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Research Paper

Ruptured retinal arterial macroaneurysm: a case report

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***Abstract** :*
***Introduction :** Retinal arterial macroaneurysms (RAMs) are acquired focal dilatations of the large retinal arteries, usually occurring within the first three bifurcations of the central retinal artery. They can develop into spontaneous sclerosis but can also be complicated by exudative signs, intraretinal edema and preretinal, intraretinal, subretinal or intravitreal hemorrhages. In the event of complications threatening the visual prognosis, photocoagulation of the macroaneurysm may be indicated.*
***Materiels and methods :** seventy year-old women, with high blood pressure who present a ruptured retinal arterial macroaneurysm in the right eye.*
***Discussion** : RAMs are seen in elderly women and are usually closely related to long-standing hypertension and arteriosclerosis. Most RAMs are located in the temporal retina, and many may regress spontaneously with a favorable prognosis. However, a marked decrease in visual acuity may result when the macula is involved by hemorrhages and exsudates.*
***Conclusion** : Treatment guidelines for RAMs and further research to assess the long-term safety and efficacy of different treatments for RAMs are needed.*
***Key words**: ruptured retinal arterial macroaneurysm, high blood pressure, observation, photocoagulation.*

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I. Introduction :

Retinal arterial macroaneurysms (RAMs) are acquired focal dilatations of the large retinal arteries, usually occurring within the first three bifurcations of the central retinal artery. Typically, RAMs are seen in elderly women and are usually closely related to long-standing hypertension and arteriosclerosis. Most RAMs are located in the temporal retina, and many may regress spontaneously with a favorable prognosis. However, a marked decrease in visual acuity may result when the macula is involved by hemorrhages and exsudates [1].

They can develop into spontaneous sclerosis but can also be complicated by exudative signs, intraretinal edema and preretinal, intraretinal, subretinal or intravitreal hemorrhages. In the event of complications threatening the visual prognosis, photocoagulation of the macroaneurysm may be indicated [2].

II. Observation :

We report the case of a 70 year-old women, diabetic type 2 treated by oral antidiabetics, high blood pressure under amlodipine 10mg (18/10mmhg at admission), and elevated triglycerides (2.5 mmol/L), who presented to the emergency room for sudden painless unilateral loss of vision in the right eye,

Visual acuity is limited to count fingers at 3 meters right eye, 7/10 left eye, Oculomotor examination is normal, pupillary reflex and relative afferent pupillary defect (RAP) was normal. Anterior segment examination shows debutante cortical cataracte both eyes.

Fundus however shows in the right eye a retinal hematoma in the macular region involving the fovea, surrounded by dry exsudates. Signes of moderate non proliferative diabetic retinopathy with atherosclerosis signs both eyes (figure 1).

Fluorescent angiography shows at right double preretinal and intraretinal mask effect not allowing visualization of the vessels opposite on all sequences, in the periphery, punctiform intraretinal hemorrhages, with mask effect, and an enhancement suggesting an occluded macroaneurysm. Both eyes shows vascular tortuosity with signs of moderate nonproliferative diabetic retinopathy (figure 2).

OCT images shows, right eye, non-visualization of the VR interface and loss of the foveolar funnel, thickening of the intraretinal layers with intraretinal hematoma and masking effect on the RPE. Left eye was normal (figure 3).

Once the diagnosis of RAM rupture was established, the patient was sent to cardiology and endocrinology for treatment. And to accelerate the regression of the RAM, laser photocoagulation with 150 mW power was carefully performed at the RAM. The duration was 0.2 s and the spot size was 200 μm using a single pulse was performed on the ram, visual acuity passed to 4/10 within 2 weeks.

