International Journal of Clinical Science and Medical Research

ISSN(print): 2770-5803, ISSN(online): 2770-582X

Volume 05 Issue 11 November 2025

DOI: https://doi.org/10.55677/IJCSMR/V5I11-07/2025, Impact Factor: 8.005

Page No : 302-305

Angioid Streaks Revealing Pseudoxanthoma Elasticum: About Three Cases

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ABSTRACT

Purpose: To describe three cases of angioid streaks revealing pseudoxanthoma elasticum (PXE) and to discuss the diagnostic, clinical, and therapeutic aspects of this uncommon association.

Methods: Observational study of three patients managed at the Ophthalmology Departments of the Specialty Hospital of Rabat and Hassan II Military Hospital of Laâyoune.

Results: Three patients (two men and one woman, aged 58, 55, and 42 years) presented with progressive bilateral visual loss. Fundus examination revealed dark, radially oriented lines emerging from the optic disc, consistent with angioid streaks. Fluorescein angiography confirmed the diagnosis. Dermatologic evaluation revealed pseudoxanthoma elasticum, while cardiovascular and skeletal investigations were unremarkable.

Conclusion: Angioid streaks represent a rare manifestation of Bruch's membrane rupture, most frequently associated with PXE (Grönblad–Strandberg syndrome). Early detection of choroidal neovascularization and prompt anti-VEGF therapy are essential to preserve visual function.

Published Online: November 17, 2025

KEYWORDS:

Angioid streaks; Pseudoxanthoma elasticum; Grönblad–Strandberg syndrome; Choroidal neovascularization; Anti-VEGF.

INTRODUCTION

Angioid streaks (AS) are linear ruptures of Bruch's membrane that appear on fundus examination as reddish-brown lines radiating from the optic disc [1]. They result from degeneration and calcification of elastic fibers, producing structural fragility. The main systemic association is pseudoxanthoma elasticum (PXE), a rare inherited disorder caused by ABCC6 gene mutations leading to aberrant mineralization of elastic tissues [2, 3]. The coexistence of AS and PXE defines the Grönblad–Strandberg syndrome, responsible for severe ocular and systemic complications [4, 5].

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Cite this Article Z. Filali, H. Boui, H. Lazaar, L.O. Cherkaoui (2025). Angioid Streaks Revealing Pseudoxanthoma Elasticum: About Three Cases. International Journal of Clinical Science and Medical Research, 5(11), 302-305

CASE REPORTS

Case 1: A 58-year-old man with high myopia presented with progressive bilateral visual loss. Fundus examination revealed radiating angioid streaks extending from the optic disc. Fluorescein angiography demonstrated alternating hyper- and hypofluorescent linear patterns confirming the diagnosis [6]. No choroidal neovascularization (CNV) was detected.

Case 2: A 55-year-old man reported gradual bilateral decrease in visual acuity. Dermatologic examination revealed yellowish papular lesions in a reticular pattern on the neck and axillae, typical of PXE [7]. Cardiovascular assessment was within normal limits.

Case 3: A 42-year-old woman with moderate myopia presented with bilateral macular pigment changes. Fundus findings: typical angioid streaks with a cracked appearance of Bruch's membrane. Fluorescein angiography confirmed the absence of CNV but showed irregularities of the membrane [8].

RESULTS

All three patients exhibited bilateral angioid streaks confirmed by angiography [6]; dermatologic confirmation of PXE [7]; and normal cardiovascular and skeletal findings. No neovascular complications were observed initially. Regular OCT follow-up was initiated for all patients.

DISCUSSION

AS often represent the first ocular sign of systemic disease, particularly PXE [9]. PXE primarily affects women (female:male ≈ 2:1) with a prevalence of approximately 1 in 25 000 [10]. Mutations in the ABCC6 gene (16p13.1) impair pyrophosphate transport—a natural inhibitor of calcification—causing ectopic mineralization of elastic fibers in skin, vessels and Bruch's membrane [2, 3, 11]. This brittleness leads to membrane rupture and AS formation [1, 12].

Clinically, AS appear as radial reddish-brown streaks radiating from the optic disc. Fluorescein angiography shows early linear hyperfluorescence with possible pigmentary hypofluorescence [6, 12]. OCT-angiography (OCT-A) enables non-invasive detection of subclinical CNV [13].

The most serious ocular complication is choroidal neovascularization (CNV), occurring in 70–80 % of AS patients [14, 15]. These CNV may cause subretinal hemorrhage and rapid visual loss [16].

