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# Macular Telangiectasia

## Introduction

Macular Telangiectasia (MacTel) is a distinct entity from the much more common condition of age-related macular degeneration (ARMD). Recognition of MacTel is important as the condition behaves differently and usually does not require aggressive treatment like neovascular ARMD except when choroidal neovascularization develops as a complication of MacTel and then also requires antiVEGF therapy.

There are two forms of MacTel: Type 1 Macular Telangiectasia (Type 1 MacTel) and Type 2 Macular Telangiectasia (Type 2 MacTel). Type 1 MacTel has more aneurysmal telangiectasias whereas Type 2 MacTel has more ectatic capillaries (without visible aneurysms) causing leakage and edema of the macula. Both have a predilection for the temporal side of the macula. Type 1 MacTel is usually diagnosed before the age of 40 and thought to be a variant of Coats disease and is similarly mostly unilateral and affects more men than women. Type 2 MacTel is most commonly diagnosed in the 4th and 5th decades and is associated with diabetes and hypertension. Type 2 MacTel is mostly a bilateral condition, but it can manifest differently in each eye. There appears to be a genetic predisposition, but there is no known cause of the disease.

Early on, there can be little to no impact of MacTel on visual acuity and function. However, over years, blurred vision and distortion can develop with variable severity. Subretinal neovascularization may develop in 9-12% of patients with Type 2 MacTel<sup>1</sup> and can lead to rapid worsening of the leakage. Eventually there can be scarring and atrophy which will then lead to permanent vision loss. Additionally, patients may develop complications of lamellar macular holes or full-thickness macular holes.

## Examination and Testing

In the early stages of the disease, there are often limited changes affecting the foveal contour. Later, in about 46% of patients, crystalline deposits may form along the internal limiting membrane, which might be identified on optical coherence tomography (OCT) as hyperreflective dots on the surface of the retina. Slightly dilated venules may be noted as they course toward the fovea without narrowing and then suddenly take “right angle” turns as they dive into the deep retina. A subtle feature of MacTel is the development of retinal “graying” where there appears to be reduced transparency of the retina in the parafoveal area. Later there can be additional pigment changes and pigment hyperplasia resulting in pigmented plaques within the retina itself that may or may not be associated with RPE atrophy.

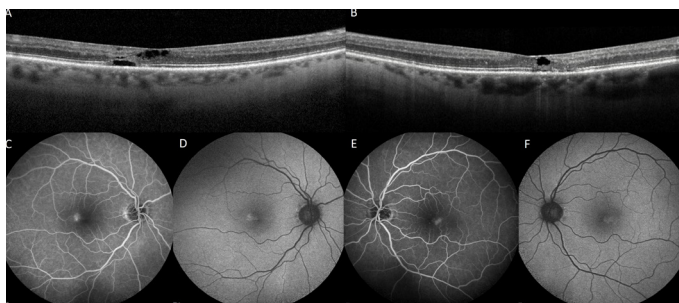
OCT and OCT-angiography can be used to identify changes suggestive of MacTel. OCT may demonstrate hyperreflective refractile dots as above, hyporefective cavitations in the foveola, and ellipsoid zone disruptions temporally. Fluorescein Angiogram may demonstrate telangiectatic vessels temporal to the foveola in early stages of the disease with diffuse hyperfluorescence in later stages.

Additional testing options include fundus autofluorescence to assess the health of the retinal pigment epithelium, confocal reflectance imaging to demonstrate loss of macular pigment, or confocal adaptive optics scanning laser ophthalmoscopy to identify individual photoreceptor and RPE cell layer loss.

## Treatment

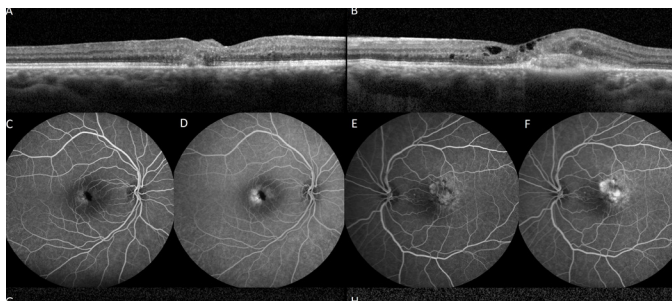
While most cases of MacTel do not require treatment due to the favorable prognosis and lack of effective treatment options, researchers are developing therapies to reverse or stall the neurodegenerative and vascular changes that occur. Most importantly, neovascularization must be identified and treated with antiVEGF therapy to reduce the likelihood of vision loss and scarring. Additionally, macular hole development may require surgery, but the surgical outcomes are less favorable than with typical macular holes.

Case 1 is a patient with typical macular changes from intermediate stage Type 2 MacTel (Figure 1). The patient has some mild blurry vision and difficulties reading. The visual acuity is: 20/30 OD and 20/20 OS. The OCT demonstrates the foveal cavitations and outer segment disruptions that are typical of Type 2 MacTel. The fluorescein angiogram demonstrates some mild late leakage in the temporal macula OU. The fundus autofluorescence demonstrates some RPE abnormalities with mixed hyper- and hypo-autofluorescence.



**Figures 1:** (A/B) Optical coherence tomography demonstrates foveal cavitations and ellipsoid zone defects in both eyes, (C/E) Late fluorescein angiogram demonstrates mild exudation from temporal parafoveal telangiectasis, (D/F) fundus autofluorescence demonstrates some temporal parafoveal RPE irregularities with hyperautofluorescence

Case 2 is a patient with bilateral choroidal neovascularization in the setting of Type 2 MacTel. He presented with decreased vision and distortions in both eyes (Figure 2). He then underwent successive intravitreal injection therapy and has recovered significantly but remains with macular changes from the underlying MacTel. Currently, excellent control of the exudation is being achieved with intravitreal injection of aflibercept every 3 months.



