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CASE REPORT

Pseudoxanthoma Elasticum-Like Papillary Dermal Elastolysis (PXE-PDE) in an Elderly Female: A Rare Diagnostic Entity

Naif Alshehri[1], Reem Alsergani[1], Abdulrahman Alqerafi[1,2], Sadan Mohammed al Harbi[3], Ahmed Alhumidi[4], Mohammed A Alfada[1]

[1]Department of Dermatology, College of Medicine, King Saud University, Riyadh, Saudi Arabia; [2]Dermatology and Dermatological Surgery Department, Prince Sultan Military Medical City, Riyadh, Saudi Arabia; [3]Department of Dermatology, College of Medicine, Imam Abdulrahman Bin Faisal University, Dammam, Eastern Province, Saudi Arabia; [4]Department of Pathology, College of Medicine, King Saud University, Riyadh, Saudi Arabia

Correspondence: Naif Alshehri, Department of Dermatology, College of Medicine, King Saud University, Riyadh, 11461, Saudi Arabia, Email n.alshehri90@gmail.com

Abstract: Pseudoxanthoma elasticum-like papillary dermal elastolysis (PXE-PDE) is a rare, acquired skin disorder characterized by multiple small papules arising from degeneration of elastic fibers in the dermis. It is often underrecognized due to its benign nature and resemblance to harmless age-related skin changes and may be misdiagnosed as conditions such as pseudoxanthoma elasticum (PXE). A 70-year-old woman presented with asymptomatic skin lesions on her neck, which had gradually increased over 2–3 years. On physical examination, multiple confluent yellowish-white papules with a cobblestone appearance were observed on the neck. Skin biopsy confirmed the diagnosis of PXE-PDE, showing loss of elastic tissue in the superficial dermis. The patient underwent two sessions of CO_2 laser treatment, with approximately 40% improvement noted by both the physician and the patient. This case underscores the importance of clinicopathologic correlation in diagnosing PXE-PDE and distinguishing it from PXE and other mimickers. CO_2 laser therapy may be considered as a cosmetic treatment option in selected patients.

Keywords: pseudoxanthoma elasticum-like papillary dermal elastolysis, PXE-PDE, fibroelastolytic papulosis, white fibrous papulosis, neck papules, CO_2 laser therapy

Introduction

Pseudoxanthoma elasticum-like papillary dermal elastolysis (PXE-PDE) is a rare, acquired skin disorder characterized by multiple small papules arising from degeneration of elastic fibers in the dermis.[1] Published case reports of PXE-PDE remain few, with fewer than 60 documented globally.[2] Clinically, it typically presents in older adults with asymptomatic or mildly itchy 2–5 mm whitish to yellow papules that may coalesce into cobblestone-patterned plaques on the neck and upper trunk.[1] While benign and primarily cosmetic in nature, its significance lies in mimicking other conditions and the potential for misdiagnosis. PXE-PDE has been documented in both men and women, it is uncommon, and its pathogenesis remains unclear but is thought to be related to intrinsic aging of the dermal connective tissue.[1]

Correctly diagnosing PXE-PDE is important for patient counseling and management. Clinically, the condition is often underrecognized due to its benign nature and resemblance to harmless age-related skin changes. Patients may live with these papules for years without diagnosis, or they may be misdiagnosed with a more serious condition like PXE. Distinguishing PXE-PDE from PXE is particularly vital – PXE carries risks of systemic disease, whereas PXE-PDE does not.[3] Mislabeling a patient with PXE could lead to unnecessary anxiety and investigations. Conversely, confusing PXE-PDE with xanthomas could prompt unwarranted evaluation for metabolic disorders. Therefore, a high index of suspicion and careful clinical evaluation are needed when encountering multiple whitish or yellow papules in an adult patient.

Histopathological examination is often required to confirm PXE-PDE, as the clinical appearance alone may be inconclusive. Skin biopsy typically reveals an absence of elastic fibers in the papillary dermis, sometimes accompanied

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by increased and thickened collagen.[4] In some cases, multiple biopsy specimens and special stains like Verheoff Van Gieson (elastic fiber stains) are needed to detect the elastolysis and rule out other conditions.[5] Once histology confirms the diagnosis, patients can be reassured of the benign course of PXE-PDE. In the absence of standardized treatment protocols, therapeutic decisions are largely cosmetic and pose no health risk.

In light of the above, recognizing PXE-PDE requires both clinical vigilance and histological confirmation. The following case illustrates the typical presentation of PXE-PDE and underscores the diagnostic challenges involved.