There is no curative therapy for PXE or AS. Management includes avoidance of ocular trauma, regular OCT monitoring, and prompt anti-VEGF injections (ranibizumab, aflibercept) for CNV [17, 18]. Anti-VEGF therapy achieves functional stabilization in more than 80 % of cases [17–19]. For extrafoveal lesions, laser photocoagulation remains an option [20]

Visual prognosis depends on early detection and control of CNV. Semi-annual ophthalmologic evaluations and systemic monitoring (dermatologic, cardiac) are recommended [9, 10]. Recognition of AS should prompt a systemic evaluation for PXE or other causes (sickle-cell disease, Paget's disease) [4, 21]. Early multidisciplinary screening improves prognosis and prevents irreversible complications [11, 22].

CONCLUSION

Angioid streaks associated with pseudoxanthoma elasticum constitute a rare but distinctive ophthalmic manifestation of systemic disease. Timely diagnosis, OCT monitoring, and early anti-VEGF therapy are key to preserving vision. The ophthalmologist plays a central role in coordinating dermatologic, cardiovascular and genetic assessments within a multidisciplinary team.

Iconography

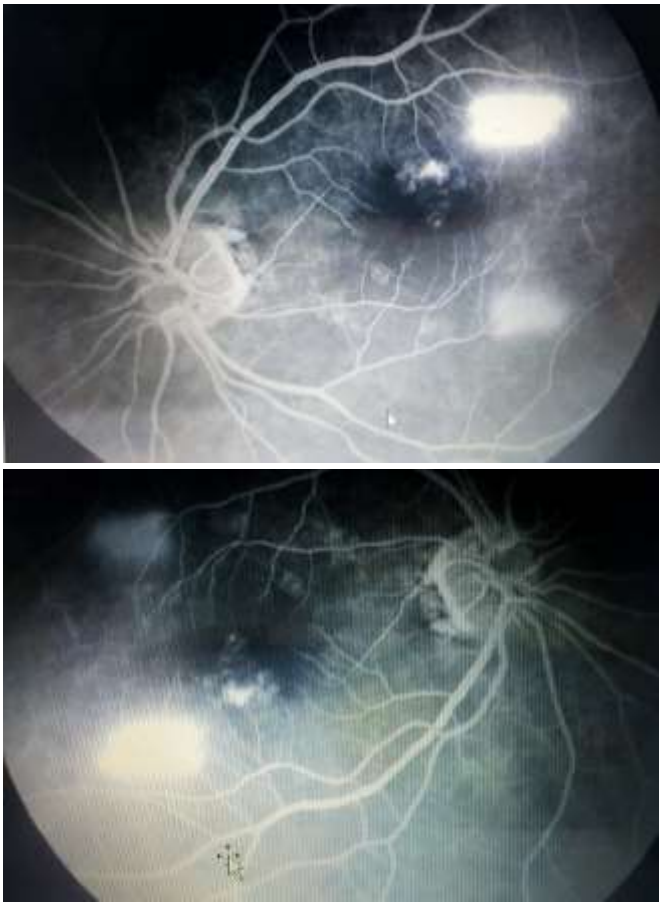


Figure 1. Bilateral fundus photograph of the first patient showing angioid streaks radiating from the optic disc in both eyes.

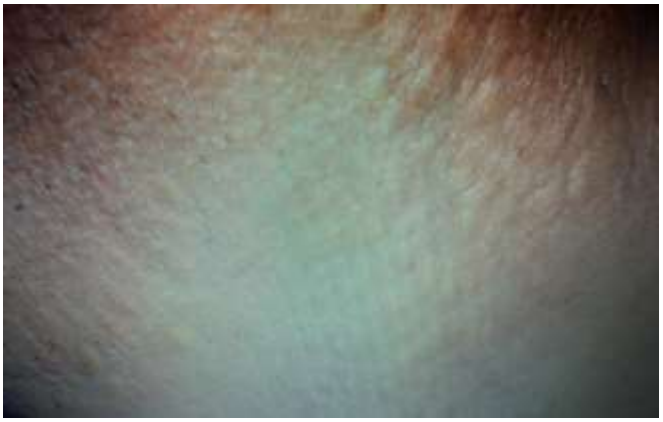


Figure 2. Neck and axillary skin lesions in the second patient, illustrating the yellowish reticulated pattern typical of pseudoxanthoma elasticum.

