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Long Term Macular Changes in an Impending Macular Hole after Vitrectomy in a Patient with Macular Telangiectasia Type 2

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Purpose: To report and review a bilateral long term follow up case of pars plana vitrectomy of macular telangiectasia (MacTel) type 2 associated impending macular hole.

Case summary: A 61-year-old male presented with decreased central vision and metamorphopsia in both eyes. His best corrected visual acuity was 20/30 in the right eye and 20/50 in the left eye. Based on fundoscopy, fluorescein angiography, and optical coherence tomography (OCT), the patient was diagnosed with MacTel type 2 combined with stage 1B impending macular hole in both eyes. A phacovitrectomy with internal limiting membrane peeling and fluid-gas exchange with SF₆ was performed in both eyes. One week after surgery, an improvement in foveal cysts and foveal defects filled with tissue were observed. However, at 1 month postoperatively, OCT findings showed near complete absence of foveal tissue resembling full thickness macular hole. One year after surgery, this loss of foveal tissue was remained.

Conclusions: In our case, the foveal structure over times has changed after vitrectomy for MacTel type 2 with impending macular hole. A case such as ours with long term follow up using OCT imaging postoperatively has not been previously reported. Our surgery failed to be reopened 1 month later. After vitrectomy, we did not find an anatomical success. However, the patient did not experience serious visual decline.

Keywords: Idiopathic juxtafoveal telangiectasis; Macular hole; Macular telangiectasia; Parafoveal telangiectasis; Pars plana vitrectomy

Introduction

Macular telangiectasia (MacTel) is a bilateral retinal disorder involving telangiectatic perifoveal retinal capillaries [1].

Idiopathic juxtafoveal telangiectasia was first described by Gass [2], and subsequently classified by both Gass [2] and Yannuzzi et al. [3] in 2006, Yannuzzi pooled MacTel types 1 and types 2 together [3].

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MacTel type 2 has a loss of transparency (graying), telangiectatic vessels in the temporal macula, right-angle venules, pigmented plaques, retinal pigment epithelial hyperplasia (RPE hyperplasia) and choroidal neovascularization [3-8]. The foveal cystic changes include lamellar, impending or full-thickness macular holes (FTMH) have been noted on optical coherence tomography (OCT) of eyes with MacTel [3-10].

Only a few reports of the outcomes from surgical intervention with pars plana vitrectomy (PPV) in this setting have been published [9-16]. This report describes the eventual structural macular changes in bilateral MacTel type 2 associated with an impending macular hole after vitrectomy during long term follow-up. We analyzed the changes in progress over a long term follow-up and we provided some information on the decision to treatment of MacTel and the changes after surgery.

Case Report

A 61-year-old male presented with decreased central vision and metamorphopsia in both eyes. The patient had no history of diabetes, hypertension, or other systemic or ocular disease. His best corrected visual acuity (BCVA) was 20/30 in the right eye and 20/50 in the left eye.

Fundus examination of the right eye showed mild grayish discoloration of the temporal to foveal area. Further, the corresponding area showed loss of retinal transparency in the right eye. RPE hyperplasia was observed in the left eye. Late fluorescein angiography (FAG) of both eyes showed late leakage from the telangiectatic vessels near the macula. OCT of both eyes revealed stage 1B impending macular holes. An OCT horizontal scan through the fovea of both eyes showed that the hyporeflectant space was characterized by a foveal cyst extension in the outer retina, as well as a disruption of the photoreceptor layer of both eyes (Fig. 1).

Four years and 8 months ago, the patient's BCVA was 20/25 in the right eye and 20/30 in the left eye. Recently he presented with decreased vision and metamorphopsia. And, OCT scan was more aggravated state and showed a disruption of the photoreceptor layer of the both eyes. The patient was counseled on his guarded or poor prognosis for visual acuity. The patient has mild nuclear sclerosis that was not considered visually significant in both eyes. A phacovit-

Figure 1. Fundus examination of both eyes reveals macular telangiectasia (MacTel) type 2. Fundus photographs of the right eye showed mild grayish discoloration in the temporal to the foveal area (A). The corresponding area showed loss of retinal transparency of the right eye (A). Retinal pigment epithelial hyperplasia is seen in the left eye (B). Late fluorescein angiography (FAG) of both eyes showed late leakage from the telangiectatic vessels near macula (C, D). Optical coherence tomography (OCT) of both eyes reveals a stage 1B impending macular hole. OCT horizontal scan through the fovea of both eyes showed hyporeflectant space characterized by a foveal cyst extending in the outer retina along with a disruption of the photoreceptor layer of both eyes (E, F). OD = oculus dexter; OS = oculus sinister.

recomy with internal limiting membrane (ILM) peeling and fluid-gas exchange with SF6 was performed sequentially in both eyes. He has postoperative face-down positioning for 7 days.

