

Idiopathic Macular Telangiectasia Associated with Large Soft Drusen and Retinal Angiomatous Proliferation

Claudio Campa^{1*}, Perri Paolo¹

1. Sant'Anna University Hospital, Ferrara, Italy

Abstract:

Idiopathic macular telangiectasia type 2 is a rare retinal condition usually characterized by parafoveal telangiectatic vessels, hyperplastic pigment plaques and seldom a full thickness macular hole or a retinal neovascularisation. Herein we describe an atypical presentation of this disease.

Methods

Review of the patient's clinical and angiographic records.

Results

A 87-year-old white man was diagnosed with idiopathic macular telangiectasia type 2. Ophthalmological and angiographic examination disclosed telangiectasia of the parafoveal capillaries together with extensive bilateral granular soft drusen and unilateral retinal angiomatous proliferation (RAP). Initially photodynamic therapy with verteporfin (PDT) and subsequently intravitreal injections of bevacizumab were used to treat RAP lesion. This approach provided visual acuity stabilization up to 24 months.

Conclusion

This case of ours shows that drusen may be a feature of macular teleangiectasia type 2. It also emphasizes that PDT and anti-VEGF therapy are both safe and effective for treating neovascular complications of this disease.

Corresponding Author: Claudio Campa, Eye Clinic, Sant'Anna University Hospital - Ferrara, Tel: +39 (0)532 236814, Fax: +39 (0)532 247365, Email: claudio.campa@unife.it

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Introduction

Macular telangiectasia type 2 is a retinal vascular disorder characterized by crystalline intra-retinal deposits in the early stages and telangiectatic vessels and hyperplastic pigment plaques in advanced stages. Herein we describe an atypical presentation of the disease, with bilateral drusen and a neovascular lesion resembling a retinal angiomatous proliferation.

Case

Idiopathic juxtafoveal retinal telangiectasis (IJT) was a term proposed by Gass and Oyakawa¹ to describe a retinal vascular disorder characterized by dilated retinal capillaries around the foveal avascular zone without any known cause in contrast to secondary telangiectasia, for example, associated with venous thrombosis, vasculitis or radiation retinopathy. These authors¹ divided patients with IJT in 4 groups according to different clinical and angiographic characteristics. The classification was updated by Gass and Blodi² in 1993 identifying only 3 groups with dissimilar biomicroscopic and fluorescein angiographic findings. Finally, Yannuzzi et al.³ proposed a new classification termed idiopathic macular telangiectasia in 2006. They simplified the description of this ocular disease recognizing 3 types. Type 2 macular telangiectasia is the most common condition and is always bilateral although sometimes asymmetrical. Early clinical manifestations consist mainly of a greyish appearance of the parafoveal retina and crystalline intra-retinal deposits⁴. Telangiectatic vessels usually become clinically evident in more advanced stages when also hyperplastic pigment plaques, a full thickness macular hole and retinal neovascularisations may occur⁴⁻⁵.

This case report describes the clinical, ophthalmological and angiographic findings of a patient with idiopathic macular telangiectasia type 2 and unilateral retinal angiomatous proliferation undergone initially photodynamic therapy and subsequently anti-

angiogenic treatment with bevacizumab.

