



Treatment of intravitreal bevacizumab combined with focal laser photocoagulation in the case of macular telangiectasia type 2 with retinal arterial macroaneurysm

Gökhan Demir¹ · Özgür Artunay¹ · Mehmet Emin Sucu¹ · Ali Demircan¹ · Dilek Yaşa¹ · Cengiz Alagöz¹ · Mevlüt Celal Öcal¹

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Abstract

Macular telangiectasia type 2 (Mac Tel 2) is a bilateral disease of unknown cause with characteristic changes of the retinal vasculature. To the best of our knowledge, we could not find any reported cases of Mac Tel 2 with retinal arterial macroaneurysm (RAM). Our aim is to report a case of Mac Tel 2 with RAM.

Keywords Macular telangiectasia · Retinal arterial macroaneurysm · Focal laser photocoagulation

Introduction

Idiopathic juxtafoveolar retinal telangiectasia (IJRT) is a rare retinal vascular malformation with tortuous, dilated, bloodfilled retinal vessels present in the inner nuclear layer and extend into the outer plexiform layer. The clinical and angiographic features and classification were first described by Gass and Oyakawa in 1982 [1]. Yannuzzi et al. added the optical coherence tomography (OCT) features to a modification of the original classification by Gass and divided IJRT into two groups: aneurysmal telangiectasia or idiopathic macular telangiectasia type 1 and perifoveal telangiectasis, also known as idiopathic macular telangiectasia type 2 (Mac Tel 2) [2].

Mac Tel 2 is a bilateral condition that usually presents during the fifth and sixth decades of life. The earliest fundoscopic manifestation of Mac Tel 2 is the loss of retinal transparency in the perifoveal region, beginning temporally and later surrounding the fovea. Parafoveal telangiectasia may not be visible clinically in early stages of the disease but can often be demonstrated more readily by red-free photography. Optical coherence tomography (OCT) findings are the formation of hyporeflective inner retinal spaces, disruption of the

photoreceptor inner-segment–outer-segment junction, plaques of pigment epithelium hyperplasia, and pseudolamellar macular holes. Metamorphopsia or a scotoma may be present in the early stages of the disease [3]. Visual impairment may be mild; however, visual acuity can be deteriorated if choroidal neovascularization (CNV) supervenes.

Retinal arterial macroaneurysm (RAM) is an acquired large arteriolar dilation that usually occurs within the first three orders of the retinal arterial vasculature bifurcations, typically in the macular or postequatorial regions [4]. This condition may cause visual deterioration secondary to macular edema or hemorrhage in any retinal layers [5].

We report a case of 77-year-old male having both idiopathic macular telangiectasia type 2 and RAM, and was treated with combination of intravitreal bevacizumab and laser photocoagulation.

Case report

A 77-year-old male was seen in January 2017 for a routine eye examination. Best corrected visual acuity (BCVA) was 20/25 in his right eye (OD) and left eye (OS). Intraocular pressure (IOP) was 15 mmHg in each eye and slit-lamp examination was unremarkable in both eyes. Dilated fundus examination of the OD revealed a large arterial macroaneurysm in the superior perifoveal region and multiple telangiectatic vessels in the parafoveal region. Fundus of the OS showed no abnormal findings (Fig. 1a–b). OCT

✉ Gökhan Demir
dr.gkhndmr@gmail.com

¹ Beyoglu Eye Training and Research Hospital, The University of Health Sciences, İstanbul, Turkey

and fundus fluorescein angiography (FFA) were performed. OCT of the OD revealed a prominent hyporeflective space nasal to the fovea and retinal thickening corresponding to the area leaking in the superionasal region of the macula. OCT of the OS was normal (Fig. 1c–e). FFA of the OD demonstrated telangiectatic and dilated parafoveal capillary network just superionasal to the fovea and a macroaneurysm on the superiotemporal arterioli that typically fills in the early arterial phase of the angiogram. FFA of the OS showed telangiectatic vessels in the temporal parafoveal region. Telangiectatic vessels were more common in the OD than in the OS in FFA (Fig. 2a–b). Asymmetric involvement was observed. Fundus autofluorescence (FAF) showed bilateral loss of the normal central attenuation. The area of late diffuse hyperfluorescence corresponded with the area of increased autofluorescence, which suggested the diagnosis of Mac Tel 2. Spectral domain OCT (SD-OCT) using horizontal scans showed macular abnormalities compatible with the presumed diagnosis. Additionally, the systematic story revealed that the patient had hypertension. Since BCVA of the patient was good and there was no visual complaint, the patient was decided to follow up without treatment. The patient was seen 2 months later and BCVA decreased 20/30 in the OD. OCT of OD showed an increase of parafoveal intraretinal cyst at the size and of retinal thickening due to leakage of macroaneurysm (Fig. 3a–b). The patient was treated with one injection of intravitreal bevacizumab (Avastin,