One week after surgery, the BCVA of the right eye was 20/50 and an improvement in the foveal cysts and foveal defects filled with tissue were observed (Fig. 2C). However, no recovery of the inner segment/outer segment layer was found. At 1 month postoperatively, the BCVA of the right eye was 20/25 and OCT findings showed the near complete absence of foveal tissue resembling an FTMH and a thin strand of outer retinal tissue spanning the central defect (Fig. 2E).

At 6 months of surgery, the BCVA of the right eye was 20/30 and the BCVA was maintained from 6 months to 1 year. The loss of foveal tissue persisted even at 6 months (Fig. 2G) to 1 year after surgery (Fig. 2I), with no definite changes in size and shape (Fig. 2A, C, E, G, I).

Figure 2. Optical coherence tomography (OCT) of a patient with macular telangiectasia treated with bilateral phacovitrectomy with internal limiting membrane (ILM) peeling. OCT images of the right eye preoperatively (A), 1 week after surgery (C), 1 month after surgery (E), 6 months after surgery (G), and 1 year after surgery (I). One week after surgery, the foveal cyst showed improvement and partial tissue filling of the foveal defect. However, no recovery of the inner segment/outer segment layer occurred. Postoperative 1 month, OCT findings showed a near complete absence of foveal tissue resembling a full-thickness macular hole. A thin strand of outer retinal tissue spanning the central defect. 6 months and 1 year after surgery, the loss of foveal tissue remained. OCT images of the left eye at the preoperative visit (B), 1 week after surgery (D), 1 month after surgery (F), 6 months after surgery (H), and 1 year after surgery (J). One week after surgery, the foveal cystic lesion disappeared and a mild hyperreflective tissue replaced all intraretinal layers at the fovea in the left eye. However, at postoperative 1 month, OCT findings of the left eye showed full thickness macular hole and an increased loss of foveal tissue the patient's at 6 months and 1 year follow-ups. OD = oculus dexter; OS = oculus sinister.

One week after surgery, the BCVA of the left eye was 20/60 and the foveal cystic lesion disappeared and a mild hyperreflective tissue replaced all the intraretinal layers at the fovea in the left eye (Fig. 2D). However, at postoperative 1 month (Fig. 2F), the BCVA was 20/40, OCT findings of left eye showed FTMH. And this loss of foveal tissue was

slightly enlarged at 6 months (Fig. 2H) and 1 year (Fig. 2J) of follow-up (Fig. 2B, D, F, H, J). The BCVA at postoperative 6 months of the left eye was 20/40 and at postoperative 1 year was 20/50.

Discussion

In our case, we performed a vitrectomy for MacTel type 2 with impending macular hole. Our surgery failed to be reopened 1 month later. After vitrectomy, we did not find an anatomical success. One year after surgery, FTMH was remained.

MacTel type 2 is a rare condition which typically presents with a slow decrease in visual acuity or metamorphopsia. Angiographically, the parafoveal capillaries mainly temporal to the fovea appear to be dilated, and a diffuse parafoveal hyperfluorescence is seen in the late phase of FAG. OCT findings are the disruption of the photoreceptor inner segment-outer segment junction and hyporeflective cavities in the inner retina [1-8].

Degeneration and atrophy of Müller cells associated with MacTel type 2 contribute to the formation of macular holes and cystic changes [6,7]. Müller cell degeneration leads to the loss of structural support, leaving a cystoid lesion that may develop into a macular hole [7]. Recently, immunohistochemical analysis of a postmortem eye demonstrated a significant reduction in the expression of specific markers for Müller cells [8]. Additionally, Charbel Issa et al. [6] postulated that the etiologies of macular holes in MacTel involved abnormal retinal vasculature, ischemia, and the physical absence or atrophy of other retinal tissues [6].

