

# A Case Report of Pseudoxanthoma Elasticum-like Papillary Dermal Elastolysis

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## Abstract

Pseudoxanthoma elasticum-like papillary dermal elastolysis is a relatively rare disorder within the literature. Increased awareness of this entity will broaden the differential diagnoses among dermatologists encountering such a condition. Pseudoxanthoma elasticum-like papillary dermal elastolysis tends to occur predominantly in elderly females. Clinically, the lesions present as papules and cobblestone plaques on the neck, resembling pseudoxanthoma elasticum. Histologically, there is absence of elastic fibers in the papillary dermis, and no calcification of the remaining fibers. Pseudoxanthoma elasticum-like papillary dermal elastolysis differs from Pseudoxanthoma elasticum by the absence of systemic manifestations, hence patients are reassured and no further workup is warranted. As of now, no effective treatments are available for this entity. Pseudoxanthoma elasticum-like papillary dermal elastolysis should be in the differential diagnosis of a dermatologist when encountering a clinical presentation of Pseudoxanthoma elasticum in an elderly patient localized to the neck, with absence of fragmented and calcified elastic fibers on histologic examination.

## Keywords

Elastic fibers; Cobblestone; Neck

## Introduction

**P**seudoxanthoma elasticum-like papillary dermal elastolysis (PXE-PDE) is a rare disorder of elastic tissue characterized by clinical lesions resembling pseudoxanthoma elasticum (PXE)<sup>[1,2]</sup>. It is an acquired condition, and tends to affect elderly women between the ages of 60 and 80<sup>[2]</sup>. These lesions are small non-follicular papules that tend to coalesce, forming cobblestone-like plaques. Their distribution is symmetrical around the neck and in the area over the clavicle, and to a lesser degree in the axilla and antecubital fossa, and on the abdomen. Unlike PXE,

PXE-PDE has no associated systemic manifestations and no increased mortality<sup>[3]</sup>.

The pathogenesis of PXE-PDE is unknown. Usually, no history of excessive prior sun exposure, urticarial reaction or inflammatory dermatosis is elicited<sup>[4-6]</sup>. Although abnormal elastogenesis has been suggested as being of importance, it has been proposed that this condition represents an intrinsic aging phenomenon<sup>[1]</sup>.

Histologically, there is a complete loss of elastic fibers in the papillary dermis. The remaining fibers are not calcified or fragmented; that is, there are no histopathologic features of PXE. Elastophagocytosis

was present in one case, suggesting that this may be the mechanism for the loss of elastic fibers<sup>[7]</sup>.

### Case History

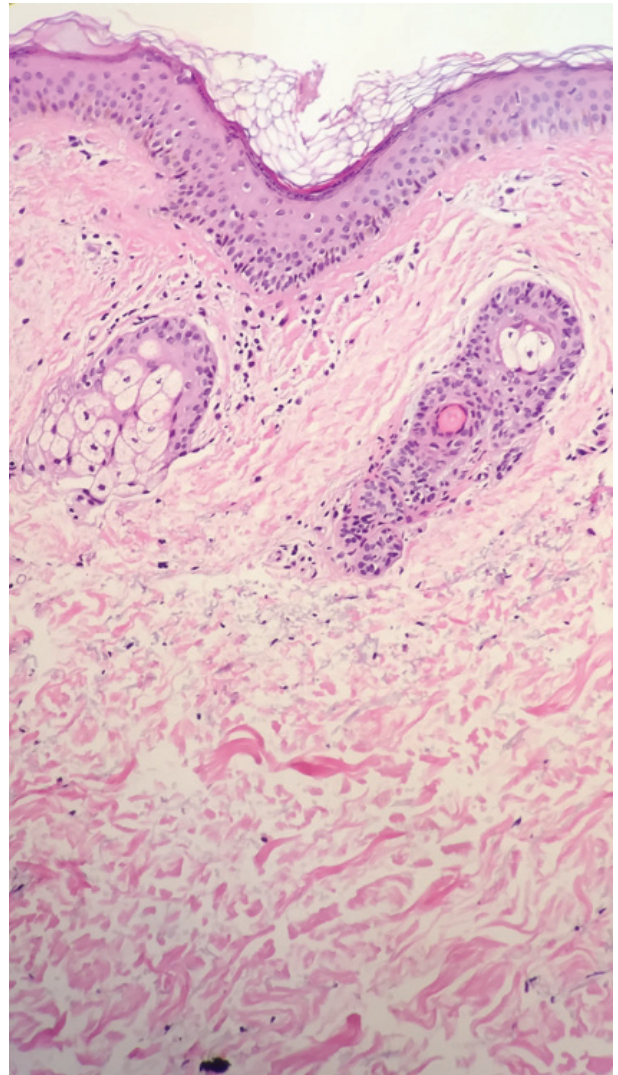
A 71 year-old woman, presented with a 1-2 year history of asymptomatic yellow colored papules coalescing into plaques on both sides of the neck (Fig. 1). No other body sites were involved. There was no family history of any similar condition in other family members. The patient was otherwise healthy, with no other systemic complaints. Given the clinical resemblance to PXE, an ophthalmology consult was performed to rule out the presence of angioid streaks. The eye examination was normal.

