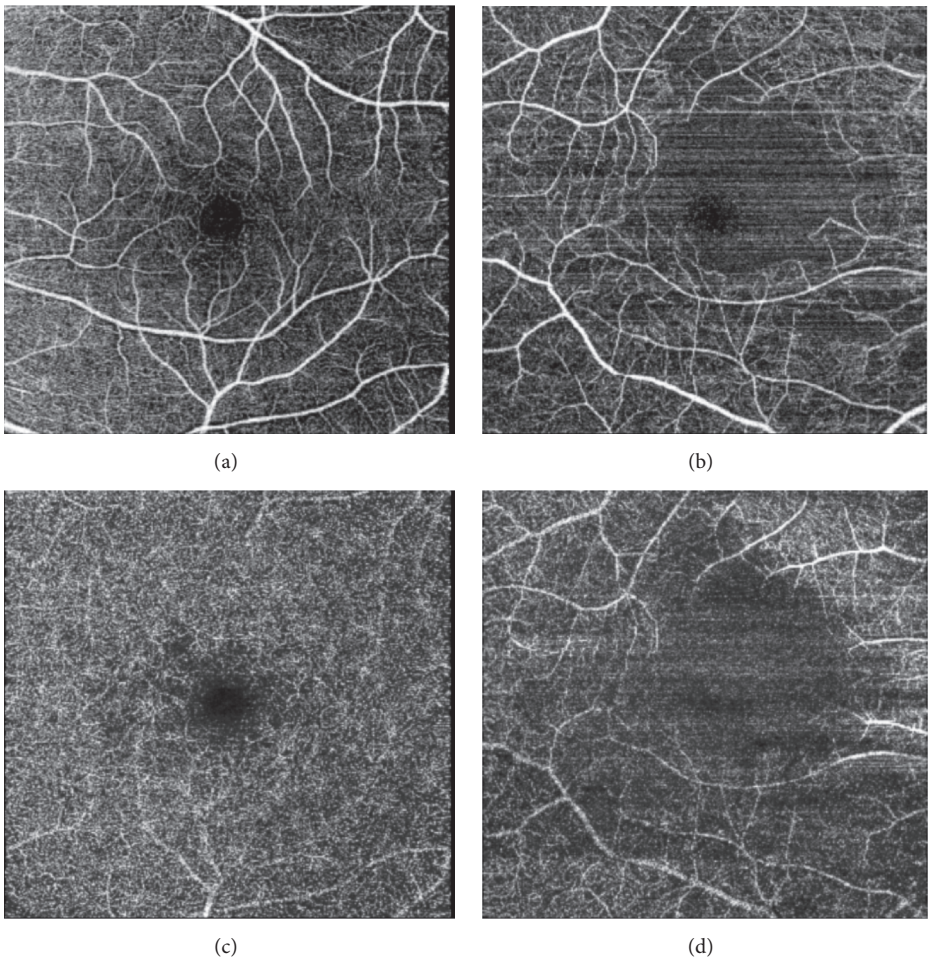
OCT showed hyperreflective and thickened inner retinal layers, a sign of ischemia in the inner retinal circulation. (Figure 2). At follow-up visits the edema decreased significantly on OCT with disruption of the inner retinal layers but also seemingly disrupted ellipsoid zone. In addition, the thickness of the retina was reduced, from 427 microns at onset to 207 microns at 6 months. A manual segmentation of the OCT layers was conducted in order to ensure correct layer identification and thus thickness calculation. Testing of the central visual field by microperimetry showed a central scotoma with decreased sensitivity in the fovea (Figure 1).

OCTA (Figure 3) revealed extensive nonperfusion in the macular area in both the superficial (Figure 3(b)) and the deep capillary plexus of the right eye (Figure 3(d)). OCTA of the left eye was with normal capillary plexus and normal foveal avascular zone (Figures 3(a) and 3(c)).

3. Discussion

Our patient had lower sensitivity in the fovea, which we found to correlate morphologically with the subfoveal atrophy found on OCT. The novelty in this case report is the description of Purtscher retinopathy on OCTA where we found nonperfusion in both the superficial and the deep capillary plexus in the macular region. OCTA allows a fast and noninvasive assessment of the retinal vascular structures and can detect vascular abnormalities without the need of fluorescein angiography [3].

Purtscher retinopathy is a rare condition that was first described by Otmar Purtscher in 1910 with findings of multiple retinal white lesions and superficial retinal hemorrhages. This was in a patient with head trauma, and the condition has since been described in various types of trauma, including seatbelt and airbag pressure, malar bone fracture, and chest trauma [4]. A similar retinal presentation has also been seen in patients without trauma but with a variety of conditions including acute pancreatitis, systemic lupus erythematosus, renal failure, and lymphoproliferative disorders. The condition is called Purtscher-like retinopathy because of the comparable clinical presentation but different etiological association.



(a) (b)

(c) (d)

Figure 3

The diagnosis of Purtscher retinopathy is made on the clinical presentation and the patient's history. The symptoms can be unilateral or bilateral and usually with immediate decrease of visual acuity. The characteristic findings are the Purtscher flecken, which are multiple cotton wool spots of varying sizes. The condition must be differentiated from other ocular disorders that may have common clinical features, for example, commotion retina, Terson's syndrome, shaken baby syndrome, and Valsalva retinopathy [5].

In our case the condition was unilateral, but the onset of the decreased vision is uncertain because of the severe general condition of the patient. The ischemia involving the inner retinal layers during the acute phase which we describe in our patient has previously been reported [6]. It is more difficult to identify the involvement of the outer retina in the acute phase because of the difficulty of its visualization. This is due to the thickened inner retina that appears to be interfering with the signal of the blood flow in the outer retina making the OCTA image not optimal. However, there are reports on photoreceptor disruption with loss of photoreceptor segments in the acute phase, recognized by multifocal electroretinography [7]. In other retinal conditions the finding of an alteration of the interface line between the inner and outer segments is an indication of suffering photoreceptors [8], and this may also be true in Purtscher retinopathy. The involvement of the photoreceptors may explain the visual field abnormalities found in this patient. The microperimetry provides the functional aspect of the morphological changes found on OCT with fundus-controlled testing allowing a precise retinal location [9].

The pathogenesis is still not completely clear, but some hypothesis exist. An increase in the thoracic pressure leads to a reflux in the venous system leading to endothelial damage. This results in incompetence of the microvascular circulation and subsequent occlusion and ischemia [5]. Another hypothesis suggests that the ischemia is the result of an emboli. Both air and fat emboli have been described due to trauma, and the source of the emboli may be thorax [10]. There is no standardized or recommended treatment, and the prognosis varies, some experience a recovery with a visual acuity of 6/12 Snellen or better; however, the prognosis is generally poor and the visual acuity may remain decreased particularly in case of foveal photoreceptor atrophy [5].

In conclusion, OCTA is valuable in the assessment of retinal vascular structures in Purtscher retinopathy and may replace invasive dye-based angiography. It also reveals ischemia at an earlier point.

Conflicts of Interest

None of the authors had any conflicts of interest and the authors have no proprietary interests.

References

[1] A. Giani, A. P. Deiro, P. Sabella, and C. M. Eandi, "Spectral domain-optical coherence tomography and fundus autofluorescence findings in a case of purtscher-like retinopathy," *Retinal Cases and Brief Reports*, vol. 5, no. 2, pp. 167–170, 2011.

[2] S. Nor-Masniwati, Y. Azhany, and E. Zunaina, "Purtscher-like retinopathy following valsalva maneuver effect: Case report," *Journal of Medical Case Reports*, vol. 5, article no. 338, 2011.

[3] V. Kumar and A. Tomar, "Optical coherence tomography angiography imaging of Purtscher retinopathy," *Canadian Journal of Ophthalmology*, vol. 52, no. 6, pp. e201–e203, 2017.

[4] A. Agrawal and M. McKibbin, "Purtscher's retinopathy: epidemiology, clinical features and outcome," *British Journal of Ophthalmology*, vol. 91, no. 11, pp. 1456–1459, 2007.

[5] Y.-C. Lin, C.-M. Yang, and C.-L. Lin, "Hyperbaric oxygen treatment in Purtscher's retinopathy induced by chest injury," *Journal of the Chinese Medical Association*, vol. 69, no. 9, pp. 444–448, 2006.

[6] A. Agrawal and M. A. McKibbin, "Purtscher's and Purtscher-like retinopathies: a review," *Survey of Ophthalmology*, vol. 51, no. 2, pp. 129–136, 2006.

[7] F. Haq, T. S. Vajaranant, J. P. Szlyk, and J. S. Pulido, "Sequential multifocal electroretinogram findings in a case of Purtscher-like retinopathy," *American Journal of Ophthalmology*, vol. 134, no. 1, pp. 125–128, 2002.

[8] F. C. Piccolino, R. R. De La Longrais, G. Ravera et al., "The foveal photoreceptor layer and visual acuity loss in central serous chorioretinopathy," *American Journal of Ophthalmology*, vol. 139, no. 1, pp. 87–99, 2005.

[9] M. Hanout, N. Horan, and D. V. Do, "Introduction to microperimetry and its use in analysis of geographic atrophy in age-related macular degeneration," *Current Opinion in Ophthalmology*, vol. 26, no. 3, pp. 149–156, 2015.

[10] S. A. Buckley and B. James, "Purtscher's retinopathy," *Postgraduate Medical Journal*, vol. 72, no. 849, pp. 409–412, 1996, http://www.ncbi.nlm.nih.gov/pubmed/8935600.

Hindawi
Case Reports in Ophthalmological Medicine
Volume 2018, Article ID 4098419, 5 pages
https://doi.org/10.1155/2018/4098419

Case Report

Evolution of Choroidal Neovascularization due to Presumed Ocular Histoplasmosis Syndrome on Multimodal Imaging including Optical Coherence Tomography Angiography

T. Y. Alvin Liu,[1] Alice Yang Zhang,[1,2] and Adam Wenick[1]

[1]*Retina Division, Wilmer Eye Institute, Johns Hopkins University, Baltimore, MD, USA*
[2]*Department of Ophthalmology, University of North Carolina at Chapel Hill, Chapel Hill, NC, USA*

Correspondence should be addressed to Adam Wenick; awenick1@jhmi.edu

Received 28 December 2017; Accepted 21 January 2018; Published 13 February 2018

Academic Editor: Stephen G. Schwartz

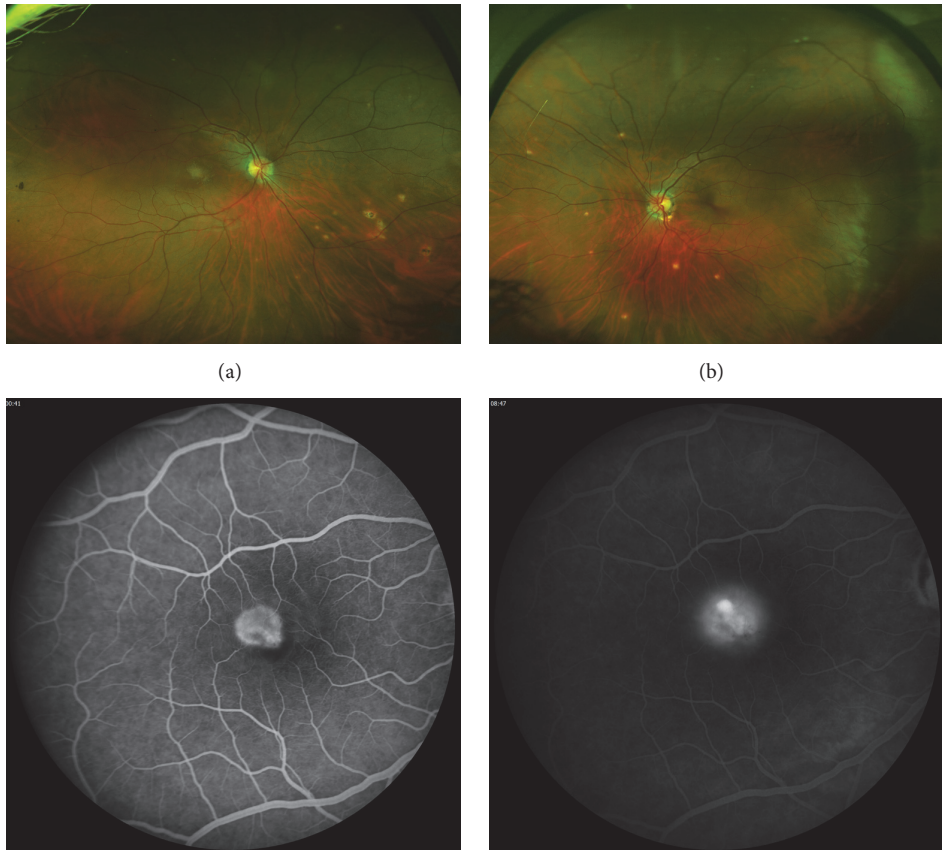
A 37-year-old Caucasian woman presented with acute decrease in central vision in her right eye and was found to have subfoveal choroidal neovascularization (CNV) due to presumed ocular histoplasmosis syndrome (POHS). Her visual acuity improved from 20/70 to 20/20 at her 6-month follow-up, after 3 consecutive monthly intravitreal bevacizumab injections were initiated at her first visit. Although no CNV activity was seen on fluorescein angiography (FA) or spectral-domain optical coherence tomography (SD-OCT) at her 2-month, 4-month, and 6-month follow-up visits, persistent flow in the CNV lesion was detected on optical coherence tomography angiography (OCTA). OCTA shows persistent vascular flow as well as changes in vascular flow in CNV lesions associated with POHS, indicating the continued presence of patent vessels and changes in these CNV lesions, even when traditional imaging of the lesion with OCT and FA indicates stability of the lesion with no disease activity. Additional cases with longitudinal follow-up are needed to assess how OCTA should be incorporated into clinical practice.

1. Introduction

Presumed ocular histoplasmosis syndrome (POHS) can lead to central vision loss with development of choroidal neovascularization (CNV). Herein, we report a case of POHS related CNV that was successfully treated with intravitreal bevacizumab. The evolution of the CNV lesion on multimodal imaging, including optical coherence tomography angiography (OCTA), will be presented.

2. Case Presentation

A 37-year-old Caucasian woman, with no past medical or ocular history, presented with acute-onset and decreased central vision in the right eye. Her left eye was asymptomatic. Visual acuities of the right eye and left eye measured 20/70 and 20/20, respectively, with no improvement with pinhole. The anterior segment was normal with no intraocular inflammation in both eyes. Dilated fundus examination of both eyes showed peripapillary hyper- and hypopigmentation and punched-out chorioretinal lesions in the midperipheral retina consistent with a diagnosis of POHS (Figures 1(a) and 1(b)). Additionally, the right eye showed foveal retinal thickening with trace subretinal hemorrhage and parafoveal intraretinal lipid. Fluorescein angiography (FA) of the right eye showed early hyperfluorescence in the fovea with intense leakage in the late frames, consistent with the presence of CNV (Figures 1(c) and 1(d)). Spectral-domain optical coherence tomography (SD-OCT) of the right eye showed a foveal fibrovascular pigment epithelial detachment (PED) with overlying subretinal hyperreflective material and cystoid macular edema (CME). A diagnosis of choroidal neovascularization (CNV) in the right eye due to presumed ocular histoplasmosis syndrome (POHS) was made. Intravitreal bevacizumab therapy was initiated with close follow-up using serial SD-OCT and OCTA images, shown in Figures 2 and 3, respectively.



(a) (b) (c) (d)

Figure 1: Ultra-wide-field color photographs of the right eye (a) and left eye (b) at presentation showing peripapillary areas of hyper- and hypopigmentation and punched-out chorioretinal lesions in the midperipheral retina, findings classic for presumed ocular histoplasmosis syndrome. 30-degree fluorescein angiography of the right eye showed abnormal early hyperfluorescence in the fovea in the early frame (c) and intense leakage in the late frame (d).

At her 1-month follow-up, the visual acuity of the right eye improved from 20/70 to 20/40, with resolution of subretinal hemorrhage and near resolution of CME. Intravitreal bevacizumab #2 was given.

At her 2-month follow-up, the visual acuity of the right eye improved to 20/32, with no subretinal fluid (SRF) or CME on SD-OCT and resolution of leakage on FA (Figure 4). However, optical coherence tomography angiography (OCTA) showed persistent flow within the CNV lesion. Hence, a decision was made to administer intravitreal bevacizumab #3.

At her 4-month follow-up, the visual acuity of the right eye improved to 20/25, with stable findings on SD-OCT and FA. Although there was persistent flow within the CNV lesion on OCTA, observation was opted given her excellent visual acuity.

At her 6-month follow-up, her right eye visual acuity further improved to 20/20, with no CNV activity seen on SD-OCT or FA. The area of vascular flow on OCTA appeared to have enlarged slightly, but cautious observation was opted for again given her excellent visual acuity.

3. Discussion

Our patient's fundus examination was consistent with classic POHS. Although she did not have a history of spelunking or living in the Mississippi/Ohio river valleys, she endorsed exposure to farm animals. The CNV in her right eye responded well to intravitreal bevacizumab treatment. Her visual acuity improving from Snellen 20/70 to 20/20 over a span of 6 months, with only 3 monthly injections initiated at her presentation. Our experience was consistent with that of published case series, which showed intravitreal antivascular endothelial growth factors (VEGF) injections to be effective for POHS related CNV; on average, patients' visual acuity improved requiring relatively few treatments (range 2.6 to 7 injections per patient per year) [1–4] as compared to neovascular age-related macular degeneration (AMD).

