Letter to the Editor
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Pseudoxanthoma elasticum-like papillary dermal elastolysis: A rare case report with dermoscopic features

Nadir görülen psödoksantoma elastikum-benzeri papiller dermal elastolizis olgusu ve dermoskopik bulguları

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To Editor,

Pseudoxanthoma elasticum-like papillary dermal elastolysis (PXE-PDE) is a rare acquired elastolytic disorder characterized by elastolysis in the papillary dermis, clinically resembling hereditary PXE but without systemic involvement[1]. Clinical diagnosis of PXE-PDE is often challenging, and histopathological evaluation plays a critical role in differential diagnosis. Due to its clinical and histopathological similarities with PXE, which tends to present at an earlier age, skin biopsy remains an essential diagnostic tool for accurate identification. However, the use of non-invasive imaging techniques that can enhance diagnostic accuracy, alongside invasive procedures such as skin biopsy, may also be beneficial. In recent years, clinical practice has increasingly defined the dermoscopic features of many dermatological conditions. Nevertheless, the contribution of dermoscopic examination to the diagnosis of elastic fiber disorders, especially rare entities such as PXE-PDE, has not been adequately described in the literature. Therefore, we aimed to describe the clinicopathological and dermoscopic features of this rare disease.

A 71-year-old female patient presented with asymptomatic, white-to-skin-colored papules symmetrically distributed on the neck for several years (Figure 1). The lesions reportedly did not progress over time. The patient's medical history was unremarkable, except for hypothyroidism, osteoporosis, and a history of prolonged occupational sun exposure. Dermoscopy revealed multiple yellowish-white, non-follicular papules accompanied by fine, linear vessels (Figure 2). A punch biopsy revealed orthokeratosis in the epidermis, pigment incontinence in the dermis, and superficial perivascular lymphocytic inflammation. Verhoeff-Van Gieson staining confirmed a marked loss of elastic fibers in the papillary dermis (Figure 3). The absence of calcification in the elastic fibers helped distinguish this condition from hereditary PXE. Based on these findings, the patient was diagnosed with PXE-PDE.

PXE-PDE is a rare elastic fiber disorder with an unknown pathogenesis. Factors such as aging, impaired elastogenesis, and chronic ultraviolet exposure have been suggested as possible contributors to its pathogenesis. Owing to their clinical and histopathological similarities, distinguishing PXE-

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Figure 1. Multiple skin-colored to white papules with a cobblestone-like pattern were symmetrically distributed on the neck, forming plaques through coalescence

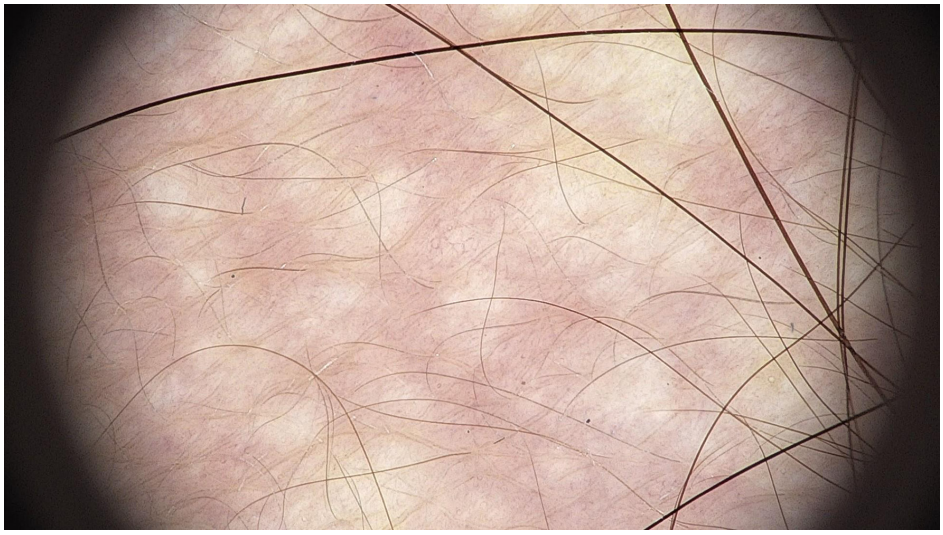

Figure 2. Pale, white, non-follicular papules forming plaques with overlying reticular and linear vessels were observed on dermoscopic examination

