Review Article

The Use of Swept Source OCT Angio in Diagnosis and Staging of Type 2 Macular Telangiectasia (MacTel 2)

Fawwaz Falih Al-Mamoori*

Medical Retina Department, Eye Specialty Hospital Amman, Jordan

*Corresponding Author: Fawwaz Falih Al-Mamoori, Medical Retina Department, Eye Specialty Hospital Amman, Jordan.

Introduction

Macular telangiectasia type 2 has also been termed idiopathic perifoveal telangiectasia or idiopathic juxta foveal telangiectasia type 2 [1-3]. it is now referred to as MacTel type 2, and it is a bilateral perifoveal vasculopathy which originates in the deep retinal capillary plexus in the temporal juxta foveal region. As it progresses, it involves the superficial retinal capillary plexus, and continues to progress anteriorly, posteriorly, and circumferentially. This is called the non-proliferative stage of the disease [4-9]. MacTel type 2 becomes pro-liferative when the vasogenic process extends under the retina, forming detachment and a retinal–retinal anastomosis. This form of the disease may eventually lead to disciform scarring.

In the early stages of the disease, fluorescein angiography (FA) imaging shows abnormal hyperfluorescence and leakage from the temporal, juxta foveal 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 [11,12]. Auto fluorescence (AF) imaging is also useful in diagnosing MacTel2 [13], Due to the depletion of luteal pigment in the temporal juxta foveal retina, a relative increase in AF is observed in this region [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.

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 pho-toreceptor inner segment/outer segment/ellipsoid (IS/OS/E) region that were not previously appreciated by FA or AF imaging [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 in-traretinal pigment plaques, and subretinal neovascularization. With the development of spectral domain OCT (SDOCT) instruments with increased scanning speeds and high speed swept source OCT (SSOCT) instruments, OCT microangiography (OMAG) imaging has emerged as a noninvasive strategy to visualize the retina and choroidal microvasculature without the use of an exogenous intravenous dye injection [26-38], 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 "SweptSource OCT Angiography of the Retinal Vasculature Using Intensity Differentiation based Optical Microangiography.

Aim and Objectives

To evaluate the central macular microvascular network in patients with macular telangiectasia type 2 (MacTel2) by using Swept Source optical coherence tomography Angiography. That will be useful in diagnosis and staging of the disease.

Patient and Method

We retrospectively review a 60 Y old female patient with bilateral MacTel2 evaluated using a swept source OCT (SSOCT). The patient underwent a comprehensive ocular examination and imaging tests as part of the evaluation of her condition. The imaging tests included color fundus imaging (Topcon, Tokyo, Japan), digital fundus AF imaging, FA & Swept Source OCT with OCT Angio (TRITON TOPCON OCT).

The patient had no any other retinal pathology such as diabetic retinopathy or pathologic myopia and not 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. The retina was segmented 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.

Results

(SS-OCT A) detected abnormal microvasculature in all MacTel2 eyes, predominantly in the middle retinal layer. These vessels correlated well with the FA alterations. The abnormal temporal, juxta foveal microvasculature in MacTel2 became apparent as the disease progressed and in later stages tended to extend circumferentially, with anastomotic vessels temporally.