Genentech, Inc., South San Francisco, CA; 1.25 mg/0.05 cm³) followed by closure laser photocoagulation due to deterioration in BCVA and OCT findings of OD. The interval between injection and laser was 1 week. Partial response to the treatment was gained based on the OCT findings of OD. It was planned to apply one more injection of intravitreal bevacizumab 3 weeks later in order to acquire a complete recovery of OCT findings of OD. Focal green laser photocoagulation of 532 nm (Topcon Pascal Streamline Laser System) was applied on macroaneurysms. Threshold laser was performed around the macroaneurysm with confluent pattern. The subthreshold laser was applied to the macroaneurysm to facilitate thrombosis and stop active bleeding until the laser spot was barely visible. It was seen that improvement in BCVA, disappearance of intraretinal cyst detected in previous OCT, and a decrease of retinal thickening due to leakage of macroaneurysm were seen after the treatment (Fig. 4a–b). The patient was seen at 2 months intervals for 10 months after the treatment, with no change in his clinical presentation.

Discussion

Mac Tel type 2 is a rare and potentially blinding macular disease usually diagnosed during the fifth to seventh decades of life [6]. The maculopathy manifests as a slowly progressive decrease in visual acuity, which may occur when degenerative

Fig. 1 **a.** Color fundus picture of OD showing slightly dilated right-angle veins with an arterial macroaneurysm. **b** Color fundus picture of OS showing no abnormal findings. **c** Macula OCT scan of OD showing intraretinal hyporeflective space. **d** Retinal thickening due to leakage of macroaneurysm. **e** Normal macula OCT scan of OS

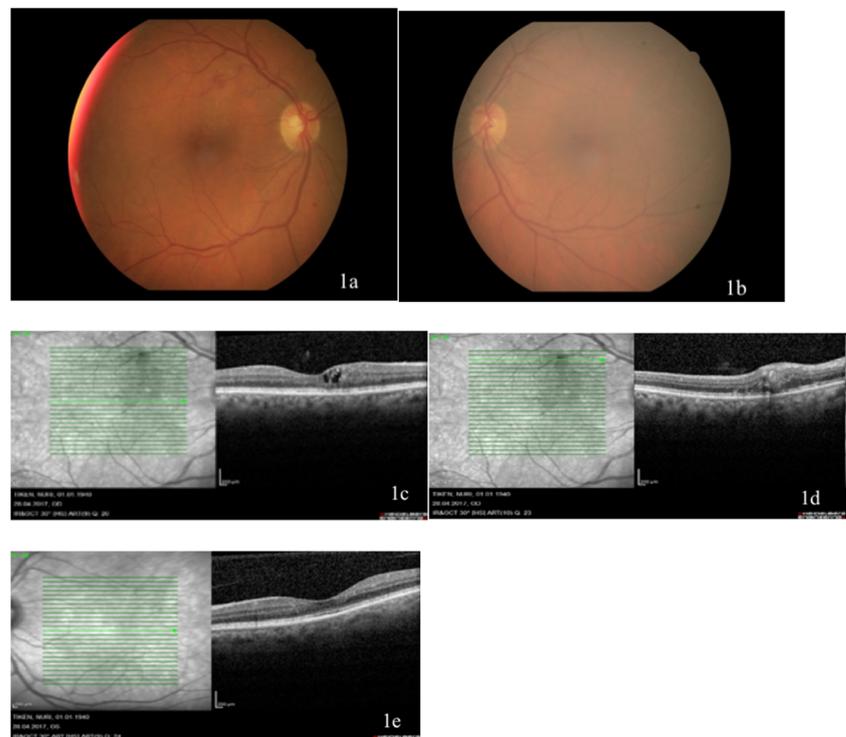
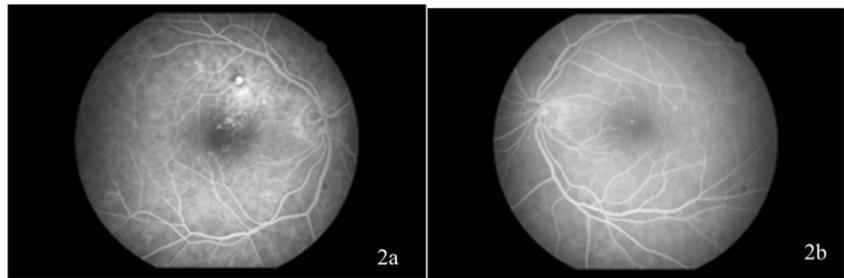


