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Case Report

Interesting case of groenblad-strandberg syndrome

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ABSTRACT

A 40-year old female came with complaints of headache for 5 days. Her general examination and vitals were within normal limits. On complete ophthalmic examination we found there are irregular streaks radiating from disc in the fundus of both the eyes suggesting of angioid streaks. We did a dermatological evaluation in view of increased stretchability of skin and papules over neck & thigh and diagnosed as pseudoxanthoma elasticum. So we diagnosed as angioid streaks with pseudoxanthoma elasticum also known as Groenblad-Strandberg syndrome.

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1. Introduction

Angioid streaks are crack-like dehiscences in brittle thickened and calcified bruch membrane. Approximately 50% of patients with angioid streaks have systemic infection such as pseudoxanthoma elasticum, Ehlers-Danlos syndrome, Paget's disease and Sickle cell trait & disease. But most commonly associated with pseudoxanthoma elasticum. They usually develop in 2nd-3rd decade and are not seen in children. Angioid streaks results from crack like breaks in Bruch's membrane, which is abnormal in its structural composition predisposing to these localized areas of rupture.¹ It is because secondary to blunt trauma or may occur spontaneously, even with minor injury.²

2. Case History

A 40-year old female came with complaints of headache since 5 days not associated with nausea and vomiting. No complaints of photophobia. Patient did not complain any visual related complaints. Not a known case of any systemic diseases like systemic hypertension, diabetes etc.

Her general examination and vitals are normal.

On ocular examination, visual acuity in both the eyes was 6/6, colour vision was unaffected, extra-ocular motility was full & free in both the eyes & anterior segment was normal. Intraocular pressure (IOP) was found to be 16 and 14 mm hg respectively in right and left eyes. Fundus examination revealed irregular streaks radiating from the disc and extending radially for a variable distance. OCT picture showed us breaks in bruch's membrane. So diagnosed as angioid streaks.

On dermatological examination, patient complaints of increased stretchability of skin and found to have tiny skin coloured papules over neck and thigh with hyperextensibility noted all over body. Histopathology was done diagnosing those skin lesions as pseudoxanthoma elasticum or groenblad-strandberg syndrome.

Patient was advised to take tablet Vitamin C 500 mg, tablet Antoxid HC and to apply sunscreen lotion over skin lesions.

Safety glasses are advised for patients because they are highly susceptible for choroidal rupture and multiple subretinal bleed following even minor blunt injury.

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Contact sports should be avoided by patients having angioid streaks.

Patient being followed up in our OPD every 3 months for her fundus status and it remains the same. Patient is still being followed up.

Table 1: On ocular examination

Anterior segment	Right eye	Left eye
Visual acuity	6/6 25/25	6/6 25/25
Colour vision		
Extraocular movements	Full and free	Full and free
Eyelids	Normal	Normal
Conjunctiva	Normal	Normal
Cornea	Clear	Clear
Anterior chamber	Normal depth	Normal depth
Iris	Normal colour and pattern	Normal colour and pattern
Pupil	3-4 mm, round and reacting to light	3-4 mm, round and reacting to light
Lens	Clear	Clear

Table 2: On ocular examination

Posterior segment	Right eye	Left eye
90 D	Media-clear Disc & vessels – normal CDR-0.3 Angioid Streaks	Media-clear Disc & vessels – normal CDR-0.3 Angioid Streaks

