

Case of the Month – September 2018

Presented by David Lazar, MD

A 75 year-old woman presented with a "several year" history of poor vision in both eyes. Best corrected vision in the right eye was 20/50. Best corrected vision in the left eye was 20/40. Anterior segment examination was unremarkable OU. Posterior segment examination is as below:



Color Fundus Photos: Right eye: Intraretinal pigment migration, juxtafoveal crystal deposits and right angle venules. Left eye: Superficial crystals concentrated primarily around the temporal macula with adjacent right angle venules.

OCT Macula: Right eye: Increased foveal contour with retinal atrophy from the outer nuclear layer to the RPE. Focal areas of intraretinal hyperreflectivity are present. Left eye: Retinal atrophy/loss with ILM draping. Intraretinal hyperreflective changes.

Differential diagnosis: Macular Telangiectasia type 2 or Mac Tel 2, Wet Age Related Macular degeneration, Diabetic Macular edema, Radiation retinopathy, Coats Disease, Lamellar Holes.

Discussion:

The patient was diagnosed with Macular Telangiectasia type 2. Macular Telangiectasia is a rare disorder which causes decreased vision or an enlarged blind spot. It has three different sub-groups and is characterized by abnormal blood vessels in the parafoveal region, lipid deposition, and vascular leakage. Type 1 is a unilateral disease, Type 2 (most common form) is bilateral, and type 3 (rarest form) is diagnosed by an occlusive telangiectasias. The pathophysiology of Macular Telangiectasia is unclear but is associated with parafoveal retinal/Muller cell death, which affects the adjacent retinal vasculature. Although some promising clinical trials are ongoing (CNTF Trial), there is currently no proven treatment for this condition. However, new abnormal blood vessels may form and often respond well to anti-VEGF medication. Smoking increases your risk of this disease. The surgeons at The Retina Partners are experienced and comfortable with diagnosing and managing this rare disease.

Take Home Points

- Macular Telangiectasia is a rare retinal disorder that can be confused with Wet Macular Degeneration
- There is no current treatment for this disorder, although upcoming clinical trials are promising
- Smoking is a risk factor for this disease





homas Hanscom Robert Engstrom



Hajir Dadgostar





Christian Sanfilippo

Amir Guerami

David Lazar