A 87-year-old white man presented with six-months-history of decreased vision and metamorphopsia in his left eye. His past medical history was positive only for angina. He had been diagnosed with open angle glaucoma 10 years before presentation and was treated with brinzolamide 1% and timolol 0.25% guttae in both eyes. He had undergone cataract surgery in both eyes 1 year before presentation. On examination best corrected visual acuity was 6/9.5 (71 Early Treatment Diabetic Retinopathy Study - ETDRS letters) in the right eye and 6/30 (51 letters) in the left eye. Ophthalmoscopy showed evidence of bilateral large granular confluent soft drusen resulting in crescentic and sinuous shapes. In addition, some capillary aneurysmatic changes together with subretinal plaques of pigmentation were detected in both eyes. Fluorescein angiography revealed retinal-retinal anastomosis and telangiectasia of the parafoveal capillaries with intraretinal leakage from these capillaries in later frames in both eyes. There was also punctate hyperfluorescence associated with the numerous basal laminar drusen in both eyes (figure 1 a-d). Moreover, in the left eye, an additional late leakage due to a vascularized pigment epithelium detachment was detected. Indocyanine angiography (ICGA) confirmed the diagnosis of bilateral idiopathic macular telangiectasia type 2 complicated by a multifocal retinal angiomatous proliferation (RAP) stage 3 in the left eye showing two hot spots in the left eye (figure 1f). RAP stage 1 was excluded in the right eye because ICGA in that eye was normal (figure 1e) and is well known that hot spot sign is crucial in the differential diagnosis because it can be observed in any stages of RAP. The patient was treated in his left eye initially with photodynamic therapy with verteporfin (Visudyne; Novartis Pharma AG, Basel, Switzerland) using standard parameters (PDT). Two PDT treatments (at baseline and 6 months later) were able to stabilize his condition for

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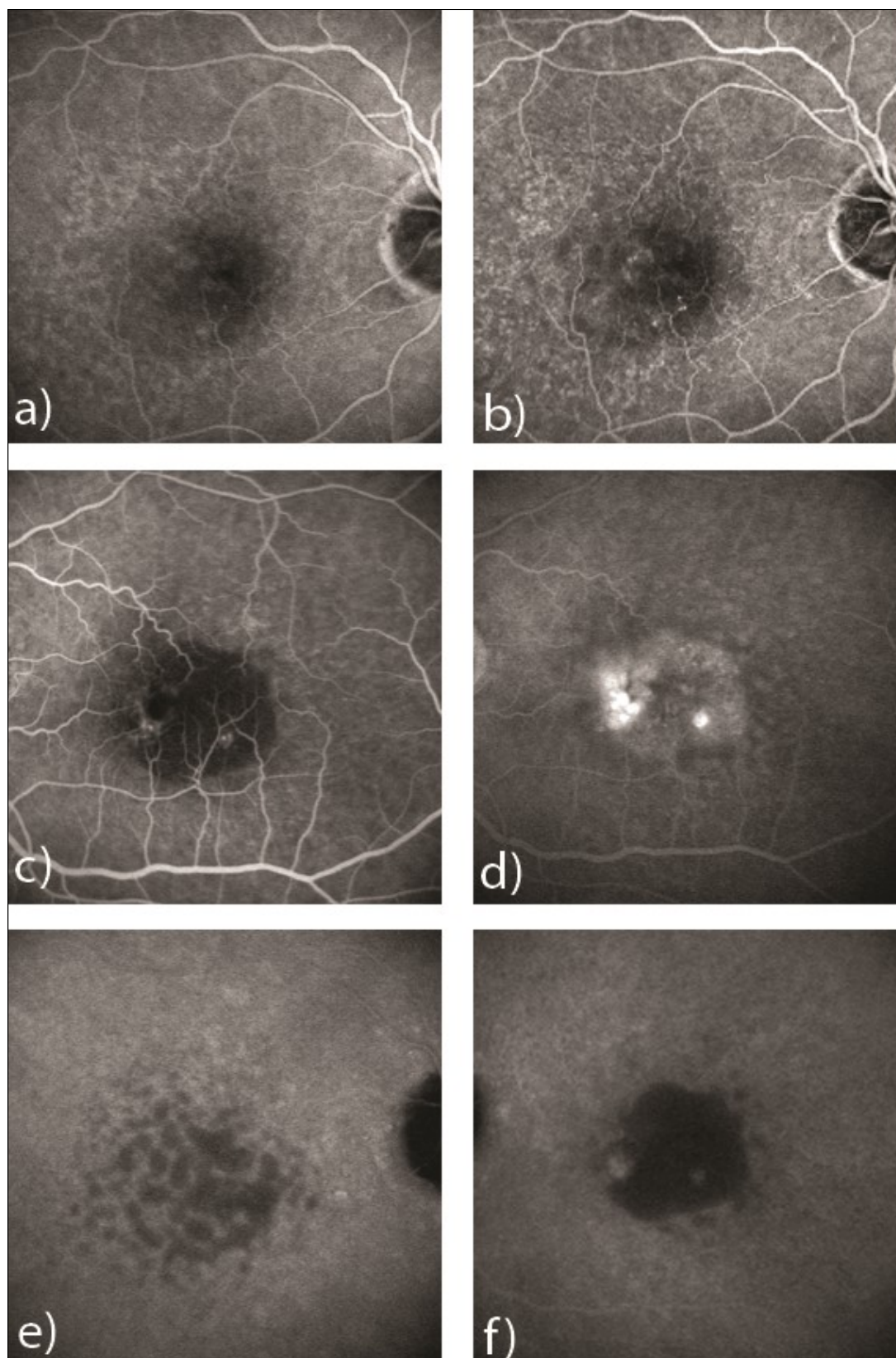


