Idiopathic macular telangiectasia type 1 treated with intravitreal ranibizumab and laser photocoagulation

Abstract

Introduction: Idiopathic Macular Telangiectasia (IMT) are a rare group of various entities presenting with incompetence, ectasia and/or irregular dilations of the juxtafoveal capillaries of one or both eyes.

Case Report: A 62 year old female with Glaucorna was sent to the Retina Department with a suspected diagnosis of retinal vein occlusion on the left eye (LE). Her best corrected visual acuity (BCVA) was 20/20 in the right eye (RE) and 20/25 in the LE. Slit lamp examination was normal; fundoscopy showed a cup-to-disc relation of 0.6 in the RE with normal macula and a cup-to-disc relation of 0.7 in the LE with multiple perifoveal microaneurysms, inferior temporal hard exudates and edema comproved with macular optical coherence tomography; fluorescein angiography revealed an area of hyperfluorescence with juxtafoveal telangiectatic vessels located inferiorly and temporally to the fovea, with progressive filling and late active leakage in the LE. The diagnosis of IMT type 1 was made and she was treated with a loading dose of intravitreal injections of ranibizumab followed by a fourth injection because of persistent macular edema. One month after, her BCVA was 20/20 in the LE with improvement of macular edema and macular árion laser photocoagulation was performed. Three months after the laser treatment her LE BCVA was 20/20 with fundoscopy showing small macular hemorrhages and microaneurysms.

Conclusion: Macular edema and exudation are the main cause of visual loss in these patients. This case suggests that ranibizumab and laser photocoagulation can be effective in the treatment of type 1 IMT.

Key Words: Retina; Idiopathic Juxtafoveal Retinal Telangiectasia; Macular Edema; Intravitreal Injections;

Introduction

Idiopathic Macular Telangiectasia (IMT) are a rare group of various entities presenting with incompetence, ectasia and/or irregular dilations of the juxtafoveal capillaries of one or both eyes, usually located temporally to the fovea. The term was first proposed by Gass and Oyakawa in 1982 based on clinical and fluorescein angiographic (FA) features.1 Later in 1993 Gass and Blodi made an update of this classification subdividing IMT into 3 groups (group 1, group 2 and group 3) with 2 subgroups (A or B) based on clinical and demographic characteristics.2 Recently Yannuzzi et al simplified this model proposing 2 distinct types, based on angiographic imaging and optical coherence tomography (OCT): type 1 or aneurysmal telangiectasia and type 2 or perifoveal telangiectasia.3 This condition should be differentiated from similar patterns such as venous occlusive diseases, diabetic retinopathy, Eales disease, radiation retinopathy, dilation of perifoveal capillaries associated with vitreous cellular infiltration or secondary to inflammatory diseases.2,3,5

Type 1 or aneurysmal telangiectasia involves unilateral telangiectasia mostly in men and is not related with systemic disease. Characteristically, multiple venular and arteriolar aneurysmal telangiectasia are mainly localized in the paramacular or juxtafoveal area, affecting deep and superficial circulations with lipid deposition. Complementary examination shows capillary ischemia with leakage into the retina, cystoid macular edema and no evidence of neovascularization.3,4 Macular edema and exudation are the main cause of visual loss and various treatment options have been used as laser photocoagulation, intravitreal steroids and antivascular endothelial growth factor (anti-VEGF) agents.3,4 Since it is a rare condition there is no sufficient data to guide for the best treatment approach.
Case report

A 62-year-old female, followed at the Glaucoma Department for Primary Open Angle Glaucoma and treated with combined travoprost with timolol in both eyes, was sent to the Retina Department due to retinal findings. She had a suspected diagnosis of retinal vein occlusion on the left eye (LE) due to the presence of hemorrhages and hard exudates in the macular area. There was no other ocular history and systemic history included a knee and gynecological surgery.

