

## RETINAL ANGIOMATOUS PROLIFERATION CASE REPORT

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### Abstract

The diagnosis of RAP is similar with the diagnosis of the AMD, but PED, exudate and superficial hemorrhages are more common in RAP.

A 75-year-old male presented himself at the Ophthalmology Department of the Emergency County Hospital in Craiova in January 2015, with a 4 months history of vision loss. In his right eye the visual acuity was 4/50 eye non-optical correctable and 5/5 with optical correction in the left eye.

According to the FA and OCT that were performed in both eyes, in the right eye was found intraretinal neovascularization and sub and intraretinal fluid. A normal aspect was found in the left eye. After the investigations we decided to start the treatment consisting in one single intravitreal injection with Triamcinolone Acetonide (IVTA) in the right eye. The VA improved 1 week after the treatment from 4/50 to 5/30. In comparison with the improved VA, the macular edema gradually resorbed 1 month after the IVTA injection.

In spite of the late presentation of the patient in the Ophthalmology Department, the VA and OCT outcome after a single dose of IVTA injection was very good.

**Key words:** Retinal angiomatic proliferation, Triamcinolone acetonide, Age related macular degeneration AMD, Intravitreal injection.

### Introduction

Retinal angiomatic proliferation (RAP) is defined as a variant of exudative age related macular degeneration (AMD) in which the retinal-choroidal neovascularization is characterized by intraretinal capillary proliferation or, if the origin of the process is in the choroid, then it is an early retinal-choroidal anastomoses (RCA) without the presence of a neovascular membrane type 1. Although dry AMD represents the majority of all diagnosed cases, wet AMD associated with RAP is responsible for the majority of the severe vision

loss and it can be very difficult to treat. The diagnosis of RAP is similar with the AMD but exudate and superficial and multiple hemorrhages are more common. [1,2,3] We describe the case of a 75-year-old male with unilateral RAP treated successfully with intravitreal triamcinolone acetonide injection (IVTA).

### Case presentation

A 75-year-old male presented in the Ophthalmology Department of the Emergency

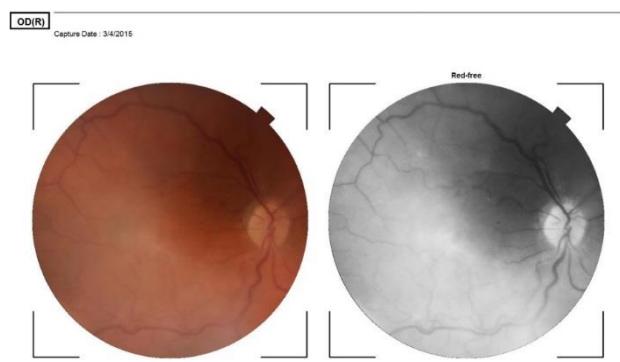
County Hospital Craiova in January 2015 with a 4 months history of vision loss in his right eye.

The medical history of the patient revealed in the right eye: a cataract surgery with a posterior chamber pseudofak 2 years ago, a laser YAG iridotomy for closed angle glaucoma 2 years ago and a macular edema; in his left eye a laser YAG iridotomy for closed angle glaucoma 2 years ago and a senile cataract in evolution. He was also diagnosed with essential arterial hypertension 10 years ago and impaired glucose tolerance 8 months ago.

At the admission his visual acuity (VA) was 4/50 in the right eye non-optical correctable and 5/5 with optical correction in the left eye.

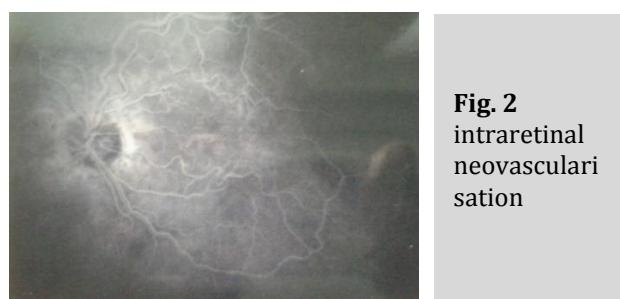
The slit lamp examination revealed laser YAG iridotomy in both eyes, posterior chamber pseudofak in the right eye, cortico-nuclear densifications in the left eye.

The indirect ophthalmoscopy examination revealed normal optic disc, a few macular hemorrhages situated in the superior-temporal sector, a white epiretinal membrane and macular edema in the right eye (**Fig. 1**). The left eye indirect ophthalmoscopy could not be performed due to the miotic pupil.



**Fig. 1** Right eye - white epiretinal membrane, macular edema

The angiofluorography (FA) in the right eye shows intraretinal neovascularization (**Fig. 2**) and late hyperfluorescence due to staining (**Fig. 3**).

