Frequency, phenotypic characteristics and progression of atrophy associated with a diseased Bruch membrane in pseudoxanthoma elasticum

Supplementary material

Martin Gliem¹,², Philipp L. Müller¹,², Johannes Birtel¹,², Doris Hendig³, Frank G. Holz¹,², Peter Charbel Issa¹,²

¹ Department of Ophthalmology, University of Bonn, Bonn, Germany
² Center for Rare Diseases Bonn (ZSEB), University Hospital of Bonn, Bonn, Germany
³ Institute for Laboratory and Transfusion Medicine, Heart and Diabetes Center North Rhine-Westphalia, University Hospital of the Ruhr University of Bochum, Bad Oeynhausen, Germany
Supplementary Figure 1: Subfoveal Choroidal Thickness in Pseudoxanthoma Elasticum Patients before the Onset of Atrophy or with Early Atrophy

The grey line represents the mean of 54 healthy controls without any eye disease and the grey area the respective 95% confidence interval.
Supplementary Figure 2: Foveal Sparing of Atrophy in Pseudoxanthoma Elasticum

Fundus color (A, D, G), fundus autofluorescence (AF) (B, E, H) and near-infrared (NIR) reflectance (C, F, I) images of representative PXE patients shown in figure 4 illustrating foveal sparing of the atrophy. Foveal sparing is best visible in NIR reflectance images (C, F, I) as visibility is obscured by macular pigment in fundus AF images (B, E, H).
Supplementary Figure 3: Variability of Disease Severity in Older Patients with Pseudoxanthoma Elasticum

Fundus color (A, C) and fundus autofluorescence (B, D) images showing the variability of disease severity in Pseudoxanthoma elasticum (PXE). Besides the typical PXE features like peau d’orange and angioid streaks a 79 years old male patient presented only some peripapillary atrophy and pattern dystrophy-like changes (A, B). In contrast, a 62 years old male patient showed widespread atrophy and central neovascular activity (C, D).