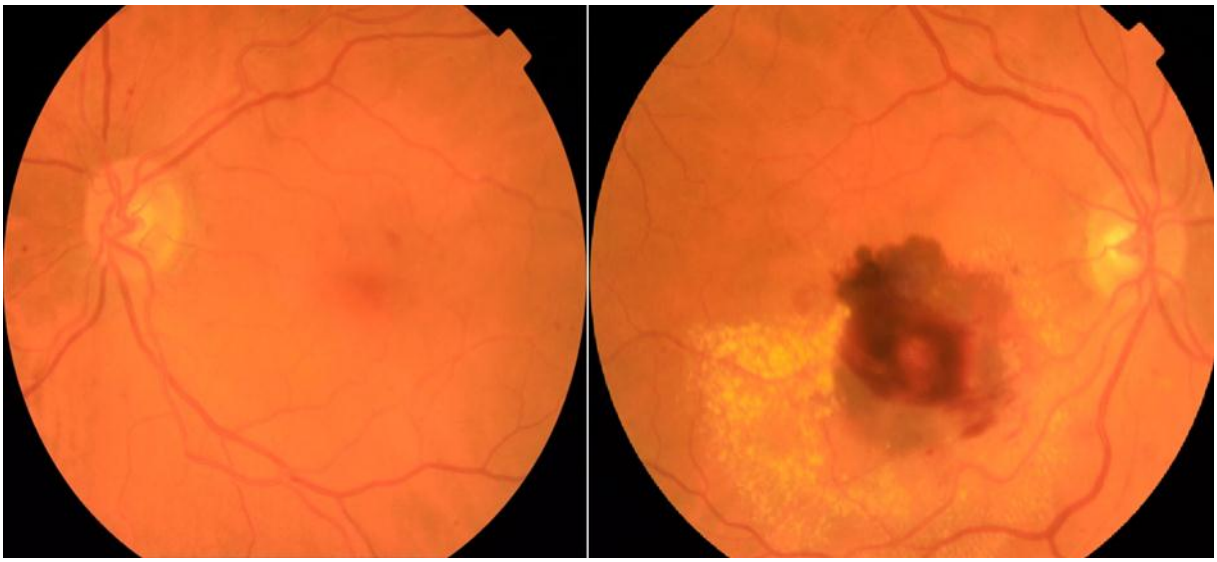

Figure 1: retinography image shows in the right eye retinal hematoma in the macular region involving the fovea, surrounded by dry exsudates. Signes of moderate non proliferative diabetic retinopathy with atherosclerosis signs both eyes.