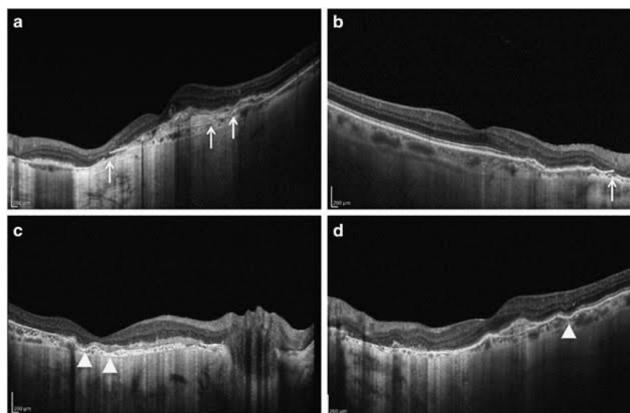


Fig. 2: OCT picture showing breaks in bruchs membrane



Fig. 3: Multiple skin coloured papules over neck and face

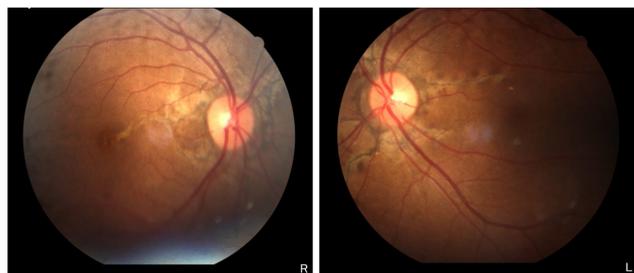


Fig. 1: Fundus photo of both right and left eye showing irregular streaks surrounding the optic disc and extending radially for a variable distance from there suggestive of Angioid streaks

Histopathology of skin biopsy showing remarkable epidermis and mid dermis shows fragmented, tortuous, amorphous to basophilic collagen bundles seen as aggregated with interwoven sclerotic areas these are seen extending to lower dermis which gives impression of pseudoxanthoma elasticum.

3. Discussion

The diagnosis of Angioid streaks was done on fundus examination where we found irregular streaks surrounding the optic disc and extending radially for a variable distance. She has multiple tiny skin coloured papules over neck and thigh with hyperextensibility noted all over body. On skin biopsy it was compatible with pseudoxanthoma elasticum. And we know that Angioid Streaks was most commonly associated with pseudoxanthoma elasticum.²

Angioid streaks are crack like dehiscences in brittle thickened and calcified bruch membrane. They are visible, linear, irregular, crack-like dehiscences of Bruch membrane, which may be unilateral or most commonly bilateral. Approximately 50% of patients with angioid streaks have a systemic disease such as pseudoxanthoma elasticum, Ehlers-Danlos syndrome, Paget's disease and

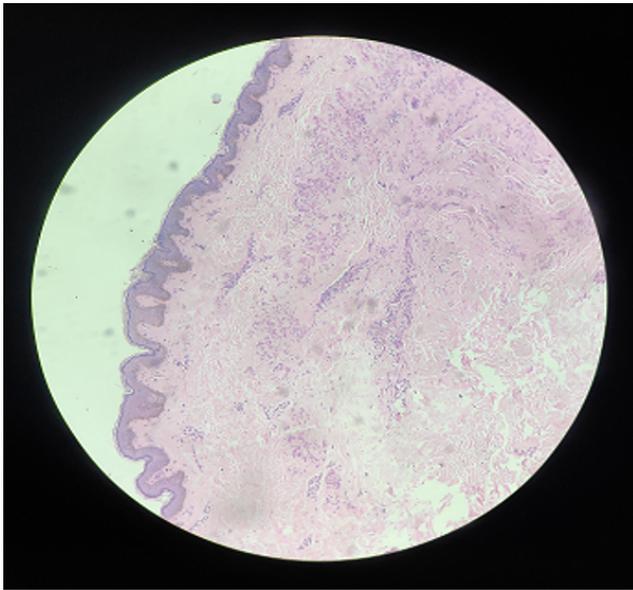


Fig. 4:

Sickle cell disease. But most commonly associated with pseudoxanthoma elasticum. They usually develop after the second decade of life.³

In pseudoxanthoma elasticum, incidence is 85% based on whether the pseudoxanthoma was diagnosed clinically or with a skin biopsy. Practically, all patients with pseudoxanthoma elasticum would have developed angioid streaks 20 years after first diagnosis.⁴

Bruch membrane is elastin and collagen-rich membrane attached to the retinal pigment epithelium (RPE) that is involved in the transport of nutrients and metabolic waste products that separates the RPE from the choriocapillaris.²

The bruch membrane elastin-rich mineralization is responsible for the pathogenesis of angioid streaks, especially in patients with pseudoxanthoma elasticum, when there is lack of systemic antimineralization factor that results in a calcification of connective tissues rich in elastic fibers, as is Bruch membrane, with profound deposition of calcium elements in the Bruch membrane.

