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CLINICAL VIGNETTE

A Case Report of Pseudoxanthoma Elasticum-like Papillary Dermal Elastolysis

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Abstract

Pseudoxanthoma elasticum-like papillary dermal elastolysis (PXE-PDE) is a rare acquired fibroelastolytic disorder characterized by soft, white-yellow papules located primarily on the neck, supraclavicular region, and axillae. Affected patients are almost exclusively elderly women. The pathogenesis of PXE-PDE has been speculated to involve various factors including ultraviolet radiation, aging, and abnormal elastogenesis. Here we present a rare case of PXE-PDE in a 73-year-old woman and review the differential diagnosis, pathophysiology, and current treatment options of this disease.

Keywords: Elastolysis; elastic tissue disorders; pseudoxanthoma elasticum-like papillary dermal elastolysis; PXE-PDE

Introduction

Pseudoxanthoma elasticum-like papillary dermal elastolysis (PXE-PDE) is a rare acquired fibroelastolytic disorder characterized by soft, white-yellow papules coalescing into "cobblestone" plaques located primarily on the lateral and posterior neck, supraclavicular region, and axillae. On histology, there is a reduction of elastic fibers in the papillary dermis.[1,2] Pseudoxanthoma elasticum-like papillary dermal elastolysis occurs almost exclusively in elderly, postmenopausal women although few cases have been reported in younger patients. Since it was first described in 1992 by Rongioletti and Rebora, only approximately 60 cases have been reported to date. Here we present a rare case of PXE-PDE in a 73-year-old woman who presented with typical clinical and pathologic findings.

Case Synopsis

A 73-year-old menopausal Caucasian woman presented with a four-year history of multiple progressive, asymptomatic papules around her neck. In the past year, the papules had spread to the superior chest, shoulders, and antecubital fossae. No other areas were involved. She had no known history of excessive sun exposure. She reports that mother had similar lesions on her back.

Her medications included rosuvastatin 20 milligrams daily, vitamin D 2000 units daily, and omega-3 fish oil 1200 milligrams daily. She had a past medical history of hypercholesterolemia, bilateral cataracts, and osteoporosis. She was otherwise healthy without any cardiovascular, bleeding diasthesis, or gastrointestinal problems. An echocardiogram performed two years ago and ophthalmology exam performed several months ago were normal. She had no history of systemic steroid exposure or high-dose steroid injections.

Physical exam revealed yellowish-to-skin colored papules coalescing to form pebbly plaques on the bilateral neck (Figure 1A) extending to the anterior neck, upper chest, bilateral trapezii, and bilateral antecubital fossae (Figure 1B). A skin biopsy revealed a decrease in elastic fibers in the papillary dermis supported by a Verhoeff-van Gieson stain (Figure 2).

Based on the clinical exam and histologic findings, a diagnosis of PXE-PDE was made.

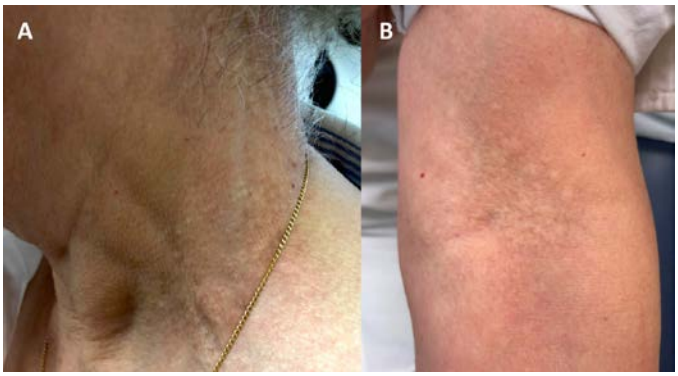

Figure 1: Clinical photographs of (A) yellowish-to-skin colored papules coalescing to form pebbly plaques on the lateral neck. (B) Similar appearing lesions were present on the bilateral antecubital fossa.

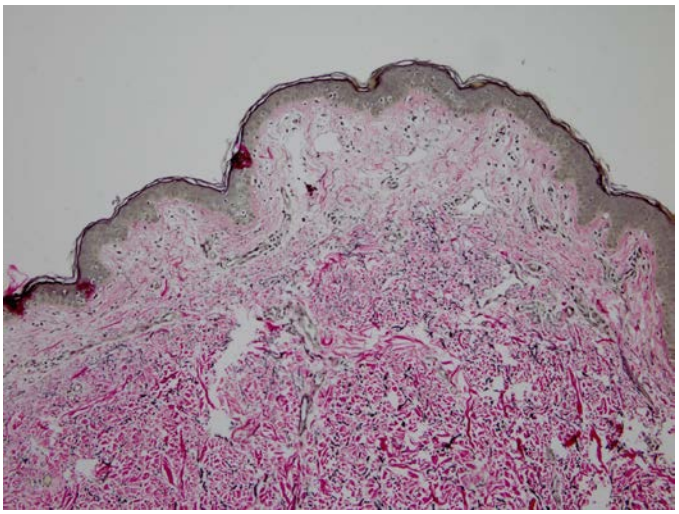

Figure 2: Verhoeff-van Gieson stain of punch biopsy specimen from neck revealed decreased elastic fibers in the papillary dermis consistent with pseudoxanthoma elasticum-like papillary dermal elastolysis (PXE-PDE).