Previously, Wang et al. [5] reported that the peripheral punched-out chorioretinal lesions in POHS corresponded to areas of focal flow loss in the choriocapillaris and deeper choroidal layers on OCTA. To the authors' knowledge, our report is the first reported case on the OCTA evolution

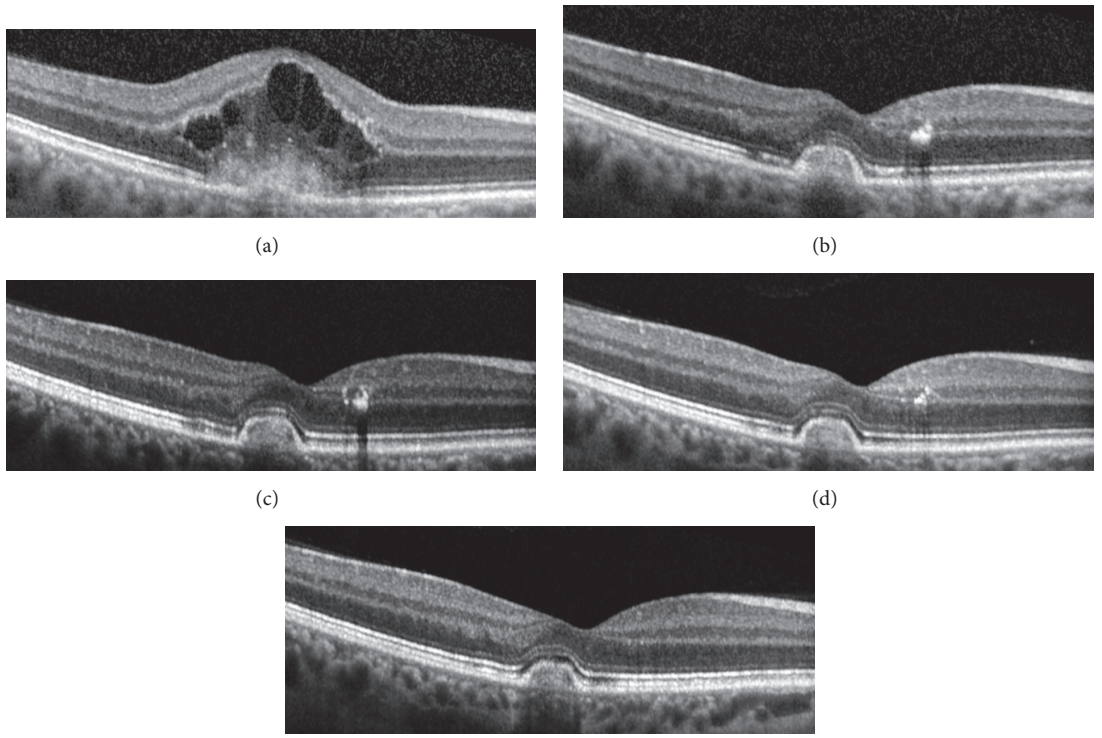


Figure 2: Spectral-domain optical coherence tomography images obtained at the patient's baseline visit (VA 20/70, CST 442, bevacizumab #1 given) (a); 1-month visit (VA 20/40, CST 263, bevacizumab #2 given) (b); 2-month visit (VA 20/32, CST 259, bevacizumab #3 given) (c); 4-month visit (VA 20/25, CST 253) (d); 6-month visit (VA 20/20, CST 257) (e). Between her 2-month and 6-month visits, the appearance of the fibrovascular PED remained stable with gradual decrease in the parafoveal intraretinal lipid. There was no recurrence of SRF or CME or change in PED size between her 4-month and 6-month visits, despite the lack of treatment. VA = visual acuity; CST = central subfield thickness; PED = pigment epithelial detachment; SRF = subretinal fluid; CME = cystoid macular edema.

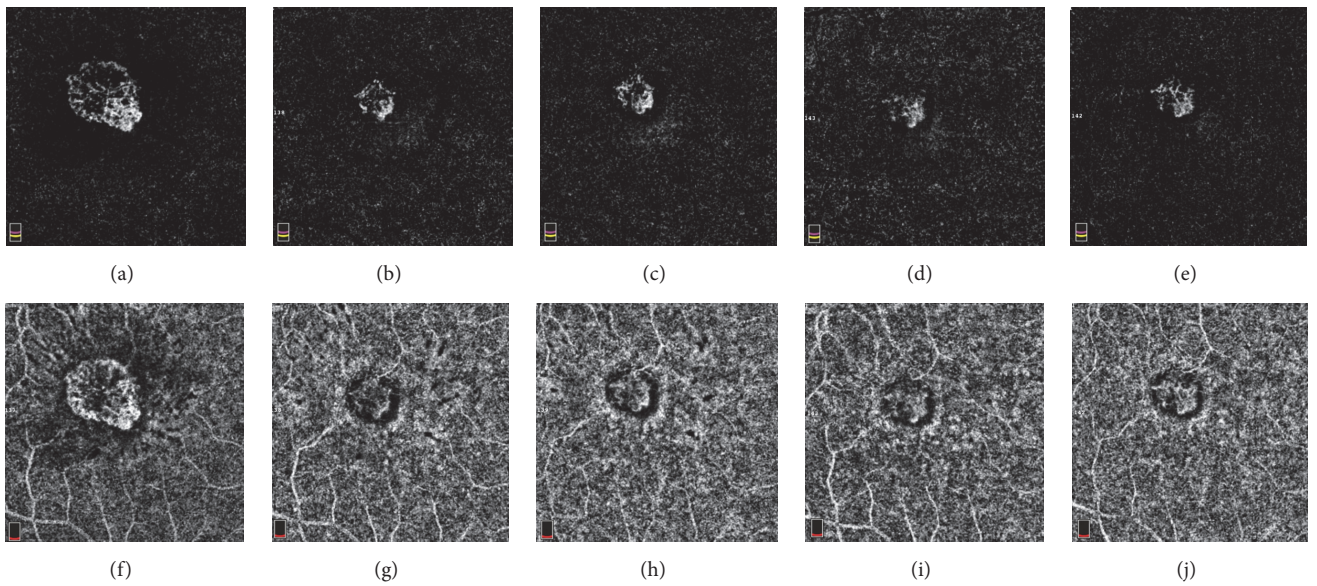
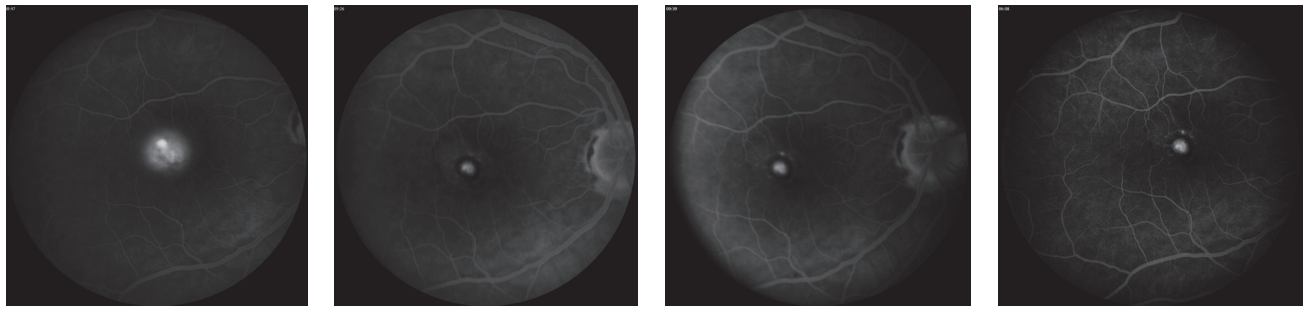


Figure 3: Optical coherence tomography angiography images obtained at the patient's baseline visit (VA 20/70, bevacizumab #1 given) (a, f); 1-month visit (VA 20/40, bevacizumab #2 given) (b, g); 2-month visit (VA 20/32, bevacizumab #3 given) (c, h); 4-month visit (VA 20/25) (d, i); 6-month visit (VA 20/20) (e, j). (a) to (e) represented the outer retinal layer, while (f) to (j) represented the choriocapillaris layer. At her 4-month visit, the superior aspect of the CNV lesion (d) continued to regress following her bevacizumab #3 treatment given 2 months prior, while there was no change in the corresponding FA and OCT images. At her 6-month visit, there was subtle expansion of the superior aspect of the CNV lesion (e), while the corresponding FA and OCT images remained stable.



(a) (b) (c) (d)

Figure 4: Fluorescein angiography (late frame) at the patient's baseline visit (VA 20/70, bevacizumab #1 given) (a); 2-month visit (VA 20/32, bevacizumab #3 given) (b); 4-month visit (VA 20/25) (c); 6-month visit (VA 20/20) (d). After the 2-month visit, there is complete resolution of leakage. VA = visual acuity.

of CNV due to POHS. Using the classification developed by Coscas et al. [6] for neovascular AMD, our patient's CNV lesion will be classified as Pattern I based on the presence of (1) a well-defined lacy-wheel CNV lesion, (2) numerous branching tiny capillaries, (3) peripheral arcade and (4) perilesional hypointense halo at the level of the choriocapillaris.

The serial multimodal images obtained for our patient also provided several interesting observations. First, although no CNV activity was seen on FA or SD-OCT at her follow-up visits, persistent flow of the CNV lesion was detected on OCTA. This suggests that OCTA is more sensitive than FA or SD-OCT in detecting the presence of persistent CNV related to POHS. This observation parallels the work published by de Oliveira Dias et al. [7], in which subclinical macular neovascularization was detected by swept source OCTA in eyes with nonexudative AMD. Second, the CNV lesion in our patient decreased substantially in size between her first visit and 1-month follow-up, after the first bevacizumab treatment. However, the central portion of the CNV lesion remained essentially unchanged between her 1-month and 4-month follow-up visits, despite 2 additional anti-VEGF injections. This is consistent with the observation by Lumbroso et al. [8] that the central trunk of CNV, as compared to the smaller peripheral vessels, can be relatively resistant to anti-VEGF treatments. Third, it was interesting to note that while no recurrence of CNV activity was seen on FA or SD-OCT at our patient's 6-month visit, expansion of the area of flow in the CNV lesion was detected on OCTA. While this was not surprising, given treatment was deferred at the patient's 4-month visit, this suggests that OCTA is more sensitive than FA or OCT in detecting very early increase or recurrence of CNV activity. However, it also shows that an increase in CNV activity on OCTA does not necessarily translate into clinically significant or visually significant changes.

4. Conclusion

To the authors' knowledge, this is the first reported case of OCTA findings in CNV related to POHS. Our case suggests that OCTA could be useful in the management of POHS related CNV, similar to neovascular AMD, in that OCTA seems to be more sensitive than FA and SD-OCT in detecting subtle changes in CNV lesions. What remains unknown is whether the treatment based on subtle OCTA findings, in the absence of fluid on SD-OCT, ultimately translates to better outcomes. Additional cases of CNV related to POHS with longitudinal follow-up imaged with OCTA are needed to assess how OCTA should be incorporated into clinical management decisions.

Conflicts of Interest

The authors declare that there are no conflicts of interest regarding the publication of this article.

References

[1] R. Ehrlich, T. A. Ciulla, R. Maturi et al., "Intravitreal bevacizumab for choroidal neovascularization secondary to presumed ocular histoplasmosis syndrome," *Retina*, vol. 29, no. 10, pp. 1418–1423, 2009.

[2] J. Hu, Q. V. Hoang, F. Y. Chau, M. P. Blair, and J. I. Lim, "Intravitreal anti-vascular endothelial growth factor for choroidal neovascularization in ocular histoplasmosis," *Retinal Cases and Brief Reports*, vol. 8, no. 1, pp. 24–29, 2014.

[3] J. S. Nielsen, T. A. Fick, D. D. Saggau, and C. H. Barnes, "Intravitreal anti-vascular endothelial growth factor therapy for choroidal neovascularization secondary to ocular histoplasmosis syndrome," *Retina*, vol. 32, no. 3, pp. 468–472, 2012.

[4] H. S. Walia, G. K. Shah, and K. J. Blinder, "Treatment of CNV secondary to presumed ocular histoplasmosis with intravitreal aflibercept 2.0 mg injection," *Canadian Journal of Ophthalmology*, vol. 51, no. 2, pp. 91–96, 2016.

[5] J. C. Wang, I. Laíns, L. Sobrin, and J. B. Miller, "Distinguishing white dot syndromes with patterns of choroidal hypoperfusion on optical coherence tomography angiography," *Ophthalmic Surgery, Lasers and Imaging Retina*, vol. 48, no. 8, pp. 638–646, 2017.

[6] G. J. Coscas, M. Lupidi, F. Coscas, C. Cagini, and E. H. Souied, "Optical coherence tomography angiography versus traditional

multimodal imaging in assessing the activity of exudative age-related macular degeneration: a new diagnostic challenge," *Retina*, vol. 35, no. 11, pp. 2219–2228, 2015.

[7] J. R. de Oliveira Dias, Q. Zhang, J. M. B. Garcia et al., "Natural History of Subclinical Neovascularization in Nonexudative Age-Related Macular Degeneration Using Swept-Source OCT Angiography," *Ophthalmology*, 2017.

[8] B. Lumbroso, M. Rispoli, and M. C. Savastano, "Longitudinal optical coherence tomography-angiography study of type 2 naive choroidal neovascularization early response after treatment," *Retina*, vol. 35, no. 11, pp. 2242–2251, 2015.

Hindawi
Case Reports in Ophthalmological Medicine
Volume 2018, Article ID 4342158, 4 pages
https://doi.org/10.1155/2018/4342158

Case Report

Optical Coherence Tomography Angiography of Combined Central Retinal Artery and Vein Occlusion

Shuo-chieh Wu, Victor M. Villegas, and Jaclyn L. Kovach

Department of Ophthalmology, Bascom Palmer Eye Institute, University of Miami Miller School of Medicine, Miami, FL, USA

Correspondence should be addressed to Victor M. Villegas; v.villegas@med.miami.edu

Received 26 August 2017; Accepted 11 January 2018; Published 12 February 2018

Academic Editor: Winfried M. Amoaku

Optical coherence tomography angiography (OCTA) is a new, noninvasive technology that enables detailed evaluation of flow in the retinal and choroidal vasculature. The authors believe this to be the first report to describe the optical coherence tomography angiography findings associated with combined central retinal artery occlusion (CRAO) and central retinal vein occlusion (CRVO).

1. Introduction

Combined central retinal artery occlusion (CRAO) and central retinal vein occlusion (CRVO) is a rare vasoocclusive entity that has been associated with multiple etiologies that can cause devastating vision loss [1–8]. In the population without age-related cardiovascular risk factors, the majority of the combined cases has been attributed to rheological causes, including thrombophilia, vessel wall inflammation, and mechanical compression [3].

Optical coherence tomography angiography (OCTA) is a new, fast, noninvasive imaging modality that allows detection of blood flow through the retinal and choroidal plexuses without intravenous dye injection [9]. The depth-resolved imaging technique affords insight regarding various retinal and choroidal diseases that is not available through other diagnostic modalities, such as fluorescein angiography (FA) [10]. OCTA is rapidly becoming an indispensable tool to describe a spectrum of pathologies, including macular degeneration, diabetic retinopathy, glaucoma, and choroidal neovascularization [10, 11]. Recently, it has been utilized as an adjunct tool to characterize retinal venous or arterial occlusion [10, 12, 13].

The authors believe this report to be the first to describe the optical coherence tomography angiography findings associated with combined central retinal artery occlusion and central retinal vein occlusion. The commercially available Cirrus 5000 with AngioPlex (Zeiss, Jena, Germany) was used, without any subsequent image modification or processing.

2. Case Report

A healthy 69-year-old female presented to the Emergency Department with sudden, painless, visual loss that started immediately following cataract surgery with retrobulbar anesthesia in the left eye (OS) nine days prior to presentation. The patient denied jaw claudication, temporal headache, scalp tenderness, or visual loss in the right eye (OD). Immediately following the event, the patient underwent a work-up which included a transthoracic echocardiogram (TTE), electrocardiogram (EKG), carotid ultrasound, erythrocyte sedimentation rate (ESR)/C-reactive protein (CRP), computed tomography (CT), and magnetic resonance imaging (MRI) of head. All tests were within normal limits.

A complete ophthalmologic exam was performed. Best corrected visual acuity was 20/40 OD and hand motion OS. Intraocular pressure measured by Tono-Pen XL (Reichert Technologies) was 18 mmHg OD and 19 mmHg OS. Full ductions were present without pain. Pupils were equally round with an afferent pupillary defect OS. Anterior segment examination in the right eye was significant for a nuclear sclerotic cataract and examination of the left eye revealed corneal edema, trace cell, +1 flare, and a well-centered intraocular lens.

Fundus examination by indirect ophthalmoscopy was unremarkable OD. Funduscopic exam OS demonstrated mild disc edema, macular edema, whitening of the macula, subtle tortuosity of vessels, and flame-shaped hemorrhages and cotton wool spots in all quadrants (Figure 1).

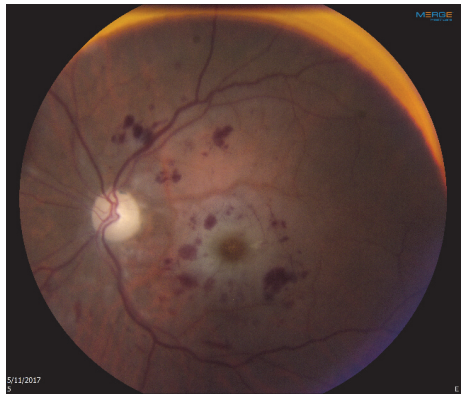


Figure 1: Fundus color photography of the left eye.

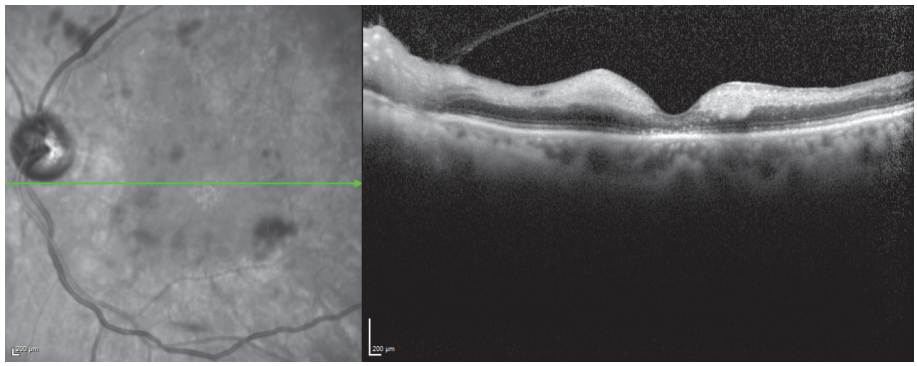


Figure 2: Spectral domain optical coherence tomography of the left eye.

Spectral domain optical coherence tomography (SD-OCT) was performed on OS and showed increased hyperreflectivity and edema of the inner retina with disruption of the ellipsoid zone (EZ) (Figure 2).

OCTA revealed an absence of flow in the foveal and perifoveal area in the superficial and deep retinal capillary plexuses (Figures 3(a) and 3(b)). In contrast, there is minimal alternation in choriocapillaris and choroidal vascular flow (Figures 3(c) and 3(d)).

3. Discussion

A combined CRAO and CRVO is a rare entity, and the etiology is incompletely understood. Although cardiovascular diseases, hypercoagulopathy, and inflammatory diseases are potential risk factors, our patient presented with a combined occlusion without any history of systemic diseases following cataract surgery with retrobulbar anesthesia [1–4]. Several studies have reported the occurrence of a combined CRAO and CRVO following retrobulbar injections, suggesting it can be a severe complication of periocular anesthesia [14–20].

The exact mechanism of combined CRAO and CRVO has not been elucidated, but there are multiple mechanisms proposed to explain the association with retrobulbar injection. Combined occlusion could result from optic nerve sheath hematoma secondary to needle penetration or direct injection into the optic nerve sheath [16, 21]. Another potential mechanism is the compromise of one circulation leading to the occlusion of the other. Brown et al. described two patients who initially presented with a CRVO and then developed a subsequent CRAO, suggesting that increased venous pressure could cross the capillary bed to impede the arterial flow and cause ischemia [18].

