Macular Telangiectasia Type 2: A Comprehensive Review

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Abstract: Macular telangiectasia type 2 (MacTel) is a gradually progressive disease that affects the quality of life by impairing both distant and near vision. It had previously been considered a vascular condition, but recent evidence suggests a neurodegenerative etiology, with primary involvement of Muller cells. Retinal pigment epithelium (RPE) hyperplasia and subretinal neovascularization (SNV) are responsible for most of the vision loss in advanced cases. Neurotrophic factors in the non-proliferative phase and intravitreal anti-Vascular Endothelial growth factor (VEGF) in the proliferative phase have shown to retard the progression of the disease. This review will discuss the pathophysiology, clinical features, important diagnostic imaging studies and available treatment options for MacTel.

Keywords: macular telangiectasia type 2, MacTel, CNTF, ciliary neurotrophic factor

Introduction

Macular telangiectasia type 2 (MacTel) is a slowly progressive disease of the macula. It had previously been considered a vascular condition, but recent evidence suggests a neurodegenerative etiology, with primary involvement of Muller cells. Retinal pigment epithelium (RPE) hyperplasia and subretinal neovascularization (SNV) are responsible for most of the vision loss in advanced cases. Neurotrophic factors in the non-proliferative phase and intravitreal anti-Vascular Endothelial growth factor (VEGF) in the proliferative phase have shown to retard the progression of the disease. This review will discuss the pathophysiology, clinical features, important diagnostic imaging studies, and available treatment options for MacTel.

Demography

MacTel is a bilateral asymmetric condition affecting patients over the age of 40 years,1,2 with a predilection for women.1,3 The Beaver Dam Eye Study reported a prevalence of 0.1% in 4790 individuals between the ages of 43 and 86 years,4 but the Melbourne Collaborative Cohort estimated a considerably lower prevalence of 0.0045% in 22,415 participants.2 MacTel is equally prevalent throughout the world with no racial predilection.5

Pathogenesis

Vascular Theory

Gass and Oyakawa proposed that stasis within the retinal veins due to arterial crossing impingement plays an etiologic role.1 They believed that chronic exudation from the telangiectasia leads to outer retinal atrophy and RPE metaplasia.

Several inconsistencies plagued the widespread acceptance of the vascular theory. MacTel usually starts in the temporal parafovea though the reason for this predilection is unclear. Anastomoses between the superior and inferior temporal arcade vessels could be associated with vascular decompensation, though the reason behind this mechanism had not been elucidated.6 Retinal thickness is substantially reduced (not increased) despite dye leakage on fluorescein...
angiography (FA) and vision loss due to photoreceptor atrophy occurs without any evidence of cystoid macular edema. Optical coherence tomography (OCT) shows abnormalities inconsistent with vascular leakage and leakage on FA often predates the development of telangiectasia.

Neurodegenerative Theory
Muller cells are important to maintain the blood retinal barrier, and providing trophic factors to the surrounding neurons. Muller cell dysfunction is central to the neurodegenerative theory of MacTel.

The Muller cells surround the neurons and provide them with nutrition. The Muller cell processes are in close contact with the retinal blood vessels in the outer plexus. Nutritional deprivation may be a major factor leading to the loss of retinal transparency commonly seen in MacTel. Muller cell dysfunction is associated with atrophy and disorganization of the outer retina, leading to hyporeflective spaces in the outer retinal layers seen on OCT. OCT and ERG changes in the form of inner lamellar holes and reduced cone responses manifested before the development of typical vascular changes. Xanthophyll, that is primarily stored in Muller cells, is diminished in early MacTel.

Vascular Proliferative Changes
Neurodegeneration leads to retinal thinning. As the disease progresses, associated capillary endothelial cell degeneration leads to hypoxia. The combination of VEGF secretion and Muller cell dysfunction leads to intraretinal edema, that when occurs with neuroretinal thinning may be associated with normal retinal thickness in the early stages.

It has been suggested that the SNV probably originates from the retinal blood vessels and Indocyanine green angiography (ICGA) has shown intraretinal anastomosis.

