

Visual and Anatomic Outcomes in Eyes with Idiopathic Juxtafoveal Macular Telangiectasia (MacTel) and Full Thickness Macular Holes Undergoing Surgical Repair

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Abstract

Purpose

To report visual and anatomic outcomes in eyes with idiopathic juxtafoveal telangiectasia (MacTel) type 2 undergoing small gauge vitrectomy surgery with gas tamponade for full thickness macular holes (FTMH).

Methods

Medical records of all adult patients with the diagnoses code for retinal telangiectasia were reviewed. Preoperative and postoperative data were obtained including visual acuity and OCT imaging. These cases were then compared with historical controls that also underwent surgical repair.

Results

Between the years of 2003-2013 34 patients met the inclusion criteria. Of the 34 patients, only 4 patients developed a FTMH. Of the 4 patients, 2 underwent surgical repair. Of these surgical cases one hole closed and one hole remained open at final follow-up

Conclusion

In our cases of FTMH in patients with MacTel type 2, we had mixed results with hole closure and preservation of visual acuity. When adding our cases to the current cases in literature, the hole closure percentage is 47% with stabilization of visual acuity with or without surgical repair. It seems reasonable in these cases to observe unless visual acuity is significantly diminished.

Introduction

Macular telangiectasia (MacTel) type 2, also known as idiopathic juxtafoveal telangiectasia type 2, is a rare disorder involving telangiectatic perfoveal vasculature typically diagnosed in mid to late adult life of unknown etiology [1]. It has been reported to occur in approximately 0.1% of the US population, typically bilaterally often with asymmetric involvement, with no apparent gender predominance [2-4].

The disease was first described by Gass and Oyakawa and the name of the disease was based on characteristic fluorescein angiographic changes in the macula [5,6]. There are additional disease signs as well, including loss of macular transparency, superficial white crystals, depletion of macular pigment, and progressive foveal thinning [6,7]. In addition, optical coherence tomography (OCT) has identified hyporeflective spaces in the inner and outer retina in some patients [4].

Visual symptoms are usually first noticed in the 5th and 6th decade of life including reading difficulties and loss of visual acuity (VA) [1-3]. During early stages of MacTel type 2, central scotopic function is reduced; more advanced cases usually develop dense scotopic and photopic scotomas in the perifovea.

Cystic macular changes, lamellar macular holes (LMH), and full-thickness macular holes (FTMH) have been reported in MacTel and generally are considered poor candidates for surgery because of the underlying degenerative process and chronic vascular leakage [1].

The purpose of this article is to report outcomes of two patients with MacTel type 2 who underwent surgical repair of FTMH and add these cases to the growing body of literature regarding surgical intervention in these rare cases.

Methods

All patients and data were treated in accordance with the declaration of Helsinki and IRB approval was obtained for a retrospective chart review. Cases included patients who presented to Casey Eye Institute at Oregon Health & Science University between 2003 and 2013 in Portland, Oregon who had a diagnosis code for retinal telangiectasia between the ages of 45-75 years. Patients were excluded if they had a known diagnosis of a retinal vascular disease other than Mactel. The description of study population is characterized in (Table 1). All patients were diagnosed with MacTel type 2 based on fundus photography, fluorescein angiography (FA), and OCT studies. Preoperative and postoperative data were obtained including visual acuity and OCT imaging to evaluate the overall visual acuity changes and the macular hole closure rates. These cases were then compared with historical controls that also underwent surgical repair of FTMH in eyes with MacTel type 2.

Table 1: Description of Patients included into the Study

Total number of Patients included	34
Total number of females	9
Total number of males	25
Average age at Diagnosis	57.8
Average age of females at Diagnosis	58.1
Average age of males at Diagnosis	57.1

Cases

Case 1

A 60-year-old white male with history of progressive loss of vision OD for three years with MacTel type 2 was referred to us for a second opinion regarding surgical intervention OD. His Past medical history is notable for hypertension. He presented with a VA of 20/200 OD and 20/40 OS. On 12/21/11 he elected to undergo PPV, no ILM peel/SF6 gas, with post-operative prone positioning for 14 days. On 1/3/12 the gas bubble had dissipated with VA OD 20/100-, however OCT demonstrated persistent and slightly larger macular hole. On 1/25/12 he subsequently underwent PPV, peeling of ILM with ICG and 12% C3F8 gas and was placed for 13 days in prone position, then right-side down for 5 days, followed-by, left-side

down for 4 days, followed by right-side down for 6 days. Post-operatively he developed a retinal detachment underwent PPV, 20% SF6 gas, scleral buckle and EL on 2/22/12. On 3/5/12 patient returned to the operating room for recurrent retinal detachment with proliferative vitreoretinopathy (PVR) and had further surgery including PPV, EL, and membrane peel (MP), and silicone oil. Three weeks post-op patient's VA was 20/400+1. On 5/16/12 patient taken back to the OR for removal of silicon oil (ROSO) and PPV; two weeks post-op, patient's VA was 20/200-1 and remained stable. Hole remains flat but open.

