OPEN

Photo Essay

Edited by Dilraj Grewal with Frank L. Brodie and Henry Feng

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

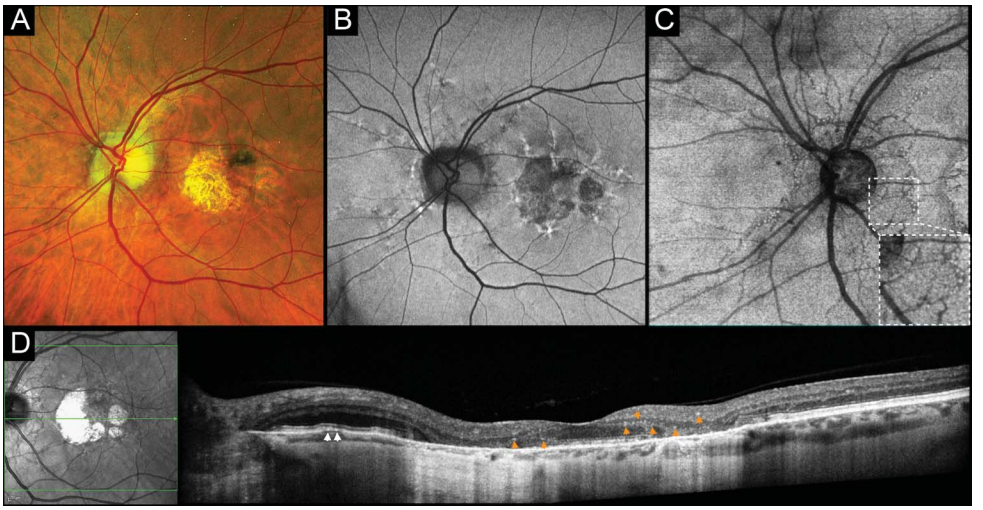

Fig. 1. A. Fundus photography showed peripapillary AS and macular geographic atrophy. **B.** Fundus autofluorescence revealed a butterfly-shaped pattern dystrophy, and AS appeared as speckled hyperautofluorescent lines. **C.** C. En face OCT at the level of the RPE-BM complex enhanced the visualization 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]

From the Centre Hospitalier Universitaire de l'Hôpital Nord, chemin des Bourrely, Marseille, France.

None of the authors has any financial/conflicting interests to disclose.

Reprint requests: Prithvi Ramtohul, MD, Centre Hospitalier Universitaire de l'Hôpital Nord, chemin des Bourrely, 13015 Marseille, France; e-mail: pramtohul@me.com

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

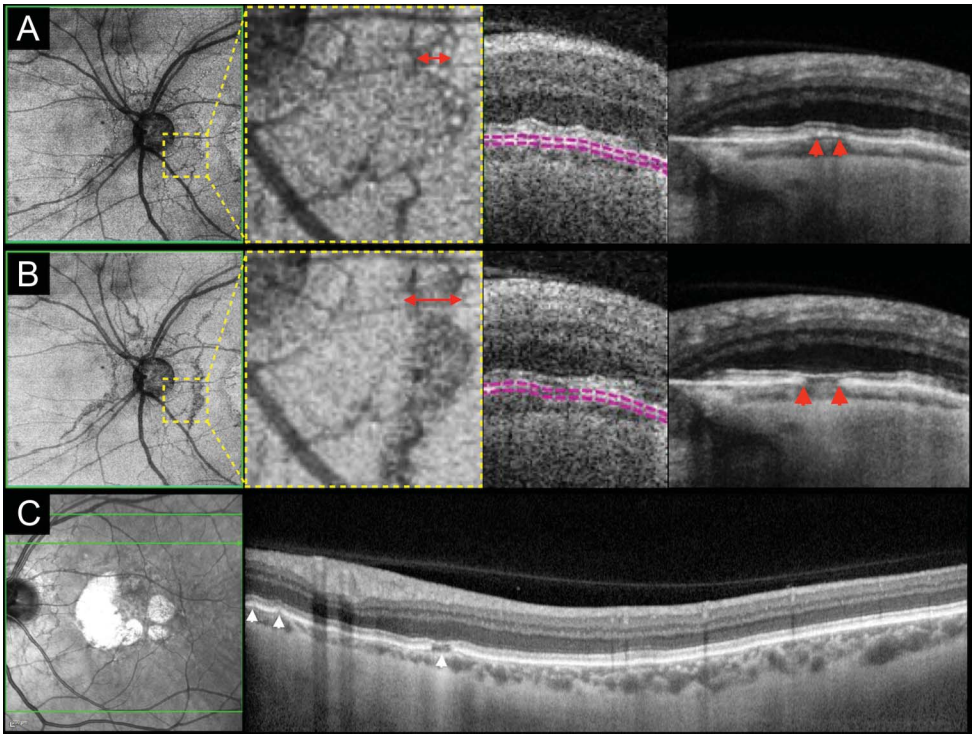

Fig. 2. Baseline (**A**) and follow-up (**B**) en face OCT images showing loss of hyperreflective dots. The red arrows in the magnified views (yellow dashed squares) highlighted the horizontal enlargement of AS. A 20-mm-thick ellipsoid zone-based contour positioned at the depth of the RPE-BM complex (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.

be derived from the RPE cells undergoing transdifferentiation.[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.

PRITHVI RAMTOHUL, MD
ALBAN COMET, MD
DANIÈLE DENIS, MD, PHD

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

RETINA® is now accepting manuscripts for consideration for publication in the Photo Essay section. For a manuscript to be considered for publication within this section, the significance of the manuscript should revolve around the photographs. The photographs should convey an important or unique clinical diagnosis, condition, or treatment. The photographs can be a combination of kodachromes, angiograms, histologic sections, or ancillary diagnostic studies (e.g., echograms, radiograms, CT or MRI studies, arteriograms), all of which are imperative in the evaluation, diagnosis, and/or treatment of the condition that is represented. Overall, the Photo Essay manuscript will be limited to 300 words, five photographs, and five references. All figures submitted in color will be published in color at the expense of the authors. Please refer to the Author Instructions for all other general requirements of manuscripts submitted to **RETINA**®.