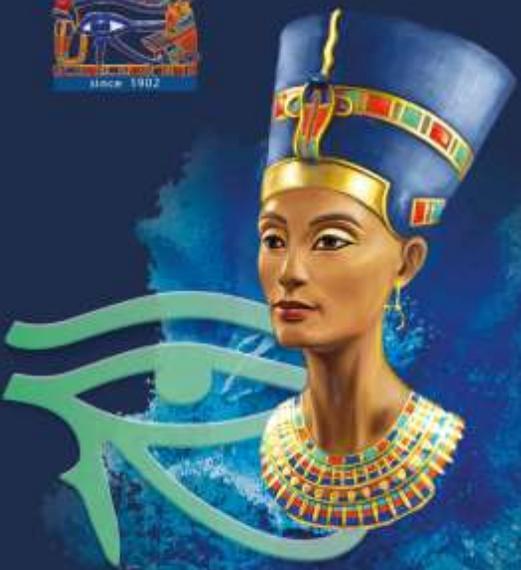


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Morphological and vascular density changes using OCT-A in Patients with macular Telangiectasia type 2

Dr. Shaymaa Hassan Salah;
Assistant Professor Cairo University

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Morphological and vascular density changes using OCT-A in Patients with macular Telangiectasia type 2

- **Many thanks** go to co-authors of the study:
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 - 2- **Hany Hamza**, MD. Ophthalmology department, Faculty of Medicine, Cairo University, Cairo, Egypt
 - 3- **Ihab Abdelaziz Mohamed**, MD. Ophthalmology department, Memorial institute of ophthalmology, Cairo, Egypt
 - 4- **Rania G. Estawro**, FRCS. Watany Eye Hospital

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Disclosure



- All authors declare that they have no financial or other conflicts of interest.



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Mac Tel-2

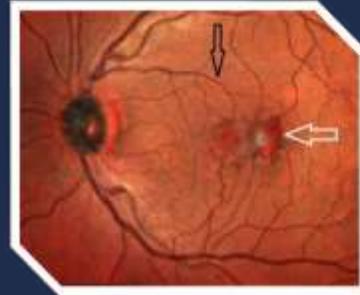
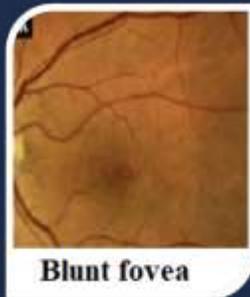
- Current study was a prospective cross-sectional analytical study that was done on fifteen patients (24 eyes) diagnosed with MacTel-2 in at least one eye and thirteen healthy controls (24 eyes) from December 2019 to December 2021.
- We conducted this study after Kasr AlAiny ethics committee approval (MD-221-2019)



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Mac Tel-2

- Eyes with MacTel-2 included in this study were selected when any clinical sign of MacTel-2 as loss of retinal transparency, retinal crystalline deposits, right-angled retinal venules, pigment clumps and/or subretinal neovessels.



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Mac Tel-2

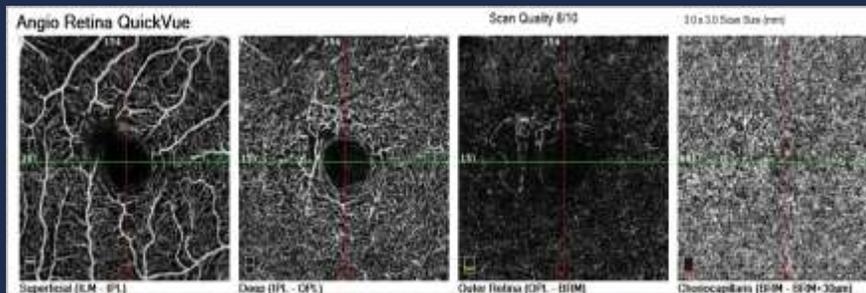
- All patients and controls underwent macular 3x3 mm OCT and OCT-A using Optovue RTVue XR Avanti (Optovue, Inc, Fremont, CA) within one week of clinical diagnosis.
- Macular 3x3-mm en face and structural OCTA images centered on the fovea were obtained with a split-spectrum amplitude-decorrelation angiography algorithm.
- Both the en face and the B-scan data were only saved and analyzed after projection artifact removal and when the signal strength is better than 7/10.

