

Angioid Streaks and Optic Disc Drusen in Pseudoxanthoma Elasticum

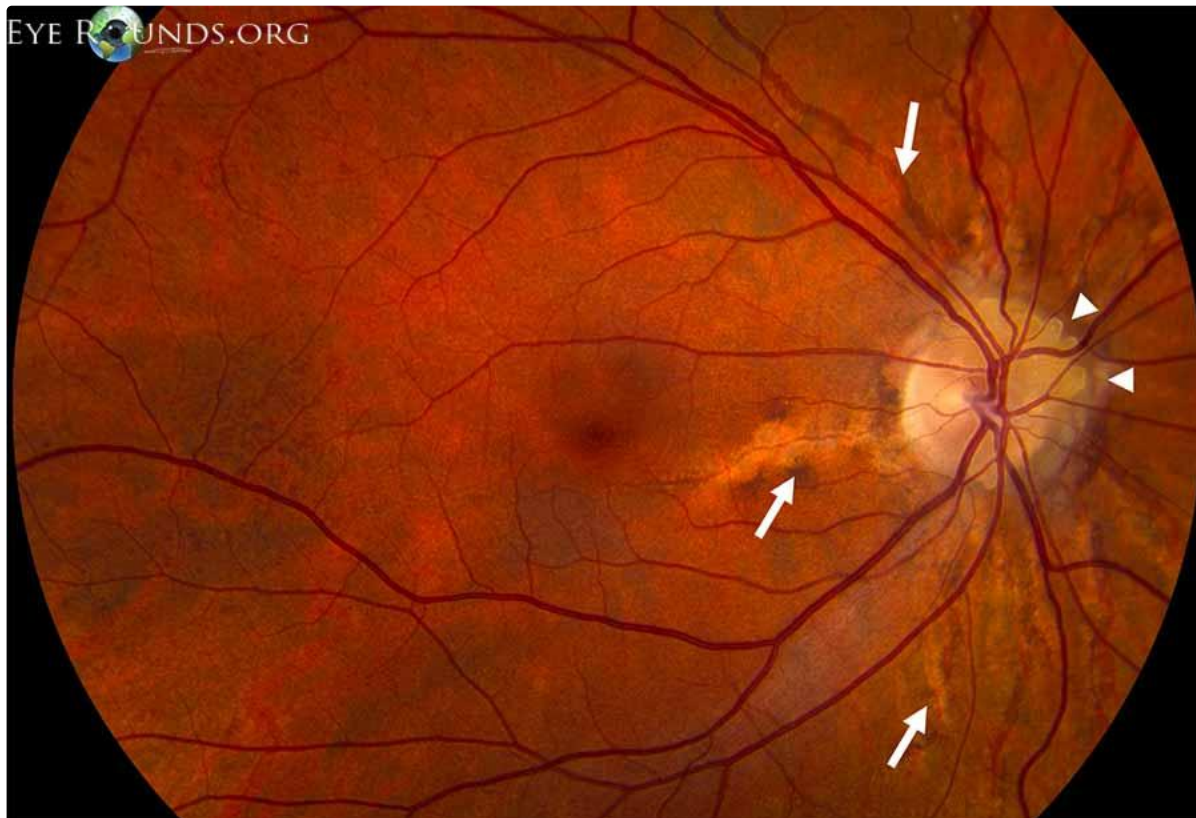
Category(ies): Genetics, External Diseases, Retina

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34-year-old female with a recent diagnosis of pseudoxanthoma elasticum (PXE) by skin biopsy, one month prior to presentation. She had no visual complaints.

- BCVA cc:
 - OD 20/20
 - OS 20/20
- IOP: OD 16, OS 18



Right Eye. Disk drusen (arrow heads); normal vessels; angioid streaks (arrows) emanating from the optic nerve

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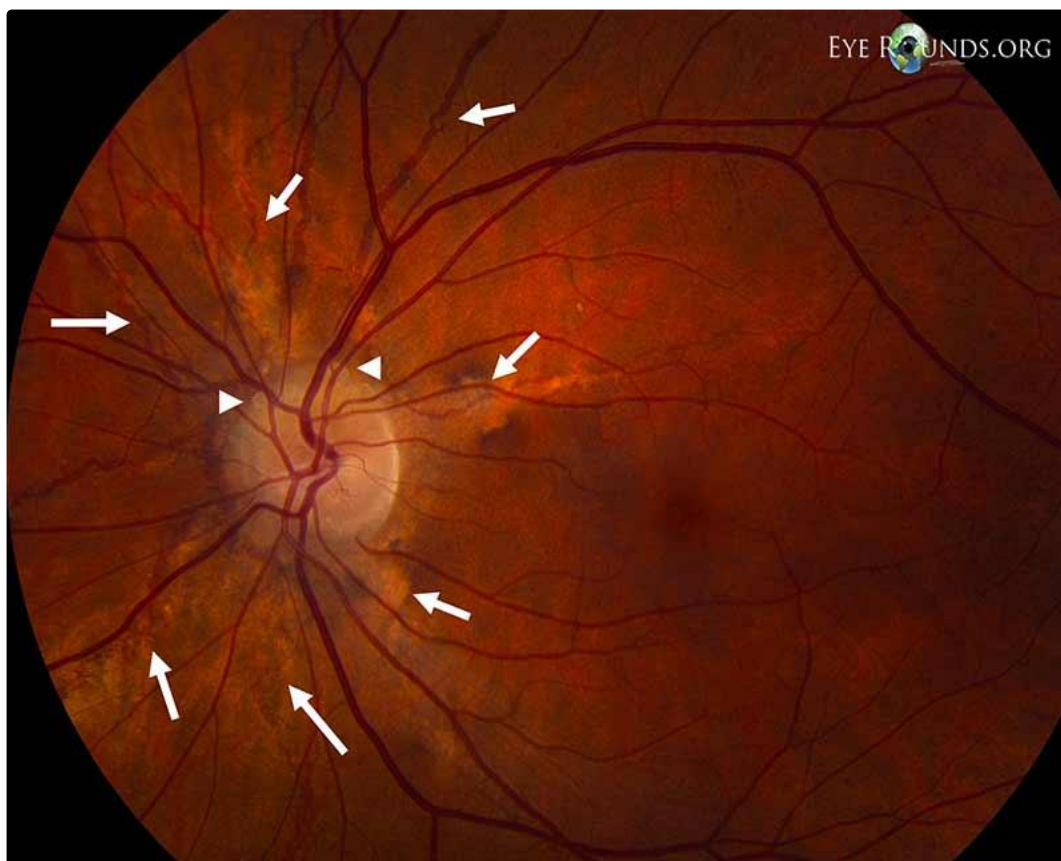