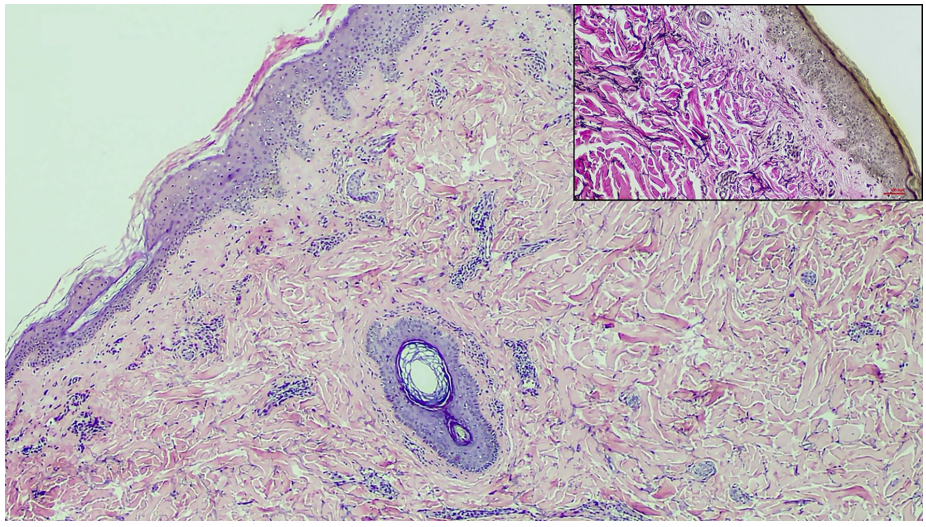

Figure 3. Hematoxylin and eosin, ×40; inset: Verhoeff-Van Gieson, ×100.Verhoeff-Van Gieson staining, at a magnification of approximately 100 in the inset, highlights the marked reduction and fragmentation of elastic fibers in the dermis

PDE from PXE can be challenging. The primary differences between PXE-PDE and PXE are the absence of systemic organ involvement and the lack of elastic fiber calcification on histopathology. Moreover, while PXE typically presents during childhood or adolescence, PXE-PDE more commonly affects elderly individuals and has a female predominance (Table 1). In our postmenopausal female patient, no ocular or cardiac abnormalities were detected during the systemic evaluation. In the differential diagnosis, other conditions such as white fibrous papulosis of the neck, mid-dermal elastolysis, and papillary dermal elastosis should be considered[2]. Dermoscopy may serve as a valuable non-invasive diagnostic tool that can help differentiate elastolytic disorders. Identifying dermoscopic features specific to PXE-PDE could support clinical diagnosis and potentially reduce the need for unnecessary invasive procedures. In this case, the marked loss of elastic fibers in the papillary dermis, along with the absence of fiber fragmentation and calcification, supported the diagnosis of PXE-PDE. Furthermore, dermoscopic examination revealed yellowish-white homogeneous areas corresponding to non-follicular papules, along with fine linear vessels. A few previous case reports have highlighted these linear vessels as potentially characteristic dermoscopic features of PXE-PDE. For instance, Ito et al.[3] described confluent, skin-colored, non-follicular papules forming plaques with prominent linear vessels in a patient with PXE-PDE. Similarly, another case report noted multiple normochromic papules and linear vessels in a PXE-PDE patient[4]. A more comprehensive characterization of dermoscopic findings through case reports in the literature may reduce the need for biopsy and expedite the diagnostic process. Although no definitive treatment exists, topical retinoids and fractional laser therapy have shown clinical improvement in some cases[5].

This case was presented to contribute to the differential diagnosis of PXE-PDE through the identification of its dermoscopic features. Dermoscopy may accelerate the diagnostic process and reduce the need for biopsy, thereby supporting the clinical management of PXE-PDE. In particular, the presence of multiple skin-colored non-follicular papules and linear vessels are dermoscopic features that may support the diagnosis of PXE-PDE. In the future, further case reports are needed to standardize dermoscopic evaluations and optimize the diagnostic processes for PXE-PDE.

Table 1. Comparison of PXE and PXE-PDE

Feature	PXE	PXE-PDE
Age of onset	Early adulthood	Postmenopausal
Systemic involvement	Cardiovascular, ophthalmologic, gastrointestinal	None
Histopathology	Basophilic, calcified elastic fibers in mid and deep dermis	Loss of elastic fibers in papillary dermis
Calcification	Present	Absent
Elastin fiber arrangement	Clumped and fragmented	Marked reduction in papillary dermis

PXE: Pseudoxanthoma elasticum, PXE-PDE: Pseudoxanthoma elasticum-like papillary dermal elastolysis

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Ethics

Informed Consent: Informed consent and permission notice was obtained from the patient to publish the person's photographs.

Footnotes

Authorship Contributions

Surgical and Medical Practices: N.S., S.D., Consept: N.S., Design: N.S., E.B.A., Data Collection or Processing: N.S., E.B.A., S.D., Analysis or Interpretation: N.S., E.B.A., S.D., Literature Search: N.S., E.B.A.,Writing: N.S., E.B.A.

Conflicts of Interest: The authors declare no conflict of interest.

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