

# ACTAS Dermo-Sifiliográficas

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## CASE FOR DIAGNOSIS

### Non-Follicular Papules with a Cobblestone Appearance

### Pápulas no foliculares en empedrado

#### Patient History

The patient was a woman aged 55 years with a history of intraductal tubular carcinoma of the breast treated with surgery, radiotherapy, and chemotherapy. She consulted for several asymptomatic lesions that had appeared in the axillas and on the neck, upper chest, and arms 2 years earlier and had extended to the wrists and lower part of the abdomen and groin. She reported no history of prolonged exposure to the sun and no family history of similar lesions.

#### Physical Examination

Multiple yellowish non-follicular papules 2 mm to 4 mm in diameter with a cobblestone appearance were present on the neck (Figure 1), axillas, arms, lower abdomen, groin (Figure 2), thighs, and wrists. The patient had no ophthalmic or cardiovascular disorders.

#### Histopathology

Histology revealed the presence of a perivascular lymphohistiocytic inflammatory infiltrate in the dermis,

associated with a loose basophilic material that dissected between the collagen fibers and that was positive on staining with colloidal iron, confirming the presence of acid mucopolysaccharides. Orcein and Verhoeff-Van Giessen stains revealed evidence of a reduction and fragmentation of the elastic fibers in the papillary and mid dermis, with loss of the branching pattern (Figure 3). Von Kossa staining was negative.



Figure 2



Figure 1

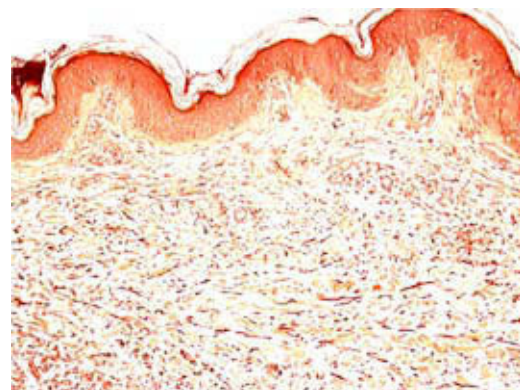


Figure 3 (Orcein stain, original magnification  $\times 10$ ).

**What is Your Diagnosis?**

## Diagnosis

Pseudoxanthoma elasticum (PXE)-like papillary dermal elastolysis.

## Clinical Course and Treatment

The patient was given no treatment due to the benign nature of the condition, the absence of symptoms, and the lack of effective treatments available. The lesions remained stable.

## Comment

PXE is an acquired disorder first described by Rongioletti and Rebora in 1992.<sup>1</sup> It is characterized by multiple papules that mimic the skin lesions of pseudoxanthoma elasticum and by a reduction or absence of elastic fibers in the papillary dermis.<sup>2</sup> It mainly affects women over 60 years.<sup>3,4</sup> The etiology and pathogenesis are unknown but the disorder has been linked to exposure to ultraviolet radiation, endogenous aging, or abnormal elastogenesis (indicated by the finding of immature elastic fibers in the upper reticular dermis). The pathogenesis of the disorder may be related to a defect in elastin and fibrillin 1<sup>3,5</sup> or elastolysis in the middle dermis linked to a deficit of elastin alone.<sup>5</sup> In our patient, we believe the onset of the lesions could be linked to chemotherapy and radiotherapy received 2 years earlier.

Clinical presentation includes the appearance of multiple yellowish non-follicular papules 2 mm to 4 mm in diameter. The lesions are asymptomatic or slightly pruritic with a cobblestone appearance<sup>2,3,5,6</sup> and are caused by herniation of the dermis through areas with altered elastic fibres.<sup>2,3,5</sup> The papules are mainly located in the cervical and supraclavicular regions although they have been known to spread to the axillas, pectoral region, antecubital folds, and lower abdomen.<sup>3,5</sup> Unlike PXE, there are no systemic ophthalmic or cardiovascular manifestations.<sup>2,3,5,6</sup>

Histopathology reveals a partial or total loss of elastic fibers in the papillary dermis, which may be associated with fragmentation and loss of the branching pattern of these fibers. The reticular dermis is normal or may show some reduction of elastic fibers. Von Kossa stain is negative.<sup>2,3,5</sup>

Differential diagnosis principally includes PXE, pseudoxanthoma-like late-onset focal dermal elastosis (increased elastic fibers in the mid and deep reticular

dermis), white fibrous papulosis of the neck (presenting thickening of collagen bundles in the papillary and midreticular dermis, mainly in the cervical region), and upper dermal elastolysis (1 published case associated with elastophagocytosis).

No effective treatment exists and the application of topical retinoids leads to no improvement.<sup>2,3</sup>

In view of their subtle histopathological differences, the similar morphology and location of the lesions, and the higher frequency among elderly individuals, we believe that PXE, white fibrous papulosis of the neck, upper dermal elastolysis, and pseudoxanthoma-like late-onset focal dermal elastolysis could form part of a single clinical spectrum. We therefore conclude that these conditions are different expressions of the skin aging process and propose that they be considered varieties of a single entity under the name of PXE-type lesions of skin aging.

## Conflict of Interest

The authors declare they have no conflict of interest.

## References

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C. Prada-García,\* I. Ruiz-González, S. Delgado-Vicente

*Servicio de Dermatología, Complejo Asistencial de León, León, Spain*

\*Corresponding author.

*E-mail address:* caminoprada@gmail.com

(C. Prada-García).