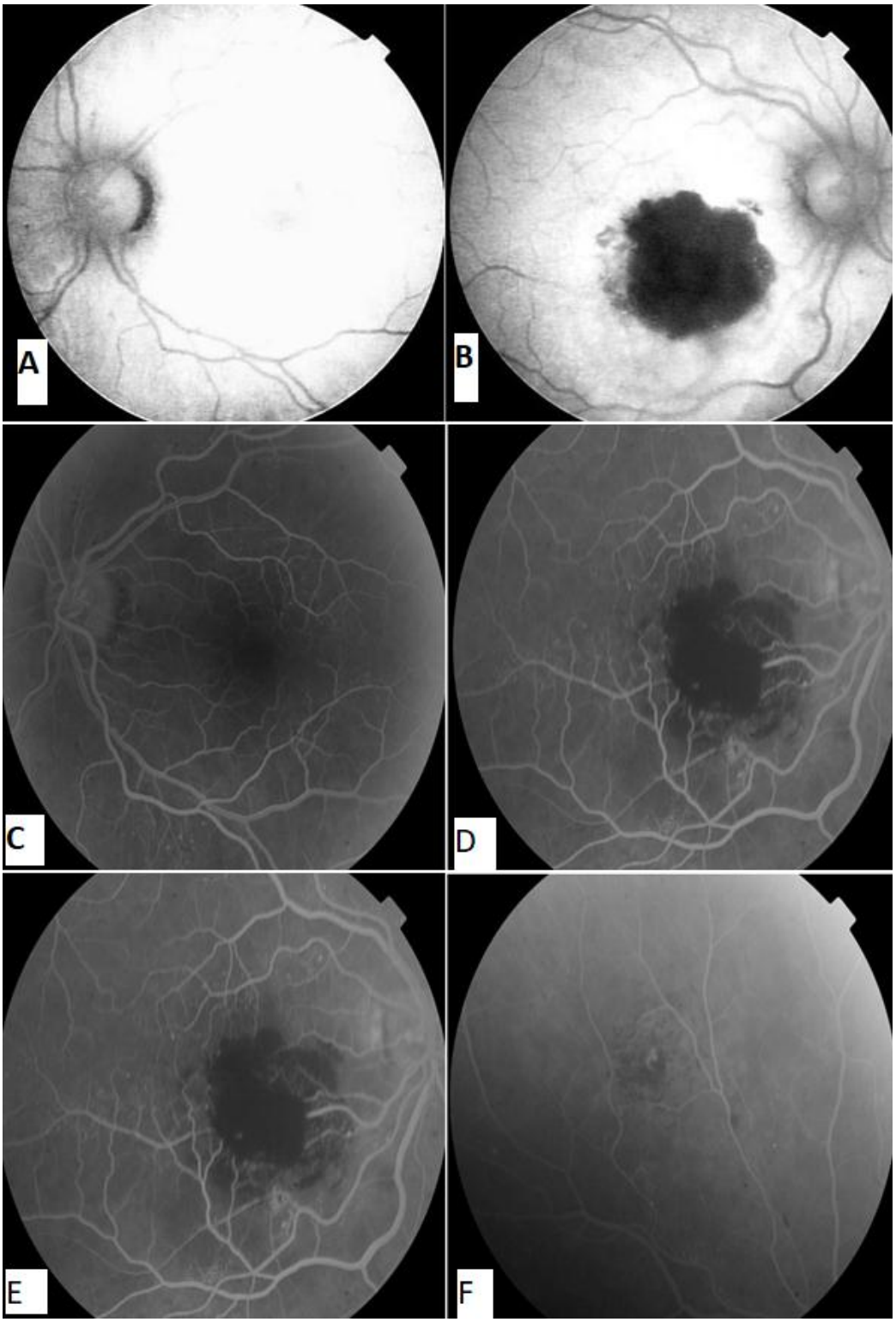

Figure 2 : autofluorescent sequences(A,B), Fluorescent angiography shows at right double preretinal and intraretinal mask effect not allowing visualization of the vessels opposite on all sequences, note in the periphery, punctiform intraretinal hemorrhages, with mask effect, and an enhancement suggesting an occluded macroaneurysm (D, E, F). left eye moderate NPRD and vascular tortuosity(C).