Genetics
Reports of familial clusters and affected monozygotic twin suggest that MacTel has a genetic component, although no hereditary pattern has yet been established. The MacTel project group suggested an autosomal dominant inheritance pattern with variable penetrance. Twin studies with asymmetric findings among twins suggest that epigenetics may be the cause for discordance among monozygotic twins. Secondary factors, such as smoking, diabetes mellitus (DM), coronary artery disease or hypertension, may increase the risk of clinical disease.

Genome-wide linkage analysis has identified a single peak on chromosome 1 at 1q41-42 that may lead to the development of MacTel. Serine biosynthesis and lipid recycling in the retina occur primarily in the Muller cells and RPE. Defects in metabolism with accumulation of deoxysphingolipids result in a loss of photoreceptors in MacTel. Eade et al have found a missense mutation in the PHGDH gene involved in serine synthesis pathway in ~3.2% of affected individuals. The PHGDH mutation decreased serine biosynthesis with accumulation of deoxysphingolipids in RPE cells. Table 1 gives details of known genes associated with MacTel.

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<td>Single peak on chromosome 1 at 1q41-42 significant LOD score</td>
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<td>Gantner et al, 2021</td>
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Ocular and Systemic Associations
Up to 45% of patients with MacTel have a systemic disorder most commonly DM, obesity, hypertension and cardio-
vascular diseases.\textsuperscript{5,29} Though Gass and Blodi recognized that DM could predispose patients to develop MacTel the incidence of diabetic retinopathy was found to be significantly lesser in diabetic patients with MacTel.\textsuperscript{11,12} MacTel has also been found to be associated with celiac sprue,\textsuperscript{30} polycythemia vera, Alport disease, CREST syndrome\textsuperscript{31} and pseudoxanthoma elasticum.\textsuperscript{32} MacTel has also been associated with ocular conditions like soft confluent drusen,\textsuperscript{33} microhemangiomas of the pupillary margin,\textsuperscript{34} full thickness macular holes (FTMH)\textsuperscript{35} and epiretinal membranes.\textsuperscript{7}

Clinical Features
Gass and Blodi categorized MacTel based on disease severity into five stages: Stage 1 – slight loss of the retinal
transparency, usually in the temporal juxtafoveal area. Stage 2 – Loss of transparency all around the fovea with
superficial crystals may be seen. Stage 3 - right-angled venules; stage 4 – hyperplastic RPE within the retina; and
stage 5 – retinal neovascularization or exudation from SNV.\textsuperscript{11} Yannuzzi revised the classification into nonproliferative
and proliferative disease.\textsuperscript{12} Nonproliferative MacTel refers to exudative telangiectasia and foveal atrophy and
Proliferative disease refers to proliferative changes with SNV and fibrosis. The Modified Mactel classification described
in 2013, divides MacTel into 5 stages.\textsuperscript{36} The first stage is nonproliferative deep capillary involvement characterised by
inner retinal thickening and a few cysts. The second stage superficial capillary shows loss of transparency and few
telangiectatic vessels temporally. Stage 3 is subinternal limiting membrane and foveal area involvement with distortion
and constriction of FAZ. Stage 4 is proliferative stage showing presence of telangiectatic vessels in deep and superficial
capillary plexus, with proliferation upto sub ILM and subretinal spaces. The stage 5 fibrovascular proliferation shows
subretinal vascular complex perfused by descending arterioles and venules. Venkatesh et al proposed a preproliferative
stage of MacTel that could help identify patients at risk of proliferative disease.\textsuperscript{37}

Retinal graying usually begins temporally and then expands in an oval fashion, with the horizontal linear dimension
being greater than the vertical dimension, usually within one disc diameter from the central fovea. The earliest clinical
feature is a mild grayish discoloration of the temporal retina due to loss of retinal transparency (Figure 1).\textsuperscript{11,38} Vascular
changes are absent or rarely evident early in the disease and FA is often required to identify them.\textsuperscript{39} Ironically, leakage of
fluorescein on FA is not associated with cystoid changes in the retina, and there is lack of lipid exudation or
haemorrhages.\textsuperscript{39}

Blunted, slightly dilated venules are seen in later disease stages of the disease with pigment hyperplasia (Figure 2).
The venules do not narrow as they approach the foveola and they appear to dive at right angles into the deeper retinal
layers. The right angled venules originate from the deep capillary plexus and form a stellate radiating pattern because of

