Swept-Source OCT Angiography of Macular Telangiectasia Type 2

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BACKGROUND AND OBJECTIVE: To evaluate the central macular microvascular network in patients with macular telangiectasia type 2 (MacTel2) using optical coherence tomography (OCT)-based microangiography (OMAG).

PATIENTS AND METHODS: Prospective, observational study of patients with MacTel2 evaluated using a swept-source OCT (SS-OCT) prototype. OMAG was performed using a 3 mm × 3 mm central foveal raster scan. The algorithm segmented the retina into three layers. Microvascular distribution was depicted as en face images, and qualitative information was compared to fluorescein angiography (FA) images.

RESULTS: OMAG detected abnormal microvasculature in all MacTel2 eyes, predominantly in the middle retinal layers with neovascularization in the outer retina. These vessels correlated well with the FA alterations. The abnormal temporal, juxtafoveal microvasculature in MacTel2 became apparent as the disease progressed and in later stages tended to extend circumferentially, with anastomotic vessels temporally.

CONCLUSION: OMAG provided detailed, depth-resolved information about the perifoveal macular microvasculature in MacTel2. In most cases, images were better using OMAG than FA. The OMAG images demonstrated that most of the leakage seen on FA appeared to arise from the abnormal perifoveal microvasculature in the middle retinal layer.


INTRODUCTION

Macular telangiectasia type 2 (MacTel2) is a bilateral retinal disease that presents with asymmetric severity during the 5th to 7th decades of life and affects the juxtafoveal region of the macula.1-3 The pathogenesis of the disease is unknown, and currently there is no proven treatment. According to the limited histologic information, the telangiectatic-appearing vascular changes are thought to originate in the deep retinal capillary plexus of the inner nuclear layer and progress to involve the superficial retinal capillary plexus during the early nonproliferative stages.4 Clinically, the disease is characterized by a change in retinal transparency to a grayish coloration in the juxtafoveal region, the deposition of superficial retinal crystals, and the formation of right-angled retinal vessels.5-9 The proliferative stages begin when neovascularization arises from the retinal vasculature and extends under the retina or forms retinal-retinal anastomoses, both of which eventually lead to disciform scarring.4 In these later stages, patients can present with depigmentation of the retinal pigment epithelium (RPE), RPE atrophy, and foveal thinning.5-9 Visual function impairments include metamorphopsia and parafoveal or central scotomas with associated visual function deficits.10

In the early stages of the disease, fluorescein angiography (FA) imaging shows abnormal hyperfluorescence and leakage from the temporal, juxtafoveal capillary plexus.7 As the disease progresses, the hyperfluorescence and leakage spreads circumferentially around the fovea. While FA provides a definitive...
diagnosis of MacTel2, it also involves the intravenous injection of a dye that can result in adverse effects such as nausea or vomiting, and rarely fluorescein can elicit an anaphylactic response.\textsuperscript{11,12} Autofluorescence (AF) imaging is also useful in diagnosing MacTel2.\textsuperscript{13} Due to the depletion of luteal pigment in the temporal juxtafoveal retina, a relative increase in AF is observed in this region.\textsuperscript{14-16} As the disease progresses, luteal pigment is lost circumferentially around the fovea and an increase in the relative hyperfluorescence is observed. In the later stages of the disease, atrophy of the RPE is observed, resulting in decreased AF within the central macula.\textsuperscript{17}

Optical coherence tomography (OCT), a noninvasive imaging modality, has revealed structural abnormalities in the inner retina such as retinal cavitation with draping of the internal limiting membrane and abnormalities in the outer retina such as disruption of the photoreceptor inner segment/outer segment/ellipsoid (IS/OS/E) region that were not previously appreciated by FA or AF imaging.\textsuperscript{3,18-25} OCT imaging has improved the early detection of MacTel2 by identifying these early subtle changes in retinal anatomy, and OCT has proven to be useful for following these alterations in macular anatomy as the disease progresses to foveal atrophy, the formation of intraretinal pigment plaques, and subretinal neovascularization.\textsuperscript{1} However, up until recently, OCT has been unable to provide information on the functional microvasculature in the central macula or about blood flow.