Combined CRAO and CRVO is an ophthalmological emergency that should be recognized as a serious postsurgical complication due to its poor outcome. Without timely intervention, combined occlusion can lead to rubeosis iridis, neovascular glaucoma, retinal necrosis, periphlebitis of the central vein, and eventually permanent vision loss [18, 22]. Various treatment modalities have been attempted to reverse the pathology with limited success, including triamcinolone, bevacizumab, and hyperbaric oxygen therapy. However, Vallée et al. demonstrated that timely intervention with fibrinolytics may restore retinal perfusion with visual improvement [2, 4, 22].

In the current report, OCTA showed that the vascular flow of the superficial and deep retinal plexuses were both interrupted OS (Figures 3(a) and 3(b)). In contrast, the choriocapillaris and choroidal vascular flow were minimally affected (Figures 3(c) and 3(d)). These results together suggest

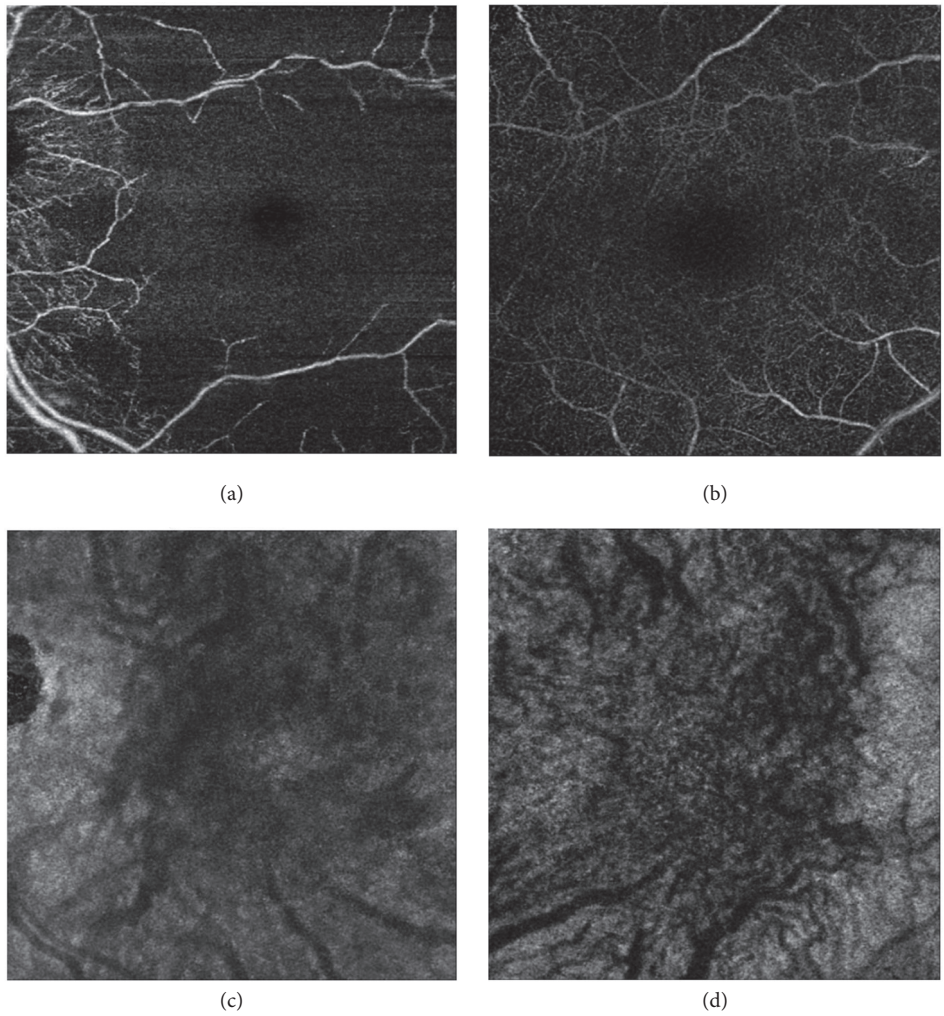


Figure 3: Optical coherence tomography angiography of the left eye. Vascular flow in the (a) superficial retinal capillary plexus, (b) deep retinal capillary plexus, (c) choriocapillaris plexus, and (d) choroidal plexus.

that the occlusion was limited to the retinal circulation without significant involvement of the choroidal circulation. Fluorescein angiography would have allowed us to assess the macular flow impairment. However, OCTA enables visualization of the flow disruption in the superficial and deep retinal capillary plexuses. With the depth of vascular disruption as a new metric for assessing disease severity, OCTA can provide more information regarding visual prognosis for this condition and other retinal vascular diseases.

This case demonstrates clinical features of combined CRAO and CRVO imaged with OCTA following retrobulbar anesthesia associated with cataract surgery. OCTA technology can facilitate diagnosis and extent of combined CRAO and CRVO as it enables discrimination between superficial and deep retinal vasculature. Additional advantages of OCTA compared to FA include faster image acquisition and no potential allergic systemic effects [9, 23, 24].

In conclusion, OCTA is a new, fast, noninvasive imaging technology that has enabled improved understanding of the pathophysiology of many retinal vascular diseases including combined CRAO and CRVO. To the best of our knowledge, this is the first reported case that describes the OCTA findings associated with combined CRAO and CRVO. Future studies with OCTA will hopefully illuminate additional features of combined CRAO and CRVO and provide a better understanding of this complex disease.

Conflicts of Interest

The authors declare that they have no conflicts of interest regarding the publication of this paper.

Acknowledgments

This work is partially supported by NIH Center Core Grant P30EY014801, Research to Prevent Blindness Unrestricted Grant, and Department of Defense.

References

[1] D. Schmidt, "Comorbidities in combined retinal artery and vein occlusions," *European Journal of Medical Research*, vol. 18, no. 1, article no. 27, 2013.

[2] J. A. Lemos, C. Teixeira, R. Carvalho, and T. Fernandes, "Combined Central Retinal Artery and Vein Occlusion Associated with Factor V Leiden Mutation and Treated with Hyperbaric Oxygen," *Case Reports in Ophthalmology*, vol. 6, no. 3, pp. 462–468, 2015.

[3] S. Desai, N. Rai, P. Kulkarni, and S. Natarajan, "Combined crvo with crao in a patient with protein C deficiency," *Retinal Cases and Brief Reports*, vol. 8, no. 2, pp. 145–149, 2014.

[4] S. G. Schwartz, M. Hickey, and C. A. Puliafito, "Bilateral CRAO and CRVO from thrombotic thrombocytopenic purpura: OCT findings and treatment with triamcinolone acetonide and bevacizumab," *Ophthalmic Surgery, Lasers & Imaging Retina*, vol. 37, no. 5, pp. 420–422, 2006.

[5] Ş. Özdek, F. Yülek, G. Gürelik, B. Aydin, and B. Hasanreisoğlu, "Simultaneous central retinal vein and retinal artery branch occlusions in two patients with homocystinaemia [4]," *Eye*, vol. 18, no. 9, pp. 942–945, 2004.

[6] S. M. Parchand, V. S. Vijitha, and D. P. Misra, "Combined central retinal artery and vein occlusion in lupus," *BMJ Case Reports*, vol. 2017, Article ID bcr-2016-218848, 2017.

[7] K. M. Nishiguchi, Y. Ito, and H. Terasaki, "Bilateral central retinal artery occlusion and vein occlusion complicated by severe choroidopathy in systemic lupus erythematosus," *Lupus*, vol. 22, no. 7, pp. 733–735, 2013.

[8] A. V. Rachitskaya, R. K. Lee, S. R. Dubovy, and E. R. Schiff, "Combined central retinal vein and central retinal artery occlusions and neovascular glaucoma associated with interferon treatment," *European Journal of Ophthalmology*, vol. 22, no. 2, pp. 284–287, 2012.

[9] A. Y. Lee, Q. Zhang, D. M. Baughman, R. Mudumbai, R. K. Wang, and C. S. Lee, "Evaluation of bilateral central retinal artery occlusions with optical coherence tomography-based microangiography: a case report," *Journal of Medical Case Reports*, vol. 10, no. 1, pp. 1–6, 2016.

[10] A. H. Kashani, C.-L. Chen, J. K. Gahm et al., "Optical coherence tomography angiography: A comprehensive review of current methods and clinical applications," *Progress in Retinal and Eye Research*, vol. 60, pp. 66–100, 2017.

[11] T. E. de Carlo, A. Romano, N. K. Waheed, and J. S. Duker, "A review of optical coherence tomography angiography (OCTA)," *International Journal of Retina and Vitreous*, vol. 1, article 5, 2015.

[12] A. H. Kashani, S. Y. Lee, A. Moshfeghi, M. K. Durbin, and C. A. Puliafito, "Optical coherence tomography angiography of retinal venous occlusion," *Retina*, vol. 35, no. 11, pp. 2323–2331, 2015.

[13] M. A. Bonini Filho, M. Adhi, T. E. De Carlo et al., "Optical coherence tomography angiography in retinal artery occlusion," *Retina*, vol. 35, no. 11, pp. 2339–2346, 2015.

[14] K. L. Sullivan, G. C. Brown, A. R. Forman, R. C. Sergott, and J. C. Flanagan, "Retrobulbar anesthesia and retinal vascular obstruction," *Ophthalmology*, vol. 90, no. 4, pp. 373–377, 1983.

[15] D. Vasavada, P. Baskaran, and S. Ramakrishnan, "Retinal vascular occlusion secondary to retrobulbar injection: Case report and literature review," *Middle East African Journal of Ophthalmology*, vol. 24, no. 1, pp. 57–60, 2017.

[16] C. M. Morgan, H. Schatz, A. K. Vine et al., "Ocular complications associated with retrobulbar injections," *Ophthalmology*, vol. 95, no. 5, pp. 660–665, 1988.

[17] W. F. Mieler, S. R. Bennett, L. W. Platt, and S. B. Koenig, "Localized retinal detachment with combined central retinal artery and vein occlusion after retrobulbar anesthesia," *Retina*, vol. 10, no. 4, pp. 278–283, 1990.

[18] G. C. Brown, J. S. Duker, R. Lehman, and R. C. Eagle Jr., "Combined central retinal artery-central vein obstruction," *International Ophthalmology*, vol. 17, no. 1, pp. 9–17, 1993.

[19] R. J. De Almeida Torres, A. Luchini, W. Weis, P. R. Frecceiro, and M. Casella, "Combined central retinal vein and artery occlusion after retrobulbar anesthesia - Report of two cases," *Arquivos Brasileiros de Oftalmologia*, vol. 68, no. 2, pp. 257–261, 2005.

[20] G. Giuffre, M. Vadala, and L. Manfre, "Retrobulbar anesthesia complicated by combined central retinal vein and artery occlusion and massive vitreoretinal fibrosis," *Retina*, vol. 15, no. 5, pp. 439–441, 1995.

[21] R. Klein, B. E. Klein, S. E. Moss, and S. M. Meuer, "The epidemiology of retinal vein occlusion: the Beaver Dam Eye Study," *American Ophthalmological Society*, vol. 98, pp. 141–143, 2000.

[22] J.-N. Vallée, M. Paques, A. Aymard et al., "Combined central retinal arterial and venous obstruction: Emergency ophthalmic arterial fibrinolysis," *Radiology*, vol. 223, no. 2, pp. 351–359, 2002.

[23] A. S. L. Kwan, C. Barry, I. L. McAllister, and I. Constable, "Fluorescein angiography and adverse drug reactions revisited: the Lions Eye experience," *Clinical & Experimental Ophthalmology*, vol. 34, no. 1, pp. 33–38, 2006.

[24] B. J. Jennings and D. E. Mathews, "Adverse reactions during retinal fluorescein angiography," *Journal of the American Optometric Association*, vol. 65, pp. 465-71, Jul 1994.

Hindawi
Case Reports in Ophthalmological Medicine
Volume 2017, Article ID 9032576, 4 pages
https://doi.org/10.1155/2017/9032576

Case Report

Cilioretinal Artery Occlusion Combined with Central Retinal Vein Occlusion: What Is the Best Imaging Modality for the Follow-Up?

Mahmut Kaya, Taylan Ozturk, Ziya Ayhan, Nilufer Kocak, and Suleyman Kaynak

Department of Ophthalmology, Dokuz Eylul University School of Medicine, Izmir, Turkey

Correspondence should be addressed to Mahmut Kaya; mahmutkaya78@yahoo.com

Received 27 July 2017; Revised 16 October 2017; Accepted 2 November 2017; Published 21 December 2017

Academic Editor: Stephen G. Schwartz

We report retinal structural changes of a 37-year-old man diagnosed with the concomitant occlusion of cilioretinal artery and central retinal vein. Comprehensive ophthalmological evaluation was performed, followed by spectral-domain optical coherence tomography (SD-OCT, Heidelberg), optical coherence tomography angiography (OCT angiography, Optovue Inc., Fremont, California, USA), fluorescein angiography, and color fundus photography. The use of OCT angiography and en face SD-OCT imaging as an adjunct test to map out correlative paracentral scotomas during follow-up allowed us to evaluate cilioretinal artery occlusion in the best way due to obtaining satisfactory images of the normal retinal vascular networks and areas of nonperfusion and congestion at various retinal levels.

1. Introduction

The cilioretinal artery occlusion is very rare and accounts for 5% of retinal artery occlusions [1]. Optical coherence tomography angiography (OCT angiography) is a new imaging technology that allows for fast, noninvasive assessment of microvascular perfusion across the macular region, offering the potential to perform quantitative assessment. OCT angiography has the capability to segment each layer of the retinal microvasculature in normal and pathological eyes without dye injection [2]. Herein, we aimed to characterize the appearance of the peri- and parafoveal macular microvasculature in a visually asymptomatic young patient with the cilioretinal artery occlusion in chronic phase using OCT-A and to compare different imaging modalities.

2. Case Report

A 37-year-old man without any history of ocular and systemic pathology presented with painless visual decrease in his right eye for 10 days. He had suffered from multiple episodes of amaurosis fugax for 30 days. Clinical examination, spectral-domain OCT (SD-OCT), OCT angiography (XR Avanti, software version 2015.1.1.98, Optovue Inc., Fremont, California, USA), fluorescein angiography, and color fundus photography, as well as systemic and laboratory assessments, were used to document findings in the patient with cilioretinal artery occlusion combined with central retinal vein occlusion. The patient presented in this report has given informed consent for this publication.

At the first visit, his best-corrected visual acuity (BCVA) was 20/60 in the right eye (RE) and 20/20 in the left eye (LE). Fundus examination of the RE demonstrated a whitening of the retina along the distribution of the cilioretinal artery. The retinal veins were mildly dilated and tortuous and accompanied by adjacent retinal haemorrhages (Figure 1(A)). Fluorescein angiography demonstrated delayed filling and emptying of this artery (Figure 1(B)). A SD-OCT examination revealed retinal hyperreflectivity and thickening with loss of distinction of retinal layers (Figure 1(C)). Based on these findings, the patient was ultimately diagnosed with a combined cilioretinal artery and central retinal vein occlusion. His blood laboratory findings, systemic physical examination, electrocardiogram, carotid ultrasound imaging, and chest

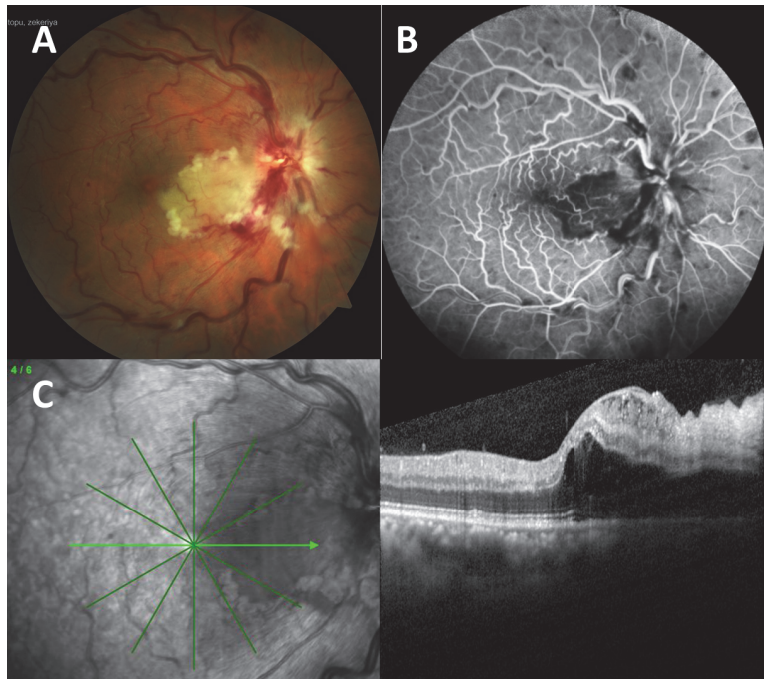


Figure 1: Color fundus photographs of the right eye (A) at the time of presentation. On fluorescein angiography (B), nonperfusion of the cilioretinal artery was shown in the area of cilioretinal at the initial presentation. Optical coherence tomography (OCT) at baseline (C) showed the presence of intraretinal and subretinal fluid and hyperreflective inner retinal layers.

X-ray were all unremarkable. Echocardiography revealed normal findings. Cryoglobulin, lupus anticoagulant, and anti-cardiolipin antibodies were all negative. Antithrombin III, protein C, and protein S activities were normal. Dexamethasone intravitreal implant (Ozurdex, Allergan Inc., Irvine, California, USA), which is the first-line treatment option according to health insurance policy in Turkey, was injected into the right vitreous cavity to at least alleviate the central retinal vein occlusion-related concomitant optic disc and macular edema. The patient was examined monthly over a 6-month period.

Upon follow-up, BCVA had increased to 20/20, the haemorrhages were absorbed, and the dilatation and tortuosity of retinal vessels had resolved. In OCT angiography, of the affected RE, there was attenuation of both the superficial capillary plexus and deep capillary plexus (Figure 2). The circulation of the retinal vessels had improved on FA (Figure 3(A)). The SD-OCT density map clearly delineates the areas of atrophy corresponding to the distribution of the sclerotic arterioles (Figure 3(B)). Spectral-domain OCT scan demonstrated diffuse thinning of the inner nuclear layer, corresponding to the central zone of the cilioretinal artery occlusion (Figure 3(C)). Color fundus photography was unremarkable.

3. Discussion

The superficial retinal capillary plexus is located in the nerve fiber layer near the disc, but is present more predominantly within the ganglion cell layer in the central macular region [3–5]. The deep retinal capillary plexus is composed of an intermediate and deep plexus located at the inner and outer planes of the inner nuclear layer [3–5]. These capillary layers are interconnected by perpendicularly oriented vessels [6] and may be disproportionately affected by ischemic retinal vascular disease. Although retinal vasculature changes in retinal artery occlusions are widely described in the literature, precise assessment and analysis of the deep retinal capillary plexus with FA is limited mostly by light scattering from the inner retinal layers. At the follow-up, not all imaging modalities are useful for retinal artery occlusion, so, for retina specialists, it is important to be well informed in order to make the best choice from all of the various available imaging modalities [4, 5].

