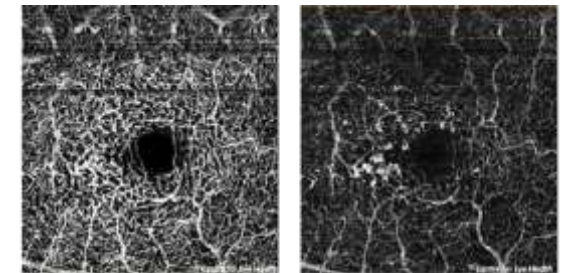
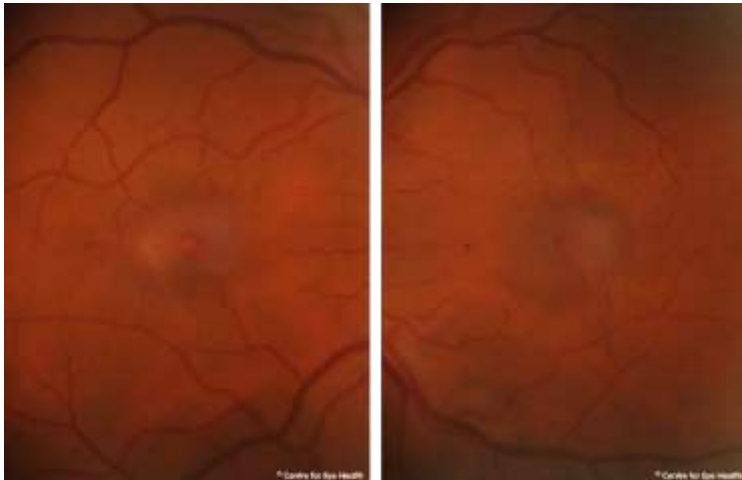


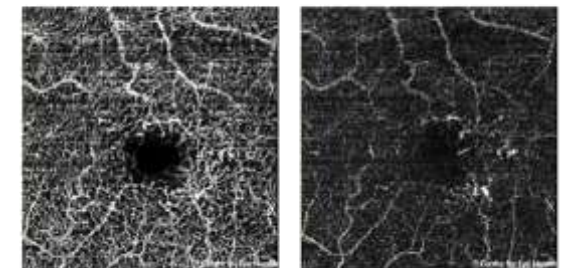
CFEH Facebook Case #142

A 70 year old male was referred to the Centre for a macula assessment. Aided acuities were 6/15+ OD and 6/7.5- OS and he noted a central distortion of the vertical lines on testing with an Amsler grid (OU). As part of the macula assessment, both OCT and OCTA were performed and the results are shown below. What condition does this patient have and what is the potential sequelae of this condition.



OD: Deep vascular plexus

OD: Avascular layers



OS: Deep vascular plexus

OS: Avascular layers

Answer

Spectralis OCT revealed inner retinal cavitation and outer retinal thinning at the fovea in both eyes. OCT Angiography demonstrates temporal juxtafoveal microaneurysms and telangiectatic vessels, more notable in the right eye. These changes are consistent with a diagnosis of macular telangiectasia type 2.

The pathogenesis of Mac Tel 2 is poorly understood, current thought is that it is a primary neuro retinal degenerative condition with a second vascular involvement. It is relatively rare with a prevalence of 0.1% in the general population based on grading of stereoscopic fundus photographs. This is likely to be underestimated as OCT and fundus autofluorescence imaging that can detect disease at an earlier stage was not performed.

Symptoms typically start in the 5th or 6th decade of life and the early signs and symptoms may be very subtle but are usually limited to mildly reduced visual acuity and metamorphopsia which increase as the disease progresses. Typical clinical findings include a dulling or loss of the foveal reflex, a greying of the parafoveal area, small foveal cystoid changes and pigment clumping. A pseudo-vitelliform lesion can also form in the central macula.

This patient is in the non-proliferative stage of this disease as there are no clinical signs of sub-retinal neovascularisation and fibrosis, consequently he was asked to return for a 6 month on-going review. While there is no treatment for the pre-proliferative stages, photodynamic therapy may be useful in decreasing vision loss in the proliferative stages.