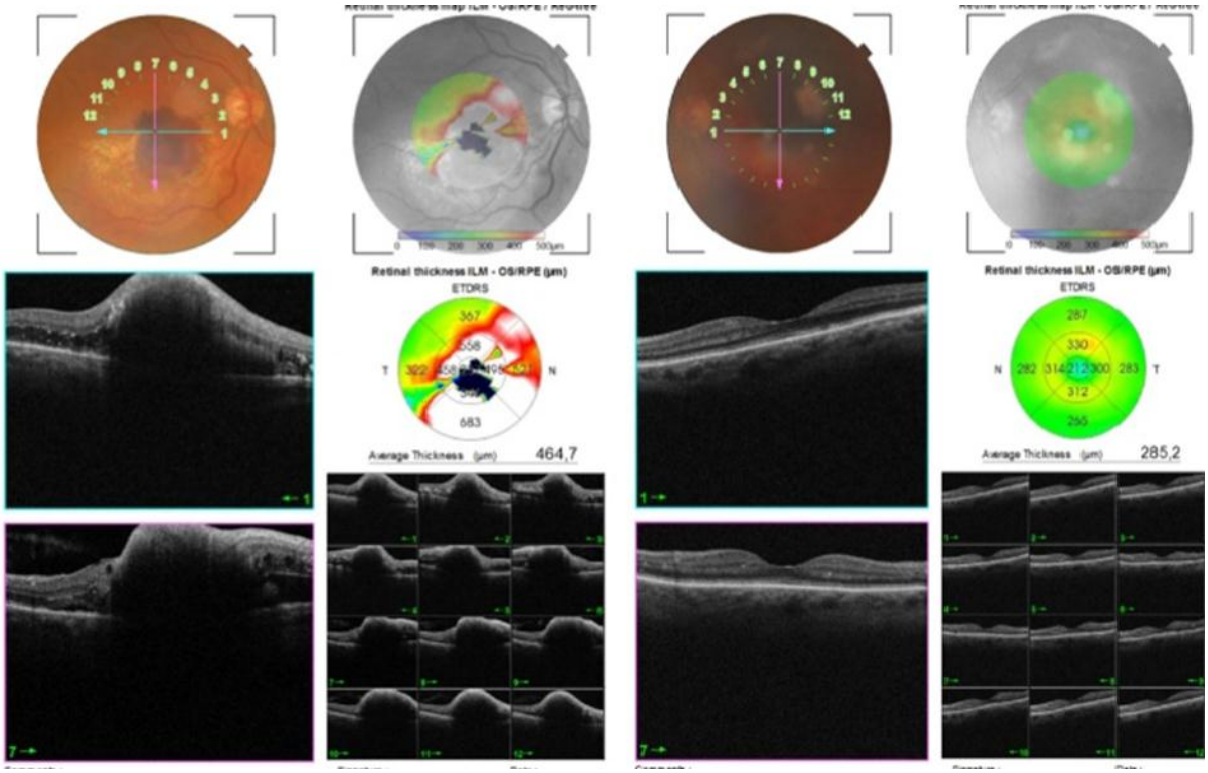

Figure 3 : OCT images, right eye, non-visualization of the VR interface and loss of the foveolar funnel, thickening of the intraretinal layers with intraretinal hematoma and masking effect on the RPE.

III. Discussion :

Retinal arterial macroaneurysms remain a rare pathology, which occurs more frequently in women, between 50 and 80 years of age. Known risk factors of RAMs include female sex, advanced age, systemic hypertension, hypercholesterolemia, polycythemia, and arteriosclerotic cardiovascular disease, among others. Previous studies have suggested that hypertension is the most important risk factor, and the proportion of hypertension in patients with RAMs ranges from 51 to 75%. Although there is no ocular risk factor that has been clearly confirmed by previous studies, several ocular diseases have been reported to be associated with RAMs, including retinal venous occlusion (RVO), retinal arterial occlusion (RAO), and macular hole (MH) [3] [4].

The precise pathophysiology of RAMs has not been well-understood to date. However, it has been recognized that RAMs are characterized by thinned blood vessel walls, decreased elasticity, and fibrosis secondary to the loss of the muscular coat, thus leading to increased susceptibility to dilatation by raised intraluminal pressure. The focal weakness of the arterial walls results in RAMs. And rupture of RAMs occurs when intravascular pressure exceeds the threshold that the fragile vessel walls can withstand. various focal or systemic factors leading to increased vessel wall susceptibility may be responsible for RAMs. For example, hyaline degeneration and arteriosclerosis of the retinal arteries are commonly seen in patients with sustained hypertension, and these conditions can partly explain why hypertension is the most important risk factor for RAMs [5].

The presentation of a patient with RAMs can be variable. RAMs can be classified into three types: hemorrhagic, exudative, and quiescent, between which the management and prognosis may differ. Hemorrhagic RAMs are caused by the acute rupture of the fragile retinal artery walls. Depending on the location of the rupture, hemorrhagic RAMs can cause multi-level retinal hemorrhages and can be observed in the preretinal, sub-ILM, intraretinal, and subretinal spaces. VHs occur when blood breaks through the ILM and is drained into the vitreous. Exudative RAMs are marked by exudates with or without macular edema. If a RAM shows both exudates and hemorrhages, the type is based on the initial predominant complication causing decreased visual ability (VA). Contrary to the first two types, quiescent RAMs are generally asymptomatic with no obvious hemorrhages or exudates. However, hemorrhages and/or exudates could also be found in quiescent RAMs when the macula is not affected. Quiescent RAMs may develop into the hemorrhagic or exudative type. Typically, RAMs are unilateral, but there are reports of bilateral RAMs or multiple RAMs occurring in the same eye. Neurosensory retinal detachment and macular edema with or without exudations can also be seen in some cases. RAMs might be found upon routine ophthalmic examination in patients without any ocular symptoms. However, most RAMs are found because of the abrupt and severe decrease in VA and loss of visual field that result from hemorrhages and exudates [5][6][7][8].

The diagnosis of RAMs is typically made based on clinical manifestations, demographic characteristics, and examinations such as FFA, ICGA, and OCT. However, RAMs can be described as a masquerade syndrome and may mimic a range of other ocular diseases. According to previous literature, RAMs are frequently misdiagnosed, especially when the underlying RAMs are obscured by hemorrhages or exudates. Considering that these studies were completed at a time when RAMs were not well understood and the means of inspection were more limited and less advanced, the current misdiagnosis rate should now be lower. Yet, it is still necessary and important to distinguish RAMs from other mimickers in clinical practice. For patients with retinal hemorrhages, mimickers such as RVO, diabetic retinopathy (DR), retinal telangiectasia (Coats' disease and Leber's miliary aneurysms), Valsalva retinopathy, idiopathic polypoidal choroidal vasculopathy (IPCV), choroidal neovascularization (CNV), retinal capillary hemangiomas, and retinal cavernous hemangioma need to be excluded. In patients with lesions covered with dense VH, mimickers including wet age-related macular degeneration (wAMD), proliferative diabetic retinopathy (PDR), retinal tear, and ischemic RVO should be ruled out before making a diagnosis [9][10][11].

