

Retinal Artery Macroaneurysm (RAMA)

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Chief Complaint: Sudden painless decreased vision in the right eye

History of Present Illness: The patient is an 85 year old woman who noted sudden-onset darkening of her central vision surrounded by a gray ring-like pattern in her right eye. She had no associated photopsias, metamorphopsia, or new floaters. She denied any eye pain, diplopia, redness, or headache. She had no history of ocular trauma, transient ischemic attacks, or neurologic symptoms.

Past Ocular History:

- Ocular hypertension
- Pseudoexfoliation
- Cataract surgery OD
- Nuclear sclerotic cataract OS

Past Medical History: Hypertension, hypothyroidism, osteopenia, hypercholesteremia

Medications: Timolol drops OU. Systemic medications include aspirin, levothyroxine, losartan, simvastatin, risedronate, tolterodine tartrate.

Family History: Noncontributory

Social History: Patient smokes half a pack of cigarettes per day and does not consume alcohol.

Physical Exam:

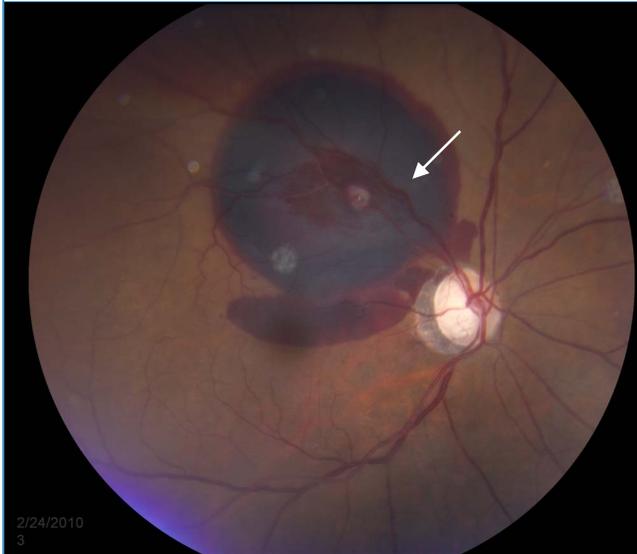
- Visual acuity (with correction)
 - 20/70+1 OD
 - 20/60 OS
- Extraocular motility: Full OU. Orthophoric in primary position.
- Pupils: 4 → 2 mm OD, OS, no relative afferent pupillary defect (RAPD)
- Intraocular pressure: 14 mmHg OD, 13 mmHg OS
- External and anterior segment exam:
 - OD: Normal other than PCIOL and pseudoexfoliation material at the pupillary margin
 - OS: Normal except for 2+ nuclear sclerotic cataract, 2+ cortical cataract, and pseudoexfoliation on the lens capsule and at the pupillary margin
- Dilated fundus exam (DFE):
 - OD: See Figure 1A
 - OS: Healthy optic nerve with cup-to-disc ratio of 0.3, normal vessels, macula, and periphery

Additional Tests:

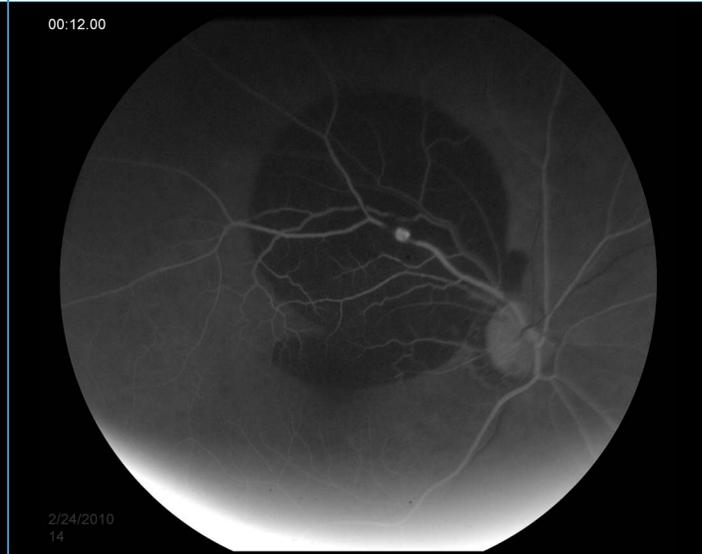
- Fundus fluorescein angiography (FFA): Aneurysm along the superotemporal retinal arteriole with surrounding area of blockage from hemorrhage (see Figure 1B-D)
- Optical coherence tomography (OCT): Subretinal fluid under the fovea OD (see Figure 2)

Figure 1

1A: Fundus photo of the right eye showing large area of subretinal hemorrhage with hemorrhage extending into the macula. Note the white spot along the superotemporal artery which corresponds to the hyperfluorescent lesion in the angiogram.



1B: FFA of the right eye in the early arteriolar phase showing early hyperfluorescence along the superotemporal arcade over the subjacent blockage from subretinal hemorrhage.