\textbf{Figure 1} Fundus photograph (A) and early (B) and late (C) fluorescein angiography, OCT (D) and OCTA (E and F) in a patient with MacTel. There is loss of retinal transparency temporal to fovea with angiographic leakage. (B and C) OCT (D) shows thinning of the parafoveal retina with hypo-reflective cavities in the outer retina. The OCTA (E and F) demonstrates dilation and telangiectasis of the deep capillary plexus temporal to fovea.
tissue contraction in the temporal macula. They may appear dilated due to increased blood flow through the telangiectatic capillaries.

Eyes with MacTel have multiple tiny, yellow, crystalline deposits on the inner retinal surface adjacent to the telangiectasis (Figure 3). Nearly half of the patients exhibit crystalline deposits (60% bilaterally), but they do not correlate with the severity of disease. The source of the crystalline deposits remains unknown, though research has suggested that they are due to degenerating Muller cells or lipofuscin-containing pigmented cells. The cells containing brown pigment that surround abnormal blood vessels may represent hyperplasia of the RPE.

Yellow foveal lesions occur in up to 5% of cases of MacTel. Focal atrophy of the foveola may create a lamellar macular hole and FTMH may be due to atrophic foveal changes, vitreomacular traction, or progression from a lamellar macular hole. Eyes with FTMH secondary to MacTel are considered poor surgical candidates.

Figure 2 Fundus photograph (A) showing blunted retinal venule and early pigmentation. The corresponding fluorescein angiogram (B) shows mild leakage in the late phase with hypofluorescence at areas of pigmentation. The OCTA (C) demonstrates dilation and telangiectasis of the deep capillary plexus temporal to fovea, and the OCT (D) shows foveal thinning with loss of the ellipsoid zone.

Figure 3 Fundus photograph (A) showing pronounced crystalline deposits, blunted retinal venules, and pigment proliferation. OCT images (horizontal and vertical cross-sections represented as (B and C) respectively) show loss of outer-retinal layers with inner and outer retinal hypo-reflective cavities. Back shadowing beneath a hyper-reflective region corresponds to pigmentary changes on the fundus photo.
Gass and Oyakawa observed that progressive loss of photoreceptors allows hyperplastic RPE cells to migrate along the right angled vessels to form stellate intraretinal hyperpigmented black plaques.\textsuperscript{1} Meleth et al used OCT to verify that pigment clumping is initiated in areas of outer retinal thinning and atrophy.\textsuperscript{46} Intraretinal pigments correlate with disease progression but the relation between plaques and retinal vascular proliferation is controversial.\textsuperscript{46,47}

Advanced MacTel is characterized by outer retinal degeneration, atrophy, and scarring. SNV (Figure 4) develops in a minority of patients\textsuperscript{3} but it can result in significant loss of vision due to subretinal hemorrhage, cystoid macular edema, hard exudates, and disciform scarring.\textsuperscript{12} If the SNV enters the subretinal space, chorio-retinal shunts may develop and neovascular membranes that resemble markedly ectatic capillaries might represent retinal–retinal anastomoses.\textsuperscript{47} Patients with SNV in one eye have an increased risk for development in the fellow eye.\textsuperscript{48}

**Visual Function**

Patients with MacTel have significant more difficulty with near vision compared to distance, and often have central visual field defects. Patients with no or minimal decrease in binocular best corrected visual acuity (BCVA) often have significantly reduced reading ability because of central field defects. For efficient reading, patients need BCVA with preservation of at least four degrees of central field to process words and perform eye movements.\textsuperscript{49}

MacTel patients often have profound paracentral scotomas that can be identified with microperimetry. Visual acuity is generally stable during long-term follow-up studies.\textsuperscript{3,6,11} Watzke et al observed that 25% of their patients maintained a stable visual acuity over 17 years\textsuperscript{6} and Gass and Blodi reported that only 33% of their patients deteriorated to a visual acuity of less than 20/200.\textsuperscript{11} The natural history of MacTel has been described in Table 2.