With the development of spectral-domain OCT (SD-OCT) instruments with increased scanning speeds and high-speed swept-source OCT (SS-OCT) instruments, OCT microangiography (OMAG) imaging has emerged as a noninvasive strategy to visual-
OMAG is a dynamic strategy capable of providing a three-dimensional reconstruction of the perfused microvasculature within the retina and choroid and identifying distinct characteristics of the capillary networks located within different layers of the retina and choroid (see “Swept-Source OCT Angiography of the Retinal Vasculature Using Intensity Differentiation-based Optical Microangiography Algorithms” in this issue). OMAG acquires images by detecting motion of scattering particles such as erythrocytes within sequential OCT cross-sectional scans performed repeatedly at the same location of the retina. The temporal changes in the OCT signal caused by erythrocyte motion generate the contrast and allow visualization of the microvasculature in OMAG. This technique to detect blood flow has been demonstrated to be useful in clinical studies involving patients with age-related macular degeneration and glaucoma. When compared with standard FA imaging, OMAG imaging is comparable if not better than FA imaging in visualizing microstructural abnormalities and has several distinct advantages over FA imaging, such as its noninvasiveness, speed, and ability to measure blood flow. While OMAG cannot detect the same type of leakage seen with FA imaging, it is possible to identify microvascular abnormalities in the retina and choroidal microvasculature without the use of an exogenous intravenous dye injection.

Figure 3. OCT microangiography (OMAG) images of a 70-year-old woman with intermediate, nonproliferative MacTel2 (case 2). (A) Horizontal central B-scan with the three layers of retinal segmentation: inner retinal layer from the ganglion cell layer to the inner plexiform layer (GCL+IPL), middle retinal layer from the inner nuclear layer to the outer plexiform layer (INL+OPL), and outer retinal layer from outer nuclear layer to the external limiting membrane (ONL+ELM layer). (B) Horizontal central B-scan shows the microvascular flow in different colors corresponding to the different segmented layers of the retina with areas of disruption of the inner segment/outer segment/ellipsoid region. (C) En face OMAG image from the GCL to the IPL shows irregular vessels and vascular drop-out temporal juxtafoveal region. (D) En face OMAG from INL to OPL shows microvascular abnormalities in the perifoveal region. (E) En face OMAG from ONL to the ELM shows the extension of the abnormal microvasculature to the outer retina.

Figure 4. OCT microangiography (OMAG) and fluorescein angiography (FA) images of a 70-year-old woman with intermediate, nonproliferative MacTel2 (case 2). (A) Early-phase FA image shows temporal juxtafoveal hyperfluorescence. (B) Late-phase FA image shows increased hyperfluorescence and leakage. (C) Magnified early-stage FA image shows a detailed view of the hyperfluorescent area that corresponds to the telangiectatic microvasculature. (D) Composite en face color-coded OMAG image demonstrates abnormal microvasculature in the middle layers (green), corresponding to the telangiectatic vessels seen on FA imaging in the temporal juxtafoveal location.
vascular abnormalities associated with leakage, and the same OCT data can be used to examine cross-sectional and en face OCT intensity B-scans to identify anatomic changes in the retina that are the hallmark of leakage, such as subretinal fluid, cystic maculopathy, and increased retinal thickness. Another strategy developed by Chin et al used a technique known as power-Doppler OCT to visualize blood flow information in cross-sectional B-scans to study eyes with different stages of MacTel2 pathology. However, the study was limited to visualization of flow in B-scans, and direct comparisons with en face FA imaging were not possible. In this study, we investigated the retinal microvasculature in patients with MacTel2 using a Carl Zeiss Meditec (Dublin, CA) 1-µm SS-OCT prototype to acquire the en face OMAG images of three distinct physiological layers in the retina. The OMAG images were then compared with the images obtained using standard FA imaging.

PATIENTS AND METHODS

Patients diagnosed with MacTel2 were enrolled in a prospective, observational study at the Bascom Palmer Eye Institute as part of the MacTel Project. All patients underwent a comprehensive ocular examination and imaging tests as part of the evaluation of their condition. The imaging tests included color fundus imaging (Topcon, Tokyo, Japan), digital fundus AF imaging (Topcon, Tokyo, Japan, and...