In our Case, the Right Eye was in Early, Non-proliferative MacTel2, Best corrected visual acuity (BCVA) in her left eye was 20/30. 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 1). With intact IS-OS Junction (Figure2), Fluorescein angiography shows telangiectatic abnormalities with mild hyperfluorescence and leakage in the temporal juxta foveal region (Figure 3).
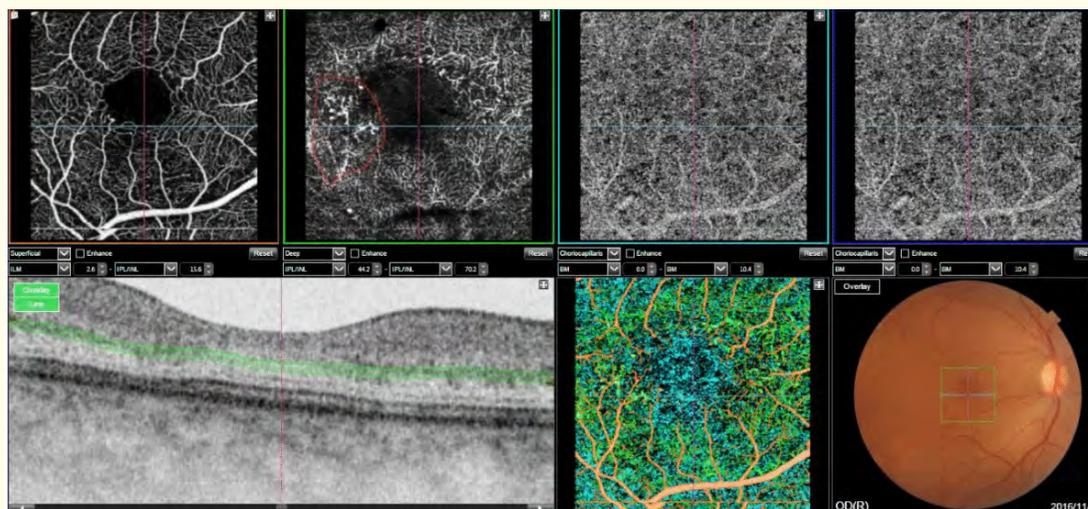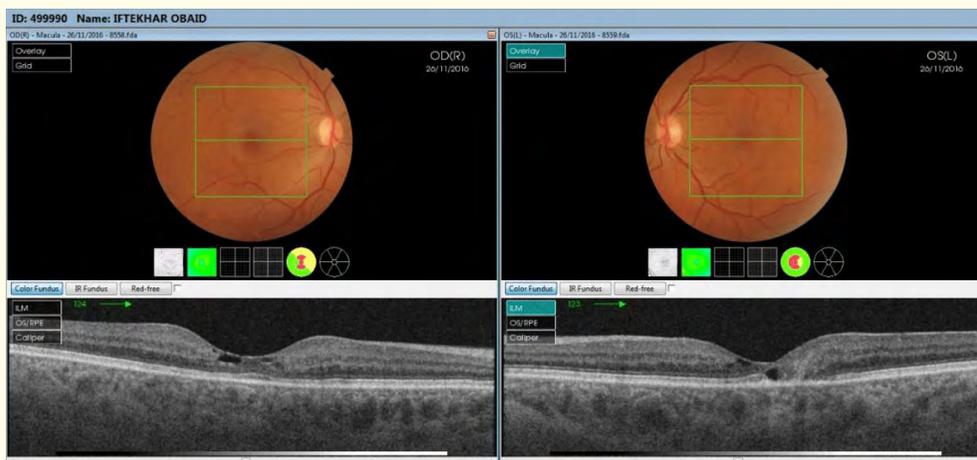