1C: Mid-venous phase of FFA shows increased hyperfluorescence.



1D: Late or washout phase of FFA shows persistent lesion hyperfluorescence.



Figure 2A: OCT of the right eye showing subretinal fluid beneath the fovea

Thickness Profile

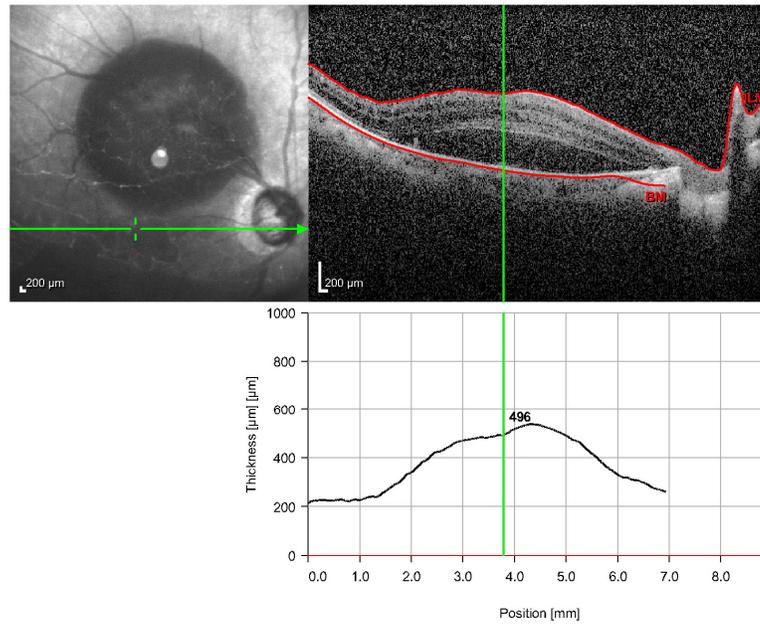
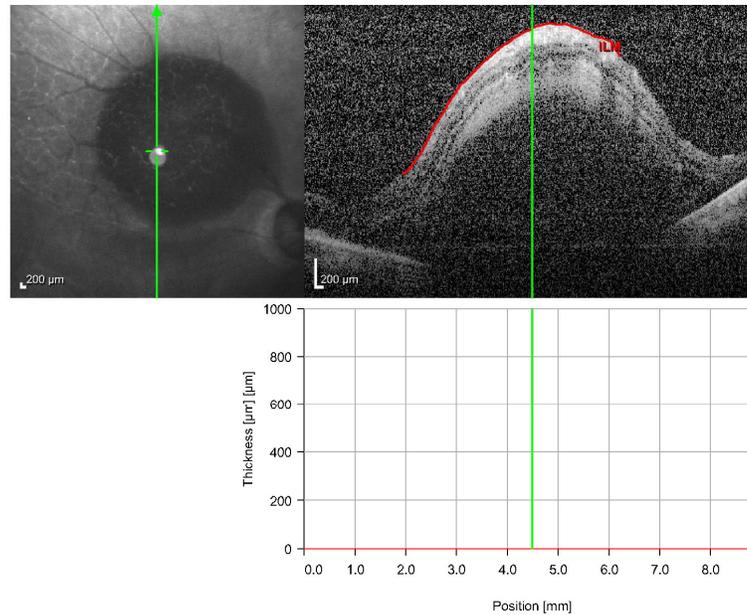


Figure 2B: OCT of the right eye showing extensive area of intraretinal edema and subretinal fluid (blood) with no choroidal abnormality

Thickness Profile



Discussion:***Pathophysiology***

A retinal artery macroaneurysm (RAMA) is usually described as an idiopathic, acquired dilation of a major retinal arteriole. Typically these develop within its first three bifurcations, at branch points, or areas of arteriovenous crossing. The most commonly reported site for RAMAs is the superotemporal arteriole. Less commonly they may develop from the cilioretinal arteries or on the optic nerve head. The underlying pathophysiology of macroaneurysms is not fully understood. One hypothesis is that arteriosclerosis leads to vessel wall fibrosis. The resulting decrease in wall elasticity, combined with elevated luminal pressure results in aneurysmal dilation. An alternative or accessory hypothesis is that emboli (which have been associated with vessels harboring macroaneurysms) or intra-arterial thrombosis leads to mechanical damage of the endothelium or adventitial vessel wall, which predisposes the vessel to aneurysm formation. Chronic venous stasis from hypertension and arteriosclerosis may also play a role (Rabb et al 1988).

