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

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. On the optical coherence tomography (OCT) B-scan, AS appear as speckled hyperautofluorescent lines. 

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.

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 co-localized 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
be derived from the RPE cells undergoing transdifferentiation. 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.

**References**

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