A 53-Year-Old Male with 1 Week of Worsening Vision in His Right Eye

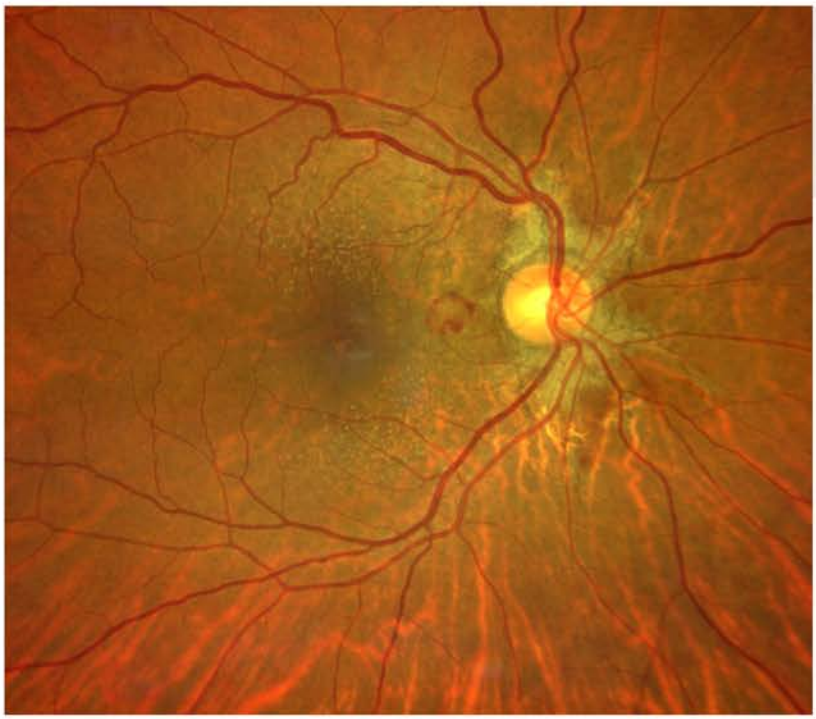
Gautam Vangipurim, MD; Richard J. Rothman, MD

Introduction:

A 53-year-old gentleman presented to The Retina Institute with 1 week of worsening vision in his right eye. He had no significant prior ocular history or prior ocular surgeries.

Exam:

Visual acuity was measured at 20/30 in both eyes. The anterior segment was notable for early cataract formation. The posterior segment exam is shown in Figure 1 and was notable for bilateral pseudodrusen and pigmented lines emanating from the optic nerve head. In the right macula, a small nasal subretinal hemorrhage was noted with no significant peripheral findings. An OCT (Figure 2) showed sub-retinal fluid and increased reflectively consistent with sub-retinal neovascularization. An OCTA (Figure 3) showed vascular flow at the level of the RPE consistent with a choroidal neovascular membrane (CNVM). The constellation of findings represents angioid streaks with the subsequent development of CNVM. Further systemic history revealed a history of beta-thalassemia and he was treated with intravitreal Bevacizumab to the right eye.



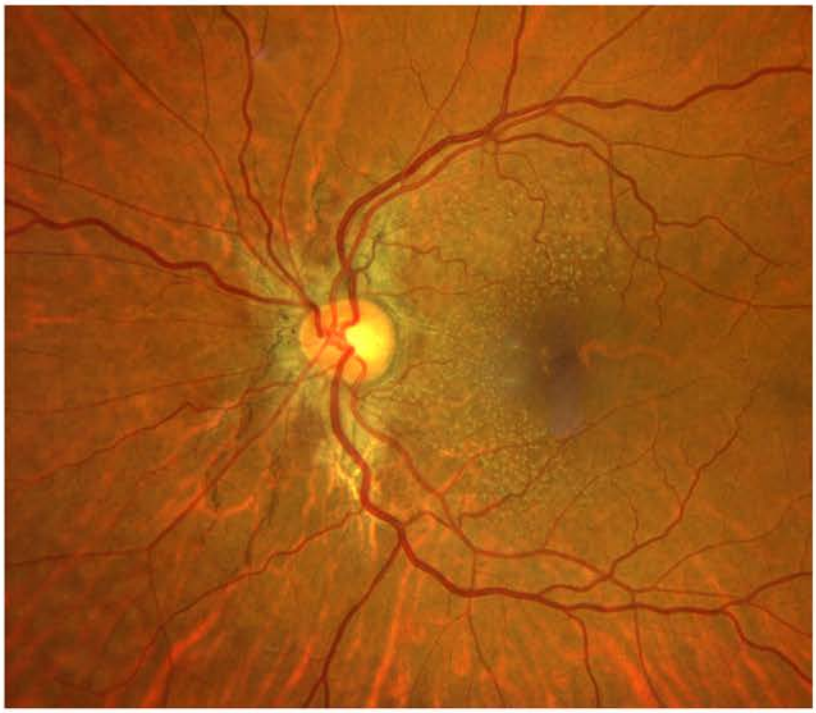


Figure 1: Color fundus photos demonstrating bilateral angioid streaks with sub-retinal hemorrhage nasal to fovea in the right eye.