**Fundus Fluorescein Angiography**

FA has been the gold standard for diagnosing MacTel, and leakage on FA may precede any other visible changes. Characteristically, FA shows dilated leaking capillaries temporal to the fovea in the early phase, and diffuse hyperfluorescence in the late phase. These vascular changes in the deep capillary plexus, as shown by stereoscopic angiographic studies, are usually bilaterally symmetric and the amount of “staining” is similar in the late phase of the angiogram.\textsuperscript{14} The leakage in MacTel is not associated with retinal edema and thickening, suggesting that partial breakdown of the blood-retinal barrier allows passage of small molecules such as fluorescein but not the larger albumin.\textsuperscript{51}

Occasionally, an area of intense focal hyperfluorescence within the area of diffuse hyperfluorescence can occur. This intense hyperfluorescence could be due to staining at the level of the outer retina or the development of intraretinal new vessels.\textsuperscript{37} SNV usually develops near migrating intraretinal pigment epithelium, appears to originate from the deep retinal circulation, and is characterized by early and late fluorescein leakage.\textsuperscript{52}

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**Figure 4** Fundus photograph (A) depicts an active SNV with subretinal hemorrhage inferior to the fovea. Prominent crystalline deposits and pigment proliferation are seen temporal to foveae. The OCT images (horizontal and vertical cross-sections represented as (B and C) respectively) show sub-retinal hyper-reflective material with overlying retinal thickening and the OCTA (D and E) demonstrates a vascular network with a hypo-reflective flow void in the deep capillary plexus.
Optical Coherence Tomography (OCT)

The earliest sign noted on OCT is temporal widening of the foveal pit, due to thinning of the temporal juxtafoveal retina. Most of the thinning occurs in the outer nuclear/Henle’s fiber layer. Charbel Issa et al showed that eyes with decreased central foveal thickness may actually have better light sensitivity than eyes with normal foveal thickness.

The most common OCT findings are hyporeflective cavities in the inner and outer neurosensory retina and disruption of the external limiting membrane (ELM), photoreceptor inner segment-outer segment border and interdigitation zone. Cystoid spaces in eyes with MacTel have more boundary irregularities and lower reflectivity than those in eyes with MacTel 1, retinal vein occlusions and diabetic macular edema. These atrophic hyporeflective spaces are usually situated in the foveal pit with a predilection for the temporal slope and may resemble lamellar macular holes. The hyporeflective spaces in the outer retinal layers are due to photoreceptor loss. Müller cell degeneration may lead to outer retinal atrophy, foveal thinning and the ILM “drape” sign. As the disease progresses, inner hypo-reflective cavities may change, possibly due to loss of supporting structures, surrounding atrophy and decreasing leakage. The hyperreflective cavities may disappear, only to reappear over time.

Eyes with disorganized inner retinal layers collapsed outer retinal layers and disrupted outer retinal hyperreflective bands have significantly worse visual acuity. The amount of ellipsoid zone loss on SD-OCT corresponds with deficits in microperimetry and may be used as a diagnostic feature and outcome measure. The extent of photoreceptor loss correlates with visual impairment, and limiting photoreceptor loss appears to be the most promising therapeutic approach to maintaining vision. Current clinical trials with ciliary neurotrophic factor (CNTF) implants use photoreceptor loss as a marker of efficacy.

Increased reflectivity in various layers due to SNV or pigment hyperplasia may be seen. Macular holes, and foveal detachment occur less commonly. Subretinal deposits, if present, may represent debris from photoreceptor elements, which could cause increased foveal autofluorescence. Hyper-reflective intraretinal lesions with back shadowing on OCT usually correspond to pigment plaques. They first begin to appear in the outer retina before migrating into the inner retinal layers, but larger, flat lesions may start within the inner retina.

Neovascular complexes may present as thick, hyper-reflective lesions on OCT, located in the outer retina. A thicker retina temporal to the fovea compared to the nasal fovea, without any retinal fluid, may indicate early SNV. In the presence of SNV, highly reflective dots can be seen in the inner and outer nuclear layers.
Atrophy of the outer retina is the most common cause of vision loss. Choroidal thinning due to ischemia has been proposed as a cause but Chhablani et al found no difference in choroidal thickness between eyes with MacTel and age-matched healthy eyes. A recent study by Chandran et al has shown that fellow eyes of patients with MacTel showed telangiectasia and FAZ changes in the superficial and deep capillary plexuses prior to any neurodegenerative changes on SD-OCT.