Case 2

A 70 year-old female was referred for decreased vision OU prior to undergoing cataract surgery, VA OD 20/70 and OS 20/40, with progressive vision loss OD with distortion. Her past medical history is notable for Grave's disease without significant thyroid eye disease and diabetes without retinopathy. OCT and FA illustrated Mactel type 2 OU with FTMH OD. On 5/8/13 she elected to undergo PPV with 20% SF6 gas OD with post-operative prone-positioning. Post-operatively the hole OD remained opened and she underwent PPV with ILM peel and C3F8 gas in August 2013. The hole remained open and a 3rd surgery was performed in December 2013 with PPV/phaco/IOL/C3F8 gas and prone positioning for 14 days, however the hole failed to close and has remained open with VA 13 months post-opcounting fingers (CF) at 2 feet.

Discussion

The first time the association of MacTel type 2 with FTMH was made was by Olson and Mandava in 2006, but without the mention of surgical intervention. Since that time, several reports of Mactel type 2 and surgical intervention have been reported and were summarized in there cent paper by Karth et al. in *Retina* 2013 in which the authors reported 4 cases of Mactel with FTMH undergoing surgery. In their report, 2 holes closed, 1 initially closed then re-opened and 1 did not close. In addition, 2 eyes had improved vision, 1 worsened and 1 was unchanged [4]. There are several other surgical cases reported in the literature and include a total of 6 other cases of mactel and FTMH by Rishi and Kothari, Charbel et al., Gregori and Flynn, and Shukla [1,8-11]. Rishi and Kothari reported 1 patient with FTMH who underwent surgical intervention, without hole closer. Charbel et al. reported 2 patients who underwent surgical intervention, and also did not have closure. Gregori and Flynn reported 2 patients with FTMH who underwent surgical intervention, resulting in 1 patient with a closed FTMH and the other patient experiencing initial closure with subsequent opening. Shukla reported 1 patient with FTMH who underwent surgical repair and did not have closure. We report 2 cases with FTMH that underwent surgical intervention, in both cases the holes remained open.

A total of 12 cases (including ours) undergoing surgical repair have been reported (Table 2). In 3 eyes the holes remained closed, 2 eyes closed then reopened, and 7 eyes did not close. The success rate of closure is 25%. As far as vision improvement, 6 eyes overall had vision improvement, 2 had worsening and 4 had no change.

Table 2: Cases of Patients with MacTel Type 2 and FTMH who underwent Surgery

Author	No. Patients	Closed	Closed initially, but reopened	Not closed
Rishi and Kothari [8]	1	0	0	1
Charbel et al. [9]	2	0	0	2
Gregori and Flynn [10]	2	1	1	0
Shukla [11]	1	1	0	0
Karth et al. [4]	4	1	1	2
Casey Eye	2	0	0	2
Total	12	3	2	7

The pathogenesis of MacTel remains unclear. Proposed mechanisms include progressive capillary endothelial cell degeneration, ischemic changes in watershed zones, and primary neurodegenerative process [1]. Koizumi et al. hypothesized that macular hole formation in MacTel may be the result of the separation of neural tissue but that the retained vision in their series suggested that photoreceptor atrophy was not in play [12]. In addition, MacTel has been hypothesized to involve the degeneration of Müller cells and loss of their vascular support. Meaning it is a degenerative condition affecting primarily glial cells and neurons. It is possible that the absence of Müller cells makes surgical closure in these cases less successful when compared with typical idiopathic macular holes where there is no tissue loss [4]. Karth et al. postulate a multifactorial pathogenesis consisting of tissue loss and tractional changes. The appearance of tissue depletion in their cases with unsuccessful closure supports the hypothesis of Powner et al. that foveal tissue (Müller cells) is missing.

Conclusion

The literature suggests that surgery for repair of FTMH in patients with MacTel type 2 may offer some visual improvement in eyes with poor visual acuity or if visual acuity decreases dramatically. Surgical intervention may not be warranted in all cases. It seems as that once the VA has deteriorated and stabilized with the development of a FTMH. This may be attributed to the anatomical and functional outcomes are less successful and this reduced surgical success could be due to the loss of Müller cells. Preoperative appearance of FTMH on OCT may provide prognostic insight into surgical outcomes as noted by Karth et al. They suggested that the appearance and extent of tissue depletion might be associated with post-operative outcomes. Further studies and a better understanding of the pathogenesis may lead us to a more definitive treatment protocol.

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