Commentary: Imaging in macular telangiectasia type 2 – Correlating structural change with vision

Macular telangiectasia, or MacTel, is a neurodegenerative disorder of the macula with a vascular component that is believed to be compensatory or secondary to the primary pathology. Classic clinical features include a loss of foveal translucency, crystalline deposits, intra-retinal cavitary lesions, loss of macular pigment, widened capillary spaces, right-angled vessels, pigment proliferation, perifoveal capillary ectasia, subretinal neovascularization (SRNV), and foveal atrophy. Two different forms, namely nonproliferative and proliferative, have been described.

The original classification of MacTel was proposed by Gass, who divided it into four stages based on the clinical appearance.[1] Further classifications were based on Fluorescein angiography (FFA) leakage, optical coherence tomography (OCT) patterns, optical coherence tomography angiography (OCTA), and multimodal imaging. Most use circumferential involvement as a feature, starting with temporal involvement alone in early stages, and temporal and nasal and eventually the entire circumference with advanced stages of the disease.

Various investigations that are performed to characterize the neurodegenerative component are

1. Confocal blue reflectance, optical coherence tomography (OCT) - Muller cell function and ganglion cell involvement. Increased reflectance with confocal blue light occurs due to Muller cell damage as these cells transmit light through the retina and reduced macular pigment whose absorption maximum is in the blue range.[2,3]
2. Macular pigment optic density assessment (MPOD), Fluorescence lifetime imaging (FLIO) – Macular pigment, photoreceptors. FLIO has been shown to detect changes in macular pigment with high contrast; thus, changes in macular pigment-specific lifetimes provide information about macular photoreceptors or possibly Müller cell loss.[4]
3. OCT, including enface OCT – Ellipsoid zone (EZ) evaluation.[5]
4. Multifocal ERG (MFERG), microperimetry – tests of function.

Investigations that evaluate the vascular or angiogenic component include

1. FFA – nonproliferative vs. proliferative MacTel, identification of SRNV
2. OCT – EZ changes[5]
3. OCTA – telangiectasia in superficial and deep capillary plexus (SCP and DCP), right-angled vessels, retinochoroidal anastomosis (RCA), SRNV, and vessel density.

The neurodegenerative and vascular changes are believed to run in parallel.[3] Photoreceptors along the margins of EZ loss may be the initial source of signaling to the overlying vasculature leading to telangiectasia, and progressive loss of these photoreceptors may alter the signal. This, combined with loss of the Muller cell control of vascular growth, may lead to worsening of telangiectasia and the development of SRNV.[6]

Methods of evaluating the severity of MacTel involve both qualitative and quantitative imaging features, in particular using OCT and OCTA. The EZ integrity measured on horizontal OCT images and the EZ area measured on en face OCT are representative of photoreceptor function. They are currently the most established markers for disease progression as well as assessment of response to interventional measures for the disease.[5] Retinal cavitations are caused by Muller cell damage and are known to change in character over time as a process of continuous remodeling. Large cavitations and extensive EZ loss are seen in eyes with poor vision, though cavitations may disappear with extensive end-stage atrophy. The lack of correlation between cavitation volume and EZ disruption has been observed, though cavitation volume has been shown to negatively correlate with BCVA.[7]

OCTA enables us to obtain three-dimensional images of the perfused microvasculature in different retinal layers and choroid in a repeatable, noninvasive manner. OCTA studies of the retinal vasculature in MacTel have observed a decrease of capillary density, dilated and telangiectatic vessels, and the presence of RCA as well as SRNV in the outer retinal layers. Quantitative OCT-A data show a progressive rarefication of the retinal microvasculature in MacTel. The deep retinal plexus showed a progressive decrease of mean vessel density (VD), skeleton density (SD), and fractal dimension (FD) in the temporal parafoveal region in all disease stages. In the superficial layer, VD, SD, and FD were significantly decreased in the temporal parafoveal region of advanced and neovascular stages. The segments of the ETDRS grid were used as guides for measurement.[8,9]

An article in this issue of the journal measured the area of vascular telangiectasia and correlated it with other clinical and structural parameters on OCT.[10] Measurement of the dimensions of the vascular abnormality has rarely been performed, possibly due to the difficulty in accurately identifying the exact borders of the same. The dynamic changes that occur in the natural history of the disease especially with respect to retinal cavitations on OCT emphasize the need to select imaging techniques and parameters that are reliable, reproducible, and easily interpreted.

