



## A Healthy 64-Year-Old Male with Difficulty Reading Fine Print

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### Introduction:

A healthy 64-year-old male presented to our clinic for difficulty with reading fine print. His past medical history was significant for Type 2 diabetes and he had no history of ocular procedures.

### Exam:

On examination, his visual acuity was 20/40 OD and 20/25 OS. Intraocular pressures were normal with no afferent pupillary defect. His anterior exam was notable only for a mild cataract in both eyes. On posterior segment examination, both maculas revealed mild foveal RPE changes without significant vascular abnormalities. OCT imaging of the macula demonstrated cavitary changes in both foveas with a near full-thickness defect in the right eye. In addition, outer retinal/ellipsoid changes were evident without the presence of a PED or frank sub-retinal fluid. Fluorescein angiography was performed demonstrating mild temporal foveal leakage of both eyes without disc leakage. Based on this patient's history and exam findings, a preliminary diagnosis was reached of macular telangiectasia without clinically significant vascular abnormalities. The patient was told to undergo a refraction and return to clinic in 1 month for repeat exam and imaging.

### Discussion:

Type 2 macular telangiectasia (Type 2 'MacTel') is a bilateral, at times asymmetric disease presenting in middle aged patients usually manifesting as decreased central vision or metamorphopsia. Characteristic fundus findings include parafoveal right angle diving venules, intraretinal crystals and temporal parafoveal

graying.<sup>1-3</sup> With progression, complications can develop including choroidal neovascular membrane (CNVM) formation and macular hole.<sup>4</sup> OCT features include pathognomonic intraretinal irregular cavitations that may be misdiagnosed as intraretinal edema from microvascular diseases such as diabetes or retinal vein occlusion. The pathophysiology of MacTel is insufficiently understood but thought to be due to a dysfunction in Muller cells of the retina, whose role is paramount in maintaining the structural and functional stability of various signal processing cells of the neurosensory retina. Our case is somewhat unique in that no characteristic vascular abnormalities were seen on fundus examination, although sub-clinical vascular pathology is possible given the late leakage seen on fluorescein angiography. In a recent retrospective review of imaging characteristics of MacTel type 2 patients, 8 of 111 (7.2%) had characteristic retinal cavitations with no

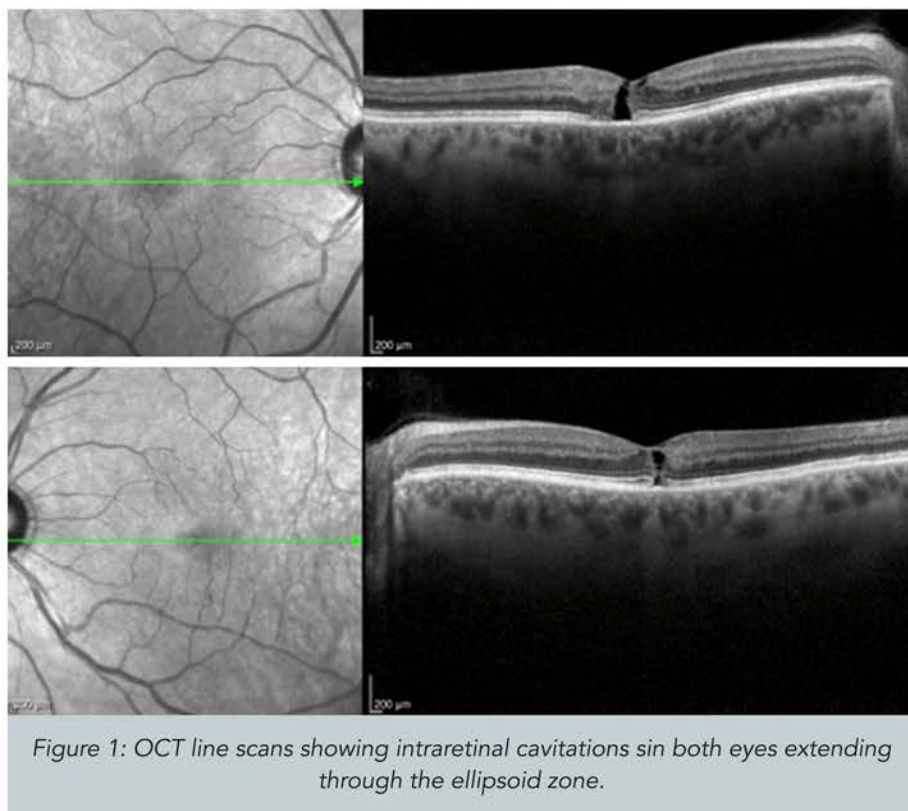


Figure 1: OCT line scans showing intraretinal cavitations in both eyes extending through the ellipsoid zone.



