

What You Should Know About Angioid Streaks

By David J. Browning, MD, PhD

The eye wall has several layers, as shown in figure 1. Proceeding from the inside of the eye to the outside, the layers are as follows:

1. *Retina* – A nerve lining which converts light into nervous impulses.
2. *Retinal Pigment Epithelium* – A layer of cells which recycles vitamin A for the retina and which removes waste produced by the retina.
3. *Bruch's Membrane* – A layer of elastic tissue and collagen, which separates the retina from the choroid.
4. *Choroid* – A layer of nutritive blood vessels providing oxygen to the retinal photoreceptors.
5. *Sclera* – The tough white outer wall of the eye.

Figure 1a. Cross-section of the Human Eye

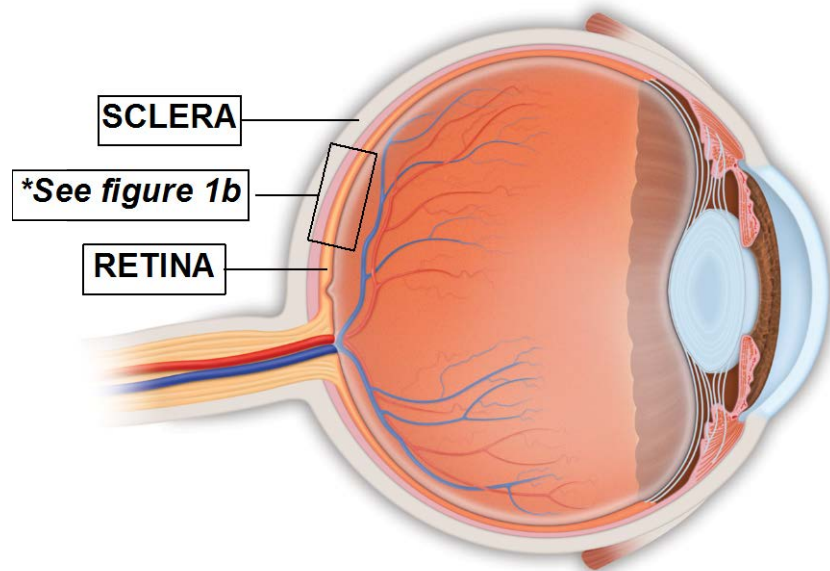
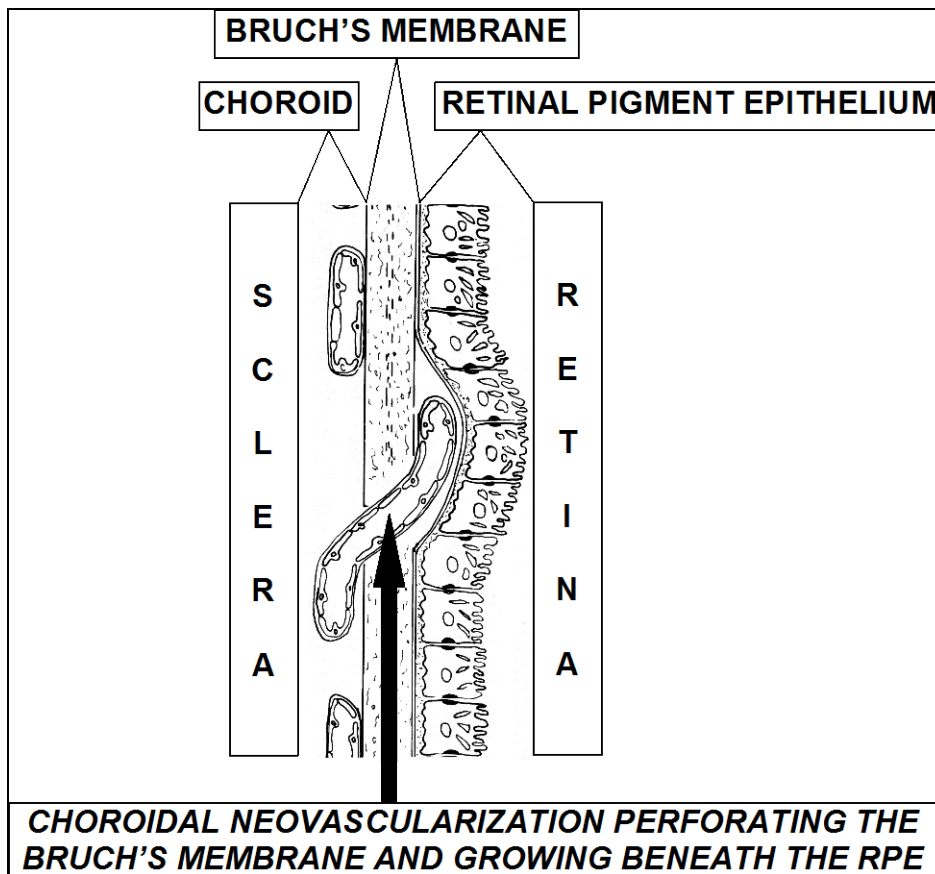