Figure 5. OCT microangiography (OMAG) images a 61-year-old woman with intermediate, non-proliferative MacTel2 (case 3). (A) Horizontal central B-scan with the three layers of retinal segmentation: inner retinal layer from the ganglion cell layer to the inner plexiform layer (GCL+IPL), middle retinal layer from the inner nuclear layer to the outer plexiform layer (INL+OPL), and outer retinal layer from outer nuclear layer to the external limiting membrane (ONL+ELM layer). (B) Horizontal central B-scan shows the microvascular flow in different colors corresponding to the different segmented layers of the retina with areas of disruption of the inner segment/outer segment/ellipsoid region. The microvascular flow is considerably more prominent in the temporal region. (C) En face OMAG image from the GCL to the IPL showing microvascular abnormalities in the juxtafoveal region. (D) En face OMAG from the INL to the OPL shows the telangiectatic and dilated vessels in the middle retinal layers. (D) En face OMAG image from the ONL to the ELM shows subtle microvascular alterations in the outer retinal layers.

Figure 6. OCT microangiography (OMAG) and fluorescein angiography (FA) images of a 61-year-old woman with intermediate, non-proliferative MacTel2 (case 3). (A) Early-phase FA image showing hyperfluorescence in the temporal juxtafoveal region. (B) Late-phase FA image with increased and diffuse hyperfluorescence and leakage. (C) Magnified early-stage FA image shows a magnified view of the hyperfluorescent area temporal to the fovea with abnormal telangiectatic microvasculature. (D) Composite en face color-coded OMAG image shows the abnormal microvasculature in the middle retinal layers (green) correspondent to the area of leakage seen with FA imaging.
Heidelberg Engineering, Heidelberg, Germany), FA imaging (Heidelberg Engineering, Heidelberg, Germany), and SD-OCT imaging (Cirrus; Carl Zeiss Meditec, Dublin, CA). In addition, selected patients underwent imaging using a modified Cirrus prototype containing a swept-source laser provided by Carl Zeiss Meditec with a central wavelength of 1,050 nm (1,000-1,100 nm full width) and a speed of 100,000 A-scans per second. The institutional review board of the University of Miami Miller School of Medicine approved the studies, and informed consent to participate in both the MacTel Project and the prospective OCT studies was obtained from all participants. The studies were performed in accordance with the tenets of the Declaration of Helsinki and in compliance with the Health Insurance Portability and Accountability Act of 1996.

Inclusion in the MacTel2 study required that patients be diagnosed with MacTel2 in at least one eye, confirmed by the study’s designated reading center. Patients were excluded from the OCT imaging study if they had any other confounding retinal pathology such as diabetic retinopathy or pathologic myopia and if they had been previously treated with photodynamic therapy (PDT), thermal laser, intravitreal injections, or any retinal surgery. Information about previous medical conditions and ocular treatments was obtained by reviewing the medical charts.

Study participants underwent SD-OCT imaging following a protocol that included both the 200 × 200 and the 512 × 128 macular raster scan patterns. In addition, each eye was scanned using the Zeiss 1-µm SS-OCT prototype following a protocol that included a 512 × 512 macular scan (12 mm × 9 mm), a 1,024 × 1,024 macular raster scan (12 mm × 9 mm), a single high-density B-scan comprised of 1,024 A-scans (12 mm long), and the OMAG scans. The OMAG scans measured 3 mm × 3 mm on the retina and were centered on the fovea. In the transverse scanning direction, a single B-scan was comprised of 300 A-scans. Four consecutive B-scans were performed at each fixed location before proceeding to the next transverse location on the retina. A total of 300 B-scan locations located 10 µm apart over a 3-mm distance were sampled. The time difference between two successive B-scans was roughly 3.8 ms, which corresponds to a B-scan acquisition rate of 263 B-scans per second. Hence, the OCT data is effectively acquired at a duty cycle of approximately 80% for this scan protocol, because it takes approximately 0.8 ms for the scanner to come back to the starting position after completion of each B-scan.

