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Fibroelastolytic papulosis: two cases of disease spectrum variants

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Abstract

Fibroelastolytic papulosis (FEP) is an acquired cutaneous disorder of elastin that encompasses both white fibrous papulosis of the neck (WFPN) and pseudoxanthoma elasticum (PXE)-like papillary dermal elastolysis (PDE). Although FEP is a benign acquired disorder, it shares overlapping clinical features with pseudoxanthoma elasticum (PXE), a genetic disorder with systemic manifestations. We report two cases of FEP, including the WFPN and PXE-like PDE variants, in elderly women. In one case, a woman in her 70s with hyperlipidemia and chronic kidney disease presented with white-to-yellow, smooth, monomorphic papules coalescing into plaques on the posterior neck and bilateral antecubital fossa. A punch biopsy and elastic stain revealed a loss of elastic fibers in the papillary dermis. Based on these findings, we diagnosed our patient with PXE-like PDE. In another case, a woman in her 60s with no significant medical history similarly presented with skin-colored, smooth monomorphic papules on the posterior and anterior neck. A punch biopsy was also performed and an elastic stain showed a loss of elastic fibers in the papillary dermis, confirming the diagnosis of WFPN. Although rare, FEP is an important entity for dermatologists to recognize and differentiate from PXE given the potential for systemic complications in PXE.

Keywords: dermatopathology, elasticum, elastin, fibroelastolytic, fibrous, genetics, neck, papulosis, pseudoxanthoma, white

Introduction

Fibroelastolytic papulosis (FEP) is characterized by two disease spectrum variants, such as white fibrous papulosis of the neck (WFPN) and pseudoxanthoma elasticum (PXE)-like papillary dermal elastolysis (PDE), [1,2]. Fibroelastolytic papulosis clinically presents as skin-colored or white-to-yellow papules coalescing into plaques along the neck and/or axillary folds or antecubital fossa. Fibroelastolytic papulosis is a benign acquired disorder, but shares overlapping clinical features with pseudoxanthoma elasticum (PXE), a genetic disorder with systemic manifestations [2-4]. Therefore, distinguishing PXE from FEP is of paramount importance. Herein, we report two cases of FEP, including the WFPN and PXE-like PDE variants, in elderly women.

Case Synopsis

A woman in her 70s with hyperlipidemia and chronic kidney disease presented to the dermatology clinic with whitish papules on the posterior neck and arms for many years. On cutaneous examination, white-to-yellow, smooth, monomorphic papules coalescing into plaques were present on the posterior neck and bilateral antecubital fossa (**Figure 1**). Total body skin examination failed to reveal any other similar lesions. She was otherwise asymptomatic, and a thorough review of symptoms was negative. We performed a punch biopsy of one of the papules along the antecubital fossa (**Figure 2A**). A Verhoeff-van Gieson elastic stain revealed a loss of elastic fibers in the papillary dermis (**Figure 2B**). Based on the clinical and histopathologic findings, we diagnosed our patient with PXE-like PDE.

A woman in her 60s with no significant medical history presented to the dermatology clinic with whitish papules on the posterior neck for approximately one year. She reported that the

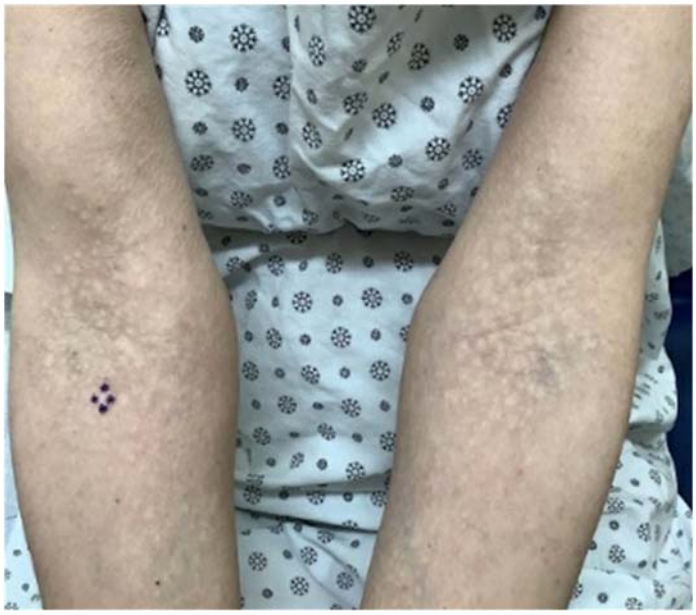

Figure 1. *Clinical image of white-to-yellow, monomorphic papules coalescing into plaques on the bilateral antecubital fossa.*

papules increased in number over the past several months and have spread to the anterior neck. The lesions were mildly pruritic, but she was otherwise asymptomatic. On cutaneous examination, skin-colored, smooth monomorphic papules were present on the posterior and anterior neck (**Figure 3**). A punch biopsy was performed (**Figure 4A**) and an elastic stain showed loss of elastic fibers in the papillary dermis, confirming the diagnosis of WFPN (**Figure 4B**).

