Long Term Result of Intravitreal Bevacizumab in a Patient Newly Transformed to Proliferative Macular Telangiectasia Type 2

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ABSTRACT

The clinical and imaging findings and therapeutic outcomes of intravitreal bevacizumab injection in a patient with macular telangiectasia type 2 are described. The patient first presented with the non-proliferative stage of the disease for 4 months, then the disease transformed to the proliferative stage. In the proliferative period, the patient was treated with intravitreal bevacizumab injections as-clinically warranted. Over a follow up period lasting 26 months, the patient received 6 intravitreal bevacizumab injections, the visual acuity improved from 20/100 to 20/40, the central retinal thickness decreased from 318 microns to 198 microns. This case implies that the patients with non-proliferative macular telangiectasia type 2 should be followed carefully for proliferative transformation, and intravitreal bevacizumab treatment seems to be effective for proliferative macular telangiectasia type 2.

Key words: Intravitreal Bevacizumab, Macular Telangiectasia, Subretinal Neovascularization

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INTRODUCTION

Macular telangiectasia (MacTel), also known as idiopathic juxtafoveolar telangiectasia, is an uncommon bilateral disease that affects the juxtafoveolar region of the macula.[1] MacTel was first described by Reese in 1956,[1] and was classified into three groups by Gass and Blodi[2] in 1993. MacTel type 1 is unilateral and associated with exudation and macular edema. MacTel type 2 is bilateral and associated with minimal macular edema, deep hyperfluorescence on fluorescein angiography (FA), loss of macular transparency, superficial white crystals, depletion of macular pigment, progressive foveal thinning and edema in the non-proliferative stages, and subretinal neovascularization (SRN) in the proliferative stage. Type 3 is less frequent type and characterized by macular ischemia.[1]

We report a patient who first presented with bilateral non-proliferative MacTel type 2, who showed a proliferative transformation in her left eye, and underwent intravitreal bevacizumab (IVB) treatment as-required with a beneficial outcome.

CASE REPORT

A 47-year-old female was admitted to our clinic with a complaint of decreased vision in the right and left eye (OU) in December 2009. She was in excellent health. On baseline examination, monocular best corrected visual acuity (BCVA) was 20/25 in both eyes. Biomicroscopic anterior segment examination was normal, and intraocular pressure was within normal limits OU. Biomicroscopic fundus examination revealed macular hole like images OU. FA, showed a horse shoe shaped hyperfluorescence on the temporal fovea OU [Figure 1a]. Optical coherence tomography (OCT) showed an internal limiting membrane (ILM) drape OU [Figure 2a]. The central retinal thickness (CRT) was 198 microns and 210 microns, in the right and left eyes respectively. Based on the clinical and imaging findings the patient was diagnosed with MacTel type 2 and advised to present for examinations every 2 months. Four months later, the patient complained of decreased vision in the left eye (OS). On the examination, BCVA was 20/100 OS,

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and fundus examination revealed a grey reflex representing a SRN beneath the fovea. FA showed a subfoveal classic choroidal neovascularization (CNV) associated with leakage OU [Figure 1b]. OCT showed a high reflective area located subfoveally, and associated with intraretinal and subretinal fluid collection, and the CRT was 318 microns OS [Figure 2b]. The patient was diagnosed with proliferative MacTel type 2 OS, and underwent 3 consecutive monthly 1.25 mg IVB injections. After these injections, BCVA increased to 20/25, and both clinical and angiographic features showed significant improvement with minimal leakage on FA and absence of intra- or sub-retinal fluid on OCT examination. The CRT was 258 microns [Figure 2c]. Follow-up examinations were scheduled monthly and treatment regimen of IVB injections as clinically required was planned. Retreatment criteria were determined as; loss of visual acuity of ≥1 line, and detection of any amount of intra- or subretinal fluid on OCT. At month 12, OCT showed subretinal fluid with a CRT of 259 microns OS. BCVA remained stable at 20/25 OS. The patient underwent another IVB injection OS. At month 24, the BCVA decreased to 20/63, and FA showed active leakage around the CNV OS. After the fifth IVB injection, BCVA improved to 20/40 OS. At month 29 as BCVA decreased to 20/126 and OCT showed subretinal fluid, a sixth IVB injection was administered. At month 30, visual acuity improved to 20/40, and OCT revealed a hyperreflective CNV scar located subfoveally, with a CRT of 198 microns OS [Figure 2d]. No adverse events were detected due to IVB injections during follow up. The right eye remained stable with a visual acuity of 20/25 for 30 months, and did not show any signs of proliferative MacTel type 2.

DISCUSSION

Macular telangiectasia type 2 is a symmetrical disease that involves the temporal side of the fovea. Several classifications are described according to clinical, FA, OCT and fundus autofluorescence findings.[1-3] The most popular classifications are the Gass-Blodi[2] and Yannuzzi[3] classifications. MacTel type 2 was divided into 5 stages by Gass and Blodi.[2] In stage 1, a grayish reflex temporal to the fovea may be present on clinical examination, FA is especially important at this stage, and in the late phases, FA shows hyperfluorescence in the perifoveal region. Stage 2 is characterized by the loss of transparency of the retina temporal to the fovea and early staining at the level of outer capillary zone and late staining in the outer retina. In stage 3, FA reveals capillary dilatation, staining and prominent right angled venues. Stage 4 shows migrating RPE cells appearing as stellate foci along the right-angled venules. Stage 5 is associated with SRN.[2-4] Yannuzzi,[3] combined the classification with OCT findings, and Wong[1] added fundus autofluorescence and microperimetry findings to the classification.