**Optical Coherence Tomography- Angiography**

OCT angiography is useful in classifying the severity of the disease and identifying neovascularisation activity in MacTel. The earliest vascular changes in MacTel occur temporal to the fovea in the deep capillary network, apparent on OCTA as reduced vascular density and telangiectatic vessels in the deep vascular plexus. Progressively, the entire capillary network around the fovea becomes involved, in both the superficial and deep plexus. The abnormalities in the deep vascular network may be identified on the OCTA, very early in the disease when the OCT is within normal limits. Abnormalities in retinal vascular density, length and diameter of vessels, and fractal dimension are seen. Foveal avascular zone (FAZ) irregularities and abnormal shape or dragging of vessels perifoveally appear in stage 3 and 4 of the disease. New vessel invasion of the outer and subretinal spaces occurs beneath areas with the greatest flow abnormalities.

**Autofluorescence**

Increased blue light fundus autofluorescence (FAF) in the fovea that precedes clinical and fluorescein angiographic finding is one of the earliest changes in MacTel. This is likely due to macular pigment depletion from the fovea rather than increased lipofuscin accumulation in the RPE or displacement of the pigment. Normal eyes show central foveal masking on 488 nm FAF due to the central accumulation of macular pigment, but reduced macular pigment density or changes in its topographic distribution may affect this. Chhablani et al showed that 93.3% of eyes with MacTel had loss of foveal autofluorescence.

The distribution of macular pigment optical density (MPOD) mirrors the amount of leakage on FA. There is an initial reduction in MPOD, presumably due to the loss of the xanthophyll pigments, temporally followed by the entire fovea and the appearance of a circular perifoveal ring. Disturbances in macular pigment occur with other ocular diseases but this pattern of MPOD is unique to MacTel.

**Microperimetry**

Microperimetry sensitivity is reduced in the early stages of MacTel but visual acuity is unaffected, suggesting that microperimetry can pick up functional changes before vision becomes affected. In more advanced disease, paracentral sensitivity usually declines two degrees temporal to the foveal center though this depends on the presence and extent of photoreceptor atrophy.

**Multi-Focal Electro-Retinography**

Narayanan et al found that in 28 eyes with proliferative MacTel both N1 and P1 amplitudes on the multifocal ERG were significantly lower compared to normal control. There was, however, no correlation with visual acuity and OCT thickness.

**Treatment**

Treatment of neovascular MacTel has improved with anti-vascular endothelial growth factor (anti-VEGF) drugs but treatment of non-proliferative MacTel remains controversial.

**Non Neovascular Mactel**

Focal and grid laser photocoagulation, photodynamic therapy (PDT), intravitreal triamcinolone acetonide (IVTA) and anti-VEGF drugs have been used to treat non-proliferative MacTel.
The visual acuity did not change with laser. Macular edema reduced in only 20% of eyes.\textsuperscript{74} Compared with untreated eyes, laser photocoagulation is associated with distortion of retinal vessels, more fibrovascular tissue, retinal pigment epithelial clumping, new draining venules, and retinal haemorrhages.\textsuperscript{6,49,75} 

PDT alone and in combination with intravitreal ranibizumab does not appear to be effective to improve visual acuity, or retinal atrophy.\textsuperscript{76,77} Similarly, IVTA has not shown to be beneficial to improve the visual acuity.\textsuperscript{78} Subthreshold micropulse laser with pulses of 15-ms duration have been used in 10 eyes of MacTel. A decrease in inner and outer retinal lacunae with preservation of retinal thickness and improvement in visual acuity by 10 letters was observed in these patients.\textsuperscript{79} 

Use of anti-VEGF drugs does not appear to alter the long-term outcomes in non-proliferative MacTel. There is a short-term benefit of reduction of leakage on FA, normalization of macular capillaries and decrease in retinal thickness. A few studies have reported gain in visual acuity,\textsuperscript{14,80} while other studies have not found substantial benefit.\textsuperscript{81,82} The reduction in retinal thickness is temporary and rebound increase in parafoveal leakage and central retinal thickness has been reported.\textsuperscript{11,83} Vascular changes may lead to deficiency of oxygen and nutrients to the neurosensory retina.\textsuperscript{14} Anti-VEGF agents may also cause photoreceptor degeneration. Hence, use of anti-VEGF drugs in the nonproliferative stage is still controversial.