Fig. 2 a. FFA of the OD demonstrated telangiectatic and dilated parafoveal capillary network just superioronasal to the fovea and a macroaneurysm on the superiotemporal arterioli. **b** FFA of the OS showed telangiectatic vessels in the temporal parafoveal region



changes or neovascularisation and subsequent scar formation reach the foveal center [1]. Although the disease is bilateral, it can manifest asymmetrically, with the fellow eye seemingly unaffected and showing only very subtle anomalies on SD-OCT, autofluorescence and fluorescein angiography [7]. The main limitation in the treatment of Mac Tel 2 is the lack of knowledge of the basic pathophysiologic mechanisms underlying this condition. FA was the earliest imaging modality used to study this disease. Because the typical angiographic findings suggested the presence of macular edema, it was thought that Mac Tel 2 was a primary retinovascular disease. Therefore, the same treatment modalities used to treat retinovascular diseases, such as macular laser photocoagulation [8], photodynamic therapy [9], and intravitreal anti-vascular endothelial growth factor (anti-VEGF) agents [9], were used to treat Mac Tel 2. None of these were particularly beneficial in eyes with nonproliferative Mac Tel 2. Even though there are some case reports showing beneficial effect of the intravitreal bevacizumab in nonproliferative Mac Tel 2, it is not proven in prospective studies [10].

The treatment of retinal arterial macroaneurysm with macular edema or hemorrhage is controversial. Although there is no consensus about the management of retinal arterial macroaneurysm, general guidelines state that unruptured RAMs should be observed [11] and treatment is advised in cases of exudative manifestations involving the fovea with visual acuity deterioration [12]. Treatment options include observation, laser, intravitreal anti-vascular endothelial growth factor (VEGF), YAG laser hyaloidotomy, and vitrectomy.

Although a few studies showed no difference in clinical outcome between observation and laser therapy for RAM, most researchers favored laser treatment due to high efficacy and few complications. Previous reports noted that visual

gains were significant after threshold or subthreshold laser photocoagulation for patients with RAM [13]. The authors preferred a more gradual closure of the macroaneurysm with multiple treatment sessions instead of a more intense whitening that was used for the microaneurysms for fear of bleeding from the macroaneurysm.

RAMs are saccular and fusiform in shape. Lavin et al. categorized RAMs as the following: hemorrhagic, exudative, and quiescent [14]. The type seen in our case was saccular and exudative form. This is more evident in hypertensive patients due to increased hydrostatic pressure [15].

The occurrence of both Mac Tel 2 and RAM in a patient at the same time has not been described in the literature. This occurrence is likely, because hypertension plays a common role in the pathogenesis of both Mac Tel 2 and RAM. Furthermore, asymmetric presentation of Mac Tel 2 is rarely seen. Our case had asymmetric presentation of Mac Tel 2 as well as hypertension.

In our case, because of a good vision and lack of any visual symptoms, follow-up without treatment was decided. During the follow-ups, the patient's visual acuity decreased. OCT showed an increase in the size of retinal hyporeflective space and of retinal thickening due to leakage of macroaneurysm. For these reasons, we decided to apply intravitreal bevacizumab and focal laser photocoagulation. We started the treatment with intravitreal bevacizumab in order to decrease retinal thickness and to increase effectiveness of focal laser photocoagulation. The impact of intravitreal bevacizumab in the resolution of macular edema due to RAM has been shown in the previous studies [16, 17]. Four weeks after the second injection, the hyporeflective space had completely disappeared and the leakage of macroaneurysm decreased.

Fig. 3 a Macula OCT scan of OD showing intraretinal hyporeflective space increasing. **b** Retinal thickening due to leakage of macroaneurysm before the treatment

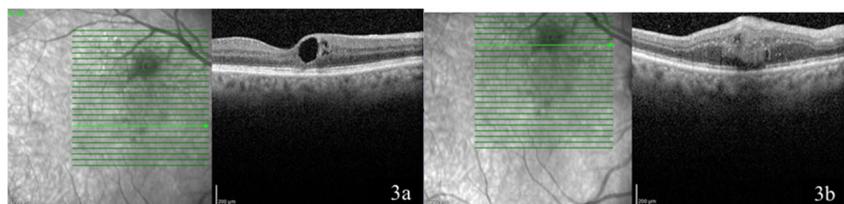
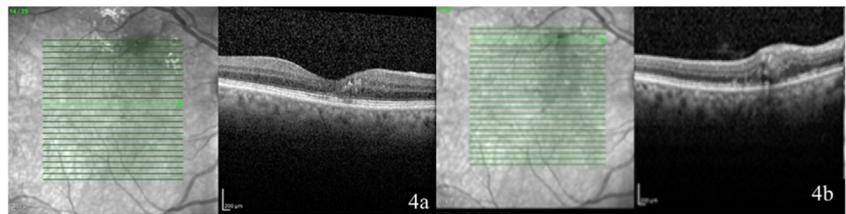


Fig. 4 **a** Disappearance of intraretinal cyst detected in previous OCT. **b** Decrease of retinal thickening due to leakage of macroaneurysm after the treatment



In conclusion, to the best of our knowledge and a detailed literature search, we could not find any reported cases of Mac Tel 2 with RAM. Focal laser photocoagulation was helpful for managing leaky RAM. An intravitreal bevacizumab injection may quickly reduce both the leakage caused by RAM and Mac Tel 2.

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