



# The Use of SWEPT SOURCE OCT ANGIO in Diagnosis and Staging of Type 2 Macular Telangiectasia (Mactel 2)

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## Abstract

**Introduction:** Macular telangiectasia type 2 has also been termed idiopathic perifoveal telangiectasia or idiopathic juxtafoveal telangiectasia type 2 [1-4]. 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 juxtafoveal 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 [5-9]. Mactel type 2 becomes proliferative 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.

**Aim & objectives:** Diagnosis and staging of macular telangiectasia type 2 (MacTel2) using Swept Source optical coherence tomography Angiography.

**Method:** We retrospectively review a 60 Y old 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.

**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, juxtafoveal microvasculature in MacTel2 became apparent as the disease progressed and in later stages tended to extend circumferentially, with anastomotic vessels temporally.

**Conclusion:** 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 juxtafoveal 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.

**Keywords:** Macular telangiectasia Type 2; Swept source oct angiography; Fluorescein angiography; Perifoveal vasculopathy

**Abbreviations:** MacTel 2: Macular Telangiectasia type 2; OCT: Optical Coherence Tomography; SS-OCT Angio: Swept Source OCT Angiography; SD-OCT: Spectral Domain OCT; FA: Fluorescein Angiography; FAF: Fundus Auto Florescence; IS-OS-E: photoreceptor inner segment/outer segment/ellipsoid; GCL+IPL: Ganglion Cell Layer Inner Plexiform Layer; INL + OPL: Inner nuclear layer-Outer Plexiform Layer; ONL + ELM: Outer Nuclear Layer+ External Limiting Membrane; BCVA: Best Corrected Visual Acuity

## Introduction

Macular telangiectasia type 2 has also been termed idiopathic perifoveal telangiectasia or idiopathic juxtafoveal 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 juxtafoveal 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 proliferative 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, juxtafoveal capillary plexus, [7] As the disease progresses, the hyperfluorescence and leakage spreads circumferentially around the fovea. While FA provides a definitive diagnosis of MacTel 2, [10] 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]. Autofluorescence (AF) imaging is also useful in diagnosing MacTel2, [13] Due to the depletion of luteal pigment in the temporal juxtafoveal 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), [17] 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 [18-25]. OCT imaging has improved the early detection of MacTel 2 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. 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 "Swept-Source OCT Angiography of the Retinal Vasculature Using Intensity Differentiation-based Optical Microangiography).

**Aim & Objectives**

To evaluate the central macular microvascular network in patients with maculartelangiectasia type 2 (MacTel2) using Swept Source optical coherence tomography Angiography.

**Patient and Method**

We retrospectively review a 60 Y old 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, juxtafoveal 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 (Figure 2), Fluorescein angiography shows telangiectatic abnormalities with mild hyperfluorescence and leakage in the temporal juxtafoveal region (Figure 3).