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Mac Tel-2

- The software automatically segmented the full-thickness retinal scans into **four** slabs: the inner and middle slabs containing the superficial and deep inner retinal vascular plexuses respectively, outer retina and choriocapillaris slabs that should be avascular, were useful in identifying outer and sub-retinal neo-vascular proliferations respectively.



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Mac Tel-2- Results

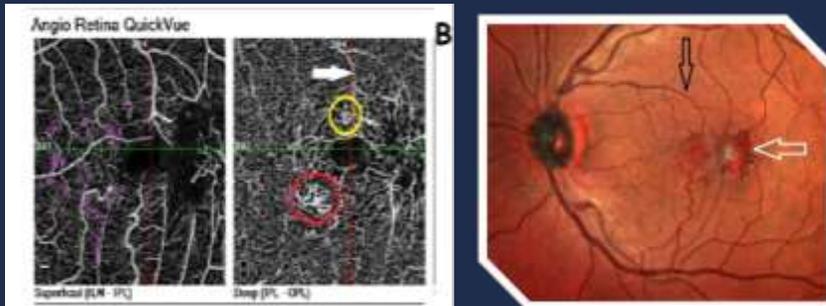
- Our study included 48 eyes divided into two groups, the first group 24 eyes (10 right, 14 left) in patients with MacTel-2 and the second group 24 eyes (13 right, 11 left) of normal age matched control group.
- The mean age in MacTel-2 group was 62.4 ± 11.9 years.
- The mean BCVA (by LogMar) in MacTel-2 group was 0.4 ± 0.12



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Mac Tel-2- Results

- In MacTel-2 eyes, 3x3 mm OCT-A scans showed common morphological features: **Ectatic Capillaries** mainly in DCP, inter-vascular **rarefaction** mainly in SCP and **Right-Angled Vessels (RAVs)**: dilation and apparent discontinuation of blunted vessel caused by a vertical deviation from superficial to deeper retinal layers, figure



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MacTel-2 group (24 eyes) using Toto et al grading system

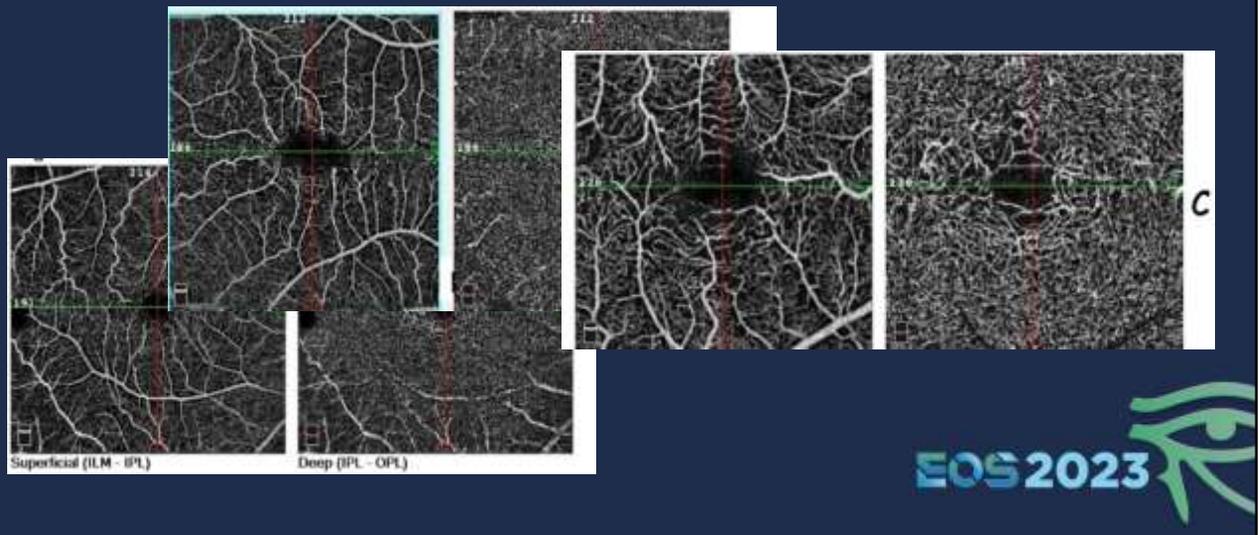
OCT-A classification in MacTel-2 group (24 eyes) using Toto et al grading system

