

Retinal Pigment Epithelium Activation in Angioid Streaks Imaged With En Face Optical Coherence Tomography

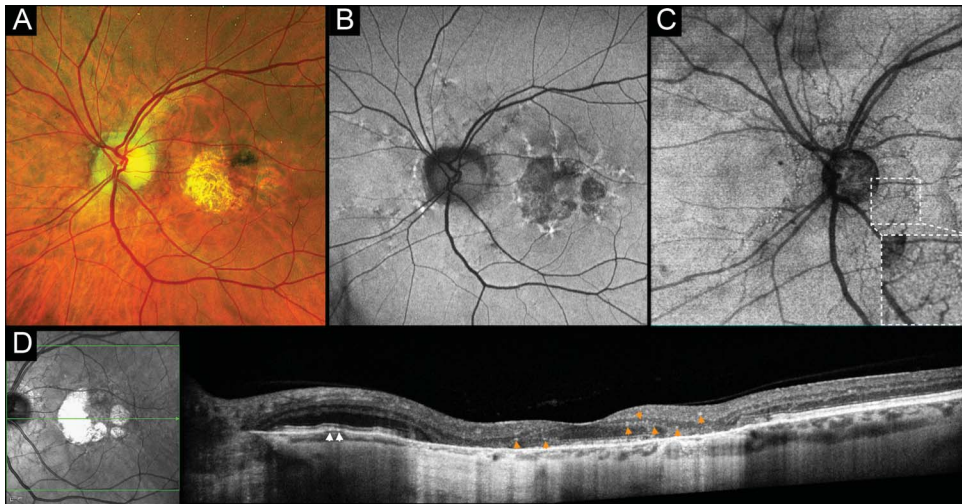


Fig. 1. **A.** Fundus photograph showed peripapillary AS and macular geographic atrophy. **B.** Fundus autofluorescence revealed a butterfly-shaped pattern dystrophy, and AS appeared as speckled hyperautofluorescent lines. **C.** En face OCT at the level of the RPE-BM complex enhanced the visualization of AS and demonstrated clustered hyperreflective dots. A 20-mm thick ellipsoid zone-based contour positioned at the depth of the RPE-BM was used to obtain en face OCT images. The dashed square is a magnified view highlighting the hyperreflective dots. **D.** The OCT B-scan through AS highlighted the mottled RPE-BM complex (arrowheads) and intraretinal hyperreflective foci in the fovea (orange

arrowheads). The green line indicates the location of the OCT B-scan.

Angioid streaks (AS) refer to linear, irregular, crack-like ruptures of a calcified Bruch membrane (BM) radially spreading from the optic nerve toward the retinal periphery. Pathogenic processes include mechanical forces exerted by the extraocular muscles on a fragile and less flexible posterior pole.¹ On the optical coherence tomography (OCT) B-scan, AS appear as a focal hyporeflective area of the retinal pigment epithelium (RPE)-BM complex.² Recently, en face OCT performed at the depth of the BM has demonstrated enhancement of the visualization and characterization of AS.³