Figure 1

Figure 2

Figure 3

The left eye was in Proliferative MacTel 2 stage, Best corrected visual acuity (BCVA) in her left eye was 20/50, The Bscan shows cavitation in the outer retina and disruption of the IS/OS/E boundary in the temporal juxta foveal region (Figure 2). The Bscan representing the microvascular flow, 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 juxta foveal capillary plexus with prominent anastomoses (Figure 4 A and 4B), FA imaging demonstrates hyperfluorescence in the temporal juxta foveal region in the earliest stage associated with late leakage (Figure 5).
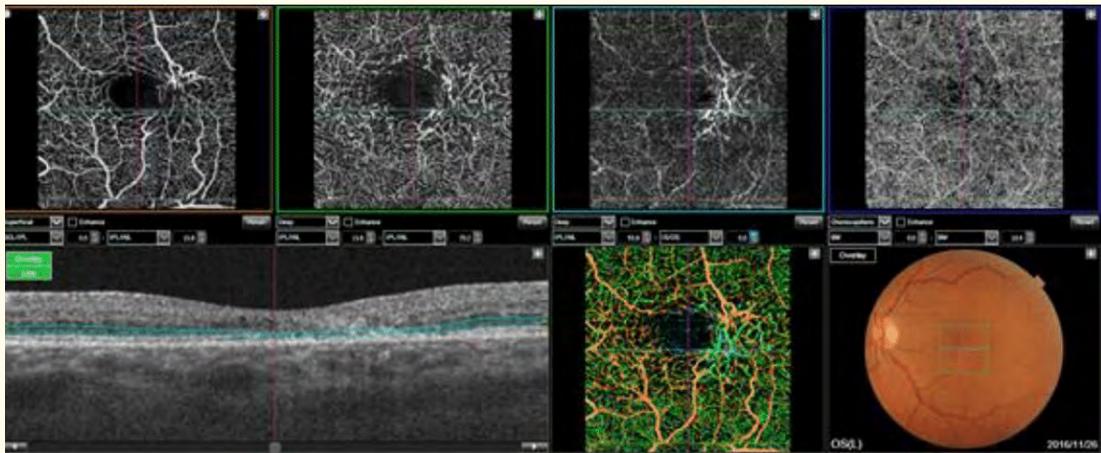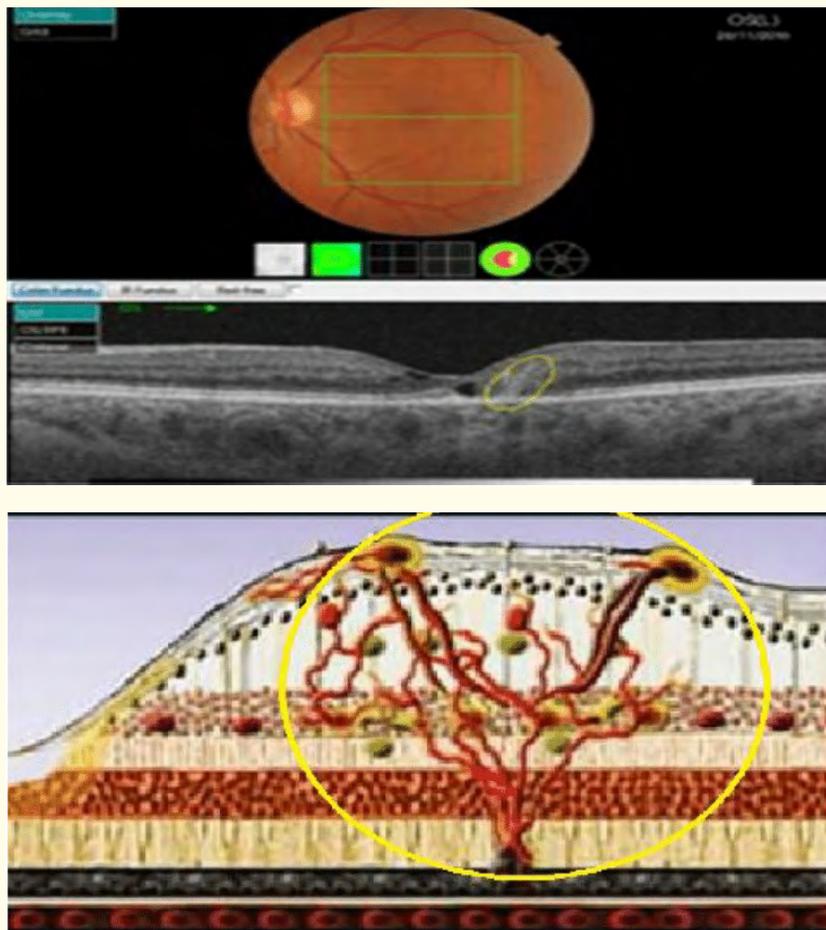
Figure 4A

Figure 4B

Figure 5

Conclusion

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 GoldmanFavre 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.