Diagnosis:

Angioid streaks present as bilateral, irregular lines that emanate in a radiating pattern from the optic disc.[1] When subtle, their appearance can be confused with the underlying choroidal vasculature, hence the name "angioid". Histopathologically,

angioid streaks represent breaks in Bruch's membrane, which normally separates the retinal pigment epithelium from rich blood supply of the underlying choriocapillaris.[2-3] These breaks are theorized to occur due to prior underlying extensive calcification and thickening of Bruch's membrane. Several disease processes are known to be associated with angioid streaks and helpfully remembered using the mnemonic: **PEPSI**: **P**seudoxanthoma elasticum (PXE), **E**hler-Danlos syndrome, **P**aget's disease of bone, **S**ickle cell disease and other hemoglobinopathies such as thalassemia, and **I**diopathic causes. Of these, the most common systemic disease etiology is Pseudoxanthoma elasticum--an inherited connective tissue disease altering elastin fibrils in skin, heart, and gastrointestinal system. A careful systemic history, family history, and physical exam is required on suspected patients paying close attention to evidence of a "chicken skin" appearance of the neck. Eye findings in PXE other than angioid streaks include optic disc drusen, "peau d'orange" appearance of the temporal macula, and comet-tail peripheral atrophic lesions.

Discussion:

Common causes of vision loss in patients with angioid streaks include choroidal rupture secondary to trivial trauma, the development of angioid streaks through the fovea, and CNVM formation. All patients with angioid streaks should be followed regularly for the development of CNVM which occurs in 72-86% of all cases.[3] Prior studies have investigated treatment of CNVM with laser photocoagulation and photodynamic therapy with minimal effect on visual acuity and recurrence rate.[4-5] However, it is reasonable to attempt initial laser photocoagulation should the CNVM be located extra-macular. More recent studies have investigated anti-vascular endothelial growth factor (anti-VEGF) in the treatment of CNVM associated with angioid streaks and found mixed results. Giacomelli et al. reported retrospectively on 52 eyes treated PRN with ranibuzumab and/or bevacizumab for angioid streak associated CNVM followed over 3 years.[6] In this study, anti-VEGF therapy appeared to slow the progression of CNVM formation however did not prevent progressive central vision loss. Another study by Tilleul et al. followed 27 patients treated with ranibizumab for CNVM associated with angioid streaks and saw stabilization of visual acuity in 62.9% of study patients and no leakage on fluorescein angiography in 77.1% of patients at the last follow up visit.[7-8]

Conclusion:

In summary, angioid streaks represent a unique ophthalmic finding usually associated with an underlying systemic disease process. Careful history and physical exam should be undertaken and coordination of care with the patient's internist is advisable. Treatment with laser photocoagulation and/or anti-VEGF agents should be considered upon the development of CNVM.