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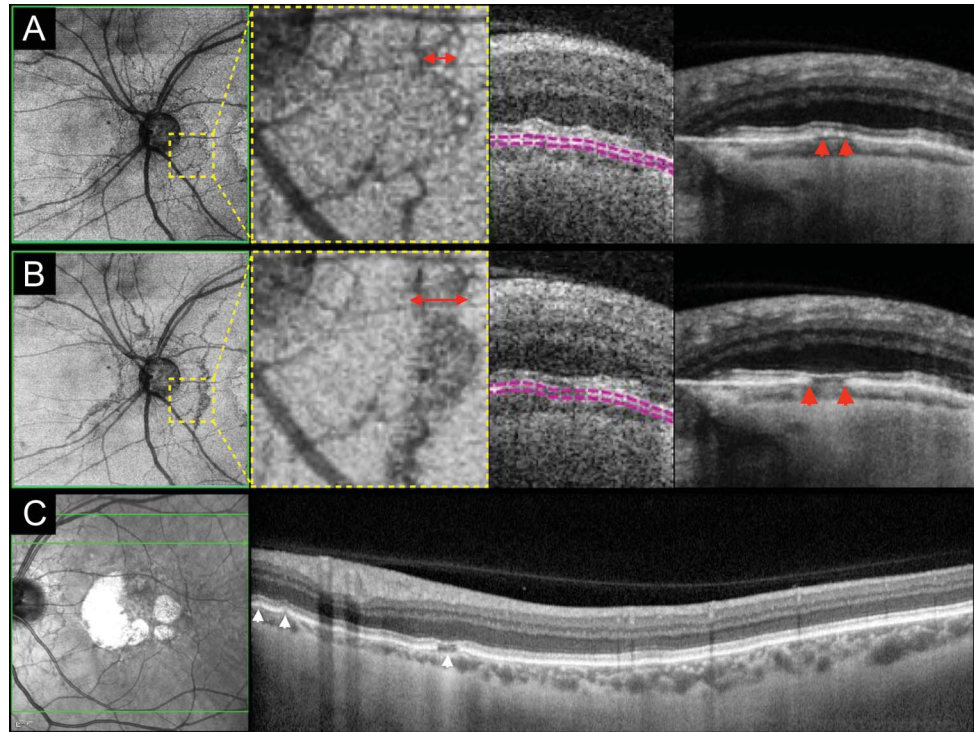
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We describe a 58-year-old man diagnosed with pseudoxanthoma elasticum. Fundus photography showed peripapillary AS and macular geographic atrophy (Figure 1A). Fundus autofluorescence revealed a butterfly-shaped pattern dystrophy (Figure 1B). Angioid streaks appeared as speckled hyperautofluorescent lines (Figure 1B). En face OCT at the level of the RPE-BM complex enhanced the visualization of AS and demonstrated clustered hyperreflective dots (Figure 1C) that colocalized with the mottled RPE-BM complex on the OCT B-scan (Figure 1D). Intraretinal hyperreflective foci were also documented in the fovea on the OCT B-scan (Figure 1D). The 6-month follow-up en face OCT demonstrated loss of these hyperreflective dots and subsequent enlargement of the AS. On the OCT B-scan, marked RPE-BM complex atrophy developed within AS (Figure 2, A and B). The OCT B-scan in the superior macula depicted varying maturity of AS, including flat elevation of the RPE and RPE-BM atrophy (Figure 2C).

Intraretinal hyperreflective foci in AS have been described using the OCT B-scan and are supposed to

Fig. 2. Baseline (A) and follow-up (B) en face OCT images showing loss of hyperreflective dots. The red arrows in the magnified views (yellow dashed squares) highlighted the horizontal enlargement of AS. A 20-mm-thick ellipsoid zone-based contour positioned at the depth of the RPE-BM complex (dashed line) was used to obtain en face OCT images. The red arrowheads delineated the RPE-BM complex atrophy. C. The OCT B-scan in the superior macula depicted varying maturity of AS, including flat elevation of the RPE and RPE-BM atrophy (arrowheads). The green line indicates the location of the OCT B-scan.



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be derived from the RPE cells undergoing trans-differentiation.⁴ In this article, we reported a novel en face OCT feature at the level of the RPE-BM complex within AS termed “hyperreflective dots.”

Key words: angioid streaks, optical coherence tomography, pseudoxanthoma elasticum, retinal pigment epithelium.

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References

1. Chatziralli I, Saitakis G, Dimitriou E, et al. Angioid streaks: a comprehensive review from pathophysiology to treatment. *Retina* 2019;39:1–11.
2. Corbelli E, Carnevali A, Marchese A, et al. Optical coherence tomography angiography features OF angioid streaks. *Retina* 2018;38:2128–2136.
3. Hanhart J, Greifner H, Rozenman Y. Locating and characterizing angioid streaks with en face optical coherence tomography: *Retin Cases Brief Rep* 2017;11:203–206.

Romano F, Mercuri S, Arrigo A, et al. Identification of hyper-reflective foci in angioid streaks. *Eye* 2019;33:1916–1923.

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