In this case in the chronic phase, on OCT angiography, we demonstrated nonperfusion of both the superficial and deep capillary plexus levels in areas with persistent ischemia. In chronic cilioretinal artery occlusion, regression of retinal edema is usually followed by development of retinal thinning and subsequent disorganization of the inner retinal architecture. En face OCT showed delineating areas of atrophy corresponding to distribution of cilioretinal arterial occlusion. However, FA did not show measurable filling delay and the capillary network seemed normally perfused.

When evaluating the acute phase of the isolated retinal artery occlusion, FA technique is an important diagnostic tool to reveal the affected arterial vasculature, with

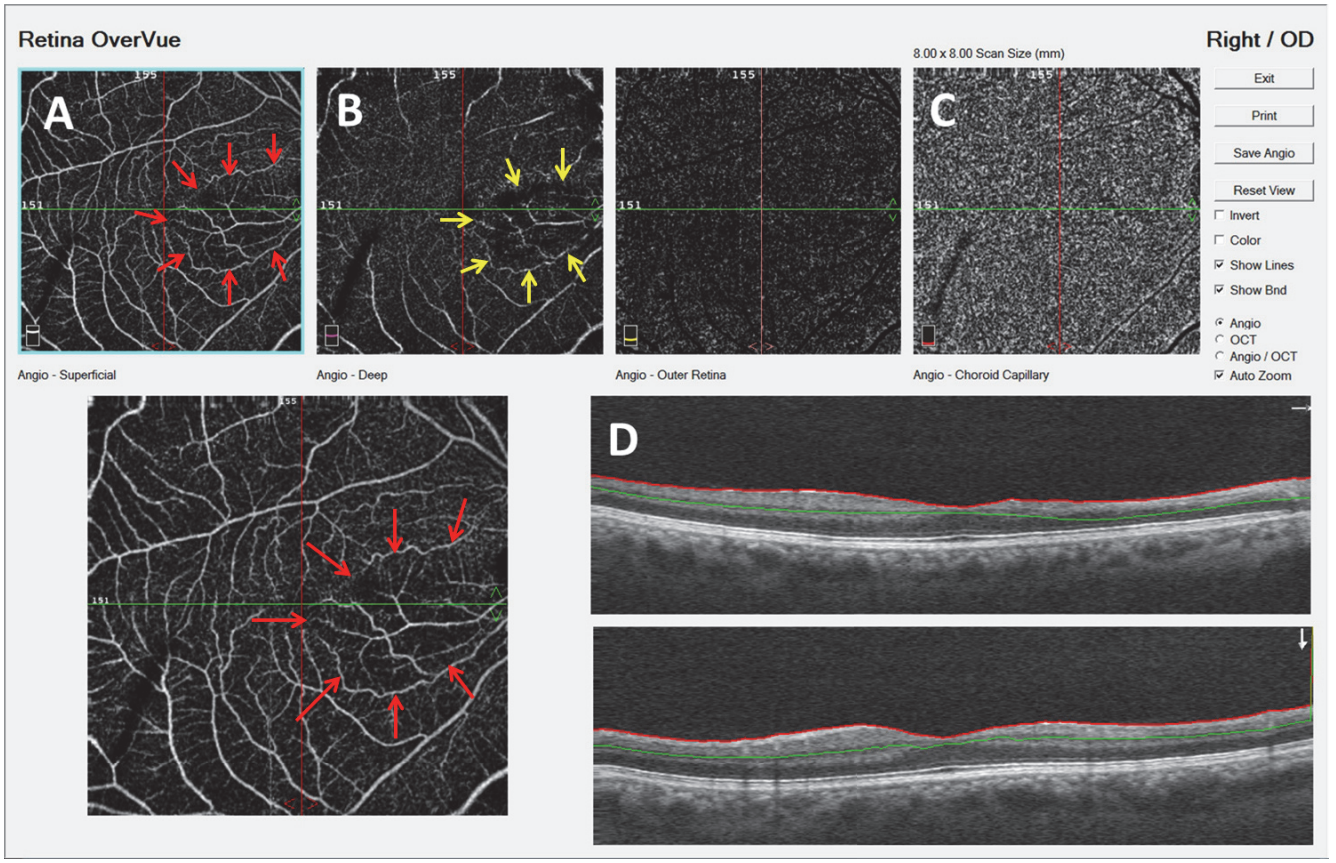


Figure 2: OCT angiography was performed 10 months later. Wedge-shaped area of capillary nonperfusion was revealed in both the superficial (A) and deep (B) retinal capillary plexus in area supplied by the cilioretinal artery and the choriocapillaris (C) was not affected.

typically normal choroidal filling. However, it is limited to imaging only the superficial vascular plexus. Fluorescein angiogram provides information regarding the localization and extent of vascular disease. In chronic phase, FA did not consistently reveal any correlation to these lesions.

In the acute phase, OCT images demonstrate the increased reflectivity and thickness of the inner retina and a corresponding decrease of reflectivity in the outer layer of the retina and retinal pigment epithelium/choriocapillaris layer. Follow-up OCT images demonstrate a decrease in the reflectivity and thickness of the inner retinal layers and a corresponding increase of reflectivity in the outer retina and retinal pigment epithelium/choriocapillaris layer compared with the baseline OCT image, suggesting a generalized atrophy of the neurosensory retina as a late finding [3]. Therefore, the use of OCT may help facilitate prompt recognition of acute and chronic cilioretinal artery occlusion. En face OCTA may prove useful to quantify and further localize the foci of retinal ischemia in retinal artery occlusive disorders as demonstrated by SD-OCT [5].

OCT angiography represents a relatively new technology with the ability to not only noninvasively image the superficial capillary plexus traditionally seen on fluorescein angiography but also capture the flow of the deeper capillary plexuses. In the chronic phase, there were pruning and dropout of the deeper plexuses on OCT angiography matching the middle retinal atrophy [2, 5]. When compared to the current standard of FA, OCT angiography is fast and noninvasive and can provide improved visualization of the microvasculature.

The use of OCT angiography and en face SD-OCT imaging as an adjunct test to map out correlative paracentral scotomas during follow-up allowed us to evaluate cilioretinal artery occlusion in the best way due to obtaining satisfactory images of the normal retinal vascular networks and areas of nonperfusion and congestion at various retinal levels. Furthermore, OCT angiography might help to assess accurately the extent of macular ischemia and vascular flow changes during the course of retinal vascular occlusions.

Conflicts of Interest

None of the authors have any financial/conflicting interests to disclose.

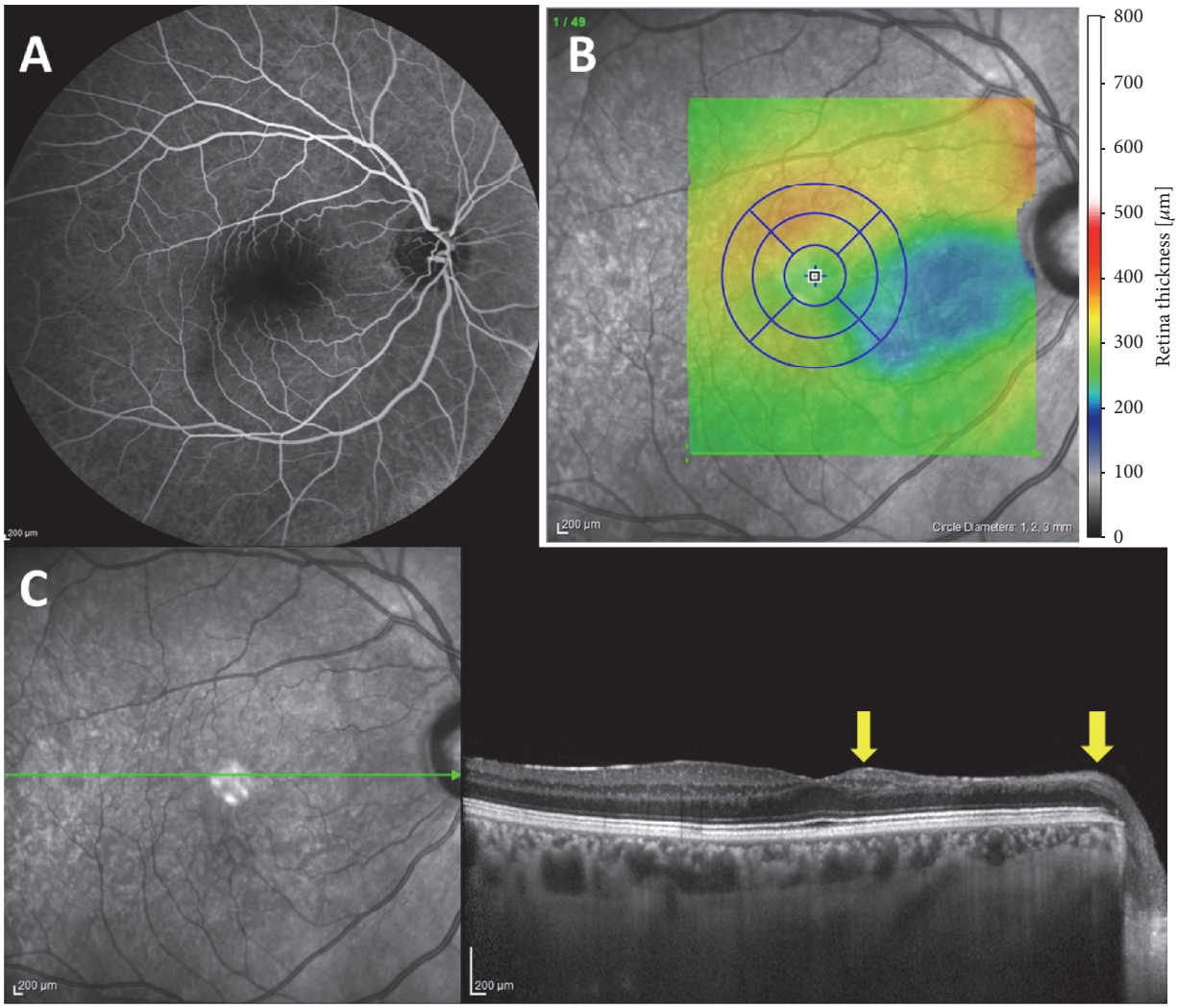


Figure 3: At follow-up, early phase of FA showing quite normal filling of the cilioretinal artery sparing (A). The retinal capillary network is well perfused on this magnification of the posterior pole. Spectral-domain OCT density map delineating areas of atrophy (blue) corresponding to cilioretinal arteriole occlusions (B). Spectral-domain OCT delineating areas of atrophy (arrows) corresponding to distribution of cilioretinal artery occlusion (C).

References

[1] G. C. Brown, K. Moffat, A. Cruess, L. E. Magargal, and R. E. Goldberg, "Cilioretinal artery obstruction," *Retina*, vol. 3, no. 3, pp. 182–187, 1983.

[2] Y. Jia, O. Tan, J. Tokayer et al., "Split-spectrum amplitude-decorrelation angiography with optical coherence tomography," *Optics Express*, vol. 20, no. 4, pp. 4710–4725, 2012.

[3] S. Yu, F. Wang, C. E. Pang, L. A. Yannuzzi, and K. B. Freund, "Multimodal imaging findings in retinal deep capillary ischemia," *Retina*, vol. 34, no. 4, pp. 636–646, 2014.

[4] D. M. Foreman, S. Bagley, J. Moore, G. W. Ireland, D. McLeod, and M. E. Boulton, "Three dimensional analysis of the retinal vasculature using immunofluorescent staining and confocal laser scanning microscopy," *British Journal of Ophthalmology*, vol. 80, no. 3, pp. 246–251, 1996.

[5] S. Yu, C. E. Pang, Y. Gong et al., "The spectrum of superficial and deep capillary ischemia in retinal artery occlusion," *American Journal of Ophthalmology*, vol. 159, no. 1, pp. 53–63.e2, 2015.

[6] D. M. Snodderly, R. S. Weinbaus, and J. C. Choi, "Neural-vascular relationships in central retina of macaque monkeys (*Macaca fascicularis*)," *The Journal of Neuroscience*, vol. 12, no. 4, pp. 1169–1193, 1992.

Hindawi
Case Reports in Ophthalmological Medicine
Volume 2017, Article ID 5151972, 4 pages
https://doi.org/10.1155/2017/5151972

Case Report

Multimodal Images of Acute Central Retinal Artery Occlusion

Parth Shah, Stephen G. Schwartz, and Harry W. Flynn Jr.

Department of Ophthalmology, Bascom Palmer Eye Institute, University of Miami Miller School of Medicine, Miami, FL, USA

Correspondence should be addressed to Stephen G. Schwartz; sschwartz2@med.miami.edu

Received 29 July 2017; Revised 19 September 2017; Accepted 31 October 2017; Published 16 November 2017

Academic Editor: Pradeep Venkatesh

Two illustrative cases of acute central retinal artery occlusion (CRAO) are presented with multimodal imaging, including fluorescein angiography (FA) and commercially available optical coherence tomography angiography (OCT-A). In both patients, retinal ischemia was imaged well using both FA and OCT-A, and the two imaging studies provided comparable pictures. OCT-A provides useful information for the diagnosis and management of patients with acute CRAO, without the need for dye injection.

1. Introduction

Central retinal artery occlusion (CRAO) results from obstruction of blood flow due to embolic, thrombotic, inflammatory, or traumatic causes. In some eyes with CRAO, visual loss is relatively less severe due to sparing of the cilioretinal artery [1].

In most patients, the diagnosis of CRAO may be made with ophthalmoscopy alone, although ancillary testing is frequently used to confirm the diagnosis and to document the findings at presentation. Multimodal imaging includes fluorescein angiography (FA), spectral domain optical coherence tomography (SD-OCT), and optical coherence tomography angiography (OCT-A). Although FA has been used traditionally to evaluate the retinal circulation, OCT-A is an emerging technology that provides clinically useful information.

The present manuscript uses OCT-A to identify the pathologic features in two illustrative cases of CRAO. In both patients, OCT-A was performed using the commercially available Cirrus 5000 with AngioPlex (Zeiss, Jena, Germany) with no subsequent image processing. A 6 × 6 mm slab was used for all images.

2. Cases

2.1. Case 1. A 77-year-old male with a history of atrial fibrillation and nonneovascular age-related macular degeneration (AMD) presented about 4 hours following acute visual loss in the left eye. Best corrected visual acuity (BCVA) was count fingers. Fundus examination revealed macular drusen as well as mild macular whitening and an early cherry red spot (Figure 1(a)). SD-OCT demonstrated thickening and hyperreflectivity of the inner retinal layers (Figure 1(b)). FA at 26.58 seconds revealed delayed retinal perfusion (Figure 1(c)). The OCT-A retina slab (Figure 1(d)), superficial slab (Figure 1(e)), and deep slab (Figure 1(f)) revealed absent flow very similar to the FA.

2.2. Case 2. An 81-year-old male with a history of hypertension presented about 13 hours following acute visual loss in the left eye. BCVA was 20/50. Fundus examination revealed macular whitening in a pattern consistent with CRAO with cilioretinal artery sparing (Figure 2(a)). SD-OCT demonstrated thickening and hyperreflectivity of the inner retinal layers temporal to the center of the macula (Figure 2(b)). FA at 18.67 seconds revealed delayed retinal perfusion consistent with the pattern of macular whitening (Figure 2(c)). The OCT-A retina slab (6 × 6 mm) revealed absent flow in the same distribution as the FA (Figure 2(d)). There was diminished signal superiorly on the OCT-A due to artifact.

3. Discussion

OCT-A characteristics of CRAO have been reported previously [2–5]. These two cases illustrate the benefits of

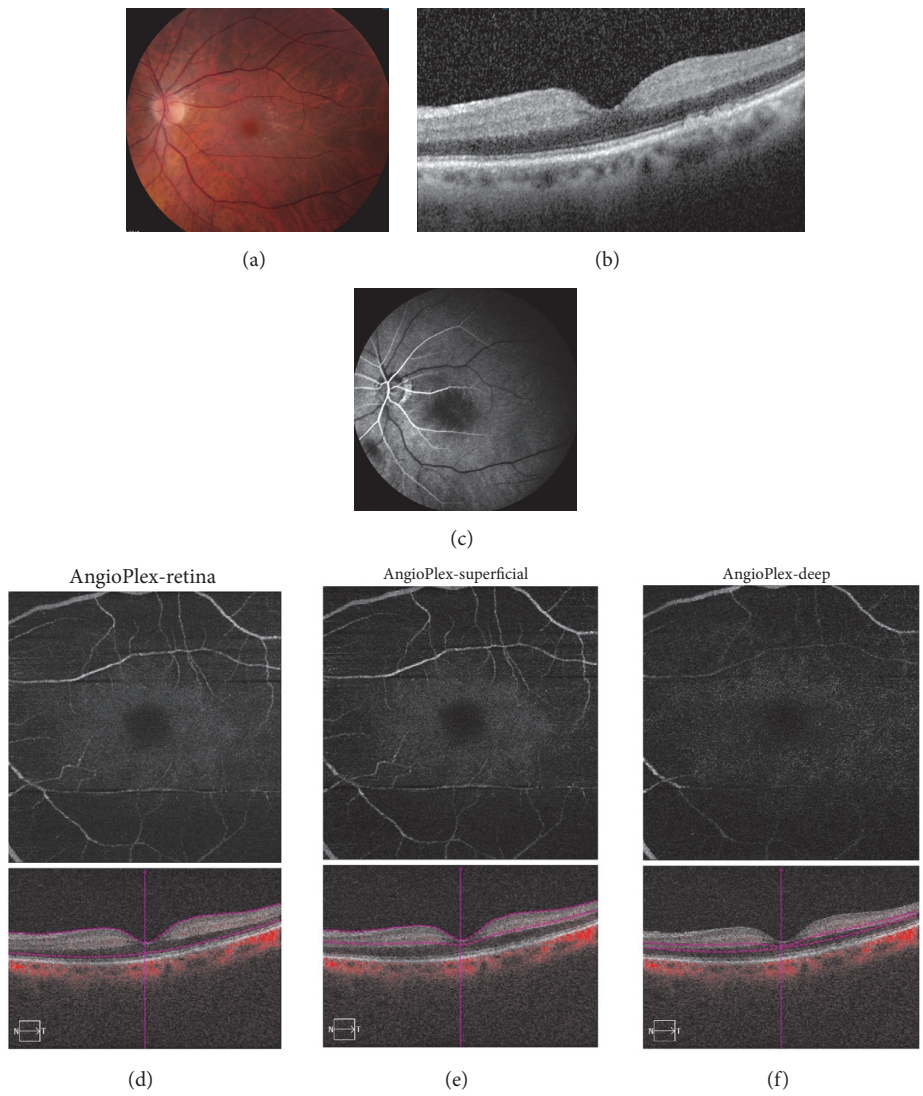


Figure 1: Acute central retinal artery occlusion, left eye. (a) Fundus photography reveals macular drusen and mild macular whitening with an early cherry red spot. (b) Spectral domain optical coherence tomography (SD-OCT) reveals thickening and hyperreflectivity of the inner retinal layers. (c) Fluorescein angiography (FA) at 26.58 seconds reveals delayed retinal perfusion. (d) Optical coherence tomography angiography (OCT-A) 6 × 6 mm retina slab reveals absent flow similar to that seen on FA. (e) OCT-A 6 × 6 mm superficial slab reveals absent flow. (f) OCT-A 6 × 6 mm deep slab reveals absent flow.

