

Clinical Image

Pseudoxanthoma Elasticum: Peau D'orange and Angioid Streaks

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A 43-year-old lady with pseudoxanthoma elasticum (PXE) presented for fundus exam. Her parents noticed rash-like lesions on the side of her neck when she was 9-years-old. She was diagnosed with PXE by skin biopsy. Genetic analysis revealed autosomal recessive mutation in the *ABCC6* gene. She was followed yearly as a child, and diagnosed with angioid streaks as a teenager. Physical exam showed small, yellowish papular lesions and cutaneous laxity mainly affecting

**Figure 1:**

the neck (A). Color fundus photography showed “peau d’orange” appearance involving Bruch’s membrane just posterior to the retina in both eyes (B and C) (Figure 1). Angioid streaks, linear breaks in Bruch’s membrane seen better on fundus auto fluorescence, radiated out from the optic nerve without involving the fovea (D and E). She has been followed for over 30 years without need for treatment. Skin lesions progressed slowly. Vision remains unaffected, but she continues to be followed for possible secondary neovascularization.