evidence of vascular abnormalities or leakage of angiography.<sup>5</sup> Gillies et al. has proposed Muller dysfunction may result in two parallel pathways of pathogenesis in MacTel type 2.<sup>6</sup> In the neuronal pathway, ellipsoid zone disruption can progress to cavitation and eventual retinal atrophy. In the vascular pathway, angiographic leakage may progress remodeling and eventual sub-retinal neovascularization. It remains unclear whether these pathways are mutually exclusive, however our case sheds light on asymmetric progression between these two routes. Treatment for MacTel type 2 unfortunately is only limited to managing its complications such as CNVM with anti-VEGF agents. However, clinical trials are ongoing to evaluate the natural progression of the disease as well as implantable therapeutic modalities.

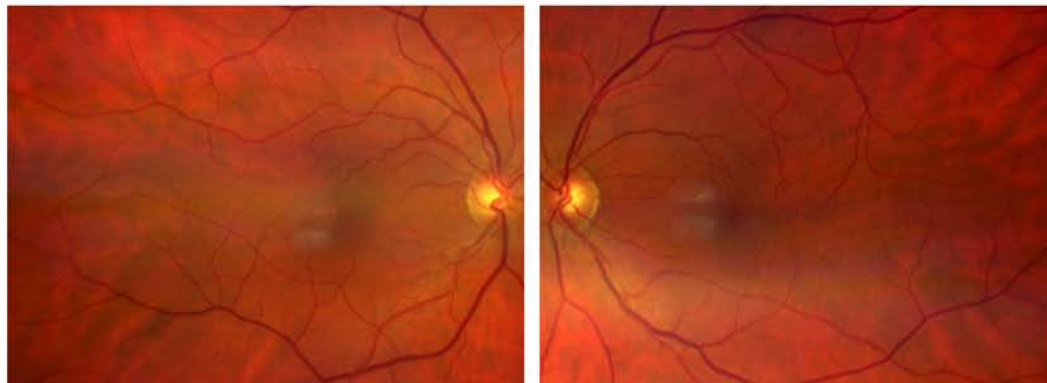


Figure 2: Color fundus photographs demonstrating mild parafoveal greying without significant vascular abnormalities.

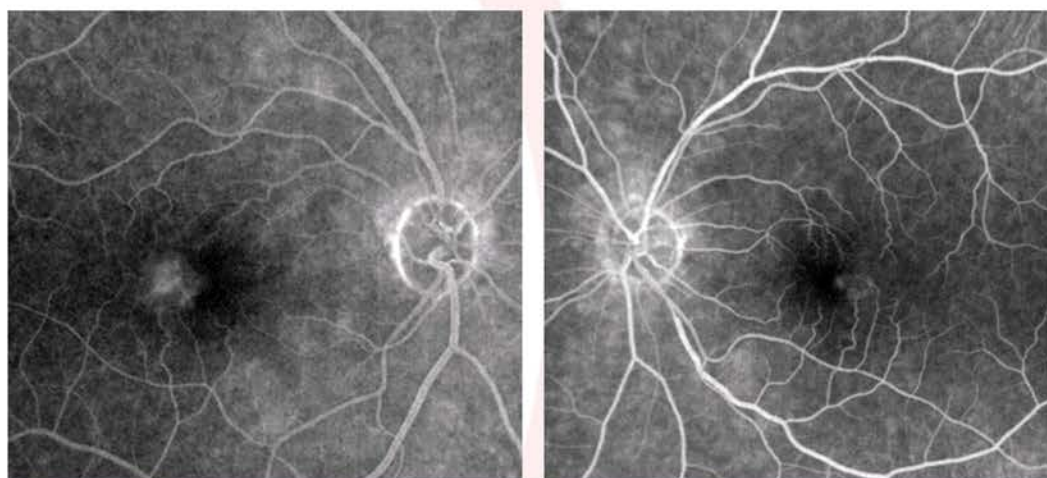


Figure 3: Fluorescein angiography showing mild temporal foveal leakage in both eyes during late phase.

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