Finding by OCT-A	Grade	Number of eyes (%)	Average BCVA by Log Mar (min-max)
Vascular anomalies in the deep and/or superficial plexus temporal to the fovea	One	8 (33.3)	0.2 (0.8-0.0)
Vascular anomalies in the deep and/or superficial plexus temporal and nasal to the fovea	Two	5 (20.8)	0.5 (0.8-0.2)
Markedly diffuse circumferential vascular anomalies in the deep and superficial plexus	Three	2 (8.3)	0.3 (0.7-0.0)
Neovascularization in the outer retina with any OCTA signs of grade 1 to 3	Four	9 (37.5)	0.8 (1.3-0.7)

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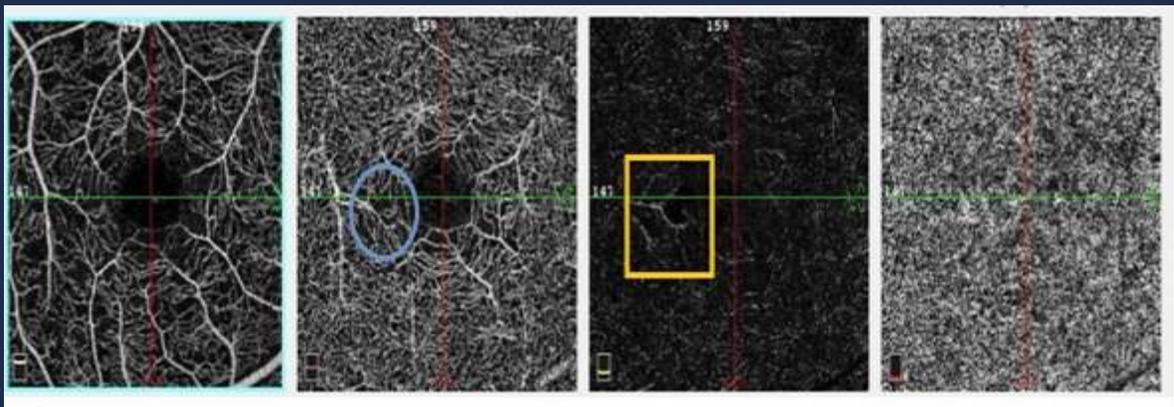
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Toto et al classification



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Mac Tel-2



retino-retinal anastomoses

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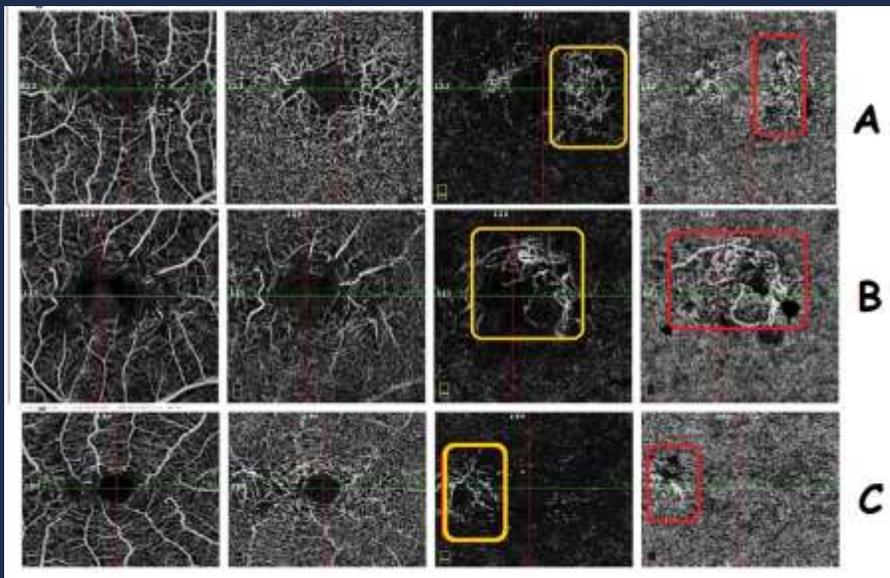
Mac Tel-2-Results