On the first visit at the Retina Department her best correct visual acuity (BCVA) was 20/20 in the right eye (RE) and 20/25 in the LE. Slit lamp examination was normal and intraocular pressure (Goldmann tonometry) was 15 mm Hg in the RE and 16 mm Hg in the LE, under treatment. Fundoscopy showed a cup-to-disc relation of 0.6 in the RE with normal macula and a cup-to-disc relation of 0.7 in the LE with multiple perifoveal microaneurysms with edema and inferior temporal hard exudates (Figure 1A). Macular OCT (Cirrus HD-OCT®, Zeiss) showed intraretinal edema with a central macular thickness (CMT) of 461 µm (Figure 1B, 1C, 1D). FA was performed and revealed an area of hyperfluorescence with juxtafoveal telangiectatic vessels located inferiorly and temporally to the fovea, with progressive filling and late active leakage, only in the LE (Figure 2). Based on clinical, tomographic and angiographic features the diagnosis of IMT type 1 or aneurysmal telangiectasia in the LE was made. The patient was treated with a loading dose of ranibizumab (0.5 mg/0.05 ml intravitreal injections in 3 consecutive months). After the loading dose an improved but persistent macular edema was observed on the macular OCT (Figure 1F, 1G, 1H) and the patient was submitted to a fourth ranibizumab injection. One month later her BCVA was 20/20 on the LE and funduscopy showed inferior temporal small hemorrhages and hard exudates, with improvement of the macular edema on the OCT (Figure 1J, 1K, 1L). One month later macular Argon laser photoocoagulation in a grid pattern was performed on the perifoveal area respecting the central avascular zone, using the

The authors present a case of a female affected by aneurysmal telangiectasia who was treated with intravitreal injection of ranibizumab followed by laser photoocoagulation.
following parameters: 15 spots, 50μm of diameter, 100mW of power and 0.1 s of time. Three months later, on the last observation, LE BCVA was 20/20 with fundoscopy showing small macular hemorrhages and microaneurysms and a slight increase on CMT of 328 μm on the OCT.

**Discussion**

The treatment of type 1 IMT remains challenging as no established treatment exists; as the vision loss in these patients results from serous and lipid deposition in the macula, this remains the target of the treatment. The gold standard treatment is until now the macular photocoagulation, usually in a grid-pattern, that seems to be successful in the resolution of exudation with VA improvement.5,6

Intravitreal injections of 4mg of triamcinolone acetonide (IVTA) were reported without great success. It has been useful in macular edema resolution due to its well known anti-inflammatory effect, stabilization of the blood-retinal barrier and inhibition of vascular endothelial growth factor (VEGF) action.4,5,7 To overcome the IVTA short-lived effect, it was recently described the use of dexamethasone intravitreal implant to treat macular edema related to IMT, showing good anatomical results with greater periods without recurrences.4 Nevertheless, both systems have important complications that include elevated intraocular pressure and the development of cataract. Those side effects were taken in account on the treatment approach in our clinical case, since it was a phakic patient with Glaucoma.

Based on the anti-VEGF agents efficacy to treat macular edema from diabetic retinopathy or retinal venous occlusion and also the documented improvement of visual and anatomical function in cases of type 2 IMT, intravitreal injections of these agents are now becoming an option to treat patients with type 1 IMT.4,5,8,9 There are some studies with bevacizumab, a humanized monoclonal antibody target against pro-angiogenic circulatory VEGF, but the functional and anatomical results are inconsistent.4,5,10 According to our knowledge there’s only two reports about the use of ranibizumab, a monoclonal antibody fragment that targets all VEGF-A isoforms, in type 1 IMT. Rouvas et al described two eyes treated with intravitreal injection of ranibizumab with a follow-up of twelve months, being re-treatment criteria the persistence or relapse of leakage combined with VA deterioration.8 One month after ranibizumab injection both eyes showed reduction of hyperfluorescent dots in early phase angiograms with a decreased leakage activity in the late phase; as in one of the eyes BCVA improved without deterioration and with no signs of disease activity, a second injection was not performed. After twelve months of follow up, both eyes presented stable BCVA with slight increase of leakage in FA late phase images.8 The authors concluded that the use of ranibizumab in IMT can reduce vascular leakage and suggested that improvement of VA depends on the structural abnormality at the level of inner/outer segments (IS/OS) of the photoreceptor layer, also supported by other authors.6,8 Ciarnella et al described an eye treated with one intravitreal injection of ranibizumab followed by laser photocoagulation. They concluded that ranibizumab was effective in reducing the macular edema leading to the improvement of VA, and that laser resolved the exudative retinopathy.9 Last year, Shibeeb et al reported an eye initially treated with a loading dose of bevacizumab followed by macular photocoagulation. Because of partial improvement of macular edema the patient was treated with five injections of aflibercept. This article contributes to demonstrate that anti-VEGF agents appear to be an option in the treatment of this condition.

In the authors clinical case it was performed a loading dose of ranibizumab followed by another injection combined after with laser photocoagulation. Anti-VEGF injection reduced the macular edema and exudation and allowed a better effect of the laser treatment. This case suggests that ranibizumab and laser photocoagulation can be effective in the treatment of type 1 IMT.

**REFERENCES**