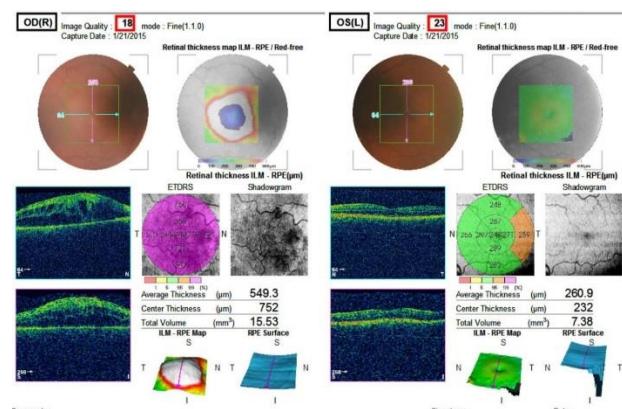


**Fig. 2**  
intraretinal  
neovascularization



**Fig. 3** late  
hyperfluoren-  
ce due to  
staining

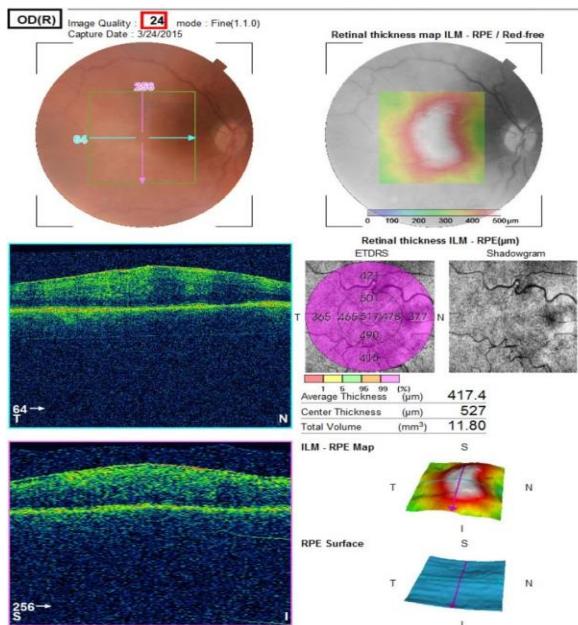
An optical coherence tomography (OCT-**Fig. 4**) was performed to the patient before the IVTA injection in both eyes as shown below. OCT revealed sub- and intraretinal fluid in the right eye with central retinal thickness of 752 µm and perifoveolar thickness of 549.3 µm and normal macular aspect with normal thickness in the center 232 µm and perifoveolar 260.9 µm.



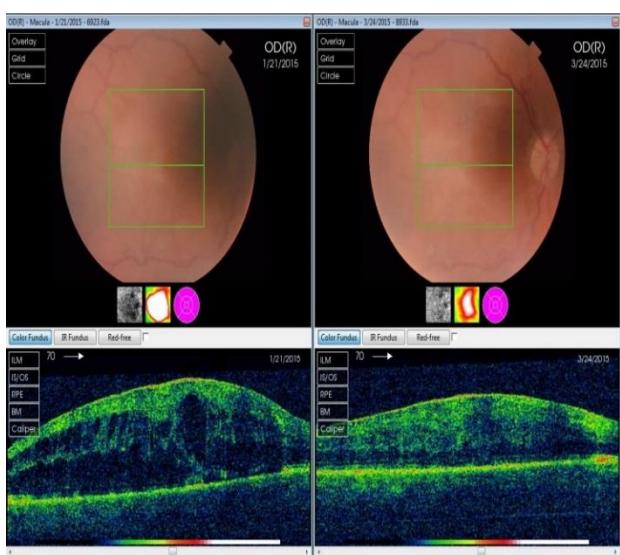
**Fig. 4** 3d macula right eye comparing to the left eye

After the investigations we decided to start the treatment consisting in one single intravitreal injection with Triamcinolone Acetonide (IVTA) in the right eye.

The follow-up visits were made at 2 days, 1 week, 4 weeks, 3 months after the IVTA injection. Two days after the IVTA injection the VA improved at about 5/40. At the 2nd follow-up visit the BCVA was 5/30. In comparison with the improved VA the macular edema gradually resorbed and 1 months after the IVTA injection the central retinal thickness was 527 µm and perifoveolar was 417.4 µm.



**Fig. 5** After 1 week from IVTA injection



**Fig. 6** Comparing before and after the IVTA

## Discussions

The first author who described the presence of anastomoses between the retinal and choroidal circulations in eyes with disciform scars was Oller over 100 years ago [4]. Chorioretinal anastomoses were later described, associated with laser photocoagulation [5],

radiotherapy [6], chorioretinal inflammatory diseases [7] and parafoveal telangiectasias [8].

Later Hartnett et al described in 1992 nine cases of retinal neovascularization, to which they referred as "deep retinal vascular anomalous complex" [9]. In 2000, Slakter et al. described chorioretinal anastomoses in eyes with pigment epithelial detachment and indocyanine green (ICG) hot spots [10].