Based on this scan protocol and system speed, the total time for a single volume acquisition was about 4.5 s, excluding the adjustment time before the data collection. The OMAG algorithm was applied to the volumetric data set, and images were extracted. These images represented the microvasculature within the scanned retinal tissue. The averaged OCT B-scans were also examined in the usual manner of OCT data to show the retinal tissue in cross-section as well as en face.

The images of microvascular networks shown here were obtained by using the previously published OMAG algorithms, and in all cases, an intensity
differentiation algorithm was applied to extract in vivo blood flow information, as described in a companion paper (see “Swept-Source OCT Angiography of the Retinal Vasculature Using Intensity Differentiation-based Optical Microangiography Algorithms” in this issue). Before the algorithm was applied, the displacements occurring between adjacent repeated B-scans caused by involuntary movement of the human eye were compensated by using a two-dimensional cross-correlation between two adjacent OMAG flow images. A semi-automated retinal layer segmentation algorithm developed by our research group at the University of Washington was used to segment the retina into three distinct physiological layers: an inner retinal layer from the ganglion cell layer to the inner plexiform layer (GCL + IPL), a middle retinal layer from the inner nuclear layer to the outer plexiform layer (INL + OPL), and an outer retinal layer from outer nuclear layer to the external limiting membrane (ONL + ELM layer). The microvasculature from the superficial capillary plexus in the inner retina is colored red, the microvasculature from the deep capillary plexus is colored green, and any microvascular structures with flow in the outer retina are colored blue. This segmentation was performed using the OCT cross-sectional structural images based on the intensity differences of the retinal layers, and the segmentation was applied to the entire three-dimensional data set. The three-dimensional structure of the retina and microvasculature were rendered and projected using Amira three-dimensional software (FEI Visualization Sciences Group, Mérygnac Cedex, France). The segmentation allowed for the visualization of the microvasculature in different layers of the retina, and the en face images were created by using a maximum projection method within the layer of interest. The qualitative OMAG en face images were compared with early and late-phase FA images. Abnormalities in OMAG images were identified based on the location, shape, size, and distribution of the microvasculature. There was no attempt to compare differences in blood flow velocity between normal eyes and eyes with MacTel2.

RESULTS

A total of 41 eyes of 22 patients with MacTel2 at different stages of disease were imaged using the Zeiss prototype 1-µm SS-OCT instrument. There was a predominance of women in the study (19 of 22; 86%). The mean age of patients was 62.8 years (range: 44 to 75 years). Six cases with MacTel2 at different stages of disease progression with characteristic findings are presented in this paper. Each case is presented with a representative OCT B-scan showing the segmented retinal layers with and without color coding of the microvasculature and with en face OMAG images showing the microvasculature along with corresponding early and late FA images.

Case 1: Early, Nonproliferative MacTel2

A 55-year-old woman noticed decreased visual acuity for 1 year in both eyes. Best corrected visual acuity (BCVA) in her left eye was 20/25. The horizontal B-scan with the retinal flow in different layers represented by colors shows the dilated vessels in the deep retinal capillary plexus found in the middle retinal layer, most pronounced in the region temporal to the fovea as observed in green (Figure 1B). The OCT B-scan and flow image also show these dilated vessels are associated with disruption of the IS/OS/E. The two-dimensional OMAG en face images demonstrate no visible alteration in the microvasculature of
the inner retina (Figure 1C) and minimal abnormalities in the middle retinal layer (Figure 1D). The dark image (Figure 1E) represents the absence of a signal, and the appearance is similar to the results observed in normal subjects. The faint microvascular structures seen in (Figure 1E) are artifacts caused by OCT signal decorrelation tails from the microvasculature in the more superficial retinal layers. These artifacts arise when the OCT light beam passes through a dynamic and high scattering region such as a blood vessel resulting in decorrelation of the OCT signal underneath, leading to the appearance of shadow artifacts.

Fluorescein angiography shows telangiectatic abnormalities with mild hyperfluorescence and leakage in the temporal juxtafoveal region (Figure 2A). The late-stage FA image shows increased hyperfluorescence and leakage in this juxtafoveal location (Figure 2B). The color-coded OMAG image illustrates the area corresponding to the subtle telangiectatic alterations, which are located in the middle retinal layer and depicted as the prominent green vessel in the temporal juxtafoveal region (Figure 2D).

