



## Streak of Concern

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### Introduction:

A 37-year-old female presents for initial consultation to The Retina Institute from an outside provider with a 1 week history of a painless distortion in the right eye. She describes seeing a 'wave' in the center of her vision. She states she had similar symptoms in her left eye prior to losing her central vision. She denies any major medical problems. A 12 point review-of-systems was otherwise negative.

### Exam:

Best corrected visual acuity was 20/40 in right eye and 20/200. No relative afferent pupillary defect was noted. Confrontation to visual field and extraocular motility was intact. Anterior segment examination was otherwise unremarkable.

DFE revealed orange-red linear irregularities extending radially from the optic nerve into the peripheral fundus in both eyes (Figures 1 & 2). These linear changes were better visualized on fundus autofluorescence (Figures 3 & 4). SD-OCT through

the macula of the left eye revealed changes consistent with fibrosis and atrophy. In the left eye, SD-OCT showed break in Bruch's membrane, sub-retinal hyper-reflective material, and associated sub-retinal fluid in the (Figure 5). Fluorescein angiography (FA) of the right eye showed a perifoveal lesion with early hyperfluorescence and late staining, consistent with a choroidal neovascular membrane (CNVM).

### Discussion:

The linear streaks depicted in this case are angioid streaks, which are defects in Bruch's membranes. Angioid streaks have many systemic associations summarized by the 'PEPSI' mnemonic – Pseudoxanthoma Elasticum Paget's disease Sickle-cell/thalassemia/spherocytosis Idiopathic. Of note, Ehlers-Danlos syndrome, once part of the mnemonic, is no longer believed to be associated with angioid streaks<sup>1</sup>.

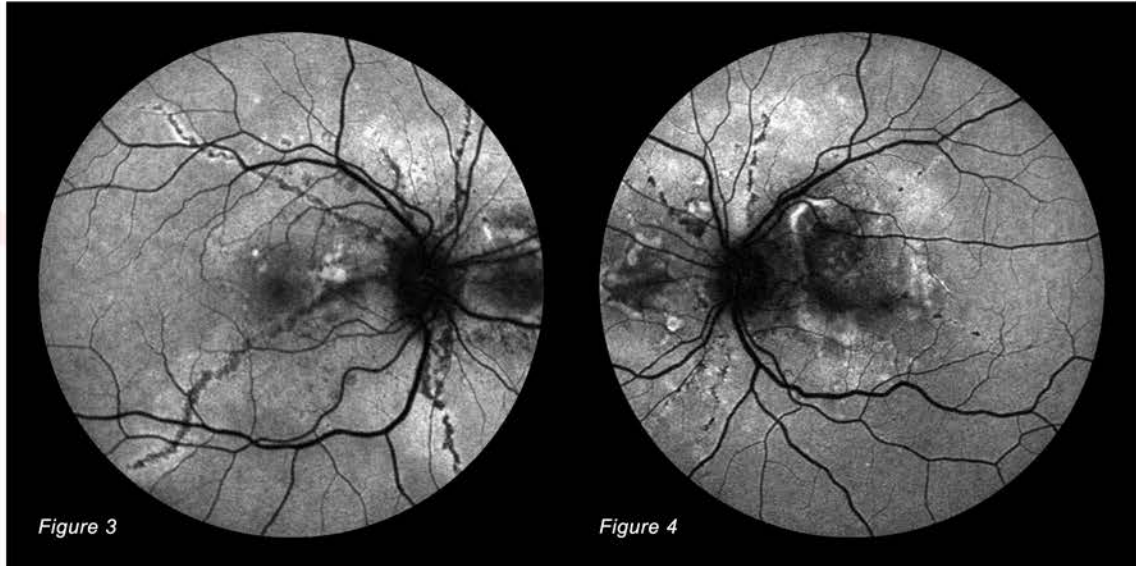
Angioid streaks may cause visual disturbances in multiple ways<sup>2</sup>. First, the streak may traverse the fovea, leading to RPE disruption.



Figure 1

Figure 2

Second, mild trauma to the eye may cause the choroid to rupture at these areas, leading to submacular hemorrhage. Third, as in our case, secondary choroidal neovascularization can occur. Figure 5 demonstrates the break in Bruch's membrane, the associated CNVM and subretinal fluid.



Our patient had characteristic skin findings on her neck which were biopsied to confirm a diagnosis of pseudoxanthoma elasticum. Pseudoxanthoma elasticum (PXE) is an inherited multi-system disorder characterized by ectopic mineralization and fragmentation of elastic fibers in the skin, the elastic laminae of blood vessels and Bruch's membrane in the eye<sup>3</sup>.

Defects in an ATP-binding cassette (ABC) transporter gene in ABCC6 on chromosome 16 are responsible for the disease<sup>4</sup>. In addition to angioid streaks, other ophthalmic manifestations of this disease include a 'peau d'orange' fundus appearance, which may appear as mottled dark spots on a lighter background.

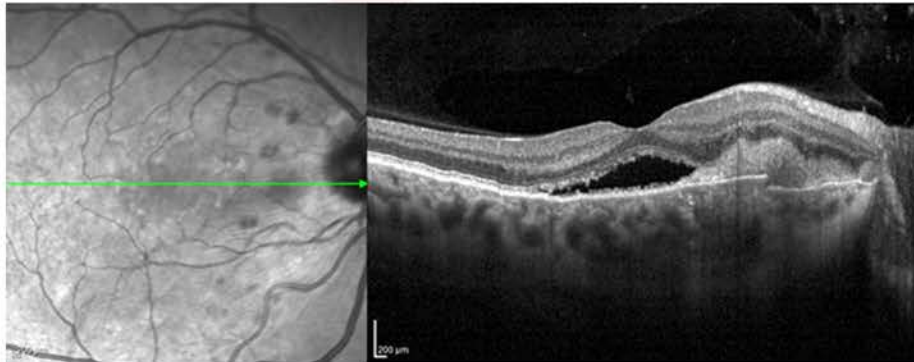


Figure 5: OCT showing subretinal fluid, the break in Bruch's membrane and associated CNVM.

Our patient had a new choroidal neovascular membrane in the right eye, and subretinal fibrosis from a prior neovascular membrane in the left eye. The patient elected for treatment with anti-VEGF agents in the right eye.

**References:**

1. Singman EL. Angioid Streaks Are Not a Common Feature of Ehlers-Danlos Syndrome. *J A M A Ophthalmology* 2018.
2. Georgalas I, Papaconstantinou D, Koutsandrea C, et al. Angioid streaks, clinical course, complications, and current therapeutic management. *Ther Clin Risk Manag* 2009;5:81-9.

3. Gliem M, Zaeytijd JD, Finger RP, Holz FG, Leroy BP, Charbel Issa P. An update on the ocular phenotype in patients with pseudoxanthoma elasticum. *Front Genet* 2013;4:14.
4. Chassaing N, Martin L, Calvas P, Le Bert M, Hovnanian A. Pseudoxanthoma elasticum: a clinical, pathophysiological and genetic update including 11 novel ABCC6 mutations. *J Med Genet* 2005;42:881-92.

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