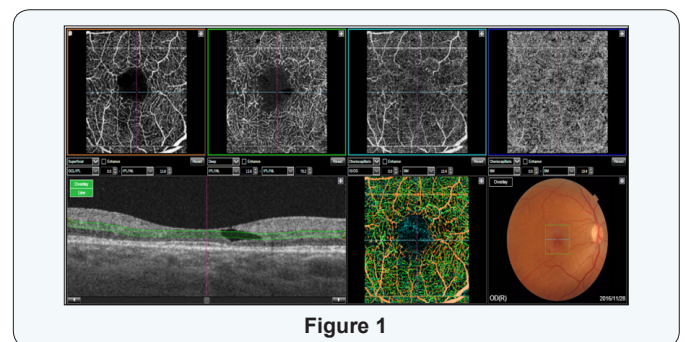


Figure 1

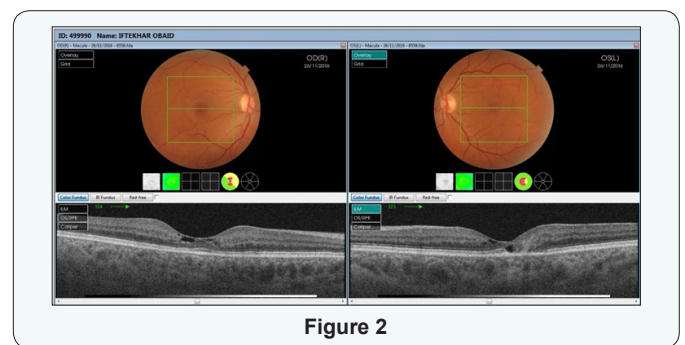


Figure 2

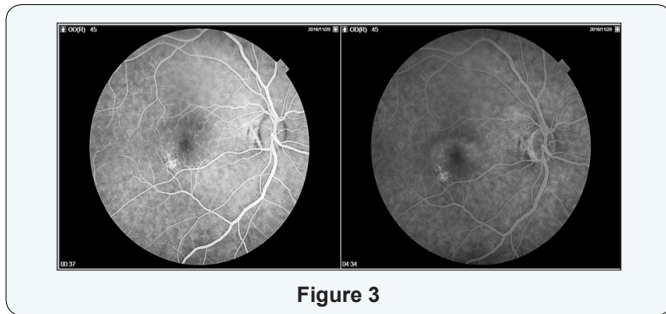


Figure 3

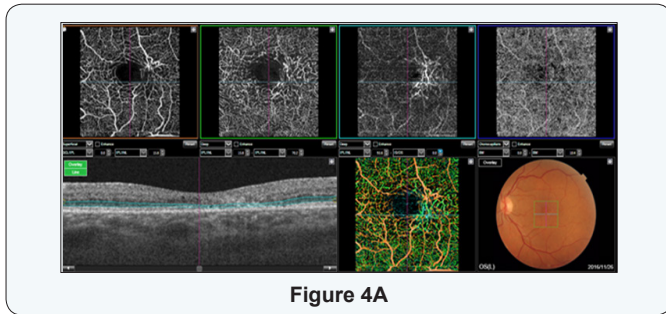


Figure 4A

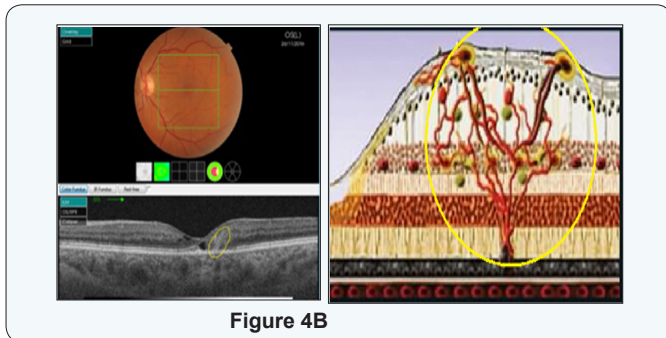


Figure 4B

The left eye was in Proliferative MacTel 2, 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 juxtafoveal region. The Bscan representing the microvascular flow (Figure 4 A&B) 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, FA imaging demonstrates hyperfluorescence in the temporal juxtafoveal region in the earliest stage associated with late leakage (Figure 5)

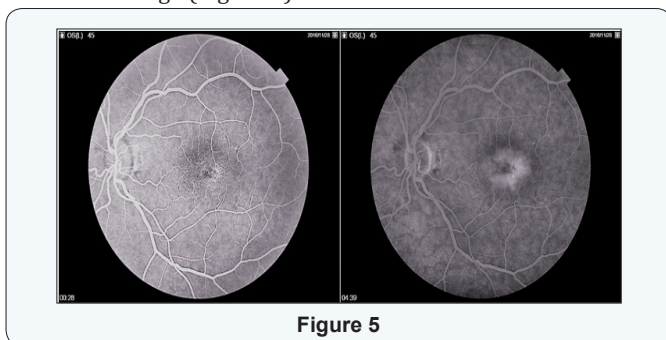


Figure 5

## Conclusion

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 juxtafoveal 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 neovascularage-related macular degeneration, diabetic macular edema, vein occlusions, and cystoid macular edema from differing conditions.

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