Figure 1b. Cross-section of the layers of the human eye



In angioid streaks, the elastic fibers in the Bruch's membrane crack, which can allow abnormal blood vessels originating from the choroid to grow up under the retina (fig. 1b). There they can bleed and form scar tissue with loss of vision.

What Are The Causes of Angioid Streaks?

Angioid streaks can be caused by many diseases. The most common cause is pseudoxanthoma elasticum (PXE), an inherited disease affecting elastic tissues throughout the body. Patients with PXE and angioid streaks often have other eye findings including disc drusen, peau d'orange, and mid-peripheral fundus comet-lesions. Disc drusen are calcium deposits that develop in the optic nerve, sometimes compromising visual field. Peau d'orange is the French term for "skin of the orange", which describes the pebbly look to the outer retina temporal to the macula on fundus examination. Comet-lesions are calcific deposits with pigmented "tails" like those of a comet. These lesions are seen on dilated fundus examination.

Patients with PXE not only have eye changes, but they have disease of the arterial walls throughout the body, and in elastic tissues of the skin and the gastrointestinal tract. They often develop skin changes of the arm crease and the neck likened to the “plucked chicken” appearance we have all seen. When skin biopsies are taken, the elastic fibers appear frayed and these microscopic changes can be detected before the yellow, “plucked chicken” dots can be noticed by the unaided eye. The arterial and gastrointestinal changes make patients with PXE more susceptible than unaffected people to problems such as stroke, heart attack, and gastrointestinal bleeding. Gastrointestinal bleeding can cause weakness, pallor of the skin, and dark, tarry stools. Patients with PXE need to know these symptoms and report to their doctor immediately should they occur. Notification of and follow-up by the family doctor or internist is crucial to reducing these risks. Patients should eat a heart healthy diet, engage in exercise, avoid obesity, and quit smoking to further reduce the risk of heart disease and stroke.

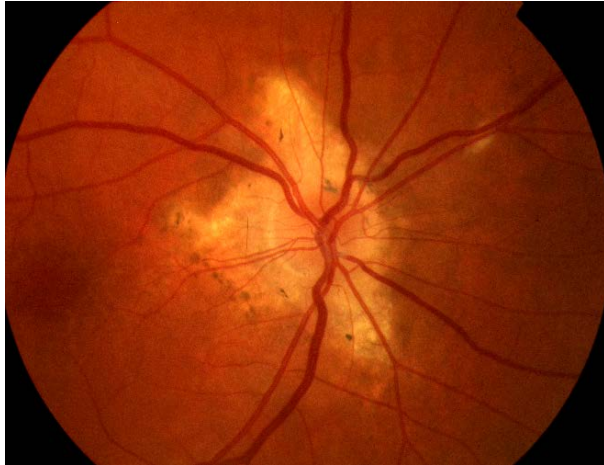
Pseudoxanthoma elasticum is a genetic disease. Mutations occur in the ABCC6 gene of chromosome 16p13.1. Some patients have an autosomal dominant form of the disease and can pass the gene to approximately half of their offspring. Most, with the recessive form, have smaller chances of transmission. Family members should be checked to help determine the inheritance pattern.¹

Other causes of angioid streaks include sickle cell disease, Paget's disease of the bone, hereditary spherocytosis, familial hyperphosphatemia, abeta lipoproteinemia, and thrombotic thrombocytopenic purpura. Approximately 2% of patients with sickle cell disease develop angioid streaks. The other conditions mentioned are exceptionally rare.