- All the 9 eyes in **stage 4** (100%) showed intraretinal neo-capillaries (neo-capillary at outer retinal slab) while 6 eyes (6/9, 66.6 %) showed both retinal and subretinal neo-capillaries (neo-capillaries at outer retinal and choriocapillaris slabs). The patterns of neo-capillaries were sea-fan, tangled or dead tree network, equally distributed among the nine eyes.
- Three eyes (33.3%) in stage 4 had history of intravitreal injections, those 3 eyes had the dead tree vascular network.



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Mac Tel-2



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Mac Tel-2-Results

- FAZ measurements regarding area, perimeter, acircularity index and density showed no statistical difference between normal and MacTel-2 group.
- In MacTel-2 group, the vessel density in the **SCP** was significantly decreased in the temporal, superior and inferior para-foveal area (p value = 0.004, <0.1001 and 0.001 respectively).
- Only the temporal parafoveal vessel density in the **DCP** showed significant decrease in the Mac Tel group as compared with the normal control group (54.01% versus 49.6%, p value =0.010).
- Regarding retinal full thickness at the 3x3 mm scan, there was statistically significant decrease in the foveal and all the para-foveal thickness



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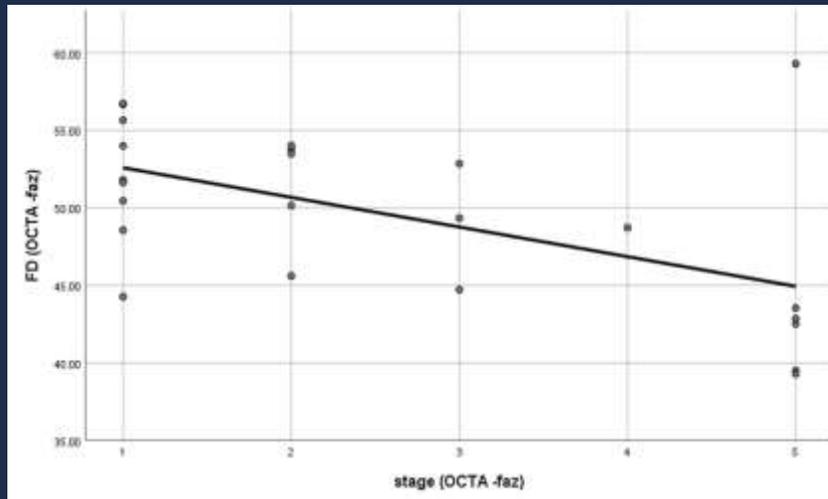
Mac Tel-2

- In MacTel-2 group, comparison between the vessel densities in both SCP and DCP shows significant decrease in the superficial vessel density in the foveal area and all para-foveal areas (p value < 0.001).
- Our study showed decrease in the foveal and para foveal vessel density with progression of MacTel-2 staging but this decrease wasn't statistically significant in neither superficial nor deep plexuses.
- There was increase in macular thickness in MacTel-2 eyes was more in stage 4 than stage 1 (positively correlated with progression of the disease), but again this correlation wasn't statistically significant.



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Mac Tel-2



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Back to literature

- Primary retinal telangiectasia comprises a group of rare, idiopathic, congenital or acquired retinal vascular anomalies characterized by dilatation and tortuosity of macular retinal blood vessels, multiple aneurysms, vascular leakage and the deposition of hard exudates.
- **Gass** in 1982 introduced the old classification based on clinical data: idiopathic juxta foveolar retinal telangiectasia (IJRT) into 4 groups then updated in 1993 to comprise three groups divided each into A and B
- **Yannuzi** Classification: Two groups: Aneurysmal (Mac Tel-1) and perifoveal telangiectasia (Mac Tel-2)



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Mac Tel-2

- Bilateral, between 40-60 years, both sex.
- Theories:
 - **Gass** proposed the telangiectatic vessels had altered structure of the capillary walls that impeded metabolic exchange. The resultant low-grade chronic nutritional damage was posited to cause degeneration and atrophy of not only the Müller cells, but also the associated photoreceptor cells.
 - **Vascular:** Watershed temporal zone → congestion → outer retinal atrophy and RPE hyperplasia.
 - **Genetic:**
 - **Neurodegenerative (Muller cell dysfunction):** Cell deletion leads to telangiectasia and neovascularization.