Figure 1 a) early phases fluorescein angiography of the right eye at the time of diagnosis; b) early phases fluorescein angiography of the right eye 6 months later; c) early and d) late phases fluorescein angiography of the left eye at the time of diagnosis; late phases indocyanine angiography of the right (e) and the left (f) eye

15 months. At the 18th month visit however, because of further visual acuity deterioration (down to 49 letters) and the recurrence of leakage at the fluorescein angiography, the decision was made to switch to intravitreal bevacizumab injections (0.5mg, Novartis, Novartis Pharma AG, Basel, Switzerland). The patient received 3 consecutive injections at 4-weekly intervals. At the final follow-up examination, 6 months after the beginning of anti-angiogenic therapy and 24 months after the initial presentation, the visual acuity remained stable (51 letters) and no clinical signs of active lesion (fluid at the OCT, leakage at the angiogram) were detected. Also the fellow eye didn't show any change during the entire follow-up.

Idiopathic macular telangiectasia type 2 is a rare condition normally causing a slow decrease in vision, metamorphopsia and positive scotoma¹⁻⁴. However in a subset of patients, a proliferative stage with secondary neovascularisation may develop later on producing an acute vision loss.

Typical findings of this disease include bilateral perifoveal telangiectasis, minimal exudation, superficial retinal crystalline deposits and right-angle venules¹⁻⁵. Gass and Oyakawa¹ also noted that sometimes a yellow central spot may be found but only two cases of macular telangiectasia type 2 associated with frank bilateral drusen has been previously reported⁶⁻⁷. The case described by Margalit et al.⁶ is represented by a 48 year old woman with evidence of basal laminar drusen in both eyes and a vitelliform lesion and parafoveal telangiectasia only in the right eye.

The case of ours is of interest because shows the presence of bilateral extensive drusen and macular telangiectasia plus a RAP lesion in one eye. Also the age of the present patient is significantly higher compared to mean age reported in previously published case series. Although we can not ascertain when the disease first developed in this patient, however this case could demonstrate that macular telangiectasia can be

asymptomatic and have an excellent VA over a long period of time.

Subretinal neovascularization represents a well recognized stage of macular telangiectasia. However a retinal-choroidal anastomosis occurs infrequently in this disease⁸. Even rarer is then the finding of a retinal pigment epithelium detachment³. For these reason we preferred classifying the neovascular lesion detected in our patient as a RAP instead of proliferative stage of macular telangiectasia. Besides Yannuzzi et al.³ have underlined that these two forms of neovascularization share some similarity. Indeed we believe that in certain stages these two entities are difficult to distinguish and in any case of RAP lesions one should be aware of macular telangiectasia because it might respond differently to treatments.

The good response of our patient initially to PDT and subsequently to anti-VEGF treatment may encourage the use of both these therapies in the management of the proliferative forms of macular telangiectasia. Our results are indeed in accordance with others authors⁹⁻¹⁰ who found PDT and anti-VEGF therapy both safe and effective for patients with this condition.

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