The surgical results of MacTel with FTMH varied from the typical idiopathic macular hole. The difference in the rate of successful closure between idiopathic macular holes and FTMH in MacTel suggests factors other than traction at work in FTMH. Poor postoperative visual acuity was attributed to neurosensory degeneration of FTMH with MacTel [10,12]. Anatomically, a high rate of successful closure of idiopathic FTMH or impending macular hole was reported [17,18]. To date, only a few reports on the surgical outcomes of vitrectomy for FTMH associated with MacTel type 2 have been published [9-14]. Of the 10 eyes reported previously, five macular holes were closed initially, two reopened later, and three were closed at the last follow-up. Rishi et al. [9], Charbel Issa et al. [10], and Gregori et al. [11] reported on patients diagnosed with MacTel and FTMH in one eye, two eyes, and one eye, respectively, diagnosed with MacTel and FTMH. Either surgery failed to close the hole or they reopened four months later. Karth et al. [13] reported that surgical treatment resulted in the closure of two to four holes initially.

Surgical closure with visual improvement after vitrectomy in patient with FTMH was reported in three eye [11-13], Shukla [12] reported good surgical results in one patient with a final postoperative vision of 6/9 and macular hole closure. Shukla [12] proposed that vitreomacular traction may contribute to macular hole formation in patients with MacTel, and that such macular holes, with a configuration similar to idiopathic macular hole, may be good candidates [12]. Further, Shukla and Venkatesh [14] described the spontaneous closure of FTMH secondary to MacTel with concomitant improvements in vision. A 70-year-old man reluctant to undergo surgery, was followed up after 6 months. Re-evaluation showed that the patient's BCVA had improved from 6/18 to 6/9, and the fundus examination suggested spontaneous closure of the FTMH. Spontaneous closure was related to the small hole size and a configuration similar to that of an idiopathic macular hole [14].

On the other hand, surgical treatment of lamellar or impending macular hole in MacTel was reported in similar cases as ours [15,16]. Five eyes of three patients with MacTel and cystoid foveal changes were treated with vitrectomy and ILM peeling and 2-year follow-ups were conducted. Although three out of the five eyes showed stable or slight visual decline postoperatively, two patients experienced significant visual recovery despite OCT evidence of continued anatomical disruption and hyporeflectant spaces within the fovea. Surgical intervention involving vitrectomy may be a treatment option for selected cases of MacTel [15,16]. Sigler et al. [15,16] hypothesized that ILM removal may be related to visual improvement. As Müller cells become depleted, alteration in tangential forces present within the ILM in the parafoveal region may lead to distortion of the remaining photoreceptors, which is a possible mechanism by which ILM removal may result in visual improvement. Furthermore, ILM removal may remove a diffusion barrier and allow transudation into the vitreous, thereby reducing the amount of extracellular fluid.

Our patient with bilateral impending macular hole associated with MacTel type 2 reported decreased vision and metamorphopsia. The OCT horizontal scan through the fovea of both eyes showed hyporeflectant space containing a foveal cyst extending in the outer retina in addition to a disruption of the photoreceptor layer of both eyes. One week after surgery, the foveal cystic lesion had disappeared and the condition of both eyes was improved. However, 1 month postop-

eratively, the OCT findings of both eyes showed FTMH and foveal tissue loss. The loss of foveal tissue was still observed at the patient's 6 months and 1 year follow-ups.

Pathophysiology determines the prognostic variation between patients with idiopathic macular holes and MacTel patients with macular holes. We hypothesize that ILM removal may influence tangential force. However, we cannot remove the cause of the hole formation, suggesting that surgical intervention may have a limited role.

The course of BCVA differs between patients with idiopathic macular holes and macular holes associated with MacTel. In our case, BCVA showed a slight decline, despite OCT findings in both eyes of full-thickness macular hole at the 1 year postoperative follow-up. Similarly, Sigler et al. [15,16] reported three to five eyes with stable or slight visual decline following surgery, and, two patients who experienced significant visual recovery despite OCT evidence of continued anatomical disruption and hyporeflectant spaces within the fovea.

In conclusion, the pathogenesis of macular hole in MacTel type 2 is likely to be multifactorial, consisting of tissue atrophy and Müller cell depletion. The prognosis of a MacTel type 2 patient undergoing vitreoretinal surgery with impending macular hole may depend on the disease progression, which can result in variable surgical outcomes.

Only a few studies have reported the outcomes of surgical intervention with PPV in this setting [9-16]. In our case, the foveal structure over times has changed after vitrectomy for MacTel type 2 with impending macular hole. A case such as ours with long term follow up using OCT imaging postoperatively has not been previously reported. Our surgery failed to be reopened 1 month later.

Statement of Ethics

The authors have no ethical conflicts to disclose.

Conflicts of Interest

The authors declare no conflicts of interest relevant to this article.

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