

Yellowish-white papules in a cobblestone pattern on the neck

A 65-year-old woman with an unremarkable medical history was referred to our department with a 2-year history of multiple pruritic lesions on the lateral and posterior parts of her neck. She denied any history of preceding trauma or prolonged sun exposure at the affected sites. None of her family members had similar skin lesions. She had no history of renal disease or intake of drugs like D-penicillamine. Physical examination revealed multiple, yellowish-white, 2 mm, non-follicular papules coalescing to form cobblestone-like plaques on her neck [Figure 1]. No other sites were involved. A representative lesion was biopsied and no histological abnormalities were found with routine hematoxylin-eosin staining [Figure 2]. Special staining with acid orcein demonstrated loss of elastic tissue in the papillary dermis and an anomalous distribution and morphology of elastic fibers in the reticular dermis [Figure 3]. Blood tests, cardiovascular and ophthalmological examinations were completely normal.



Figure 1: Multiple yellowish-white non-follicular papules forming cobblestone-like plaques on the neck

WHAT IS YOUR DIAGNOSIS?

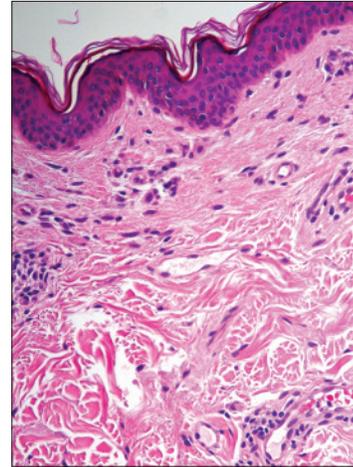


Figure 2: Panoramic view showing apparently normal skin (H and E x20)

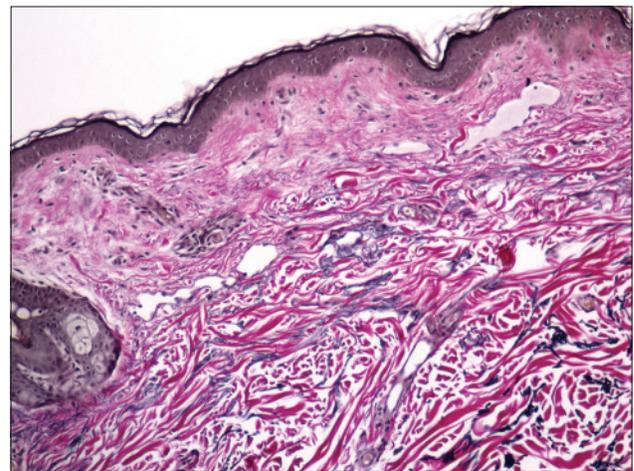


Figure 3: Orcein stain reveals loss of elastic tissue in the papillary dermis with an anomalous distribution and morphology of elastic fibers in the reticular dermis (x400)

This is an open access article distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 3.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as the author is credited and the new creations are licensed under the identical terms.

For reprints contact: reprints@medknow.com

How to cite this article: Conde-Montero E, Chavarría Mur E, Rivera García T, de la Cueva Dobao P. Yellowish-white papules in a cobblestone pattern on the neck. Indian J Dermatol Venereol Leprol 2016;82:464-6.

Received: June, 2015. **Accepted:** December, 2015.

Access this article online	
Quick Response Code:	Website: www.ijdvl.com
	DOI: 10.4103/0378-6323.181504

ANSWER

Pseudoxanthoma elasticum-like papillary dermal elastolysis.