Ocular Signs of Angioid Streaks

The most ominous sign is the growth of abnormal blood vessels through the streaks and under the retina. This growth, called choroidal neovascularization, can cause distortion and blurred spots in the field of vision. Figure 2 shows angioid streaks and figure 3 shows choroidal neovascularization from a streak. Laser treatment can be used to halt these abnormal blood vessels, but they have a high chance of recurrence – perhaps 75% over a 3-year period. Moreover, there is a high chance that

the second eye will develop choroidal neovascularization once one eye has – perhaps 60-70% over a 3-year period. More recently, and with greater effectiveness, patients developing choroidal neovascularization have been treated with serial injections of drugs into the eye that inhibit vascular endothelial growth factor. The most commonly used drug is bevacizumab



(brand name Avastin). Ranibizumab and aflibercept are two other drugs used for this purpose. They are much more expensive than bevacizumab.^{2,3} By checking a grid of lines, called an Amsler Grid, at home, and notifying the ophthalmologist promptly if changes in the appearance of the grid are noted, loss of vision may be minimized by timely injections of bevacizumab into the eye.

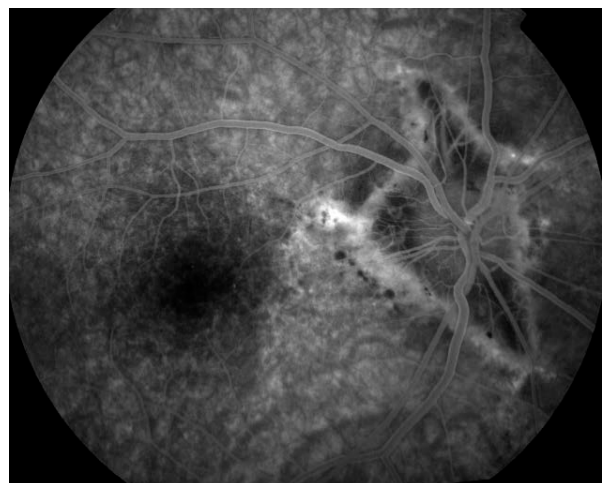
Figure 2. Color photo of angioid streaks.

Identification of the source and extent of choroidal neovascularization requires fluorescein angiography. This test is performed by a Certified Retinal Angiographer and involves two parts. First, they inject a solution of approximately 2 tablespoons of food dye into one of the veins of the arm. Then, they use a special camera to photograph the eye as the dye circulates through and highlights the problem areas. These photographs are essential to the ophthalmologist in developing a treatment plan. The dye from this test will temporarily discolor the urine and give the skin a slight yellow tinge. The dye is not related to iodine and the photographs are not x-rays.

Figure 3. Fluorescein angiogram of angioid streaks with choroidal neovascularization.

Final Comments

Angioid streaks render the eye more susceptible to hemorrhaging after minor trauma. Patients with angioid streaks should avoid rubbing



the eyes and should wear protective eyeglasses when engaging in sports in which blows to the eye might occur. All patients with angioid streaks should check the Amsler Grid periodically and notify the ophthalmologist if changes occur.

If you would like to pursue further information about angioid streaks and other medical conditions, an excellent resource is the website of the National Library of Medicine. You can find the database of medical publications by typing the term Pubmed into any search engine, or by using the following address: <http://www.ncbi.nlm.nih.gov/entrez/query.fcgi>.

Updated 11-12-2016

References

1. Tan MH, Vanakker OM, Terry SF, et al. Angioid streaks with severe macular dysfunction and generalized retinal involvement due to a homozygous duplication of the ABCC6 gene. *Eye* 2012;26:753-755.
2. Vadala M, Pece A, Cipolla S, Monteleone C, Ricci F, Boscia F, Cillino S. Angioid Streak-Related Choroidal Neovascularization Treated by Intravitreal Ranibizumab. *Retina* 2010;30:903-907.
3. Gliem M, Finger RP, Fimmers R, Brinkmann CK, Holz FG, Iss PC. Treatment of choroidal neovascularization due to angioid streaks, a comprehensive review. *Retina* 2013;33:1300-1314.