OCT-A in providing clinically useful information in the management of patients with acute CRAO without the need for fluorescein injection. In both patients, there is substantial concordance between the findings of the FA and the OCT-A performed on the same day. A similar concordance has been reported between FA and OCT-A for patients with chronic branch retinal vein occlusion [6].

OCT-A offers several advantages compared with traditional FA. OCT-A is noninvasive and has no risks of allergy [7]. In most patients, OCT-A can be obtained faster than FA. However, OCT-A is expensive and the image quality is affected by the patient's ability to fixate. In patients with poor vision, such as those with acute CRAO, it may not be possible to obtain good OCT-A images.

In patients with acute CRAO in whom an adequate OCT-A can be obtained, FA may not be necessary. Since many of these patients have serious systemic vascular diseases, OCT-A is an easily performed, quick, noninvasive alternative to FA.

Conflicts of Interest

Dr. Schwartz declares that he has received consulting fees within the last three years from Alimera Sciences, Bausch + Lomb, and Welch Allyn. All other authors declare that there are no conflicts of interest regarding the publication of this paper.

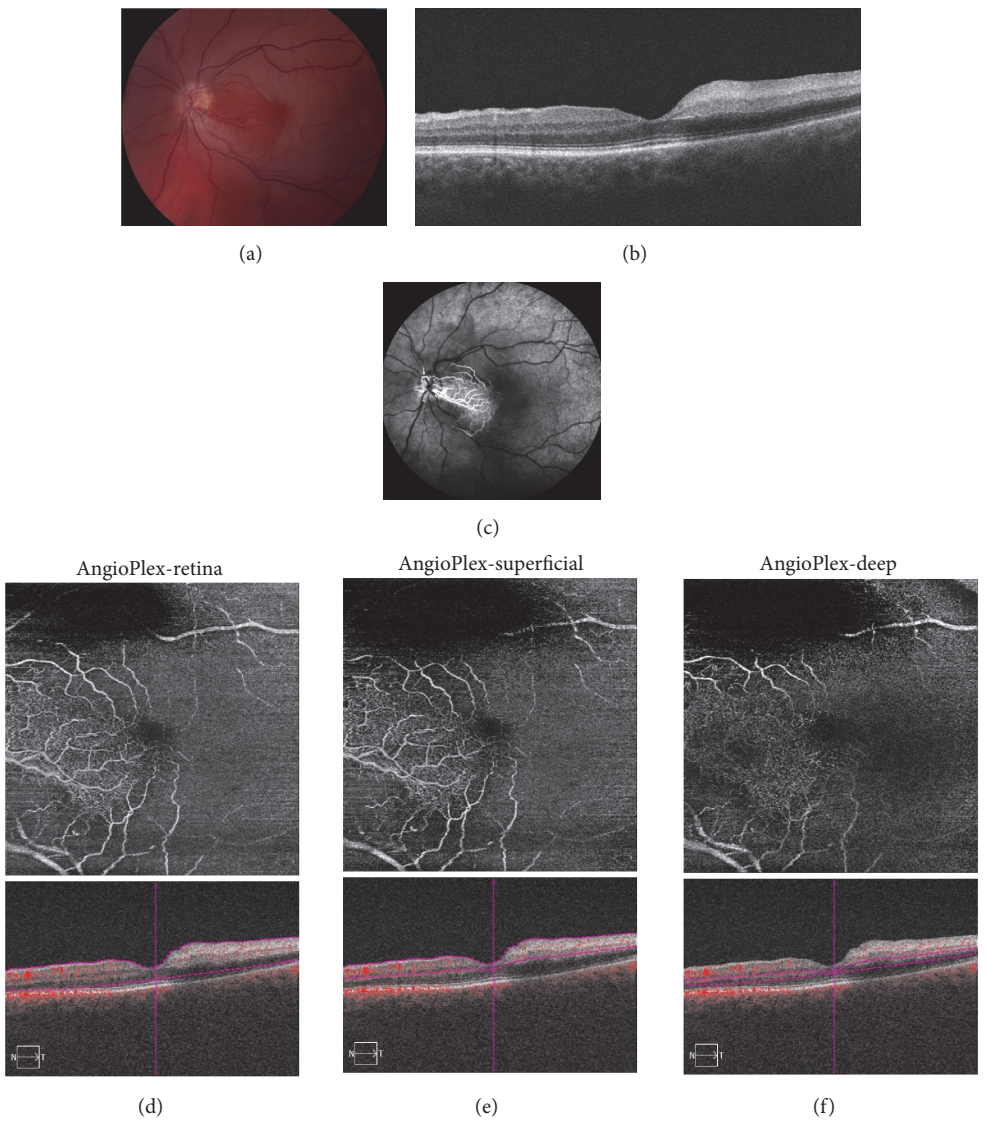


Figure 2: Acute central retinal artery occlusion with cilioretinal sparing, left eye. (a) Fundus photography reveals macular whitening with cilioretinal artery sparing. (b) Spectral domain optical coherence tomography (SD-OCT) reveals macular thickening and hyperreflectivity of the inner retinal layers temporal to the center of the macula, consistent with cilioretinal artery sparing. (c) Fluorescein angiography (FA) at 18.67 seconds reveals delayed retinal perfusion with cilioretinal artery sparing. (d) Optical coherence tomography angiography (OCT-A) 6 × 6 mm retina slab reveals absent flow similar to that seen on FA. There is diminished signal superiorly due to artifact. (e) OCT-A 6 × 6 mm superficial slab reveals absent flow. There is diminished signal superiorly due to artifact. (f) OCT-A 6 × 6 mm deep slab reveals absent flow. There is diminished signal superiorly due to artifact.

Acknowledgments

This work is partially supported by NIH Center Core Grant P30EY014801 and an Unrestricted Grant from Research to Prevent Blindness.

References

[1] D. D. Varma, S. Cugati, A. W. Lee, and C. S. Chen, "A review of central retinal artery occlusion: clinical presentation and management," *Eye*, vol. 27, no. 6, pp. 688–697, 2013.

[2] E. Philippakis, B. Dupas, P. Bonnin, R. Hage, A. Gaudric, and R. Tadayoni, "Optical coherence tomography angiography shows deep capillary plexus hypoperfusion in incomplete central retinal artery occlusion," *Retinal Cases and Brief Reports*, vol. 9, no. 4, pp. 333–338, 2015.

[3] G. Damento, M. H. Chen, and T. Leng, "Spectral-domain optical coherence tomography angiography of central retinal artery occlusion," *Ophthalmic Surgery, Lasers and Imaging Retina*, vol. 47, no. 5, pp. 467–470, 2016.

[4] C. K. Hwang, A. M. Kolomeyer, and A. J. Brucker, "Optical Coherence Tomography Angiography of a Central Retinal Artery Occlusion Before and After Anterior Chamber Paracentesis," *Ophthalmology*, vol. 124, no. 5, p. 608, 2017.

[5] E. A. Novais, L. Roisman, P. R. C. De Oliveira et al., "Optical coherence tomography angiography of chorioretinal diseases,"

Ophthalmic Surgery, Lasers and Imaging Retina, vol. 47, no. 9, pp. 848–861, 2016.

[6] S. G. Schwartz, A. Monroig, and H. W. Flynn Jr., "Multimodal images of chronic branch retinal vein occlusion," *International Medical Case Reports Journal*, vol. Volume 10, pp. 159–162, 2017.

[7] P. J. Rosenfeld, M. K. Durbin, L. Roisman et al., "ZEISS Angioplex™ Spectral Domain Optical Coherence Tomography Angiography: Technical Aspects," *Developments in Ophthalmology*, vol. 56, pp. 18–29, 2016.

Hindawi
Case Reports in Ophthalmological Medicine
Volume 2017, Article ID 8186134, 4 pages
https://doi.org/10.1155/2017/8186134

Case Report

Optical Coherence Tomography Angiography of Macular Telangiectasia Type 2 with Associated Subretinal Neovascular Membrane

Victor M. Villegas and Jaclyn L. Kovach

Department of Ophthalmology, Bascom Palmer Eye Institute, University of Miami Miller School of Medicine, Miami, FL, USA

Correspondence should be addressed to Victor M. Villegas; v.villegas@med.miami.edu

Received 27 July 2017; Accepted 22 October 2017; Published 9 November 2017

Academic Editor: Hsin-Yi Chen

Optical coherence tomography angiography (OCTA) is a recently established noninvasive technology for evaluation of the retinal and choroidal vasculature. The literature regarding the findings in macular telangiectasia type 2 (MacTel2) is scarce. We report the OCTA findings associated with a subject with MacTel2 and secondary subretinal neovascularization (SNV). The commercially available Cirrus 5000 with AngioPlex (Zeiss, Jena, Germany) was used, without any subsequent image modification or processing. Subretinal neovascularization was detectable with OCTA at the level of the outer retina and choriocapillaris. Microvascular abnormalities associated with MacTel2 were present mostly in the deep capillary plexus of the retina temporally.

1. Introduction

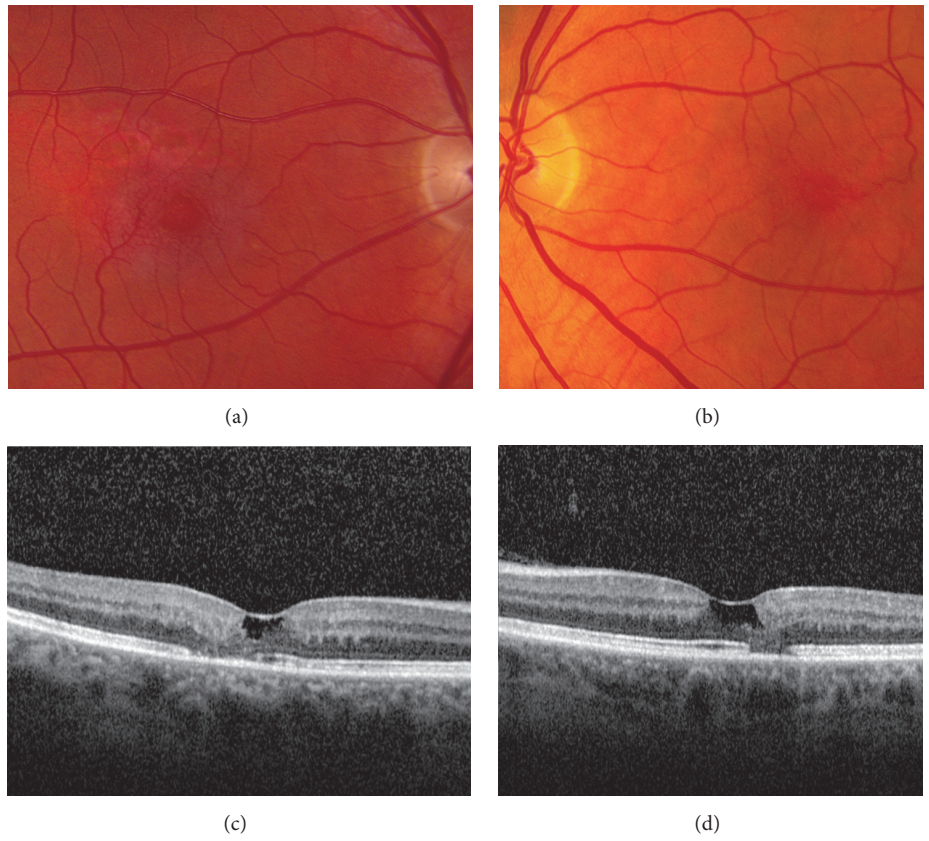
Macular telangiectasia type 2 (MacTel2) is a bilateral retinovascular disease that is typically acquired during middle age and may lead to visual loss [1–3]. The hallmark of the disease is retinal vascular ectasia and neural atrophy of the macula [4]. This condition may pose a diagnostic challenge when evaluated with indirect ophthalmoscopy due to the subtle foveal findings. Initially, the only finding on fundus examination could be a decrease in retinal transparency temporal to the fovea. In the early nonproliferative phase of this condition, other clues to diagnosis can be an increase in central foveal autofluorescence related to a reduction in macular pigment. Fluorescein angiography typically demonstrates leakage from abnormal retinal vessels temporal to the fovea. Spectral domain optical coherence tomography (SDOCT) has offered great insight into the pathogenesis of this condition and shows atrophic abnormalities throughout the retinal layers in the foveal including an inner lamellar cyst with internal limiting membrane drape and disruption of the ellipsoid zone. These changes are thought to be due to degeneration of Müller cells and photoreceptors [5, 6]. Secondary subretinal neovascularization (SNV) can arise in the proliferative phase and form connections with choroidal vessels [7]. The presence of these vessels can be challenging to identify on FA in the setting of temporal foveal leakage.

Optical coherence tomography angiography (OCTA) is a new, fast, noninvasive imaging modality that allows detection of blood flow through the retinal and choroidal plexuses without intravenous dye injection [8]. Various retinal and choroidal diseases have been described using OCTA, including macular telangiectasia [9–20]. Currently, the literature regarding OCTA characteristics of MacTel2 patients with SRNV is limited.

The purpose of this report is to discuss the OCTA features of nonproliferative and proliferative MacTel2 imaged with OCTA. The commercially available Cirrus 5000 with AngioPlex (Zeiss, Jena, Germany) was used, without any subsequent image modification or processing.

2. Case Report

A 63-year-old male with history of well-controlled type 2 diabetes mellitus, essential hypertension, and heart disease was referred to our clinic due to visual loss in the right eye (OD). Prior ophthalmic history included radial keratotomy



(a) (b) (c) (d)

Figure 1: Macular telangiectasia type 2 initial presentation. (a) Fundus photograph of the right macula shows foveal pigment mottling and changes in the perifoveal vasculature. (b) Fundus photography of the left macula shows foveal pigment mottling. (c) Spectral domain optical coherence tomography (SDOCT) reveals an atrophic cyst and ellipsoid zone disruption in the right macula. (d) Similar SDOCT changes are present in the left macula.

and LASIK in both eyes (OU). He also had uneventful cataract surgery with intraocular lens implantation OD and YAG capsulotomy. Best-corrected visual acuity was 20/50 OD and 20/20 in the left eye (OS). Manifest refraction was −1.00 + 1.50 × 52 OD and +1.50 + 1.75 × 42 OS. Intraocular pressure was 17 mmHg in both eyes (OU). Pupils were equally round and equally reactive to light. Anterior segment examination was remarkable for radial keratotomy with pseudophakia OD and nuclear sclerosis +1 OS. No evidence of intraocular inflammation was seen.

Fundus examination was remarkable for foveal pigment mottling OU. No diabetic or hypertensive retinopathy was present. SDOCT revealed atrophic cysts and ellipsoid zone disruption OU (Figure 1). Given the characteristic SDOCT findings, a diagnosis of macular telangiectasia type 2 was made and the patient was monitored every 6 months. Two years after initial examination, the patient developed acute visual loss to the level of 20/150 and presented with macular hemorrhage OD (Figure 2).

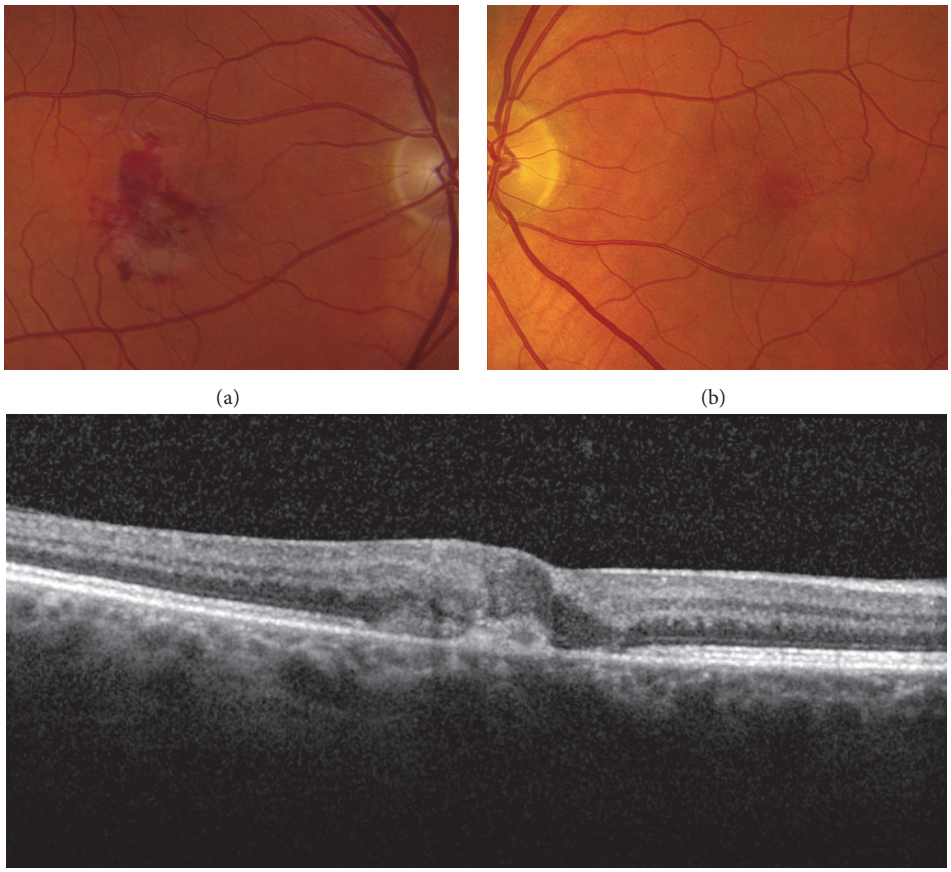
Macular OCTA was performed bilaterally. OCTA OD at the level of the outer retina and choriocapillaris shows subretinal neovascularization (Figure 3). OCTA OS reveals microvascular abnormalities in the deep capillary plexus of the retina most prominent temporally.

3. Discussion

This case demonstrates clinical features of nonproliferative and proliferative MacTel2 imaged with OCTA. Early in this condition changes in the retinal microvasculature begin temporal to the fovea in the deep capillary plexus. These abnormalities then extend circumferentially and into the superficial capillary plexus. Anastomoses form between both plexuses and retinal atrophy progresses. This can progress to SRN that can form connections with choroidal vessels [21].

OCTA technology can facilitate diagnosis of all stages of MacTel2 and essentially obviate the need for FA which does not discriminate between superficial and deep retinal vasculature. Also, the detection of subretinal neovascularization in cases in which there is no macular hemorrhage and when temporal leakage on FA is prominent can become challenging on FA.

