Efficacy of Intravitreal Aflibercept as a First Line Therapy in Macular Telengiectasia Type 1

Tip 1 Maküler Telenjiektazili Olguda İlk Seçenek Tedavi Olarak İntravitreal Afliberceptin Etkinliği

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ABSTRACT

We present a 66 year old male patient who was admitted to our clinic with blurred vision in the right eye, which commenced 6 months prior. Ophthalmic examination of the patient revealed type 1 macular telangiectasia (MACTEL). As a first line therapy, two occasions of intravitreal aflibercepts were administered a month apart. For the first and second post-treatment examinations, visual acuity was 0.7 and 1.0 while the thickness of the macula was measured at 295 µm and 254 µm, respectively. At the 12th month post-treatment, visual acuity was stabilized and sub-intra retinal fluid was not detected. This case study suggests that intravitreal aflibercept injection may be an effective therapy for the treatment of macular edema due to type 1 MACTEL.

Key Words: Macular telangiectasia type 1, Aflibercept, Juxtafoveal telengiectasia.

ÖZ

66 yaşında erkek hasta 6 ay önce başlayan bulanık görme ve görme azlığı şikayetleri ile kliniğimize başvurdu. Oftalmoljik değerlendirme sonrası Tip 1 maküler telenjiektazi tanısı konan olguya ilk seçenek tedavi olarak birer ay ara ile 2 kez intravitreal aflibercept enjeksiyonu uygulandı. Enjeksiyon sonrası vizitlerde görme keskinliği snellen eşeli ile 0.4 düzeyinden sırasıyla 0.7 ve 1.0 düzeyine yükselirken makula kalınlığı 425 µm'den sırasıyla 295 µm ve 254 µm'ye geriledi. Tedavi sonrası 12. ay kontrolünde görme keskinliğinin stabil olduğu gözlendi. İntraretinal ve subretinal sıvı izlenmedi. Olgumuz, tip 1 maküler telenjiektaziye bağlı makula ödemi olgularında intravitreal aflibercept tedavisinin etkili bir yöntem olabileceğini göstermektedir.

Anahtar Sözcükler: Maküler telenjiektazi tip 1, Jukstafoveal telenjiektazi, Aflibercept.

INTRODUCTION

Juxtafoveolar retinal telangiectasia is a rare vascular disease in which a dilatation of the capillaries occurs within the fovea. It was first described by Gass and Oyakawa in 1982, while Yanuzzi later classified the disease as type 1 (aneurysmal telangiectasia) and type 2 (perifoveal telangiectasia). The disease generally accompanies inflammatory and vascular pathologies, however idiopathic telangiectasias are also observed.[1] Aneurysmal telangiectasia, also known as type 1 telangiectasia, largely affects males unilaterally.[2] The size of the lesion is approximately 1-2 discs in diameter and tend to involve the temporal side of the macula. Vision loss occurs secondarily to the cystoid macular edema and exudation. Treatment options include laser photocoagulation, photodynamic therapy, intravitreal steroids (triamcinolone and dexametasone implants) and intravitreal anti-VEGF therapy.[3-6] Aflibercept has been used in the treatment of macular edema in diabetic patients, retinal vascular occlusions and age re-

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lated macular degeneration; however, studies about its use for the treatment of retinal telangiectasia remain limited.[7]

To our knowledge, we provide the first evidence for the succesful treatment of type 1 macular telangiectasia using aflibercept injections as a first line therapy.

CASE REPORT

We present a 66 year old male patient who was admitted to our clinic with blurred vision in the right eye, which commenced 6 months prior. The patient did not present with any significant medical history apart from systemic hypertension. In his ophthalmic examination, visual acuity was 0.4 on the right eye and 1.0 on the left eye, while anterior segments were normal. Upon examination of the fundus, we observed exudation and microhemorrhages in the superior-temporal edge of the macula with an optic disc diameter of 0.5 (Fig. 1a). Left fundus appeared normal.