Epidemiology

RAMAs usually develop in patients after the 6th decade of life (mean age of 57-71 years). However, there are published reports of patients with RAMAs in patients as young as 16 (Fernandez 1920, Rabb et al 1988). Females have a strong predilection toward the development of RAMAs. In larger studies 71-80% of patients were female (Rabb et al 1988). RAMAs are usually singular and unilateral. Ten percent of cases are reportedly bilateral and 20% present with multiple aneurysms along the same vessel or on multiple vessels. Systemic hypertension is the most commonly associated risk factor. This factor is present in 31%-81% of affected patients depending on the study cited (Fernandez 1920, Rabb et al 1988). Other risk factors reported in the literature have not been consistently observed in all studies and include elevated lipid levels, systemic vasculitis such as polyarteritis nodosa, sarcoidosis, diabetes, rheumatoid arthritis, and Raynaud's phenomenon.

Symptoms/Prognosis

Classically, patients with RAMAs present with sudden painless loss of vision. Many, however, are asymptomatic, and their RAMAs are discovered on routine exam, especially if the aneurysm is not associated with exudates, edema, or subretinal hemorrhage involving the macula. Vision loss from a RAMA is usually attributable to macular involvement including macular edema or hemorrhage in any of the retinal layers or vitreous. RAMAs may remain unchanged for many years, but most will eventually undergo thrombosis, fibrosis, and/or involution. Most patients have preservation or return of their vision unless they have extensive subfoveal hemorrhage or chronic macular edema. Ultimately the site and severity of the hemorrhage and macular edema determine visual outcomes (Rabb et al 1988).

Clinical Findings

The key fundoscopic finding is the presence of blood at multiple layers including the preretinal, intraretinal, subretinal, and sub-ILM spaces and the vitreous or the presence of an hourglass

hemorrhage (described by Schatz et al), which represents simultaneous preretinal and subretinal hemorrhage. Other common but less pathognomonic findings include exudative retinopathy consisting of yellow/white hard lipid exudates in a circinate pattern surrounding the aneurysm. Pulsations of the aneurysm can be appreciated in 10% of cases (Rabb et al 1988, Schatz et al 1980). Macular edema without exudates is less commonly seen, as are neurosensory detachments. Fluorescein angiography typically demonstrates an immediate filling of the aneurysm, which may leak throughout the study. If retinal hemorrhage is present, an area of blocked fluorescence around the aneurysm may be observed (see Figure 2). The involved artery may be narrowed and irregular, and the surrounding capillaries may demonstrate leakage (Rabb et al 1988).

Treatment

Because many patients spontaneously recover a significant amount of vision and the majority of aneurysms involute without intervention, most patients can safely be observed. Hypertension and other systemic risk factors should be treated adequately. Indications for laser treatment include vision loss due to chronic macular exudates or edema (Rabb et al 1988). Laser photocoagulation directly to the macroaneurysm using xenon arc, argon, or dye yellow has been documented in the literature to improve vision in some patients (Rabb et al 1988, Abdel-Khalek and Richardson 1986, Hudomel and Imre 1973). However, some studies note that direct photocoagulation to the macroaneurysm does not improve visual outcomes and may lead to branch retinal artery occlusion (Brown et al 1994). Indirect laser treatment to the area surrounding the macroaneurysm may also improve visual outcomes in some patients with macular edema (Robertson 1973, Palestine et al 1982, Francois 1979). Laser hyaloidotomy using a neodymium-doped yttrium aluminium garnet (Nd:YAG) laser for subhyaloid hemorrhage has been reported but is controversial due to the risk of vitreous hemorrhage or damage to the macula (Tassignon et al 1989). In the setting of vitreous hemorrhage of unclear etiology, pars plana vitrectomy has been performed. Surgical evacuation of subretinal hemorrhage has also been utilized (Ibanez HE et al 1995, Humayun M et al 1998).

Diagnosis: Retinal Arteriolar Macroaneurysm (RAMA)

<p style="text-align: center;">EPIDEMIOLOGY</p> <ul style="list-style-type: none"> • Typically occurs in patients older than 60 years of age • More frequently seen in women • Typically occurs unilaterally • Associated with hypertension 	<p style="text-align: center;">SIGNS</p> <ul style="list-style-type: none"> • Hemorrhage at multiple layers—preretinal, intraretinal, subretinal, sub-ILM, or vitreous • Aneurysmal dilation of arteriole, usually superotemporally • May be single or multiple aneurysms • Spontaneous pulsations can be seen • Macular exudates • Macular edema
<p style="text-align: center;">SYMPTOMS</p> <ul style="list-style-type: none"> • Asymptomatic • Sudden painless decrease in vision • Scotoma 	<p style="text-align: center;">TREATMENT</p> <ul style="list-style-type: none"> • Usually observation as most aneurysms involute and vision improves • Consider laser treatment for macular edema, exudates, or neurosensory detachments.

Differential Diagnosis:

- Traumatic multilayer hemorrhage
- Branch retinal vein occlusion
- Angiomatosis retinae (von Hippel Lindau)
- Capillary hemangioma
- Cavernous hemangioma
- Arteriovenous malformation
- Diabetic retinopathy
- Diabetic macular edema
- Exudative age related macular degeneration
- Retinal telangiectasia
- Coat’s disease (adult onset form)

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