**Case 2: Intermediate, Nonproliferative MacTel2**

A 70-year-old woman with BCVA of 20/25 in the left eye presented with decreased vision. The OMAG images (Figures 3B, 3D, and 4D) show dilated, irregular vessels in the temporal re-

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**Figure 9.** OCT microangiography (OMAG) images of a 54-year-old woman with late, proliferative MacTel2 (case 5). (A) Horizontal central B-scan with the three layers of retinal segmentation: inner retinal layer from the ganglion cell layer to the inner plexiform layer (GCL+IPL), middle retinal layer from the inner nuclear layer to the outer plexiform layer (INL+OPL), and outer retinal layer from outer nuclear layer to the external limiting membrane (ONL+ELM layer). (B) Horizontal central B-scan shows the microvascular flow in different colors corresponding to the different segmented layers of the retina. Prominent abnormal flow is observed temporally in the middle and deep retinal layers. Disruption of the inner segment/outer segment/ellipsoid region and retinal cavitation are present. (C) En face OMAG image from the GCL to the IPL shows retinal vascular anastomoses in the temporal juxtafoveal region with distortion of the capillary plexus. (D) En face OMAG image from the INL to the OPL shows the microvascular abnormalities. (E) En face OMAG image from the ONL to the ELM shows evidence of the same microvascular alterations extending from the inner and middle retinal layers.

**Figure 10.** OCT microangiography (OMAG) and fluorescein angiography (FA) images of a 54-year-old woman with late, proliferative MacTel2 (case 5). (A) Early-phase FA showing hyperfluorescence in the temporal juxtafoveal region. (B) Late-phase FA shows leakage in the corresponding area. (C) Magnified early-stage FA image shows a detailed view of the hyperfluorescent area with microvascular abnormalities. (D) Composite en face color-coded OMAG image demonstrates abnormal microvascular flow characteristics that correspond to the abnormal microvascular area with leakage seen on FA imaging.
region, with the abnormal vessels residing within the middle retinal layer (green) and extending to the outer retina in an area corresponding to the disruption of the IS/OS/E region. The microvascular network depicted in the different layers of retina (Figures 3D and 4D) shows an abnormal microvasculature in the temporal juxtafoveal region.

FA imaging revealed telangiectatic capillaries in the temporal juxtafoveal location characterized by hyperfluorescence in the early phase, becoming more diffuse over time, with leakage in the late phase of the angiogram. The presence of leakage prevents an appreciation of the abnormal microvasculature within this area. The color-coded OMAG en face image (Figure 4D) corresponds well with the hyperfluorescent area observed in the early stage of the FA image. These abnormal vessels are green in the OMAG color-coded image and are localized within the middle retinal layer (Figure 4D).

Case 3: Intermediate, Nonproliferative MacTel2

A 61-year-old woman presented for evaluation, with BCVA of 20/25 in the right eye. The OCT B-scan and flow image (Figures 5A-B) show disruption of the IS/OS/E, and the microvascular networks observed in the inner, middle, and outer retinal layers (Figures 5C, 5D, 5E, and 6D) show multiple, telangiectatic, microaneurysmal-like dilated vessels (green/blue) in the temporal juxtafoveal region associated with the disrupted IS/OS/E region.

FA imaging demonstrates hyperfluorescence in the temporal juxtafoveal region where the telangiectatic capillaries are located (Figures 6A and 6C). Leakage from these telangiectatic vessels in the late stage of FA (Figure 6B) correspond to the green-colored abnormal microvasculature in the middle retinal layer temporal to the fovea in the color-coded OMAG image (Figure 6D).