Case Presentation

A 70-year-old woman with a medical history of hypothyroidism, vitamin D deficiency, dyslipidemia, and chronic insomnia presented with asymptomatic skin lesions on her neck. These lesions had first appeared approximately 2–3 years prior and gradually increased in number and extent. She denied any itching, pain, or other associated symptoms. Her medications included levothyroxine, vitamin D supplementation, atorvastatin 20 mg daily, and amitriptyline 25 mg daily for insomnia. On physical examination, multiple confluent yellowish-white papules with a cobblestone appearance were observed covering the entire neck region (Figure 1).

Our differential diagnosis includes xanthoma, steatocystoma multiplex, mucinosis, pseudoxanthoma elasticum (PXE), and Pseudoxanthoma elasticum-like papillary dermal elastolysis (PXE-PDE). A skin biopsy was performed from one of the lesions over the neck. The result of the biopsy was conclusive, showing loss of elastic tissue in superficial dermis while present in deep dermis, confirming the diagnosis of PXE-PDE (Figure 2). Our patient underwent two sessions of

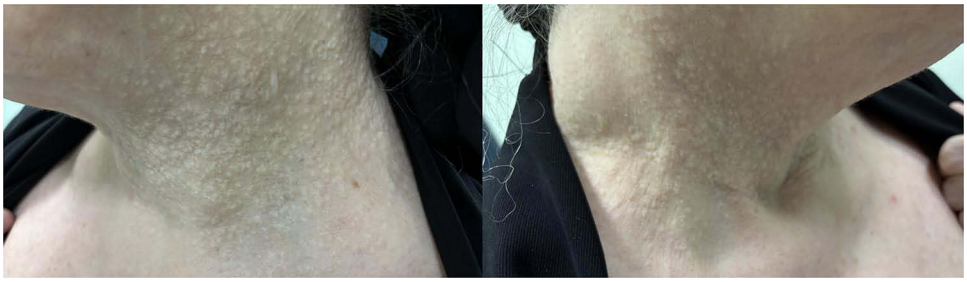

Figure 1 Clinical photograph demonstrating multiple confluent yellowish-white papules with a cobblestone pattern on the patient's neck.

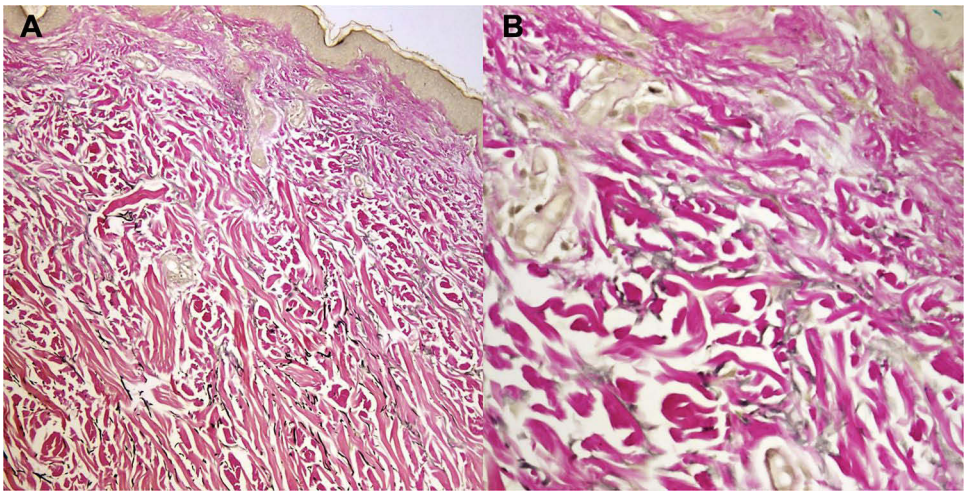

Figure 2 (**A**) Photomicrograph of high power view showing loss of elastic tissue in the superficial dermis while present in the deep dermis (Verhoeff–Van Gieson [EVG] stain, original magnification×100). (**B**) Photomicrograph of high power view of superficial/papillary dermis confirming loss of elastic tissue (Verhoeff–Van Gieson [EVG] stain, original magnification×400).

CO_2 Laser treatment, with about 40% improvement according to both physician and patient assessment. The patient is currently continuing treatment with additional sessions planned.

Discussion

In 1985, Shimizu et al was the first to report white fibrous papulosis of the neck. In their series, typical patients' presentation was asymptomatic whitish papules predominantly located around the neck.[6] Subsequently, in 1992, Rongioletti and Rebora described a similar entity as pseudoxanthoma elasticum-like papillary dermal elastolysis (PXE-PDE), highlighting its clinical resemblance to inherited pseudoxanthoma elasticum (PXE), though without associated systemic involvement.[7] Later, Balus et al (1997) recognized substantial clinical and histopathological overlaps between PXE-PDE and white fibrous papulosis of the neck (WFPN), suggesting the term "fibroelastolytic papulosis" as a disease spectrum.[8]