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Mac Tel-2

- Early manifestations of the disease include loss of retinal transparency, superficial retinal crystalline deposits, right-angled venules, and presence of intraretinal cystoid spaces in the fovea.
- In later stages, the intraretinal pigment migrates, forming pigment plaques, and subretinal neovascular membrane develops.
- **Fundus fluorescein angiography (FFA)** has been considered for many years the gold standard for diagnosis of MacTel type 2. The classic FFA finding is telangiectatic vessels that leak dye in the parafoveal area with fluorescein leakage not related to the cystic spaces



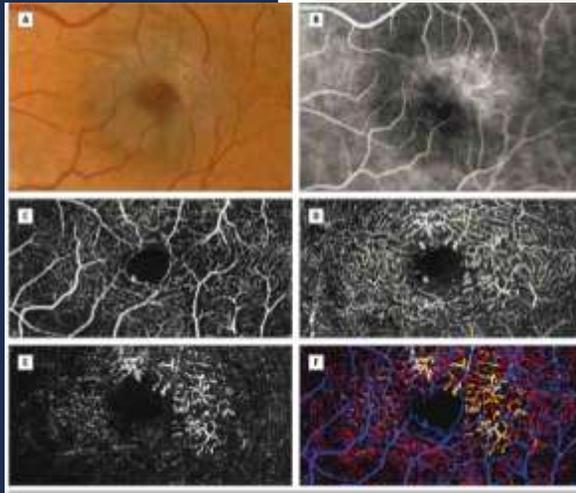
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OCT-A

Original Investigation

Retinal Vascular Layers in Macular Telangiectasia Type 2 Imaged by Optical Coherence Tomographic Angiography

Richard F. Spaide, MD, James M. Klancnik Jr, MD, Michael J. Cooney, MD



A, Color photograph; B, Fluorescein angiogram showing staining; C-F, Optical coherence tomographic angiography of the inner vascular plexus (C), deep plexus (D), outer retina (E), and color stack image (F).

Quality

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OCT-A and Mac Tel-2

- Foveal vessel density and PFVD of the superficial plexus were significantly lower in MacTel 2 group than in the control group
- Foveal vessel density of the deep plexus was significantly lower in MacTel 2 group than in the control group
- Parafoveal vascular density in the deep plexus was not significantly different between the two groups

Special Issue

Multimodal Imaging of Macular Telangiectasia Type 2: Focus on Vascular Changes Using Optical Coherence Tomography Angiography

Lisa Toto,¹ Luca Di Antonio,¹ Rodolfo Mastropasqua,² Peter A. Matti,¹ Paolo Carpineto,¹ Enrico Borrelli,¹ Marco Rispoli,³ Bruno Lambroso,³ and Leonardo Mastropasqua¹

¹Department of Medicine and Science of Aging, Ophthalmology Clinic, University "G. d'Annunzio" Chieti-Pescara, Chieti, Italy
²Ophthalmology Clinic, University of Verona, Verona, Italy
³Italian Macular Center, Rome, Italy

Quantification
and
Qualification

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OCT-A and Mac Tel-2

- capillary proliferation in the outer retina corresponding to areas of EZ loss, which could be a useful marker to monitor the efficacy of possible treatments in MacTel2 disease.

Apart from dilated DCP, additional layer of newly formed capillaries invades ONL. This is not a segmentation or projection artifact but newly formed capillaries not having same pattern as DCP.