Fundus fluorescein angiography (FFA) showed a diffuse leakage where the exudation exist; this blockage was caused by late stage hemorrhages and cystoid macular edema (Fig. 1b). Optic coherence tomography (OCT) showed intraretinal cysts and neurosensorial detachment (Fig. 1c). Central macular thickness (CMK) was 425 µm in the right eye. With these findings, we diagnosed the patient with type 1 macular telangiectasia and administered two occasions of intravitreal aflibercepts a month apart. In his first examination a month following the first injection, visual acuity was 0.7 and CMK was measured at 295 µm (Fig. 2a). Intraretinal fluid completely receded while subretinal fluid still minimally persisted. In his second follow-up, a month after the second injection, visual acuity improved from 0.7 to 1.0 as assessed through the Snellen scale, while subretinal fluid totally resolved (Fig. 2b). Monthly follow-ups were performed without the need to provide further injections. At the 12th month post-treatment, the patient's VA was stabilized with neither

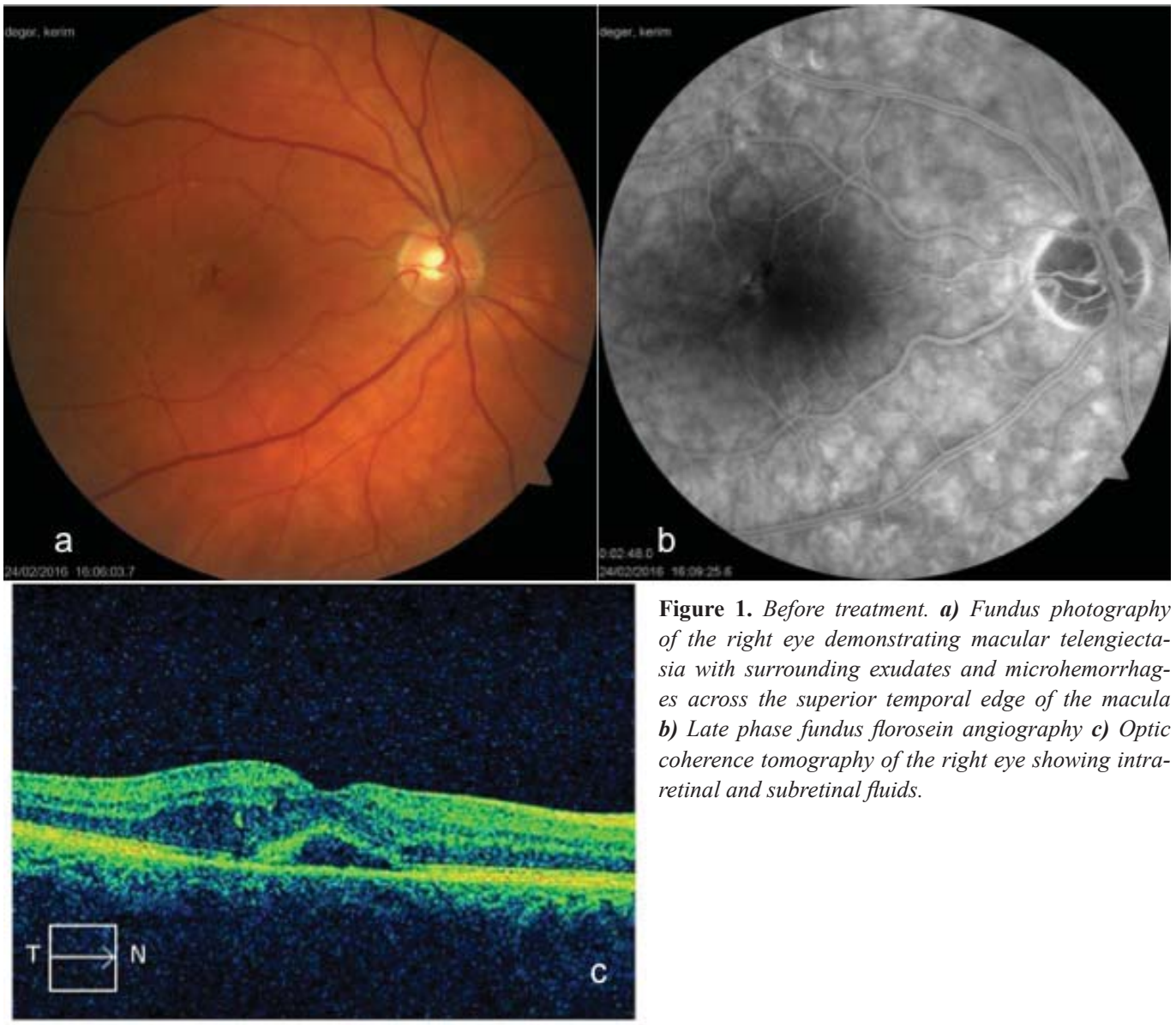


Figure 1. *Before treatment.* ***a)*** *Fundus photography of the right eye demonstrating macular telengiectasia with surrounding exudates and microhemorrhages across the superior temporal edge of the macula* ***b)*** *Late phase fundus florosein angiography* ***c)*** *Optic coherence tomography of the right eye showing intraretinal and subretinal fluids.*

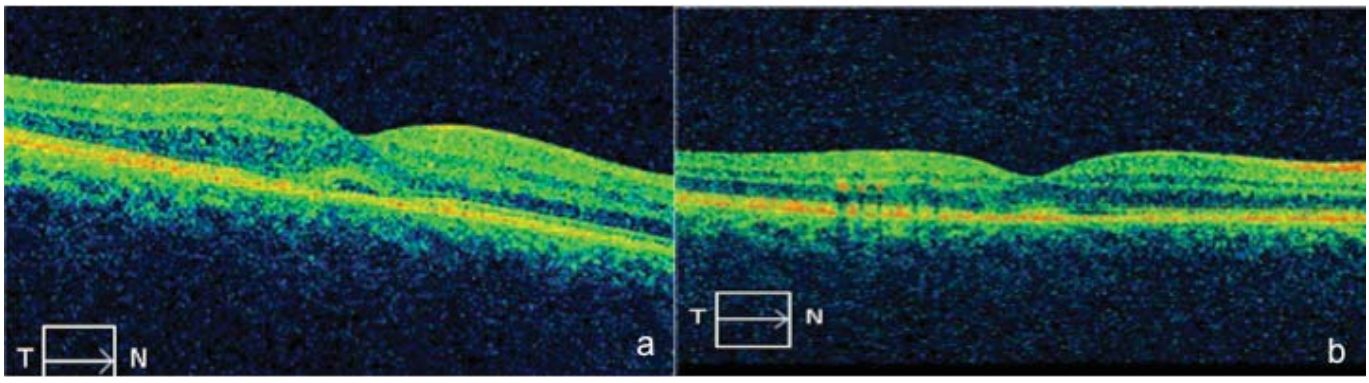


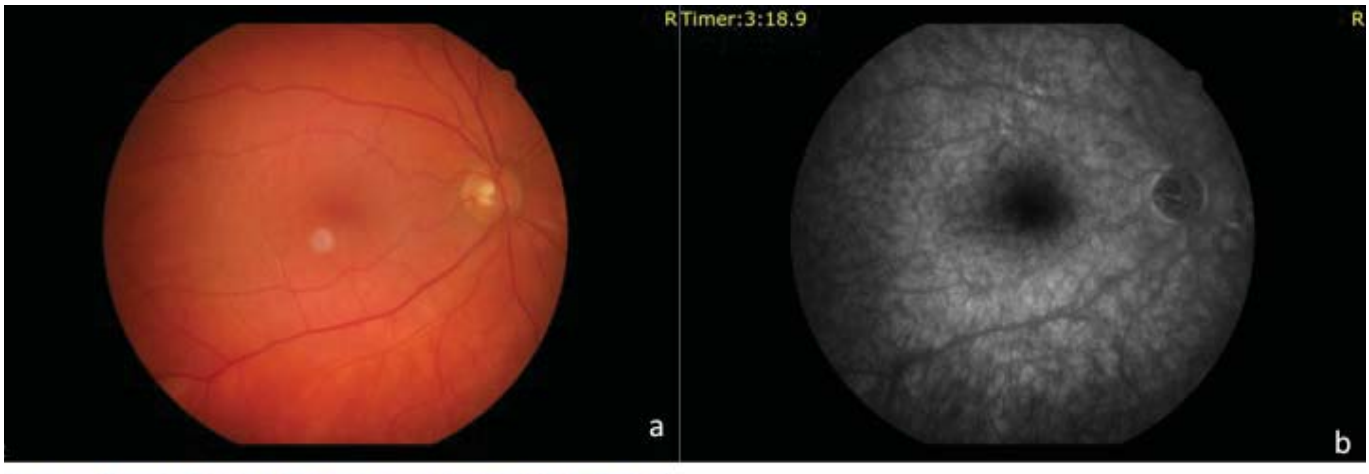
Figure 2. a) *One month following first injection of intravitreal aflibercept, intraretinal fluid completely resolved while subretinal fluid minimally persisted,* ***b)*** *One month following second injection of intravitreal aflibercept, both intraretinal and subretinal fluids thoroughly receded.*