PXE-PDE predominantly affects middle-aged to older adults, typically presenting after the age of 50, with a clear female predominance.[9] While initially considered rare, the actual prevalence of PXE-PDE might be underestimated given the asymptomatic nature of the condition, leading to fewer diagnoses and reports in medical literature.[10] Although cases have been reported across various geographic regions, no significant ethnic predisposition has been firmly established, and the condition appears globally distributed.[8]

Atypical presentations have also been reported in the literature. Gambichler et al described a case of early-onset PXE-PDE in a young female, which deviates from the typical demographic.[11] Similarly, another study documented a younger individual affected by PXE-PDE, reinforcing that it can also present before the typical age of onset.[12] Another study by Shawa et al suggests a potential role for immune or inflammatory triggers in the fibroelastolytic papulosis spectrum (FEP). The study highlighted a wolf isotopic response where FEP onset followed a herpes zoster infection.[13] Several reports have noted involvement of less common sites such as the axillae, chest, and upper back, as well as presentations with mild pruritus, pigmentary changes, or subtle histopathological findings.[9,10,14] These variations, while uncommon, expand our clinical understanding of this condition.

PXE-PDE is characterized histologically by a band-like loss of elastic fibers in the papillary dermis, which may require special stains like Verhoeff–Van Gieson (VVG) or amyloid P immunostaining, which will show positive staining along altered elastic fibers in the papillary dermis, supporting its utility as a complementary diagnostic tool in ambiguous or subtle cases.[15] Furthermore, Ohnishi et al demonstrated that the primary defect in elastogenesis is in elastin itself, rather than in fibrillin-1, the two main components of elastic fibers.[16]

The exact pathogenesis of PXE-PDE remains unclear. Proposed contributing factors include intrinsic aging, ultraviolet (UV) radiation, hormonal influences, and inflammatory triggers.[2] Since the condition most often arises in older individuals, further supports the idea that age-related degeneration of dermal elastic fibers plays a central role. Some authors have hypothesized that UV exposure contributes to elastic fiber loss, although cases in sun-protected areas like the axilla challenge this theory. Also, a reported case in a hijab-wearing woman with minimal sun exposure, like in our case, further argues against UV damage being a major etiological trigger.[17] Furthermore, familial cases, such as the one documented by Orlandi et al, raise the possibility of a genetic or inherited predisposition. In their report, two sisters presented with similar clinical findings with histological confirmation of the diagnosis.[18] This familial pattern strengthens the theory of a hereditary component in selected cases. Altogether, current literature supports a multifactorial etiology involving a combination of intrinsic aging, environmental exposure, and possibly genetic susceptibility.

Although dermoscopy is not routinely used for diagnosing PXE-PDE, several case reports have described helpful patterns. As reported by multiple studies, the most consistent findings include multiple whitish or yellowish-white papules that coalesce with overlying linear or arboriform vessels.[17,19] These features, while not specific, may assist in distinguishing this disease from clinically similar conditions and support the decision to pursue biopsy when the diagnosis is uncertain.

There is no standardized treatment for PXE-PDE, and management is usually pursued for cosmetic reasons. Rongioletti et al reported case series in which topical retinoids were the treatment of choice in most cases and resulted in minimal improvement.[9] Moreover, Maione et al reported a clinical improvement after the treatment with topical tretinoin.[20] Another reported case described no clinical response after two courses of intralesional triamcinolone

acetonide (0.1 mL of 5 mg/mL), suggesting limited benefit from corticosteroid injections.[3] Moreover, Foering et al described the use of non-ablative fractional laser, which led to both clinical and histological improvement after a total of three sessions.[21] Furthermore, CO_2 ablative laser has been reported as a treatment option. Chong et al demonstrated a marked improvement in papular elevation and skin texture in their patient after multiple CO_2 laser sessions.[22] In another case, a 63-year-old man experienced complete resolution of pruritus and lesion flattening after three treatments, with no recurrence at six-month follow-up.[23] Our case adds to the growing evidence by demonstrating clinical improvement following CO_2 laser therapy, further supporting its potential role in the management.

Conclusion

PXE-PDE is a benign, often overlooked condition that can mimic other disorders, making the diagnosis unclear without histology. Our case adds to the understanding of its clinical spectrum and supports CO_2 laser as a potential cosmetic treatment option. Further case series and longitudinal follow-up studies are needed to better characterize treatment outcomes and inform evidence-based management. The importance of clinicopathologic correlation and individualized care becomes clearer as more cases emerge.

Data Sharing Statement

All data generated or analyzed during this study are included in this article. Further enquiries can be directed to the corresponding author.

Statement of Ethics

This case report does not require institutional review board approval in accordance with King Saud University or national guidelines.

Declaration of Patient Consent

Written informed consent was obtained from the patient for publication of the details of their medical case and any accompanying images.

Disclosure

The authors declare that there are no conflicts of interest.

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