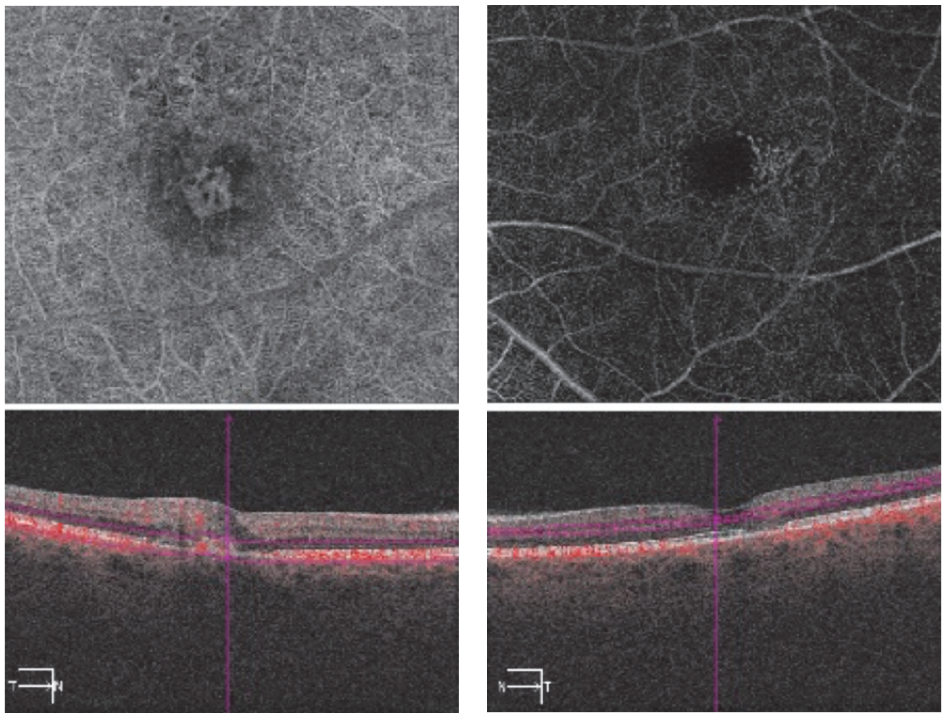
MacTel2 etiology continues to be poorly understood. Perifoveal Müller abnormalities may be a common pathway associated with the disease [1, 4]. Loss of regulation of these cells may lead to photoreceptor death, dysregulation of angiogenic and inflammatory factors, vascular ectasia, and subsequent subretinal neovascularization [22]. Histopathologic studies have shown thickening of retinal capillaries and



(a) (b)

(c)

Figure 2: Fundus photography 2 years after initial presentation. (a) Macular hemorrhages are present in the right eye. (b) Stable foveal pigment mottling is present in the left eye. (c) SDOCT of the right macula reveals a subretinal neovascular membrane.



(a) (b)

Figure 3: (a) OCTA OD at the level of the outer retina and choriocapillaris shows subretinal neovascularization. (b) OCTA OS demonstrates microvascular abnormalities in the deep capillary plexus of the retina most prominent temporally.

loss of Müller cells in subjects with macular telangiectasia [6, 23]. Recent studies have suggested that subjects with diabetes mellitus and hypertension are more likely to have MacTel2 [24]. This may be due to ischemic changes at the level of Müller cells. However, the mechanisms behind systemic disease and MacTel2 remain poorly understood. Further studies will elucidate the mechanisms that explain such associations.

In conclusion, OCTA is a new, fast, noninvasive imaging technology that has enabled improved understanding of the pathophysiology of many retinal vascular diseases including MacTel2. For this condition, OCTA not only facilitates diagnosis but also enables the clinician to monitor disease progression and quantify response to anti-VEGF therapy. Future studies with OCTA will hopefully illuminate additional features of MacTel2 and provide a better understanding of this complicated disease.

Conflicts of Interest

The authors declare that they have no conflicts of interest.

Acknowledgments

This case report is partially supported by NIH Center Core Grant P30EY014801, Research to Prevent Blindness Unrestricted Grant, and the Department of Defense.

References

[1] J. D. M. Gass and B. A. Blodi, "Idiopathic juxtafoveolar retinal telangiectasis. Update of classification and follow-up study," *Ophthalmology*, vol. 100, no. 10, pp. 1536–1546, 1993.

[2] L. A. Yannuzzi, A. M. C. Bardal, K. B. Freund, K.-J. Chen, C. M. Eandi, and B. Blodi, "Idiopathic macular telangiectasia," *JAMA Ophtalmology*, vol. 124, no. 4, pp. 450–460, 2006.

[3] J. L. Kovach and P. J. Rosenfeld, "Bevacizumab (avastin) therapy for idiopathic macular telangiectasia type II," *Retina*, vol. 29, no. 1, pp. 27–32, 2009.

[4] M. B. Powner, M. C. Gillies, M. Tretiach et al., "Perifoveal müller cell depletion in a case of macular telangiectasia type 2," *Ophthalmology*, vol. 117, no. 12, pp. 2407–2416, 2010.

[5] S. M. Cohen, M. L. Cohen, F. El-Jabali, and S. E. Pautler, "Optical coherence tomography findings in nonproliferative group 2A idiopathic juxtafoveal retinal telangiectasis," *Retina*, vol. 27, no. 1, pp. 59–66, 2007.

[6] J. D. M. Gass and W. R. Green, "Histopathologic study of presumed parafoveal telangiectasis [2] (multiple letters)," *Retina*, vol. 20, no. 2, pp. 226-227, 2000.

[7] M. J. Potter, S. M. Szabo, E. Y. Chan, and A. H. C. Morris, "Photodynamic therapy of a subretinal neovascular membrane in type 2A idiopathic juxtafoveolar retinal telangiectasis," *American Journal of Ophthalmology*, vol. 133, no. 1, pp. 149–151, 2002.

[8] Y. Jia, O. Tan, J. Tokayer et al., "Split-spectrum amplitude-decorrelation angiography with optical coherence tomography," *Optics Express*, vol. 20, no. 4, pp. 4710–4725, 2012.

[9] N. Takase, M. Nozaki, A. Kato, H. Ozeki, M. Yoshida, and Y. Ogura, "Enlargement of foveal avascular zone in diabetic eyes evaluated by en face optical coherence tomography angiography," *Retina*, vol. 35, no. 11, pp. 2377–2383, 2015.

[10] N. Suzuki, Y. Hirano, M. Yoshida et al., "Microvascular abnormalities on optical coherence tomography angiography in macular edema associated with branch retinal vein occlusion," *American Journal of Ophthalmology*, vol. 161, pp. 126–132e1, 2016.

[11] F. Coscas, A. Glacet-Bernard, A. Miere et al., "Optical coherence tomography angiography in retinal vein occlusion: evaluation of superficial and deep capillary plexa," *American Journal of Ophthalmology*, vol. 161, pp. 160–171e2, 2016.

[12] K. Sambhav, S. Grover, and K. V. Chalam, "The application of optical coherence tomography angiography in retinal diseases," *Survey of Ophthalmology*, 2017.

[13] M. K. Durbin, L. An, N. D. Shemonski et al., "Quantification of Retinal Microvascular Density in Optical Coherence Tomographic Angiography Images in Diabetic Retinopathy," *JAMA Ophthalmology*, vol. 135, no. 4, p. 370, 2017.

[14] N. A. Yannuzzi, N. Z. Gregori, L. Roisman, N. Gupta, B. E. Goldhagen, and R. Goldhardt, "Fluorescein angiography versus optical coherence tomography angiography in macular telangiectasia type i treated with bevacizumab therapy," *Ophthalmic Surgery, Lasers & Imaging Retina*, vol. 48, no. 3, pp. 263–266, 2017.

[15] S. G. Schwartz and J. W. Harbour, "Multimodal Imaging of Astrocytic Hamartomas Associated With Tuberous Sclerosis," *Ophthalmic Surgery, Lasers & Imaging Retina*, vol. 48, no. 9, pp. 756–758, 2017.

[16] S. Y. Chan, Q. Wang, Y. X. Wang, X. H. Shi, J. B. Jonas, and W. B. Wei, "Polypoidal choroidal vasculopathy upon optical coherence tomographic angiography," *Retina*, p. 1, 2017.

[17] M. Pellegrini, F. Corvi, E. A. Say, C. L. Shields, and G. Staurenghi, "Optical coherence tomography angiography features of choroidal neovascularization associated with choroidal nevus," *Retina*, p. 1, 2017.

[18] M. A. Gonzalez, D. Shechtman, J. M. Haynie, and L. Semes, "Unveiling idiopathic macular telangiectasia: clinical applications of optical coherence tomography angiography," *European Journal of Ophthalmology*, vol. 27, no. 4, pp. 0–0, 2017.

[19] R. F. Spaide, J. M. Klancnik Jr., and M. J. Cooney, "Retinal vascular layers imaged by fluorescein angiography and optical coherence tomography angiography," *JAMA Ophthalmology*, vol. 33, no. 1, pp. 45–50, 2015.

[20] L. Chidambara, S. G. K. Gadde, N. K. Yadav et al., "Characteristics and quantification of vascular changes in macular telangiectasia type 2 on optical coherence tomography angiography," *British Journal of Ophthalmology*, vol. 100, no. 11, pp. 1482–1488, 2016.

[21] L. Roisman and P. J. Rosenfeld, "Optical Coherence Tomography Angiography of Macular Telangiectasia Type 2," *Developments in Ophthalmology*, vol. 56, pp. 146–158, 2016.

[22] W. Shen, M. Fruttiger, L. Zhu et al., "Conditional müller cell ablation causes independent neuronal and vascular pathologies in a novel transgenic model," *The Journal of Neuroscience*, vol. 32, no. 45, pp. 15715–15727, 2012.

[23] W. R. Green, H. A. Quigley, Z. de la Cruz, and B. Cohen, "Parafoveal retinal telangiectasis," *Retina*, vol. 25, no. Supplement, pp. 162–170, 2005.

[24] T. E. Clemons, M. C. Gillies, E. Y. Chew et al., "Medical characteristics of patients with macular telangiectasia type 2 (MacTel Type 2) MacTel Project Report No. 3," *Ophthalmic Epidemiology*, vol. 20, no. 2, pp. 109–113, 2013.

Hindawi
Case Reports in Ophthalmological Medicine
Volume 2017, Article ID 4754231, 3 pages
https://doi.org/10.1155/2017/4754231

Case Report

Optical Coherence Tomography Angiography of Punctate Inner Choroidopathy

Blake M. Hampton,[1] Christopher M. Aderman,[2] Harry W. Flynn Jr.,[1] and Jayanth Sridhar[1]

[1]*Bascom Palmer Eye Institute, University of Miami, Miami, FL, USA*
[2]*Mid Atlantic Retina, The Retina Service of Wills Eye Hospital, Philadelphia, PA, USA*

Correspondence should be addressed to Jayanth Sridhar; jsridhar@med.miami.edu

Received 2 August 2017; Accepted 25 September 2017; Published 5 November 2017

Academic Editor: Hsin-Yi Chen

Purpose. To report a case of bilateral choroidal neovascularization (CNV) in punctate inner choroidopathy (PIC) visualized utilizing optical coherence tomography angiography (OCT-A). *Methods*. Case report. *Results*. A 29-year-old woman presented with new visual symptoms in both eyes. Fundoscopic exam revealed bilateral multifocal, small, well-defined lesions consistent with PIC. Optical coherence tomography demonstrated subretinal fluid and retinal pigment epithelium detachments (RPEDs) in both eyes. OCT-A revealed bilateral abnormal increased flow within the RPEDs consistent with CNV. Fluorescein angiography confirmed the presence of bilateral CNV. *Conclusion*. CNV secondary to PIC may be identified using noninvasive optical coherence tomography angiography.

1. Introduction

Punctate inner choroidopathy (PIC) is a posterior uveitis belonging to the group of idiopathic white dot syndromes. It tends to present in young to middle aged women with myopia [1]. Choroidal neovascularization (CNV) may develop in the setting of PIC, leading to visual impairment [2]. Optical coherence tomography (OCT) and fluorescein angiography (FA) are most commonly utilized to identify and monitor CNV in patients with PIC.

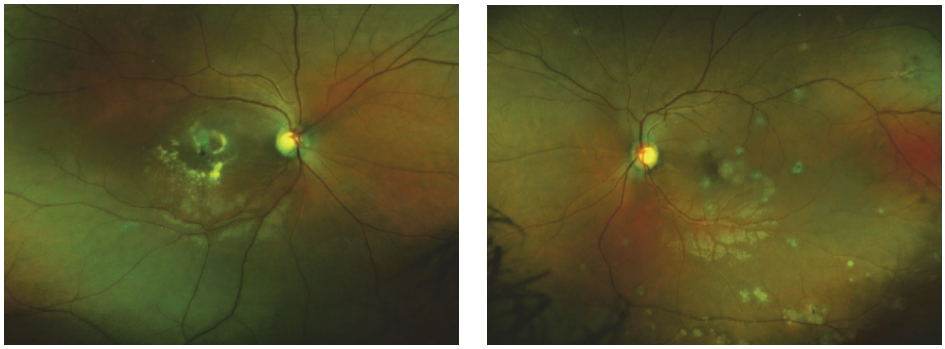
Optical coherence tomography angiography (OCT-A) is a relatively new, noninvasive imaging modality that had been previously shown to successfully identify CNV in patients with diseases such as neovascular age-related macular degeneration (AMD) before a recently published series demonstrated its utility in PIC [3, 4]. This report highlights the usefulness of OCT-A in cases of PIC to guide management.

2. Case Presentation

A 29-year-old woman with no prior medical history presented with a new black spot in her right eye and wavy lines in her left eye. Best corrected Snellen visual acuity was finger count in the right eye and 20/40 in the left eye. Anterior chamber examination was unremarkable and trace vitreous cell was noted in both eyes. Fundoscopic exam revealed multiple, small, punched-out variably pigmented lesions in the posterior pole of both eyes with scattered peripheral lesions (Figure 1). OCT (Spectralis HRA + OCT, Heidelberg Engineering, Inc., Heidelberg, Germany) demonstrated RPE disruption and deposition with intraretinal fluid with retinal atrophy in the right eye (Figure 2(a)) and RPE disruption with subretinal and intraretinal fluid in the left eye (Figure 2(b)). OCT-A (RTVue-XR Avanti, Optovue, Fremont, CA) at the level of the RPE disruption in both eyes revealed increased abnormal flow consistent with bilateral CNV (Figure 3). FA (Optos, Optos Inc., Dunfermline, United Kingdom) demonstrated late leakage corresponding to the CNV (Figure 4). The patient received bilateral intravitreal injections of bevacizumab and was lost to follow-up after a single treatment.

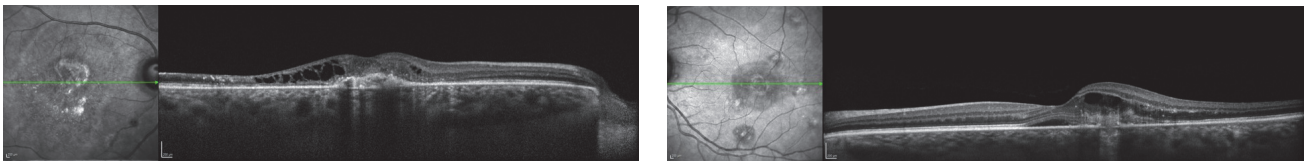
3. Discussion

FA and OCT are the current gold standard for identifying and monitoring CNV. One limitation of these techniques is



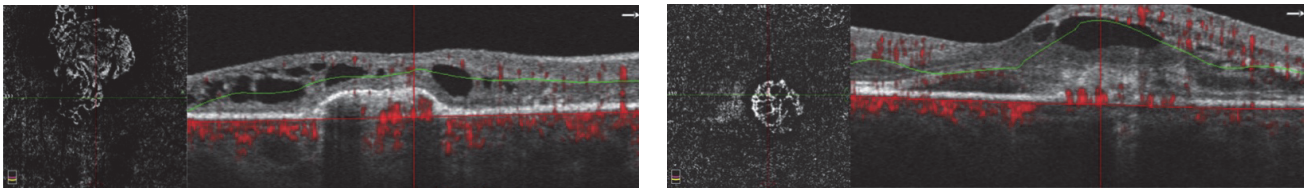
(a) (b)

Figure 1: Fundus photography of the right (a) and left (b) eyes demonstrating multiple posterior small variably pigmented lesions consistent with punctate inner choroidopathy.



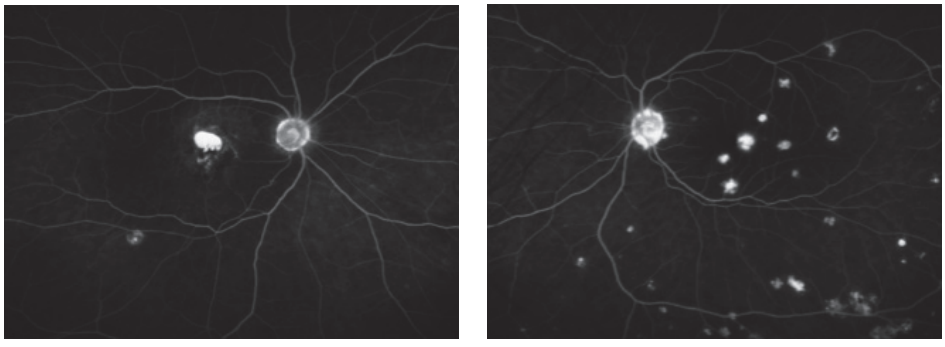
(a) (b)

Figure 2: (a) Right eye optical coherence tomography revealing retinal pigment epithelium disruption and elevation and subretinal deposition with intraretinal cystic changes. (b) Left eye optical coherence tomography revealing retinal pigment epithelium disruption with associated subretinal and intraretinal fluid.



(a) (b)

Figure 3: Optical coherence tomography angiography at level of retinal pigment epithelium elevation in right (a) and left (b) eyes demonstrates abnormal flow consistent with choroidal neovascularization.



(a) (b)

Figure 4: Fluorescein angiography of right (a) and left (b) eyes reveals late leakage posteriorly consistent with choroidal neovascularization.

that CNV and inflammatory lesions may present similarly with elevation of the retinal pigment epithelium (RPE) and subretinal fluid on OCT and leakage on FA [5]. It is important to be able to differentiate the two, as CNV typically requires prompt treatment with intravitreal antivascular endothelial growth factor (anti-VEGF) agents whereas treatment of inflammatory lesions relies on immunosuppressant therapy [5]. A recently published case series demonstrated the value of OCT-A in detecting CNV in PIC. In some instances, FA was unable to distinguish active inflammation from CNV while OCT-A clearly revealed abnormal flow corresponding to CNV [4]. OCT-A carries the additional advantage of being noninvasive and avoiding potential rare side effects such as anaphylaxis from fluorescein dye.