intraretinal nor subretinal fluids detected. There was no observed leakage in the FFA, while all exudation and hemorrhage resolved upon examination of the fundus (Fig. 3).

Discussion.

Despite several studies and case reports conducted on the treatment of type 2 macular telangiectasia, the number of studies regarding type 1 IMT is inadequate. Although laser treatments have been succesful for the improvement of exudations, insufficient recovery in visual acuity and the risk of expanding the laser spot caused a shift towards the use of intravitreal injections.

There were only a few cases in which satisfactory results were obtained for CMT and VA from the use of bevacizumab or ranibizumab.[6,8,9] Two recent cases reported patients who were partially and non-responsive to bevacizumab, and demonstrated favourable results upon switching to aflibercept.[7,10] While ranibizumab and bevacizumab specifically inhibit isoforms of VEGF-A, aflibercept targets VEGF-A, VEGF-B and the plasental growth faktor (PIGF). A murine



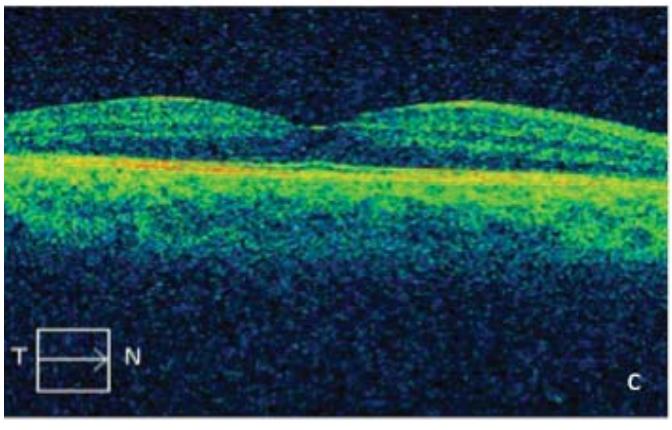


Figure 3. *12 months post-treatment.* ***a)*** *Exudates and microhemorragies were resolved* ***b)*** *Late phase FFA of the right eye show the absence of leakage* ***c)*** *Normal oct findings.*

study showed that PIGF provoked dilatation, tortuosity and aneurysmal formations in retinal vessels,[11] and promoted the effects of VEGF in pathologic angiogenesis.[12] In cases where resistance to bevacizumab and ranibizumab developed, the improvement in anatomic and functional outcomes through the use of intravitreal aflibercept may be due to the inhibition of both VEGF and PIGF.

Another advantage of aflibercept over bevacizumab and ranibizumab is its abilty to bind endothelial cells, pericytes and vascular basal membranes.[13] Considering the underlying pathology of aneurysmal telangiectasia, the impact of aflibercept on vascular events may have played an important role on the treatment.

In conclusion, intravitreal aflibercept injections have been effective for the treatment of a patient with macular edema, which developed from a type 1 MACTEL, and the beneficial effects were observed to last for 12 months. Although aflibercept was successfully used as a promising therapeutic agent, studies with larger populations are needed to elucidate the effects of aflibercept on type 1 MACTEL.

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