Case Discussion

Few case reports of FEP, including both PXE-like PDE and WFPN, have been reported in the literature [1-3,5-7]. Most cases have been reported in patients over 40 years of age [1]. The lesions typically present gradually over months to years and are often

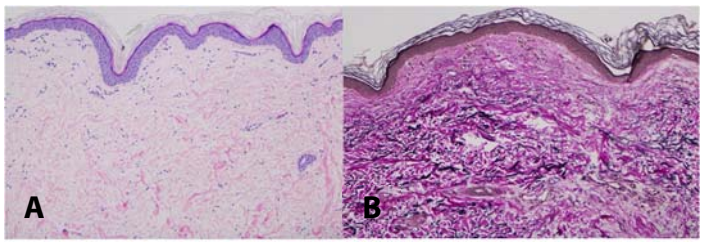

Figure 2. ***A)*** *Histopathology of the punch biopsy performed showing thickened collagen bundles in the papillary dermis. H&E, 200×.* ***B)*** *Verhoeff-van Gieson elastic stain was applied and revealed a loss of elastic fibers in the papillary dermis, 400×.*

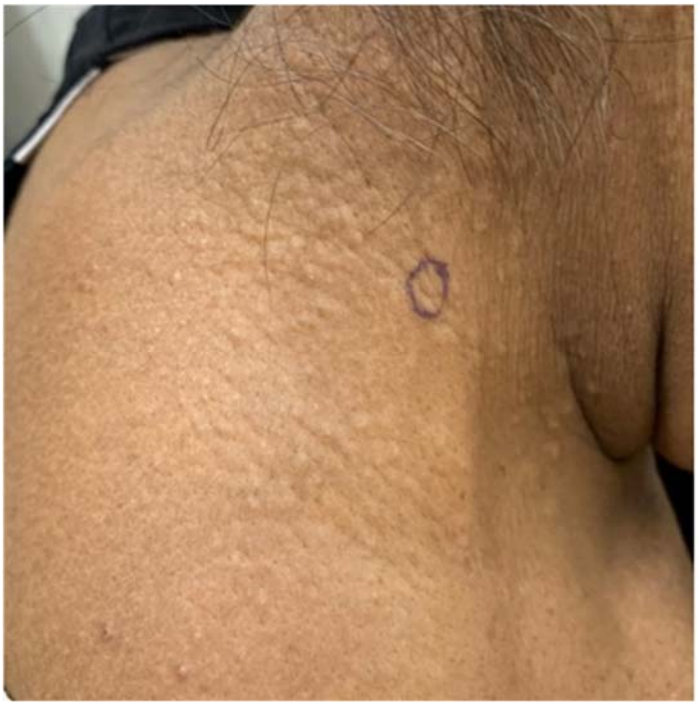

Figure 3. *Clinical image showing skin-colored papules coalescing into plaques on the posterior neck.*

asymptomatic or mildly pruritic. The etiology of FEP is unknown but is considered to be related to intrinsic skin aging [1]. White fibrous papulosis of the neck can be distinguished from PXE-like PDE by anatomic location. Although WFPN only occurs on the neck, PXE-like PDE may occur on the neck, supraclavicular area, axillary folds, antecubital fossa, scalp, and lower inguinal area [1,3,5-7].

The most important condition in the differential diagnosis for FEP is PXE, which is an autosomal recessive genetic disorder that typically presents in a younger patient population and has a female preponderance [4,8]. Biopsy is critical for differentiating FEP from PXE. On histopathology, FEP is characterized by decreased elastic fibers in the

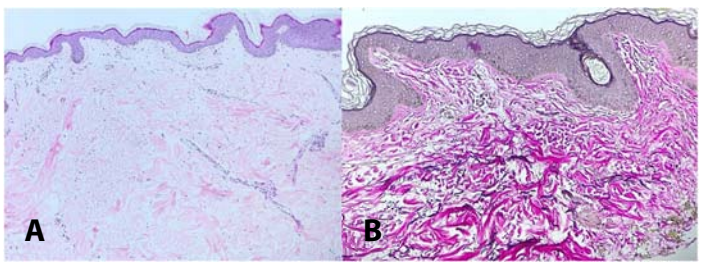

Figure 4. ***A)*** *Histopathology of the punch biopsy performed showing thickened collagen bundles in the papillary dermis. H&E, 200×.* ***B)*** *Verhoeff-van Gieson elastic stain showed loss of elastic fibers in the papillary dermis, 200×.*

Table 1. *Demographic and clinical characteristics of pseudoxanthoma elasticum (PXE) and the white fibrous papulosis of the neck (WFPN) and pseudoxanthoma elasticum (PXE)-like papillary dermal elastolysis (PDE) variants of fibroelastolytic papulosis (FEP).*