In spite of different classifications, MacTel type 2 is mainly divided into 2 stages, in case of treatment. The first stage is the non-proliferative stage, which is characterized by slow, progressive visual loss in association with outer retinal atrophy. The second stage is the proliferative stage, which is characterized by the presence of SRN.[1-4] Several treatment options were studied for non-proliferative stage; however, most were reported to be ineffective because of the degenerative and progressive nature of this disease.[4] In contrast, the proliferative stage was successfully treated with photodynamic therapy, intravitreal anti-vascular endothelial growth factor (VEGF) injections, and combination therapies.[1]

Engelbrecht *et al*.,[5] showed that the natural course of proliferative MacTel type 2 resulted in very poor prognosis including visual acuity of 1/20 or less in 80% of cases.

A retrospective case series by Potter *et al*.,[6] showed that PDT was effective for stabilizing the vision in 5 of 7 eyes. However, improved vision was achieved only in 3 of 7 eyes, and there was no improvement in median visual acuity over a mean follow up time of 21 months.

Anti-VEGF agents,[1] have rapidly become the mainstay of therapy against any type of CNV in various diseases, including proliferative MacTel type 2.[1,7-10] A case series of 6 eyes, reported that median

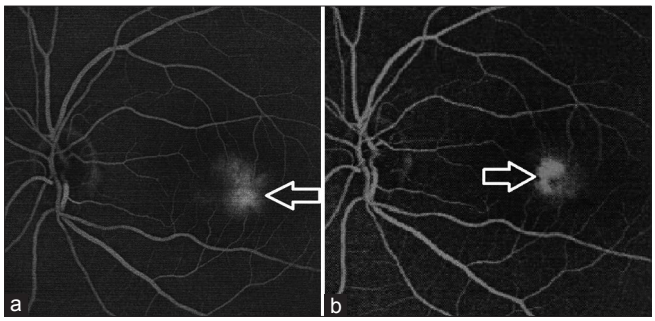

Figure 1: Fluorescein angiography images of the patient (a) horse shoe shaped hyperfluorescence temporal to the fovea (white arrow), (b) subfoveal classic choroidal neovascularization with active leakage (white arrow).

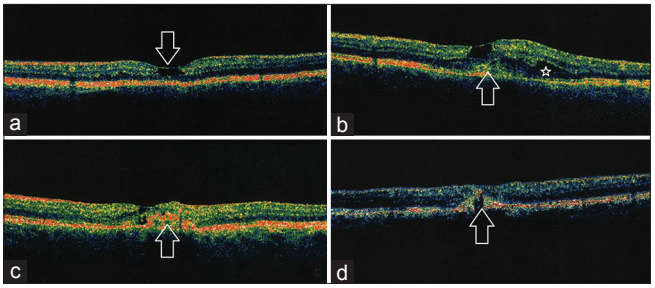

Figure 2: Optical coherence images of the patient (a) Internal limiting membrane drape (white arrow) at baseline, (b) hyperreflective subfoveal classic choroidal neovascularization (white arrow), and subretinal fluid (white star) at the 4th month, (c) resolution of subretinal fluid and small hyperreflective scarring secondary to classic choroidal neovascularization, after the first three intravitreal bevacizumab injections (white arrow), (d) small hyperreflective scarring secondary to classic choroidal neovascularization (white arrow) at the last follow-up visit.

visual acuity improved from 20/200 to 20/100, and the mean CRT decreased from 263 microns to 201 microns over a mean follow up time of 4.2 months.[8] These results were fairly good; however, the follow-up time was very limited.[8] Roller *et al*.,[9] also reported beneficial results of intravitreal bevacizumab in 9 patients. In their[9] study, after a mean follow-up time of 17 months, and mean of 4.9 bevacizumab injections (range 1 injection and 15 injections), the mean visual acuity improved 1.1 lines, and mean central macular thickness decreased by 48 microns.[9] In addition, the visual acuity either remained stable or improved In 7 of 9 eyes (78%), and the CNV resolved completely in 8 of 9 eyes (89%) after treatment.[9]

Karagiannis *et al*.,[10] showed that monthly injections of ranibizumab were effective in a patient with proliferative MacTel type 2. They reported that, the visual acuity improved from 1/20 to 3/10 after 12 monthly intravitreal ranibizumab injections.

In our case, the patient was treated with intravitreal bevacizumab on an as needed treatment regimen. Two different stages of the disease were observed during follow-up. In the first 4 months, the non-proliferative stage of the disease was seen which was followed by a proliferative stage over the following 26 months. The visual acuity decreased from 20/25 to 20/100, and CRT increased by 108 microns while the disease was transforming from the non-proliferative to the proliferative stage. After the intravitreal bevacizumab therapy for the proliferative phase, the visual acuity increased from 20/100 to 20/40, and CRT decreased from 318 microns to198 microns. The visual acuity improved by 4 lines with intravitreal bevacizumab therapy after the transformation of the disease. To our knowledge this is the first reported case of a patient who showed newly proliferative transformation of MacTel type 2, and was immediately treated with intravitreal bevacizumab injections on an as needed treatment regimen associated with beneficial results (based on a PubMed and MEDLINE searches).

In conclusion, non-proliferative MacTel type 2 patients should be informed about the symptoms as decreased vision and metamorphopsia are associated with proliferative transformation, and followed carefully. Intravitreal bevacizumab injections on an as needed treatment regimen may be effective in proliferative MacTel type 2. Although investigation of large series is difficult due to the relative paucity of patients with proliferative MacTel type 2, further studies are required to evaluate the safety and efficacy of intravitreal anti-VEGF agents in this group of patients.

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