Right Eye. Disk drusen

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Left eye: Disk drusen (arrow heads); normal vessels; angioid streaks (arrows) emanating from the optic nerve

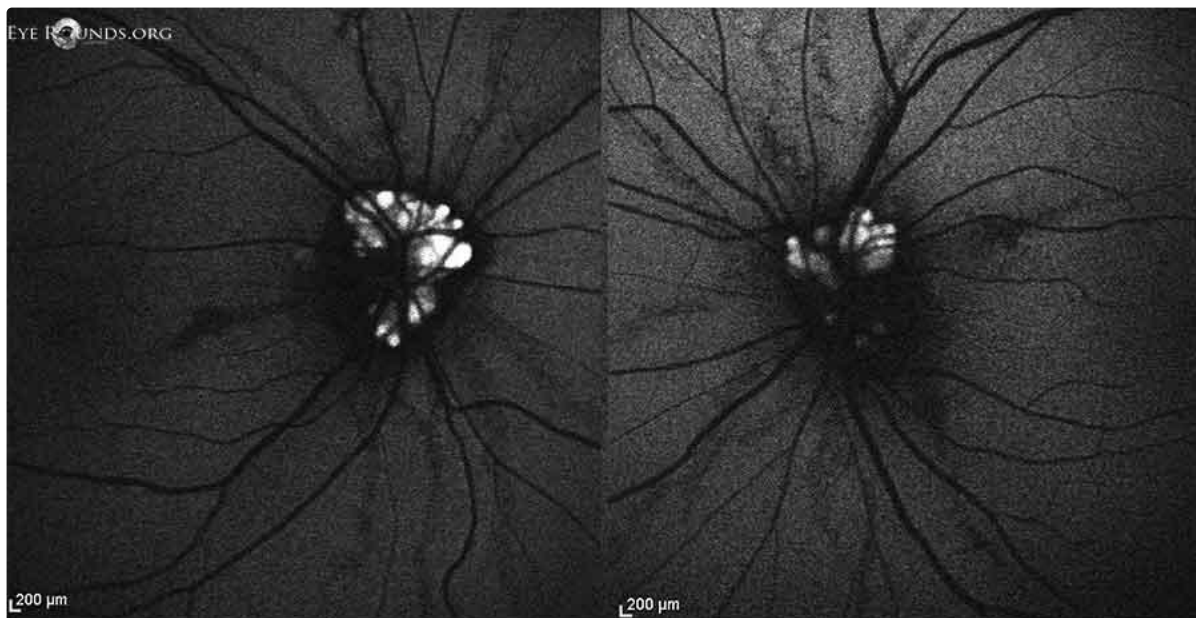
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Left Eye. Disk drusen

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Autofluorescence, both eyes. Areas of increased autofluorescence at the nerve corresponding to the disk drusen; radial areas of hypo-autofluorescence corresponding to angiod streaks. (click image for higher resolution)

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External Photo. Neck, right side view. Small papular lesions and cutaneous laxity of the neck ("plucked chicken skin") consistent with pseudoxanthoma elasticum. (click image for higher resolution)

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Angioid Streaks

- Named because of their resemblance to blood vessels.
- Result from crack-like breaks in weakened Bruch's membrane ("peau d'orange" appearance) that radiate out from the optic nerve.
- Seen with pseudoxanthoma elasticum (most commonly), Ehler's Danlos syndrome, Paget's disease, sickle cell anemia, or Idiopathic ("PEPSI"). Other associations include abetalipoproteinemia, acromegaly, diabetes mellitus, facial angiomas, hemochromatosis, hemolytic anemia, hereditary spherocytosis, hypercalcinosis, hyperphosphatemia, lead poisoning, myopia, neurofibromatosis, senile elastosis, Sturge-Weber syndrome, and tuberous sclerosis.
- Associated optic disc drusen are common.
- Usually asymptomatic even if they cross into the foveal area, but complications include choroidal neovascularization and choroidal rupture.

Pseudoxanthoma Elasticum

- Also known as Gronblad-Strandberg syndrome.
- Caused by an autosomal recessive mutation in the ABCC6 gene on chromosome 16p13.1
- Results in a fragmentation and mineralization of elastic-containing fibers in connective tissue.
- Small, yellowish papular lesions form and cutaneous laxity mainly affects the neck, axilla, groin, and flexural creases.
- Eye findings are present in almost all PXE patients, and usually noticed a few years after the onset of cutaneous lesions.
- Gastrointestinal and cardiovascular systems may also be affected.

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