



DERMATOPATHOLOGY

PAPULAR DERMAL FIBRO-ELASTOPATHY: A UNIFYING CONCEPT OF PSEUDOXANTHOMA ELASTICUM-LIKE DISORDERS

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Background: Pseudo xanthoma elasticum-like diseases are rare acquired elastic tissue disorders characterized clinically by yellowish-to-white papules most commonly occurring on the posterior neck, axillae and antecubital fossae of otherwise healthy elderly patients. The term fibroelastolytic papulosis has been proposed to encompass similar conditions previously reported as pseudoxanthoma elasticum-like papillary dermal elastosis (PXE-like PDE) and white fibrous papulosis of the neck (WFPN). Papillary dermal elastosis (PDE) has been described as an entity with similar clinical appearance and the histopathologic finding of foci of clumped elastic tissue alternating with foci of decreased elastic fibers in the papillary dermis. The term Late-onset focal dermal elastosis (LOFDE) has been used to describe a PXE-like eruption characterized by elastotic changes within the reticular dermis. We hereby present a case of a patient with a unique clinico-pathological picture, suggesting the theory that all these entities exist as variants along the same spectrum.

Observations: a 49 years old otherwise healthy female presented with an asymptomatic, generalized eruption of small white-to-yellow papules with a cobblestone appearance. The histology showed subtle elastolytic changes in the papillary dermis and thickened, hyperplastic collagen bundles with marked elastotic changes in the mid and deep reticular dermis. These findings are consistent with a clinico-pathological diagnosis of all the abovementioned elastopathic disorders.

Key message: This case is of special interest as it supports the need of a unifying concept of PXE-like disorders. We suggest the term "Papular dermal PXE-like fibro-elastopathy", in order to highlight the similar clinico-pathological presentation beyond the nature of collagen and elastic fiber changes and the level of the dermis in which they take place. Both dermatologists and pathologists should be familiar with these entities that closely resemble pseudoxanthoma elasticum, but unlike the latter are skin-limited conditions, and cause only aesthetic discomfort.

