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REVIEW

# Ruptured retinal arterial macroaneurysm: Diagnosis and management

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Retinal arterial macroaneurysm;
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**Abstract** Retinal arterial macroaneurysm is an acquired, focal dilation of a retinal artery, typically occurring within the first three bifurcations of the central retinal artery. The clinical presentation of a retinal arterial macroaneurysm is highly variable, making initial diagnosis difficult and differentials many. Identification of retinal arterial macroaneurysms is crucial to appropriately co-manage with the primary care physician for hypertension control. Prognosis is generally good and observation is often an adequate treatment. However, in cases of macular threat or involvement, some treatment options are available and referral to a retinal specialist is indicated.
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**PALABRAS CLAVE**
Macroaneurisma de la arteria retiniana;
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**Rotura de macroaneurisma de la arteria retiniana: diagnóstico y manejo**

**Resumen** El macroaneurisma de la arteria retiniana es una dilatación focal y adquirida de una arteria retiniana, que se produce normalmente en de las primeras tres bifurcaciones de la arteria central de la retina. La presentación clínica del macroaneurisma de la arteria retiniana es altamente variable, lo que dificulta el diagnóstico inicial dadas las muchas características diferenciales. La identificación de dichos macroaneurismas es esencial para poder coordinar con el facultativo de atención primaria el control de la hipertensión. El pronóstico es generalmente bueno, siendo a menudo la observación el tratamiento adecuado. Sin embargo, en casos de amenaza o afectación macular, las opciones de tratamiento son variables, recomendándose la derivación al especialista de la retina.
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## Introduction

Retinal arterial macroaneurysm (RAM) is described as an acquired, focal dilation of a retinal artery, typically occurring within the first three bifurcations of the central retinal artery.[1–4] RAMs can present with a wide variety of retinal findings and are often misdiagnosed at initial presentation.[3] Formation of a RAM is believed to result from a combined process of arteriosclerosis and hypertension. Up to 75% of patients presenting with RAM are shown to have hypertension.[1,3,4] Accurate identification of RAM is crucial to appropriately co-manage these patients with the primary care physician.[3,4] Prognosis for RAM is favorable; most undergo spontaneous resolution over a period of months.[1,2,4] In many instances observation is adequate management. However, in cases of macular involvement referral to a retinal specialist is often warranted as a variety of treatment options exist.[3,4] In this review, we aim to highlight the variable presentation of this frequently misdiagnosed condition and provide an update on current treatment options. We also include a case report of a ruptured RAM with macular involvement, co-managed between optometry and ophthalmology.

## History and epidemiology

Clinical descriptions of retinal arterial aneurysms have been reported in the literature since the late 1800s,[1] but in 1973 Robertson[2] was the first to describe the clinical characteristics and apply the term retinal arterial macroaneurysm. Retinal arterial macroaneurysms are described as an acquired, focal dilation of a retinal artery, typically occurring within the first three bifurcations of the central retinal artery.[4] They are commonly found at arteriovenous crossings[4,6] or directly at a bifurcation.[5] Although RAMs are usually a solitary, unilateral finding, multiple RAMs may be observed in 15–20% of cases and bilateral disease occurs in up to 10% of cases.[5] It is believed that less structural support of arteries exist at arteriovenous crossings due to the absence of the adventitial layer, making these areas more prone to aneurysm formation.[6] Two types of RAM are described in the literature, fusiform (cuffed) and saccular (blowout).[7] Fusiform RAMs are described as an uniform widening of the retinal artery while saccular RAMs are described as a localized outpouching of the arterial wall.[8]

RAMs are most commonly observed in elderly females, with most studies reporting an age range from 66 to 74 years old, and a female preponderance of about 70%.[3,6,8–11] Hypertension is the most common systemic condition associated with RAM; approximately 75% of presenting patients have hypertension.[1,6,11] Other associated conditions include arteriosclerosis and abnormal lipid levels.[3]

## Pathophysiology

The pathophysiology behind the formation of RAMs is not fully understood. RAMs result from a focal weakness in the arterial wall, which is believed to be the combined result of aging and atherosclerosis. With aging, atrophy of the muscularis layer leads to thinning and fibrosis of the vessel wall causing decreased elasticity within the artery.[1] This sequence of events creates an increased susceptibility to arterial dilation from raised hydrostatic pressure, as seen in hypertension.[1,8] The reason and the localization of these focal, as opposed to widespread, weaknesses develop are not entirely understood. It is suggested that these are areas where emboli have lodged or local thrombosis has occurred. Histological studies of RAMs have shown both thrombus and cholesterol crystals partially filling the macroaneurysm, lending support to these theories.[1]

## Clinical signs and symptoms

The clinical presentation of RAM varies greatly and is frequently described as a masquerade syndrome. A retrospective study by Lavin and colleagues in 1987[6] found RAMs to be misdiagnosed at a rate of 75% at first presentation and a more recent study by Moosavi et al. reported only 4 of 14 cases (28%) listing RAM as the initial diagnosis.[8] Patients may present with an acute or insidious loss of vision when hemorrhage or edema involves the macula or when vitreous hemorrhage is present. But often RAMs are discovered on routine ophthalmic examination, presenting without any symptoms at all.[5]

The clinical appearance of RAMs is also highly variable. Traditionally, we expect to see blood in multiple layers, including subretinal, intraretinal, preretinal and vitreal.[4,12] This is expected as arteries are high flow vessels, thus when an aneurysm ruptures, it does so under significant pressure, pushing blood into many retinal layers. Often the macroaneurysm can be seen as a rounded dilation within an artery[1] and spontaneous pulsations have been documented. There is some thought that spontaneous pulsation could be an indication of pending rupture, however this idea has been challenged.[5] Exudation may also be present, usually seen in a circinate pattern surrounding the aneurysm but may also be found in the macular region.[3] Macular edema can occur with or without exudation and neurosensory detachments may also be seen.[5]

## Differential diagnoses

Differential diagnoses of RAM vary depending on the clinical presentation of the case. The predominating feature – abnormal vessel structure, hemorrhage, or exudates, and the retinal location – is going to alter the differential diagnoses considered in each case. Submacular blood and exudation are often confused with age related macular degeneration (ARMD).[5] The bilateral nature of ARMD can help rule this out; however in patients who may additionally have ARMD, a fluorescein or indocyanine green angiogram can help determine a diagnosis.[3,5] Dense subretinal blood may be flat or elevated and mimic a malignant melanoma. In such a case, ultrasound, fluorescein angiography (FA), magnetic resonance imaging and observation can be considered to help determine a diagnoisis.[3] Differentials for the appearance of pre-retinal hemorrhage in the macular region, such as seen in our case, include valsalva retinopathy, posterior vitreous detachment with secondary preretinal hemorrhage, proliferative diabetic retinopathy and exudative ARMD. Valsalva retinopathy presents with pre-retinal hemorrhage near the macula and may be unilateral or bilateral in nature.

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