DISCUSSION

Pseudoxanthoma elasticum-like papillary dermal elastolysis was first described in 1992 by Rongioletti and Rebora.^[1] It is a rare, acquired elastic tissue disorder that normally affects adult or elderly women and is characterized by multiple, tiny, flesh-colored or yellowish, non-follicular papules coalescing to form cobblestone-like plaques resembling pseudoxanthoma elasticum. Distribution is usually symmetrical and may involve the posterior and lateral aspects of the neck, axillae, flexor forearms, lower abdomen and inframammary folds. Pruritus maybe an associated symptom.^[2] Unlike pseudoxanthoma elasticum, pseudoxanthoma elasticum-like papillary dermal elastolysis is not associated with any systemic manifestations. Histopathological findings on hematoxylin-eosin staining are non-specific and elastic tissue stains reveal a band-like area of elastolysis along the papillary dermis. Melanophages may be present in the same zone.^[3] Some authors have postulated that the clinical appearance of lesions is due to the herniation of dermis through areas where elastic fibers are reduced or absent whereas others believe that the lesions are secondary to elastotic changes, rather than the elastolytic process.^[4]

The factors that influence the causation of pseudoxanthoma elasticum-like papillary dermal elastolysis remain unclear. Different theories have been proposed such as ultraviolet radiation damage, intrinsic aging, genetic or inheritable factors and defective elastogenesis.^[4]

Differential diagnosis of pseudoxanthoma elasticum-like papillary dermal elastolysis should include other cutaneous disorders of elastic tissue. History of renal disease and D-penicillamine intake has to be specifically ruled out, as this drug interferes with desmosine cross-linking of elastin. The other clinical differential diagnoses would be pseudoxanthoma elasticum, white fibrous papulosis of the neck, late-onset focal dermal elastosis and elastoderma. Moreover, from a histopathologic point

of view, mid-dermal elastolysis and upper dermal elastolysis should also be considered.

Pseudoxanthoma elasticum is an inherited disorder of elastic fibres which appears at a younger age and is characterized by the fragmentation and calcification of elastic fibers with associated systemic symptoms, especially, ocular and cardiovascular manifestations. White fibrous papulosis of the neck presents clinically as white non-confluent papules on the neck and histologically with focal thickening of collagen bundles in the upper dermis. Late-onset focal dermal elastolysis is a rare disorder, clinically similar to pseudoxanthoma elasticum, but characterized by the presence of increased normal-appearing elastic tissue in the mid-dermis and reticular dermis. Elastoderma is an uncommon acquired disorder which presents with increased laxity and loss of recoil of the affected skin. Unlike other elastolytic disorders that produce increased skin laxity, elastoderma is characterized by an accumulation of elastin without calcification. Upper dermal elastolysis is characterized by a papular eruption on the neck, shoulders, upper part of the chest and back with selective loss of elastic tissue in the upper dermis. Mid-dermal elastolysis typically presents with patches and plaques of finely wrinkled skin and focal loss of elastic tissue in the mid-reticular dermis.^[2-4]

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

**Elena Conde-Montero,
Eva Chavarría Mur, Teresa Rivera García¹,
Pablo de la Cueva Dobao**

Departments of Dermatology and ¹Histopathology, Hospital Universitario Infanta Leonor, Madrid, Spain

Address for correspondence: Dr. Elena Conde-Montero,
Department of Dermatology, Hospital Universitario Infanta Leonor,
Madrid, Spain.
E-mail: elenacondemontero@gmail.com

REFERENCES

1. Rongioletti F, Rebora A. Pseudoxanthoma elasticum-like papillary dermal elastolysis. *J Am Acad Dermatol* 1992;26:648-50.
2. Vázquez-Osorio I, Rosón E, Suárez-Peñaranda JM, Vázquez-Veiga H. Pseudoxanthoma elasticum-like papillary

- dermal elastolysis. *Actas Dermosifiliogr* 2015;106:333-6.
3. 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;67:128-35.
 4. Revelles JM, Machan S, Pielasinski U, Camacho D, Vallés L, Santonja C, *et al.* Pseudoxanthoma elasticum-like papillary dermal elastolysis: Immunohistochemical study using elastic fiber cross-reactivity with an antibody against amyloid P component. *Am J Dermatopathol* 2012;34:637-43.