Case 4: Late, Proliferative MacTel2

A 66-year-old woman, followed up for 4 years in the MacTel Project, presented for her annual visit with BCVA of 20/25 in the right eye. The B-scan (Figures 7A-B) shows marked retinal thinning and disruption of the IS/OS/E boundary in the temporal juxtafoveal region. The B-scan representing the microvascular flow (Figure 7B) shows the presence of abnormal vessels (green and blue) corresponding to an area with retinal vascular anastomoses. The abnormal microvasculature extends to the outer retina where the IS/OS/E is disrupted. Microvascular abnormalities, such as a distorted juxtafoveal capillary plexus with prominent anastomoses, are evident in the depth-resolved OMAG en face images of all the segmented layers (Figures 7C-E).

FA imaging demonstrates early hyperfluorescence with late leakage in the temporal juxtafoveal region (Figures 8A-C). The hyperfluorescent microvascular abnormalities seen in the early-stage FA image correspond well with the microvascular abnormalities seen in the color-coded OMAG image (Figures 8C-D). In this patient, the early transit images were obtained...
on the fellow eye, so images with earlier transit times are not available.

**Case 5: Late, Proliferative MacTel2**
A 54-year-old woman presented with BCVA of 20/16 in the left eye. The B-scan (Figures 9A-B) shows cavitation in the outer retina and disruption of the IS/OS/E boundary in the temporal juxtafoveal region. The B-scan representing the microvascular flow (Figure 9B) details the presence of abnormal microvasculature (green and blue) corresponding to an area with retinal vascular anastomoses. Disruption of the microvasculature extends into the outer retina where the IS/OS/E is disrupted. Microvascular abnormalities, such as a distorted juxtafoveal capillary plexus with prominent anastomoses, are evident in the depth-resolved OMAG en face images of all segmented layers (Figures 9C-E).

FA imaging demonstrates hyperfluorescence in the temporal juxtafoveal region in the earliest stage associated with late leakage (Figures 10A-C). The hyperfluorescent microvascular abnormalities seen in the early stage FA image correspond well with the microvascular abnormalities seen in the color-coded OMAG image (Figures 10C-D).

**Case 6: Late, Proliferative MacTel2**
A 51-year-old woman presented with BCVA of 20/640 in the left eye and complained of worsening vision. The B-scan shows a thickened retina with a subretinal hyperreflective plaque in the temporal juxtafoveal location (Figures 11A-B). Vascular flow was detected within this hyperreflective plaque (blue), and the OMAG en face images show abnormal microvasculature in the superior half of the juxtafoveal region, with a more pronounced alteration in the size and disposition of the vessels in the temporal juxtafoveal location (Figures 11C-E).

Early fluorescein angiographic imaging demonstrates an intense hyperfluorescence in the juxtafoveal region representing an extensive subretinal neovascular complex involving the outer retina as well as anastomotic vessels located temporally (Figures 12A-C). The angiographic early hyperfluorescence and late leakage (Figures 12A-B) correspond well with the color-coded en face OMAG image representing the abnormal retinal microvasculature in red and green and the subretinal neovascularization in blue (Figure 12D).

**DISCUSSION**
In this study, we used the OMAG technique to investigate eyes with MacTel2 using a Zeiss 1-µm SS-OCT prototype system with a scanning speed of
To extract the blood flow information and visualize the microvasculature of the central macula, we employed an OMAG algorithm based on an intensity differentiation method to provide precise qualitative vascular details while effectively suppressing the noise. There was obvious agreement between the OMAG en face images and the FA images obtained using a Heidelberg Spectralis instrument. Overall, the central macular microvasculature was visualized better using OMAG than with FA imaging. One explanation could be the fact that the OMAG scanning protocol uses a very high sampling density of the retina, and coupled with the high confocality provided by the single-mode fiber-based collection of the backscattered light, OMAG imaging results in significant enhancement of the signal from the retinal microvasculature in the eye compared with FA imaging. In addition, the better visualization of the juxtafoveal microvasculature with OMAG may also be due, in part, to the absence of leakage on OMAG imaging, and it is this leakage that could obscure the normal vasculature seen on routine FA imaging. Luteal pigment also attenuates the excitation of fluorescein in the central macular region, which could obscure the microvasculature, but this luteal pigment would have no effect on OMAG imaging that typically uses illumination light in the near infrared wavelength region. However, this only explains part of the OMAG advantage, because OMAG imaging was routinely better at identifying the microvascular networks within the central macula even when images were compared outside the area where the luteal pigment resides.

