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A rare case report: Pseudoxanthoma elasticum-like papillary dermal elastolysis

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Introduction & Objectives:

Pseudoxanthoma elasticum-like papillary dermal elastolysis (PXE-PDE) is a rare, acquired elastic tissue disorder characterized by asymptomatic, yellowish, non-follicular papules, typically located on the posterior neck. It most commonly affects postmenopausal women over the age of 40. Despite its clinical similarity to pseudoxanthoma elasticum (PXE), PXE-PDE lacks systemic involvement and is distinguished by its specific histopathological findings. The exact pathogenesis remains unclear, but ultraviolet exposure, aging, and abnormal elastogenesis have been suggested as contributing factors. In this case report, we aim to present a 73-year-old woman with clinical and histological features consistent with PXE-PDE, contributing to the limited number of documented cases in the literature.

Materials & Methods:

A 73-year-old woman presented with a 3–4-year history of asymptomatic papules on the posterior neck, gradually extending laterally. Her medical history included type 2 diabetes mellitus and hypertension, managed with metformin, losartan/hydrochlorothiazide, and amlodipine. There was no family history of similar lesions. Physical examination revealed multiple yellowish 1–2 mm non-follicular papules on the posterior and lateral aspects of the neck. A punch biopsy was performed. Histopathological analysis included H&E, Elastic Van Gieson, Masson's Trichrome, and Alcian Blue staining. Routine biochemical and hematological tests were also conducted. Treatment with three sessions of intralesional triamcinolone acetonide (10 mg/mL) was initiated.

Results:

Histological findings revealed focal reduction and fragmentation of elastic fibers (Elastic Van Gieson), melanin incontinence and mild edema in the papillary dermis (H&E), and no thickening of collagen bundles (Masson's Trichrome). Alcian Blue staining was negative. Laboratory results were within normal limits. Based on clinical and histopathological features, the diagnosis of PXE-PDE was confirmed. No significant response was observed after corticosteroid injections.

Conclusion:

PXE-PDE is a rare and under-recognized condition that requires clinical suspicion and histopathological confirmation. It should be differentiated from PXE to avoid unnecessary systemic investigations. Current treatment options remain limited, and patient response is generally poor. This case aims to enhance awareness of PXE-PDE and its diagnostic features among dermatologists.