ALBERTO M. KRIVOSIC, MD, VALÉRIE KRIVOSIC, MD, RAMIN TADAYONI, MD, PhD

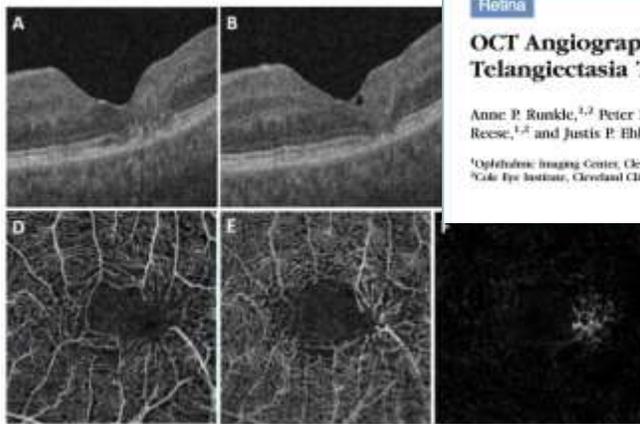
- This usually occurs in front of the area of EZ loss

Methods: Thirty eyes of 15 patients with MacTel2 were examined using optical coherence tomography angiography. Images of the superficial and deep capillary plexus were analyzed and compared with en-face angio-flow images of the outer retina and en-face image of the EZ.
Results: Twenty-one eyes were available for the study, including 12 with invasion of the outer retina capillary on optical coherence tomography angiography but without subretinal neovascularization. The outer retina capillary proliferation had a radial pattern in eight cases, formed loops in four, and was circumscribed to an area of EZ loss on en-face images. In nine cases, there was no outer capillary proliferation including two cases with loss of EZ loss.
Conclusions: Optical coherence tomography angiography shows capillary proliferation in the outer retina corresponding to areas of EZ loss, which could be a useful marker to monitor the efficacy of possible treatments in MacTel2 disease.
 RETINA 35:2300-2306, 2015.



OCT-A and Mac Tel-2

OCT-A and EZ Mapping in DIT



Retina

OCT Angiography and Ellipsoid Zone Mapping of Macular Telangiectasia Type 2 From the AVATAR Study

Anne P. Runkle,^{1,2} Peter K. Kaiser,^{1,2} Sunil K. Srivastava,^{1,2} Andrew P. Schuchat,² Jamie L. Reese,^{1,2} and Justin P. Ehlers,^{1,2}

¹Ophthalmic Imaging Center, Cleveland Clinic, Cleveland, Ohio, United States
²Cole Eye Institute, Cleveland Clinic, Cleveland, Ohio, United States



OCT-A and Mac Tel-2

- RAVs were defined as : in fundoscopy/fundus photography visible, blunted, slightly dilated vessels that are mainly located in the temporal parafovea, and that seem not to narrow towards the foveola, but suddenly dive at a right angle into deeper retinal layers (definition according to the morphological criteria described by Gass and Blodi).
- By means of OCT-A, vessels reflecting the morphological characteristics of RAVs could already be detected in early disease stages (stages 1 to 2) and before they became visible in fundoscopy.



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OCT-A and Mac Tel-2

RETINAL-CHOROIDDAL ANASTOMOSIS IN MACULAR TELANGIECTASIA TYPE 2

RICHARD F. SPAIDE, MD,* LAWRENCE A. YANNUZZI, MD,* PETER M. MALOCCÀ, MD†

The anastomotic vessels are not particularly evident in en face images largely because they were small vessels seen in cross-section. They were seen in both the B-scan cross-sections and volume-rendered images.



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Our study limitations:

- Difficulty in segmentation of retinal layers in diseased eyes especially with dragged vessels.
- Poor fixation increases motion artifacts.
- Highly magnified 3x3 mm images give limited vascular analysis.
- Rarity of diseased eyes justifies the use of chronic, treated eyes.



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Take home message

- **In our study:**
 - Common features in OCT-A: dendritic bulbs of DCP, rarefaction in SCP and RAV.
 - Temporal vascular density is highly reduced
 - Neo-capillaries are 100 % intraretinal due to Retino-retinal anastomoses and 66 % of them have Retino-choroidal anastomoses.
- **Generally:**
 - Optical coherence tomography angiography is useful for retinal vasculature characterization in MacTel type 2 patients.



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Ophthalmology Department - Cairo University

THANKS

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