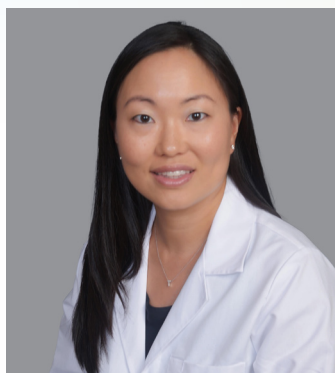
**Figures 2:** (A/B) Optical coherence tomography (OCT) demonstrates at presentation demonstrate temporal choroidal neovascularization (CNVM) with exudation OS > OD, (C-F) early and late fluorescein angiogram photographs demonstrate early demarcation of the choroidal neovascular membrane and late exudation from the CNVM in both eyes, (G/H) OCT after treatment demonstrates regression of the CNVM and the slight presence of more typical foveal cavitations and temporal parafoveal ellipsoid zone defects, particularly in the right eye.

## Summary

Macular telangiectasia is a degenerative condition of the macula with similarities to age-related macular degeneration. In Type 2 MacTel, examination may demonstrate pigmentary changes and vascular abnormalities, and OCT may demonstrate foveal cavitations and temporal parafoveal ellipsoid zone disruptions. Both conditions increase the risk of choroidal neovascularization and exudation that can be treated with anti-VEGF therapy. Type 2 MacTel carries some increased risk of developing a full-thickness macular hole with unique characteristics that reduce the success of typical macular hole repair surgery. Eye care providers should be aware of this condition and its features in order to accurately diagnose and counsel the patient.

## References:

1. Leung I, Sallo FB, Bonelli R, Clemons TE, Pauleikhoff D, Chew EY, Bird AC, Peto T; MacTel Study Group. "Characteristics of pigmented lesions in type 2 idiopathic macular telangiectasia." *Retina* 2018 Jan;38 Suppl 1(Suppl 1):S43-S50.
2. Yannuzzi LA, Bardal AMC, Freund KB, Chen K, Eandi CM, Blodi B. "Idiopathic macular telangiectasia." *Archives of Ophthalmology* 2006 Apr:450-460.
3. Gass JD, Blodi BA. "Idiopathic juxtafoveolar retinal telangiectasias. Update of classification and follow-up study." *Ophthalmology* 1993;100:1536-46.



**Tamara Lee Lenis M.D., Ph.D.**

## NJRetina Welcomes Tamara Lee Lenis, MD, Ph.D. to our medical staff

### Medical Training

**Fellowship:** New York Presbyterian/Weill Cornell Medical Center, NY - Vitreoretinal Surgery Fellowship

**Residency and Ph.D. Training:** University of California Los Angeles, CA  
Eye STAR Program (Specialty Training and Advanced Research) - Stein Eye Institute  
Internship: University of California Los Angeles - Olive View Medical Center, CA

### Education

**Ph.D.:** University of California Los Angeles, CA - Molecular, Cellular, and Integrative Physiology  
**MD:** Cleveland Clinic Lerner College of Medicine, Case Western Reserve University, OH  
**MS:** Clinical Research - Cleveland Clinic Lerner College of Medicine, Case Western Reserve University, OH  
**BA:** Columbia College, Columbia University, NY

### What is your philosophy of care?

Listen. Treat the whole patient. Treat my patients as I would a family member.

### What made you choose the Retina field as your specialty area?

Retina is an exciting specialty because it spans medicine and surgery, and because you can quite dramatically help improve quality of life through vision. As a retina specialist, you often help your patient develop agency for their health and in turn their vision. There is also a lot of exciting research in retina that lends hope and treatment options for conditions with previously limited treatments. The imaging in retina is also amazing and helps the doctor (and the patient) better understand the current state of the eye.

### Why did you choose NJRetina?

I chose NJRetina because of its commitment to excellent, evidence-based, and compassionate patient care. After getting to know the group, it is also very clear that everyone shares a commitment to embracing diversity, work-life balance, and coming to work with a smile.

### What are some of your personal interests?

Yoga, running, hiking, cooking, travel, and spending time with family.

### Are you fluent in any language aside from English?

I am conversant in Spanish and Korean.



## At the forefront of clinical research

NJRetina continuously conducts clinical trials at key locations. Our clinical research coordinators will be happy to discuss the inclusion/exclusion criteria or any other aspect of these studies with you or your patients. If you have any questions, please feel free to contact:

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## Enrolling Studies:

### Wet AMD

#### Edison

**Opthea Coast:** A Phase 3, Multicenter, Double-masked, Randomized Study to Evaluate the Efficacy and Safety of Intravitreal OPT-302 in Combination with Aflibercept, Compared with Aflibercept Alone, in Participants with Neovascular Age-related Macular Degeneration (nAMD)

#### Toms River

**Opthea Shore:** A Phase 3, Multicenter, Double-masked, Randomized Study to Evaluate the Efficacy and Safety of Intravitreal OPT-302 in Combination with Ranibizumab, Compared with Ranibizumab Alone, in Participants with Neovascular Age-related Macular Degeneration (nAMD)

#### Teaneck

**Elevatum:** A Phase IIIB/IV, Multicenter, Open-Label, Single-Arm Study to investigate Faricimab treatment in response to treatment-naïve, underrepresented patients with Diabetic Macular Edema.

#### Teaneck

**Luna:** A Multi-Center, Randomized, Double-Masked Phase 2 Study to Assess Safety and Efficacy of ADVN-022 (AAV.7m8-aflibercept) in Anti-VEGF Treatment Experienced Patients with Neovascular (Wet) Age[1]related Macular Degeneration

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