As the technology continues to evolve, OCT-A should continue to provide more information about classically described disease processes such as PIC. In the same case series, OCT-A showed the progression of CNV activity as measured by flow while OCT imaging remained stable [4]. Beyond its ability to simply visualize CNV, in the future OCT-A may ultimately help elucidate how those new vessels respond to anti-VEGF treatment [6].

Conflicts of Interest

None of the authors has any relevant financial conflicts of interest.

References

[1] R. C. Watzke, A. J. Packer, J. C. Folk, W. E. Benson, D. Burgess, and R. R. Ober, "Punctate inner choroidopathy," *American Journal of Ophthalmology*, vol. 98, no. 5, pp. 572–584, 1984.

[2] T. W. Olsen, J. Capone A., J. Sternberg P., H. E. Grossniklaus, D. F. Martin, and S. Aaberg T.M., "Subfoveal choroidal neovascularization in punctate inner choroidopathy: Surgical management and pathologic findings," *Ophthalmology*, vol. 103, no. 12, pp. 2061–2069, 1996.

[3] T. E. de Carlo, M. A. Bonini Filho, A. T. Chin et al., "Spectral-domain optical coherence tomography angiography of choroidal neovascularization," *Ophthalmology*, vol. 122, no. 6, pp. 1228–1238, 2015.

[4] A. L. Levison, K. M. Baynes, C. Y. Lowder, P. K. Kaiser, and S. K. Srivastava, "Choroidal neovascularisation on optical coherence tomography angiography in punctate inner choroidopathy and multifocal choroiditis," *British Journal of Ophthalmology*, vol. 101, no. 5, pp. 616–622, 2017.

[5] D. Huang, Y. Jia, M. Rispoli, O. Tan, and B. Lumbroso, "Optical coherence tomography angiography of time course of choroidal neovascularization in response to anti-angiogenic treatment," *Retina*, vol. 35, no. 11, pp. 2260–2264, 2015.

[6] R. Amer, E. Priel, and M. Kramer, "Spectral-domain optical coherence tomographic features of choroidal neovascular membranes in multifocal choroiditis and punctate inner choroidopathy," *Graefe's Archive for Clinical and Experimental Ophthalmology*, vol. 253, no. 6, pp. 949–957, 2015.

Hindawi
Case Reports in Ophthalmological Medicine
Volume 2017, Article ID 4057615, 4 pages
https://doi.org/10.1155/2017/4057615

Case Report

Multimodal Imaging Analysis in a Case with Congenital Fovea-Involving Retinal Macrovessel and Excellent Visual Acuity

Vishal Shah,[1] M. Ashwin Reddy,[1,2] and Vasilios P. Papastefanou[1]

[1]*Ophthalmology Department, The Royal London Hospital, Whitechapel, London E1 1BB, UK*
[2]*Department of Paediatrics, Moorfields Eye Hospital Foundation Trust, London EC1V 2PD, UK*

Correspondence should be addressed to Vishal Shah; vbshah@doctors.org.uk

Received 29 July 2017; Accepted 27 September 2017; Published 19 October 2017

Academic Editor: Stephen G. Schwartz

Purpose. Congenital retinal macrovessels (CRM) represent rare aberrant vasculature of the retinal vessels that can supply or drain the macula. In this report, the optical coherence tomography angiography features of a congenital retinal macrovessel are discussed. *Methods*. The history and examination findings are presented alongside swept-source OCT angiography with corresponding B scan and en face OCT imaging. *Patients*. The case is a 12-year-old female patient with excellent best-corrected visual acuity in both eyes. *Results*. Swept-source OCT angiography demonstrated considerable loss of the foveal avascular zone at the levels of the superficial and deep capillary plexus. *Discussion*. In this case there was no detrimental effect on vision despite anatomical loss of the foveal avascular zone.

1. Introduction

Congenital retinal macrovessel was first described by Mauthner as "an aberrant retinal vessel" in 1869 [1]. Aberrant retinal vessels were noted to be rare and small in size with little or no branching and passing only to the macular region. Most were purported to arise directly from the short ciliary vessels, with occasional cases noted to communicate directly with the choroidal circulation. They were also noted to arise at both the nasal and temporal disc margins [2]. Aberrant macrovessels crossing the fovea can cause visual impairment but this is not always the case [3]. The pathophysiology is unknown; however one theory proposes a role for an intrauterine hypoxic stimulus that may then permit vascular proliferation reaching the center of the foveola [4]. In this report we describe OCT-A features in a case with congenital retinal macrovessel (CRM).

2. Case Presentation

A 12-year-old girl presented to the department having been referred from her opticians with an incidental finding of aberrant vasculature in the right fundus. She had no history of trauma or ocular surgery and no family history of ophthalmic disease. On examination the vision was 20/66 unaided correcting to 20/20 with glasses in the right eye and 20/16 with glasses in the left eye. Near vision was N5 bilaterally. Refraction performed at referral revealed that she was moderately myopic bilaterally (−3.50 diopters). Anterior segment examination of both eyes and intraocular pressures were normal (11 and 16 mmHg in the right and left eyes, resp.). Examination of the right fundus revealed a healthy optic disc, with an aberrant retinal macrovessel (Figure 1) arising from the inferotemporal major vascular arcade and extending towards the fovea. There were no exudates or haemorrhages and no epiretinal membrane. The appearances of the remaining retinal vessels and peripheral fundus were unremarkable. The left fundus was completely normal.

Swept-source optical coherence tomography angiography (SS-OCT-A) imaging of the right fundus (Triton, Topcon) (Figure 2) demonstrated the aberrant vessel encroaching into the foveal avascular zone (FAZ) but not passing through the center of the fovea. The anatomy of the neurosensory retina appeared preserved, despite the elevation of the foveal depression. No evidence of retinal layer loss, cysts, leakage, or exudation was noted. There was no evidence of disturbance in the vitreoretinal interface and no disturbance of the RPE-choriocapillaris complex (Figure 3).

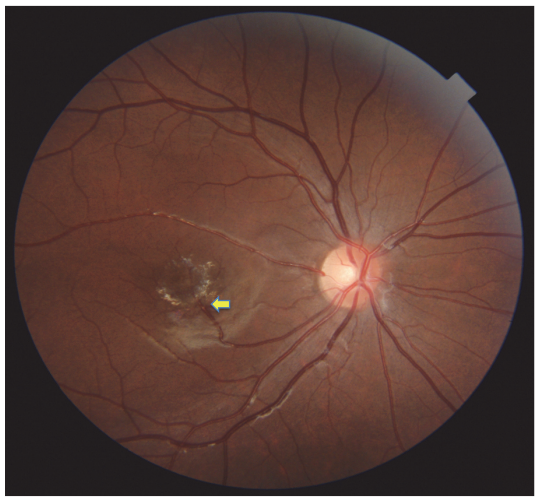


Figure 1: Right fundus photograph of a 12-year-old patient with a congenital retinal macrovessel (arrow) extending towards the fovea in a superotemporal course. The foveal reflex appears disturbed by the macrovessel.

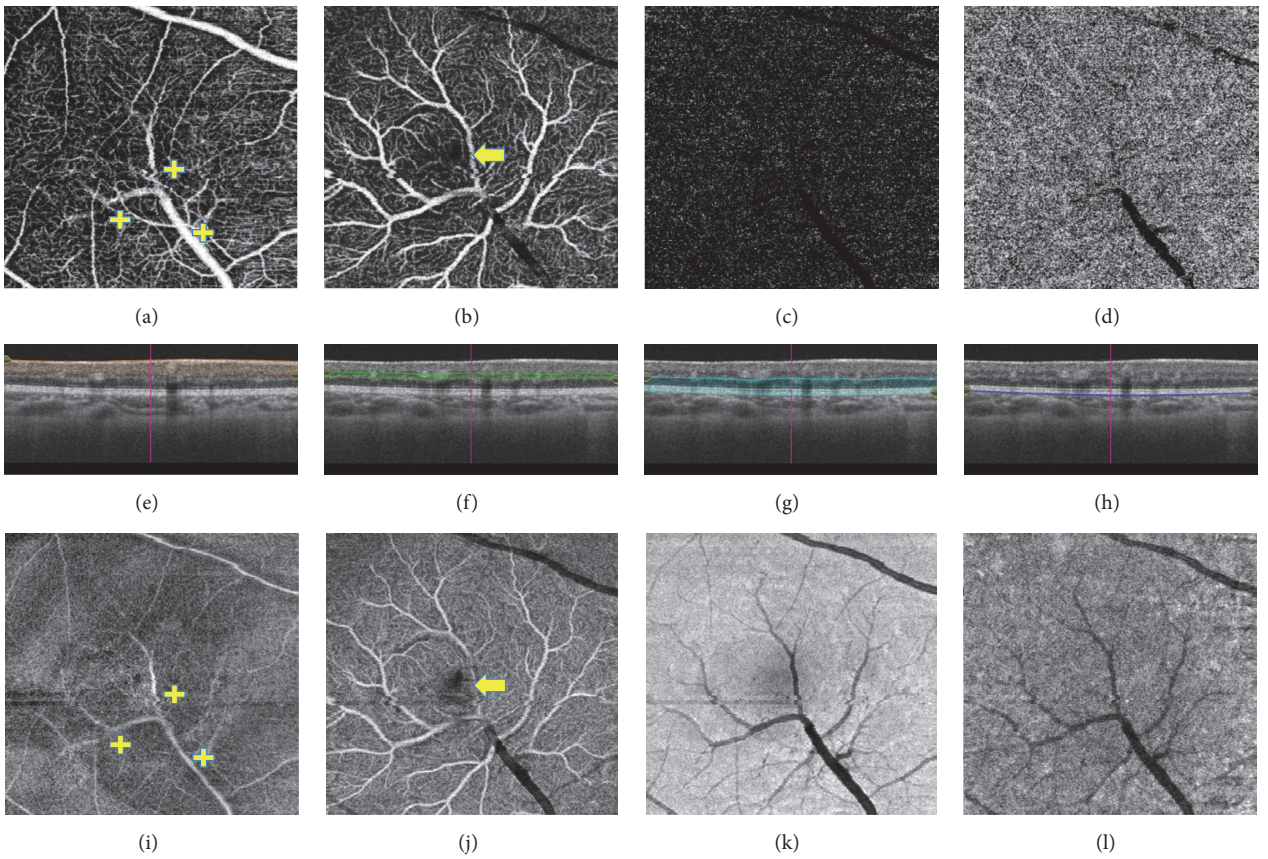


Figure 2: Swept-source OCT angiography (a, b, c, and d) with corresponding segmentation B scan (e, f, g, and h) and en face OCT imaging (i, j, k, and l) of the right eye of a 12-year-old patient with congenital retinal macrovessel (a, e, and i). At the level of the superficial capillary plexus the principal stem of the macrovessel and two main bifurcations are noted (+). Microvascular interconnections obliterate the foveal avascular zone (a). Corresponding SS-en face OCT does not indicate any leakage from the macrovessels (b, f, and j). At the level of the deep capillary plexus there are multiple branches emanating from the principal stem and the bifurcations directed at the level of the outer plexiform layer. The latter extend and bifurcate further horizontally prior to fusing with the capillaries of the deep vascular plexus. The remaining foveal avascular zone is also depicted (arrow), considerably reduced in extent. En face OCT also clearly demonstrates the aberrant vasculature and no evidence of leakage at this level. Scans at the level of outer retina (c, g, and k) and choriocapillaris (d, h, and l) demonstrate projection artefacts of major vessels without evidence of coexisting pathology.

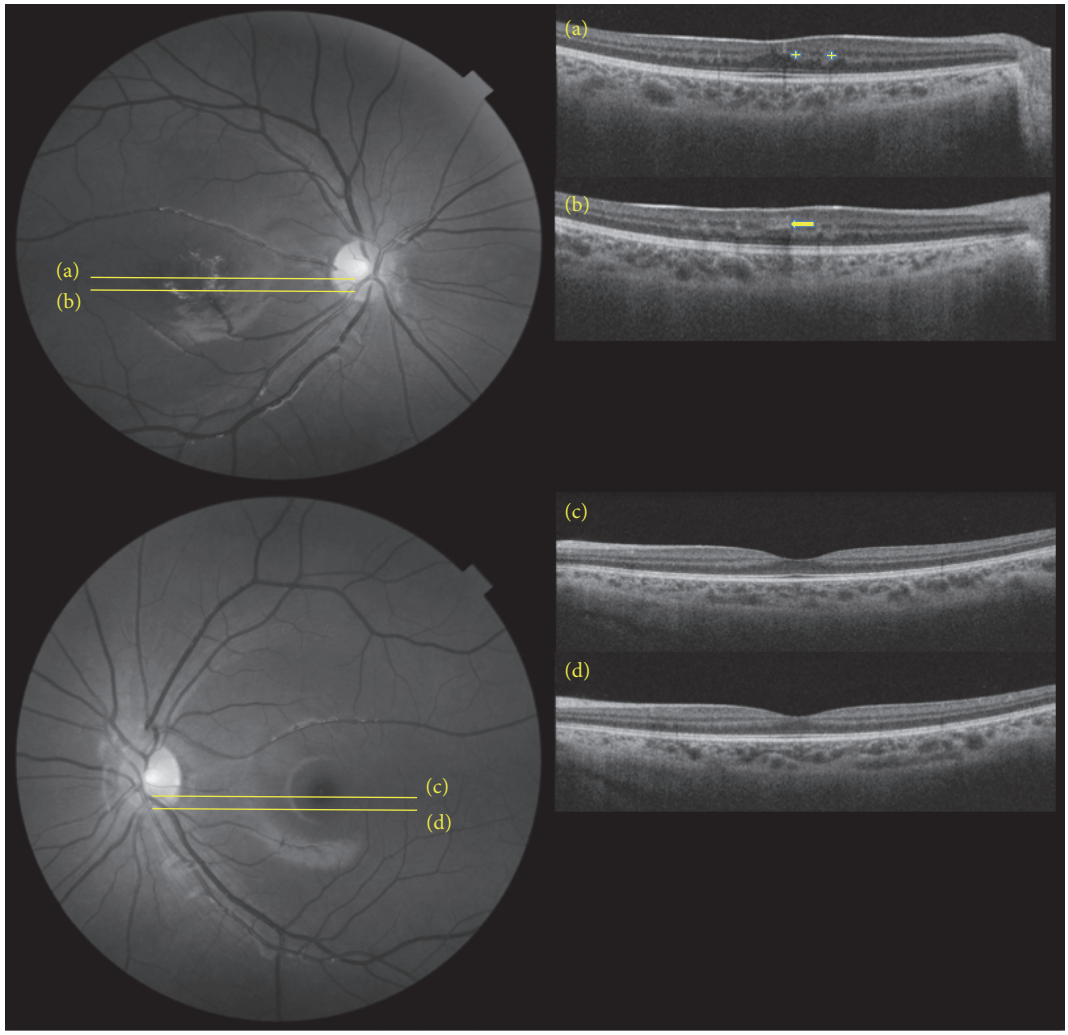


Figure 3: Swept-source OCT at the level of the right fovea (a) and inferior to the fovea (b) in a 12-year-old patient with congenital retinal macrovessels. The stem of the vessel is depicted at the level of the inner nuclear and plexiform layers (arrow) and its branches at the level of the outer nuclear layer (+). The foveal anatomy is largely preserved, though the foveal depression is reduced. There is no evidence of disturbance of the RPE-choriocapillaris complex also confirmed by OCT-A in the previous image. Corresponding swept-source OCT scans at the level of the left fovea (c) and inferior to the fovea (d) are included for comparison.

Swept-source OCT angiography demonstrated considerable loss of the foveal avascular zone at the levels of the superficial and deep capillary plexus (a). Outer retinal and choriocapillaris SS-OCT-A did not yield any abnormal findings. Corresponding en face SS-OCT again confirmed no leakage from the macrovessels (b, f, and j).

3. Discussion

Archer et al. [5] classified congenital vessels into three categories based on the caliber of communicating vessels, presence of capillary plexus bridging the vessels, and the grade of visual impairment. Group 1 is defined as anomalous arteriovenous communications being localized to a single sector of the retina, most frequently the macula. Group 2 demonstrates direct arteriovenous communications that are larger in size than the vessels in Group 1 but in the absence of interposition of arteriolar or capillary elements. In Group 3 the arteriovenous vessels are large and entangled and give rise to retinal complications and consequent severe visual impairment. Chawla and colleagues recently reported a similar case of CRM [6]. This case was associated with poor BCVA in the affected eye since childhood and they attributed this to the distorted FAZ. This effect of aberrant retinal vasculature on vision was also emphasized in a previously reported case of a white male with two macular arteriovenous anastomoses and a small venous tributary crossing the fovea. The authors associated reduced visual acuity with the loss of the normal architecture of the fovea [7]. Goel et al. recently reported a case of reduced visual acuity due to vitreous haemorrhage secondary to congenital retinal macrovessel, highlighting the potential complications of such aberrant retinal vasculature [8]. In this case, there was no detrimental effect on vision, despite anatomical loss of the foveal avascular

zone. Interestingly this disturbance of the vascular status of the macula did not considerably reflect in the anatomy of the neurosensory retina.

Disclosure

The authors alone are responsible for the content and writing of the paper.

Conflicts of Interest

The authors declare that there are no conflicts of interest regarding the publication of this article.

References

[1] L. Mauthner, Lehrbuch der ophthalmoscopie, Vienna, Austria: Tendler; 1869: 249.

[2] B. A. Randall, "Cilio-Retinal or Aberrant Vessels," *Transactions of the American Ophthalmological Society*, vol. 4, pp. 511–517, 1887.

[3] G. C. Brown, L. A. Donoso, L. E. Magargal, R. E. Goldberg, and L. K. Sarin, "Congenital Retinal Macrovessels," *JAMA Ophtalmology*, vol. 100, no. 9, pp. 1430–1436, 1982.

[4] N. Ashton, "The mode of development of the retinal vessels in man," in *The William MacKenzie Centenary Symposium on the ocular circulation in Health and Disease*, J. S. Cant, Ed., pp. 7–17, CV Mosby Co, St Louis, Missouri, 1969.

[5] D. B. Archer, A. Deutman, J. T. Ernest, and A. E. Krill, "Arteriovenous communications of the retina," *American Journal of Ophthalmology*, vol. 75, no. 2, pp. 224–241, 1973.

[6] R. Chawla, R. Bypareddy, K. Tripathy, S. S. Daggumili, and A. S. Tomar, "Optical coherence tomography angiography imaging of congenital retinal macrovessel," *Ophthalmic Surgery, Lasers and Imaging Retina*, vol. 47, no. 10, pp. 972-973, 2016.

[7] G. de Crecchio, G. Pappalardo, F. Pascotto, and R. Forte, "Aberrant congenital macular vessel crossing the fovea: Evaluation with optical coherence tomography," *Eye*, vol. 20, no. 12, pp. 1464–1466, 2006.

[8] N. Goel, V. Kumar, and B. Ghosh, "Congenital retinal macrovessel associated with vitreous hemorrhage," *Journal of American Association for Pediatric Ophthalmology and Strabismus*, vol. 21, no. 1, pp. 83–85, 2017.

