Macular Telangiectasia Type 2 with Submacular Vitelliform Deposits

Dr. Apoorva Ayachit MS, DNB, FICO(UK), FVRS(MMJEI), FAICO(VR), **Dr. Suresh Babu N., Dr. Guruprasad A.S., Dr. Srinivas M. Joshi, Dr. Laxmipriya Uday Reddy**

Department of Vitreoretina, M.M. Joshi Eye Institute, Hubballi, India

Abstract: *Macular telangiectasia (Mac Tel) is typically bilateral and presents with near vision problems and bilateral foveal and parafoveal graying of the retina, superficial crystals and neuronal degeneration seen as cavitations on optical coherence tomography. We present a case of a middle aged woman who presented to us with unusual yellowish deposits subretinally along with typical features of mactel on multimodal imaging.*

Macular telangiectasia type 2 (Mac Tel) also known as juxtrafoveal telangiectasia is characterized by parafoveal telangiectasia, graying of retinal area involved, hyperplasia of retinal pigment epithelium, right angled retinal venules and can be complicated by choroidal neovascular membranes (CNVM). It is typically a bilateral disease. It is known to be associated with empty spaces- intraretinally and subretinally and rarely neurosensory detachments. We present a rare association of Mac Tel type 2 with submacular vitelliform deposits.

CASE REPORT

A 46 year old woman presented to us with diminution of vision in both eyes. On examination best corrected visual acuity (BCVA) in both eyes was 6/9, N6. Anterior segment in both eyes was unremarkable. Fundus examination of right eye showed perifoveal greying and small, multiple, yellowish vitelliform lesions subfoveally and left eye showed perifoveal depigmentation and a solitary yellowish subfoveal vitelliform lesion (Figure 1). Optical coherence tomography (OCT) in both eyes showed disorganized outer retinal layers, disrupted ellipsoid zone and subfoveal hyperreflective echoes with multiple pseudocysts and intact retinal pigment epithelium (Figure 2). Fundus fluorescein angiography showed parafoveal leakage in both eyes suggestive of Mac Tel type 2 (Figure 3). OCT angiography in both eyes showed dilated capillaries temporal and nasal to fovea in both superficial and deep vascular layer typical of Mac Tel type 2 (Figure 4). Full field electroretinogram (FF- ERG) and electrooculogram (EOG) were normal (Arden ratio- 2.8). There was no evidence of CNVM in either eye.

Figure 1: *Multicolor fundus photo of both eyes showing parafoveal graying and yellowish vitelliform lesions at the fovea.*

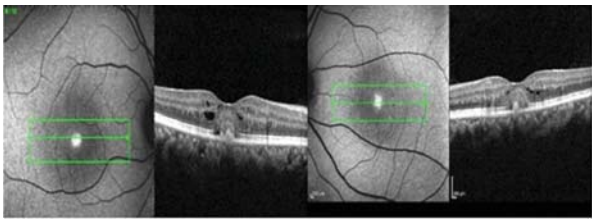

Figure 2: *SD-OCT both eyes shows intact RPE, disrupted ellipsoid zone, parafoveal intraretinal cystic spaces and disorganized outer retinal layers.*

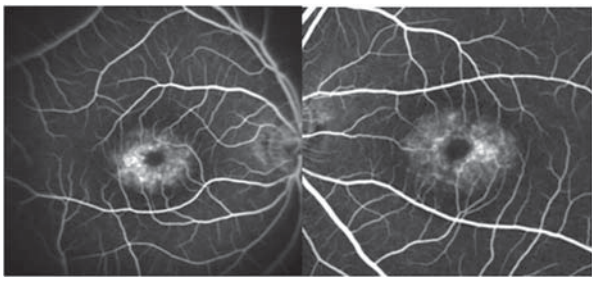

Figure 3: *FFA showing parafoveal capillary leakage typical of Mac Tel. The vitelliform material is not getting stained.*

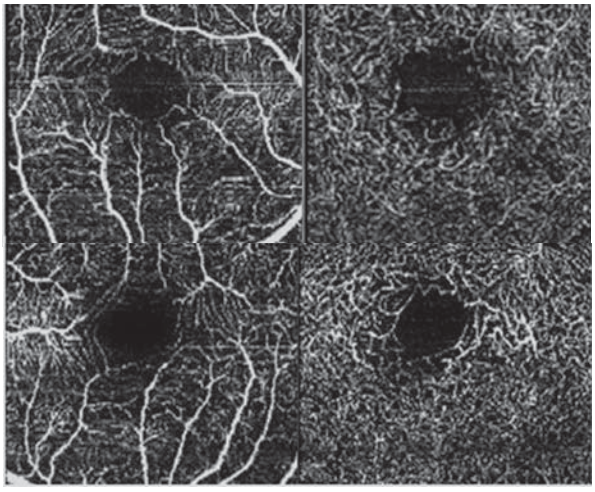

Figure 4: *OCTA showing dilated ectatic and pruned capillaries especially in the deep vascular plexus in both eyes. (right column) compared to the SVC (left column).*

DISCUSSION

The prevalence of Mac Tel type 2 was approximately 1:1000 in the Beaver Dam population, older than 45 years[1]. Mac Tel type 2 can present with superficial crystals, foveal yellow deposits, focal juxtafoveolar pigment changes, dipping venules. Later in the course it can present with inner and outer lamellar cavitations with intact ILM (ILM drape sign)[2].

A case of macular telangiectasia with unusual subretinal vitelliform lesion was described by Lekha et al in 2015[3]. Similar to our case they also encountered subretinal vitelliform deposits and performed electrophysiological tests to distinguish Mac Tel from adult vitelliform foveomacular dystrophy (AVFD). However vitelliform deposits in their case did not have any peculiar characteristics on fluorescein angiography which only showed characteristic perifoveal staining typical of Mac Tel. Cherapanoff et al described subretinal debris in 8 eyes of 6 patients of macular telangectasia. Electron microscopy of the subretinal debris consisted of degenerate photoreceptor outer segments which is similar to AVFD. However, in AVFD there is reaction to this material in the form of RPE hypertrophy, hyperpigmentation and/or anterior migration of RPE. In AVFD phagocytosis of this material leads to increased amount of lipofuscin in RPE which will show hyperautofluorescence on short wave autofluorescence and late staining on FFA[4]. Yellowish lesions seen in Mac Tel can be mistaken for AVFD. OCT features of these vitelliform lesions include hyperreflective material between ellipsoid region and RPE and focal loss of overlying photoreceptor layer but the RPE remains intact in Mac Tel unless complicated by CNVM. The exact reason behind the accumulation of these debris in Mac Tel is not known as the Bruch membrane remains intact and RPE is not the site of pathogenesis. The hypothesis for the accumulation of these debris is 1) phenotypic variation in the phagocytic potential of RPE or microglia, or 2) disrupted Muller cell-RPE and/or Muller cell-microglia interactions[5].

Our patient did not show any features of CNVM and hence was not treated. The typical features of Mac Tel 2 along with the OCT and FFA features correlate with literature. Electron microscopy of such eyes can give further information regarding the nature of subfoveal deposits. Since our patient does not have a long term follow up, we cannot describe the effect of these deposits on the vision or the natural course of this rare association with Mac Tel 2.

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Correspondence to:
Dr. Suresh Babu N.
M.M. Joshi Eye Institute, Hubballi, India