The pathogenetic pathway for the development of angioid streaks includes: at the start, a thickening of Bruch membrane and/or a decrease of the pigment granules, pigment stripping, mottling or formation of pigment clumps in the RPE exist.

These findings can occur with or without concurrent disruption in the overlying layers of the retina and the underlying choriocapillaris. The disease can progress to the development of choroidal neovascularization (CNV), emanating from the growth of fibrovascular tissue through the localized defect.^{3,5}

Specifically, if the mechanical integrity of Bruch membrane is interrupted, it can result in modification

of growth factor agents interacting with a potential communication between the retina and choroid, leading to Choroidal neovascularisation formation. At the final stage of the disease's natural course, the Choroidal neovascularisation can result in a disciform scar with detrimental implications for the vision.²

There is relationship between angioid streaks and peripapillary chorioretinal atrophy or peripapillary choroidal sclerosis because these entities may be precursors of the subsequent development of angioid streaks.

Angioid streaks results from crack like breaks in Bruch's membrane, which is abnormal in its structural composition predisposing to these localized areas of rupture. This process may occur spontaneously or could be secondary to blunt trauma, even very minor. In a individual with angioid streaks, scleral depression should generally be withheld as this could theoretically contribute to the formation of additional breaks in Bruch's membrane.⁴

They are generally asymptomatic but can cause visual impairment when involvement of macula causes secondary complications such as choroidal neovascularization or subretinal hemorrhages.^{1,2}

Angioid streaks may occur in association with other fundus features such as Peau De orange appearance, choroidal neovascularization and disciform macular degeneration. Visual impairment occurs eventually in over 70% of patients.

The systemic disease most commonly associated with angioid streaks is pseudoxanthoma elasticum or Gronblad-Strandberg syndrome, a predominantly autosomal recessive disorder. Gronblad-strandberg syndrome, characterised by dystrophic calcification of elastic tissues and progressive abnormal mineralization.⁴

Histopathologic breaks in Bruch's membrane correspond to the clinical location of angioid streaks. Ingrowth of fibrovascular tissue may occur from the choroid into the subretinal pigment epithelial space, which is abnormally thin in the region of angioid streak, causing subretinal hemorrhage, choroidal neovascularization [CNV] and disciform scarring, Regardless of any underlying systemic association, the pathology of angioid streaks is similar.²

Regarding treatment angioid streaks are usually asymptomatic requiring observation only. When associated with Choroidal neovascularisation, laser photocoagulation and photodynamic therapy have been previously used with poor results and frequent recurrences. Advent of intravitreal anti-VEGF treatments led to more promising results.¹

4. Conclusion

Ophthalmic examination of a patient confirmed Angioid Streaks and skin biopsy of tiny skin coloured papules over neck and thigh gives impression of pseudoxanthoma elasticum. Angioid streaks must be differentiated from pseudo-angioid streaks and lacquer cracks seen in

pathological myopia because the latter is not associated with any systemic disease.

Safety glasses are advised for patients because they are highly susceptible for choroidal rupture and multiple subretinal bleed following even minor blunt injury, especially during participation in contact sports. Contact sports should be avoided by patients having angioid streaks.

Medical consultation is indicated to evaluate for systemic manifestations of pseudoxanthoma elasticum, calcific arteriosclerosis and gastro intestinal and cerebrovascular bleeding.

As angioid streaks is associated with systemic diseases, whenever a patient is diagnosed with pseudoxanthoma elasticum, ehler danlos syndrome, pagets disease etc. should undergo ophthalmic evaluation eventhough patient doesn't have any complaints.

5. Source of Funding

None.

6. Conflict of Interest

None.

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