In 2001, Yannuzzi et al. described chorioretinal anastomosis as neovascular proliferation with origin in the retina, and proposed the designation of RAP - retinal angiomatic proliferation [2]. Several authors [11-16] maintained the designation of chorioretinal anastomosis, proposing a choroidal origin for this clinical entity. In 2008, Yannuzzi et al. described 5 cases of RAP with the neovascular complex originating in the choroid instead of the retina, and proposed that RAP should be called type 3 neovascularization [17].

According to Yannuzzi et al. [2] classified RAP initial neovascularization process in 3 stages:

### 1) Stage I- Intraretinal neovascularization.

This stage is mainly diagnosed when the second eye is already affected. The presence of associated small retinal hemorrhages constitutes a very useful sign in early RAP diagnosis. A small elevation of the inner/intermediate retina caused by angiomatic tissue may be observed under a slit lamp; this elevation may extend tangentially, assuming telangiectasic appearance. In FA, the angiomatic complex is observed as a hyperfluorescent area in front of the RPE, identical to that occurring in classic choroidal and, more frequently occult neovascularization. Dilated retinal vessels may perfuse and drain the intraretinal neovascularization (IRN) and form retino-retinal anastomoses. ICG may reveal a hot spot with staining and leakage.

### 2) Stage II- Involvement of the subretinal space with localized neurosensory detachment

of the retina, edema and retinal hemorrhages at the edges may already be observed in fundus color photography. The angiomatic formation draining vein becomes visible, originating in the deep retina. PED is observed in 94% of eyes; FA may reveal well-delimited hyperfluorescence in the diffuse leakage area associated with PED, identical to that occurring for a minimally classic

choroidal neovascular membrane. ICG clearly establishes stage II diagnosis as the presence of a hot spot with leakage, as well as a hyperfluorescent area associated with serous pigment epithelium detachment (SPED). Leakage might not be revealed by FA, probably because fibrin from retinal exudates cannot be impregnated with fluorescein, contrary to what occurs with ICG [18]. FA is rarely useful in differential diagnosis between classic, occult or minimally classic membranes and stages I and II [2], contrary to ICG, where a slow-growing extra, juxta or subfoveal hot spot, sometimes asymptomatic, is observed in stages I and II. Retino-retinal anastomoses may also be observed in stage II.

**3) Stage III-** Choroidal neovascularization is clearly visible, sometimes with the appearance of vascularized PED.

According to Gass [19], RAP would progress in 5 stages instead of 3, easily identifiable by FA and ICG:

**I) Pre-clinical stage-** Atrophy of the outer retina, with retinal capillaries moving closer to a choroidal neovascular complex located below the RPE – type 1 choroidal neovascularization – and no clinical signs of chorioretinal anastomosis. ICG would be necessary to identify type 1 neovascularization.

**II) Early clinical signs-** Anastomosis between dilated capillaries of the deep retina and the choroidal neovascular complex is associated with small intraretinal haemorrhages, which would constitute the first clinical sign of chorioretinal anastomosis.

This stage may occur weeks or months before stage 3, where subretinal choroidal neovascularization is already observed.

**III) Proliferation of choroidal neovascularization over the RPE-** subretinal neovascularization – type 2 CNV.

**IV) Appearance of serous PED caused by activation of newly formed subepithelial vessels.**

**V) Mixed neovascularization-** piggyback-type neovascularization, with two levels – type 1 and type 2 with cicatricial disciform lesion, making chorioretinal anastomosis visible.

Stage 3 in Gass's classification corresponds to stage I in Yannuzzi's classification, with stages 4 and 5 in Gass's classification corresponding to stages II and III, respectively.

Currently, Yannuzzi's classification is the most widely used.

In our case, we diagnosed the patient with RAP stage I after Yanuzzi and stage II/III after Gass.

The average and median ages of a series of 108 patients studied by Yannuzzi et al. [2] were 80 and 81 years respectively.

Both eyes were similarly affected in 51.9% of patients, with RAP occurring in the second eye with an average of 15 months after appearing in the first eye.

At 3 years, both eyes were affected in 100% of patients [20,21].

In our case, the patient was diagnosed with RAP in only one eye but we will be monitoring him every six months.

Most of the published studies use intravitreal injection with ranibizumab in treating RAP as a form of exudative AMD. Due to the inflammatory process involved in RAP pathogenesis, recent research has proved the efficacy of IVTA injection in the treatment of this condition.

Saito M et al observed in a retrospective, observational case series over a 3 years period, that the combined therapy intravitreal bevacizumab (IVB) plus PDT was more effective than IVTA plus PDT in the treatment of RAP. [22]

Adnaan Haq et al demonstrated in a case report the successful RAP treatment with combined intravitreal ranibizumab and IVTA. [23]

## Conclusions

In spite the patient late presentation in the Ophthalmology Department (4 months after he noticed the vision loss in the right eye) the VA and OCT outcome after a single dose of IVTA injection was very good.

Further studies are required to establish the difference between monotherapy with IVTA or combined therapy IVTA with ranibizumab as the optimal treatment for RAP.

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