### Histological Findings

A skin punch biopsy was performed, and the Hematoxylin and Eosin sections showed a normal appearing epidermis and dermis (Fig. 2). An elastin stain was performed and revealed absence of elastic fibers in the papillary dermis (Fig. 3).



**Figure 1.** Skin with yellow colored papules on the lateral side of the neck.



**Figure 2.** Biopsy specimen of the relatively normal appearing epidermis and dermis (Hematoxylin and Eosin, X20).

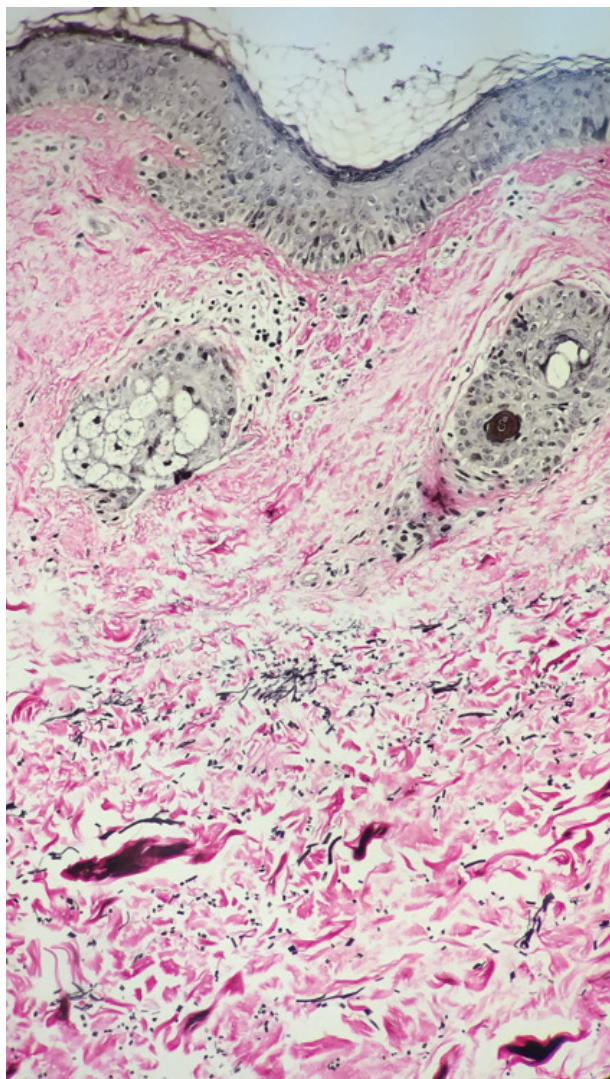
### Follow-up and Management

The patient was seen on follow up, and felt reassured that no systemic manifestations were associated with this disorder. The lesions were stable and asymptomatic. We did not pursue any treatment modalities, as the current literature shows no effective treatment as of now.