	PXE	FEP	
Variants		**WFPN**	**PXE-PDE**
Inheritance [1,4]	Autosomal recessive mutations of the *ABCC6* gene	Not inherited	Not inherited
Male/female ratio [8,9]	1:2	1:1	1:1
Age of onset [1,3,4,6,7]	Teens or young adulthood	Adulthood: all cases to date have been described in those >39 years of age	Adulthood: all cases to date have been described in those >39 years of age
Location of lesions [1,3,6,9]	May include skin folds, such as the neck, supraclavicular area, axillary folds, antecubital fossa	Neck	May include skin folds, such as the neck, supraclavicular area, axillary folds, antecubital fossa, scalp, lower inguinal region
Morphology [3,4,6,9]	White to yellow, monomorphic papules coalescing into plaques	White to yellow, monomorphic papules coalescing into plaques	White to yellow, monomorphic papules coalescing into plaques
Pathology (H&E and special stains [3,4,6,9]	Calcification and fragmentation of elastic fibers in the papillary dermis using Verhoeff-van Gieson stain for elastic fibers, or Von Kossa or alizarin red calcium stains	Thickened collagen fibers bundles on H&E and decreased elastic fibers in the papillary dermis with Verhoeff-van Gieson stain for elastic fibers	Thickened collagen fibers bundles on H&E and decreased elastic fibers in the papillary dermis with Verhoeff-van Gieson stain for elastic fibers
Associations [4,6,9]	Angioid streaks in the retina, retinal hemorrhages, gastrointestinal bleeding, arteriosclerosis, cerebrovascular accidents, myocardial infarction, blindness	No comorbidities associated	No comorbidities associated
Treatment [6-9]	No treatments available for systemic disorder	Limited efficacy of available treatments, such as tretinoin, fractionated lasers, and surgical excision	Limited efficacy of available treatments, such as tretinoin, fractionated lasers, and surgical excision

papillary dermis and thickened collagen bundles, whereas PXE is marked by calcification and fragmentation of elastic fibers [1,3,4,8]. Unlike FEP, PXE is associated with numerous comorbidities [4,6,8]. In addition to the skin, organs that contain elastic may be also affected in PXE, such as blood vessels, the retina, and gastrointestinal tract. These alterations to elastic may manifest as angioid streaks in the retina, retinal hemorrhages, gastrointestinal bleeding, and arteriosclerosis, that can increase the risk for cerebrovascular accidents and myocardial infarction [4,8]. Therefore, a diagnosis of PXE necessitates an extensive work-up to evaluate for systemic involvement, unlike fibroelastolytic papulosis. A comprehensive comparison of the demographic and clinical characteristics of PXE and the WFPN, and PXE-like PDE variants of FEP appears in **Table 1**.

Conclusion

Treatment of fibroelastolytic papulosis is not required, though patients may seek treatment to improve the appearance of the lesions and/or reduce the associated pruritis. Topical retinoids, fractionated lasers, and surgical excision have been utilized to improve the appearance of the affected skin with varying success [6,9]. Although rare, FEP is an important entity for dermatologists to recognize and differentiate from PXE given the potential for systemic complications in PXE.

Potential conflicts of interest

The authors declare no conflicts of interest.

References

1. Jagdeo J, Ng C, Ronchetti IP, et al. Fibroelastolytic papulosis. *J Am Acad Dermatol*. 2004;51:958-964. [PMID: 15583589].
2. Cohen PR, Tschen JA. Linear lumbar localized lysis of elastic fibers: a distinctive clinical presentation of mid-dermal elastolysis. *J Clin Aesthet Dermatol.* 2013;
3. 6:32-99. [PMID: 23882313].
4. Rongioletti F, Izakovic J, Romanelli P, Lanuti E, Miteva M. Pseudoxanthoma elasticum-like papillary dermal elastolysis: a large case series with clinicopathological correlation. *J Am Acad Dermatol*. 2012;67:128-135. [PMID: 22018757].
5. Luo H, Faghankhani M, Cao Y, Uitto J, Li Q. Molecular genetics and modifier genes in pseudoxanthoma elasticum, a heritable multisystem ectopic mineralization disorder. *J Invest Dermatol*. 2021;14:1148-1156. [PMID: 33341249].
6. Shimizu H, Kimura S, Harada T, Nishikawa T. White fibrous papulosis of the neck: a new clinicopathologic entity? *J Am Acad Dermatol*. 1989;20:1073-1077. [PMID: 2666457].
7. Nowicka D, Woźniak Z, Maj J, Szepietowski J. White fibrous papulosis of the neck. *Postepy Dermatol Alergol.* 2019;36:775-777. [PMID: 31998010].
8. Maguire E, Kelly C, Walsh NM, Gruchy J. White papules on the neck of an older woman: answer. *Am J Dermatopathol.* 2020;42:140-141. [PMID: 31990701].
9. Marconi B, Bobyr I, Campanati A, et al. Pseudoxanthoma elasticum and skin: clinical manifestations, histopathology, pathomechanism, perspectives of treatment. *Intractable Rare Dis Res.* 2015;4:113-22. [PMID: 26361562].
10. Ho D, Jagdeo J. Fractionated carbon dioxide laser treatment of fibroelastolytic papulosis with excellent cosmetic result and resolution of pruritus. *J Drugs Dermatol*. 2015;14:1354-1357. [PMID: 26580887].