Hindawi
Case Reports in Ophthalmological Medicine
Volume 2017, Article ID 1368581, 4 pages
https://doi.org/10.1155/2017/1368581

Case Report

Optical Coherence Tomography Angiography of Two Choroidal Nevi Variants

Victor M. Villegas, Armando L. Monroig, Lazaro H. Aguero, and Stephen G. Schwartz

Department of Ophthalmology, Bascom Palmer Eye Institute, University of Miami Miller School of Medicine, Naples, FL, USA

Correspondence should be addressed to Victor M. Villegas; v.villegas@med.miami.edu

Received 20 July 2017; Accepted 24 August 2017; Published 9 October 2017

Academic Editor: Alexander A. Bialasiewicz

Optical coherence tomography angiography (OCT-A) is a recently established noninvasive technology for evaluation of the retinal and choroidal vasculature. The literature regarding the findings in choroidal nevi is scarce. We report the OCT-A findings associated with two different variants. Subject one had decreased vascular flow signal in the choroidal, choriocapillaris, deep retinal, and superficial retinal layers. Subject two had decreased vascular flow signal in the choroidal, choriocapillaris, and deep retinal layers with a normal vascular flow signal in the superficial retinal layer. To our knowledge, these patterns of decreased vascular flow signals have not been previously reported using OCT-A. This may be due to blockage from the choroidal nevus, true diminished blood flow (ischemia), or other factors.

1. Introduction

Optical coherence tomography angiography (OCT-A) is a recently established technology that allows visualization of the chorioretinal vasculature without intravenous dye injection [1–4]. The application of this noninvasive technique is mainly designed to evaluate blood flow [1]. Various retinal and choroidal diseases have been described using OCT-A, including some intraocular tumors [2–10]. However, the literature regarding OCT-A characteristics of choroidal nevi is relatively limited [11].

The purpose of this report is to illustrate some of the OCT-A characteristics associated with two lesions diagnosed clinically as benign choroidal nevi. In both cases the commercially available Cirrus 5000 with AngioPlex (Zeiss, Jena, Germany) was used, without any subsequent image modification or processing.

2. Case Report

2.1. Subject 1. A 65-year-old female with history of polymyalgia rheumatica presented for scheduled follow-up of a choroidal nevus of the left eye (OS). Visual acuity was 20/20 in each eye (OU). The examination was normal except for a juxtapapillary pigmented choroidal nevus OS, with overlying drusen, and without orange pigment or subretinal fluid (Figure 1). OCT-A 6 mm × 6 mm showed a decreased vascular flow signal in the choroidal, choriocapillaris, deep retinal, and superficial retinal layers (Figure 2).

2.2. Subject 2. A healthy 60-year-old male presented for scheduled follow-up of a choroidal nevus of the right eye (OD). Visual acuity was 20/20 in both eyes (OU). The examination was normal except for a pigmented choroidal nevus OD with a depigmented halo around it. No drusen, orange pigment, or subretinal fluid was present (Figure 3). OCT-A 6 mm × 6 mm showed a decreased vascular flow signal in the choroidal, choriocapillaris, and deep retinal layers with a normal vascular flow signal in the superficial retinal layer (Figure 4).

3. Discussion

The evaluation of choroidal nevi can be a diagnostic challenge because in some cases a distinction between a benign nevus and a small choroidal melanoma is not readily apparent [12]. Multiple studies have reported hypervascularity using fluorescein angiography (FA) in patients with uveal melanoma

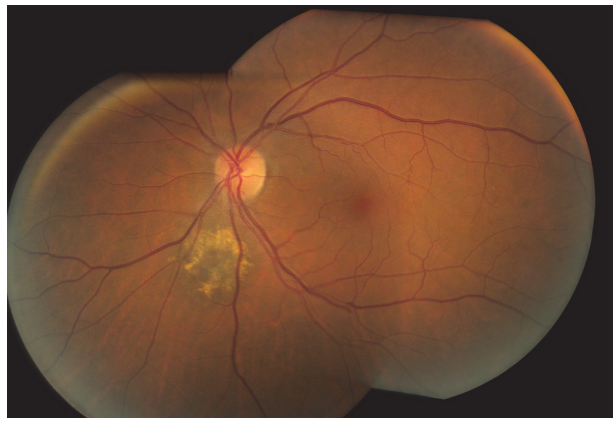


Figure 1: Fundus photography OS shows juxtapapillary pigmented choroidal nevus.

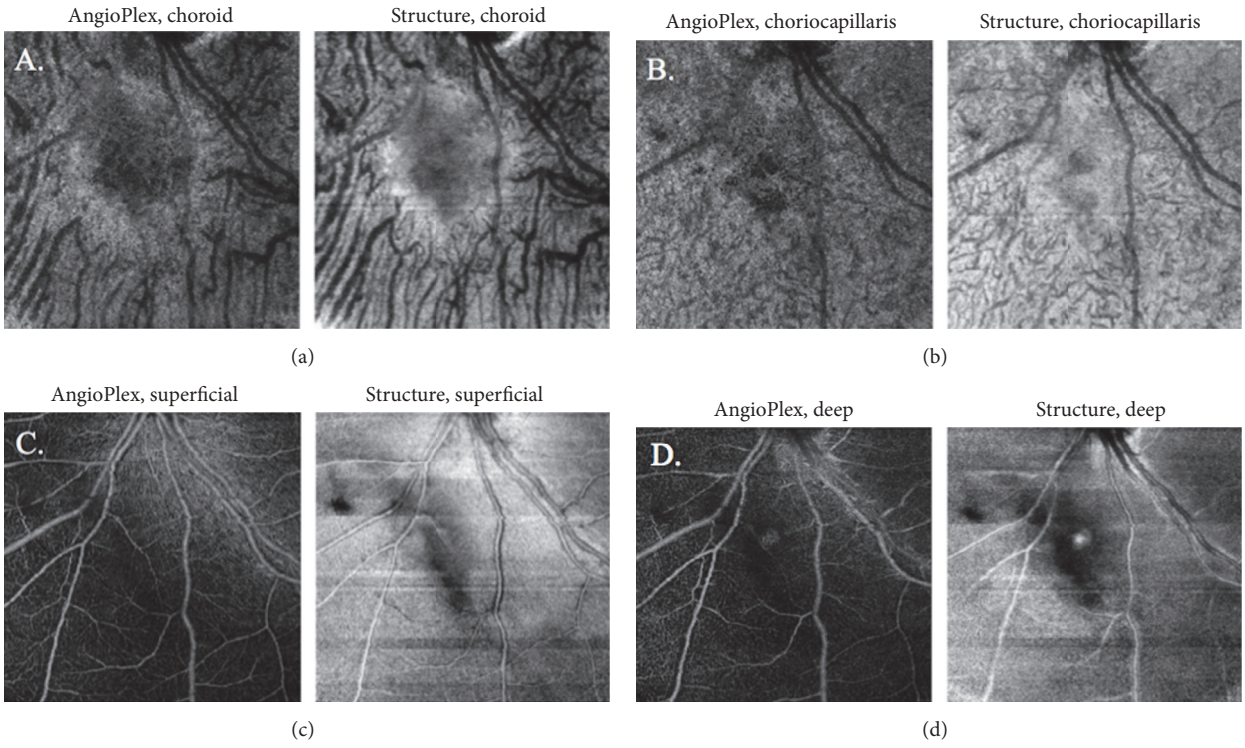


Figure 2: AngioPlex OCT-A 6 mm × 6 mm. (a) Choroidal plexus layer showing a decreased vascular flow signal. (b) Choriocapillary plexus layer showing a decreased vascular flow signal. (c) Superficial retinal plexus layer showing a subtle decreased vascular flow signal. (d) Deep retinal plexus layer showing a subtle decreased vascular flow signal.

[13, 14]. Increased vascularity in solid tumors is a hallmark of malignant transformation [15]. For example, a recent study that evaluated subjects with iris melanomas with OCT-A reported increased vascularity, with disorganized and tortuous intratumoral vascular patterns and increased vessel density [16].

OCT-A may offer certain advantages over FA in this situation. OCT-A is fast and noninvasive and has no risk of allergy. OCT-A can readily distinguish different layers of the retina and choroid, which cannot be performed with FA [17, 18]. A previous study that imaged choroidal nevi with OCT-A reported decreased vascular flow signal only in the deep retinal layer [11]. Subject 1 in the current series showed decreased vascular flow signal in the superficial, deep, and subretinal layers. Subject 2 had normal vascular flow signal in the superficial retinal plexus but decreased vascular flow signal in the deep and subretinal layers.

To our knowledge, these patterns of decreased vascular flow signals have not been previously reported using OCT-A. This may be due to blockage from the choroidal nevus, true diminished blood flow (ischemia), or other factors. If ischemia is truly present, then diminished blood flow in the

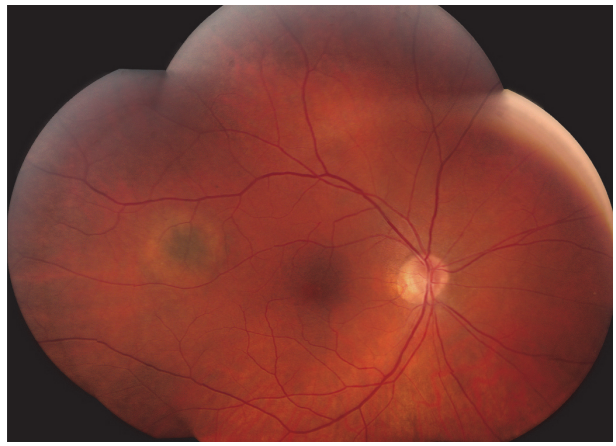


Figure 3: Fundus photography OD shows a temporal halo choroidal nevus.

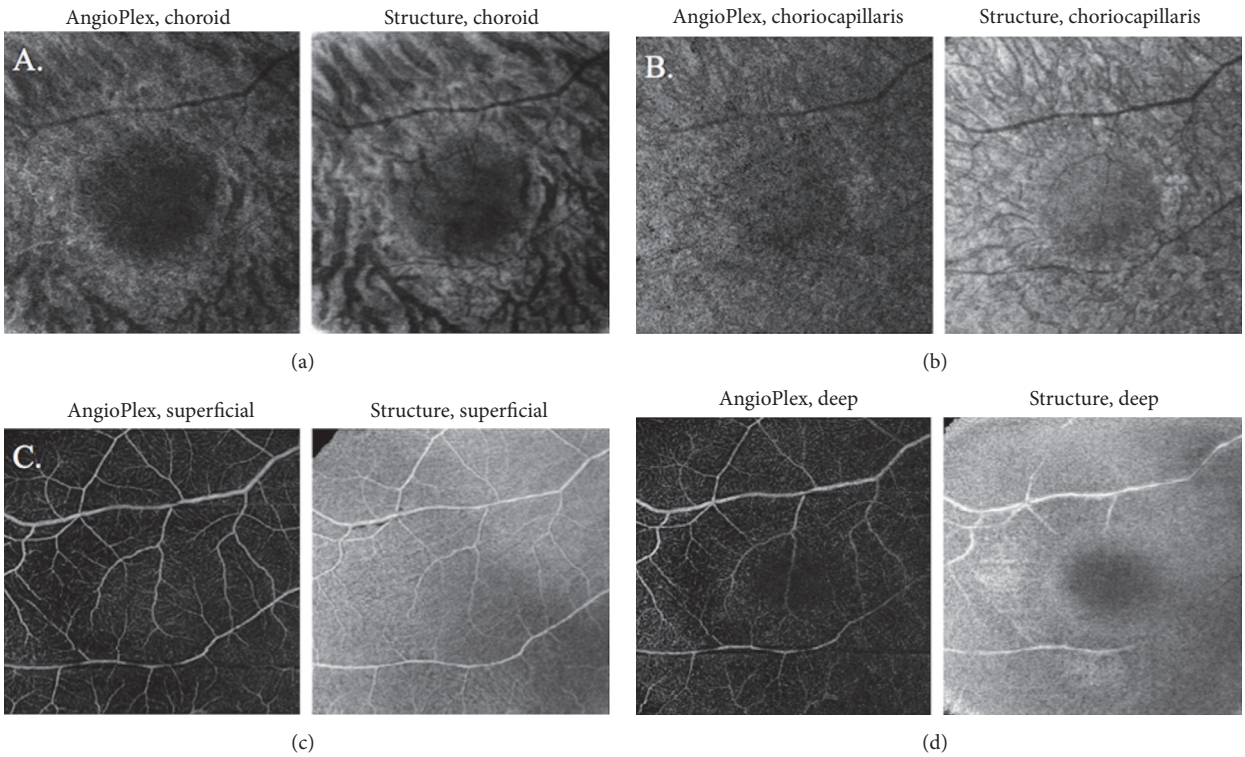


Figure 4: AngioPlex OCT-A 6 mm × 6 mm. (a) Choroidal plexus layer showing a decreased vascular flow signal. (b) Choriocapillary plexus layer showing a decreased vascular flow signal. (c) Superficial retinal plexus layer showing a normal vascular flow signal. (d) Deep retinal plexus layer showing a subtle central decreased vascular flow signal.

superficial retina may explain the atrophic changes that are typically seen in the retina overlying some nevi [19].

OCT-A has some limitations, including expense and the relatively long image acquisition time, approximately 3 seconds. Some patients cannot hold their gaze for this long, especially because most nevi are not in the macula. Furthermore, inadequate penetration through highly pigmented or thick tumors may limit the use of this technology to small nevi/melanomas (less than 2 mm in thickness). It might be interesting to compare OCT, OCT-A, and B-scan but echography was not performed on these two patients because the lesions were clinically flat.

In summary, OCT-A may provide a simple, quick, and safe way to monitor choroidal nevi. OCT-A may be particularly useful in cases with high-risk features to detect changes in vascularity. As more clinical examples are collected, our understanding of using OCT-A to image pigmented lesions should increase.

Conflicts of Interest

Dr. Stephen G. Schwartz discloses personal fees from Alimera, Bausch + Lomb, and Welch Allyn unrelated to this work. All other authors declare that they have no conflicts of interest.

Acknowledgments

This paper was partially supported by NIH Center Core Grant P30EY014801, Research to Prevent Blindness Unrestricted Grant, Department of Defense.

References

[1] Y. Jia, O. Tan, J. Tokayer et al., "Split-spectrum amplitude-decorrelation angiography with optical coherence tomography," *Optics Express*, vol. 20, no. 4, pp. 4710–4725, 2012.

[2] N. Takase, M. Nozaki, A. Kato, H. Ozeki, M. Yoshida, and Y. Ogura, "Enlargement of foveal avascular zone in diabetic eyes evaluated by en face optical coherence tomography angiography," *Retina*, vol. 35, no. 11, pp. 2377–2383, 2015.

[3] N. Suzuki, Y. Hirano, M. Yoshida et al., "Microvascular abnormalities on optical coherence tomography angiography in macular edema associated with branch retinal vein occlusion," *American Journal of Ophthalmology*, vol. 161, pp. 126–132e1, 2016.

[4] F. Coscas, A. Glacet-Bernard, A. Miere et al., "Optical coherence tomography angiography in retinal vein occlusion: evaluation of superficial and deep capillary plexa," *American Journal of Ophthalmology*, vol. 161, pp. 160–171e2, 2016.

[5] K. Sambhav, S. Grover, and K. V. Chalam, "The application of optical coherence tomography angiography in retinal diseases," *Survey of Ophthalmology*, 2017.

[6] M. K. Durbin, L. An, N. D. Shemonski et al., "Quantification of Retinal Microvascular Density in Optical Coherence Tomographic Angiography Images in Diabetic Retinopathy," *JAMA Ophthalmology*, vol. 135, no. 4, p. 370, 2017.

[7] N. A. Yannuzzi, N. Z. Gregori, L. Roisman, N. Gupta, B. E. Goldhagen, and R. Goldhardt, "Fluorescein angiography versus optical coherence tomography angiography in macular telangiectasia type i treated with bevacizumab therapy," *Ophthalmic Surgery, Lasers and Imaging Retina*, vol. 48, no. 3, pp. 263–266, 2017.

[8] S. G. Schwartz and J. W. Harbour, "Multimodal Imaging of Astrocytic Hamartomas Associated With Tuberous Sclerosis," *Ophthalmic Surgery, Lasers & Imaging Retina*, vol. 48, no. 9, pp. 756–758, 2017.

[9] S. Y. Chan, Q. Wang, Y. X. Wang, X. H. Shi, J. B. Jonas, and W. B. Wei, "Polypoidal choroidal vasculopathy upon optical coherence tomographic angiography," *Retina*, p. 1, 2017.

[10] M. Pellegrini, F. Corvi, E. A. Say, C. L. Shields, and G. Staurenghi, "Optical coherence tomography angiography features of choroidal neovascularization associated with choroidal nevus," *Retina*, p. 1, 2017.

[11] G. Cennamo, M. R. Romano, M. A. Breve et al., "Evaluation of choroidal tumors with optical coherence tomography: enhanced depth imaging and OCT-angiography features," *Eye*, vol. 31, no. 6, pp. 906–915, 2017.

[12] A. D. Singh, P. Kalyani, and A. Topham, "Estimating the risk of malignant transformation of a choroidal nevus," *Ophthalmology*, vol. 112, no. 10, pp. 1784–1789, 2005.

[13] J. K. Dart, R. J. Marsh, A. Garner, and R. J. Cooling, "Fluorescein angiography of anterior uveal melanocytic tumours," *British Journal of Ophthalmology*, vol. 72, no. 5, pp. 326–337, 1988.

[14] B. L. Hodes, M. Gildenhar, and E. Choromokos, "Fluorescein Angiography in Pigmented Iris Tumors," *JAMA Ophtalmology*, vol. 97, no. 6, pp. 1086–1088, 1979.

[15] J. Folkman, "Tumor angiogenesis: therapeutic implications," *The New England Journal of Medicine*, vol. 285, no. 21, pp. 1182–1186, 1971.

[16] A. H. Skalet, Y. Li, C. D. Lu et al., "Optical Coherence Tomography Angiography Characteristics of Iris Melanocytic Tumors," *Ophthalmology*, vol. 124, no. 2, pp. 197–204, 2017.

[17] R. F. Spaide, J. M. Klancnik Jr., and M. J. Cooney, "Retinal vascular layers imaged by fluorescein angiography and optical coherence tomography angiography," *JAMA Ophthalmology*, vol. 33, no. 1, pp. 45–50, 2015.

[18] Q. Zhang, A. Zhang, C. S. Lee et al., "Projection Artifact Removal Improves Visualization and Quantitation of Macular Neovascularization Imaged by Optical Coherence Tomography Angiography," *Ophthalmology Retina*, vol. 1, no. 2, pp. 124–136, 2017.

[19] G. Espinoza, B. Rosenblatt, and J. W. Harbour, "Optical coherence tomography in the evaluation of retinal changes associated with suspicious choroidal melanocytic tumors," *American Journal of Ophthalmology*, vol. 137, no. 1, pp. 90–95, 2004.