Pars plana vitrectomy (PPV) combined with ILM peeling does not improve the anatomical or functional outcome in MacTel.\textsuperscript{54} PPV and ILM peeling is commonly performed in eyes with ERM or full-thickness macular holes, and it has been suggested that vitreous adhesion and traction may cause foveal changes in MacTel. PPV is not routinely recommended in MacTel.

Since the earliest finding in MacTel is redistribution of macular carotenoid pigment beginning in the temporal perifovea, supplementation with lutein, xeaxanthin and meso-zeaxanthin has been studied. However, this failed to show any benefit.\textsuperscript{85,86} The role of carotenoid supplementation remains uncertain and needs further study.

CNTF appears to reduce photoreceptor loss in animal models of outer retinal degeneration. CNTF supplementation with the Neurotech 501 cell implant (Neurotech Pharmaceuticals, Inc, Cumberland, RI) is being evaluated.\textsuperscript{87–89} This innovative, cell-based drug delivery system is surgically anchored at the pars plana with a titanium loop (Figure 5).

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**Figure 5** Representative Illustration of Ciliary Neurotrophic factor (CNTF)-secreting implant (A) using Encapsulated cell technology. The implant consists of genetically bioengineered CNTF secreting cells enclosed in a semipermeable membrane. It is surgically anchored at the pars plana using a suture clip (B). The cross section image shows the membrane that allows nutrients to diffuse in and CNTF agent to diffuse out while not allowing immune cells to reach the CNTF secreting cells (C).
Genetically modified human RPE cells are encapsulated in a semipermeable membrane and programmed to synthesize CNTF that is released into the vitreous.\(^\text{90}\)

Phase 2 trials with 99 eyes demonstrated that CNTF significantly slowed the progression of retinal degeneration.\(^\text{91}\) Sham treatment showed greater progression of neurodegeneration, decrease in retinal sensitivity and reading speed compared to eyes treated with CNTF. There have been no safety concerns with the use of CNTF. Phase 3 trials are under way in the US, Europe, and Australia.

### Neovascular Mactel

Active SNV is clinically suspected in the presence of subretinal hemorrhage, thickening of the retina and/or visible membrane at the macula. SNV is confirmed with FA and OCTA imaging.

Subretinal surgery to remove the neovascular membrane has been performed but its strong adherence to the overlying neurosensory retina leads to poor outcomes.\(^\text{92}\) Parafoveal scarring and impaired reading ability have limited the use of thermal laser photocoagulation\(^\text{93}\) and PDT has not been shown to improve visual acuity.\(^\text{94,95}\) Transpupillary thermotherapy (TTT) led to regression of the neovascular network in 11 of 13 eyes at 3 months.\(^\text{96}\)

Anti-VEGF drugs are able to improve visual acuity and control the CNV.\(^\text{97–99}\) In a retrospective series of 16 patients followed over 12 months, Narayanan et al reported improvement in mean visual acuity from 20/120 to 20/70.\(^\text{48}\) They also found that the number of injections was significantly lower (1.9 injections per eye) than needed for other diseases such as age-related macular degeneration. In another study of 25 patients, the mean BCVA improved (20/91 to 20/62) and the central macular thickness decreased.\(^\text{100}\)

Earlier diagnosis of SNV with high-resolution OCT may lead to better visual outcomes. Treatment outcomes of SNV have been described in Table 3.

### Conclusion

MacTel is a neurodegenerative disease that is found among people over the age of 40 years throughout the world. The neurodegenerative mechanism proposes that dysfunction of the parafoveal Müller cells leads to retinal thinning and subsequent vascular changes. However, the pathogenesis of the disease till remains elusive. MacTel causes limited vision loss and may not progress after a few years. Treatment with CNTF to slow disease progression holds promise and anti-VEGF therapy is often able to control SNV.

### Disclosure

The authors report no conflicts of interest in this work.
References