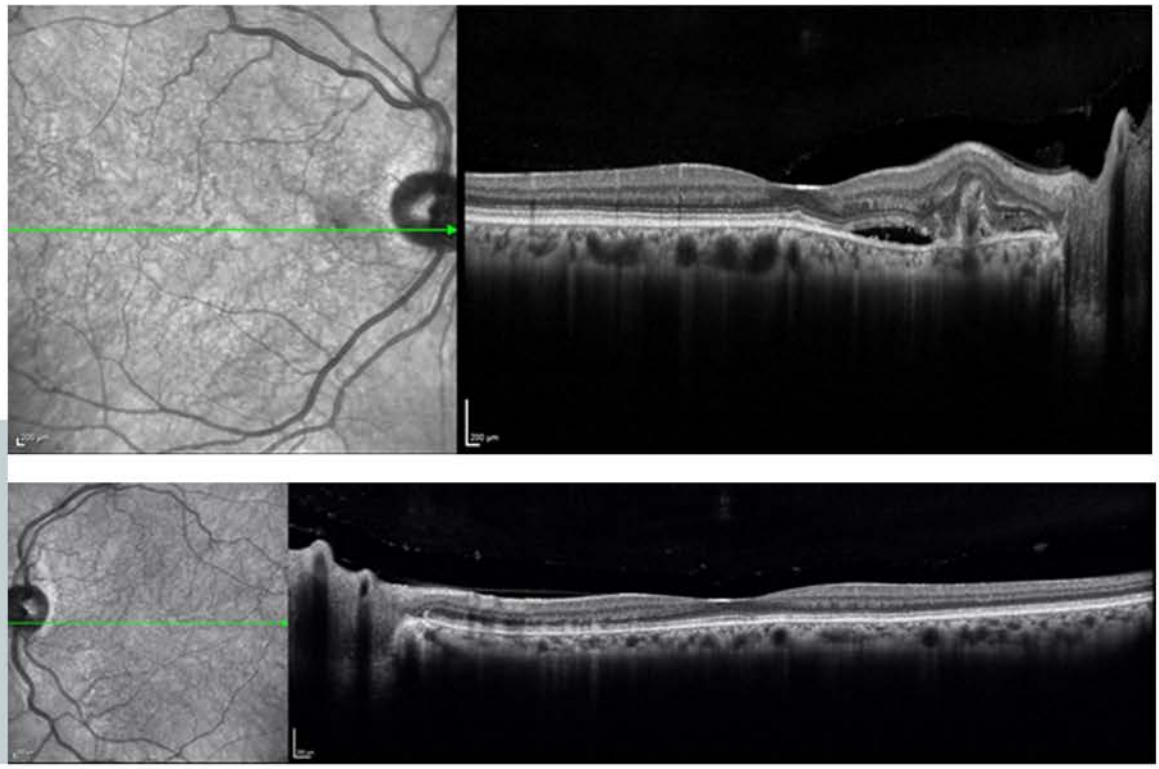


Figure 2: OCT macula of both eyes demonstrating right sub-retinal fluid and hyperreflectivity consistent with leakage from an active CNVM.

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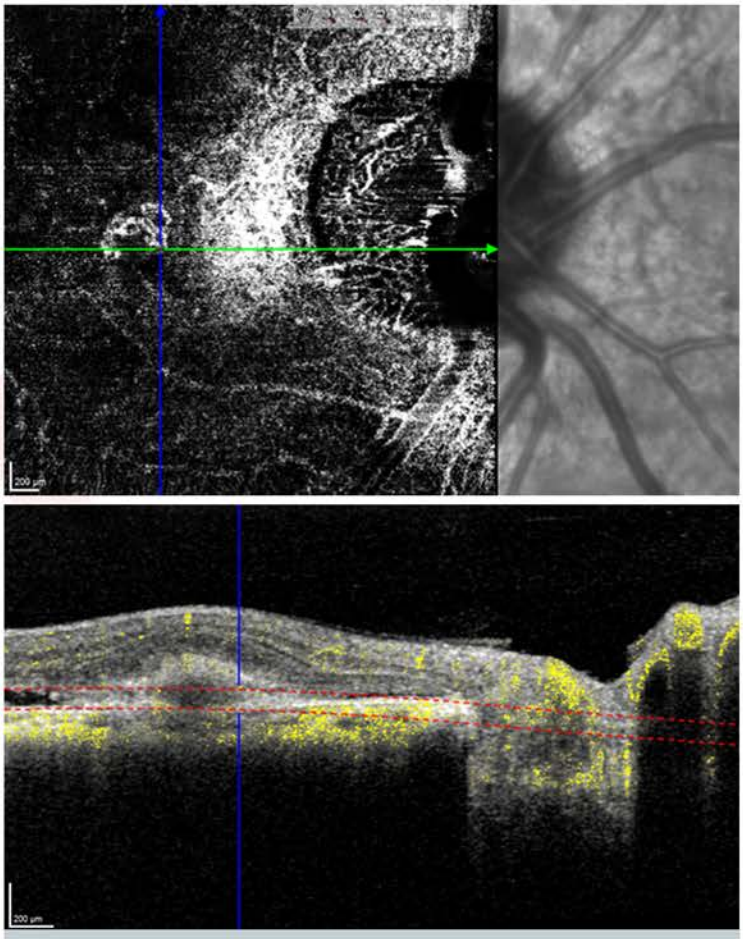

Figure 3: OCTA of the right eye demonstrating sub-retinal flow and branching pattern consistent with a macular CNVM

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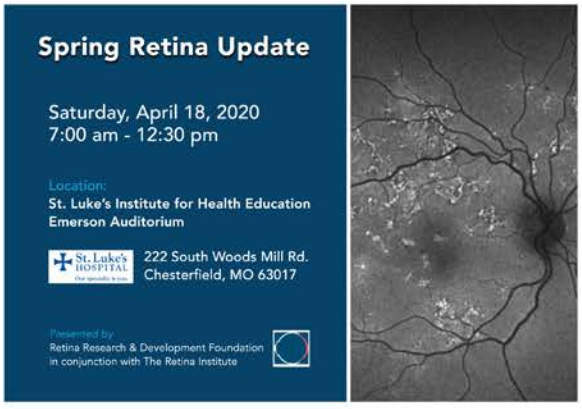

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