While OMAG provides detailed structural blood flow information, albeit from a small, 3 mm × 3 mm area, it cannot identify leakage from the retinal microvasculature. This inability of OMAG to identify leakage may be fully compensated by the unique ability of OCT to provide information about abnormal vascular perfusion and correlate these areas with structural changes seen on registered B-scans. For example, in eyes with MacTel2, the B-scan images alone are often sufficient to make the diagnosis by identifying the characteristic structural abnormalities in the retinal layers. With the introduction of OMAG imaging, we are now able to identify abnormal microvasculature in the perifoveal region, and we can combine the abnormal B-scan findings with the abnormal microvasculature findings to help confirm the diagnosis. In addition, by using OMAG imaging and taking the maximum projection of the inner, middle, and outer retinal layers and adjusting the color and contrast to highlight the vasculature from these layers, it is easier to identify the abnormal vasculature, where it is located, and qualitatively correlate this abnormal microvasculature seen on the en face OMAG images with the leakage seen on FA. It appears as though the dilated, truncated abnormal vessels seen in green and blue from the middle and outer retina are associated with leakage on FA. A definitive statement about whether these abnormal vessels correlate with the leakage seen on FA images will require a more quantitative approach. However, we are encouraged by the apparent qualitative association between the abnormalities observed in the perifoveal microvasculature and leakage seen on FA imaging in eyes with MacTel2.

In other retinal diseases in which leakage seen on FA correlates with an increased retinal thickness on OCT imaging, such as in macular edema from diabetes and vein occlusions, the ability to correlate abnormal vasculature seen on OMAG imaging with an abnormal OCT B-scan should adequately replace the

**Figure 12.** OCT microangiography (OMAG) and fluorescein angiography (FA) images of a 51-year-old woman with late, proliferative MacTel2 (case 6). (A) Early-phase FA image shows extensive hyperfluorescence. (B) Late-phase FA image shows extensive leakage. (C) Magnified early stage FA image showing a detailed view of the hyperfluorescent area corresponding to subretinal neovascularization and temporal juxtafoveal telangiectatic and anastomotic vessels. (D) Composite en face color-coded OMAG image demonstrates abnormal microvascular flow in the outer retina (blue) that corresponds to the subretinal neovascularization observed in the FA images.
need for information about leakage obtained from FA imaging alone. In addition to being a noninvasive imaging strategy, OMAG has advantages over FA in that the image quality is less affected by the presence of cataract and OMAG is able to segment the retinal layers in three dimensions. By using this ability to extract and visualize these retinal layers in MacTel2 and other diseases, OMAG imaging may help facilitate the early diagnosis of disease and provide a better understanding of disease progression and the efficacy of treatments. The greatest disadvantages of OMAG compared with FA imaging include the small imaging area and the motion artifacts that are introduced with subtle eye movements. However, as imaging speeds improve and eye tracking is introduced, larger scan areas will become common and motion artifacts will become less of a problem.

In conclusion, OMAG is a noninvasive imaging strategy that holds great promise for the evaluation of eyes with MacTel2 and other diseases affecting the retinal microvasculature associated with fluorescein angiographic leakage, such as neovascular age-related macular degeneration, diabetic macular edema, vein occlusions, and cystoid macular edema from differing conditions. In addition, OMAG should be useful in investigating the pathophysiology of angiographically silent cystoid macular edema, such as the microvascular changes associated with the taxane class of medications (paclitaxel and docetaxel), vitreomacular traction, epiretinal membranes, niacin maculopathy, juvenile retinoschisis, retinitis pigmentosa, and Goldman-Favre disease. However, different patterns of retinal segmentation may be needed to highlight the underlying microangiopathy using OMAG, and all the different segmentations are derived from one three-dimensional data set, which takes about 4.5 seconds to acquire and has none of the potential adverse events associated with FA. This study demonstrated the ability of OMAG to image the perifoveal microvasculature in eyes with different stages of MacTel2, but further studies are needed to quantitate vascular caliber, density, and blood flow in the central macula and identify whether changes in these parameters predict disease progression or response to future therapeutic interventions.

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