OCT angiography classification of type 2 idiopathic macular telangiectasia and correlation with OCT biomarkers

Ivan Sychev, Anitha Shankar, Antonios Ragkousis, David Parry, Paula Burke, Savita Madhusudhan

St Paul's Eye Unit, Royal Liverpool University Hospital, UK

Introduction

Type 2 idiopathic macular telangiectasia (MacTel) is a bilateral acquired neurodegenerative condition associated with loss of Muller cells, causing gradual decline in central vision.
Characteristic vascular abnormalities mainly affect the deep vascular plexus (DVP) in non-proliferative (NP) stages, and development of subretinal neovascularisation (SNV) is seen in the proliferative (PL) stage.
More recently with widespread use of optical coherence tomography angiography (OCTA), a pre-proliferative (PP) stage has been described.

Aims

To categorise eyes with type 2 MacTel into non-proliferative (NP) stage, pre proliferative stage (PP) and Proliferative (PL) stages based on OCTA and describe their association with key OCT features, in order to facilitate identifying eyes at risk of developing acute/ subacute severe sight loss from macular neovascularisation.

Methods

•Single-centre retrospective case series analysis.
•Electronic patient record system at a tertiary UK NHS hospital.
•Treatment-naïve patients with a confirmed diagnosis of type 2 MacTel
•Bilateral macular OCTA and OCT images (Heidelberg Spectralis) from the same visit were graded at Liverpool Ophthalmic Reading Centre for pre-defined established features of type 2 MacTel and secondarily adjudicated by a senior clinician.
•Eyes with Deep Vascular Plexus (DVP) bunching and absent SNV were assigned to the PP group (Fig. 1 and 2), eyes with a neovascular complex in the avascular slab on OCTA to the PL group and the rest to the NP group.
•Patients with other macular co-pathology and images affected significantly by artefacts were excluded.

Results

•Twenty-six patients (52 eyes). Mean age of 65 (40-83) yrs
•Females :15 (58%), Males : 11 (42%)
•81% had the same stage bilaterally
•There were 26 (50%), 7 (13.5%) and 19 (36.5%) eyes with NP, PP and PL MacTel
•Mean BRVA of 74 (27-95, SD 13.83), 78 (57-88, SD 9.91), 62 (5-83, SD 19.73) ETDRS letters and total macular volume (TMV) of 7.57, 5.84, 7.06 mm^3 with NP, PP and PL MacTel mirroring CSFT findings (Fig. 3)
•In the PL group, 100% of eyes had disorganised outer retinal hyperreflective material and inner retinal hyporeflective spaces, 95% had outer retinal cavitation and 79% had retinal pigment epithelial elevation and disruption.

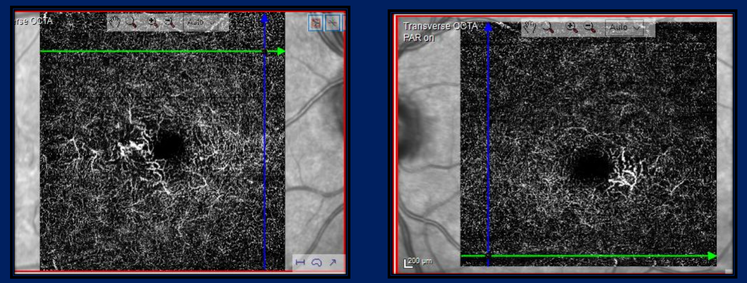


Figure 1. OCTA of right and left eye showing dilatation and bunching at the level of the deep capillary plexus (DCP) in bilateral pre-proliferative Mac Tel.

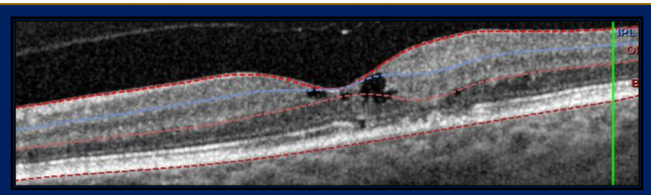


Figure 2. OCTA showing inner retinal hyporeflective spaces and outer retinal cavitations in a patient with pre proliferative disease.

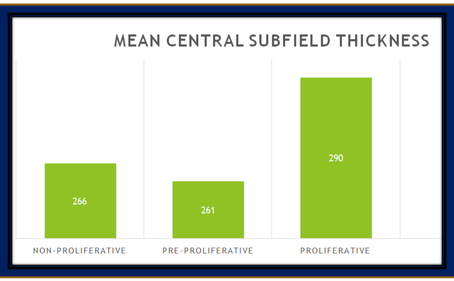


Figure 3. Mean central subfield thickness (CSFT) on macular OCT in different categories of MacTel.

Results (continued)

•In the NP and PP groups 96% and 100% of eyes had inner retinal hyporeflective spaces, 58% and 29% had hyperreflective middle retinal layers and 50% and 71% had outer retinal cavitation respectively .
•On OCTA in the NP group 31% and 73% of eyes, and in the PP and PL groups, approximately 85% and 100% of eyes had dilated perifoveal vessels in the superficial vascular plexus and DVP layers respectively.
•Eyes with foveal avascular zone (FAZ) irregularity increased from 46% to 86% in the NP to PP group.

Conclusion

•The most prevalent OCT biomarkers seem to be inner retinal hyporeflective spaces and outer retinal cavitation across the 3 groups. With progressive stages of MacTel the latter became almost universally prevalent.
•TMV reduced in the pre-proliferative group, likely due to more advanced thinning of the neural retina compared to the non-proliferative group but increased in the proliferative group.
•There was no association between worsening degree of dilated vessels in DVP on OCTA in the pre-proliferative group and the prevalence of hyperreflective middle retinal layers on OCT.
• Even in eyes with minimal DVP bunching on *enface* OCTA there was an obvious increase in flow signal on cross-sectional B-scan, and localised distortion of FAZ boundary, both of which might serve as surrogate markers to identify pre-proliferative MacTel and undertake closer monitoring.
• Disorganised hyperreflective material disrupting retinal layers is universally present on OCT in eyes with proliferative MacTel; further evaluation with OCTA to confirm the diagnosis promptly and offer timely treatment is advisable.

References

1) Venkatesh R, Reddy NG, Mishra P, et al. The preproliferative stage in type 2 macular telangiectasia (MacTel type 2). Graefes Arch Clin Exp Ophthalmol. 2022 Jan;260(1):121-132.
2) Villegas VM, Kovach JL. Optical Coherence Tomography Angiography of Macular Telangiectasia Type 2 with Associated Subretinal Neovascular Membrane. Case Rep Ophthalmol Med. 2017;8186134.