We used the SS-OCT Angio technique to investigate eyes with MacTel2 using a TRITON SSOCT. To extract the blood flow information and visualize the microvasculature of the central macula, the central macular microvasculature was visualized better than with FA imaging. In addition, the better visualization of the juxta foveal microvasculature with SS-OCT Angio may also be due, in part, to the absence of leakage on OCT Angio imaging, and it is this leakage that could obscure the normal vasculature seen on routine FA imaging. By using this ability to extract and visualize these retinal layers in MacTel2 and other diseases, SS-OCT Angio imaging may help facilitate the early diagnosis of disease and provide a better understanding of disease progression and the efficacy of treatments and to differentiate MacTel2 from 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.

Bibliography

1. Gass JD and Blodi BA. "Idiopathic juxtafoveolar retinal telangiectasis. Update of classification and follow-up study". *Ophthalmology* 100.10 (1993): 1536-1546.

2. Gass JD and Oyakawa RT. "Idiopathic juxtafoveolar retinal telangiectasis". *Archives of Ophthalmology* 100.5 (1982): 769-780.

3. Yannuzzi LA., *et al*. "Idiopathic macular telangiectasia". *Archives of Ophthalmology* 124.4 (2006): 450-460.

4. Yannuzzi LA. "The Retinal Atlas". *Oxford, UK: Saunders Elsevier* (2010).

5. Aung KZ., *et al*. "The prevalence estimates of macular telangiectasia type 2: the Melbourne Collaborative Cohort Study". *Retina* 30.3 (2010): 473-478.

6. Klein R., *et al*. "The prevalence of macular telangiectasia type 2 in the Beaver Dam eye study". *American Journal of Ophthalmology* 150.1 (2010): 55-62e2.

7. Clemons TE., *et al*. "Baseline characteristics of participants in the natural history study of macular telangiectasia (MacTel) MacTel Project Report No. 2". *Ophthalmic Epidemiology* 17.1 (2010): 66-73.

8. Sallo FB., *et al*. "Retinal crystals in type 2 idiopathic macular telangiectasia". *Ophthalmology* 118.12 (2011): 2461-2467.

9. Zhu M., *et al*. "The relationship between inner retinal cavitation, photoreceptor disruption, and the integrity of the outer limiting membrane in macular telangiectasia type 2". *Retina* 33.8 (2013): 1547-1550.

10. SchmitzValckenberg S., *et al*. "Structural and functional changes over time in MacTel patients". *Retina* 29.9 (2009): 1314-1320.

11. LopezSaez MP., *et al*. "Fluoresceininduced allergic reaction". *Annals of Allergy, Asthma, and Immunology* 81.5 (1998): 428-430.

12. Lira RP., *et al*. "Adverse reactions of fluorescein angiography: a prospective study". *Arquivos Brasileiros De Oftalmologia* 70.4 (2007): 615-618.

13. Charbel Issa P., *et al*. "Macular telangiectasia type 2". *Progress in Retinal and Eye Research* 34 (2013): 49-77.

14. Theelen T., *et al*. "Analysis of visual pigment by fundus autofluorescence". *Experimental Eye Research* 86.2 (2008): 296-304.

15. Wong WT., *et al*. "Fundus autofluorescence in type 2 idiopathic macular telangiectasia: correlation with optical coherence tomography and microperimetry". *American Journal of Ophthalmology* 148.4 (2009): 573-583.

16. Ooto S., *et al*. "Highresolution photoreceptor imaging in idiopathic macular telangiectasia type 2 using adaptive optics scanning laser ophthalmoscopy". *Investigative Ophthalmology and Visual Science* 52.8 (2011): 5541-5550.

17. Balaskas K., *et al*. "Associations between a autofluorescence abnormalities and visual acuity in idiopathic macular telangiectasia type 2: MacTel Project report number 5". *Retina* 34.8 (2014): 1630-1636.

18. Mathew R., *et al*. "Agreement between timedomain and spectraldomain optical coherence tomography in the assessment of macular thickness in patients with idiopathic macular telangiectasia type 2". *Ophthalmologica* 230.3 (2013): 144-150.