A large part of RAMs are of no severe threat to vision and may regress spontaneously with a favorable prognosis. For asymptomatic patients without hemorrhages or exudations related to RAMs, a visit every six months until regression would suffice. It is suggested that patients without macular involvement should be observed, and those with vision loss secondary to vitreous, intraretinal, or preretinal hemorrhages (macula not involved) may also be observed for spontaneous regression for a few months before the treatment [5] [12].

Both direct and indirect laser photocoagulation treatment can be used, especially when exudation or edema is vision-threatening. Direct laser photocoagulation is performed directly at the RAM, aiming to speed the regression of the RAM and fundamentally reduce the leakage. As another option, indirect laser photocoagulation is applied to the surrounding retina of the RMA to order to keep the leakage away from the macula. The main complications of laser photocoagulation are arteriolar occlusion, retinal traction, increased exudation, capillary dropout, and subretinal scarring. By reducing the duration of laser exposure and using sub-visible clinical endpoints, subthreshold laser treatment (STLT) can achieve similar treatment outcomes while reducing the complication rate when compared with conventional laser photocoagulation. For sub-ILM hemorrhage secondary to RAM rupture, Nd: YAG laser membranotomy should be considered as a method for draining the sub-ILM hemorrhages into the vitreous for quicker absorption and has been proven to be both safe and effective [5] [13].

Anti-VEGF therapy has been frequently studied as a promising treatment for RAMs with hemorrhage or macular edema. Anti-VEGF agents may close the involved pathologically permeabilized retinal arteries by normalizing the vessel walls and inducing thrombosis within the RAMs. Several previous studies indicate that anti-VEGF agents are an effective therapy for complicated hemorrhagic and/or exudative RAMs, resulting in quick and safe improvement of vision with fewer complications. It has also been noted that combined laser and anti-VEGF therapy can rapidly reduce macular exudation by RAM [5] [14].

Pars plana vitrectomy (PPV) should be considered in the following cases: submacular hemorrhages that cannot be treated by laser photocoagulation; non-clearing VHs after observation for over three months, as the natural resorption process usually lasts less than three months; VHs that obscure the macula, making the diagnosis difficult, especially when other lesions are suspected; VHs that interfere with necessary treatment needed for vision recovery; and removal of pre-retinal hemorrhages if quicker vision recovery is needed. In appropriate circumstances, PPV can help to remove the pre-retinal hemorrhages in a timely manner, reducing the toxic effects on the retina and reducing the probability of ERM formation [15] [16].

The combination of tPA with submacular surgery or pneumatic replacement has been effective in the treatment of submacular hemorrhage (SMH). Submacular surgery includes surgical drainage of the submacular hemorrhage through retinotomy, fluid-gas exchange, and insufflation with intravitreal gas. Pneumatic displacement is less invasive and includes perfluorocarbon gas injection and downward gaze positioning in order to remove the hemorrhage from the macula. These two approaches both require patients to adopt a face-down position for several days after the operation. Potential complications of pneumatic replacement include failure of displacement, VH, endophthalmitis. Complications of submacular surgery are similar to those of PPV. [17] [18].

IV. Conclusion :

Retinal arterial macroaneurysms remain a rare pathology, which occurs more frequently in women, between 50 and 80 years of age. Systemic arterial hypertension is associated in more than half of the cases. These conditions are unilateral in 90% of cases but can be multifocal.

They can develop into spontaneous sclerosis but can also be complicated by exudative signs, intraretinal edema and preretinal, intraretinal, subretinal or intravitreal hemorrhages. In the event of complications threatening the visual prognosis, photocoagulation of the macroaneurysm may be indicated.

Treatment guidelines for RAMs and further research to assess the long-term safety and efficacy of different treatments for RAMs are needed.

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