### Discussion

When faced with an elderly patient presenting with yellowish colored papules on the neck and other flexural areas, the first clinical impression among most dermatologists is PXE. However, PXE is hereditary, tends to present within the first and second decades of life,





**Figure 3.** An elastin stain showing decreased elastic fibers in the papillary dermis (X20).

and virtually all patients have ophthalmologic findings of Angioid streaks at the time of presentation<sup>[8-12]</sup>. Histologically, there are fragmented and calcified elastic fibers in the mid-dermis<sup>[13]</sup>. Another important differential diagnosis to keep in mind is focal dermal elastosis (FDE). This entity tends to present with the same age, distribution, and clinical presentation of PXE-PDE. However, on histological examination, there is an increase in the amount of elastic fibers in the mid and deep dermis. The elastic fibers are neither fragmented/calcified in the mid-dermis (as in PXE) or absent in the papillary dermis (as in PXE-PDE). This entity has no systemic involvement as well<sup>[14]</sup>.

## Conclusion

Whether PXE-PDE is truly a rare condition, or simply under-reported is yet to be confirmed. That being said, when facing a patient presenting with yellow colored papules on the neck, the following should be considered. If the patient is young, and the skin punch biopsy results show calcified and fragmented elastic fibers in the mid-dermis, a diagnosis of PXE is highly likely and an ophthalmology consultation is warranted. If on the other hand the patient is elderly, then the differential diagnosis includes either FDE or PXE-PDE. If the biopsy results show an increased amount of elastic fibers in the mid and deep dermis, a diagnosis of FDE is favored. However, if elastic fibers were absent in a band-like pattern in the papillary dermis, this would support the diagnosis of PXE-PDE. Patients should be reassured and no further systemic workup is needed.

## Conflict of Interest

The author has no conflict of interest.

## Disclosure

The author did not receive any type of commercial support either in forms of compensation or financial for this study. The author has no financial interest in any of the products or devices, or drugs mentioned in this article.

## Ethical Approval

Ethical approval was obtained with reference number 432-16 from the Unit Biomedical Ethics Research Committee, Faculty of Medicine, King Abdulaziz University.

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## تقرير حالة ورم أصفر كاذب مرن مثل انحلال الجلد النسيجي المرن

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**المستخلص.** ورم أصفر كاذب مرن مثل انحلال الجلد النسيجي المرن هو اضطراب نادر مع عدد قليل من الحالات المبلغ عنها. وزيادة الوعي لهذا الكيان سوف يوسع التشخيص التفريقي لأطباء الأمراض الجلدية عند مواجهة حالة مماثلة لها. ورم أصفر كاذب مرن مثل انحلال الجلد النسيجي المرن يميل إلى أن يحدث في الغالب في النساء المسنات. سريريا، والأفة بشكل حطاطات ولويحات المرصوفة بالحصى على الرقبة، تشبه ورم أصفر كاذب مرن. تشريحيًا، هناك غياب الألياف المرنة في الأدمة الحليمية، وعدم التكلس في الألياف المتبقية. ورم أصفر كاذب مرن مثل انحلال الجلد النسيجي المرن يختلف عن ورم أصفر كاذب مرن بعدم وجود أي ضرر على الأعضاء الداخلية، وبالتالي يتم طمأننة المريض وعدم الخوض في فحوصات أخرى. حالياً، لا توجد أي علاج فعال لهذا المرض. في الختام، يجب أن يكون ورم أصفر كاذب مرن مثل انحلال الجلد النسيجي المرن في التشخيص التفريقي السريري لأطباء الأمراض الجلدية عند مواجهة حالة مماثلة لورم أصفر كاذب مرن في مريض مسن في رقبته، مع عدم وجود الياف مرنة مجزأة ومتكلسة في الفحص النسيجي