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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[1] 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 parafoveolar 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.

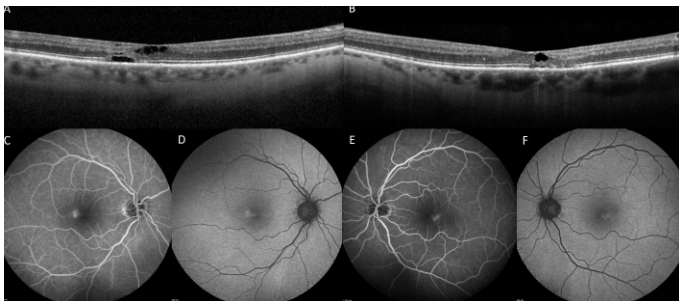
OCT and OCT-angiography can be used to identify changes suggestive of MacTel. OCT may demonstrate hyperreflective refractile dots as above, hyporeflective cavitations in the foveola, and ellipsoid zone disruptions temporally. Fluorescein Angiogram may demonstrate telangiectatic vessels temporal to the 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

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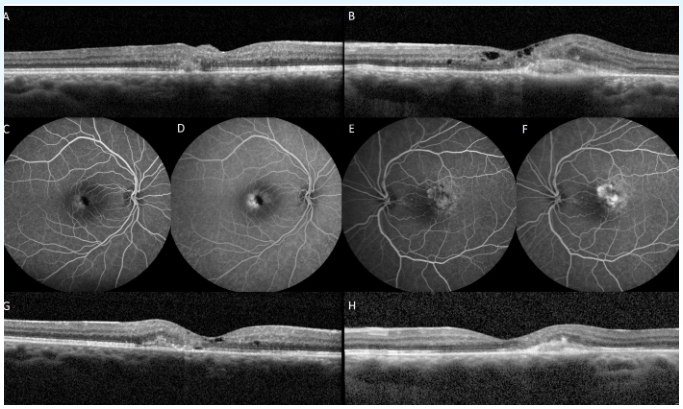
Case 2 is a patient with bilateral choroidal neovascularization in the setting of Type 2 MacTel. He presented with decreased vision and distortions in both eyes (Figure 2). He then underwent successive intravitreal injection therapy and has recovered significantly but remains with macular changes from 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.

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.

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At the forefront of clinical research

The Retina Care Center continuously conducts clinical trials at our Baltimore, MD office. 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:

Dry AMD

Golden: A Phase 2, Randomized, Placebo-Controlled, Double-masked Study to Assess Safety and Efficacy of Multiple Doses of IONIS-FB-L, an Antisense Inhibitor of Complement Factor B in Patients with Geographic Atrophy Secondary to Age-related Macular Degeneration (AMD)

Explore: A Phase 2, Outcomes assessor-Masked, Multicentre, Randomised Study to Evaluate the Safety and Efficacy of Two Doses Of GT005 Administered as a ASA Single Subretinal Injection in Subjects with Geographic Atrophy Secondary to Age-related Macular Degeneration

Horizon: A Phase II, Open-label, Outcomes-assessor Masked, Multicenter, Randomized, Controlled Study to Evaluate the Safety and Efficacy of Two Doses of GT005 Administered as a Single Subretinal Injection in Subjects with Geographic Atrophy Secondary to Dry Age-related Macular Degeneration

Wet AMD

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

Belvedere- ML43000: A Phase III/IV, Multicenter, Open-Label, Single-Arm Study of The Efficacy and Safety of the Port Delivery Systemwith Ranibizumab in Patients with Neovascular Age-related Macular Degeneration Previously Treated with Intravitreal Agents other than Ranibizumab

RGX-314 Atmosphere: This Randomized, Partially Masked, Controlled, Phase 2b/3 Clinical Study will Evaluate the Efficacy and Safety of RGX-314 Gene Therapy in Participants with nAMD. The Study will Evaluate 2 Dose Levels of RGX-314 Relative to an Active Comparator. The Primary Endpoint of this Study is Mean Change in Best-corrected Visual Acuity (BCVA) of RGX-314 Relative to Ranibizumab. Approximately 300 Participants who Meet the Inclusion/Exclusion Criteria, will be Enrolled into One of 3 Arms.

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