Severe vision loss is rare in MacTel and is mostly due to the neurodegenerative component, rarely the neovascular process. Findings in eyes with very poor vision from MacTel Natural History Observation Registration Study showed foveal photoreceptor layer atrophy with or without associated subretinal fibrosis; an affected area, termed MacTel area, limited to a horizontal diameter not exceeding the distance between the temporal optic disc margin and foveal center, and the vertical diameter not exceeding approximately 0.8 times this distance (except in eyes with large SRNV); reduced retinal thickness measures within the MacTel area; and less frequent retinal greying and more frequent hyperpigmentation compared with eyes with better BCVA.[11]

Imaging in MacTel has given us a greater understanding of the pathogenesis of the disease, the presence of two parallel pathways of neurodegeneration and angiogenesis, and reliable biomarkers to assess response to treatment. Combined with our recent understanding of genetic influences that could dictate the phenotype, treatments could be directed toward managing the proliferative and nonproliferative forms of the disease.

Muna Bhende

Shri Bhagwan Mahavir Vitreoretinal Service, Medical Research Foundation, Sankara Nethralaya, Chennai, Tamil Nadu, India

Correspondence to: Dr. Muna Bhende, Shri Bhagwan Mahavir Vitreoretinal Service, Medical Research Foundation, Sankara Nethralaya, Chennai, Tamil Nadu, India. E-mail: drmuna@snmail.org

References

1. Gass JD, Blodi BA. Idiopathic juxtafoveolar retinal telangiectasis. Update of classification and follow up study. Ophthalmology 1993;100:1536-46.
2. Charbel Issa P, Berendschot TTJM, Staurenghi G, Holz FG, Scholl HPN. Confocal blue reflectance imaging in type 2 idiopathic macular telangiectasia. Invest Ophthalmol Vis Sci 2008;49:1172-7.
3. Pauleikhoff D, Gunnemann F, Book M, Rothaus K. Progression of vascular changes in macular telangiectasia type 2: Comparison between SD-OCT and OCT angiography. Graefes Arch Clin Exp Ophthalmol 2019;257:1381-92.
4. Solberg Y, Dysli C, Wolf S, Zinkernagel MS. Fluorescence lifetime patterns in macular telangiectasia type 2. Retina 2020;40:99-108.
5. Heeren TFC, Kitka D, Florea D, Clemons TE, Chew EY, Bird AC, *et al*. Longitudinal correlation of ellipsoid zone loss and functional loss in macular telangiectasia type 2. Retina 2018;38(Suppl 1):S20-6.
6. Micevych PS, Lee HE, Fawzi AA. Overlap between telangiectasia and photoreceptor loss increases with progression of macular telangiectasia type 2. PLoS One 2019;14:e0224393.
7. Cai CX, Choong J, Farsiu S, Chiu SJ, Chew EY, Jaffe GJ. Retinal cavitations in macular telangiectasia type 2 (MacTel): Longitudinal structure-function correlations. Br J Ophthalmol 2021;105:109-12.
8. Spaide RF, Klancnik JM, Cooney MJ. Retinal vascular layers imaged by fluorescein angiography and optical coherence tomographic angiography. JAMA Ophthalmol 2015;133:45-50.
9. Tzaridis S, Wintergerst MWM, Mai C, Heeren TFC, Holz FG, Charbel Issa P, *et al*. Quantification of retinal and choriocapillaris perfusion in different stages of macular telangiectasia type 2. Invest Ophthalmol Vis Sci 2019;60:3556-62.
10. Kelkar AS, Kelkar JA, Tidke S, Agarwal A, Bolisetty M, Kelkar S. Does size of telangiectasia on optical coherence tomography angiography influence vision in eyes with type 2 macular telangiectasia? Indian J Ophthalmol 2021;69:3570-6.
11. Heeren TFC, Chew EY, Clemons T, Fruttiger M, Balaskas K, Schwartz R, *et al*. Macular telangiectasia type 2: Visual acuity, disease end stage, and the MacTel area: MacTel project report number 8. Ophthalmology 2020;127:1539-48.

Access this article online

Quick Response Code:

Website: www.ijo.in

DOI: 10.4103/ijo.IJO_2086_21

Cite this article as: Bhende M. Commentary: Imaging in macular telangiectasia type 2 – Correlating structural change with vision. Indian J Ophthalmol 2021;69:3577-8.