Case Discussion

Acquired disorders of elastic tissue can be divided into those involving an increase (elastosis) or decrease (elastolysis) of elastic tissue in the dermis. The diseases of elastolysis include PXE-PDE, nevus anelasticus, papular elastorrhexis, perifollicular elastolysis, anetoderma, acquired cutis laxa, post-inflammatory elastolysis and cutis laxa, white fibrous papulosis of neck (WFPN), mid- and upper dermal elastolysis, granulomatous slack skin, and acrokeratoelastoidosis.[3] Various fibroelastolytic diseases of the skin share some common clinical features, so histopathologic analysis may be necessary for definitive diagnosis. Special stains for elastic tissue such as the Verhoeff-van Gieson stain are helpful given that the routine hematoxylin and eosin stain lacks the specificity required for diagnosis.

In particular, PXE-PDE and WFPN can both present as asymptomatic white-to-yellow papules on the neck. Clinically, WFPN is distinguished by the lack of confluent lesions, paler appearance of lesions, and occurrence predominantly in elderly men.[3] The histologic hallmark of PXE-PDE is marked reduction or absence of oxytalan and elaunin fibers in the papillary dermis. There is no increase in fibers of the reticular dermis and dermal fibrosis is not seen, distinguishing PXE-PDE from WFPN.[2,3] However, it has been proposed that PXE-PDE and WFPN exist on the same spectrum of disease, encompassed by the term fibroelastolytic papulosis of the neck (FEPN), based on several cases with similar clinical and histologic features.[4-6]

The pathophysiology of PXE-PDE is speculated to be due to a combination of factors including ultraviolet radiation, aging, and abnormal elastogenesis.[2] Ultraviolet radiation has been implicated given the frequent involvement of the posterior neck, upper back, and supraclavicular areas.[2,3] A suggested mechanism is ultraviolet-induced activation of matrix metalloproteinases in keratinocytes and fibroblasts that degrade collagen.[2,7] However, ultraviolet light is unlikely to be an independent factor as many patients do not report a history of excessive sun exposure and lesions can present in sun-protected skin such as the axilla.[8-12] Additionally, only one case of PXE-PDE has been reported with solar elastosis and actinic damage.[13]

Intrinsic aging is thought to be contributory given that affected patients are typically elderly with an average age of 61.8 years.[2] Aging results in a decrease of oxytalan fibers in the papillary dermis, similar to that seen in PXE-PDE.[14] However, several cases of PXE-PDE have been reported in younger females, the two youngest at 28 years of age, suggesting that aging may play a smaller role than previously thought.[8,15,16] This is supported by an immunohistochemical study demonstrating loss of both elastin and fibrillin-1 in PXE-PDE in contrast to loss of fibrillin-1 alone in normal aged skin.[17]

To date, all 52 reported cases of PXE-PDE have been in women. The reason for this sexual bias is unknown, but may indicate the presence of some unidentified genetic, hereditary, or hormonal factor. Our patient reported her mother had similar lesions on her back. While this assertion cannot be confirmed, a familial case of PXE-PDE in two sisters has been reported, providing further evidence for genetic factor.[18] Alternatively, some have suggested that the predisposition for one sex is due to women more frequently seeking medical care for cosmetic concerns than men, rather than related to the pathophysiology of the disease.[17,19] Given that the disease is asymptomatic and has subtle manifestations, it is likely that the true incidence of PXE-PDE is underreported.

Steroid therapy is currently the only drug exposure implicated in PXE-PDE. A case series of four patients all had been exposed to long-term systemic methylprednisolone or intralesional triamcinolone for rheumatologic conditions during which they developed the lesions of PXE-PDE.[20] The proposed mechanism involves decreased expression of the elastin gene and elastin messenger ribonucleic acid as demonstrated in cultured human skin fibroblasts.[21]

At present, there are no effective treatments for PXE-PDE. Tretinoin has been shown to decrease the activity of matrix metalloproteinases and collagenase and improve the appearance of photoaged skin.[7] However, the reported efficacy of topical retinoids in PXE-PDE is minimal.[2] Despite the implication of steroids in the pathogenesis of PXE-PDE, low-dose intralesional steroid injections have been attempted given their reported efficacy in papular elastorrhexis, another elastolytic disorder.[20,22] Unfortunately, no clinical improvement was noted.[19] There are several case reports of successful treatment with non-ablative fractional resurfacing (NAFR) and fractionated carbon dioxide lasers.[16,23,24] However, additional studies are required to ensure the results are replicable. As such, patients with PXE-PDE should be counseled on the absence of definitive treatment.

Conclusion

Pseudoxanthoma elasticum-like papillary dermal elastolysis is an underreported cutaneous fibroelastolytic disorder that occurs exclusively in women. Multiple pathogenic mechanisms have been proposed, including aging, ultraviolet light, abnormal elastogenesis, and genetic or hereditary factors although further studies are required to elucidate the details. Treatment is cosmetic with no effective therapies to date.

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