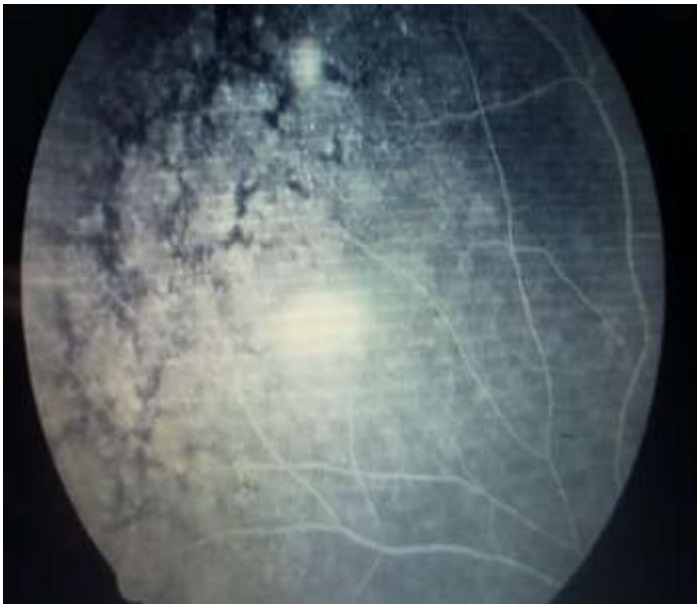
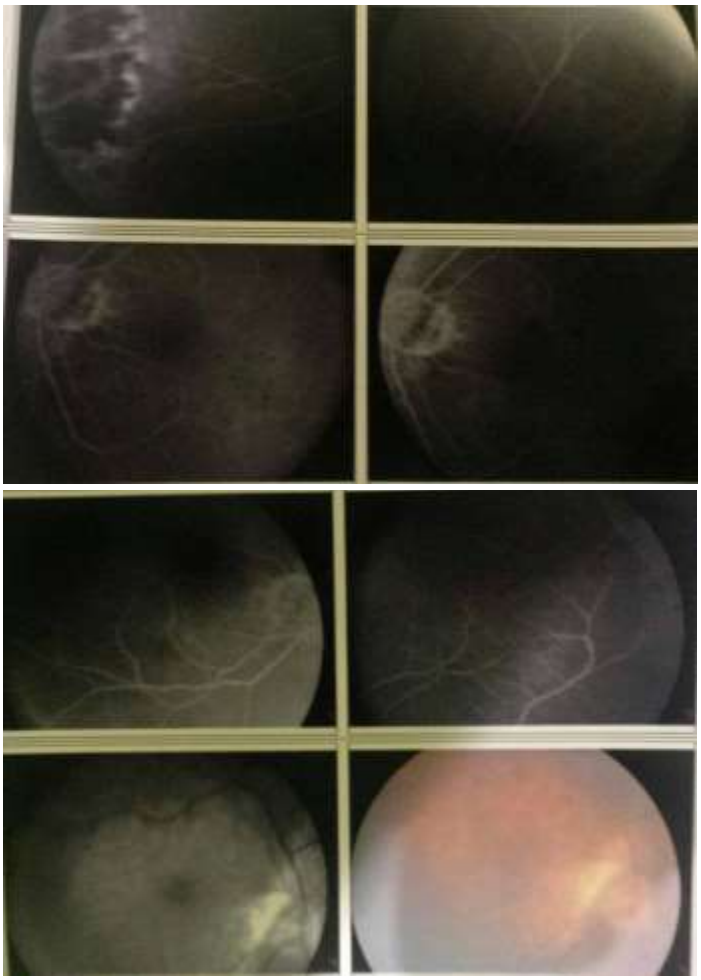


Figure 3. Cracked-like retinal appearance in the second patient consistent with membrane fragility and Bruch's membrane breaks.



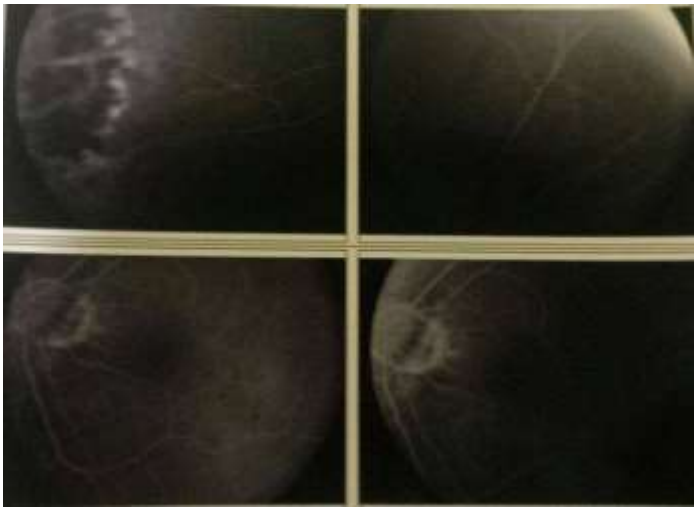


Figure 4. Fluorescein angiography of the third patient showing linear hyperfluorescence corresponding to angioid streaks without evidence of choroidal neovascularization.

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