19. Surguch V., *et al*. "Optical coherence tomography findings in idiopathic juxtafoveal retinal telangiectasis". *Graefe's Archive for Clinical and Experimental* 245.6 (2007): 783-788.

20. Gaudric A., *et al*. "Optical coherence tomography in group 2A idiopathic juxtafoveolar retinal telangiectasis". *Archives of Ophthalmology* 124.10 (2006): 1410-1419.

21. Paunescu LA., *et al*. "Idiopathic juxtafoveal retinal telangiectasis: new findings by ultrahighresolution optical coherence tomography". *Ophthalmology* 113.1 (2006): 48-57.

22. Koizumi H., *et al*. "Morphologic features of group 2A idiopathic juxtafoveolar retinal telangiectasis in threedimensional optical coherence tomography". *American Journal of Ophthalmology* 142.2 (2006): 340-343.

23. Baumuller S., *et al*. "Outer retinal hyperreflective spots on spectraldomain optical coherence tomography in macular telangiectasia type 2". *Ophthalmology* 117.11 (2010): 2162-2168.

24. Sallo FB., *et al*. "The IS/OS junction layer in the natural history of type 2 idiopathic macular telangiectasia". *Investigative Ophthalmology and Visual Science* 53.12 (2012): 7889-7895.

25. Sallo FB., *et al*. "En face" OCT imaging of the IS/OS junction line in type 2 idiopathic macular telangiectasia". *Investigative Ophthalmology and Visual Science* 53.10 (2012): 6145-6152.

26. Wang RK., *et al*. "Depthresolved imaging of capillary networks in retina and choroid using ultrahigh sensitive optical microangiography". *Optics Letters* 35.9 (2010): 1467-1469.

27. An L., *et al*. "Optical microangiography provides correlation between microstructure and microvasculature of optic nerve head in human subjects". *Journal of Biomedical Optics* 17.11 (2012): 116018.

28. Wang RK. "Optical Microangiography: A label free 3d imaging technology to visualize and quantify blood circulations within tissue beds in vivo". *IEEE Journal of Selected Topics in Quantum Electronics* 16.3 (2010): 545-554.

29. Wang RK., *et al*. "Three-dimensional optical angiography". *Optics Express* 15.7 (2007): 4083-4097.

30. Choi W., *et al*. "Choriocapillaris and choroidal microvasculature imaging with ultrahigh speed OCT angiography". *PLoS One* 8.12 (2013): e81499.

31. Jia Y., *et al*. "Quantitative optical coherence tomography angiography of choroidal neovascularization in agerelated macular degeneration". *Ophthalmology* 121.7 (2014): 1435-1444.

32. Wei E., *et al*. "Parafoveal retinal vascular response to pattern visual stimulation assessed with OCT angiography". *PLoS One* 8.12 (2013): e81343.

33. Braaf B., *et al*. "Angiography of the retina and the choroid with phaseresolved OCT using intervaloptimized backstitched Bscans". *Optics Express* 20.18 (2012): 20516-20534.

34. Schwartz DM., *et al*. "Phasevariance optical coherence tomography: a technique for noninvasive angiography". *Ophthalmology* 121.1 (2014): 180-187.

35. Kim DY., *et al*. "Optical imaging of the chorioretinal vasculature in the living human eye". *Proceedings of the National Academy of Sciences of the United States of America* 110.35 (2013): 14354-14359.

36. Motaghiannezam SM., *et al*. "Differential phasecontrast, sweptsource optical coherence tomography at 1060 nm for in vivo human retinal and choroidal vasculature visualization". *Journal of Biomedical Optics* 17.2 (2012):026011.

37. Kim DY., *et al*. "Noninvasive imaging of the foveal avascular with high-speed, phase-variance optical coherence tomography". *Investigative Ophthalmology and Visual Science* 53.1 (2012): 85-92.

38. Thorell MR., *et al*. "Swept-source OCT angiography of macular telangiectasia type 2". *Ophthalmic Surgery, Lasers and Imaging Retina* 45.5 (2014): 369-380.